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A Comparative Study of 5% Topical Lidocaine-Prilocaine Cream (EMLA) and 1% Lidocaine Infiltration of Local Anesthesia in Myringotomy with Ventilation Tube Insertion*

Gabriel O. Te, MD**
Ronaldo A. Reyes, MD**
Jose L. Montilla, III, MD**
Edilberto M. Jose, MD***

The effectiveness of 5% topical lidocaine-prilocaine cream (EMLA) as a local anesthetic agent in myringotomy with ventilation tube insertion was compared with infiltration of 1% lidocaine. Fifteen patients with bilateral chronic serous otitis media were included in the study. The patients' responses to (1) preoperative ear cleaning and antisepsis (2) introduction of the anesthetic agent: 5% topical lidocaine-prilocaine cream (EMLA) in one ear and 1% lidocaine infiltration in the opposite ear, (3) myringotomy (4) ventilation tube insertion and (5) postoperative cleaning were recorded using a visual analogue scale. Statistical analysis using the Wilcoxon signed rank test showed 5% topical lidocaine-prilocaine cream (EMLA) to have distinct advantages over infiltration of 1% lidocaine in myringotomy with ventilation tube insertion: (1) a greater anesthetic effect, (2) avoidance of the painful needle infiltration and (3) its ease of administration.

INTRODUCTION:

Myringotomy with ventilation tube insertion was first employed in the 19th century to relieve serous otitis media. The procedure fell into disfavor due to the high rate of infection and failure. The emergence of antibiotics and newer tube designs have resurrected this procedure. Today, it is one of the most common ear operations performed.

Myringotomy with ventilation tube insertion is the definitive surgical procedure for serous otitis media. This condition is characterized by the development of a non- purulent effusion in the middle ear which may present acutely or develop into an insiduous chronic course.

The underlying cause of serous otitis media is a eustachian tube dysfunction which may be due to varied causes ranging from an upper respiratory infection, allergy, enlarged tonsils and adenoids to tumors of the nasopharynx.

The normal middle ear mucosa continuously absorbs air. With obstruction of the eustachian tube, there is a failure of normal ventilation resulting in a vacuum and negative pressure inside the middle ear cavity. The negative pressure causes a transudate from blood vessels resulting in accumulation of fluid inside the middle ear cavity.

Clinical features include deafness, ear fullness, occasional pain, demonstration of middle ear effusion as evidenced by an air-fluid level visible through a bulging or retracted tympanic membrane. Occasionally, the only otoscopic picture is that of a yellowish tinge to the affected membrane as compared to the normal side. Tuning fork tests and pure tone audiograms generally demonstrate a conductive deafness. Impedance audiometry will invariably show a flattened curve with a shift to the negative side or a totally flat tracing.

Medical treatment consists of inflation of the middle ear by Valsalva's maneuver, politzerization or catheterization of the eustachian tube and the administration of systemic or topical decongestants directed towards the eustachian tube orifice. Antihistamines can be added if an allergic etiology is strongly considered. Surgical treatment is directed towards evacuation of the effusion. The surgical procedure of choice is a myringotomy. For acute conditions, the above procedure is adequate. For chronic diseases, a myringotomy is accompanied by insertion of a tube transtympanically into the middle ear cavity to ventilate the middle ear thereby restoring normal middle ear pressure and allowing mucosal edema and consequent effusion to subside.

In a myringotomy, a small incision is made through the tympanic membrane, either anteroinferiorly or posteroinferiorly, to evacuate the middle ear effusion. Of the two, an anteroinferior quadrant incision is preferred because of two reasons, namely: spontaneous extrusion of a ventilation tube is delayed and the avoidance of residual hearing

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** Residents, Dept. of ENT, UP-PGH.
*** Consultants, Dept. of ENT, UP-PGH

impairment that may occur if a tube is placed near the round window as in a posteroinferior quadrant myringotomy. The superior quadrants of the tympanic membrane are avoided since the ossicles may be injured. A ventilation tube may be inserted through the incision and left in place to restore normal ventilation and pressure. The tubes are allowed to extrude spontaneously.

Due to the confined operating space, i.e. the dimensions of the external auditory canal, myringotomy with ventilation tube insertion requires a quiet and cooperative patient. To ensure this, anesthesia is needed.

The choice of anesthesia depends on the surgeon and the patient. For pediatric patients, general anesthesia is invariably required. For adults, a local anesthetic agent is adequate for the procedure. The standard anesthetic technique used in our institution consists of infiltration of 1% to 2% lidocaine hydrochloride with a small gauge needle at the junction between the cartilaginous and bony ear canal at several points along its circumference. This technique provides adequate analgesia albeit with several drawbacks. Infiltration of lidocaine along the circumference of the external auditory canal elicits considerable pain. In addition, lidocaine hydrochloride is inherently irritating to the skin and subcutaneous tissues. This initial pain of infiltration causes varying degrees of discomfort and agitation. Improper infiltration may result not only in anesthetic failure but also cause unwanted swelling of the canal skin thereby occluding the tympanic membrane and limiting the operative field. To remove this distracting initial pain, other authors have advocated the use of iontophoretic application of lidocaine. Iontophoresis is a procedure in which charged molecules or ions are induced to migrate through tissues under the influence of a direct electrical current. With this technique, a small quantity of lidocaine solution is introduced over the tympanic membrane and direct electrical current is applied to the solution. This technique has produced conflicting results. Still other investigators have used phenol applied to the myringotomy site. Nevertheless, with either procedure, tympanic membrane pain thresholds are unpredictable and myringotomy and ventilation tube insertion may still cause discomfort and pain.

The availability of a lidocaine-prilocaine cream (EMLA) for topical analgesia offers great promise in its possible use in minor surgery of the tympanic membrane. EMLA 5% cream consists of a mixture of 25 mg/ml of lidocaine base and 25 mg/ml of prilocaine base together with an emulsifier, a viscosity-increasing agent and water.

This topical anesthetic has been widely used in minor dermatologic surgery and to relieve the pain of venipuncture. In a study by **Smith et al** (1990) comparing EMLA with lidocaine infiltration to relieve venipuncture pain, EMLA was found to be more effective than lidocaine infiltration.

Previous studies have shown that the minimal application time for effective anesthesia is 45 minutes although 60 minutes is necessary in children (**Hallen et al**, 1984). Anesthesia was effective for up to five hours after application (**Hopkins et al**, 1988). EMLA has also been used to relieve the pain of otitis externa and to allow pain-free ear cleaning in these patients (**Premachandra**, 1990).

Objective:

To compare the effectiveness of 5% topical lidocaine-prilocaine cream (EMLA) versus 1% lidocaine infiltration for anesthesia of the tympanic membrane and external auditory canal in myringotomy with ventilation tube insertion.

METHODOLOGY:

A. Participants.

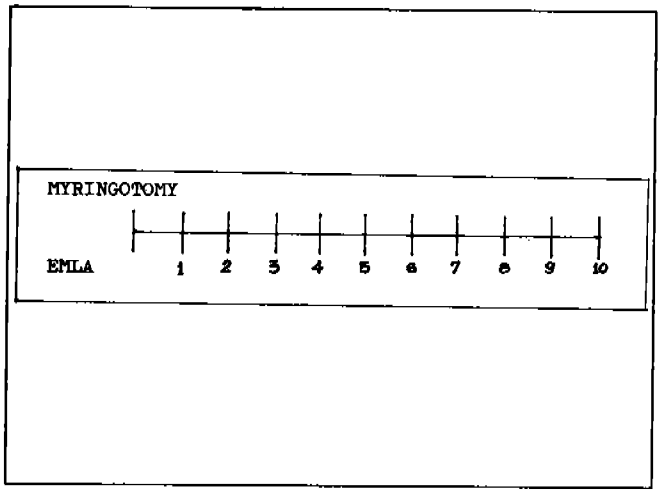
The study involved 15 cooperative adult patients with an age range of 21 to 50 years diagnosed to have bilateral chronic serous otitis media through a detailed history and thorough ENT examination. Patients with concommitant otitis externa and/or an acute suppurative otitis media and those with medical contraindications to surgery and anesthesia were not included in the study. A properly informed consent was obtained from all the study participants.

B. Procedure.

All the study participants were randomly assigned numbers from 1 to 15. Odd-numbered patients were anesthesized with 5% topical EMLA cream in the left ear and infiltration of 1% lidocaine hydrochloride in the right ear. Even-numbered patients, on the other hand, were anesthesized with infiltration of 1% lidocaine hydrochloride in the left ear and 5% topical EMLA cream in the right ear. The topical EMLA cream and 1% lidocaine solution are manufactured by ASTRA Pharmaceuticals (Phils.), Inc.

A simple coin toss decided which ear would receive the intended treatment first with "heads" for the left ear and "tails" for the right ear. The same coin (50c) was used throughout the study.

A rest period of one hour was given between treatment procedures to allow for dissipation of any residual pain experienced during the first treatment procedure.



(FIGURE 1) VISUAL ANALOGUE SCALE

The patients were comfortably placed in a reclining position and properly draped prior to any aural manipulation.

C. Pain Recording.

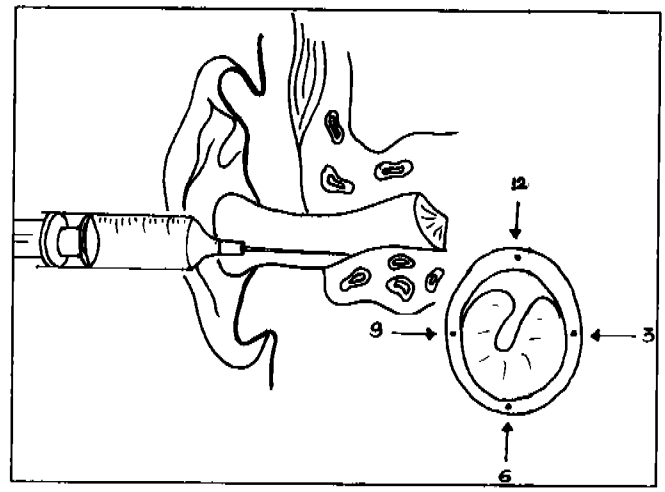
The patients' reactions to discomfort and pain were recorded using a visual analogue scale during each of the following stages in the procedure: (1) cleaning and antiseptic preparation of the ear canal and tympanic membrane with Povidone-iodine antiseptic (2) introduction of the intended anesthetic agent (3) myringotomy (4) ventilation tube insertion and (5) postoperative cleaning.

Through each of the following steps, a visual analogue scale was shown to the patients to record the level of pain and discomfort experienced from a scale of 0 (no pain) to 10 (extreme pain comparable to the most physically painful experience ever felt by the patient in his life), (Fig. 1). A new visual analogue scale was shown in each step of the procedure to avoid patient bias. Moreover, only one investigator performed all the procedures in the entire study.

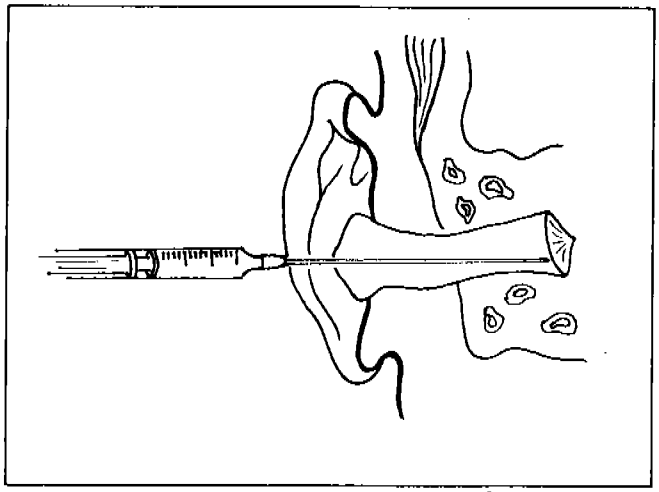
D. Introduction of anesthetic agent.

Application of 5% Topical lidocaine-prilocaine cream (EMLA).

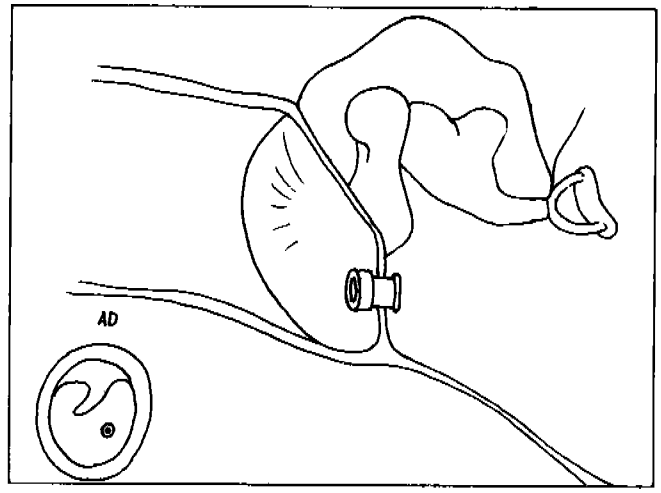
After proper cleaning and antiseptic preparation of the ear canal and tympanic membrane, 0.5 cc of EMLA cream was applied on the tympanic membrane and then along the inner two-thirds of the external auditory canal using a gauge 16 intravenous catheter attached to a tuberculin syringe, (Fig. 2). The external auditory meatus was, then, lightly occluded with a piece of sterile cotton for one hour to await adequate anesthesia.



(FIGURE 3) INJECTION OF 1% LIDOCAINE AT FOUR QUADRANTS 1, 2, 3, 6, & 9 O'CLOCK AT JUNCTION OF CARTILAGINOUS AND BONY EAR CANAL



(FIGURE 2) APPLICATION OF EMLA 5% ALONG TYMPANIC MEBRANE AND BONY PORTION OF EAR CANAL



(FIGURE 4) VENTILATION TUBE AT THE ANTEROINFERIOR QUADRANT

Needle Infiltration of 1% lidocaine solution

After adequate cleaning and antisepsis, 1.0 cc of 1% lidocaine hydrochloride was infiltrated with a gauge 26 needle attached to a tuberculin syringe at the junction of the outer cartilaginous third and the inner bony two-thirds of the external auditory canal at the 12, 3, 6, and 9 o' clock positions, (Fig. 3). The external auditory meatus was then lightly occluded with sterile cotton for 10 minutes for adequate anesthesia.

E. Myringotomy and Ventilation Tube Insertion

With the aid of an operating microscope, the incision was placed in the anteroinferior quadrant of the tympanic membrane, (Fig. 4). An electric sucker was used to evacuate fluid inside the middle ear cavity.

A polyethylene tube shaped like a Grommet ventilation tube was then inserted into the myringotomy incision.

F. Postoperative Care

Postoperatively, the ear canal and tympanic membrane was suctioned clean of blood and fluid and in the EMLA group, of remnants of EMLA cream.

The patients were advised to avoid aural manipulation and to keep ears dry. Decongestants and prophylactic oral antibiotics were given.

G. Statistical Analysis

The degree of discomfort and pain experienced by the patients with the use of lidocaine infiltration and application of EMLA in opposing ears as measured by the visual analogue scale was subjected to statistical analysis using the Wilcoxon signed ranks test through each of the five steps in our procedure using critical values for a two- tailed distribution in step 1 and one-tailed distribution from step 2 to step 5.

RESULTS:

Step 1 :Cleaning and antiseptic preparation of the ear.

The patients' reactions to ear cleaning and antisepsis as reflected by their visual analogue scores (VAS) were not significantly different between the two groups. ($a = .01$; $p = 0.1250$). No anesthetic agent had been introduced at this point. From this lack of significant difference, pain thresholds are presumed equal in opposing ears and validates the use of the patient as his own control.

Step 2 :Introduction of anesthesia - 1% lidocaine needle infiltration vs. topical application of 5% lidocaine-prilocaine (EMLA) cream.

Lidocaine infiltration into the ear canal resulted in markedly high visual analogue scores ($X = 6.00$) which were significantly different from the lower VAS ($X = 1.83$) recorded from EMLA-treated ears ($a = .01$; $p = 0.0000$). The greatest significant difference in VAS between the opposing treatment groups was seen in this step of the procedure.

Step 3: Myringotomy

Myringotomy on ears anesthesized with infiltration of 1% lidocaine elicited higher visual analogue scores ($X = 4.33$) than on ears anesthesized with 5% EMLA cream ($X = 1.8$). The difference between the VAS of the opposing groups was significant ($a = .01$; $p = 0.0001$).

Step 4: Ventilation tube insertion

Ventilation tube insertion in ears anesthesized with 5% EMLA cream had lower visual analogue scores ($X = 2.33$) than ventilation tube insertion in ears infiltrated with 1% lidocaine ($X = 4.40$). Again, the difference between the two groups was significant ($a = .01$; $p = 0.0067$).

Step 5: Postoperative cleaning

No significant difference was noted between the VAS of the lidocaine infiltration group ($X = 1.93$) and the EMLA group ($X = 1.47$) at an $a = .01$ and $p = 0.0313$.

Among the 30 ears operated on in our sample of 15 patients, only one ear had a complication. This ear was part of the EMLA group and developed a suppurative otitis media three days after the operation causing extrusion of the ventilation tube. VI.

DISCUSSION:

The ideal anesthetic technique for minor surgery of the tympanic membrane and ear canal is one which would have the following features: (1)ability to provide adequate anesthesia of the ear canal and the tympanic membrane without needle infiltration of the anesthetic agent, (2) a topically applied anesthetic agent capable of penetrating the barrier of the tympanic membrane epithelium and anesthesizing the nerve endings in the lamina propia without causing damage to the external auditory canal, tympanic membrane, middle and inner ear structures and (3) ease of administration.

Topical 5% lidocaine-prilocaine cream (EMLA) seems to fulfill all of the above criteria.

The first criterion cited is clearly demonstrated in

APPENDIX

Wilcoxon signed rank test (two-tailed distribution)

Table 1. Visual analogue scores recorded in Step 1 (Ear Cleaning and Antisepsis) in ears infiltrated with 1% lidocaine (A) and in ears applied with 5% EMLA cream (B)

Sample #	Anesthetic agent A	B	Difference (D)	Rank of /D/	Signed +	Rank -
1	0	0	0	dropped	-	-
2	1	1	0	dropped	-	-
3	3	3	0	dropped	-	-
4	1	1	0	dropped	-	-
5	1	1	0	dropped	-	-
6	1	1	0	dropped	-	-
7	0	0	0	dropped	-	-
8	2	1	1	2.5	2.5	-
9	2	1	1	2.5	2.5	-
10	1	0	0	dropped	-	-
11	0	0	0	dropped	-	-
12	1	1	1	2.5	2.5	-
13	1	0	0	dropped	-	-
14	1	1	1	2.5	2.5	-
15	1	0	0	dropped	-	-
TOTAL	16	12			10	-
X	1.06	0.8				

H_O : There is no significant difference in pain threshold of the opposing ears.
H_A : There is a significant difference in pain threshold of the opposing ears.

If $a = 0.01$; $p = 0.125 > 0.01$

Decision : Cannot reject H_O
Conclusion: There is not enough evidence to say that there is a significant difference in pain threshold of the opposing ears.

Wilcoxon signed rank test (one-tailed distribution)

Table 2. Visual analogue scores recorded in Step 2 (application of anesthesia: 1% lidocaine infiltration (A) and 5% EMLA application (B)

Sample #	Anesthetic agent A	Anesthetic agent B	Difference (D)	Rank of /D/	Signed +	Rank -
1	8	0	8	15	15	-
2	7	1	6	13.5	13.5	-
3	10	5	5	11	11	-
4	7.7	4.5	3	5	5	-
5	6	3	3	5	5	-
6	5	2	3	5	5	-
7	3	1	2	1.5	1.5	-
8	6	1	5	11	11	-
9	7	1	6	13.5	13.5	-
10	5	1	4	8.5	8.5	-
11	3	0	3	5	5	-
12	5	2	3	5	5	-
13	5	3	2	1.5	1.5	-
14	6	1	5	11	11	-
15	6	2	4	8.5	8.5	-
TOTAL	89.5	27.5			120	-
X	6.00	1.83				

H_O : There is no significant difference in pain and discomfort experienced between infiltration of 1% lidocaine and topical application of 5% EMLA.

H_A : There is a significant difference in pain and discomfort experienced between infiltration of 1% lidocaine and topical application of 5% EMLA in that 5% EMLA results in a significantly lesser degree of pain and discomfort than infiltration of 1% lidocaine.

If $a = 0.01$; $p = 0.0000 < 0.01$

Decision : Reject H_O

Conclusion: There is a significant difference in pain and discomfort experienced between infiltration of 1% lidocaine and topical 5% EMLA in that topical application of 5% EMLA results in a significantly lesser degree of pain and discomfort than infiltration of 1% lidocaine.

Wilcoxon signed rank test (one-tailed distribution)

Table 3. Visual analogue scores recorded in step 3 (myringotomy) in ears infiltrated with 1% lidocaine (A) and in ears applied with 5% EMLA cream (B).

Sample #	Anesthetic agent A	B	Difference (D)	Rank of /D/	Signed +	Rank -
1	0	0	0	dropped		-
2	4	2	2	4.5	4.5	-
3	8	3	5	14	14	-
4	8	5	3	10	10	-
5	2	0	2	4.5	4.5	-
6	3	1	2	4.5	4.5	-
7	2	0	2	4.5	4.5	-
8	6	2	4	12.5	12.5	-
9	5	1	4	12.5	12.5	-
10	5	3	2	4.5	4.5	-
11	3	1	2	4.5	4.5	-
12	4	2	2	4.5	4.5	-
13	5	3	2	4.5	4.5	-
14	6	3	3	10	10	-
15	4	1	3	10	10	-
TOTAL	65	27			105	-
X	4.3	1.80				

H_o : There is no significant difference in effectiveness of 1% lidocaine infiltration and topical application of 5% EMLA as a local anesthetic in myringotomy.

H_A : There is a significant difference in effectiveness of 1% lidocaine infiltration and topical application of 5% EMLA in that 5% EMLA is more effective as a local anesthetic agent than 1% lidocaine infiltration in myringotomy.

If $a = 0.01$; $p = 0.0001 < 0.01$

Decision : Reject H_o

Conclusion: There is a significant difference in effectiveness of 1% lidocaine infiltration and topical application of 5% EMLA in that 5% EMLA is more effective as a local anesthetic agent than 1% lidocaine infiltration in myringotomy

Wilcoxon signed rank test (one-tailed distribution)

Table 4. Visual analogue scores recorded in step 4 (ventilation tube insertion) in ears infiltrated with 1% lidocaine (A) and in ears applied with 5% EMLA cream (B).

Sample #	Anesthetic agent A	B	Difference (D)	Rank of /D/	Signed + | - |	Rank
1	0	0	0	dropped		-
2	4	2	2	4.5	4.5	-
3	8	3	5	13	13	-
4	9	6	3	10	10	-
5	2	4	-2	-	-	-
6	4	2	2	4.5	4.5	-
7	2	0	2	4.5	4.5	-
8	4	1	3	10	10	-
9	6	3	3	10	10	-
10	5	3	2	4.5	4.5	-
11	3	2	1	1	1	-
12	4	2	2	4.5	4.5	-
13	4	2	2	4.5	4.5	-
14	6	3	3	10	10	-
15	5	2	3	10	10	-
TOTAL	66	35			91	-
X	4.40	2.33				

H_o : There is no significant difference in effectiveness of 1% lidocaine infiltration and topical application of 5% EMLA as a local anesthetic in ventilation tube insertion.

H_A : There is a significant difference in effectiveness of 1% lidocaine infiltration and topical application of 5% EMLA in that topical application of 5% EMLA is more effective as a local anesthetic agent than 1% lidocaine infiltration in ventilation tube insertion.

If $a = 0.01$; $p = 0.0067 < 0.01$

Decision : Reject H_o

Conclusion: There is a significant difference in effectiveness of 1% lidocaine infiltration and topical application of 5% EMLA in that topical application of 5% EMLA is more effective as a local anesthetic than 1% lidocaine infiltration in ventilation tube insertion.

Wilcoxon signed rank test (one-tailed distribution)

Table 5. Visual analogue scores recorded in step 5 (postoperative cleaning) in ears infiltrated with 1% lidocaine (A) and in ears applied with 5% EMLA cream (B)

Sample #	Anesthetic agent A	B	Difference (D)	Rank of /D/	Signed +	Rank -
1	0	0	0	dropped	-	-
2	1	1	0	dropped	-	-
3	3	3	0	dropped	-	-
4	4	4	0	dropped	-	-
5	0	0	0	dropped	-	-
6	1	1	0	dropped	-	-
7	1	1	0	dropped	-	-
8	3	1	2	4.5	4.5	-
9	4	2	2	4.5	4.5	-
10	2	2	0	dropped	-	-
11	2	1	1	2	2	-
12	1	1	0	dropped	-	-
13	1	1	0	dropped	-	-
14	3	2	1	2	2	-
15	3	2	1	2	2	-
TOTAL	29	22			15	-
X	1.93	1.47				

H_o : There is no significant difference in effectiveness of 1% lidocaine infiltration and topical application of 5% EMLA as a local anesthetic in ear cleaning.

H_A : There is a significant difference in effectiveness of 1% lidocaine infiltration and topical application of 5% EMLA in that topical application of 5% EMLA is more effective as a local anesthetic agent than 1% lidocaine infiltration in ear cleaning.

If $a = 0.01$; $p = 0.0313 > 0.01$

Decision: Cannot reject H_o

Conclusion: There is not enough evidence to say that there is a significant difference in effectiveness of 1% lidocaine and topical application of 5% EMLA in that topical application of 5% EMLA is more effective as a local anesthetic agent than 1% lidocaine infiltration in ear cleaning.

step 2 of the procedure (introduction of the anesthetic agent: 1% lidocaine infiltration vs. topical 5% lidocaine-prilocaine cream). The highest visual analogue scores, at any point of the study, were obtained when the patients' ears were infiltrated with 1% lidocaine as compared to the low VAS recorded when 5% EMLA was applied into the ear canal. The greatest significant statistical difference between the two treatment groups was also evident in this step. Apparently, the most painful part of the surgical procedure arises from the initial prick of the needle and the secondary irritation from the subsequent infiltration of lidocaine. With the use of EMLA, this most painful step is eliminated.

Topical 5% lidocaine-prilocaine cream (EMLA) has also demonstrated anesthesia superior to that of lidocaine infiltration as reflected by the statistically significant differences between the VAS of the two groups in steps 3 and 4 (myringotomy and ventilation tube insertion).

It is the authors' opinion that a large part of the success of 5% EMLA in inducing superior anesthesia can be attributed to its ease of application in contrast to lidocaine infiltration. Lidocaine infiltration along the prescribed areas of the external auditory canal needs a certain degree of technical expertise which may result in less than optimal anesthesia or even anesthetic failure in some instances. Topical lidocaine-prilocaine cream (EMLA), on the other hand, can be applied easily with the minimal amount of manipulation and obtain adequate anesthesia.

It is interesting to note that in one of our patients, insertion of a ventilation tube in the ear anesthesized with EMLA resulted in greater pain (VAS of 4) than in the ear anesthesized with infiltration of 1% lidocaine with a VAS OF 2. (Appendix B, Table 4, Sample #5).

This isolated recording could be due to subjectivity on the patient's part however, noting her consistent responses in the other steps of the procedure, the possibility of anesthetic failure with EMLA is a strong consideration. The insertion of a ventilation tube entails considerable manipulation on the tympanic membrane as well as along the confines of the narrow ear canal. If EMLA has not been applied adequately to all prescribed areas, then pain and discomfort will surely be greatest in this step of the procedure. It is our strong recommendation that EMLA be applied evenly to all areas in question and that the patient tilt his head with the involved ear up to allow gravity to help distribute the cream along the membrane and canal as well as to maintain adequate contact.

It also appears that there is no significant difference in recorded VAS between treatment groups in step 5 (postoperative cleaning). There is minimal manipulation at this stage of the procedure and this may be the reason behind the lack of significant difference. While the advantages of EMLA as a local anesthetic for minor ear surgery are attractive, there are several disadvantages to be considered.

One obvious drawback to its use is the prolonged waiting time for optimal anesthesia. Forty-five minutes to an hour is recommended whereas only five to ten minutes is needed for lidocaine infiltration. A second disadvantage is its cost. A 5 g tube of EMLA cost approximately P225 as compared to a carpule of 1% lidocaine costing approximately P10 only. If possible, a 5 g tube of EMLA can be shared among several patients to minimize the cost.

Lastly, there are complications in the use of topical lidocaine-prilocaine cream which include skin blanching which invariably disappears 30 minutes after application. There is also the possibility of the cream causing contamination of other structures by virtue of it being applied throughout the tympanic membrane and the ear canal if an aseptic technique is not followed.

Among a total of 30 ears operated on in our sample of 15 patients, one ear in the EMLA group developed an infection three days after operation. The possibility of contamination during operation has been strongly considered as well as a secondary infection either from aural manipulation or from a rhinogenic focus.

In summary, the authors have shown that 5% topical lidocaine-prilocaine cream (EMLA) is a viable local anesthetic agent for myringotomy with ventilation tube insertion. It is easy to apply, offers superior anesthesia compared to lidocaine infiltration and eliminates the need for painful needle infiltration.

It is the recommendation of the authors that further investigation be done to determine if the anesthetic effect of EMLA is still adequate for a myringotomy with ventilation tube insertion even with drastically reduced waiting periods.

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TUBERCULOUS OTITIS MEDIA: IS IT AROUND OR NOWHERE TO BE FOUND?*

Ma. Cristina S. Lopez, MD**
Marilyn Ong-Mateo, MD**
Norberto V. Martinez, MD***

ABSTRACT

Tuberculous Otitis Media is today an uncommon disease, which, if left untreated, causes great damage to the middle ear and its surrounding structures. This paper has proven that it is definitely not easy to diagnose and thus, the otolaryngologist shquld be made aware of its features and maintain a high index of suspicion. Twenty-five patients, 14 males and eleven females with aural discharge were included in this study. Presenting signs and symptoms were carefully noted. AFB staining and Bactec culture were done. In six patients, Mycobacterium Tuberculosis was isolated. Chemotherapy has proven satisfactory results. A mere suspicion in the clinician's mind should be sufficient enough to reach a diagnosis of tuberculous otitis media.

INTRODUCTION

Despite advances in pharmacology, tuberculosis still remains as one of the most common lethal infectious disease especially in underdeveloped countries. Statistics ranks it as the fifth leading cause of morbidity and fourth as a cause of mortality. Because of the adverse effects of tuberculosis on the individual, the family, as well as the community, it has been considered a grave problem to health officials, to practicioners and to the general public.

The incidence of tuberculous ear disease is quite low particularly in advanced and industrialized countries. In Great Britain, only eleven out of 23,000 cases of suppurative otitis media were reported for the years 1950-1959. In the United States, only 22 cases of chronic otitis media caused by Mycobacterium tuberculosis during a 30 - year period were available. In the British Isles, there is a 0.5% incidence of tuberculous otitis media reported. Diagnosis can be quite difficult because the signs and symptoms may mimic other ear infections. A careful history of the onset and the presentation of the findings may sometimes disclose that discharging ears which have failed to respond to the usual management are considered tuberculous in nature.

Infection of the middle ear may occur as follows: a) Primary - in which there is no other focus of tuberculosis in the patient. In these cases, the infection is usually due to the bovine strain of the tubercle bacillus contracted by drinking infected cow's milk. The anatomical differences between the adult and infant eustachian tubes have been implicated in the higher incidence of involvement in the younger age group. The infant's tube is shorter, lumen is greater and nearly more horizontal. Reflux of milk has been suggested as a route of contamination. b) secondary - occurring in adults who are suffering from active pulmonary tuberculosis with human type bacilli in the sputum. The bacillli may each the middle ear either by hematogenous spread in acute miliary tuberculosis, or via the eustachian tube from the nasopharynx during a cough or a sneeze, or through regurgitation. The mucous membranes of the eustachian tube may also become infected from tuberculous adenoids, and infection may spread to the middle ear. The initial lesion, the Ghon tubercle, involves lymphatics with a secondary lymphadenitis. If the infection continues, there is seeding of the bloodstream.[16] In a survey conducted among 32 Otolaryngologists in Metro Manila, only six (18.7%) consider tuberculous otitis media as a differential diagnosis whenever they encounter patients with otorrhea. Tuberculous otitis media is still considered an under diagnosed disease throughout the world. It is a variable, erratic, and puzzling infectious disease which may remain undiagnosed or confused with other acute or chronic ear conditions if appropriate diagnostic studies are not performed. The index of suspicion worldwide is quite low even in the Philippines where tuberculosis is endemic. This attitude of indifference manifests well in the local literature where studies on tuberculous otitis media are lacking. It is primarily for this reason why this study was initiated. It is believed that no other country has enough cases of

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** Residents, Dept. of ENT, UST
*** Consultants, Dept. of ENT, UST

tuberculosis to deal with and this research will probably uncover another salient aspect of tuberculosis as a multisysyem disease.

HISTORICAL REVIEW

Tuberculous involvement of the temporal bone was first described by **Jean Louis Petit** in the 18th century. In 1835, **Romberg** and **Geissler** associated tuberculous mastoiditis with pulmonary tuberculosis. The clinical signs were first outlined by **Wilde**, who in 1853 described the painless otorrhea and the appearance of the tympanic membrane. **Schwartze** reported the cheesy infiltration and the tubercles of mucosa. **Politzer** and **Meniere** pointed out the destructive nature of this disease. In 1882, **Koch** demonstrated the tubercle bacillus, and in 1883 **Esche** isolated the bacillus in the aural discharge. **Habermann** in 1885 was able to show giant cells and bacilli in the mucosa of the tympanic cavity, and ossicular destruction and extrusion were reported by **Oppenheimer** in 1900.[3]

Earlier reports in the literature divide tuberculous otitis media into primary and secondary types depending upon whether the disease was found elsewhere. **Milligan** in 1903 stated that primary infection was not so uncommon as has been supposed, in that it could occur in cancellous spaces of the temporal bone. In 1911, he cited numerous reports to support his claim.[3]

Proctor and **Lindsay** in 1942 stated that most of the reported cases of primary tuberculosis of the temporal bone had not been studied completely to exclude tuberculous foci elsewhere. They emphasized that the majority of these cases in the temporal bone were primary only in its clinical sense.

Several reports from foreign literature were made regarding tuberculous otitis media. Clinical records at the Department of Otolaryngology, University of Tubingen in West Germany reviewed 4,000 cases from the years 1967-1979, 14 of which were proven to be cases of middle ear tuberculosis through biopsy specimens.[9]

Hospital records at the Massachusetts Eye and Ear Infirmary were likewise reviewed for the years 1962-1984 for cases of tuberculosis of any site. Relevant clinical and laboratory data were extracted from these records. Mycobacterium tuberculosis using the Lowenstein-Jensen media based on biochemical parameters were isolated.[9]

OBJECTIVES

GENERAL: To describe the clinical picture of tuberculous otitis media among sample Filipinos.
SPECIFIC: To identify the signs and symptoms of tuberculous otitis media as proven clinically and/or through laboratory results. To describe the otoscopic findings of tuberculous otitis media as proven clinically and/or through laboratory findings. To identify what laboratory examinations are needed in coming up with a diagnosis of tuberculous otitis media.
To determine the efficacy of management of cases of tuberculous otitis media.

MATERIALS AND METHODS

An initial study of 39 patients chosen from ENT Out-patient Department of the Sto. Tomas University Hospital was made however, fourteen were lost to follow-up, thus twenty-five patients, 14 males and 11 females, ages one year to 63 years old with aural discharge were included in this study.

The criteria for inclusion were the following: otorrhea of three months or longer but were never treated; or those treated with antibiotics but did not respond; otorrhea of less than three weeks but developed complications; those with positive exposure and/or positive family history; and those presenting with the constitutional symptoms of tuberculosis.

Excluded from this study were patients undergoing anti-koch's therapy for the past three months. Patients currently on antibiotics were asked to stop for a week to make the aural discharge free from any antibiotic reaction.

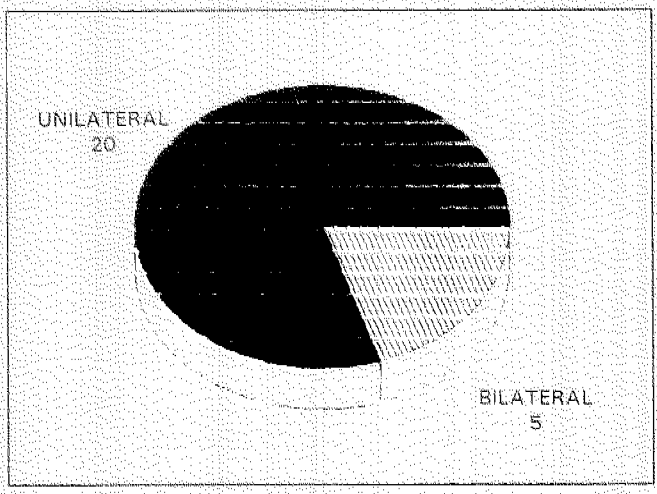
Subjects suspected of having a tuberculous disease or those with suspicious otoscopic findings were selected from the ENT Out-Patient Department of the Santo Tomas University Hospital. They were reviewed from the months Jun 1991 to February 1992. Presenting signs and symptoms, such as otorrhea, otalgia, decreased hearing, as well as the presence of perforation and granulation tissues were carefully noted. A careful history of the onset as well as the presenting symptoms were established. The ear canal was initially cleansed with a sterile cotton pledget soaked in 70% alcohol solution. With a sterile collecting bottle attached to a suction, discharge was collected. Specimen was wiped on glass slides for AFB staining while the remaining specimen were sent for culture where it was incubated at 37 degrees centigrade for 24 hours after which they were read. Chest x-ray, mastoid x-ray, and pure-tone audiometry were done. In those who underwent surgery, intraoperative findings were noted and granulation tissues were sent for histopath. All subjects were started on anti-koch's therapy. (Rifampicin, Isoniazid, Pyrazinamide). Dosages were computed based on body weight and were prescribed for six months. The degree of improvement was noted by the use of a score index. (1=dry, 2=less discharge, 3=no change, 4=more

discharge). No attempts whatsoever were made to differentiate the primary from the secondary form of tuberculosis.

RESULTS:

I. CLINICAL SYMPTOMATOLOGY

Twenty five (25) patients, 14 (56.0%) males and 11 (44%) females were included in this study. The peak age incidence was in the second decade of life seen in Table IA. With regards the side of involvement, 20 (80%) were unilateral and 5 (20%) were bilateral, (Fig. 1). All patients had positive exposure to active pulmonary tuberculosis, of which, 9 (36%) had past history of

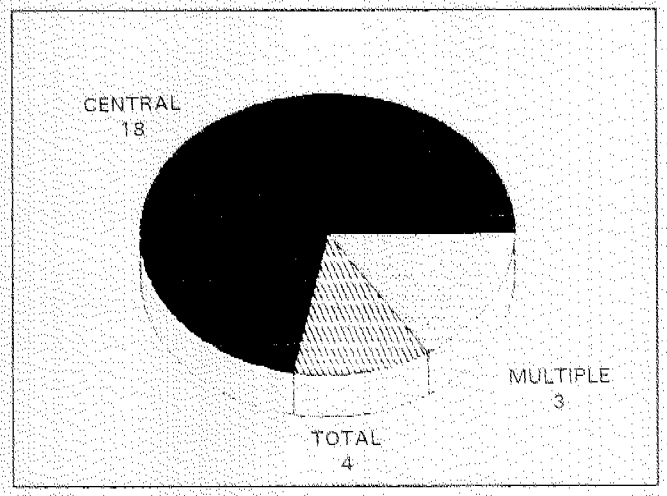


(FIGURE 1) SITE OF INVOLVEMENT

pulmonary tuberculosis.

The duration of symptoms prior to presentation is shown in Table IIA. Otorrhea was present in all cases with the discharge varying from scanty to profuse (Table IIB). There were 17 (68%) patients who had painless otorrhea often of long duration, however 2 (28%) of the eight cases who presented with otalgia were acute cases. This shows an 83.33% sensitivity and a 36.80% specificity, with a false positive of 63.75% and false negative result of 16.66% (Table IIIB).

Likewise, otoscopic findings were noted such as the type of perforation and the presence of granulation tissues. Of the 25 ears affected, only three presented with multiple perforations and yet, these were the cases which did not turn out to be positive in the culture (Figure 2). A sensitivity of 0.00%, specificity of 84.20%, false positive of 15.78%, false negative of 100% were noted with regards the type of perforation. While a sensitivity of 66.66%, a specificity of 21.50%, false positive of 78.94%, and a false negative of 33.33% was seen



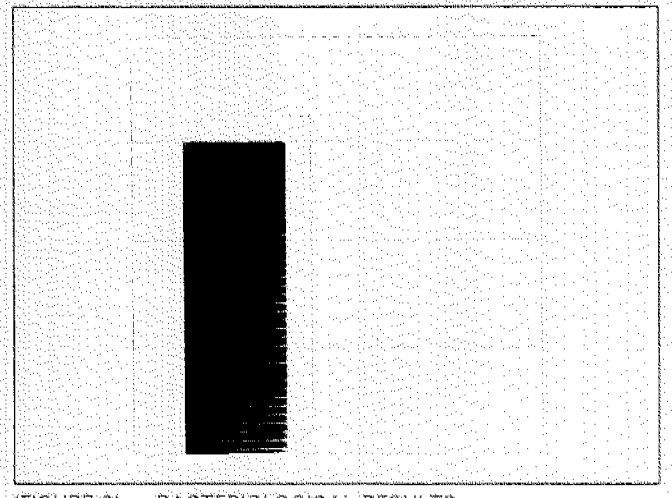
(FIGURE 2) TYPE OF PERFORATION

regarding the presence of granulation tissues (Table IIIB). Conductive hearing loss was a consistent finding in 15 (60%) of patients. Three (12%) showed sensorineural hearing loss and five (20%) had mixed hearing loss. Two were not tested. Three (12%) patients presented with facial paralysis.

II. LABORATORY AND ANCILLARY PROCEDURES:

Results of bacteriological studies are shown in Figure 3. Aural discharge sent for AFB staining all turned out to be negative, however, six (24%) of the 25 ears which were cultured were positive for Mycobacterium Tuberculosis.

X-ray of the mastoids revealed mastoiditis in all 25 cases. Of these, three had evidences of bone destruction



(FIGURE 3) BACTERIOLOGICAL RESULTS

TABLE 1 A: SEX OF PATIENTS

	NO.	PERCENTAGE
Males	14	56%
Females	11	44%

TABLE I B: AGE OF PATIENTS

	NO.	PERCENTAGE
< 9	5	20%
10 - 19	7	28%
20 - 29	5	20%
30 - 39	5	20%
40 - 49	1	4%
50 - 59	0	0%
> 60	2	8%

TABLE II A: DURATION OF SYMPTOMS BEFORE PRESENTATION

DURATION (MONTHS)	NO.	PERCENTAGE
< 3	6	24%
3 - 6	5	20%
7 - 12	7	28%
13 - 18	1	4%
19 - 24	5	20%
25 - 30	0	0%
> 30	1	4%

TABLE II B: PRESENTING SYMPTOMS

	NO.	PERCENTAGE
Otorrhea alone	11	44%
Otorrhea, Otalgia	1	4%
Otorrhea, Decreased Hearing	6	24%
Otorrhea, Otalgia Decreased Hearing	7	28%

TABLE II C: TYPE OF HEARING LOSS

Ave. Hearing Loss (dB) (WHO Classification)	CHL	SNHL	MIXED
0 - 25	0	0	0
26 - 39	2	0	0
40 - 49	6	3	3
50 - 59	5	0	2
>60	2	0	0
	15	3	5
	23		
Not Assessed	2		
TOTAL	25		

TABLE III A

A. SYMPTOMATOLOGY

	TB CULTURE	
OTORRHEA	+	-
PAINLESS	5	12
PAINFUL	1	7
PERFORATION	+	-
MULTIPLE	0	3
SINGLE	6	16
GRANULATION TISSUES	+	-
PRESENT	4	15
ABSENT	2	4
CHEST X-RAY	+	-
POSITIVE	4	12
NEGATIVE	2	7

TABLE III B:

	SPECIFICITY	SENSITIVITY	FALSE(+)	FALSE(-)
OTORRHEA	36.80%	83.33%	63.75%	16.66%
PERFORATION	84.05%	00.00%	5.78%	100.00%
GRANULATION TISSUES	21.05%	66.66%	78.94%	33.33%
CHEST X-RAY	36.84%	66.66%	63.15%	33.33%

and four (16%) showed presence of sclerosis.

Chest radiographs showed tuberculous changes in 16 (64%) patients, but only one (4%) showed cavitation. Eleven (44%) showed presence of minimal tuberculosis and four (16%) were moderately advanced. Nine (36%) were negative. A specificity of 36.84%, sensitivity of 66.66%, false positive of 63.15% and false negative of 33.33% were noted.

III. TREATMENT AND FOLLOW-UP:

Of the 25 patients, eight (32%) underwent mastoidectomy. The most common finding is the presence of abundant, pale granulation tissue. Osssicular chain was noted to be intact in one case (12.5%) partially missing in five cases (62.5%), and in two cases (25%), the ossicular chain was completely missing. Granulation tissues were sent for histopathological examination and revealed non-specific chronic inflammation in all eight cases. All were started on anti-koch's treatment after surgery. With this treatment, significant improvement was noted in two weeks time and six (75%) of the ears stopped discharging within two months.

Among the six patients who were diagnosed to have tuberculous otitis media as proven by the culture, anti-koch's treatment was likewise given for a duration of six months. All showed improvement within two weeks and ears were noted to be free from discharge within three to four weeks.

Of the eleven cases who presented with clinical signs and symptoms suspicious of a tuberculous type of otitis media, had positive chest x-ray, and presented with the constitutional symptoms of tuberculosis but showed negative results in AFB stain and culture, anti-koch's treatment was likewise started. Of these, 8 (72.7%) ears showed marked improvement within three weeks, and 5 (45.5%) were dry within three months.

DISCUSSION:

In the past, it has been mentioned that the presence of multiple perforations, painless otorrhea, abundant granulation tissues, early severe hearing loss as well as bone necrosis are features of the disease. However, the clinical picture of tuberculous otitis media has changed over the years. Thus it is an important etiology to consider in the presence of otorrhea.

In this study, 17 (68%) had painless otorrhea, often of long duration, but two (28.6%) of the eight cases who presented with otalgia were acute cases. The amount of discharge varied considerably from scanty to profuse and a factor affecting this as well as the otalgia may be the amount of superinfection present. The type and the site of perforation likewise varied, but it was generally central. Of the 25 ears affected, only three presented with multiple perforations. It has been previously noted that multiple perforations were only a feature of early pathology because at a later stage they fuse to form a large perforation. Three of our patients were seen within the first 14 days of symptoms and yet, multiple perforations were not observed. A feature of some significance seems to be the presence of pale, abundant, granulation tissues.

The degree of hearing loss may not be proportional to the damage especially in early cases. The incidence of facial palsy although not very high (3/15) ((12%) should raise the suspicion that the ear infection is tuberculous. A past or a positive family history of tuberculosis is a good diagnostic indicator and active tuberculosis of any site in the patient is most suspicious.

Chest x-ray is of great value but since it does not often show positive results in uncomplicated cases. it could not be considered a routine investigation in tuberculous otitis media. All 25 cases showed the presence of mastoiditis. Presence of bone destruction was seen in three cases. Evidence of sclerosis was observed in four patients and this may be attributed to ears in which otitis began in early childhood.

With regards bacteriology, six (24%) yielded positive culture for tubercle bacillus. It can be assumed that a majority of the tuberculous infection in our study seems to be secondary to an existing chronic infection of the middle ear. All acid - fast bacillus stains were negative, thus, it could be said that bacteriological staining may not be totally reliable since it may need considerable amount of organisms to be present in the specimen. Culture methods are similarly not very reliable since incubation periods are measured often in weeks making the investigation more confirmatory than diagnostic.

The early diagnostic of tuberculous infection in the middle ear is of utmost importance as prompt institution of anti- tuberculous treatment has a decisive influence upon the cause and its possible complications. Since otorrhea was the consistent feature in all 25 cases, the progression of improvement of this symptom was utilized as the basis of response to chemotherapy. Hearing will not be expected to resolve in such a short span of time, thus it was not used as a parameter regarding response to treatment. In 18 (72%) patients, the progress of otorrhea halted within four weeks. Twelve (48%) were dry after the first month of treatment. The high success rate may be attributed to the fact that most of our patients were diagnosed in the not so late phase of the disease.

Often the diagnosis is delayed when the tuberculous middle ear infection is the primary focus. It seems

to be difficult to isolate Koch's bacillus on culture from the middle ear discharge, presumably because of superinfection. Therefore, the diagnosis must be based primarily upon clinical findings.

Tuberculous otitis media can generally be distinguished from the usual middle ear infection by its onset, which is wihout pain or fever.[10] The secretion is usually serous initially but later on becomes purulent. A progressive loss of auditory acuity subsequently occurs. Hoowever, it is often difficult to culture the tubercle bacilli from the middle ear discharge, therefore the clinician must maintain a high degree of suspicion, perform multiple cultures and search diligently for evidence of tuberculous infection in other organisms.

In the early stages of tuberculous otitis media, the drum may present as dull in character with some dilated vessels.[18] The tympanic membrane then becomes thickened and landmarks become obliterated. Caseous tubercles in the membrane liquefy and cause single or multiple perforations, usually in the lower half of the drum. If the infection remains unchecked, the entire drum may get involved.

Myerson's experience demonstrated that "discharge from the middle ear appearing without pain in a tuberculous individual should be considered tuberculous".[3] Thus, the time and mode of onset becomes most important in the diagnosis.

A mere suspicion in the clinician's mind should be sufficient enough to reach the diagnosis of tuberculous otitis media. Provided the patient has not been treated with anti-tuberculous medications or with antibiotic ear drops, staining and culture of the aural discharge should be performed which may probably confirm the diagnosis.

CONCLUSION:

Tuberculous otitis media is today an uncommon disease which, if left untreated causes great damage to the middle ear and its surrounding structures. This paper has proven that it is definitely not easy to diagnose and thus the otolaryngologist should be made aware of its features and maintain a high index of suspicion. Although it may not be a common cause of chronic otitis media, it is still an important etiology that should be considered particularly in developing countries such as the Philippines.

The incidence of tuberculous otitis media varies considerably depending on the socio-economic status and race. This study was conducted among patients belonging to the lower socio-economic strata with 18 (72%) found to be living in crowded conditions.

Mycobacterium tuberculosis was isolated in six (24%) ears using BACTEC culture. All had positive exposure to tuberculosis, majority has painless otorrhea with the discharge described to be serous, non-foul in character. Otoscopic findings showed presence of perforation and abundant granulation tissues. Audiometric studies done revealed presence of conductive hearing loss in two cases and mixed hearing loss in one. Acid-fast bacillus staining of submitted specimen of aural discharge all turned out to be negative. This is primarily due to the fact that a considerable amount of the organism should be present in the specimen. Since there are other organisms which can cause secondary infection to the disease and can interfere with the growth of the tubercle bacilli, as well as its fastidious nature, staining may not be regarded as reliable.

Likewise, we should be able to consider that the false negatives for culture often occur, due both to the nature of the tubercle bacilli as well as other organisms in the specimen which may interfere with the growth of the bacillus. The slow growth and unreliability of cultures limit their diagnostic value and makes thorough examination and maintenance of a high index of suspicion mandatory.

Chest radiographs on the other hand were found to be negative in three of the six confirmed cases. This may be attributed to the fact that the ear problem might be the primary focus and as earlier mentioned, there are cases where it may not turn out to be positive especially in uncomplicated cases.

Chemotherapy has been considered to be the cornerstone of management. Satisfactory results have been obtained in this study. More often than not, the correct diagnosis is usually made with the failure of treatment and the chronic recurrence of symtpoms.

Early diagnosis is difficult because of the following reasons:

1. That only 0.4% of chronic suppurative otitis media cases are of tuberculous origin,[5] thus the index of suspicion is often low.
2. The clinical signs of tuberculous otitis media have changed in recent years making it more difficult to recognize.[9,15]
3. False negative cultures occur because of the nature of the tubercle as well as the other bacteria in the specimen which may interfere with the growth of the bacillus.[17,18]

It is then worth mentioning that the Otolaryngologist should be ever mindful to include tuberculous otitis media in his differential diagnosis in chronic ear infections.

The lack of awareness of the manifestations of tuberculous otitis media have resulted in a decreased suspicion of the tuberculous etiology for many symptoms. As a result, tuberculosis continues to flourish in certain parts of the world.

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THE EXPRESSION OF INTERMEDIATE FILAMENT PROTEINS IN MALIGNANCIES OF THE HEAD AND NECK.
Preliminary Report*

GERARD C. AGCAOILI, M** • GLORIA DE CASTRO-BERNAS, MS
ANTONIETA C. LIMJOCO, MD • EDGARDO B. PERALTA, MD
RAMON L. SIN, MD

ABSTRACT

Twenty seven surgically excised and biopsied tumor specimens were investigated immunohistochemically for the presence of intermediate filament proteins using the immunoperoxidase method and a commercial kit (Ultra). All these tumors were taken from head and neck malignancies previously diagnosed using the standard Hematoxylin and Eosin staining method. From the results obtained, it appears that intermediate filaments cytokeratin and vimentin are the two most widely expressed tumor markers appearing in almost all head and neck specimens gathered. A high specificity has likewise provided a more definitive diagnosis of undifferentiated tumors and those that present with constitutional characteristics. This initial study, however limited, has shown that intermediate filament proteins have a significant role in the determination of tissue origin, specifically in cases which present with diagnostic dilemmas.

INTRODUCTION

Neoplasia represents the most baffling complex of diseases yet to confront medical science.

The era of modern thinking concerning the nature of cancer and its etiologic factors began with the development of the achromatic microscope in 1824. There followed a systematic study of specific types of cancer s and their classification into major groups on the basis of histopatologic characteristics. The tendency to regard common histologic types as representing the same disease process, regardless of the organs or the anatomic sites in which they occurred, led to the concept that "tumors fall into a limited number of grand classes".

The accuracy in diagnosis of this tumors are only as good as the pathologists or clinician who identiffies or describes them. The diagnosis of cancer, therefore, has been limited to the morphology and structure of these tumor cells, until Green and Brunnet, and Miller and Miller in 1952 postulated the presence of certain specific cell antigens in cancer cells which they termed "self-markers" (1).

Tumor diagnosis in surgical pathology is still based largely on morphologic aspects of tissues and cells. Depending on the experience of the pathologist, most tumors can be typed by routine histologic staining technique applied to paraffin-embedded sections. However, a certain proportion of neoplasms may confront the histopathologist with serious problems in diagnosis. In such cases, additional techniques are applied to obtain information relevant to the tumor-type and the tissue origin. Such techniques may include study of the ultrastructure of the tumor as well as its biochemical characteristics. It is, therefore, not surprising that immunohistochemistry is being used increasingly in tumor pathology (2). Particularly when performed with well-defined monoclonal antibodies, these procedures complement morphologic studies and may be a particularly strong tool because fine needle aspiration biopsy specimens, sputum smears or cytologic preparations from body cavity effusions may be difficult to interpret. The advantage of immunohistochemistry over routine cytologic staining is that biochemical processes command the fate of the cells and occur ahead of morphologic changes. Changes in morphology are actually late consequences of all cellular biochemical processes. Thus, if products of the biochemical reaction are to be assayed, these would be more representative of cellular changes induced by malignant processes. This would also reflect the direction towards which a neoplasm will differentiate.

During embryogenesis, cells undergo proliferation, differentiation and progressive specialization. Microtubules, microfilaments and intermediate filaments are present in these cells. The type of intermediate filament expressed depends on the degree of differentiation and direction towards which the cells will differentiate.

The first intermediate filament to be expressed is

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** Resident, Dept. of ENT, UST

vimentin. As the cells become more specialized, they also start to express other intermediate filaments in combination with vimentin, depending on the direction towards which the cells will differentiate. As they become more differentiated, they finally express only one intermediate filament.

The role, therefore, of tumor markers comes of utmost importance and the attention on its uses, particularly of the intermediate filament proteins, has for this purpose, become very vital. Most normal cells contain 3 classes of filamentous structures that form part of the cytoskeleton: 1) Microfilaments - 5 to 6 nm in diameter, 2) Microtubules - 25 nm in diameter, are present in all cells and show no changes in composition, and 3) Intermediate Filaments - cytoplastic, polymeric structures that are 10-12 nm in diameter. They may be present in most mammalian cells and have been shown to be tissue-specific to a certain extent. Five major types of intermediate filament proteins have been distinguished and the distribution of these proteins has been shown to follow well-known histologic patterns.

Intermediate filament proteins can be divided into five major biochemically and immunologically distinct classes. The expression of the intermediate filament subclasses is developmentally regulated and shows a remarkable tissue and cell-type specificity. Thus, cytokeratin occurs in epithelial cells, vimentin in mesenchymal cells, desmin in muscle cells, neurofilament proteins in neural cells, and glial fibrillary acidic protein in glial cells (Table 1) (3).

TABLE 1. BIOCHEMICAL DIFFERENTIATION OF INTERMEDIATE FILAMENT PROTEINS

Interm Fil.	Diameter (nm)	Mol. Wt. (Daltons)	Distribution
Keratins	8-10	40-65	Epithelial and Meso- epithelial cells
Desmins	8-10	50	Muscle Cells, Fibroblasts
Vimentin	8-10	52	All Mesenchymal Cells
Glial Filament	8-10	51	Glial Cells
Neuro Filament	8-10	68;160;210	Neurons

Monoclonal antibodies raised against intermediate filament proteins are used to determine their expression in fixed cell lines and tumor sections.

This tissue-specific distribution of intermediate filament proteins, therefore, permits their use as markers in histopathology.

Monoclonal and polyclonal antibodies to the different types of intermediate filament proteins have been prepared by several groups and many of these are commercially available. It has been mentioned that these antibodies react in a tissue-specific manner, such that in general, antibodies to cytokeratin react only with epithelial tissues, antobodies to desmin, only with muscle tissues, glial fibrillary acidic proteins, only with glial cells and neurofilament antibodies, only with glial and certain neuroendocrine cells. Antibodies to vimentin normally react only with cells and tissue of mesenchymal origin, but for this type of intermediate filament proteins, several important exceptions have to be recognized. For example, mesothelial cells have been shown to coexpress cytokeratins and vimentin, a phenomenon also seen in some other types of epithelial cells. The different types of tissues listed in Table 2 can be distinguished by immunohistochemical methods, using specific antibodies to intermediate filament proteins (4).

TABLE 2. TISSUE AND TUMOR SPECIFICITY TO INTERMEDIATE FILAMENTS

Type of Intermediate Filament Proteins	Tissue Type	Tumor Type
Cytokeratin	Epithelial	Carcinomas
Vimentin	Mesenchymal	Lymphomas, Sarcomas
Desmin	Muscle	Myocarcinomas
Glial Fibrillary	Astroglial	AstrocytomaAcidic Protein
Neurofilament Protein	Nerve	Neural Tumors

Monoclonal anti-cytokeratin is immunospecific for cytokeratins 13 and 16 as demonstrated by immunoblotting analysis on isolated cytokeratin polypeptides. The antibody specifically stains non-keratinizing squamous epithelial, basal layer of pseudostratified epithelia and transitional epithelium. It does not react with epidermis of simple epithelia.

Each epithelial tissue has a specific and stable pattern of expression of some of these cytokeratin subunits. Epithelium-derived tumors maintain the expression of the cytokeratins found in the normal tissue of origin. Monoclonal anti-keratin may be used in the differentiation of epithelia tumor origin.

Monoclonal anti-Vimentin is immunospecific for vimentin as determined by indirect immunoflourescent staining. When used with immunoperoxidase or imunoflourescent labeling of frozen tissue section, it localizes vimentin in fibroblasts, endothelial cells, lymphoid tissues, melanocytes, etc. Similarly, the antibodies stain tumors derived from these cells, including sarcomas, lymphomas, melanomas, etc. and their metastatic lesions.

Anti-desmin is an antiserum to smooth muscle desmin, the protein sub-unit of muscle-type intermediate filaments. Desmin is an intermediate filament found to exist exclusively in skeletal muscle, cardiac muscle, visceral smooth muscle and smooth muscle of some blood vessels. The expression of desmin is therefore useful as a marker for muscle differentiation in a tumor.

Monoclonal anti-neurofilament stains fibrous profiles in neuronal perikarya, dendrites and axons. The antibody does not cross-react with other intermediate filament proteins, and reacts specifically by recognizing neurofilaments in the central and peripheral nervous system.

Using immunoperoxidase labeling on alcohol and formalin fixed paraffin embedded or frozen tissue sections, the antibody localized glial fibrillary acidic protein in astrocytes, glia cells (Bergmann glia), gliomas and other glial-cell derived tumors. It has proven to be a valuable tool for immunochemistry localization of GFAP in normal central nervous system tissue and certain tumors, likewise metastases of glial origin.

Proper and accurate diagnosis of cancer has thus remained vital to patient management. The aim of this study is to determine the pattern of expression of intermediate filaments among various tumors, particularly in this field of head and neck surgery.

MATERIALS AND METHODS

Twenty seven (27) surgically excised and biopsied tumor specimens were investigated immunohistochemically for the presence of intermediate filament proteins. All these tumors were taken from head and neck cancers earlier diagnosed using the standard Hematoxylin and Eosin Staining Method.

Tissues were fixed in 10% formaldehyde and embedded in paraffin, and serial sections were cut at 4 micra. Immunohistochemical procedures were performed with the use of the immunoperoxidase method and a commercial kit (Ultra).

Initial staining was carried out using monoclonal mouse antibody (Ultra) and incubated overnight in a moist chamber at room temperature. A second antibody, rabbit anti-mouse, was then introduced and allowed to stand for one hour, followed by a third antibody, swine anti-rabbit (Ultra) and again incubated for one hour. The specimens were treated with a substrate reagent (Amino-ethyl carbazole + acetate buffer (pH = 5.0) + peroxide) and color developments were monitored until optimal color reaction was noted. The reaction was stopped by immersing the slides in distilled water. The specimen was then mounted using water-base glycerol-gelatin solution.

TABLE 3. EXPRESSION OF SQUAMOUS KERATIN & VIMENTIN IMMUNOREACTIVITY IN HEAD AND NECK MALIGNANCIES

	Squamous Keratin	Vimentin
Basal Cell Carcinoma	1/1	1/1
Adenocarcinoma	2/2	1/2
Fibrosarcoma	1/1	1/1
Undifferentiated CA	0/1	1/1
Malignant Lymphoma	0/1	1/1
Squamous Cell CA		
Well Differentiated	5/7	7/7
Moderately Differentiated	1/2	2/2
Poorly Differentiated	0/1	1/1
Thyroid Carcinoma		
Papillary Carcinoma	4/7	6/7
Follicular Carcinoma	1/3	3/3
Anaplastic Carcinoma	0/1	0/1

RESULTS

The main results are tabulated in Table 3.

Squamous Cell Carcinoma, Poorly Differentiated

Forty percent of the tumor cells reacted with molecular antibodies against squamous keratin. One hundred percent of the overlying suprabasal epithelium reacted intensely (+++). Other layers were non-reactive.

Sqaumous Cell Carcinoma, Moderately Differentiated

There was an almost complete staining of cells for both squamous keratin and vimentin, with a likewise even distribution of strong, moderate and weakly staining characteristics.

Squamous Cell Carcinoma, Well Differentiated

Six tissue sections were made to react with squamous keratin and vimentin antibodies. Majority of the samples showed 90% - 100% staining with anti-Squamous Keratin. One sample showed only 50% reaction with anti-squamous keratin. Reaction with vimentin is 60% in all tissue sections.

Basal Cell Carcinoma

Ninety percent of all cells reacted with anti-squamous keratin antibodies and 50% with anti-vimentin.

Adenocarcinoma (Cervical LN)

Ninety percent reacted with anti-squamous keratin and another 90% reacted with anti-vimentin antibody. Another specimen, however, showed only a 70% reaction with anti-squamous keratin, while there was no reaction with anti-vimentin antibody.

Fibrosarcoma

Sixty percent of the tumor cells reacted with anti-squamous keratin, while 70% of the same tumor cells reacted with anti-vimentin antibody.

Undifferentiated Carcinoma

Only 40% of the tumor cells reacted with anti-squamous keratin antibody, presenting with a weak staining characteristic. Only 65% however, of these tumor cells reacted strongly with anti-vimentin antibody.

Malignant Lymphoma

Eighty percent of cells reacted strongly with anti-vimentin antibody, while 10% reacted weakly with anti-squamous keratin antibody.

Anaplastic Carcinoma, Thyroid

Thirty percent of the tumor cells reacted with anti-squamous keratin while 20% reacted with Anti-vimentin antibody.

Papillary Carcinoma, Thyroid

Four out of five specimens showed that the papillary lesion had a 100% reaction with anti-squamous keratin. One specimen gave an 80% reaction to the same antibody.

Follicular Carcinoma, Thyroid

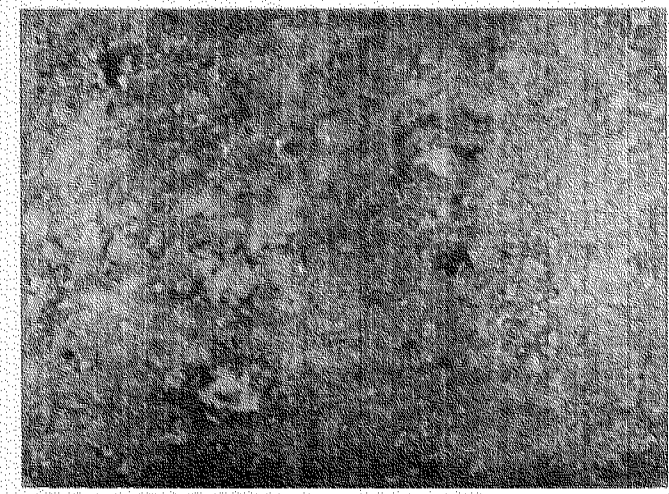
Only one of the three specimens reacted with anti-squamous keratin antibody by 50%. Anti-vimentin reactions was seen in all of these three. Two specimens (slide) did not react with anti-squamous keratin. This however, showed that 60% of the tumor cells reacted with anti-vimentin antibody.

DISCUSSION

In this study, an analysis of the cytoskeletal intermediate filament proteins in a random group of tumors was done as a preliminary evaluation into their patterns of expression among the head and neck malignancies gathered from a three year survey of cases at the Clini... Division of Santo Tomas University Hospital.

Specimens prepared as earlier described were individually read and interpreted under light microscopy. Positive results were presented by the presence of red deposits brought about by peroxidase activity and enhanced by aminoethyl-carbazole as indicator compound (5).

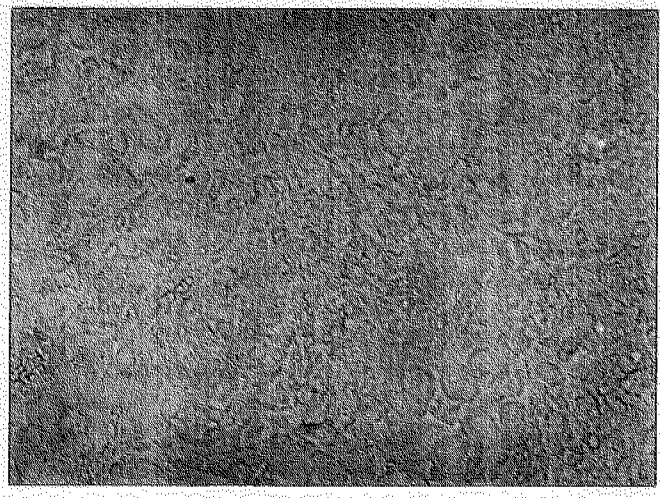
From the results obtained, it appears that among the major groups of intermediate filament proteins,



(FIGURE 1) MODERATELY DIFFERENTIATED SQUAMOUS CELL CARCINOMA (HPO), REACTION WITH ANTISQUAMOUS KERATIN ANTIBODY

cytokeratin annd vimentin are the two most widely expressed, appearing in almost all the head and neck tumors gathered.

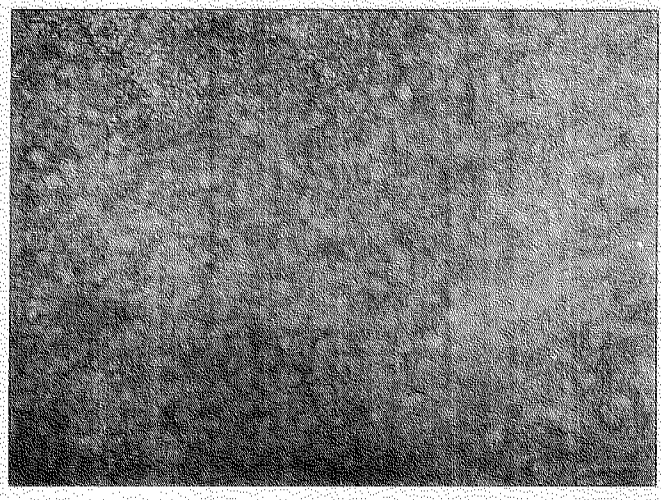
Cytokeratin as mentioned earlier is distributed among epithelial and mesoepithelial cells. It is not unusual for such an intermediate filament protein to be strongly manifested among malignancies of the head and neck considering the embryologic nature of these tumors. A very considerable majority of head and neck malignancies are carcinomas and are, thus, highly ex-



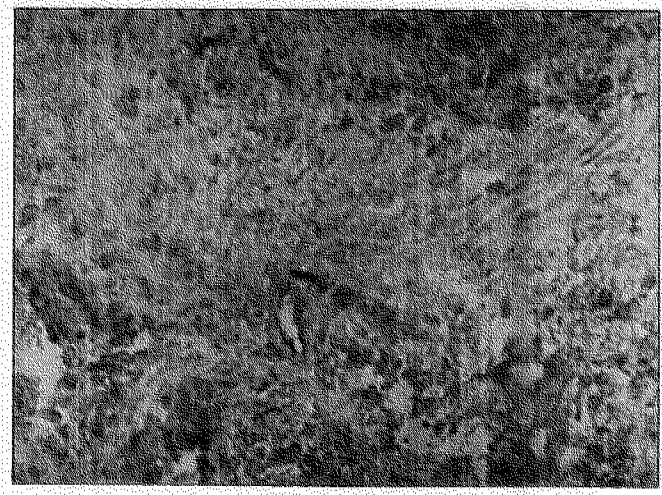
(FIGURE 2) UNDIFFERENTIATED CARCINOMA, TONSIL REACTION WITH ANTICYTOKERATIN ANTIBODY (HPO)

pressive of cytokeratin.

It is worth noting that several monoclonal antibodies to the different cytokeratin have been prepared, and diffferent reactivities of such antibodies with different subtypes of epithelial tissues and epithelial tumors have



(FIGURE 3) UNDIFFERENTIATED CARCINOMA, TONSIL REACTION WITH ANTI-VIMENTIN ANTIBODY (HPO)



(FIGURE 4) MALIGNANT LYMPHOMA, PAROTID GLAND REACTION WITH ANTI-VIMENTIN ANTIBODY

been observed. In addition to broadly cross-reacting antibodies which stain virtually all types of epithelial tissues, monoclonal cytokeratin antibodies which cross-react with only one of the 19 cytokeratin polypeptides, and which show a more specific staining pattern, have been developed.

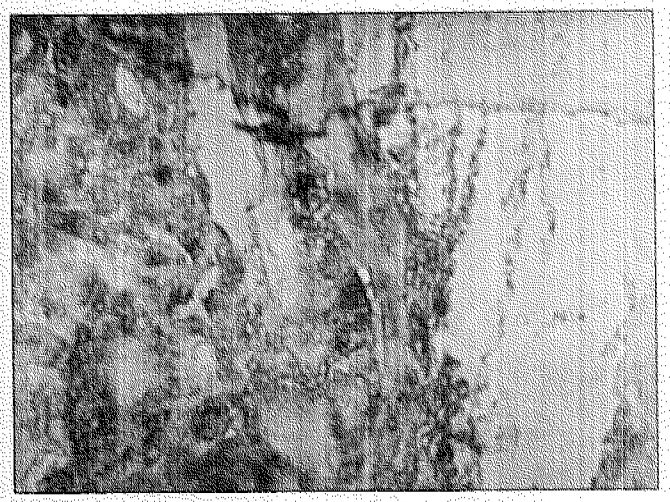
This is clearly exemplified in Figure 1, which shows this sharp contrast in stain between the moderately differentiated squamous cell carcinoma of the

(FIGURE 5) MALIGNANT LYMPHOMA, PAROTID GLAND REACTION WITH ANTISQUAMOUS KERATIN ANTIBODY

nasopharynx, in contrast to its negative control which does not take any stain at all. Very distinct is its brick red color that signifies a highly positive result in comparison to the brownish, faded presentation of the control.

In relation to what has just been mentioned, a very strong indication of these tumor markers has been in the diagnostic dilemma posed by poorly differentiated and

(FIGURE 6) MALIGNANT LYMPHOMA, PAROTID NEGATIVE CONTROL



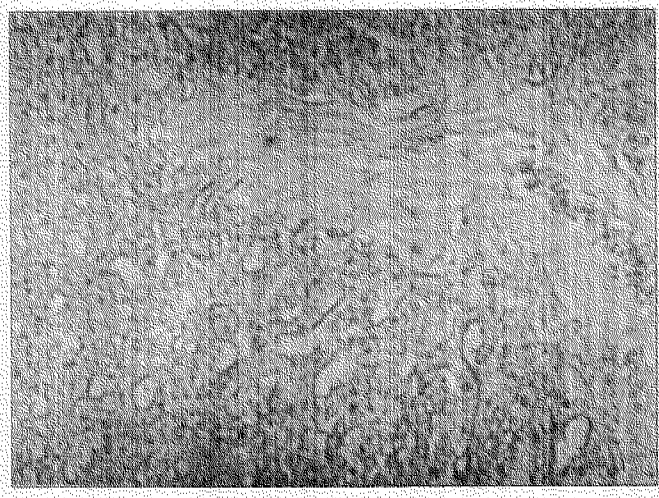
(FIGURE 7) PAPILLARY CARCINOMA, THYROID GLAND REACTION WITH ANTISQUAMOUS KERATIN ANTIBODY

undifferentiated cancers. A good example is seen in the following specimen of undifferentiated carcinoma of the tonsils. As expected, the negative control showed a very faint, somewhat brownish stain. This is compared with the following slide, (Fig. 2) stained for cytokeratin, showing a minimal stain uptake. A review, however, of its corresponding slide stained with anti-vimentin antibody shows a strongly positive result (Figure 3), evidence that the undifferentiated carcinoma has a greater pattern of expression towards a lymphoma than a carcinoma. This diagnosis would therefore greatly affect patient managemennt and prognosis.

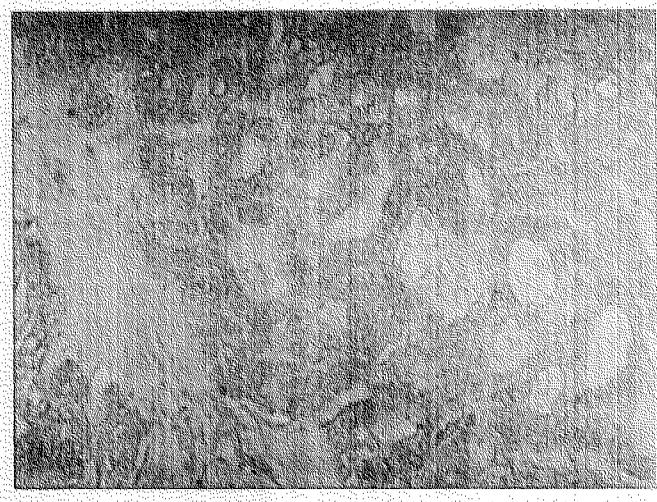
Similarly, Figure 4 shows the highly positive expression of vimentin on a previously diagnosed case of malignant lymphoma of the parotoid gland, in comparison to the degree of expression of squamous keratin for the same specimen (Fig. 5).

It is worth mentioning at this point that in contrast to normal adult human tissue, certain neoplastic tissues may co-express different types of intermediate filament protein. When the tumor cells contain more than one intermediate filament protein, vimentin is one of them in most cases. In epithelial tumors, vimentin has been found in addition to cytokeratin as in pleomorphic adenomas of the parotid gland, adenoid cystic carcinomas of the salivary gland, some adenocarcinomas of the lungs, thyroid carcinomas and mesotheliomas (4).

Despite the fact that malignant lymphomas should react only with anti-vimentin antibodies, the presence of an immunohisto- chemical reaction with antisquamous keratin, antibodies, however weak, can be explained on the basis of the histological nature of the basal cell component of ductal cells. Despite all these, the factremains that vimentin, in such a tissue, is still the

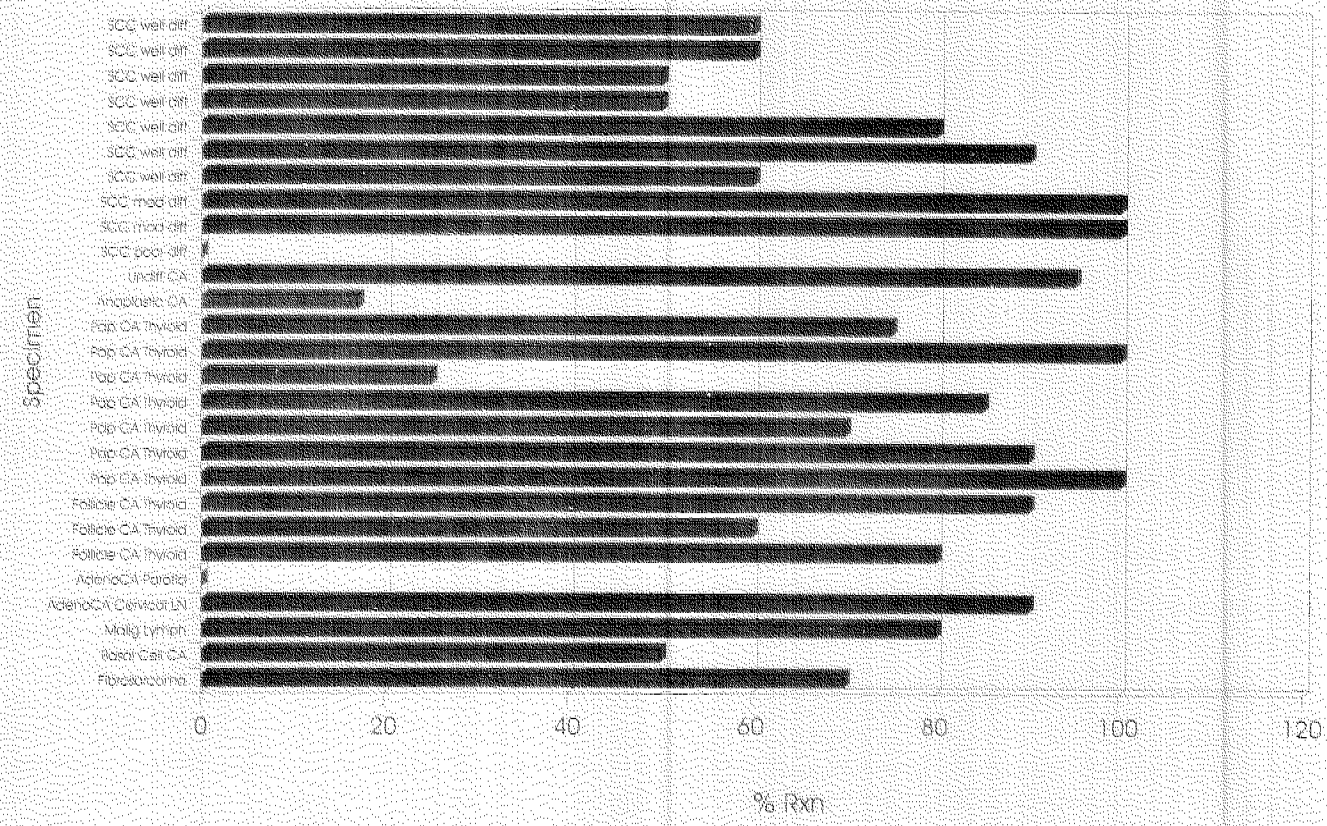


(FIGURE 8) PAPILLARY CARCINOMA, THYROID GLAND,

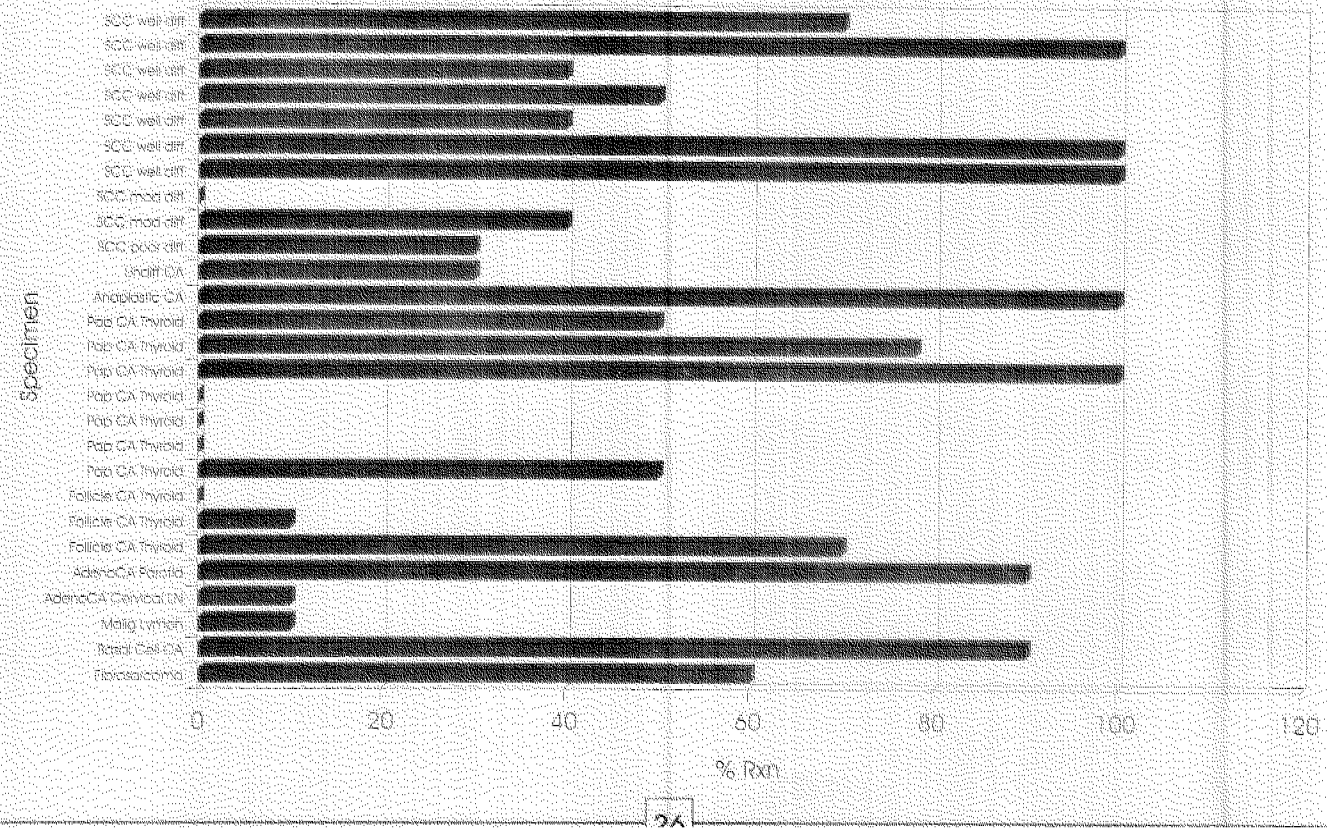


(FIGURE 9) PAPILLARY CARCINOMA, THYROID GLAND, RE-

IMMUNOHISTOCHEM STAINING RXN OF H&N MALIGNANCIES TO VIMENTIN



IMMUNOHISTOCHEM STAINING RXN OF H&N MALIGNANCIES TO SQUAMOUS KERATIN



primary intermediate filament to consider.

Adenocarcinomas may likewise present such a condition. All adenocarcinomas, regardless of the cell of origin express keratins that are readily detectable with broadly reactive keratin monoclonal antibodies. Statements to the contrary in earlier reports are attributable to the use of antisera of restricted specificity or low sensitivity, or to the ommission of protease digestion when the tissues are fixed with formalin. These findings indicate that broadly reactive keratin monoclonal antibodies could be used as a tool for the microinvasion or micrometastases, since even single neoplastic cells can be demonstrated readily in organs that do not normally contain keratin such as lymph nodes.

Malignancies of the thyroid gland on the other hand present with an even more unstable pattern of expression. The need, however, for an immunohistochemical test such as this is due to the fact that most papillary and follicular carcinomas of the thyroid gland, though easily diagnosed on the basis of histologic features, may occasionally show some features characteristic of both tumor types (7). As presented in our study and in other researches, the difference in expression of cytokeratins provides additional aid in the differential diagnosis between these tumors. The difference in cytokeratin expression between papillary and follicular carcinomas suggest that these tumors originate from different types of epithelial cells, with the possibility that follicular carcinomas from follicular epithelial cells lacking reactivity with the antiepidermal prekeratin antibodies. Figure 7 shows a strongly positive reaction of Papillary Thyroid Carcinoma with anti-squamous keratin antibody in contrast to its negative control (Fig. 8) and a weakly reacting sample stained with anti-Vimentin antibody. (Fig. 9).

CONCLUSION

Intermediate filament proteins are still considered to be the most reliable markers of tissue origin. (8) Particularly in the field of head and neck surgery, accurate tissue diagnosis is necessary for corresponding patient management. A need, however, for a more detailed study into the specific tumor types in the head and neck is still necessary for a more definitive analysis into the pattern of expression among these intermediate filaments. This initial study, though limited, has shown that the intermediate filaments have a significant role in the determination of tissue origin, specifically in those cases which present with diagnostic dilemmas.

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SAM BERNAL, MD, PhD -
Boston University, Mass. USA

TERESITA TUAZON, MD -
Head, Laboratory Medicine,
Pathology Department, National Kidney Institute, Quezon City

DEPARTMENT OF BIOCHEMISTRY,
University of Sto. Tomas
Faculty of Medicine and Surgery

DEPARTMENT OF ANATOMIC PATHOLOGY,
University of Santo Tomas
Faculty of Medicine and Surgery

NATIONAL KIDNEY INSTITUTE,
Special Research Laboratory

MANDIBULAR TUMOR: A PATHOLOGIC DILEMMA *

Deo Talanay, MD **

INTRODUCTION:

"Things are never what they seem." Such is the dictum encountered often in everyday life. This is applicable, even more so in the art of medicine. The Otolaryngologist-Head and Neck Surgeon is not spared from such a plight. Oftentimes, tumors of the head and neck appear easy to diagnose at first glance. However, in a few cases, once one investigates deeper, the more baffling it gets. One is then left in a quandary as discrepancies between clinical and pathologic diagnosis arises. One of the "Gray Zones" in histopathologic diagnosis includes soft tissue tumors, specifically those found in the head and neck. It is in these conditions that controversies often arise. At times, there would be discrepancies between clinical and pathologic diagnoses in which case, proper management is compromised.

The case being presented now is a perfect example of such a dilemma. Initially diagnosed as an undifferentiated malignant mesenchymal tumor, surgical extirpation of the lesion was done. However, final histopathologic confirmation exhibited an entirely different entity, one possessing "pleomorphic round cleaved cells".

CASE HISTORY:

A 47 year old male consulted the out patient department of Ospital ng Maynila due to a mass over the left mandibular area of three (3) months duration. Initially, the lesion was about 2 x 2 cm. firm, movable, with well delineated borders, associated with left hemifacial pain and toothache. Dental consultation was sought for which Bacampicillin 800 mg BID and Mefenamic Acid 500 mg QID was given. No relief was noted. The mass persistently grew in size to about 11 x 9 cm with its superior border encroaching the left malar area. Significant weight loss and anorexia were noted. As the mass enlarged, it became exquisitely tender and assumed a whorl-like consistency.

On admission, pertinent physical examination findings revealed limited mouth opening, mobile ipsilateral mandibular molars, nodulations along the buccal area with an intact mucosal surface, and an 11 x 9 cm mass along the angle of the left hemimandible. The mass was firm, tender, with a whorl-like consistency, its outer borders encroaching the parotid compartment, malar and submental regions of the head. No cervical lymphadenopathies were noted. Sensory and motor functions of the left hemifacial region were intact. Admitting impression was a Left Mandibular Mass to consider Malignancy.

Pertinent radiographic evaluation of the maxillofacial bony framework showed erosion of the left hemimandible up to the level of the mentum. A soft tissue density was evident over the lateral aspect of the left hemimandible. Fine needle aspiration cytology studies were inconclusive. Wedge incision biopsy was done over the left buccal area and histopathological studies revealed an undifferentiated malignant mesenchymal tumor.

The patient underwent wide excision of the mass with left hemimandibulectomy and left total parotidectomy with facial nerve sacrifice. No surgical reconstruction was done and the defect was closed primarily.

Grossly, the specimen obtained from the left hemimandible weighed 250 gm and measured 10 x 8 cm. The outer surface was dark brown with severe areas of congestion. Cut sections showed a fleshy, soft cut surface. Microscopic sections disclosed a very cellular tumor consisting of a monotomy of pleomorphic round cells. The individual cells varied in size and showed vesicular nuclei with distinct nucleoli and ample amount of cytoplasmic ream. Most of the cells were hyperchromatic and showed clefts. Some of the lesions showed a surrounding fibrocollagenous capsule and muscular infiltrations. The final diagnosis was given as PRIMARY INTRAOSSEOUS MALIGNANT LYMPHOMA, MANDIBLE (DIFFUSE TYPE, LARGE AND SMALL CLEAVED CELLS).

The postoperative clinical course was unremark-

* Presented PSO-HNS Interesting Case Contest held at Hotel Nikko, Manila Garden, July 9, 1992.

** Residents, Dept. of ENT, OM - PLM

able. Consultation with a medical oncologist was sought and postoperative adjuvant chemotherapy or radiotherapy was suggested. The patient underwent a full course of radiotherapy six (6) weeks after the operation.

DISCUSSION:

From the preceding discussion, one can observe glaringly the pitfalls of histopathologic diagnosis. When one depends on histopathologic results to be able to decide on subsequnet management, it is imperative that an accurate assessment of the lesion be obtained. Otherwise, the patient's well-being is sacrificed and the surgeon would be put on the spot. However, this point is raised not to condemn anyone. In this particular case, neither the pathologist nor the surgeon is at fault. It is the nature of the lesion itself which made diagnosis difficult.

Under the heading "Aggressive Mandibular Newgrowths", about four entities come into mind: (1) Osteosarcoma, (2) Osteoblastoma, (3) Histiocytosis X and (4) Primary Malignant Lymphoma. Of these, osteosarcoma appears to be the most logical choice as an initial primary consideration, it being the most common in terms of overall incidence. However, if prevalence in terms of anatomic location is taken into consideration, it is not that common in the head and neck.

In terms of clinical presentation, the above conditions usually have similar features of palpable mass, pain and tooth mobility. In other words, one has to ultimately rely on microscopy to clinch a diagnosis. Again, this is easier said than done and would depend on the skill of the histopathologist.

Osteosarcomas would present with mature, spindle-shaped osteocytes with malignant characteristics. In order to say it is osteosarcoma, demonstrable osteoid directly synthesized by the malignant osteocytes should be present.

Osteoblastomas, though containing osteoid, display proliferating round osteoblasts instead of the more mature osteocytes.

Histiocytosis X and lymphomas are reticuloendothelial tumors which have similar features. The former, however, presents with proliferating malignant histiocytes and macrophages, while the latter will show malignant lymphocytes.

In a review of 116 reported cases of intraosseous Non-Hodgkin's Lymphoma, only five (5) were reported to be solitary lesions in the mandible. At present, no local literature has reported a similar case of primary malignant lymphoma of the mandible. It is interesting to note that inspite the aggressiveness of the lesion, patients with this condition, in comparison to other osseous malignancies, possess a relatively good well-being and favorable prognosis after appropriate treatment modalities have been instituted.

From the above discussion, it is thus easy to understand why diagnosis in these particular lesions is quite hard, and discrepancies thus often are encountered. This would, perhaps, be compounded if an inadequate, unrepresentative sample of the tumor is obtained for preoperative diagnosis.

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PNEUMOCEPHALUS
A DREADED COMPLICATION OF NASOPHARYNGEAL CATHETERS*

Rhodora E. Llamanzares-Ballestero, MD **

ABSTRACT

A case of pneumocephalus caused by a nasopharyngeal oxygen catheter in a one year old boy who had been receiving oxygen inhalation through the right nostril for nine days is presented. Pneumocephalus due to this etiology has been reported once in a foreign journal, but after extensive research, it has yet to be reported locally. Since this unusual injury maybe associated with serious complications, it must be promptly recognized and treated.

INTRODUCTION

Pneumocephalus was first described in 1884 by Chiari as an autopsy finding in a patient who died of ethmoiditis. Luckett was the first to make the roentgenographic diagnosis of pneumocephalus in 1913. Since then, more than 370 such cases have been described. Between 1914 and 1918, there were frequent reports of pneumocephalus secondary to war injuries. In Markham's series of 295 cases, trauma accounted for 73.9% and neoplasm for 12.9% of cases. Surgery in the sitting position, nitrous oxide anesthesia, and cerebrospinal fluid draining intraoperatively all have been implicated. Other causes include intracranial extension, as in sinusitis, otitis media and primary gas containing abscesses. It has also been reported secondary to a high-pressure water injury to the nose during a water-skiing fall.

Local literature review revealed no reported case of pneumocephalus caused by a nasopharyngeal oxygen catheter. Thus, we are presenting this unusual and serious complication of nasopharyngeal oxygen catheters.

CASE HISTORY:

J.G. is a 1 year old male from Laguna who was admitted for the first time at UERM Memorial Medical Center because of sudden forward protrusion of the right eye of a few minutes duration.

Patient was on his twenty-first day of confinement at the Philippine Heart Center for Asia because of bronchopneumonia and status epilepticus secondary to metabolic encephalopathy. He was brought to UERM to undergo cranial C.T. Scan. During the procedure, he suddenly developed proptosis of the right eye. C.T.Scan showed moderate amount of air density occupying the right anterior cranial fossa limited by the anterior falx medially and the frontal cortex posteriorly. The right frontal lobe was pushed posteriorly producing some deformity of the right frontal horn. Air was likewise noted at the area of the right cribiform plate of the ethmoid and right orbit producing displacement of the eyeball anterolaterally. No demonstrable retroorbital mass lesion was noted. An incidental finding of right maxillary sinusitis was also noted. Emergency referrals to Otolaryngology, Ophthalmology and Neurosurgery were made and patient was subsequently admitted at UERM for further management.

Pertinent physical examination revealed an awake, crying patient in moderate respiratory distress and decerebrate posture. Head was normocephalic. The right upper and lower eyelids were swollen, pupils were equal and briskly reactive to light. Fundoscopy was unremarkable. Hertel's exophthalmometry revealed the following results: OD-23 mm, OS-19 mm, Base-85 mm. Schiotz' tonometry showed the following intraocular pressure readings: TOD-15.9 mm Hg, TOS-10.2 mm Hg. A nasopharyngeal catheter for oxygen inhalation was noted on the right nostril. Anterior rhinoscopy revealed blood clots with watery nasal discharge on the right nasal cavity. The length of the nasal catheter inserted was around 6 centimeters and the tip was noted to be lodged in the ethmoid area. The left nasal cavity was unremarkable.

COURSE IN THE WARD:

Upon finishing the C.T.Scan, the nasal catheter on the right nostril was removed and transferred to the left nostril. Vidisic eye gel, Tears Naturale and Tobramycin ophthalmic drops were applied to the right eye.

* Second Prize, PSO-HNS Interesting Case Contest held at Hotel Nikko, Manila Garden, July 1992.

** Resident, Dept. of ENT, UERM

Oxymetazoline nasal drops and brompheniramine maleate were also given. Proptosis of the right eye gradually subsided and was completely resolved within 48 hours. Follow-up C.T. Scan done 3 days after revealed considerable diminution of the amount of air in the cranial cavity. The Neurosurgery survice deemed that no surgical intervention was needed at that moment but advised close monitoring of the patient for any signs of neurological deterioration.

DISCUSSION:

Air within the cranium usually implies a connection with the atmosphere, directly or via air-containing structures. Positive pressure in the upper airway is assumed to have forced air into the skull through a weak spot in the cranial structure, most likely the cribiform plate.

The usual manifestations of pneumocephalus are headache, dizziness, cranial nerve palsies and seizures. Management includes bed rest, analgesia, avoidance of coughing, sneezing, nose blowing and straining due to physical activity and laxative use to minimize the increase in intraabdominal pressure during a bowel movement. The most effective method for decreasing intracranial pressure is repeated lumbar taps with the removal of cerebrospinal fluid. The course is generally benign since the majority will heal during the first week. However, those who do not are at high risk for meningitis and may require surgical intervention.

The length of the catheter inserted was around 6 centimeters and it has a side and an endhole. Upon removal, erosion of the mucosa was noted. In this patient, pneumocephalus developed 9 days after insertion of the nasopharyngeal catheter.

The presumed mechanism of injury was a perforation of the cribiform plate of the ethmoid. Most likely, the nasopharyngeal catheter was positioned in such a way that it caused local pressure on the mucous membrane. The dry oxygen from the endhole may have contributed to a focal necrosis with subsequent perforation of the cribiform plate. Since oxygen flow was increased to around 8 liters per minute due to moderate respiratory distress of the patient, there was rapid enhancement of the proptosis and orbital emphysema which were evident on C.T.Scan. Hertel's exophthalmometry showed a significant difference of 4 millimeters proptosis on the right eye. Immediately upon removal of the catheter, there was evident gradual resolution of the proptosis of the right eye.

Since this condition maybe associated with serious and fatal complications, it must be promptly recognized and treated. It is therefore suggested that if ever a nasopharyngeal catheter for oxygen inhalation is to be used, the length of the catheter inserted should not exceed 5 centimeters. The direction should be downwards hugging the floor of the nasal cavity to avoid perforation of the cribiform plate of the ethmoid.

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TERATOMA OF THE ORO-ANTRAL CAVITY IN GOLDENHAR SYNDROME: A CASE REPORT*

Reynaldo Z. Astorga, MD** • Angel Villoria, MD**
Gerardo E. Cruz, MD** • Rene Lacanilao, MD**
Milagros S. Lopez, MD*** • Remigio I. Jarin, MD***

ABSTRACT

Presented with a full term female infant with multiple congenital anomalies such as oro-antral mass, bilateral auricular dysplasia, cleft palate, micrognathia, vertebral abnormalities, thymic aplasia, congenital heart disease, and corpus callosum agenesis. This is diagnosed as Goldenhar Syndrome. The oro-nasal mass was excised and was determined histopathologically to be a benign teratoma. The dilemma here lies on the timing og surgery.

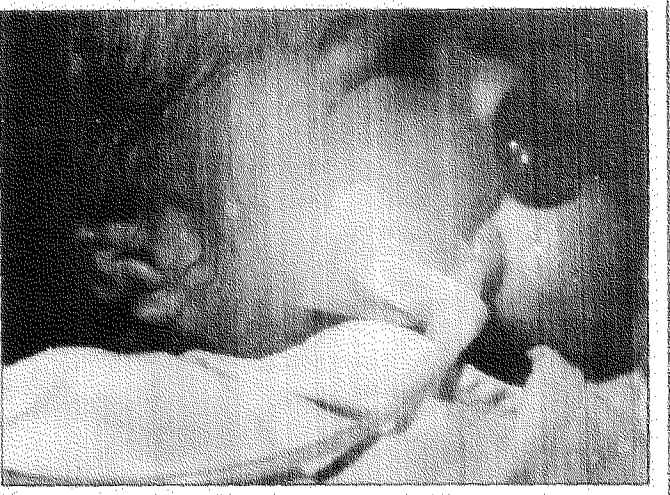
INTRODUCTION

The complexities of evaluating infants with congenital anomally have often caused an understandable dilemma among physicians towards an immediate and correct diagnosis. Some clear-cut syndrome may easily be recognized but others are so rare and complex as to be a "once in a lifetime" encounter. One may leaf through the volumes of books, journals, atlasses, and catalogues or seek consult with an expert if available. There are subjects with striking and unusual dysmorphic features which defy classification. This case about to be discussed stresses the importance of a thorough investigation on the presence of a single anomaly for other anomalies may exist.

CASE REPORT

A.A., newborn, live baby girl, full term, 38-39 weeks by Ballard's born to a 26 y/o G2P1 mother, delivered by Cesarean Section secondary to arrest in cervical dilatation, apgar score of 9 at 1 minute and 10 at 5 minutes, birth weight of 3657 grams, borderline LGA, moderate meconium-stained. BL was 50 cm. (50th percentile), HC was 36 cm. (90th percentile), CC was 36 cm. (97th percentile), and with an abdominal circumference of 34 cm.

At birth she was pinkish, active, with good cry, with a CR of 162/min., RR of 42/min., and a Temp. of 36.8°C. No skin rashes nor hematoma were noted. There was molding of the head, with no cephalohematoma. Anterior and posterior fontanels were flat, soft, and open. Bilateral ear deformities (microtia) were noted but more



(FIGURE 1) MICROTIA, RIGHT EAR. NOTE ALSO THE LARGE ORO-NASAL MASS. TAKEN FEW DAYS AFTER BIRTH.

on the right (Fig. 1). The external auditory canals were both patent and the TM intact bilaterally. External eyes were unremarkable. External nose and left nasal cavity were both unremarkable. A round, reddish, fluctuant, sessile mass measuring about 2.5 x 2.5 cm. protrudes out of the right nasal cavity. The mass is attached to the right nasal cavity floor via a narrow stalk. Likewise, a 5 x 5 cm. smooth, round, doughy, movable mass with a skin-like external surface protrudes from the oral cavity. Its peduncle measuring 2 cm. in diameter extends onto the area of the palate. A bony prominence near the attachment of the mass can be felt on palpation. An incomplete cleft of the 2° palate and micrognathia were also noted (Fig. 2-6). There was an anterior chest deformity (Barrel-shaped) however there was symmetrical expansion. Air exchange was clear. Cardiac rhythm was regular. A Grade 2/6 harsh Systolic murmur radiating

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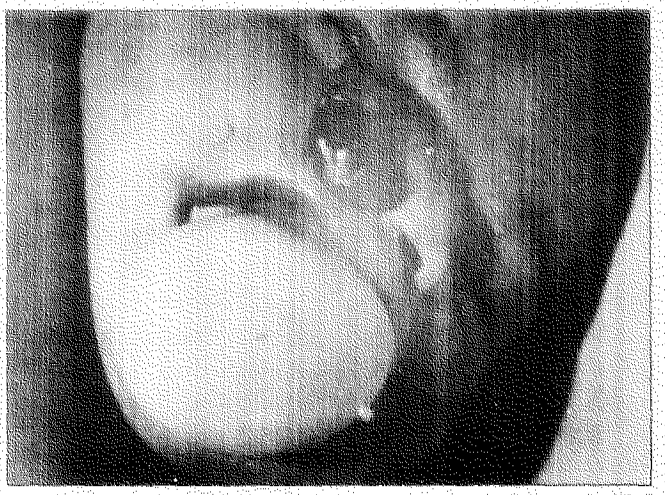
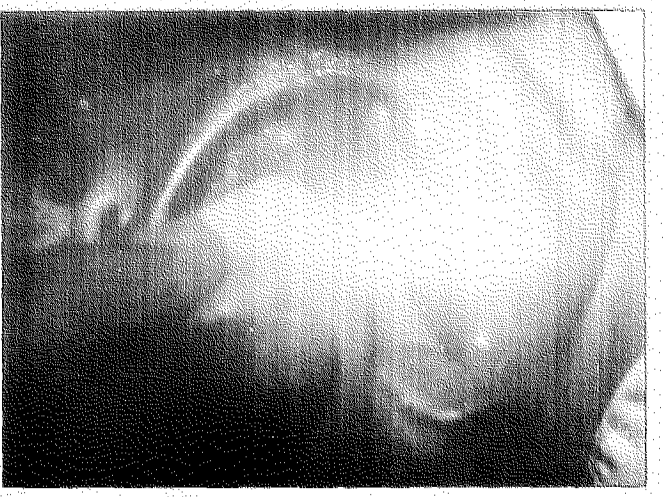
** Residents, Dept. of ENT, EAMC
*** Consultants, Dept. of ENT, EAMC



(FIGURE 2) ORO-NASAL MASS EXTENDING BEYOND THE ORAL AND NASAL CAVITY.



(FIGURE 4) ATTACHMENT OF THE MASS THROUGH A CLEFT PALATE.

to the apex was heard best at the LLSB. The abdomen was flat with NABS. No organomegaly was noted. The patient had good muscle tone. The umbilical stump and nailbeds were meconium-stained. Extremeties were unremarkable. External genitalia was grossly normal. Moro reflex was good and the anus was patent.

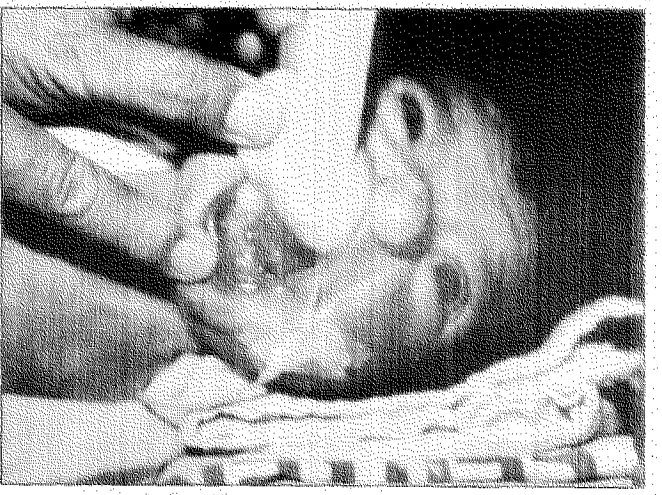
Maternal history revealed that the mother had only 2 pre-natal check-ups. She had cough and colds for 2 days at the 8th month of pregnancy. There was neither a history of drug intake nor exposure to radiation at anytime during the pregnancy.

Except for hypertension and heart disease in her paternal grandfather and aunt respectively, there were no pertinent heredofamilial diseases noted.

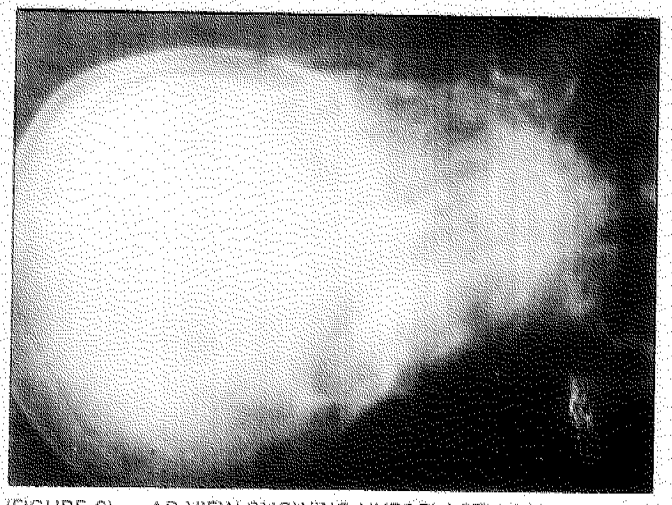
The Admitting Diagnosis was NB, LBG, FT 38-39 weeks by Ballard's born to a 26 y/o G2P1(1001) mother, delivered by CS secondary to arrest in cervical dilatation, AS 9^1 and 10^5, BW 3657 grams, borderline LGA, moderte meconium-stained. Multiple congenital anomalies: microtia, bilateral, micrognathia, barrel chest; congenital heart disease probably VSD vs. PDA to consider a multiple congenital anomally syndrome. Soft tissue tumor, oro-nasal region, probably oral teratoma and nasal cavity hemangioma R/O meningocoele for both.

After routine newborn care was done, the patient was admitted to the IMCU and OGT feedings were started. Initial CBC revealed leukopenia with a WBC of 6.80 which became persistent as shown by succeeding counts - 3.8, 5.9, 4.35, etc. Radiographic studies revealed cardiomegaly with lifting of the apex, diffuse hemivertebral abnormalities of the thoraco-lumbar spine (Fig. 9), and micrognathia. There was no hypertelorism noted by X-ray. Radiographs of the cranial vault were

(FIGURE 3) ORO-NASAL MASS. INFANT COMFORTABLE WITH MASS.



(FIGURE 5) NOTE COLOR AND APPEARANCE OF NASAL PORTION OF THE MASS, ALMOST SIMILAR TO THE ORAL PORTION SEVERAL WEEKS AFTER BIRTH.



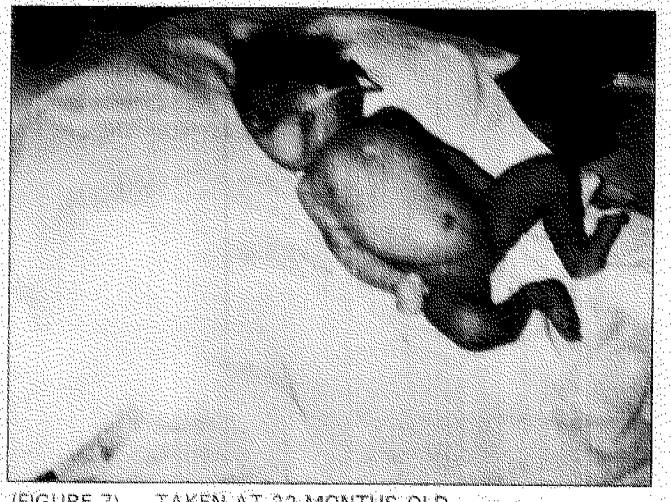
(FIGURE 6) AP VIEW SHOWING HYPOPLASTIC MANDIBLE.

unremarkable.

Examination with a 30° telescope revealed that the nasal mass was attached to the peduncle of the oral mass which in turn was attached to the inferior portion of the posterior end of the nasal septum. An incomplete cleft of the 2° palate was likewise noted (Fig. 4).

Magnetic resonance imaging showed two separate extracranial soft tissue lesions, one in the right paranasal region and the other within the oral cavity. There were no intracranial communications. Corpus callosum agenesis was noted. Other intracranial findings were unremarkable (Fig. 9-10).

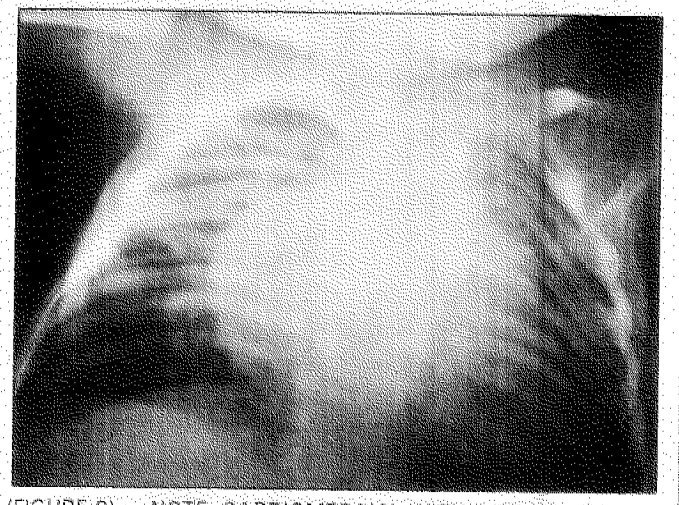
2-D echocardiogram demonstrated VSD, FDA, and mild PAH. Karyotyping was done, however the sample failed to grow on the special culture medium. The Alpha Fetoprotein was determined at 296.48 micrograms/ml.



(FIGURE 7) TAKEN AT 22 MONTHS OLD.

The patient was able to tolerate tube feedings and despite the huge oro-nasal mass, no airway obstruction was noted. The patient however developed pneumonia which became persistent despite adequate antibiotic coverage. The oro-nasal mass then was considered to be a major factor as far as the persistence of the pneumonia was concerned. For the infection to resolve the mass had to be removed.

On the 80th day of life, because of and despite the pneumonia, the patient underwent excision of the mass under close intra-operative monitoring. Induction of anesthesia was done smoothly through an oro-tracheal tube. Intra-operatively, the previous endoscopic findings of the oral and nasal mass connected to each other and its attachment to the inferior side of the posterior



(FIGURE 8) NOTE CARDIOMEGALY WITH LIFTING OF THE APEX. ALSO IS THE DIFFUSE HEMIVERTEBRAL ABNORMALITY OF THETHORACO-LUMBAR SPINE.

end of the nasal septum was confirmed. No connection with the nasopharyn naso pharynx was noted. A bony prominence at the area of the attachement of the mass was drilled off. The lines of excision were sutured using 4-0 Dexon sutures on opposite sides. Staphylorrhaphy was done (Fig. 11). The entire operation was uneventful. The procedure was completed in less than 2 hours with minimal blood loss (necessitating no transfusion).

On the 2nd post-op day, she again started to show signs of infection. The patient's clinical picture worsened on the 3rd post-op day and the patient went into CP arrest. The patient subsequently expired.

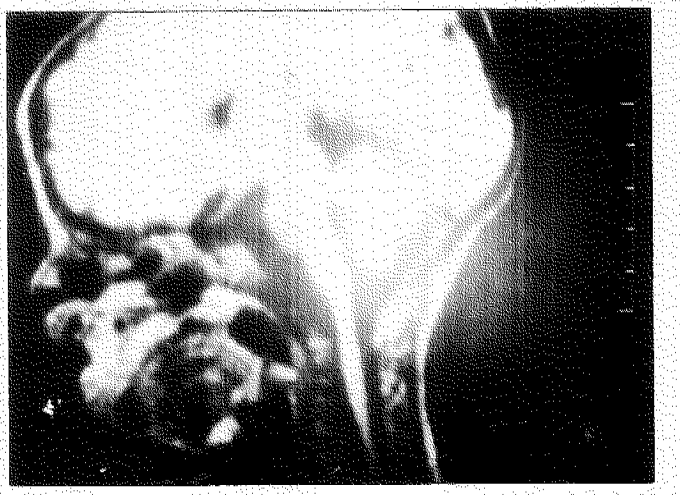
Post-mortem exam was done which revealed the following: Interstitial pneumonia, bilateral PDA (0.4 cm) with secondary RVH and pulmonary artery dilatation (Fig. 12), thymic aplasia, pulmonary congestion with atelectasis and hemorrhage bilaterally, focal necrosis with cloudy swelling of the liver, cloudy swelling and congestion of both kidneys, and congestion of

both the spleen & intestines. The cause of death was sepsis.

Fixed tissue slides were passed to different institutions for review with a total of twelve pathologists examining the slides. Histopathologic diagnosis was benign teratoma.

CASE DISCUSSION

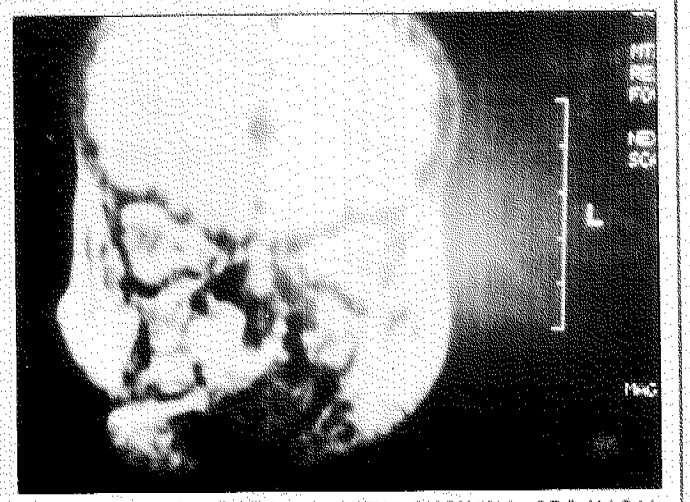
The case of a full term female infant with multiple congenital anomalies which includes an oro-nasal mass, bilateral auricular dysplasia (microtia), cleft palate, micrognathia, vertebral abnormalities, thymic aplasia, a congenital heart defect, and corpus callosum agenesis is presented. This constellation of malformations does not appear to have been reported previously. Presented with such a patient with dysmorphic features, interest naturally centered on the obviously abnormal features. Nevertheless, a detailed familial, gestational, and peri-



(FIGURE 9) MRI, SAGITTAL VIEW SHOWING CORPUS CALLOSUM AGENESIS.

natal history must never be overlooked. The predominant defects in this spectrum of anomalies represent problems in morphogenesis of the 1st and 2nd branchial arches.

The principal features of the head and neck develop primarily from the branchial or pharyngeal arches. These arches appear in the 4th and 5th week of development. The auricle develops from six mesenchymal proliferations located at the dorsal ends of the 1st and 2nd pharyngeal arches. The maxillary and mandibular processes, which contribute greatly to the formation of the facial skeleton through membraneous ossification, are likewise derived from the 1st branchial arch. During the 4th day of embryogenesis, the vertebral column

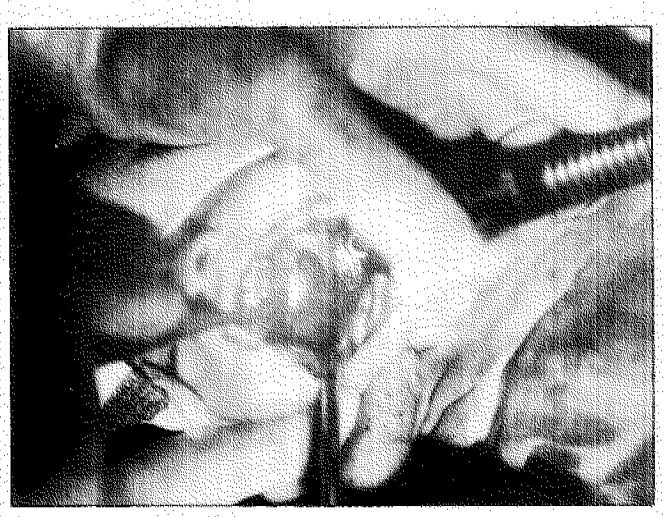


((FIGURE 10) MRI, CORONAL VIEW SHOWING ORO-NASAL MASS. NO CONNECTION WITH INTRACRANIAL COMPARTMENT.

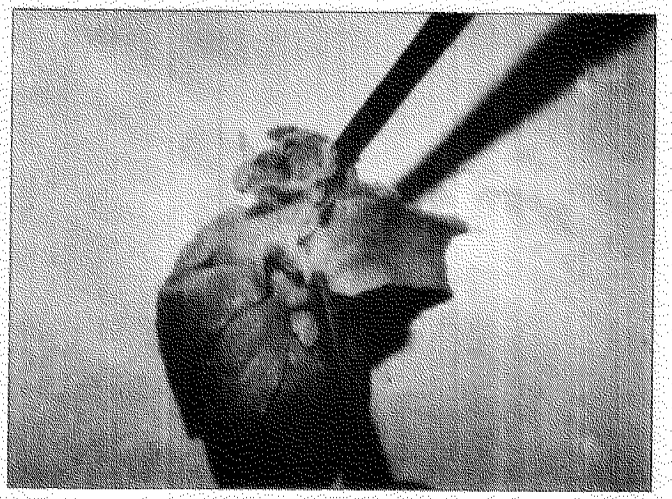
starts to develop as well. Here the cells of the sclerotomes shift their position to surround the spinal cord and the notochord. The formation and subsequent rearrangement of the segmental sclerotomes into the definitive vertebrae is a complex process, and it is not uncommon that 2 successive vertebrae fuse asymmetrically or that half a vertebrae is missing.

Study of the subject was done with the following included in the data base: Physical examination, Radiographic evaluation. Genetic mapping via karyotyping was attempted but was unsuccessful. A noted Pediatric Clinical Geneticist was consulted in this case. Likewise a computerized data record of recognizable congenital syndromes was used. By carefully studying the data at hand and all the available literature, a final diagnosis of Goldenhar Syndrome was made.

The syndrome, first described by Goldenhar in



(FIGURE 11) INTRAOPERATIVE PROCEDURE.



(FIGURE 12) POST-MORTEM EXAM SHOWING PDA (0.4 cm.)

1952[22], is also known as the first and second branchial arch syndrome. The term Oculo-Auriculo-Vertebral dysplasia (OAV) was suggested by Borlin, et al. in 1963 to describe patients with microtia, mandibular hypoplasia, vertebral anomalies, and epibulbar dermoids or lipodermoids[22]. Goldenhar's Syndrome, a non-chromosomal entity, is a rare symptom complex. The predominant defects in the nonrandom association of anomalies represent problems in morphogenesis of the 1st and 2nd branchial arches producing malformations accompanied by vertebral anomalies with or without ocular abnormalities. However, various combinations and gradations of this pattern of anomalies do occur. Thus, they are classified at times under the general description of facio-auriculo-vertebral spectrum[1].

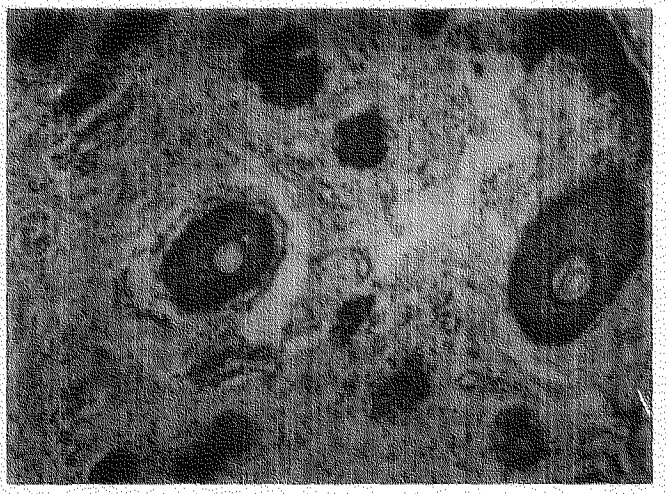
Our case however is unique in the sense that she presented with features not typical of the syndrome. Our patient had a huge oro-nasal mass, agenesis of the corpus callosum, thymic aplasia, and a congenital heart defect. In the study done by Rollnick, et al, in 1987, patients with OAV dysplasia were noted to have an increase in the incidence of cleft palate and congenital heart disease[15]. Scheurle et al, in 1990, in his study of patients with pharyngeal pouch malformations, including this syndrome, indicated that patients with any pharyngeal pouch malformation may have an abnormality of the immune system in the form of thymic aplasia or hypoplasia[18]. Involvement of the corpus callosum was likewise mentioned in the study of Thommen in 1986[20], and by Beltinger in 1988. Bustamante, et al. in 1989 reported three cases of the Goldenhar Syndrome with associated cardiac malformations such as TOF, transposition of the great vessels, and anomalies of the pulmonary venous collection[4,7]. More recently, other malformations have been documented in associations with the Goldenhar Syndrome including CNS, cardiac, pulmonary, and renal anomalies[9]. Nevertheless, auricular dysplasia plus vertebral malformations are pathognomonic and are the cornerstone of this syndrome[15]. Vertebral fusion was the most prevalent malformation encountered. Microtia, vertebral abnormalities, mandibular hypoplasia, and ocular abnormalities are the four main features. Variations do occur. Auricular and mandibular malformations, both derivatives of the 1st and 2nd branchial arches, plus the vertebral defect is the most common combination.

Goldenhar Syndrome occurs sporadically. Its etiology is unknown. Estimated occurence in the first degree relative is about two percent. When unilateral it tends to be right-sided. Most of the patients are of normal intelligence. Mental deficiency is more common in association with microphthalmia. Deafness should be tested at an early age. Cosmetic surgery is usually indicated.

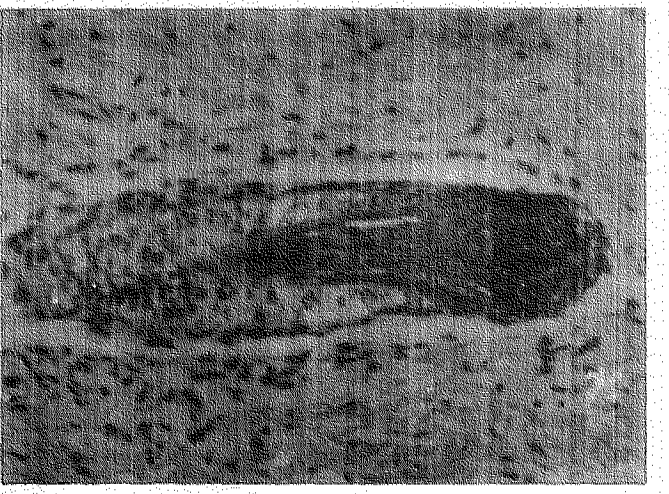
Indeed, Goldenhar Syndrome is a rare symptom complex. In her clinical experience our geneticist has encountered only 5 local cases so far presenting with its classical features. Our patient fulfills the minimum diagnostic criteria. However, in addition to this she presented with several abnormalities which has never been reported together in any patient with the syndrome. Literature reviewed mentioned the occurrence of these variable abnormalities but each defect occurred alone with the typical features. Moreover, our case distinguishes itself in that the patient presented with an oronasal mass which has never been reported in any other case, local or foreign. Nor has there been mention of tumors in any part of the body occurring with or being part of the syndrome.

There is quite a long list of tumors occurring in the neonatal period. Most of these are benign in nature. Surgery is indicated but it is usually the location rather than the histologic nature that determines the timing of surgery. Delay of surgery has always been the dictum as far as benign neonatal tumors are concerned. This dictum would have worked well for the patient and the oronasal mass. Surprisingly, there was no airway obstruction which was relevant considering the location of the tumor. However, because of the persistent pneumonia, this wasn't forthcoming and the mass was excised on the earlier permissible moment. Unfortunately, because of sepsis the patient expired on the 3rd post-op day.

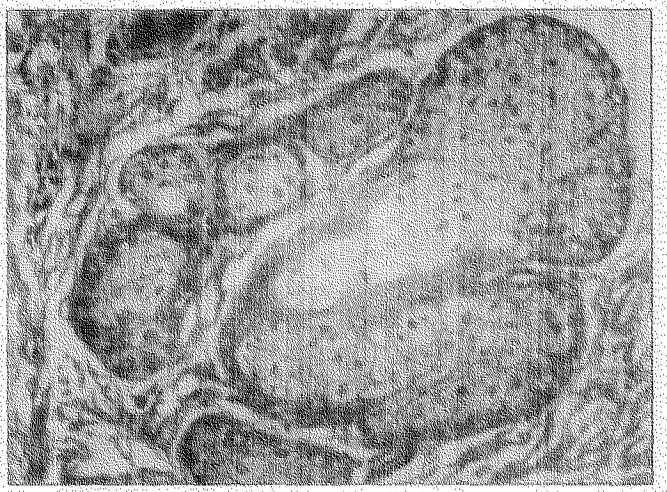
Post-mortem examination revealed thymic aplasia. A review of chest radiographs revealed the initially unnoted absence of the thymic shadow. Involvement of the thymus and cellular immune system in craniofacial malformation syndromes has been reported by Scheuerle, et al. in 1990[18].



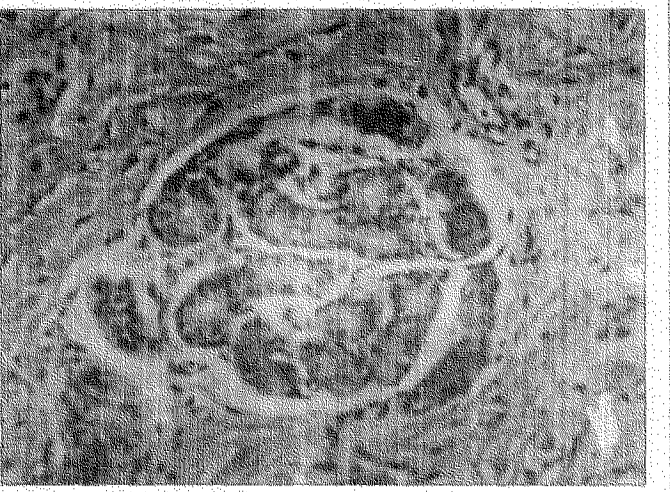
(FIGURE 13) MICROSCOPIC SECTION OF THE MASS LINED BY BENIGN STRATIFIED SQUAMOUS EPITHELIUM WITH NORMAL MATURATION (ECTODERMAL DERIVATIVE). THE SUBJACENT STROMA IS FIBROCARTILAGINOUS AND SCATTERED IN IT ARE THE ADNEXAL APPENDAGES. ALSO SHOWN ARE CROSS-SECTION OF HAIR FOLLICLES.



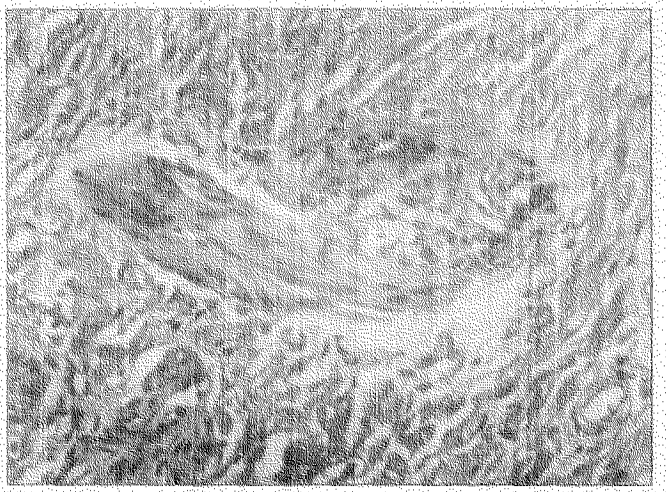
(FIGURE 14) CLOSER VIEW OF A LONGITUDINAL SECTION OF A HAIR FOLLICLE SHOWING A DEVELOPING HAIR TUFT (ECTODERMAL DERIVATIVE).



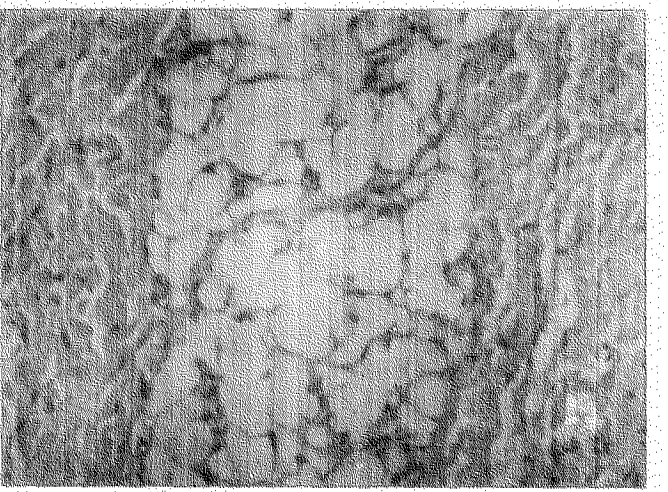
(FIGURE 15) SHOWN HERE ARE GROUPS OF GLANDULAR STRUCTURES (SWEAT GLANDS) (ECTODERMAL DERIVATIVE).



(FIGURE 16) SEBACEOUS GLAND SHOWING ITS TYPICAL MULBERRY-LIKE APPEARANCE (ECTODERMAL DERIVATIVE).



(FIGURE 17) SLIPS OF SMOOTH MUSCLE TISSUE (ELONGATED SMOOTH MUSCLE CELLS) (MESODERMAL DERIVATIVE).



(FIGURE 18) MATURE FAT CELLS WITH ABUNDANT CLEARED-OUT CYTOPLASM DISPLACING THE NUCLEI INTO THE PERIPHERY, LOCATED IN BETWEEN SHEETS OF FIBRO-COLLAGENOUS TISSUE. (MESODERMAL DERIVATIVE).

Microscopic studies of the mass revealed the presence of a benign stratified squamous epithelial lining with fibrocollagenous stroma. Also present were hair follicles, groups of glandular structures such as sweat and sebaceous glands, smooth muscles and vascular structures. Mature fat cells located in between sheets of fibrocollagenous tissues, neuroglial tissue, bone, ganglion cells, and neurons. The histologic picture of both the oral and nasal masses were similar except that the nasal portion imparted a loose quality of the adjacent stroma and was more vascular. Slides were submitted to the different institutions for review. A total of 12 pathologists reviewed the slides and came out with a consensus of BENIGN TERATOMA (Fig. 13-22).

Histopathologic diagnosis was benign teratoma since the tumor contained histologic components not normally present in the site of the tumor. The histologic components are derivatives mainly of the ectodermal and mesodermal germ layers. Failure to identify any histologic derivative of endodermal germ layer did not prevent the pathologist from labelling it as a teratoma, since they thought that this may just be an artifact of sectioning and it is the histopathologic description best suited for the microscopic examination.

True teratomas contain tissue elements derived from all 3 germ layers[16]. The cells found in the lesion may be in any state of differentiation and when cells are quite immature, malignancy is suggested. This however is probably quite rare and most likely reflects only the immaturity of the tissue. They very likely arise from embryonic tissue about the primitive streak and notochord after an escape from external governing influences.

During the embryonic period, approximately from

(FIGURE 19) LINING EPITHELIUM NASAL MASS. (BENIGN STRATIFIED SQUAMOUS EPITHELIUM). ADJACENT STROMA (MESODERMAL DERIVATIVE) IS OF LOOSE QUALITY.

(FIGURE 20) ELONGATED CELLS WIDELY SEPARATED BY LOOSE STROMA WITH OCCASIONAL VASCULAR CHANNELS (MESODERMAL DERIVATIVE) REMINISCENT OF NEUROGLIAL TISSUE (ECTODERMAL DERIVATIVE).

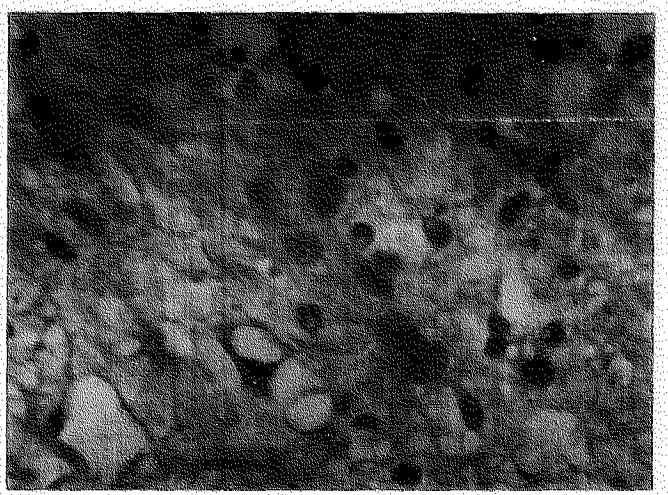
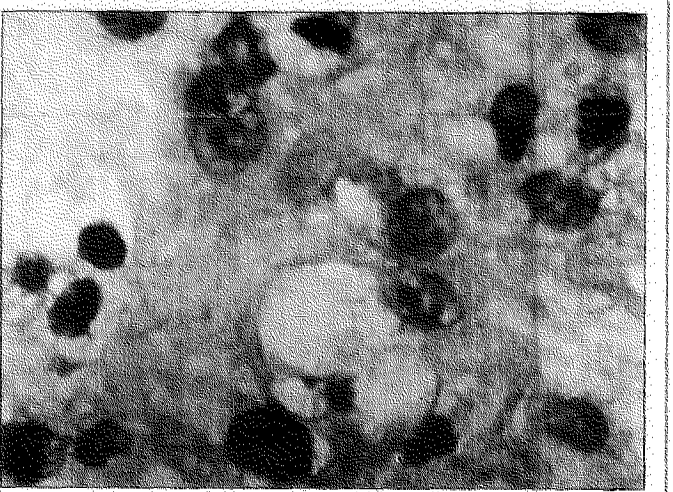


FIGURE 21) CLOSER VIEW SHOWING THE PREVIOUSLY MENTIONED ELONGATED CELLS SHOWING PROMINENT NUCLEUS. STROMA HAS A NEUROFIBRILLARY QUALITY (NEURONS) (ECTODERMAL DERIVATIVE).



(FIGURE 22) CELLS WITH PROMINENT NUCLEOLI AND COARSELY GRANULAR CYTOPLASM (GANGLION CELLS). DEEPLY BASOPHILIC NUCLEUS ARE GLIAL CELLS (ECTODERMAL DERIVATIVE).

the 4th-8th week of development, the 3 germ layers give rise to a number of specific tissues and organs. By the end of this period, the main organ systems have been established.

In true teratomas, failure to demonstrate more than two germ layer components may represent only a section artifact and hence won't justify a separate category. In recent decades however, the definition of teratoma as containing tissue elements of tridermal lineage has become less stringent with the acceptance of examples that are composed of only bidermal ingredients[10]. Teratoid has sometimes been applied to tumors lacking trigeminal histologic appearance[21].

Pathogenesis of this lesions remain obscure. The teratoma may arise from epithelium that has been enclaved in the tissue either on closure of embryogenic process or from traumatic implantation[16].

Teratoma occurs in less than 5% in the head and neck region. The most common site in the head and neck is the cervical region followed by the nasopharynx. Even nasopharyngeal teratomas are considered rare, which usually present with life threatening respiratory embarassment. Our case presented as an oro-nasal teratoma, a location that is rare for such a tumor to occur. A review of foreign literature came up with only one case of a teratoid tumor in the oral cavity[19]. Due to their rarity, reviews often have relied upon compilations of isolated case reports. We report our experience of the early outcome of our case to demonstrate the high morbidity and mortality that these benign but critically placed lesions have because of local mass effect, this of course without taking into consideration the immunodeficient state of our patient.

At this point one can probably draw out some thoughts from the case just presented. First, the need for a thorough investigation to search for all defects is needed in patients with congenital anomalies as it may affect the final outcome of surgery. Second, as mentioned in previous studies, infants with any branchial arch anomaly usually has involvement of the thymus and cellular immune system as well[18]. Third, tumor location and size rather than histologic type were the most significant factors affecting the immediate clinical course.

We have just been presented an infant with an expanded Goldenhar Syndrome. Expanded since it has other more significant congenital anomalies not included in the spectrum of the syndrome. It presents with an oro-nasal teratoma which is a rarity in itself as far as the histodiagnosis and location is concerned. In all the literature concerning the Glodenhar Syndrome, no association with any form of tumor has ever been documented. Given all these manifestations, it may even be right to suspect that the case presented is a totally new and previously unreported syndrome. Our geneticist concurs. Perhaps this case shall not be one to be regarded as an insignificant apostrophe in the broad continuum of reproductive wastage.

SUMMARY

In summary, we are presented with a full term female infant with multiple congenital anomalies, including the oro-nasal mass, bilateral auricular dysplasia, cleft palate, micrognathia, vertebral abnormalities, thymic aplasia, congenital heart defect, and corpus callosum agenesis. Most represent problems in morphogenesis of the first and second branchial arches

After evaluating the data at hand and with help from a clinical geneticist, the diagnosis of Goldenhar Syndrome was arrived at. First described by Goldenhar in 1952, its features include microtia, mandibular hypoplasia, vertebral and ocular abnormalities. This case presented with more than just the minimum criteria for the diagnosis.

The oro-nasal mass for one has never been reported in association with the syndrome. Second, congenital teratoma has always been a rarity as far as the oro-nasal region is concerned. The most common location being the saccrococcygeal region with the head and neck being the most unlikely. If they do occur in this area the most common site is the cervical and nasopharyngeal area.

This dysmorphic accident could have happened anytime from the 4th week of embryogenesis onwards primarily in the period when the branchial arches starts to appear and the germ layers starts to develop.

In this particular case, the authors were faced with the dilemma regarding the timing of surgery of the tumor. The infant developed an uncontrollable pneumonia secondary to the oro-nasal mass. The pnemonia secondary to the oro-nasal mass. The pneumonia in itself plus the patient's heart condition and subsequent pulmonary hypertension prevented the surgeon from doing the procedure. When the decision to operate was arrived at, the anesthesia and surgery was performed with relative ease. However, the patient did succumb to sepsis and eventually expired. This could be explained by the immunocompromised state of the patient secondary to the thymic aplasia.

To the unsuspecting clinician or surgeon, it is noteworthy to draw out some thoughts from this rare and interesting case presented. The search for anomalies and eventually the syndrome even in the absence of literature is never futile. The suspicion of other anomalies which may influence the surgery should always be kept in mind.

ACKNOWLEDGEMENT

The authors would like to thank all those who have given their most valuable time in this case especially Dr. Carmencita Padilla, a Pediatric Clinical Geneticist. Special thanks goes to all the patholgoists from the different institutions:

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COMPLETE EXTERNAL OPHTHALMOPLEGIA AN UNUSUAL PRESENTATION*

Roel Pidlaoan, MD**
Bernardo Dimacali, MD***

ABSTRACT

A 30 year old male with complete external ophthalmoplegia presented with a left parasellar mass and left maxillary sinusitis on C-T scan. Exploratory Caldwell-Luc and Transantral ethmoidectomy revealed a mass obstructing the maxillary ostium; the histopathologic diagnosis of transitional cell papilloma of the maxillary sinus was given. The patient gradually improved after the surgical intervention and was pronounced fully recovered, one month postoperatively. A possible pathophysiology in the development of complete external ophthalmoplegia associated with maxillary sinusitis was presented.

The author recommended that cases presenting with complete external ophthalmoplegia should be carefully evaluated for the possibility of an otolaryngologic etiology.

INTRODUCTION

Ophthalmoplegia is a general term for paralysis of the ocular muscles. If we want to refer to a particular group of muscles affected, we can describe it as follow: (1) internal ophthalmoplegia when it involves the iris and ciliary body muscles, (2) external ophthalmoplegia when if affects only the extraocular group of muscles, namely, the four recti, two obliques and levator muscle, or (3) Total ophthalmoplegia when it involves the paralysis of all the ocular muscles (**Duane**, 1986).

Complete external ophthalmoplegia will therefore be characterized by the eyeball frozen in the primary position and the pupils remaining equal and reactive. Diplopia manifests in all secondary gazes because when the unaffected eye focuses in one direction, while the affected eye is immobile, two images are sent to the occipital cortex. Ptosis is also seen due to the paralysis of the levator palpebrae superioris.

The above signs and symptoms are caused by varied entities that usually arise in and around the vicinity of the ocular apparatus.

Papilloma, one of the more common benign tumors of the oral and upper respiratory tract, has been broadly and loosely defined as an epithelial reactivity to tissue injury (**Batsakis, 1986**). The possibility of an ordinary papilloma to cause a symptom complex like complete external ophthalmoplegia may, at first appear far-fetch.

A review of the available literatures mentioned the possibility of maxillary sinusitis to cause ophthalmoplegia, however, no pathophysiology has ever been presented. The first reported local case (Villanueva-Sarenas, 1990) proposed three possible pathways for the spread of the infection from the maxillary sinus to the region of the orbit. At present, this is only the second reported case of ophthalmoplegia associated with maxillary sinusitis. This paper, therefore, is being presented to gain a deeper understanding regarding another orbital complication of maxillary sinusitis.

CASE REPORT

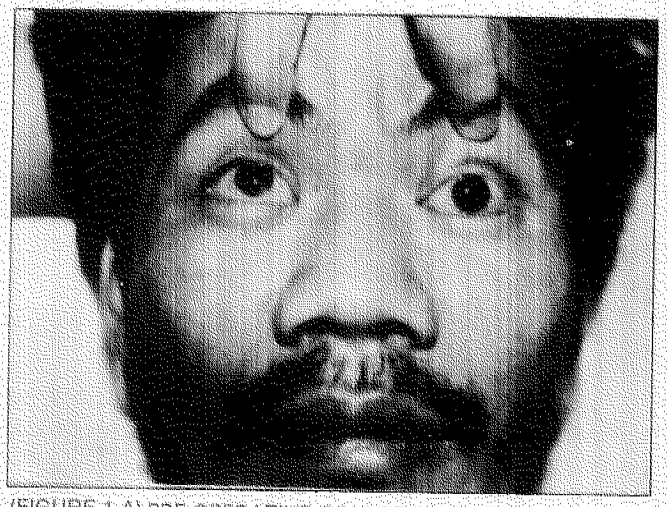
This is a case of a 30 y/o male who presented with the chief complaint of double vision of one week duration. The present illness apparently started two weeks prior to admission as rapid development of redness and swelling of the left eye, followed by intermittent, deep-seated, boring left eye pain and vague left-sided facial pain. He then consulted a private physician who prescribed unrecalled oral antibiotics.

One week prior to admission, diplopia was noted. Later on there was resolution of the swelling of the left eye but the left-sided facial pain persisted. A physician suggested a C-T scan of the head which revealed a parasellar mass, left and maxillary sinusitis, left. He was subsequently referred to the hospital for admission.

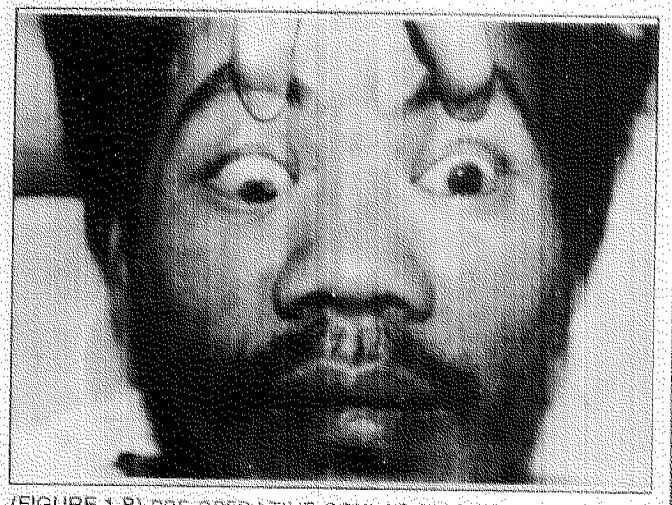
His past medical history were non-contributory. His personal and social history revealed a smoking history of 8.5 pack years and chronic alcohol intake.

On admission, the pertinent physical findings were

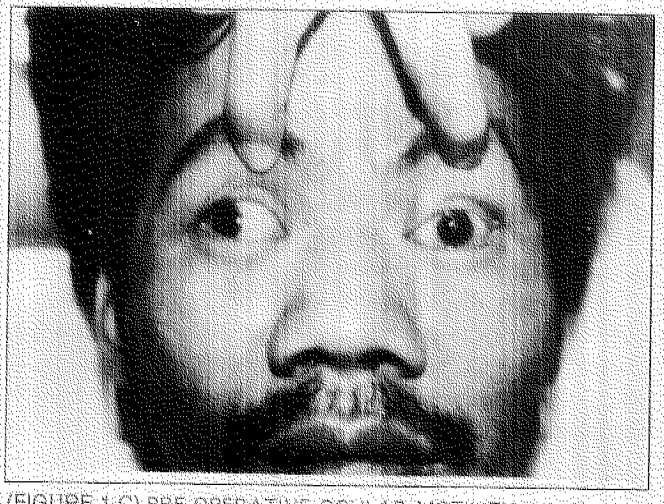
* Presented at the PSO-HNS Interesting Case Contest held at Hotel Nikko, Manila Garden Hotel, July 9, 1992.
** Resident, Dept. of ENT, FEU.
*** Consultant, Dept. of ENT, FEU



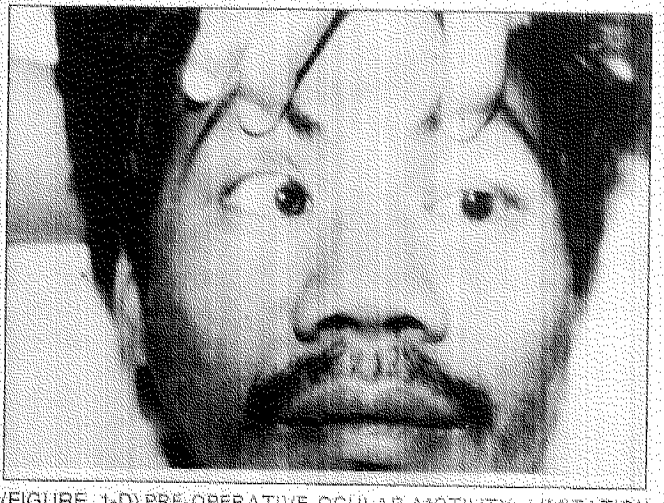
(FIGURE 1-A) PRE-OPERATIVE OCULAR MOTILITY: LIMITATION OF THE UPWARD GAZE.



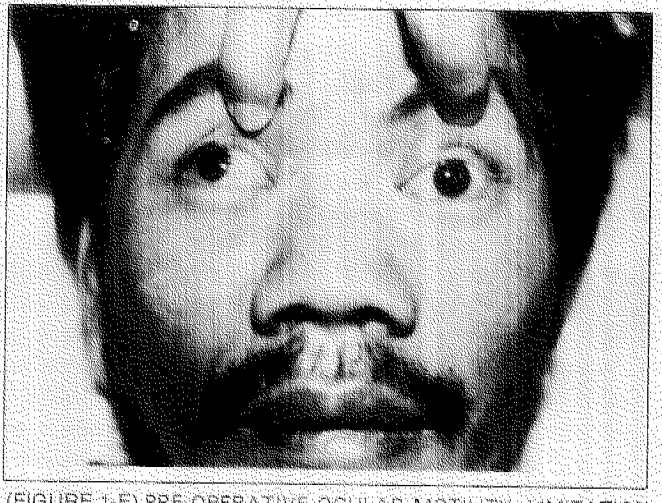
(FIGURE 1-B) PRE-OPERATIVE OCULAR MOTILITY: LIMITATION OF DOWNWARD GAZE.



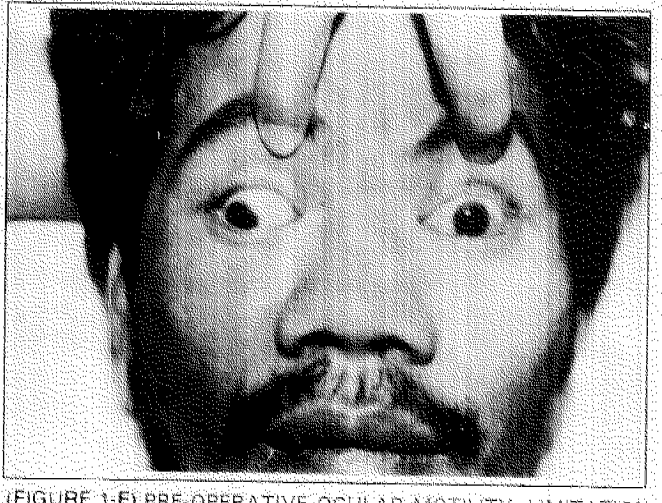
(FIGURE 1-C) PRE-OPERATIVE OCULAR MOTILITY: LIMITATION OF LATERAL GAZE.



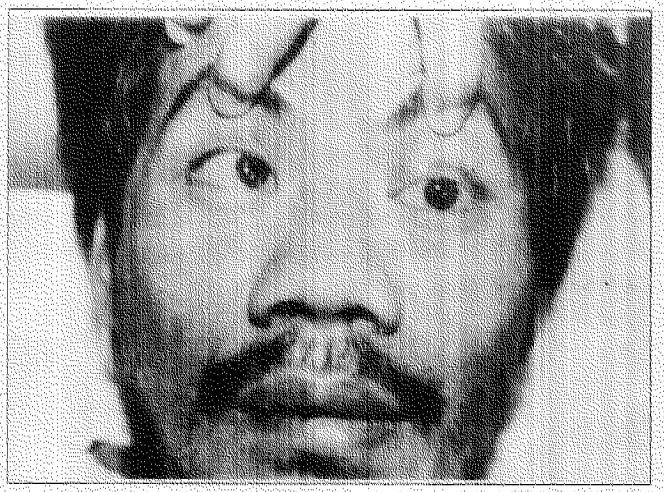
(FIGURE 1-D) PRE-OPERATIVE OCULAR MOTILITY: LIMITATION OF MEDIAL GAZE.



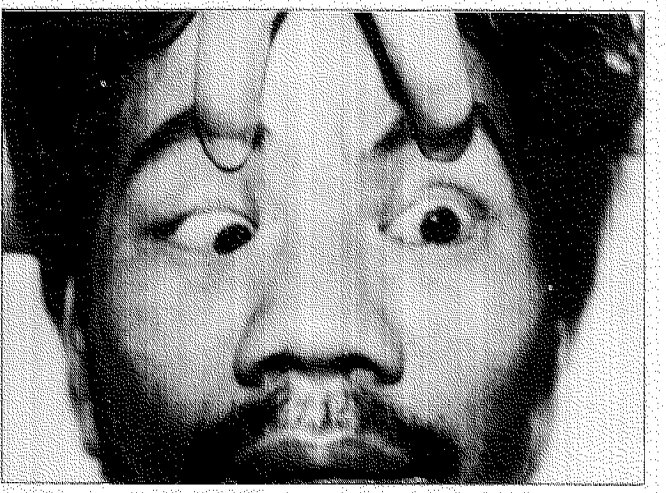
(FIGURE 1-E) PRE-OPERATIVE OCULAR MOTILITY: LIMITATION OF SUPERO-LATERAL GAZE.



(FIGURE 1-F) PRE-OPERATIVE OCULAR MOTILITY: LIMITATION OF INFERO-LATERAL GAZE.



(FIGURE 1G) PRE-OPERATIVE OCULAR MOTILITY: LIMITATION OF SUPERO-MEDIAL GAZE.



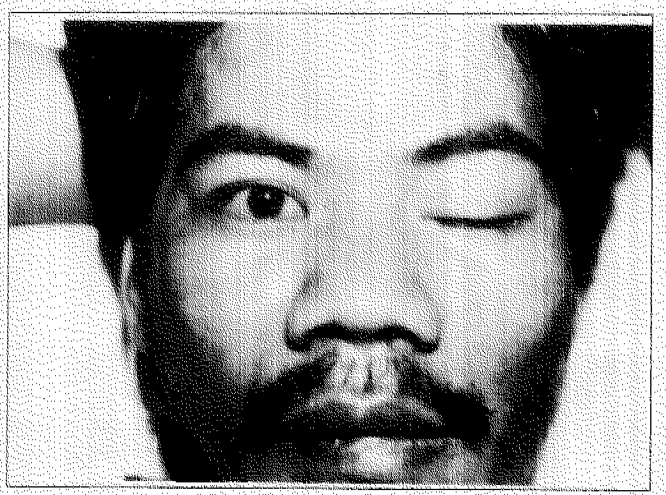
(FIGURE 1H) PRE-OPERATIVE OCULAR MOTILITY: LIMITATION OF INFERO-MEDIAL GAZE.

the following: limitation of the superior, inferior, lateral and medial gaze (Fig. 1 a to h); ptosis (Fig. 2); and a negative forced duction test. The visual acuity was 20/20. Fundoscopic examination was within normal limits. On anterior rhinoscopy, no nasal congestion and nasal discharge were noted. Posterior rhinoscopy revealed the nasopharyngneal mucosa to be slightly congested with whitish post nasal discharge, no masses noted. The rest of the ENT findings were unremarkable.

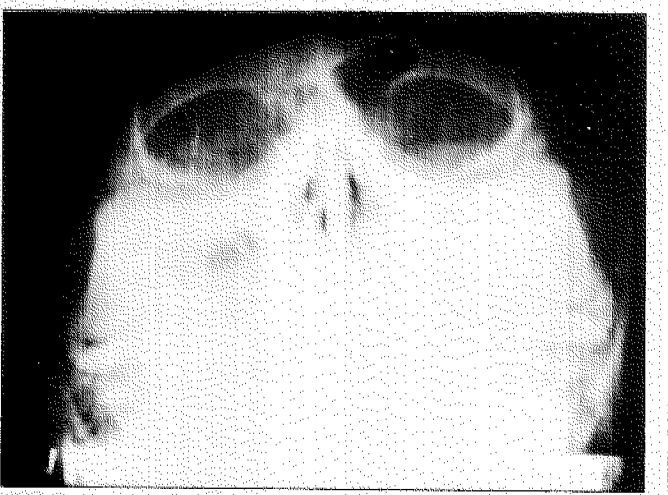
The pertinent neurological findings were as follows: paralysis of the left cranial nerves III (occulomotor), IV (trochlear), and VI (abducens); pupils equally reactive to light; and involvement of the first and second branches of the left cranial nerve V (trigeminal). The rest of the neurological findings were unremarkable.

The patient stayed in the ward for 5 weeks. He was initially admittted under the service of the Department of Surgery with the diagnosis of parasellar mass, left and maxillary sinusitis, left. He was given the following medications: ampicillin/cloxacillin, clemastine, vitaminn B-complex, mefenamic acid and nalbuphine. CBC, ECG and Chest-X-ray were done and showed the results to be within normal limits.

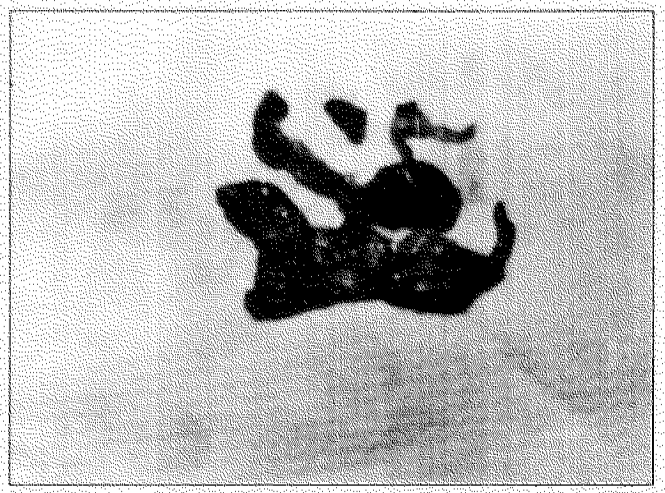
On the 5th H-day, he was referred to the Department of ORL-HNS for further evaluation and management of the left maxillary sinusitis. The assessment of the service were the ff.: 3rd, 4th, 5th and 6th cranial nerve palsy probably secondary to the left parasellar mass, which may be a metastatic tumor of nasopharyngeal origin or an infection arising from the left maxillary sinus. X-ray of the paranasal sinuses and random nasopharyngeal biopsy were subsequently suggested. The X-ray (Fig. 3) showed marked opacification of the left maxillary sinus and slight hazi-



(FIGURE 2) PTOSIS OF THE UPPER EYELID (L).



(FIGURE 3) PARANASAL X-RAY (WATER'S VIEW).



(FIGURE 4) CROSS SPECIMEN OF MASS TAKEN FROM THE POSTERO-SUPERO-MEDIAL WALL OF THE MAXILLARY SINUS (L).

ness of the right maxillary sinuses, hypoplastic frontal sinuses and normal sphenoidal sinuses. The histopathologic report of the random nasopharyngeal biopsy revealed chronic nonspecific inflammatory process. There was no sign of malignancy. The service then recommended for an exploratory Caldwell-Luc operation.

On the 11th, H-day he was referred to the Department of Medicine (Neurology Servicce) which gave the ff. assessment; parasellar mass, left involving the 3rd, 4th, 5th and 6th cranial nerve. Suggested medication were carbamazepine, low-dose prednisone and cimetidine. Despite the above medications, the patient persistently complained of severe intermittent left-sided facial pain and fronto-temporal headache.

On the 12th H-day, blood chemistry determination were requested which included FBS, BUN, Total Cholesterol, Uric Acid and Potassium, the result of which were within normal limits.

On the 20th H-day, the patient, underwent exploratory Caldwell-Luc and Transantral ethmoidectomy. The intraoperative findings were as follows; thickened maxillary sinus mucosa with yellowish mucopurulent antral discharge and a grayish mass measuring 4x3x2 cm. at the postero-supero-medial wall of the maxillary antrum. No bone destruction was noted. The histopathologic report of the mass showed a hyperplastic transitional epithelium with loose edematous stroma and moderate inflammation. This was signed-out as transitional cell papilloma of the left maxillary sinus (Fig. 4). Gram stain of the antral discharge showed few gm (+) cocci in chains and in clusters with plenty of pus cells. The culture report was positive for *Streptococcus* (non-enteroccoci) and *Staphylococcus epidermis*. Following surgery, massive parenteral antibiotics were started (ampicillin/cloxacillin) and the steroid was discontinued.

On the 6th postoperative day, slight mobility of the left eye was noted. From then on his condition gradually improved. On follow-up, 1 month after surgery, he had fully regained the mobility of his left eye. (Fig. 5 a to d). The ptosis, diplopia and pain also completely resolved. (It was unfortunate that post-operative C-T scan was not done to document whether the parasellar mass disappeared with the intervention due to patient's financial constraints.)

DISCUSSION

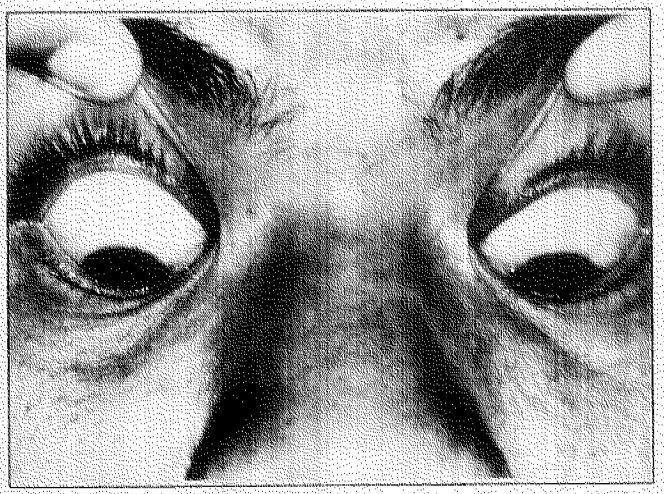
Painful ophthalmoplegia are most commonly caused by either inflammatory or neoplastic lesions. A previous study showed that while tissue enhancement following intravenous contrast administration provided a substantial additional information, in some instances C-T scan did not reliably differentiate inflammatory from neoplastic lesion (**Unger**, et al, 1986). Since modern diagnostic tools were of little help, the author resorted to diagnosis by exclusion.

There are three general mechanisms by which extraocular muscle paralysis may occur (**Villanueva-Sarenas**, 1990). The first is mechanical, wherein some factors may interfere with ocular muscle movement. This mechanism was readily excluded by the negative forced duction test of the patient. The second mechanism is myogenic, which includes disorders that primarily affect the muscle, as exemplified by myasthenia gravis. This disorder should present with a clear-cut history of episodic attacks associated with fatigue, which was definitely absent with the patient.

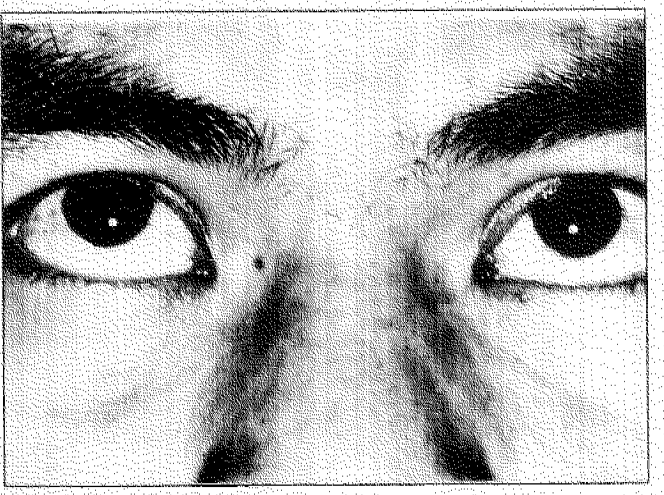
The third mechanism, and the one which is of particular concern is the neurogenic mechanism. Under acquired neurogenic causes are five groups of disorders. Of these, trauma was excluded since he had no history of trauma. A metabolic disorder was also thought of to be highly unlikely as his blood chemistry were all normal.

The vascular disorders pertaining to CVA's or TIA's, and the infectious disorders, referring to meningitis and encephalitis, were of remote considerations.

The space occupying lesions which include aneurysm, localized inflammatory lesions and newgrowth, were the primary considerations in this case because of the presence of the parasellar mass on C-T scan. A newgrowth was an unlikely candidate since newgrowth usually produce gradually progressive disorders, with the cranial nerve becoming involved one after the other. Furthermore, nasopharyngeal carcinoma, which have been associated with over 20% of parasellar syndromes (**Duane**, 1986), was eliminated by the negative nasopharyngeal biopsy. Aneurysms more commonly



(FIGURE 5-A) POSTOPERATIVE OCULAR MOTILITY: DOWNWARD GAZE.



(FIGURE 5-B) POSTOPERATIVE OCULAR MOTILITY: UPWARD GAZE.

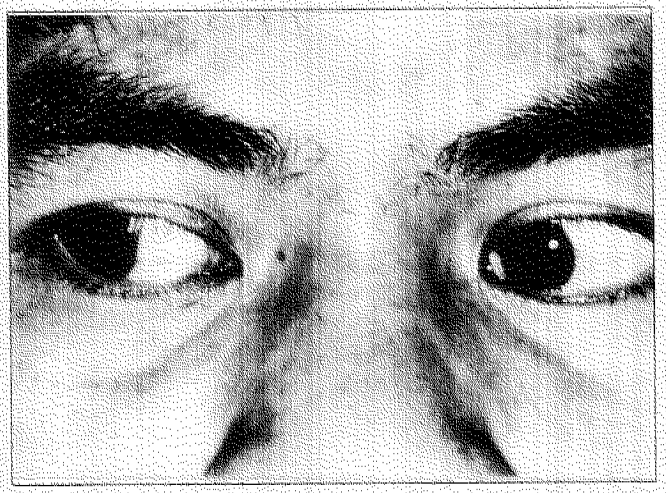
produce visual defects and are thus excluded since the vision was unaffected in this patient. Hence, the mass was most probably a localized inflammatory process.

The next step was to pinpoint the probable location of the mass. Cranial nerve III (occulomotor), IV (trochlear) annd VI (abducens) and the 1st and 2nd branches of the V (trigeminal) nerve lie in close proximity only within the cavernous sinus and in and around the superior orbital fissure.

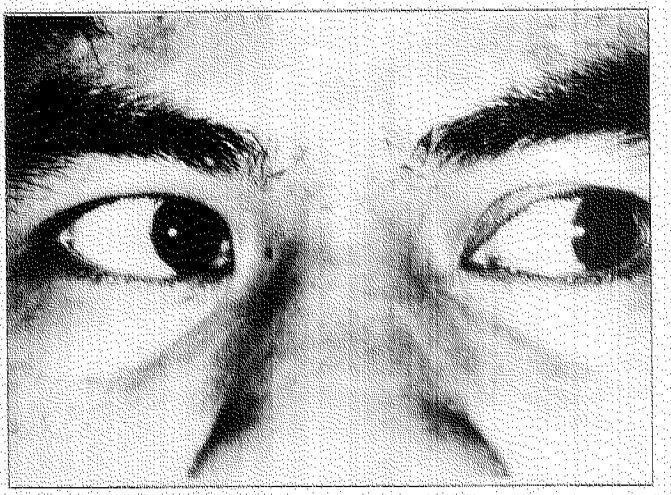
The following conditions were taken into consideration because of their presenting characteristics: Cavernous sinus thrombosis could produce ophthalmoplegia but proptosis, chemosis of the conjunctiva, blurred vision and systemic signs and symptoms would also be present. Similarly, an orbital cellulitis or abscess, could also produce a painful ophthalmoplegia but it would likewise manifest chemosis, proptosis and blurred vision. Furthermore, an intraorbital lesion would not be compatible with the C-T scan. Therefore, a retro-orbital involvement of the lesion adjacent to the area of the superior orbital fissure would match the patient perfectly. (Fig. 6).

Since the superior orbital fissure contains the 3rd, 4th, 6th and the 1st and 2nd branches of the 5th C.N.'s, the patient most probably developed a localized infection at this site hence, the involvement of the extraocular muscles. This location is also compatible with the parasellar mass seen on Ca-T scan.

Although others may argue that chronic inflammation must be present for the papilloma to have developed, this was not readily apparent in the patient. Nevertheless, we have arbitrarily identified the papilloma as the aggravating factor in the development of the opthalmoplegia, since in its abscence the course of



(FIGURE 5-C) POSTOPERATIVE OCULAR MOTILITY: LATERAL GAZE.



(FIGURE 5-D) POSTOPERATIVE OCULAR MOTILITY: MEDIAL GAZE.

this sinusitis could have undoubtedly been different.

In any case, the postero-supero-medial location of the papilloma probably prevented the drainage of the maxillary ostium. As noted in a previous study, once the ostium has been obstructed, intrasinus air is absorbed by the mucosa and negative intrasinus pressure develops, leading to increase transudation, which can provide nutrients for bacterial growth. The absorption of air during the initial phase moderately reduces the oxygen tension within the sinus fostering the growth of aerobic facultative organisms (**Brook**, 1981; and **Carenfelt** and **Lundberg**, 1977). The history of chronic alcohol intake of the patient could also have weakened his constitution, predisposing him to develop a severe form of sinusitis. Although there was an absence of nasal discharge in anterior rhinoscopy, this was most likely a reflection of the degree of ostial obstruction caused by the papilloma.

As his antrum became full of purulent material, positive pressure was created forcing the pus into the venous system. The sinus infection could then spread to the adjacent structure through an intricate system of valveless communication between the nose, paranasal sinuses, orbit and cavernous sinus (**Batson**, 1936). Maxilary sinusitis can spread to an adjacent orbit thru its venous drainage, the inferior ophthalmic vein. The inferior ophthalmic vein is formed by a confluence of veins in the floor and medial wall of the orbit. This inferior ophthalmic vein drains into the superior ophthalmic vein then to the cavernous sinus (**Duane**, 1986).

The point when the patient noted redness and swelling of his left eye, followed by pain, within and around the left orbit radiating to the fronto-temporal area, probably heralded the spread of the infection to the region of the superior orbital fissure.

Another consideration in the disemmination of the infection was via direct extension through the incomplete central portion of the orbital floor which is traversed by the inferior orbital fissure. This fissure houses the inferior orbital nerve, the inferior ophthalmic vein with the tributaries to the superior ophthalmic vein. Another preformed path-way is the connection between the inferior and superior orbital fissure which contains the nerve supply to the extraocular muscle (**Duane**, 1986). The author, therefore, agreed with an earlier local study (**Villanueva - Sarenas**, 1990) which proposed the above pathways for infections in the maxillary sinuses to cause ophthalmoplegia.

What could have happened in this patient was that, he probably developed phlebitis of the superior ophthalmic vein which could have produced inflammatory edema to the adjacent III, IV and VI cranial nerve causing the ophthalmoplegia of the patient. The above pathogenesis could also explain the presence of facial pain with the involvement of the 1st and 2nd branches of trigeminal nerve.

Partial treatment of the infection with the oral antibiotics probably caused the resolution of the redness and swelling of the left eye. However, the dose of the antibiotics was not enough to eradicate the infection; instead, localization occured at the retro-orbital area adjacent to the superior orbital fissure.

This abscess could explain the small left parasellar mass seen at the C-T scan. An earlier study described it as such: phlebitis in the sinus mucosa may extend to the vein in the periorbita, resulting in subsequent rupture of the vessels. Vessel rupture may result from congestion or actual erosion of the vessels by the infectious process. The resultant hematoma may undergo suppurative changes, producing an abscess (**Harris, Kay & Nilles**, 1978).

Involvement of the III, IV and VI cranial nerves of the affected side resulted in external ophthalmoplegia, and since the other eye was un affected, two images are thus formed causing diplopia in all secondary gazes.

Timely surgical intervention, massive antibiotics and possibility steroid therapy all contributed to the gradual improvement of the patient.

SUMMARY AND CONCLUSION

The report presented a case of chronic maxillary sinusitis aggravated by maxillary ostium blockade by a papilloma, presenting as complete external ophthalmoplegia. The condition resolved completely leaving no residual neurologic deficit following a Caldwell-Luc operation and transantral ethmoidectomy.

The proposed pathophysiology in the development of ophthalmoplegia in this patient started with chronic left maxillary sinusitis which, in turn, precipitated the development of papilloma. The papilloma aggravated the sinusitis by causing ostial obstruction. Subsequently, the increasing pressure inside the maxillary antrum caused the infectious process to spread into the venous drainage, the inferior ophthalmic vein which in turn drains into the superior ophthalmic vein. The superior ophthalmic vein probably developed phlebitis which produced inflammatory edema to the adjacent cranial nerves III (occulomotor), IV (trochlear) and VI (abducens), causing the ophthalmoplegia and facial pain, respectively The left parasellar mass in this patient was probably an abscess which resulted from the rupture of an infected vessel (phlebitis). The resultant hematoma underwent suppurative changes producing the abscess.

In conclusion, complete external ophthalmoplegia can be an orbital complication of maxillary sinusitis, hence, careful evaluation of cases presenting with this manifestation should be done in order to institute the proper management.

Table 1. Localization of signs and symptoms referable to the cavernous sinus and superior orbital fissure.

Signs and Symptoms	Cavernous Sinus	Superior Orbita Fissure intra orbital	Superior Orbita Fissure retro orbital	The Patient
Complete externa ophthalmoplegia and ptosis	+	+	+	+
Proptosis	+	+++	-	-
Chemosis	+++	+	-	-
V Vision	+	+	-	-
Systemic S/S	+	+ / -	+ / -	+
Compatible with C-T scan	+	-	+	+

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ORODIGITOFACIAL SYNDROME: A Report of a Case
"The Girl with a Forked Tongue"*

Ramon V. Gonzalez, MD**
Natividad S. Guevara, Jr., MD**
Eutrapio S. Guevara, Jr., MD***

ABSTRACT

A 7 year old girl presented with physical findings consistent with the diganosis of orodigitofacial syndrome. She demonstrated lobulated tongue, hypertrophic frenula, cleft palate, alopecia, notched upper lip, missing mandibular lateral incisors and frontal bossing. Family history was positive for cleft lip and palate and tongue deformity. Karyotyping demonstrated 46 XX genetic constitution. The malformation closely matched Doege's description. Excision biopsy and repair of the deformed tongue was done. Histopathologic report revealed multiple squamous papillomas, which is compatible with choristoma of the tongue.

INTRODUCTION

In the daily practice of otolaryngology, patients with diseases of the tongue are often seen; from infectious to newgrowths, from ulcerating to fungating types. Once in a while, something different is encountered and for a moment the specialist is caught off-guard and scramble for medical books and references.

This case is being presented in the hope that one may limit this unguarded moment.

CASE REPORT

The patient is a 7 year old female who consulted at the Department of ENT-HNS for a deformity of the tongue. Initial assessment of the patient revealed an active and apparently healthy child.

On physical examination, the patient had: flattened face with a broad nasal bridge, slight hypertelorism, frontal bossing, bifid nasal tip and notching of the upper lip. Examination of the oral cavity revealed widely spaced serrated teeth, absent mandibular lateral incisors and presence of submucous cleft palate. The tongue was lobulated at the tip with nodular, yellowish, smooth, firm masses measuring about 5 x 5 mm masses in between clefts. Another lobulae was noted at the right lateral edge of the tongue. These lobules form a trifid tongue that would fan out whenever the tongue was protruded. There was presence also of multiple hyperplastic frenula, however, there was normal tongue mobility and no speech difficulty. The rest of the physical examination and laboratory examinations were normal. Karyotyping revealed a 46, XX genetic constitution.

DISCUSSION

The tongue which has been described as the window of the digestive system, is an excellent indicator of the constitutional state of the patient, but is seldom diagnostic of any specific diseases (Keyes). Our initial concern was whether this anomaly was confined to the tongue, or whether it was an "indicator" of the patient's constitutional state.

One of the most common anomaly is that of the fissured or scrotal tongue. In this condition, the tongue has grooves which are variably deep, although, usually symmetrical. The condition is frequently familial with no pathologic significance and spontaneously resolves.

Although the patient's mother also had some sort of tongue deformity, these were only nodules on the lateral aspects of the tongue. Initially, it was thought that this might be a variant of a fissured tongue. However, the absence of the grooves and failure of the lesions to resolve spontaneously, was rather disturbing.

Embryologically, the anterior 2/3 of the tongue develops from 2 symmetric swellings at the ventral ends of the first pharyngeal arches. According to Paparella and Shumrick, a bifid tongue results when there is incomplete fusion of these mentioned lateral halves.

Although bifid tongue is a rare anomaly, it is said to have been mentioned in literature as early as 1921, together with cleft lip and palate. The first detailed

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* Presented at the PSO-HNS Interesting Case Contest held at Hotel Nikko, Manila July 9, 1992.
** Residents, Dept. of ENT, SLMC
*** Consultants, Dept. of ENT, SLMC.

description, however, was done in 1954 by **Papillon-League** and **Psaume** as part of the Oro-digito-facial-syndrome. It is claimed that this anmaly is a hereditary malformation of the buccal mucosa. Clinical features include short and narrow upper lip with median notching and shortened alar cartilages. The tongue is often bifid or lobulated. Other features include abnormality of the digits, namely polydactyly, syndactyly or clinodactyly. It is inherited as an X-linked dominant gene and almost always fatal in males. This syndrome is also called the **Papillon-Leage** and **Psaume** Syndrome in honor of the authors who first described the entity.

The question arises as to whether the patient only had an incomplete manifestation of the syndrome, as has been reported by others. **Gorlin** and **Psaume** (1962) reported only 3 patients with radiologic abnormalities out of a total of 22 patients, while **Ruess** et al. in 1962 reported it in 5 out of 12 patients. This dilemma could best be settled by chromosomal analysis.

Ruess et al. believed that the syndrome is caused by a partial trisomy, the extrachromosomal material being undetectable in others. **Kuschnick** et al, reported a case with findings consistent with the diagnosis of oro-digital-facial syndrome. The karyotype showed trisomy probably involving chromosome no. 1. **Wahrman** described a male infant showing all major features of the syndrome who had an XXY chromosomal constitution. It was thought that this XXY male was viable because of the normal alleles in one X chromosome.

Therapeutically, excision biopsy and repair of the patient's deformed tongue was done. Histopath revealed: multiple squamous papillomas which were almost completely covered by squamous epithelium. Some central portions contain mucous glands while others have dense connective tissue cores. This report was histologically compatible with a papilloma or generally, a choristoma of the tongue.

The findings closely matched **Doege's** description of the tongue in the oro-digital-facial syndrome. It is bifid or lobulated with occasional pedunculated masses or tumors attached to its dorsal surface and fixed to the alveolar ridges of the mandible by fibrous bands and to the floor of the mouth by thickened frenula. Further, the tumor of the tongue are benign choristomas consisting of aberrant mucous glands, smooth muscle, connective tissues and covered by hyperplastic keratinizing epithelium- just like our own patient.

CONCLUSION

Based on literature reports, this is probably the first ever reported case of orodigitofacial syndrome in the Philippines. This syndrome, rare as it is, is also baffling. The diagnosis involves the scrutiny of a **Sherlock Holmes** and the management, the patience of Job. It is an investigation involving four to five generations and requires a multi-disciplinary approach namely: Otolaryngology, Plastic Surgery,Pediatric Psychology, Genetics and Genetic Counselling, Orthodontics and Radiology. Hopefully, the efforts in bringing forth the presence of such malformation in the country would stimulate awareness and greater understanding of this syndrome. Only in such awareness that efforts will be given in providing such a child with a healthier and more acceptable social life.

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CASE REPORT: INTRACRANIAL MENINGIOMA WITH EXTRACRANIAL COMPONENT*

MARIA RINA T. REYES, MD**
RHODORA A. DEL ROSARIO, MD**
FREDERICK O. LEH, MD**
ALFREDO Q.Y. PONTEJOS, JR., MD***

ABSTRACT

A meningioma with intra-and extracranial components in a 24 year old female is presented.

She had a three year history of a lateral neck mass, aural polyp and parapharyngeal bulge, associated with tinnitus and hearing loss. The meningioma was excised via a suboccipital craniectomy by the neurosurgeon and base of the skull surgery by the otolaryngologist.

INTRODUCTION:

Meningiomas are benign tumors arising from the arachnoid cells. They are relatively common tumors accounting for about 14-18% of all intracranial tumors and 25% of primary intraspinal tumors (1). In a review of 100 cases from 1980-1989, in our institution, meningiomas were found to be more common in females (2:1), with an age distribution following a bell-shaped curve peaking at the fourth decade(2). However, extracranial meningiomas whether primarily extracranial or an extension from an intracranial lesion is quite rare, comprising about 2% of all meningiomas(3). The most frequent sites in which extracranial meningiomas are found are the bones of the skull, the scalp, orbit, nose, paranasal sinuses and middle ear. There have even been reports of meningiomas in the cheek, deep tissues of the neck and parotid gland (1). The following case report describes a meningioma with an intracranial component and an extracranial part presenting as a lateral neck mass, aural polyp with a parapharyngeal bulge.

CASE HISTORY:

A 24 year old female was admitted last May 23, 1991 for a left infra-auricular mass. Review of history shows that three years prior to admission, she noted a mass measuring 1 x 1 cm from the left infra-auricular area described as firm, nontender and painless, associated with tinnitus and hearing loss on the same side. There was no headache, vertigo nor ear discharge. The patient consulted a private physician and the mass was treated as reactive lymphadenopathy due to dental caries. Tooth extraction was done and antibiotics given, however the mass did not decrease in size. Two years prior to admission, there was an increase in the size of the mass. The patient consulted another private physician, and, in addition to the left infra-auricular mass, the physician also noted a polyp in the left ear. Polypec tomy was done, however specimen was not sent for histopathologic studies. One year prior to admission, due to the persistence of the mass, the patient consulted another physician. Aside from the two previous findings, there was note of a left parapharyngeal bulge. Biopsy of the aural polyp was done, and the report was "suggestive of a benign tumor". Three months prior to admission, the patient started experiencing left temporoparietal headache described as mild to moderate in severity, throbbing, lasting for 3 to 5 minutes, occuring two to three times a week, and occasional vertigo, lasting for a few seconds, not associated with changes in head position, nausea and vomiting. She subsequently consulted at our institution, where she presented with a 2 x 2 cm doughy, nontender fixed mass in the left infra-auricular area, a pinkish mass occluding the left auditory canal, and a bulge in the left parapharyngeal area. The rest of the ENT examination was essentially normal. The cranial nerves were tested and found intact. The following diagnostic procedures were done: mastoid series showed chronic mastoiditis, left; pure tone audiometry/speech test revealed a mild conductive hearing loss, AD, and severe to profound mixed hearing loss, AS (Figure 1). Fine needle aspiration biopsy of the left infra-auricular mass was read as "groups of benign acini with occasional lymphocytes". No evidence of malignancy was found from the punch biopsy of the parapharyngeal bulge. Histologic examination of the polyp on the left ear showed meningioma. A CT scan of the head was then done and this revealed a hyperdense enhancing mass in the left parapharyngeal and parotid region with intracranial extension to the left

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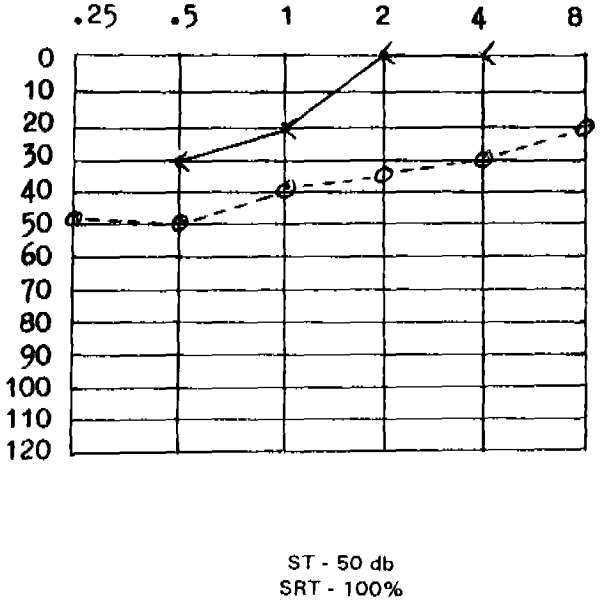
** Resident, Dept. of ENT, UP-PGH
*** Consultant, Dept. of ENT, UP-PGH.

FIGURE 1: PURE TONE AUDIOMETRY OF THE PATIENT



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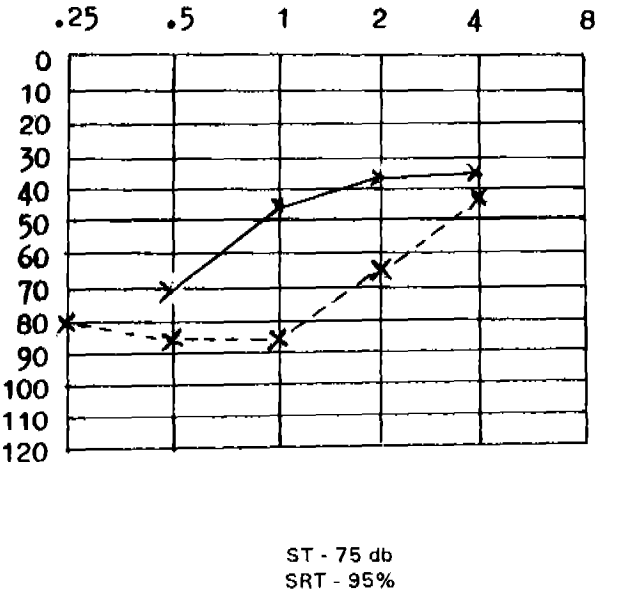

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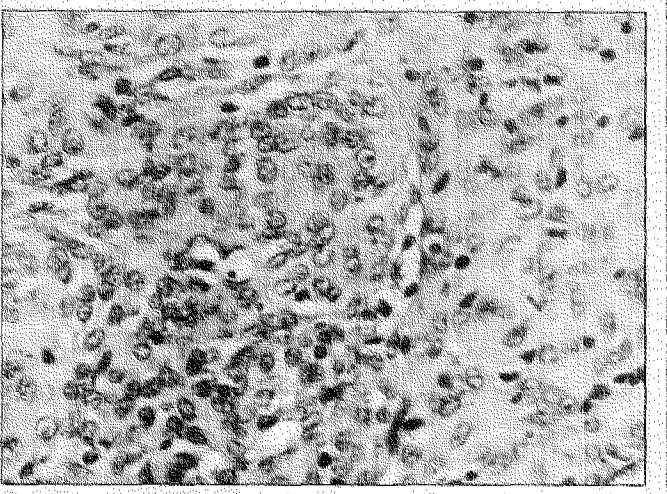
posterior fossa. The diagnosis was intracranial meningioma with extracranial extension. The patient was referred to neurosurgery for management of the intracranial portion, prior to excision of the extracranial component by the otolaryngologist via base of the skull approach. The patient subsequently underwent suboccipital craniectomy with excision of the tumor by Neurosurgery last June 6, 1991. Intraoperatively, a 3 x 3 x 4 cm grayish-pink tough, well-encapsulated mass was found adherent to the dura with bony erosion, and moderate vascularity. The tumor was attached to the posterior portion of the petrous ridge and occipital bone, stretching the lower cranial nerves. The posterior fossa was likewise eroded. The post operative course was uneventful, but the patient had facial paralysis, tongue deviation to (R) and indirect laryngoscopy showed a paralyzed left vocal cord. Three weeks later, she underwent base of the skull surgery with superficial parotidectomy and radical mastoidectomy. The incision used was an extended Y incision. Intraoperative findings showed a smooth encapsulated mass anterior to the sternocleidomastoid, and extending into the parapharyngeal space. The main trunk of the facial nerve was identified and preserved. The spinal accessory and the hypoglossal nerves were infiltrated with tumor and were thus sacrificed. The mass was traced retromandibularly towards the level of the jugular foramen where it was dissected out. During mastoidectomy, the tumor was noted to extend to the middle ear via the hypotympanum. The mastoid air cells and attic were filled with granulation tissue. The post operative course was uneventful and the patient was eventually discharged on July 5, 1991. She had deficits involving the following cranial nerves: VII, X, XI, and XII. Five months postoperatively; she has remained asymptomatic, the facial palsy has resolved, and the voice has improved. The final histopathologic report: meningioma.

HISTOPATHOLOGY:

Microscopic view of the tumor on low power shows a cellular tumor with cells arranged in islands and lobules. Higher magnification reveals that the individual tumor cells have poorly defined cytoplasmic borders with moderate amount of eosinophilia and cytoplasm. There are vesicular nuclei with prominent nucleoli. No mitotic figures are seen. (See figures 2 and 3).

(FIGURE 2: LOW POWER VIEW OF THE MENINGIOMA.)



(FIGURE 3: HIGH POWER VIEW OF THE MENINGIOMA)

DISCUSSION:

This is the case of a 25 year old female presentng with three year history of a left infraauricular mass, aural polyp and parapharyngeal bulge. These signs were associated with tinnitus, hearing loss and headache. The differential diagnosis considered for this case were: parotoid gland tumor and jugular paraganglioma.

Parotid gland tumors may be present as a pre-, infra- or post auricular mass depending on the area of gland affected. If the deep lobe is involved, a bulge in the oropharynx (often pushing the palatine tonsil medially) may be seen. These tumors can gain access to the external auditory canal via the foramen of Huschke, fissures of Santorini or the hypotympanum. Tinnitus or hearing loss may also be present. However, there has been no report of intracranial spread by extension or metastasis. The most common benign tumor of the parotid gland is the benign mixed tumor or pleomorphic adenoma; on the other hand, mucoepidermoid cancer and acinic cell carcinoma are the more common malignancies involved(1).

An aural polyp with a parapharyngeal bulge may be seen in a jugular paraganglioma. This is a benign neoplasm of the middle ear and may spread to the parapharyngeal area following the course of the jugular vein. The patient may also have tinnitus, hearing loss and dizziness as well as cranial neuropathies involving the facial, glossopharyngeal, vagal, spinal accessory and hypoglossal nerves. Also known as glomus jugulare or glomus tympanicum, this tumor originates from the adventitia of the dome of the jugular bulb. This is a well encapsulated newgrowth composed of epithelial cells divided and surrounded by a richly vascular stroma. There is a familial tendency and women of the middle age group are more often affected(4).

To the surprise of the clinician, the biopsy results of the aural polyp was meningioma. Further investigation showed that indeed there was a primary intracranial lesion albeit the absence of neurological signs.

Meningiomas are tumors made up of the meningeal cellular elements and its derivatives in the meningeal spaces. These include the arachnoid villi cells, dural fibroblasts, and pial cells(5). These tumors are characteristically well defined and lobulated. They may be homogenously pink to gray in color, and firm in consistency. They may also present "en-plaque", meaning that the mass has a diffusely spreading pattern. They are seldom infiltrative, but may compress adjacent nerves and cause hyperostosis of bone. Four microscopic patterns are recognized, but these have no prognostic significance (6). These tumors may pass through the skull foramina and bony defects into the nasopharynx, paranasal sinuses and infratemporal parotid region. Patients therefore may show signs and symptoms pertaining to area of spread: a) ear: discharge, hearing loss, tinnitus, and vertigo; b) nose: nasal congestion and obstruction, epistaxis and anosmia; c) parapharyngeal area: lateral neck mass, dysphagia, dysarthria and dysphonia. They may be clasified into four groups:

A. Extracranial extension of a meningioma with an intracranial origin (secondary)

B. Extracranial extension of a meningioma arising in a neural foramen (primary)

C. Ectopic without any connection either to a foramen of a cranial nerve or to an intracranial structures (primary)

D. Extracranial metastasis from an intracranial meningioma (secondary)

However, only 20% of intracranial meningiomas present with an extracranial extension(3).

This patient thus had an intracranial meningioma with an extracranial extension (which is by far the most common subtype). Spreading extracranially via the

skull foramina, itinvolved the spinal accesory and hypoglossal nerves, as well as the ear, and parapharyngeal area, thus accounting for the signs and symptoms presented by the patient.

Surgery is the treatment of choice and in this case with the collaborative efforts of the neurosurgeon and otolaryngologist, a complete surgical excision was done. However, meningiomas are also known to recur. Even with complete removal of tumor there is still an 8-11% recurrence within 10 years. This increases to 29-44% if the tumor is incompletely excised. Radiotherapy and hormonal therapy are adjunctive modalities available for patients with incompletely resected tumors, recurrences and those who are poor surgical risks.

Post-operatively, the patient had neurologic defects involving cranial nerves VII, X, XI and XII. After five months, there was partial recovery of the facial nerve, and the vocal cord had compensated. There were no signs of recurrence. In summary, this is the case of a meningioma with an intra and extracranial component in a 24 year old female presenting with a three year history of a left infraauricular mass, aural polyp and parapharyngeal bulge associated with hearing loss, tinnitus and bitemporal headache. Excision of the tumor was done first via a suboccipital craniectomy by the neurosurgeon followed by base of skull surgery by the otolaryngologist. Five months postoperatively there are no signs of recurrence. Although this is a rare entity, meningiomas should be included in the differential diagnosis of all middle ear mass and an intracranial component ruled out with a CT scan.

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"not one of them is without meaning"

A SERIES OF CASE REPORTS ON THE SPECTRUM OF OROMANDIBULAR-LIMB HYPOGENESIS*

Felipe Federico O. Pio, MD**
Ma. Cristina S. Lopez, MD**
Constantine O. Cu, MD**

ABSTRACT

Patients with multiple congenital anomalies present probably the most complex diagnostic and therapeutic challenges encountered in almost all fields of Medicine. The major portion of this report is devoted to identify the patterns and scope of malformations in the spectrum of oromandibular-limb hypogenesis. The spectrum includes primarily limb deficiency, hypoglossia and micrognathia. Three cases were reviewed, of which there were two males and one female all of w h o m presented with a variety of anomalies involving the tongue, mandible and extremeties, in all instances an intraoral band was present. Clinical appraisal of the cases suggests that their separation as different entities may be artificial; they may be considered as one group of anomalies. It appears therefore that variations in expression could exist. The etiology is still unknown, although experimental teratology suggests that drugs or maternal illnesses could have been the cause. The otolaryngologist's expertise is increasingly sought in the identifiction of these cases. It may be said however, that no single subspecialty is eminently qualified to deal with the multidimensional problems presented by these malformations.

INTRODUCTION

Every malformation represents an inborn error in morphogenesis. Just as the study of inborn metabolic errors has extended our understanding of normal biochemistry, so the accumulation of knowledge concerning defects in morphogenesis may assist us in further unraveling the story of structural development.

Patients with multiple congenital anomalies present probably the most complex diagnostic and therapeutic challenges encoun tered in almost all fields of medicine. Comprehensive management of these patients reaches beyond the traditional confines of any single subspecialty.

The otolaryngologist by virtue of his skills in head and neck medicine surgery, is naturally prepared to contribute significantly to the study and care of craniofacial anomalies.

The major portion of this report is devoted to identify the patterns and scope of malformations in the Spectrum of the Oro- mandibular-limb Hypogenesis. It is hoped that the presentation of this entity will lend itself to practical clinical application for a better understanding of alteration in its morphogenesis.

Accurate diagnosis of a specific syndrome among babies born with multiple malformations is a necessary prerequisite in pro viding a prognostic evaluation and plan of management for the affected infants and to include probably genetic counselling for the parents.[1]

In 1932, **Rosenthal** described aglossia and associated malfor mations.[2] More recently, **Kaplan** et al., have emphasized a commu nity or spectrum of face-limb malformation syndromes and have suggested common elements in modes of developmental pathology.[3] Oromandibular-Limb Hypogenesis Spectrum (Hypoglossia-Hypodactyly Syndrome, Aglossia-Adactyly Syndrome, Glossopalatine Ankylosis Syndrome, Moebius Syndrome, Charlie M. Syndrome, Facial-Limb Disruptive Syndrome) primarily includes Limb deficiency, Hypo glossia and Micrognathia.

The natural history of this spectrum pertains to several abnormalities. Early feeding and speech difficulties may occur. Orthopedic and Plastic Surgery may be indicated for the limb problems. Intelligence and stature may be normal. Serious problems with hyperthermia can occur in children with four-limb amputation.

The etiology is still unknown, though this is usually spo radic. The hypothesis that the abnormalities are the disruptive sequence of hemorrhagic lesions has experimental backing of **Poswillo** in his works on the Pathogenesis of the First and Second Branchial Arch Syndrome. The presumed vascular problem which has been mentioned, is more likely to occur in distal regions, such as the distal limbs, tongue and occassionally parts of the brain.[4]

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* First Prize, PSO-HNS Interesting Case Contest held at Hotel Nikko, Manila Garden, Garden, July 9, 1992.

** Residents, Dept. of ENT, UST.

CASE REPORTS

CASE 1

After a male infant was born in 1985 following an "uncomplicated" delivery by a traditional midwife, the parents immediately noted anomalies of the hands, feet and mouth.

The pregnancy was a full term one, the mother had cough and colds on the first trimester. There was a history of intake of oral contraceptive pills for four years and an interval of around three to four months pill-free period prior to conception. The mother denies any history of exposure to Roentgen rays.

The father and the mother were 32 and 27 years old respectively, unrelated and apparently in good health. The mother has had three normal full term deliveries and had no history of abortion. The matermal family history was normal, however the second degree cousin of the father had congenital amputation of the four extremities.

D.A., was three years old when first seen. The birth weight was unknown and the weight at three years was 13 kgs. The mandible was small and receding. On examination of the oral cavity, there was a midline oro-palatal band about 2.0 cm x 0.5 cm in size, absence of tongue, absence of lower teeth and a small oral vestibule. The tympanic membrane was perforated with yellowish discharge over the left ear. The neck, heart, lungs, abdomen, genitalia and rectum were normal.

He had terminal transverse amputation from the proximal third (below the elbow) of the left upper extremity, with terminal transverse amputation of the second and third distal phalanges of the right upper extremity. Likewise, a terminal transverse amputation from the proximal third (below the knee) of the left lower extremity and terminal transverse amputation of the second toe of the right lower extremity.

Radiographic examination of the extremities were compatible with the physical examination. The maxilla and temporomandibular joints were normal on x-ray but the mandible was hypoplastic.

At operation, the oro-palatal band was released uder general anesthesia. The lower buccal sulcus was absent and the palate was found to be intact. Postoperatively, our patient was able to take his feedings well and was gaining weight on subsequent follow-ups.

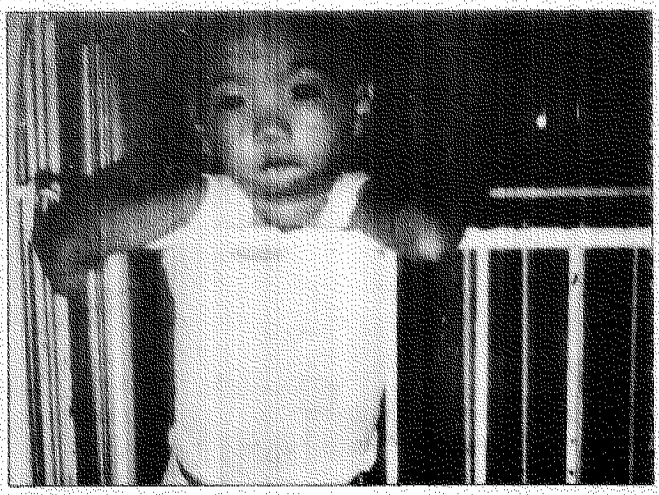
CASE NO. 2

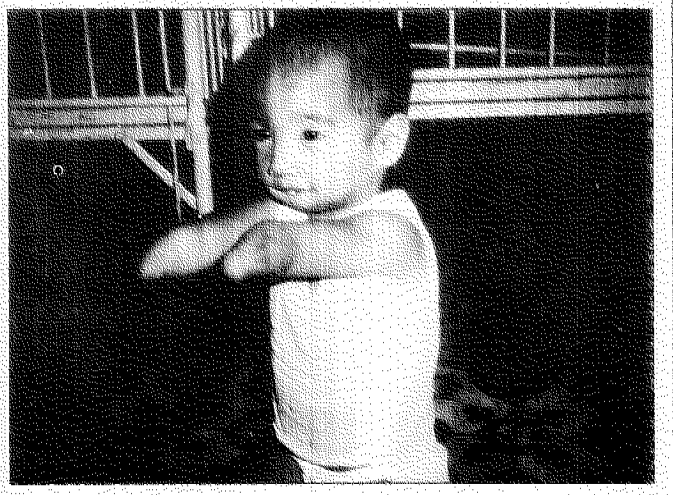
J.S., a male infant was brought to us on February 14, 1990 because of multiple congenital anomalies. He was subsequently admitted. The patient was left in an orphanage with no known perinatal, natal nor postnatal history.

Physical examination revealed an awake, alert, and active infant, the lower lip was attached to the hard palate, and there was micrognathia. The patient has complete transverse amputation of the left and right upper extremities and left lower extremity. The remainder of the physical examination were unremarkable.

The patient was referred to Rehabilitation Medicine. Radio graph examination of the extremities were requested and showed that both the upper extremities had congenital absence of the mid and distal third of the forearm including that of the hand. The radius and ulna were also markedly shortened. Left lower extremity shows absence of the bone structures below the ankle joint with no stump of soft tissue. The right lower extremity has no demonstrable congenital abnormality. Placing of stabbies over both upper extremities were contemplated, when patient will be six month old or when he is able to sit. The mandible was small and

CASE NO. 1: DEMPSEY





CASE NO. 1: JOSEPH MARTIN

underdeveloped on radiographic examination.

Likewise, the patient was referred to Nutritional Rehabili tation Unit-Pediatric service and was given a dietary prescrip tion and suggested that feeding will be given per nasogastric tube.

The gingivo-labio-palatal band was released and noted that there was hypoglossia. Postoperatively, our patient was able to take his feedings from a nipple for the first time and he was discharged one week after surgery.

CASE NO. 3

G.P., a 15 month old girl was presented in January 1991 because of multiple congenital anomalies. At birth the patient was noted to have abnormalities of the mouth, hand and feet. Birth history revealed that she was born to a 34 year old G6P5 mother. She was delivered full term by normal spontaneous deliv ery at a provincial hospital with a birth weight of 3.5 kgs.

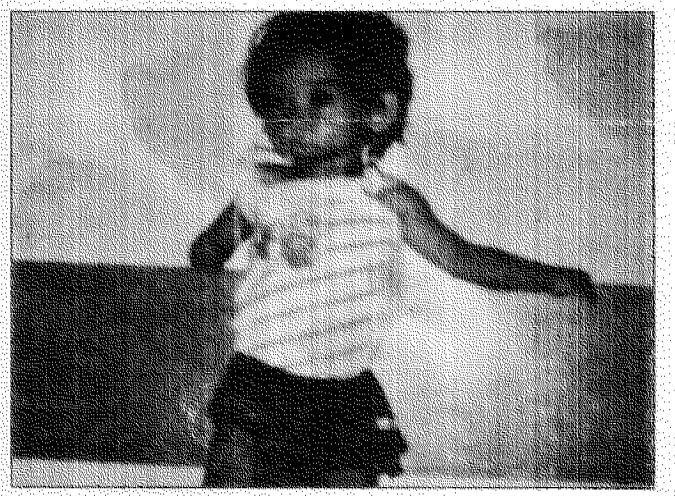
Prenatal check up of the mother started at 5 months AOG. There was a history of intake of oral contraceptives two months prior to conception. The mother developed fever and rashes on the fifth month of pregnancy for which she took Paracetamol. A history of dysuria on the 8th month of pregnancy was known without intake of any medications. There was no history of exposure to Roentgen rays.

G.P. stayed at the nursery for a week. A nasogastric tube was inserted for feeding. On admission, the patient had normal vital signs. She was noted to have transverse terminal amputa tion of the right arm, absent last three fingers with syndactyly of the 4th and 5th fingers of the left hand and terminal amputation of the right. The mandible was hypoplastic, a labio-gingi val-palatal band was present and there were 4 upper incisors. The heart, lungs and abdominal examination were normal.

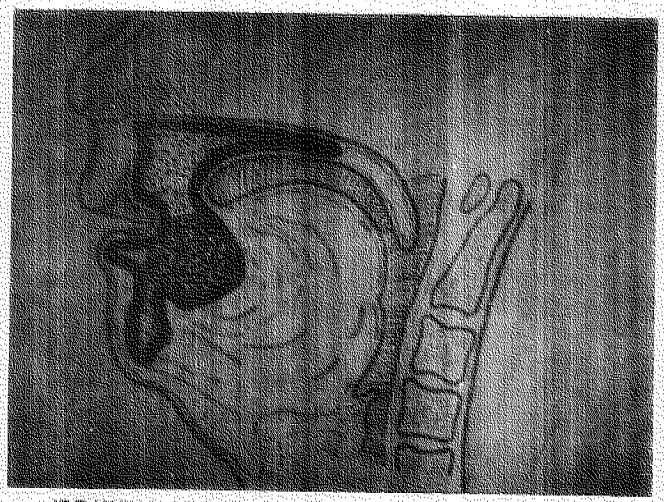
On the first hospital day, several laboratory work-ups were done. Radiographic examination of the skull, mandible and extremities were done and revealed congenital absence of the phalanges of the first and second digits of the left hand, trans verse linear bands of density in the distal aspect of both femur and proximal aspects of both tibia. She was then referred to ENT and Pediatrics for further evaluation and co-management. Apparently the patient was doing well.

On the 29th hospital day, chest x-ray was done and revealed Primary pulmonary infection. She was then started on Anti-Koch's medications.

She was scheduled for release of the oro-palatal band on the 52nd hospital day. The bony and soft tissue adhesions connecting the lower labial and mandible to the hard palate and alveolar ridge was released using



CASE NO. 1: GODNESS



(DRAWING OF PREOPERATIVE CONDITION, SHOWING PALATO-GLOSSAL ANKYLOSIS)

blunt dissection and removal of bony adhesions using mallet and chisel. VY lengthening of the lower labial area was done and silastic oral airway was inserted. OR findings also revealed absence of the floor of the mouth and tongue tissue were noted. Operatively, our patient developed high temperature ranging from 37.7°C to 40.7°C. Four hours post-op, the patient developed dyspnea with grunting, head bobbing, intercostal and subcostal retractions. Tracheostomy was done. After an hour, she became bradycardiac, went into cardiorespiratory arrest. Cardiopulmonary resuscitation was done but to no avail she was not revived. At that time, metabolic/electrolyte imbalance and malignant hyperthermia were suspected causing the demise of the patient. No autopsy report was done.

DISCUSSION

Morphogenesis is a truly and sequential process in which a defect of one structure may compromise the formation of subse quent structures.

Review of the three cases presented has shown a variety of anomalies involving the tongue, mandible and extemities. In all instances an intraoral band was present.

Kramer is credited with first describing Glossopalatine Ankylosis in 1911.[5] Very few reports exists concerning this syndrome that has appeared in the world literature. Critical appraisal of these case suggests that their separation as differ ent entities may be artificial: they may be considered as one group of anomalies, characterized by hypoplasia of the oromandib ular area and of the distal extremities.

Possible embryonic mechanism are complicated and knowledge of them is incomplete. From days 20 to 30, paraxial mesoderm on each side of the notochord becomes segmented and forms the basis for the development of the axial skeleton and musculature. During this period, the branchial arches and clefts appear and development of the face begins. By day 25, the mandibular and maxillary process of the first arch bound the stomodeum; some what later, the buccopharyngeal membrane completely ruptures, resulting in continuity between the ectoderm and endoderm.[6]

The tongue is well formed by the 7th week. The formation of the palate commences between the 7th and 8th weeks; about the 9th week, the tongue drops down, allowing the palatal shelves to change from vertical to horizontal and to fuse with each other.

In the adult, the buccopharyngeal membrane is represented by a line in the posterior region of the oropharynx between the anterior pillars. Possibly during the development, the posterior migration is interrupted, resulting in the persistence of mem brane in a more anterior position. This remnant could cause micrognathia by a briddling effect on the growth, an apparent hypoglossia by preventing fusion of the ventromedial portion of the first arch. But still, the pathogenesis of these oral mani festations is unknown.

The presence of intraoral band accompanied by micrognathia, hypoglossia and hypoplastic anomalies of one or more extremities were observed in all patients presented. Almost all anomalies represent a hypoplasia of the involved elements, like that of the extremities which has always been distal to the humerus or the femur. The limb buds appear by day 32. By day 37, the upper limb has subdivided into arm, forearm and hand while the hind limb is still paddle-shaped. By day 46, the digits of the hand are separating, while those of the foot still appear as united rays. In general, the upper limb is about one week ahead of the lower limb in its development.[8]

Experimental teratology suggests that drugs (progesterone, etc.) and maternal illness which are present in the history of our two patients may have been blamed for various congenital malformations. This indicates perhaps that this is probably a true spectrum and not just a set of malformations that have occured by chance. Although variance could still exist in the extent of abnormality (expression) among the individuals belonging to the same syndrome is a usual phenomenon.[9]

The primary hypoplasia or dysplastic growth of the mandible could have created a problem since this will prevent the normal descent of the tongue between the palatal shelves, thereby producing the oro-palatal band. There should be caution on the release of the band because the tongue may fall downward and backward due to the lack of jaw support. Some authors have tried glossopexy, but it is advised that a tracheostomy be made with decannulation after 18 months. One sugges-

tion was mandibular osteotomy to correct the hypoplastic mandible. It has been demonstrated in literature that subsequent mandibular growth catches up so that a normal profile is achieved by 4 to 6 years of age, so, we deem this unnecessary in all our cases.[10]

The three cases we have reviewed highlights not only the otolaryngologist's role in diagnostic and therapeutic endeavors in the care of patients with these kinds of anomalies, but is therefore to be hoped that a multidisciplinary study and management be done which may become a prototype in the optimal health care of complex clinical problems.

SUMMARY

I have presented three case with common anomalies of the mandible and distal limbs and presence of intraoral bands. The pertinent literature has been reviewed.

It appears therefore that variations could exist in the spectrum of Oro-Mandibular-Limb hypoplasia.

The Otolaryngologist's expertise is increasingly sought in the identification and management of patients with craniofacial anomalies. It may be said, however, that no single subspecialty is eminently qualified to deal with the multidimensional problems presented by malformations.

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We ought not to set them aside with idle thoughts or idle words about "Curiosities" or "Chances". Not one of them is without meaning; not one that might not become the beginning of excellent knowledge, if only we could answer - why is it rare? Or being rare, why did it in this instance happen?

James Paget, 1992

INTERESTING CASE PRESENTATION
"The Case of the Forgotten Foreign Body"

Mary Arlene A. Ardena, MD*
Norberto V. Martinez, MD**
Benjamin S.A. Campomanes, Jr., MD**
Eutrapio S. Guevara, Jr., MD**

ABSTRACT

A 17 year old female was diagnosed by the Pediatrics department to have upper respiratory tract infection, asthmatic bronchitis, bronchopnemonia and primary complex for the last 9 years prior to ENT consultation. Medications afforded temporary relief. Consultations with EENT service on 2 occasions 6 years after the first symptoms occurred gave the impression of nasopharyngitis. About 2 months prior to consultation, patient complained of severe nasal stuffiness and examination by ENT resident revealed multiple pale, glistening masses in both nasal cavities consistent with nasal polyps. Bilateral polypectomy, ethmoidectomy, antrostomy and Caldwell-luc operation were done. Aside from the nasal polyps, an incidental foreign body was was seen embedded along the mucosa of the septum on the right side. The patient recalled placing the cork of a softdrink crown on her 8th birthday.

INTRODUCTION

Nasal Polyposis was first recognized in India and by 1000 BC currettes had been devised to remove them (Vancil, 1969). There has been a number if different theories put forward for the pathogenesis of nasal polyps which includes the Bernoulli phenomenon, polysaccharide changes, vasomotor imbalance, infection, and allergy. All may contribute to polyp formation, but none can be universally incriminated.
Our case exemplifies a rather unusual etiology.

CASE REPORT

This is the case of a 17 year old female from Quezon City who was brought to the SLMC Pediatric OPD 9 years ago because of cough and colds of 2 days duration. This was associated with fever and multiple cervical lymphadenopahty. Patient was diagnosed to have an Upper Respiratory Infection R/O Primary Complex. Medications which consist of mucolytic and antipyretic was prescribed. PPD was ordered which was later found to be negative. There was persistence of symptoms and after a week's follow-up, patient was noted to have crepitant rales and wheezing. She was then diagnosed to have asthmatic bronchitis and beginning bronchopneumonia. An antibiotic regimen with an expectorant was prescribed. This was taken for 10 days and symptoms noted to resolve spontaneously. However, cough and colds was noted to recur after 2 months this time with no fever. Despite the supportive medications given her, the patients continued to complain of cough and colds and had her regular monthly follow-up at the OPD.

Eight years prior to consultation, patient was diagnosed to have bronchopneumonia with asthmatic component because of persistence of cough and colds. This resolved after a month of treatment however, colds was noted to recur with post-nasal drip.

Seven years prior to consultation, patient was again noted to experience cough and colds. She was diagnosed to have Acute Upper Respiratory Infection with primary focus. An anti-PTB regimen was started with supportive mucolytic.

Five years prior to consult, patient was noted to visit OPD 3 times with same complaints and was still diagnosed to have bronchopnemonia. Different antibiotics were prescribed. Symptoms would just resolve temporarily.

Four years prior to consult, patient visited the OPD twice with the same complaints. The diagnosis was Nasopharyngitis. The patient was then referred to the EENT service for the first time. On examination, the ENT resident noted congested turbinates, bilateral, with pus coming out from the middle meatus. No other findings of note was reported. Patient was made to continue her antibiotic regimen and a decongestant was prescribed.

Two years prior to consult, patient went back to the ENT OPD because of on and off nasal discharge noted

* Resident, Dept. ENT, SLMC.
** Consultants, Dept. of ENT, SLMC.

to be yellowish, foul smelling, however, sometimes noted to be whitish and mucoid. Tonsillo-pharyngeal congestion was likewise noted. A diagnosis of Acute Upper Respiratory Infection was made. After 1 month, the patient developed cough and colds and was again referred to the Pediatric service. She was again diagnosed to have Bronchopneumonia with asthmatic component. Some antibiotic regimen was given.

Two months prior to consult, patient was noted to develop severe nasal congestion and persistence of nasal discharge. She went back to the ENT OPD and was found to have multiple, pale, glistening masses occupying the nasal cavities. This was associated with yellowish and foul smelling nasal discharge. Nasal polyposis was considered probably secondary to infection. She was then advised to undergo polypectomy, ethmoidectomy, antrostomy, and Caldwell-Luc.

Past Medical History, Personal and Social History, Family History were unremarkable.

The physical examination findings revealed multiple, pale, glistening masses coming from the middle meatus as well as the postero-superior portion of nasal cavity. Pus was likewise noted to come out from the same area. The turbinates were congested. There was no septal deviation noted. The right tonsil was slightly hypertrophied but not congested. The rest of the P.E. findings were unremarkable.

All laboratory work-ups done on this patient were unremarkable except for the X-ray of the Paranasal sinus which revealed "Haziness of the Maxillary area." The rest of the paranasal sinus are well-aerated and developed. An impression of Bilateral Maxillary Sinusitis was given.

Intraoperatively, both nasal cavities were filled with polyps. The sinuscopes were used to view origin of polyps and it showed to be coming out from the ethmoids. Proceeded with polypectomy and afterwhich a foreign body was seen embedded along the mucosa of the septum at the right side.

Culture and sensitivity of nasal discharge taken that same day showed heavy growth of *Proteus Mirabilis.*

The rest of the patient;s hospital stay was unremarkable. Final diagnosis: Nasal polyposis, maxillary sinusitis, and foreign body at the right nasal cavity. She was discharged on the 7th day.

DISCUSSION

Some foreign bodies are inert and may remain in the nose for years without mucosal changes. **Maran** also emphasized that other foreign bodies develop into rhinoliths. Many however, lead to inflammation and infection of the mucous membrane, which in turn leads to production of foul-smelling and mucopuruelnt discharge. A piece of cork found in the softdrink crown was recovered from the patient's nose after the operation. Immediately after the operation, the surgeons showed the piece of cork to the patient. It was recalled that when the patient celebrated her 8th birthday, she placed the cork inside her right nasal cavity. Because of fear that her parents might scold her, she refrained from telling anybody about the cork and decided to forget all about it. It was then documented that the patient started experiencing the symptoms (cough and colds) 16 days after the patient's birthday.

For 9 years, the patient has been seeing the SLMC-OPD and was examined by the Pediatric residents. She was most of the time diagnosed to have bronchopneumonia and primary complex of which she was given the corresponding medications. The pediatric residents somehow failed to examine the nose properly and the focus of attention that time was on the findings at the chest. The multi-disciplinary approach was not given due importance because the ENT service has not played a role in the first 6 years that this patient first experienced the symptoms.

The patient was referred to the ENT service when it became clear that the main problem came from her nose. No foreign body was seen in the nose the first time that the patient was seen since it had been 6 years that the cork was already embedded at the septal mucosa. According to **Ballantyne**, mucosal edema or granulation may hide the foreign body.

Several work-ups were done including X-ray of the paranasal sinuses which revealed maxillary sinusitis. Multiple polyposis were noted. At the OPD, the patient was subjected to rigid Hopkins telescope and no foreign body was seen. The diagnosis of nasal polyposis was confirmed by the histophatologic result. Polyposis of the respiratory mucosa may result either from vascular derangement in the mucosa or less commonly, from mechanical obstruction. **Eggston and Wolff** (1943) considered that nasal polups were produced by perophlebitis and perilymphangitis leading to edema of the lamina propria. **Tos** and **Mogenses** (1977) left that there must be rupture of epithelium and formation of granulation tissue following inflammation. Since nasal polyps had been classified as either infectious or allergic, the first group being characterized by nasal neutrophilia and purulent secretions, I would like to think of this as an infectious process.

The effect of this long-retained foreign body for 9 years was described by **Cauna** (Nasal Allergy, 2nd edition) as such that it primarily becomes the focus of infection. The infectious process served as a nerve damaging factor and this leads to vasomotor degeneration (gland denervation). The author demonstrated the blood vessels to be denerved and the mast cells to be

largely degranulated. The final step could be attained via denervation and histamine release. As this point, there is an increase vascular permability which would provide extravasation of plasma and is tantamount to the formation of edema and polyp.

Denervation of blood vessels leading to vasolidation and edema formation is certainly a factor of significance to polyp formation. Possible chronic inflammation in the upper airway mucosa as a result of foreign body reaction play a part in the formation of this neutrophil polyp, but our understanding of the association between polyps, foreign bodies, and allergies is imperfect and confusing, as is our knowledge of polyp formation.

Foreign bodies in the nose do not feature eagerly in otolaryngologic literature. Yet on occasions, they may constitute a considerable challenge to both the diagnostic and surgical skills of the Otolaryngologist. As in this case, the forgotten foreign body was characterized by chronic cough and colds, symptoms which could signify several disease entities, and by which proper history-taking would be found to be most helpful. Were it not for the polyp formation secondary to these chronic symptoms, and the subsequent surgical intervention done on this patient, the foreign body, as the primary cause of the patient's malady, would never have been discovered and the disease process would have progressed further.

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ESOPHAGEAL SLOT MACHINE

Ernestine Marie I. Domingo, MD*
Marlon del Rosario, MD**
Alberto F. Calderon, MD**

INTRODUCTION:

In the field of bronchoesophagology, some of the most intriguing cases encountered are those of foreign bodies in the upper aerodigestive tract. In adults, these foreign bodies usually come in the form of unchewed food or dentures. In children, things get more interesting. This is due to the wide assortment of foreign bodies which may be ingested or aspirated, most probably because of children's insatiable curiosity. As such, case endoscopic extraction becomes challenging indeed and is never done using one technique alone.

In some cases, even though what can be seen is obvious, what is retrieved during actual endoscopy can still be astounding. There have been many cases mentioned in the literature of ingested foreign bodies such as padlocks, necklaces, buttons, pins, toy jack-stones to name a few, and the clever ways in which they were retrieved. With this the Department of Otolaryngology-Head and Neck Surgery of the Ospital ng Maynila, would like to present this simple case of a five year old child, who, because of her curiosity, accidentally swallowed a fifty centavo coin.

CASE HISTORY:

A five year old girl was brought to our institution on March 3, 1992, a referral from a private hospital. History revealed that the patient ingested a fifty centavo coin three hours prior to admission. Shortly after this the patient became cyanotic, although only transient. The mother then noted drooling of saliva for which the patient was rushed to a private hospital in Las Pinas, where AP views of the neck, upper chest, and abdomen were made. These revealed a circular radio opaque foreign body, roughly the size of a fifty centavo coin in flat presentation at the level of the 4th and 6th cervical vertebrae. Endoscopic extraction was contemplated and the patient was referred to our institution for the procedure. On examination the patient was noted to be restless, apparently trying to vomit something previously swallowed. Further evaluation was not possible. After assessment of the X-ray plates and owing to the urgency of the situation due to the great discomfort of the child, an esophagoscopy was done. Using a rigid esophagoscope, extraction of the foreign body then commenced. And lo and behold, viewed "on edge" was fifty centavos all right, but not in one piece--but as two twenty five centavo coins, one on top of the other. The patient's mother was correct! The doctors were somehow deceived--particularly with the AP film showing what appeared to be a fifty centavo coin from all appearances. Post esophagoscopic X-rays were normal.

** Resident, Dept. ENT, PLM-OM.
*** Consultants, Dept. of ENT, PLM-OM

DISCUSSION:

Once in a while, a simple problem comes to otolaryngologists, and yet leaves physicians stupefied. Owing to the great multitude of foreign bodies which one can encounter in pediatric esophagoscopy, something like this novel case was bound to happen. Many factors both controllable and uncontrollable occur to further mislead physicians.

The perfect roentgenographic illusion created by the magnification of the aligned twenty five centavo coins, amazingly made it look like a single fifty centavo coin. Pre-endoscopic impressions of a fifty centavo coin could mislead an endoscopist to use a shorter scope with a bigger lumen--easier to work with but with a big disadvantge. It disables the endoscopist from following through should the foreign body progress downwards.

It was easy enough to localize the coin in the esophagus. After all that dictum that any flat foreign body, such as a coin, will always place its greatest diameter in the coronal plane of the body, if it is in the esophagus and in the sagittal plane if it is in the trachea. This has yet to be disproven. It is for this reason that no lateral view was taken.

The endoscopist should always maintain a certain degree of cautiousness in order to detect novelties such as this. One should also be skillful enough to employ the necessary variations in the removal of foreign bodies, each method being tailor made for a specific type. One should not confine himself to textbook procedures in these aspects personal ingenuity will always be welcome.

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OTOMYCOSIS: ITS MYCOLOGY AND A COMPARISON OF TWO TREATMENT REGIMEN*

Guia C. Tan, MD** • Rhodora A. del Rosario, MD**
Ramon E. Gonzales, MD** • Rina T. Reyes, MD**
Jose U. Hardillo, MD** • Josel V. Mijares, MD**
Generoso T. Abes, MD***• Joselito C. Jamir, MD***

ABSTRACT

Thirty three patients with otomycosis were seen at the Out-patient Clinic of ENT over a one-year period. The predominant age range of the patients were in the second to fourth decade of life with the disease more common in females. The most common presenting symptom was pruritus and the greatest predisposing factor was the use of unconventional instruments in the cleaning of the ear. Otoscopic findings were mainly that of hyphal elements or ear discharge. The most common fungal isolates obtained were Candida and Aspergilus. Fifteen ears were put on Clotrimazole and fifteen ears were put on guava extract. All fifteen ears with Clotrimazole resolved after one week of therapy. Ten patients treated with guava leaf extract got well after one week and four patients responded after two weeks. One patient did not respond at all. The data showed a positive trend in the use of guava leaf extract which will hopefully provide a cheaper, readily available alternative in the management of otomycosis.

INTRODUCTION:

Otomycosis is a fungal infection of the external ear commonly encountered by the otolaryngologist in places with hot and humid temperatures like the Philippines. Its mycology has been studied extensively abroad and foreign literature reports *Aspergillus niger* and *Candida albicans* as the most common etiologic organisms isolated. Patients with the disease usually present with irritation of the ear canal in the form of ear fullness, itchiness or pain and otoscopic findings reveal hyphal elements or creamy, cheesy debris typical of the etiologic organisms. Several treatment modalities have also been experimented with of which the antifungals, Nystatin and Clotrimazole have been found to be the most effective.

In the Philippines, there has been no local study of the mycology of otomycosis. Furthermore, there have been anecdotal reports of the effectiveness of guava leaf extract, a cheap and readily available alternative, in the treatment of the disease. This study, therefore, was conducted with the following objectives:

1. To describe the local mycology of the disease.
2. To describe the population of those affected with the disease, the symptomatology and the otoscopic findings of otomycosis.
3. To determine the response of otomycosis to guava leaf extract in comparison to its response to Clotrimazole.

MATERIALS AND METHODS:

The study was conducted at the Out-Patient Department of the Department of Otolaryngology from September 1991 to August 1992. Patients, 15-60 years old, with a diagnosis of otomycosis based on symptoms of itchiness, sensation of ear fullness, pain, and ear discharge and otoscopic findings of hyphal elements, canal narrowing or discharge were included in the study. The exclusion criteria were (1) perforated tympanic membrane, (2) concomitant chronic otitis media and (3) those who had undergone previous mastoidectomy. A uniform data sheet was used to document the preceding data. Fungal cultures were then obtained after which the ears were suctioned and cleaned. The ears of patients were then consecutively and alternately assigned to two treatment regimens namely, Clotrimazole and guava leaf extract after informed consent was given.

Patients in the Clotrimazole group were given Canesten liquid solution (Bayer) (1% in a 15 ml solution) and instructed to apply it 3 drops 3 times a day in the affected ear. Those assigned to the guava leaf decoction were treated with an extract previously prepared by boiling 10 g of guava leaves in 100 ml of Water for 15 minutes. This solution was strained and allowed to cool and then stored in a sterile container in the refrigerator for a maximum of 5 days. The patient placed 3 drops of

* Presented at the Bachringes-Ingelheim Clinical Research Contest held at Anahaw Room, Silahis Hotel, Oct. 23, 1992, Manila
** Residents, Dept. of ENT, UP-PGH
*** Consultants, Dept. of ENT, UP-PGH

the extract in the involved ear 3 times a day.

The patients were then followed up on weekly basis. Success of the treatment was substantiated by the resolution of the ear symptoms and the ear discharge after one week of therapy. Those who had resolution of the ear discharge and other symptoms but had persistence of other symptoms (mainly itchiness), were considered partially treated and were continued on the guava leaf extract for another week. Those patients who had no change in the baseline status after a week or who had persistence of symptoms after 2 weeks of guava leaf extract treatment were considered to have failures of treatment and were given Clotrimazole for another week.

RESULTS:

A total of 33 patients were enrolled in the study. There were 7 dropouts. The age distribution of the patients is shown in Table 1. Majority of the patients fell in the 3rd - 4th decade of life. There were more females with a female:male ratio of 2.3:1. The left ear was affected more and 33.3% were bilateral infections (Table 3).

Pruritus was the most common presenting symptom followed by pain and sensation of ear fullness (Table 4). Otoscopic findings frequently revealed hyphal elements as shown in Table 5. Table 6 lists predisposing factors associated with otomycosis. The most common of these was the use of unconventional instruments for cleaning the ear like matchstick, hairpin, cerumen spoon and feathers.

Fungal cultures were done but the yield was not very high. *Asperigillus fumigatus* and *Candida albicans* were the most common fungal isolates (Table 7). It should be noted, however, that 14 cases did not grow any fungus. Results of the treatment regimen are shown in Table 8. All 15 patients treated with Clotrimazole responded after 1 week of treatment. Using the guava leaf extract, only 10 patients got well after 1 week, 4 patients responded after 2 weeks of treatment, while 1 patient did not respond at all.

TABLE 1. Age Distribution of patients with otomycosis

AGE	N	PERCENT (%)
15-20	2	6.0
21-25	1	3.0
26-30	7	21.2
31-35	6	18.1
36-40	1	33.3
41-45	1	3.0
46-50	2	6.0
51-55	1	3.0
56-UP	2	6.0
TOTAL	33	100

TABLE 2. Sex distribution of patients with otomycosis

SEX	N	PERCENT (%)
Female	23	69.7
Male	10	30.3
TOTAL	33	100

TABLE 3. Laterality of Otomycosis in the population studied

LATERALITY	N	PERCENT (%)
Right	9	27.3
Left	13	39.4
Bilateral	11	33.3
TOTAL	33	100

TABLE 4. Frequency of Presenting Symptoms in Patients with Otomycosis

PRESENTING SYMPTOM	N	PERCENT (%)
pruritis	28	39.4
pain	17	24
sensation of fulness	16	22.5
ear discharge	10	14.1
TOTAL	71	100

TABLE 5. Frequency of Otoscopic Findings in Patients with Otomycosis

OTOSCOPIC FINDING	N	PERCENT (%)
hyphal elements	26	59
ear discharge	9	20.5
narrowing of EAC*	9	20.5
TOTAL	44	100

* External auditory canal

TABLE 6. Frequency of Predisposing Factors in Patients with Otomycosis

PREDISPOSING FACTOR	N	PERCENT (%)
use of unconventional instruments	16	41.0
freq. manipulation	9	23.1
ears getting wet	7	17.9
use of otic drop	6	15.4
systemic disease	2	5.1
TOTAL	44	100

TABLE 7. Results of Fungal Cultures of Otomycosis

ISOLATE	N	PERCENT (%)
Asperigillus		
A. fumigatus	4	12.1
A. niger	2	6
A. feanus	1	3
Candida		
C. albicans	4	12
Candida sp. (not albicans)	2	6
No fungus isolated	13	39
No culture done	7	21
TOTAL	33	100

DISCUSSION:

Otomycosis is a ubiquitous disease and often bothersome for both the patient and the physician. Factors present in the external auditory canal such as moisture, warmth and some proteins or carbohydrates favor the growth of many fungi and bacteria (Yehia, et al., 1990), given the humidity in the Philippines.

Findings in this study showed that otomycosis was more frequently encountered in the 20-40 age range. This is comparable to those found by Yechia (1990) at 16-30 and by Muligston and O'Donoghue (1985) at 21-50. The disease is usually unilateral, with little difference in laterality. There is a 2.3:1 female to male ratio. Yehia (1990) noted a similar ratio and explained that the females in the study were mostly housewives who frequently cleaned and swept floors and the resulting dust contained fungal spores and act as the predisposing factor. However, in Bahrain, Paulose (1989) found the disease more common in males who often wear a customary head dress.

The most common presenting symptom present was itching (39.4%). This was likewise noted by Paulose (1989) and Yehia (1990). Oftentimes, this caused an intolerable urge to self-clean the ear inducing further trauma to the external auditory canal. Other symptoms include pain, sensation of fullnenss and, occasionally, ear discharge.

Otoscopic findings frequently revealed hyphal elements, and varying degrees of signs of trauma like narrowing of the canal. Sometimes, there is a characteric appearance in the ear which can be correlated to the etiologic agent. In *Candida albicans* infection, the auditory canal was filled with a cheesy moist material while aspergillus showed characteristic mycelia with conidiopores.

Associated factors with the disease were likewise investigated. (Table 6). Many patients used "unconventional" instruments for cleaning their ears like matchstick, hairpin and feather and cerumen spoon. There was 17.9% of patient whose ears got wet because of swimming. Only 2 cases had systemic diseases, one had diabetes, and the other, leprosy.

The fungal isolate grown are shown in Table 7. The most common isolates were *Aspergillus fumigatus* and *Candida albicans*. These findings were similar to those found by other authors (Yehia, 1992, Paulose, 1989, Muligston and O'Donogue, 1985). *Aspergilli* are common airborne saprophytes and can readily be isolated from house dust. Yassin (1954) demonstrated that the commonly pathogenic *Aspergilli* have their optimum growth rate in the pH range of 5-7. Furthermore, they grow maximally at temperature of 37°C, a fact clinically supported by the predilection of fungi to grow in the inner one-third of the ear canal and thus near to the case temperature, as found in a study conducted in a temperate country. Fourteen cases did not grow fungus. This was attributed to a possible technical error as the reproductive part of the fungus might not have been included in the swab.

Conley (1984) showed that the external auditory canal fulfills many of the conditions for fungal growth: moisture, warmth and some protein or carbohydrate. Furthermore, the pH of the normal ear canal is on the acidic side of neutrality. This situation coupled with the high humidity level in the Philippines makes otomycosis a very common ENT problem. Two treatment regimens were compared in this study: clotrimazole and the guava leaf extract. Clotrimazole (Canesten(R)) is an antifungal agent whose mode of action is the alteration the cell membrane of the fungus so that amino acid transport is changed, impairing subsequent protein synthesis. It is commercially available in cream and liquid solution. Clotrimazole is active against Candida, Aspergillus, Trichomonas and the dermatophytes. Clotrimazole was shown to have greater zone of inhibition in in vitro studies versus fungi and bacteria in comparison to nystatin, amphotericin B, miconazole and natamycin thus prompting its choice over nystatin, the only other drug which comes in otic solution preparation.

As a traditional herbal medicine, the guava leaf extract has been used in a wide variety of common ailments. These are: as an astringent, antidiarrhetic, febrifuge and antispasmodic. In a study by the College of Dentistry (1986), they used guava leaf decoction as an antiseptic in tooth extraction.

The components of the guava leaf are: pectin, essential oils - eugenol, malic acid, tannic acid, saponins, amygdalin, fixed and volatile oils. In an unpublished report by Aloha, et al (1982), they documented the antifungal properties against *Candida albicans* using the Kirby - Bauer disc diffusion method.

There were a total of 30 ears treated, divided equally

between clotrimazole and guava leaf extract. The results of the study are shown in Table 8. The 15 ears treated with clotrimazole responded completely after one week of treatment. Using the guava leaf extract, 10 ears got well after one week ; 4 cases became asymptomatic after 2 weeks and only one did not respond and thus was shifted to clotrimazole. Using the Fisher exact test (p=0.04), the data shows that the response rate of clotrimazole and guava leaf extract are not the same. However, the data on the whole shows a positive trend and increasing the sample size will probably establish a statistically significant pattern.

While clotrimazole continues to be the gold standard of treatment of otomycosis, the guava leaf extract is valuable as an alternative mode of therapy. Even in the urban setting where clotrimazole (or Nystatin for that matter) is difficult to come by, the guava decoction is readily accessible to the willing consumer. The patient also benefits cost-wise. Definitely, it is more available to the patient in the province. Clotrimazole has also been shown to cause mucosal irritation when applied topically, although none of the patients in this study exhibited this side effect. However, in this study, it is emphasized thet the guava leaf decoction is used in intact tympanic membrane cases.

LIMITATIONS AND RECOMMENDATIONS:

1. The study could not be blinded due to the distinct smell of the guava leaf extract as compared to the relatively odorless clotrimazole solution.
2. The sample population should be increased.
3. Perhaps it would also be of clinical interest to find out if there is any difference in those patients who are treated solely by mechanical cleansing versus those treated with clotrimazole or any trial drug.

SUMMARY:

In summary, 33 patients diagnosed to have otomycosis were seen at the Out-Patient Department of ENT over a one-year period. The predominant age range of the patients were in the 2nd to 4th decade of life with the disease more common in females than in males. The most common presenting symptom was pruritus and the largest predisposing factor was the use of uncconventional instruments in the manipulation of the ear canal. Otoscopic findings were mainly that of hyphal elements or ear discharge. The most common fungal isolates were Candida and Aspergillus.

Fifteen ears were put on Clotrimazole and 15 ears were put on guava leaf extract in a consecutive randomized manner. All 15 treated with clotrimazole resolved after one week of therapy. Ten patients treated with guava leaf extract got well after one week and 4 patients responded after two weeks. One patient did not respond at all. The data shows a positive trend in the use of guava leaf extract. Hopefully, the guava leaf extract will be a cheaper, readily available alternative in the therapy of otomycosis.

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PRIMARY MALIGNANT MELANOMA OF THE PARANASAL SINUSES
A Case Report*

Anna Marie Fong, MD**
Rafael Perlas, MD**
Ma. Clarissa Fortuna, MD**

ABSTRACT

Malignant melanoma is not exclusively a pathology of the skin but it is noted to occur also in the paranasal sinuses. Approximately 6.3 - 10% of melanomas occur in the mucous membranes with 0.5% -3.6% occurring in the nasal cavities and sinuses. Like other paranasal sinus malignancies, these tumors present commonly with facial swelling, nasal obstruction and epistaxis. Accurate diagnosis is made with proper identification of intracellular melanin pigment and the junctional activity. Total surgical excision remains the treatment of choice whereas radiotherapy, chemotherapy and immunotherapy have mainly palliative roles. What makes malignant melonoma of the paransal sinuses a formidable disease is that not only it is rare and more agressive than its cutaneous counterpart, but it also carries a poor prognosis and an unpredictable course. For these reasons, it is worthwhile to report our first-hand experience with primary malignant melanoma of the paranasal sinuses.

INTRODUCTION

Few of us realize that melanocytes are found nor only in the skin but also in the mucous membranes of the head and neck. **Zak** and **Lawson**,[1] in their study, showed that dendritic cells containing melanin are present in the respiratory epithelium and within the stroma of the nasal septum as well as both inferior and middle turbinated in man. **Golman et al**,[1] demonstrated the presence of melanocytes in the laryngeal mucosa in both Caucasians and Negros. These cells are present on the mucous membranes in far fewer numbers than on the cutaenous surfaces of the head and neck, yet they still have the ability to undergo malignant transformation. Mucosal melanomas in the head and neck are most likely primary lesions that arise de novo. Only rarely does melanoma metastasize to the head and neck region.[2]

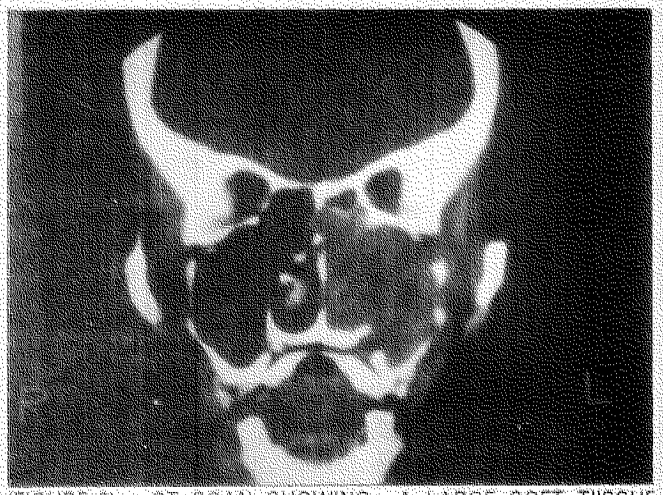
All previous foreign publications agree that malignant melanoma of the upper respiratory tract is an infrequently encountered disease. On review of literature, true incidence rates are not available and the incidence of malignant melanoma of the nose and paranasal sinuses iss best expressed as the frequency with which it occurs within a large group of patients suffering from this neoplasm. Approximately, 10-20% of all melanomas occur in the head and neck region,[3,4,5] with 6.3 - 10% occurring in the mucous membranes,[3,6] Of these, 0.5 - 3.6% were noted to occur primarily in the nasal cavity and paranasal sinuses.[3,6] Although infrequent, the clinician must be aware of the occurrence of this disease entity in these anatomical areas. The purpose of this paper is to present a documented case of primary malignant melanoma in the paranasal sinuses.

CASE REPORT

A 57 year old male from Cabalantian Bacolor, Pampanga was admitted last July 1991 because of a left sided facial swelling that started 4 months prior to

(FIGURE 1) PATIENT PRESENTING WITH LEFT FACIAL SWELLING.

* Presented at the PSO-HNS Interesting Case Contest held at Hotel Nikko Manila Garden, July 9, 1992.
** Residents, Dept. of ENT, UST.



(FIGURE 2) CT SCAN SHOWING A LARGE SOFT TIISSUE MASS IN THE LEFT MAXILLARY SINUS WITH DEFENITE BONE DESTRUCTION.

admission. This was accompanied by occasional blood stained nasal discharge and vomiting of fresh blood. Two weeks PTA, he developed left nasal obstruction and tearing of the left eye.

The patient was hypertensive but non-diabetic. He worked for 30 years at a cigarette factory as a machine operator. He had a 20 pack years of smoking history and at the same time drank alcoholic beverages excessively. Review of systems was non-contributory.

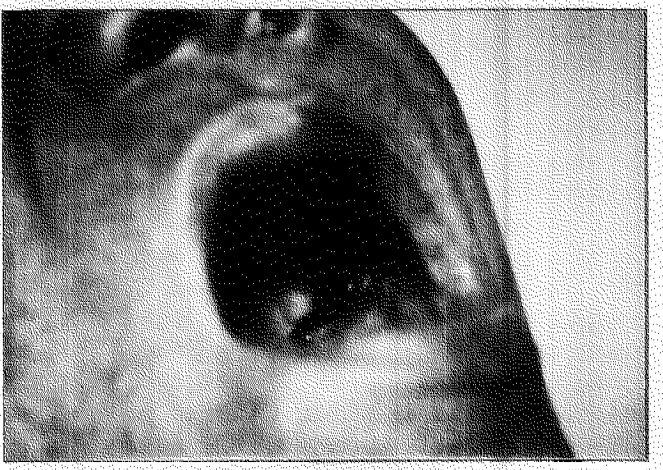
Anterior and posterior rhinoscopy revealed a friable, dark colored mass within the left nasal cavity which appeared to originate from the middle meatus. Local examination of the oral cavity revealed bulging mucosal irregularities along the junction of the hard and soft left palatal area. There was swelling of the left side of the face involving the cheek and the nasolabial fold (Figure 1). Laryngoscopic examination was normal and there was no evidenced of cervical lymphadenopathy. Ophthalmological examination showed proptosis of the left eye as evidence by a widened palpebral fissure and greater than normal difference between the length of both eyes on Hertel's exophthalmometry. There was restriction of extraocular muscle movement on lateral, inferolateral and inferior gaze of the left eye. Visual fields and acuity were normal. A brownish well demarcated flat nevus measuring 1 x 2 mm was noted over the left medial bulbar conjunctiva. Systemic examination was normal except for spider angiomatas noted over the chest. The patient did not have any neurological deficits nor other evidence of melanoma elsewhere.

CT scan of the paranasal sinuses revealed a fairly large soft tissue mass in the left maxillary sinus that has extended to the left nasal cavity, ethmoid, sphenoid and frontal sinuses. The mass likewise extended to the inferior intraorbital compartment. Bone setting showed destruction of the walls of the left maxillary sinus (Figure 2).

A biopsy of the nasal mass was made and the histopathologic result was malignant melanoma. Chest X-ray, CBC, and urinalysis were normal. Liver function tests were likewise normal except for slightly lowered total proteins and decreased A/G ratio (1.8:1).

The patient was then readmitted to the hospital for definitive surgical treatment 3 weeks later. This time, in addition to the previous findings, a 0.5 cm. exophytic black mass was seen at the left soft palate area adjacent to the third molar (Figure 3).

Left subtotal maxillectomy with preservation of the orbital floor was performed (Figures 4 & 5). Intraoperatively, the anterior maxillary region was noted to be hyperemic and bulging. Grossly, the tumor was brown-black and friable, measured 5.5 x 4.0 x 3.0 cm. and occupied the entire maxillary sinus with erosion of



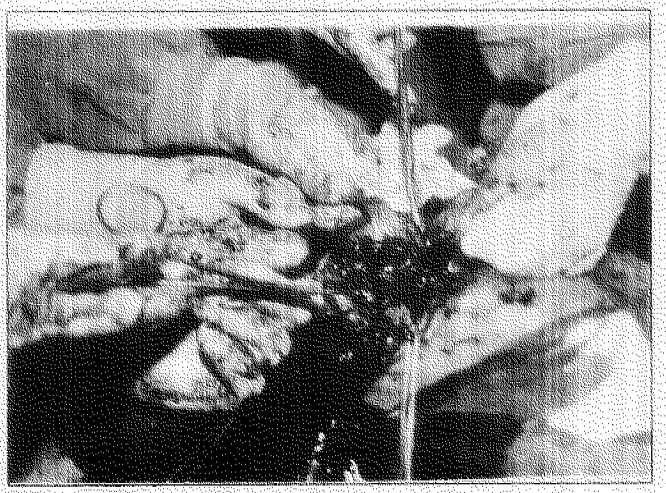
(FIGURE 3) THE 0.5 cm. EXOPHYTIC BLACK MASS OVER THE LEFT PALATAL AREA WHICH APPEARED 2 WEEKS AFTER NASAL BIOPSY.

the anterior and superomedial sinus walls (Figure 6). Induration along the left palate and alveolus was likewise noted.

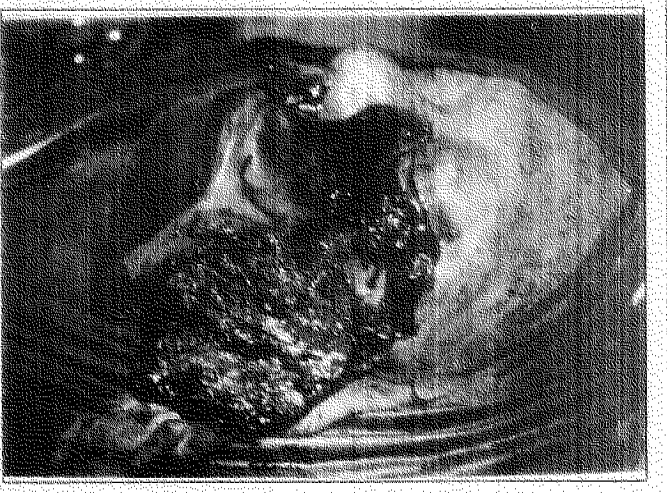
Microsections from the mass showed a poorly differentiated neoplasm consisting of ovoid to spindle shaped cells with large hyperchromatic nuclei and abundant melanin pigment in the cytoplasm. The histopath result was officially signed out as Malignant melanoma of the maxillary sinus (Figure 6).

Post-operatively, the patient had three episodes of passage of coffee-ground material per NGT. An impression of upper gastro-intestinal bleeding secondary to stress ulcer with a high probability of a chronic liver disease, was given upon consultation with GI Medicine. He did well and was discharged on the 9th post-operative day.

The patient underwent cobalt radiation therapy during the succeeding 3 weeks. After a span of two



(FIGURE 4) EXPOSURE OF THE TUMOR VIA WEBER-FERGUSSON INCISION.



(FIGURE 5) LEFT SUBTOTAL MAXILLECTOMY WITH PRESERVATION OF THE ORBITAL FLOOR.

months, local recurrence in the maxillary area was noted. However, there was no apparent evidence of regional and distant metastasis. Five months after the diagnosis, the patient died at Makabali Provincial Hospital from massive hematemesis probably secondary to ruptured esophageal varices brought about by a chronic liver disease. No autopsy was done.

DISCUSSION

Melanomas are malignant tumors originating from melanoblasts, which are cells that are capable of producing melanin pigments. This tumor may occur in the skin or in the mucosa, however, its presentation, treatment and prognosis differ markedly. Almost all cutaneous malignant melanoma arise from a precancerous lesion such as a junctional nevus or lentigo maligna unlike primary malignant melanomas of the aerodigestive mucosa, which nearly always take origin ex novo from the normal mucosa.[7] In primary cutaneous melanoma, size is directly related to prognosis, but in mucosal melanomas, this trend is not obvious. Satellitosis, frequently seen in cutaneous melanomas either present with regional node metastasis or develop nodal metastasis during the course of the disease.[5] Nodal metastasis is infrequent in mucosal melanomas.

Malignant melanoma involving the upper aerodigestive tract may present as either primary or metastatic disease. Although both are uncommon, metastatic involvement of the mucosal surfaces of the head and neck is more rare and has a different site of predilection. Primary mucosal melanoma constitutes 1.7 - 3.0% of all primary melanomas of which 70 percent occur in the nasal cavity. The paranasal sinuses account for majority of the remaining cases. On the other hand, melanoma metastatic to the mucosa of the head and neck has been encountered most often in the larynx, tongue, and tonsil, sites rarely involved by primary melanoma.[2]

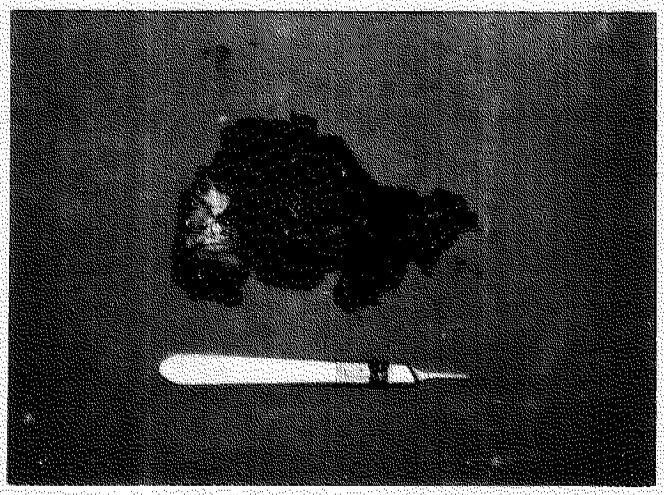
Clinical features. Age and Sex. Male preponderance is more striking at a 2:1 ratio. Approximately 73% of the patients are over 50 years of age with approximately 53.7% having onset between 50 to 69 years.[3] Youngest and oldest age reported were 17 and 84 respectively.[8]

Symptomatology. Symptoms are generally not seen until the tumor is advanced. The first and most prevalent symptom is nasal obstruction which is usually unilateral. The second most common symptom is epistaxis which is either abundant or in the form of nasal drip. In cases in which the tumor originated in the sinuses, nasal obstruction is not the present symptom. Instead, these patients usually experience epistaxis and pain felt as a sensation of pressure, blockage or paresthesia over the region of the involved sinus. Other less common findings are facial, nasal and palatal swelling, proptosis, tearing, purulent nasal discharge, chronic sinusitis and headache.[4,8] The duration of symptoms is quite variable. Majority of patients place the onset of their symptoms between 3 and 24 months with only a few having dated it between 1 week or 2 months.

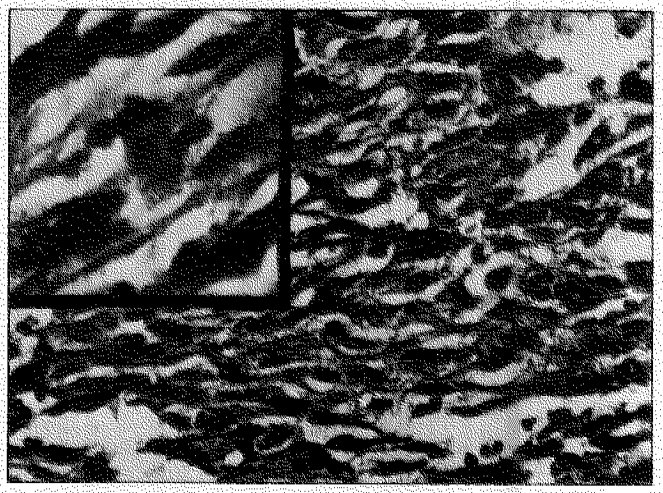
Sites of Lesion. The most common site of origin is the nasal septum, closely followed by the lateral wall, middle turbinate, inferior turbinate, and then the maxillary and ethmoid sinuses.[3,4,5,8]

Gross Pathologic Features. Mucosal malignant melanomas are usually non-encapsulated solid, polypoid, bleeding masses of varying color from white to pick, red gray, brown or black. The consistency can be described as firm, friable necrotic, granular, gelatinous or cystic.[8]

Histopathologic Features. Malignant melanomas



(FIGURE 6) GROSS APPEARANCE OF THE SPECIMEN.



(FIGURE 7) MICROSECTION (H AND E) OF THE MASS DEMONSTRATING THE OVOID AND SPINDLE SHAPED CELLS WITH HYPERCHROMATIC NUCLEI AND ABUNDANT MELANIN PIGMENT IN THE CYTOPLASM. INSET SHOWS SPECIMEN AT HIGHER MAGNIFICATION.

of the mucous membrane of the nasal fossa and paranasal sinuses do not differ much basically in thier histologic appearance from the melanomas arising from other parts of the body. On the other hand, like melanomas of the upper respiratory tract in general, they tend to show much greater pleomorphism and resemble a spindle-cell sarcoma rather than a carcinoma. For this reason, it would appear, most of them have been called "melanotic sarcomas".[9] True primary melanomas in these locations should show junctional changes in the overlying or adjacent lateral mucosa, or both.[2] In contrast, a secondary or metastatic melanoma must have both intact overlying mucosa and adjacent mucosa devoid of junctional changes. However, it is not always possible to be certain of jucntional activity in a largely ulcerated advanced tumor as in this particular case. In recent report, **Henderson and Robbins**[2] have stated that the frequency with which junctional change is noted depends on the extent of the ulceration, the amount of tissue submitted for pathologic examination and the number of sections examined. An ulcerated mucosal melanoma without demonstrable junctional activity may be considered primary if the following conditions are met: (a) it is the dominant lesion (b) it has local or regional metastasis consistent with the purported primary site; and (c) there is no history of primary cutaneous or ocular melanoma or of a nevus that underwent spontaneous regression.[2,10] In the case presented, although no junctional activity was indentified, gross and microscopic features were highly characteristic of malignant melanoma. The lesion was mainly concentrated in the left maxillary sinus and there was no other demonstrable primary site.

Malignant melanoma may histologically mimic many types of tumor such as fibrosarcoma and anaplastic carcinoma.[11] This may lead to a delay in arriving at a correct diagnosis especially if the tumor appears to lack pigment, which occurs in about 1/2 of the cases.[12] Therefore, absence of melanin is not a reliable basis for excluding the diagnosis of mucosal melanoma. Situations may be further complicated by the presence of hemorrhage and occurence of brown pigment (hemosiderin) in these tumors. In these instances, special staining techniques may be necessary to help in supporting or refuting the impressions of the pigment's nature. Melanin is characterized by (a) a positive staining with Fontana silver stain; (b) bleaching by permanganate oxalate, and (c) negative staining with Prussian blue, which is a special stain for hemosiderin.[1] None of these special techniques were necessary in this case since melanin was readily demonstrable in abundant amounts in the cytoplasm of the malignant cells, allowing the autho to arrive at the correct histopathologic diagnosis promptly.

Prognosis. Malignant melanomas of the nasal cavity and paranasal sinuses have an unpredictable course and a rather poor prognosis. Patients may die in 4 months of widespread disseminated disease or in 20 years with long periods of dormancy in the disease process. Lund estimated that about 30% will probably be alive at 5 years when regional or systemic metastasis are not found. This is much worse than the outlook in cutanoeus malignant melanoma where 77% can expect to be alive at 5 years.[13] Others report a 35.6 - 46.2% and 22.0 - 30.9% rates at three and five years respectively.[4,5]

Several different factors have been mentioned to account for the poor survival of patients with mucosal melanoma especially in comparison with their cutaneous counterpart. Cutaneous lesions are easily seen while those involving the romucous membranes are hidden from view and are inaccessible on routine examination,allowing them time for deeper penetration

and metastasis. Patients also present quite late due to fairly innocuous symptoms. The condition may be confused by previous benign polyps causing patient to be unalarmed by recurrence of similar symptoms. All of these in effect cause delay in treatment. In addition, mucosal melanoma has a more histologically aggressive bahavior, being more anaplastic, pleomorphic and angioinvasive. They are more liable to infection and ulceration than those in the skin. Lastly, most of the time, total surgical excision is technically difficult and rarely possible of the compressed and intricate surgical anatomy.[4,12,13]

Prognosis was found to be better in patients with melanomas arising from the nose than those arising from the sinuses. In cases of mucosal melanoma, review of literature failed to show any prognostic correlation with age, sex duration of symptoms, tumor cell type, tumor size, pigmentation and mitotic index.[4] However, a trend is observed with increasing depth of invasion and decrease of survival.[5]

Freedman plotted a survival graph showing a straight line relationship between the risk of death and time. Unlike other conditions, the risk of death does not increase precipituously, indicating a constant risk of death from melanoma no matter how long the patient lives after treatment.[4,13]

Treatment. Radical surgery remains to be the treatment of choice, and the resection should be as wide as possible. It is, however, difficult in this anatomical location to obtain the margins of 2 to 4 cm grossly healthy tissue recommended in the management of melanomas in the skin.[14] Exenteration of the orbit is not likely to be justified in patients with vision since the fate of the patient lies partly in adequate local resection, and partly influenced several immunological factors. Patients treated surgically were found to have a 3 year survival rate of 75% and a 5 year survival rate of 61.3%.[13]

The role of elective cervical node dissection has been recently much debated on but this problem remains open. In fact, some authors advocate this surgical procedure for staging reasons while others argue that non-involved nodes could be a further barrier to metastatic spread. Moreover, a low incidence of subsequent development of lymph node metastasis in the absence of local or distant metastasis was reported, and for this reason, prophylactic neck dissection was not recommended.[15]

According to many authors, melanoma is a radioresistant tumor. However, studies of experimental melanoma of cell lines, and culture suggest that melanoma cell may not be radioresistant per se. In fact, this malignant cell may have a high capacity to repair sublethal radiation damage and so, to achieve a tumoricidal dose, it is necessary to give a larger dose per fraction radiotherapy.[7] In a review of literature, Harwood and Cummings noted that 72% of those patients who received radiation therapy had a complete local remission. Those patients in whom the radiation technique was used involved a large dose per fraction (greater than 4 Gy per treatment) and had an 86% rate of local control at 18 months.[6]

Regarding chemotherapy, the most effective single agent has been Decarbazine, that, in Siegler's experience, used in combination with Vincristine, Lomustine, and Bleomycin showed a 10% complete remission rate and a greater than 50% remission in 30% of patients with metastatic melanoma.[7]

Even if still in the experimental stage, recent data on adoptive immunotherapy of malignant melanomas appear quite interesting. Modulating the immune response with interferon, BCG therapy, and monoclonal antibodies has led to a regression of some melanoma tumors.[7] In 1975, **Mastrangelo** et al, reported the use of intralesional BCG in producing regression of both local and metastatic melanomata. In 1982, **Saito** et al, introduced oral BCG immunotherapy combined with other treatments. Results showed that these treatments were effective in extending patient's survival time and that metastasis to the lung was more responsive than abdominal metastasis.[16] Similarly, while **Shah** et al. noted a conspicuous abscence of peripheral lymphoid reaction to the tumor in histologic sections of mucosal melanomas, **Barton**reported increased lymphocyte activity in patients who had prolonged survival after treatment with cryosurgery. He speculated that creating such a reaction by cryotherapy may have played an immunoprotective role. This variability in the host immune response may be responsible for the unpredictable nature of mucosal melanomas.[6]

Recurrence and metastasis. Malignant melanomas of the nasal cavity and sinuses are characterized by early and repeated recurences. They come in the form of local recurrence (48.2%), cervical node depositis (29.2%) and distant metastasis (14.3%) at any time after their first initial treatment. With 67% occurring in the first year and 90% by five years. Only 8.9% of patients show no recurrence. It is therefore evident that regardless of the mode of treatment, the major factor in failure of treatment of mucosal melanoma is local recurrence. **Freedman** states that it is more important to search carefully for satellite areas, second primaries or mucosal metastasis. In other words, the surgeon must be aware that the disease often is expressed as multicentric mucosal involvement and within reason, the more mucosa that is removed, the less chance there will be of recurrence.[4] Adequate repeated excision of local recurrences can offer better survival prognosis.

Widespread regional node metastasis is not a com-

mon feature of nasal and paranasal sinus melanoma. Furthermore, according to the experience of Blatchford et al., the presence of regional lymphatic involvement in mucosal melanomas of the head and neck does not seem to affect the prognosis.[17]

Distant metastasis occur in the lungs, brain, adrenal gland, liver and skin, in descending order of frequency. In some cases of extensive hematogenous dissemination, the melanogens are excreted in the urine as colorless soluble substance which on standing, are oxidized to a dark melanin precipitate.[1]

Deaths. Distant metastasis to liver, lungs and brain are the most frequent cause of death.[7] A small percentage die of disease entities.

The patient's immune sytem is believed to play a role in doubling the survival and preventing the risk of developemnt of metastasis.[7] Those whose immunological control is so disturbed and depressed by any interference are believed to be unsalvageable. It appears that the host-tumor balance is crucial to the natural history of the disease and explains the variable pattern of behaviour in this tumor.

CONCLUSION

Primary malignant melanoma is not exclusively seen in the skin. It also occurs in the paranasal sinuses. Such a disease entity should be considered as a differential diagnosis to the more commonly occurring tumors in these anatomical areas, whose clinical characteristics show great similarities with it. Accurate diagnosis may be arrived at only through proper histologic identification of melanin pigment and junctional activity. Total surgical excision remains to be the most favourable treatment modality, whereas radiation, chemotherapy and immunotherapy currently have mainly palliative roles. Prognosis is generally poor. However, advances in immunology have established a possible effect of the host's immunologic status on tumor control and surival. It was very unfortunate that the patient died from another disease entity. However, tempting as it may be, short of an autopsy, together with a strong clinical evidence of prolonged liver dysfunction, his death could be attributed to a possible distant metastasis but was most probably secondary to decreased immunocompetence and chronic hepatic disease.

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CHRONIC SUPPURATIVE TYMPANO-MASTOIDITIS WITH CHOLESTEATOMA AND SUBPERIOSTEAL ABSCESS: ITS PATHOGENESIS AND OTHER ASSOCIATED COMPLICATIONS (A SERIES OF 100 CASES)

Manuel G. Lim, MD, MSc, FPCS*
Frederick O. Leh, MD**
Manuel Anthony C. Lim, Jr., MD**
Nelson G. Magno, MD**

ABSTRACT

This study has two aims, namely: to show the pathogenesis of the different forms of subperiosteal abscess in chronic supppurative tympano-mastoiditis with cholesteatoma and to reveal other associated complications which are life-threatening to these patients.

Although this disease entity is virtually unheard of in the developed countries, it is encountered almost daily in clinical practice in Third World nations like the Philippines. Most of the patients suffering from this illness come from the lowest income brackets. Malnutrition and ignorance contribute to the chronicity and seriousness of the disease.

From July 1990 to March 1992, 100 cases of chronic suppurative tympano-mastoiditis with subperiosteal abscess (CTM-SPA) were admitted as emergency cases in the Philippine General Hospital and operated on as soon as possible. The various findings in the study of these cases led to a significant conclusion. In the past, it was believed that the destruction of the mastoid cortex was due to pressure necrosis or to enzymes from the enlarged cholesteatoma. However, in seven percent (7%) of the cases studied, only granulation tissues were found. This led to conclusion that, because of the blockage of the aditus ad antrum by either cholesteatoma or granulation tissues, osteomyelitis resulted. This produced the cortical defect of the post-auricular region, intracanal area or mastoid tip.

INTRODUCTION:

In medical literature, chronic suppurative tympanomastoiditis with cholesteatoma and subperiosteal abscess or Bezold's abscess is very seldom reported. Acute mastoiditis with subperiosteal abscess is seen only occasionally in the developed countries. However, in the Philippines and in other developing countries, chronic suppurative tympanomastoiditis is seen daily in the otolaryngologic clinic. **Ibekwe** and **Okaye** (1988) reported 16 cases over a five-year period in Nigeria. **Shenoy** (1987) and **Lee** (1991) reported a high incidence of chronic suppurative otitis media with cholesteatoma in India and Asia.

Acute suppurative otitis media is seldom seen in the clinic. Because of the poor socio-economic standing, these patients do not come for consultation during this stage of the disease. It is only when the disease process has progressed and complications have set in that they will come for treatment.

From July 1990 to March 1992 (a period of 21 months), one hundred (100) cases of chronic suppurative tympano-mastoiditis with cholesteatoma and subperiosteal abscess or Bezold's abscess were admitted to the University of the Philippines - Philippine General Hospital, and exploratory mastoidectomies were performed soon after admission.

OBJECTIVES:

The aims of this study are:

1. To show the probable pathogenesis of subperiosteal abscess, either post-auricular, intracanal or mastoid tip (Bezold's abscess).
2. To describe the clinical and surgical findings in these cases.
3. To reveal the frequency of the associated complications of the disease entity.

MATERIALS AND METHODS

From July 1990 to March 1992, one hundred (100) cases of chronic suppurative tympano-mastoiditis with cholesteatoma and subperiosteal abscess or Bezold's abscess were admitted, and exploratory mastoidectomies were performed soon after admission. Most of the patients were operated on as emergency procedures, without the benefit of radiographic examinations and audiologic studies. The x-ray study

* Chairman, Department of Otolaryngology
Philippine General Hospital
University of the Philippines
** Residents, Dept. ENT, UP-PGH

becomes only an academic procedure and is not a real necessity.

During the operation, pieces of cortical bone with granulation tissues from the edges of the cortical breaks were taken for histo-pathological examinantion. The antral cavities enlarged by cholesteatoma were measured accurately with a caliper. A CSTM-SPA form was completed after each surgery to provide all the necessary information on the ear condition before and during operation. These data comprise the results of the study.

RESULTS

The clinical features and surgical findings of one hundred (100) cases of chronic suppurative tympanomastoiditis with cholesteatoma and subperiosteal abscess and other associated complications are summarized in Table I.

Chronic Suppurative Otitis Media

All one hundred (100) cases presented had chronic suppurative otitis media or tympano-mastoiditis. At the time of examination, the discharge varied from viscid mucopurulent foul-smelling to viscid purulent foul-smelling in character. All patients in this series manifested total or near total perforation of the eardrums together with the huge polyps or thick polypoid mucosa partly covered or lined with squamous epithelium and a great deal of epithelial debris in the attic region hanging down to the mesotympanum. The foul-smelling discharge was obnoxious, an indication of severe necrosis of tissues.

Age

The youngest patients was an eleven month old baby. The oldest was 35 years old. The majority of patients were found between the ages of six (6) to fifteen (15), and they made up fifty-seven percent (57%) of the series (see Table II).

Sex

The incidence of sex distribution was very insignificant. There were forty-nine (49) male patients and fifty-one (51) female patients included in this study.

Duration of Disease

All cases had a history of chronic discharging ear/s varying from six (6) months' to 31 years' duration (depending upon the age of the patient) before the onset of the subperiosteal abscess or fistula.

Subperiosteal Abscess

Sixty (60) out of one hundred (100) cases had post-auricular subperiosteal abscess. Of these, 7 cases had both post-auricular subperiosteal abscess and intracanal subperiosteal abscess;

16 cases had intracanal subperiosteal abscess as manifested by the marked sagging of the postero-superior canal wall with tenderness; 4 cases had Bezold's abscess and during the exploratory mastoidectomies, the mastoid tips of these four cases were necrotic with granulation tissues.

Twenty-seven cases had post-auricular fistulas most of which had persistent foul discharge with granulation tissues around the fistulas.

Cortical Breaks

During the exploratory mastoidectomies, the cortical breaks were found behind the spine of Henle on the cribiform area in varying sizes, from 0.5 to 2.8 cm. in diameter, with plenty of granulation tissues in and around these cortical breaks. These were found among the post-auricular subperiosteal abscesses and post-auricular fistulas. In intracanal subperiosteal abscesses, the cortical breaks were found always in the postero-superior bony canal wall in varying sizes. Some of the defects measured almost 2 cm. in length along the canal wall.

Cholesteatoma in Antrum and Attic

Ninety-three percent (93%) of the patients in this series had cholesteatoma in the antrum. The explored antral cavities varied in sizes, in terms of cubic centimeters. The smallest antral cavity was 0.6 c.c. while the largest was 20.6 c.c., and the average was 7.71 c.c. (see Table III). Total or near-total perforation of the eardrums were seen in all. When the matrix of cholesteatoma was peeled off from the antral cavity, very often granulation tissues were seen outside of the matrix attached to the mastoid bone.

Seven cases (7%) did not have cholesteatoma in the antral cavities; of these, four (4) has slightly enlarged antral cavities; and three (3) cases had no enlarged antral cavities. All seven (7) cases had only granulation tissues in the antrum. Two (2) cases had only granulation tissues in the attic, and (5) cases had cholesteatoma in the attic.

Ninety - eight (98) cases had cholesteatoma in the attic. Most had granulation tissues also in the attic together with cholesteatoma. Only 28 had only cholesteatoma without granulation tissues in the attic.

Findings in the Middle Ear

The surgical findings in the middle ear among the one hundred (100) cases were more variable. Many of them had plenty of granulation tissues scattered all around the middle ear, particularly over the region of the footplate, which was always fully covered. Between the

granulation tissues, the middle ear spaces were lined with migratory squamous epithelium. In others, polypoid mucosa or aural polyp with areas of squamous epithelium lining the medial wall of the middle ear were seen. In all cases, most of the ossicles were destroyed, and, if any were left, were too necrotic to be used. Cholesteatoma was present in fifty-four cases in the middle ear.

Other Complications Associated with Subperiosteal Abscess

Aside from the various forms of subperiosteal abscess, many developed other complications (see Table IV).

Table IV. Other Complications of Chronic Suppurative Tympano-Mastoiditis with Subperiosteal Abscess

1. Erosion of the facial canal without paralysis	28 cases
2. Erosion of the facial canal with paralysis	6 cases
3. Erosion of tegmen with granulation tissues on dura	2 cases
4. Erosion of tegmen with epidural abscess	6 cases
5. Erosion of lateral sinus plate with granulation tissues on lateral sinus wall	4 cases
6. Erosion of lateral sinus plae with peri-sinus abscess	2 cases
7. Erosion of lateral sinus plate with lateral sinus thrombophlebitis	2 cases
8. Suppuratiive meningitis	2 cases
9. Brain abscess	2 cases
10. Fistula to horizontal semicircular canal	1 case

DISCUSSION

Chronic suppurative tympano-mastoiditis with cholesteatoma and subperiosteal abscess, which at times may be associated with other serious complications, is frequently seen in the otolaryngologic clinic in the Philippines. This can be attributed to two main factors: the poor socio-economic standing of these patients, and these patients are largely ignorant of the disease process. Very often the condition becomes very serious due to the failure to bring the patient to the physician in the early stage of the disease.

Among all the subperiosteal abscesses of the post-auricular region (60%), or the postero-superior intracanal areas (16%), the post-auricular fistula (27%) and the mastoid tip or Bezold's abscess (4%), choolesteatoma was found in the mastoid antrum in ninety-three percent (93%) of the cases and granulation tissues were found in the antrum in seven percent (7%) of the cases. Cholesteatoma was found in the attic and aditus ad antrum in ninety-eight percent (98%) of these cases; only two percent (2%) had granulation tissues in the attic and aditus ad antrum. Only three percent (3%) of the cases had no enlarged antrum; the rest had huge enlarged antrum. The volume in cubic centimeters is arrived at by measuring the widest and the deepest diameter in millimeters, but this is definitely not the actual figure, because the surfaces and contours of the mastoid antrum are very irregular; this is just an approximate measurement for purposes of comparison (see Table III).

The fact that granulation tissues only were found in the antrum in seven percent (7%) without cholesteatoma means that pressure necrosis or enzymatic action of cholesteatoma is not the main cause of the cortical breaks in the formation of subperiosteal abscesses of any form. It is believed that the presence of granulation tissues and/or cholesteatoma in the aditus ad antrum and attic prevents the trapped pus in the antral cavities from draining. This may lead to osteomyelitis of the cortical bone or the mastoid tip, producing the cortical breaks in the post-auricular region, in the supero-posterior intracanal area, or in the mastoid tip. This was demonstrated and confirmed by the histo-pathological findings of chronic osteomyelitis from the removed bone edges of the cortical bone defects. However, it is difficult to resolve the problem of the pathogenesis of subperiosteal abscess. Perhaps it will be safe to say that a combination of factors is responsible for subperiosteal absccess.

All cases had total or near total perforation of the eardrums, mostly of the pars tensa with cholesteatoma of various degrees in the attic (98%). Lee (1991) and Raffin (1988) state that cholesteatoma occuring in children is mostly of the attic retraction pocket type. Shenoy's study (1987) confirms the findings of the present paper namely that the migration type of cholesteatoma is more common. The latter type is more aggressive than the attic retraction pocket type.

In this study, the most common complication found among the one hundred (100) cases aside from the suubperiosteal abscess is the erosion of the horizontal facial canal by granulation tissues without facial paralysis. This is true for twenty eight percent (28%) (see Table IV). In six (6) cases, erosion of the horizontal or vertical (mastoid) canal was associated with facial paralysis.

TABLE II

AGE DISTRIBUTION OF PATIENTS

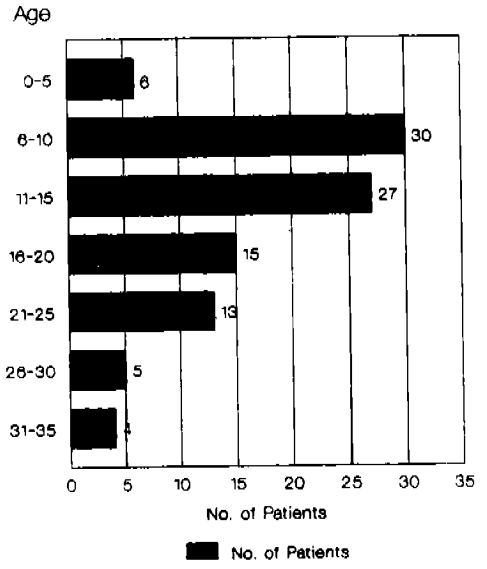
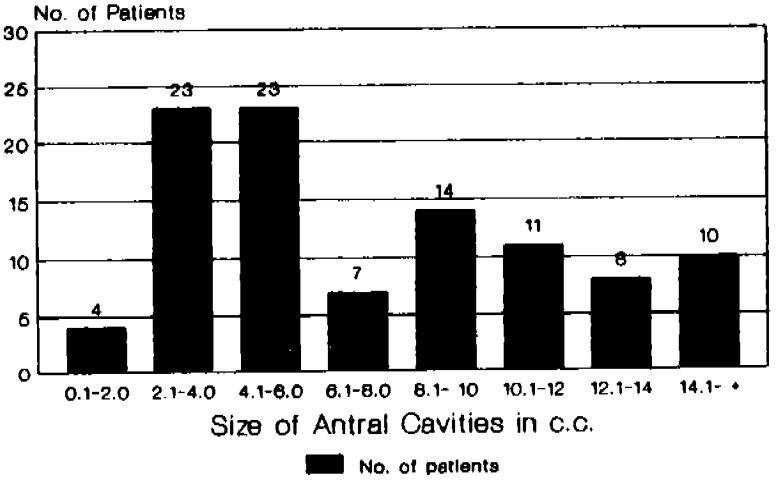


TABLE III

SIZE OF ENLARGED ANTRAL CAVITIES



Ave. size in c.c=7.71;No.of patients=100

Twelve (12) cases had erosion of the tegmen with granulation tissues on the dura but without epidural abscess or other intracranial complications. Six (6) cases developed erosion of the tegmen with epidural abscess. Erosion of the lateral sinus plate with thrombophlebitis was seen in two (2) cases. Two (2) cases manifested erosion of the lateral sinus plate with perisinus abscess. Fistula to the horizontal semicircular canal was seen in only one (1) case. Suppurative meningitis was seen seen in two (2) cases. Two (2) cases had brain (cerebellar) abscess; one had erosion of the lateral sinus plate with perisinus abscess and granulation tissues but without lateral sinus thrombophlebitis; the other developed cerebellar abscess without erosion or the lateral sinus plate.

All cases underwent exploratory mastoidectomies soon after admission. Because of the extensive destruction of the middle ears and the severe associated complications, and of the poor socio-economic circumstances of these patients, which made the follow-up treatment very difficult and uncertain, classical radical mastoidectomies were performed on all to ensure dry and safe ears after surgery. (.pa)

SUMMARY

A series consisting of one hundred (100) cases of chronic suppurative tympano-mastoiditis with cholesteatoma, post-auricular subperiosteal abscess, subperiosteal abscess of the postero-superior canal wall and/or Bezold's abscess was reported. Huge cholesteatoma was found in ninety-three (93) enlarged antral cavities, and granulation tissues were found in the other seven (7) antral cavities. Analysis leads one to believe that the cortical breaks are secondary to the infection of the cortical bone (osteomyelitis). Many other associated complications were also observed. Because of severe destruction by the disease and because of the poor socio-economic standing of these patient, radical mastoidectomy was performed in all cases.

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UNILATERAL UPPER LATERAL NECK MASS A DIAGNOSTIC AND THERAPEUTIC CHALLENGE*

NELSON G. MAGNO, M.D.**
ALBERT R. TIOTUYCO, M.D.**
RONALDO A. REYES, M.D.**
JOSE L. MONTILLA, III, M.D.**

ABSTRACT

A large, fixed, ulcerated, 7 x 6 x 4 cm mass of 2 years duration at the right upper lateral neck area was seen in a 40 year old female. The mass was suspected to be a parotid tumor. Repeated biopsies of the mass showed undifferentiated carcinoma, metastatic. Repeated Nasopharyngeal biopsies only revealed chronic inflammation. Panendoscopy revealed no definitive mass to identify a primary. The patient underwent wide excision and radical neck dissection on the right with a pectoralis major myocutaneous flap reconstruction. Post-operative histopathologic results showed a mucoepidermoid carcinoma of the parotid.

This case illustrates that tumors located at the periauricular area can confuse the clinician preoperatively, especially if a lot of consideration is given to the histopathology. Furthermore, the parotid gland should be the prime suspect in cases of tumors arising from this area, whether or not histopath reports are consistent with the diagnosis. Management of such cases is usually surgical, the other option being radiation therapy.

INTRODUCTION

There are times when an Otorhinolaryngologist Head and Neck Surgeon is challenged in the diagnosis and management of a unilateral lesion in the upper lateral neck area. This is particularly true because of the many diagnostic possibilities in that area. One can consider a parotid tumor, NPCA with metastasis or a metastatic lymph node with another primary. Following is a case presentation of a unilateral upper lateral neck mass which served a diagnostic and therapeutic challenge.

CASE REPORT:

CB, 40 year old, female, married, from Ilocos Sur was admitted for the first time on March 14, 1992 for an ulcerating right infra-auricular mass. Her history started 2 years prior to admission when she noted a 2 x 2 cm right infra-auricular mass. The mass was non-tender, movable, and progressively enlarging. She consulted a physician who prescribed her several unrecalled antibiotics and antiflammatory agents. Despite adequate compliance, the mass progressively grew in size.

Five months prior to admission the mass became erythematous, formed a small vesicle which later exuded serous discharge. She consulted at a provincial hospital where she was given cloxacillin and was advised to undergo surgery.

The patient was initially seen at the OPD with a 7 x 6 x 4 cm hard, fixed, upper, lateral neck mass. The overlying skin had a 1 x 2 cm ulceration. There were no other accompanying signs and symptoms (See Fig. 1). Thorough ENT examination revealed no other masses. Tentative diagnosis was right upper lateral neck mass, probably parotid malignancy with skin involvement.

A punch biopsy of the ulcerated area revealed metastatic undifferentiated carcinoma (See Fig. 2). Guided with this result, an initial attempt to identify the primary site of the carcinoma was sought. A four quadrant nasopharyngeal biopsy under local anesthesia was done three times consecutively, all revealing chronic inflammation. Thinking that the mass could be primarily parotid, a wedge biopsy with deeper bites was done. The result still showed metastatic undifferentiated carcinoma. The patient was subsequently admitted with a preoperative impression of right upper lateral neck mass, metastatic undifferentiated carcinoma (by biopsy), primary unknown.

The patient underwent panendoscopy under general anesthesia which did not reveal any definitive mass in the larynx, nasopharynx, oropharynx, esophagus and bronchi. Surgery proceeded with total parotidectomy, facial nerve sacrifice, radical neck dissection on the right, pectoralis major myocutaneous

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* Presented at the PSO-HNS Interesting Case Contest held at Hotel Nikko, Manila Garden, July 9, 1992.
** Residents, Dept. of ENT, UP-PGH.

flap reconstruction and static sling procedure.

The postoperative course was uneventful. Final histopathologic report was read as mucoepidermoid carcinoma, high grade, parotid gland. Positive for tumor: all 10 superior cervical lymph nodes, biopsy specimens from panendoscopy. (See Fig. 3)

The patient was subsequently discharged and is currently on regular follow-up showing no evidence of recurrence. She will undergo postoperative radiotherapy.

DISCUSSION:

Patients presenting with an isolated cervical mass pose a diagnostic and therapeutic challenge to the otolaryngologist-head and neck surgeon. Table I shows the variety in the differential diagnoses of such neck masses.

The case of a 40 year old female with a 7 x 6 x 4 cm right upper lateral neck mass was presented to illustrate the diagnostic dilemmas and options that can be encountered. Management of the adult patient with a persistent, unilateral cervical swelling should be based on the precept that malignancy is the most probable diagnosis until definite histopathological evidence to the contrary is obtained (Martin and Romieu, 1952; Shaw, 1976). After having established this through biopsy reports, the next question is whether to consider the tumor a primary malignancy or a metastatic lesion.

Majority of tumors in the parotid gland, (about 80%) arise from the superficial and caudal parts of the gland. Most frequently the presenting sign is a mass in the periauricular area. It is good clinical practice to consider every "lump" or tumor thus localized, as a probable tumor of the parotid gland. (Batsakis, 1979)

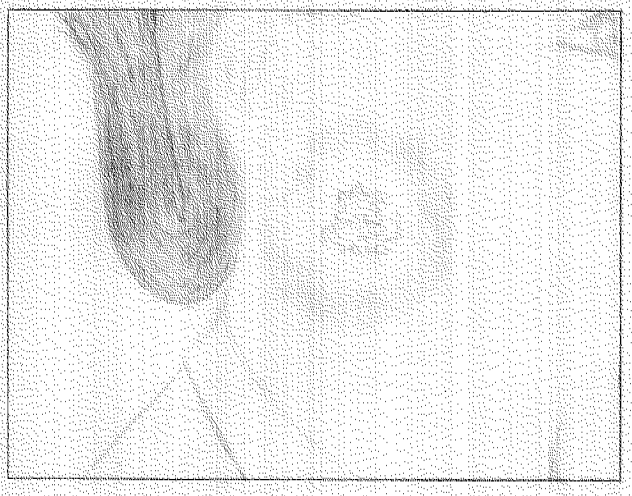

Figure 1: The patient with an ulcerated right upper lateral neck tumor.

In a study done on a series of 288 patients with a mass in the upper lateral neck by Solem, Schroder and Mair (1981), 109 were neoplasms; 62 (52%) were located within the parotid gland, 8 out of 62 were malignant. However, out of 109 neoplasms, 10 were cases of lymphoma, 7 were non-parotid benign tumors and 30 were metastatic lesions.

Considering the location of the mass, therefore, an investigator should think of 3 possibilities. The parotid gland as a primary source of the tumor, a lymphoma and a metastatic lymph node.

Aspiration biopsy may play an important role in the initial investigation of patients with cervical masses especially in the establishment of the diagnosis of malignancy (Shaha, Webber and Marfi, 1986). This was not done in the case presented because the cervical mass was already ulcerated and the possibility of seeding into the skin is moot. A direct punch biopsy via the ulceration showed metastatic undifferentiated carcinoma. With this report, the clinicians thought of a metastatic lesion as the cause of the tumor and decided to investigate along this line. A thorough and repeatedly done physical examination of the head and neck with visualization and palpation revealed no other mass. Routine chest x-ray was essentially normal. Skull AP-L, Mandible AP-O were unremarkable except for the neck mass.

Miller in 1980 mentioned that there is often a palpable upper neck node in cases of nasopharyngeal carcinoma even if no tumor is seen at the time of the diagnostic operative procedure. Two thirds of patients present with an upper neck mass at the first office visit.

Four quadrant nasopharyngeal biopsy was done three consecutive times, one of which was guided with a fiber optic scope. All specimens however, were reported as chronic inflammation. Because a primary for a metastatic lesion could not be found, the clinicians again considered the possibility of a parotid malignancy. A wedge biopsy of the tumor was performed but the result was the same as the previous one: undifferentiated carcinoma, metastatic. The clinicians were then faced with the problem of giving a lot of weight to the biopsy reports and consider this case as a metastatic CA with an unknown primary or to go with a clinical diagnosis of parotid malignancy.

Ideally, other diagnostic work up could be done in this case to settle the issue. CT Scan, sialography, panendoscopy are procedures that may be of great help in order to make a more definitive diagnosis. However, these ancillary procedures are supposed to complement rather than be the sole basics for a diagnosis. The clinicians acumen and experience should still be the best guide in such decisions.

More important than trying to establish a diagnosis is to be able to offer a logical form of management based on the available information and situation. After all, the

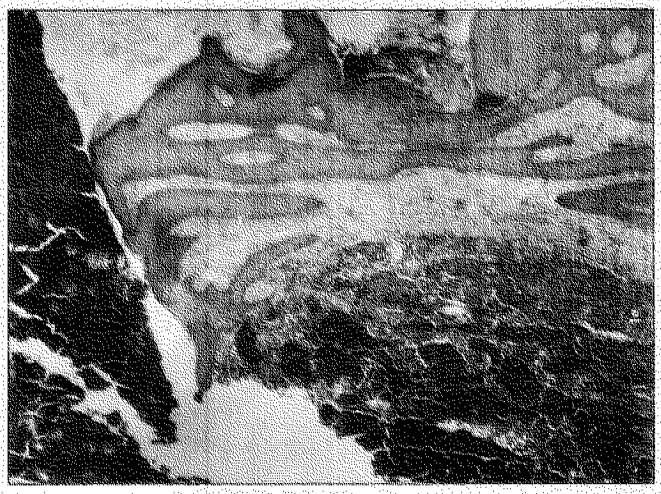

Figure 2a: Overlying epidermis is benign with acanthosis and hyperkeratosis.

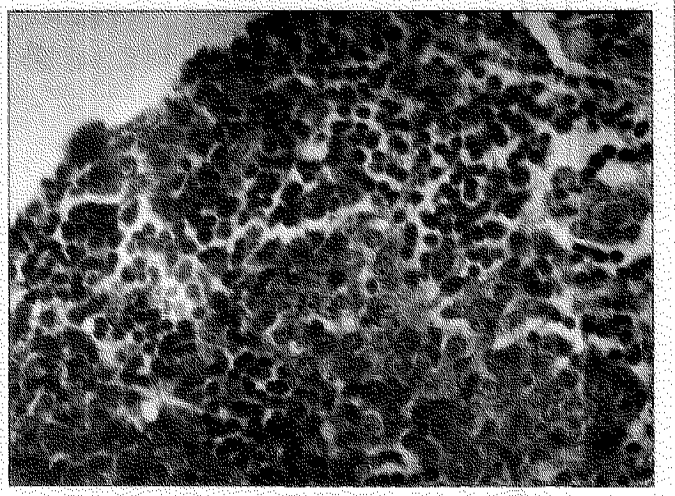

Figure 2b: Subepidermis, several islands and nest of cell with poorly defined cytoplasmic borders, scanty to moderate amount of eosinophilic cytoplasm with vesical nuclei.

essence of the medical profession is to be able to provide a more tolerable way of life for patients.

Management options for diagnostic dilemmas have been written by a lot of renowned physicians.

For patients with metastatic carcinoma in the head and neck with no detectable primary site, surgical treatment is most effective (Batsakis, 1979). Radical surgical excision appears justified when the metastasis is a squamous cell carcinoma especially when the lymph node involved from the occult primary is high in the neck.

Bataini, et al. in 1987, proposed postoperative irradiation to the entire neck and presumed site of occult primary. The control rate in the neck of patients so treated is about 85 % in 3 years and an overall survival of 65% and 55% in 3 and 5 years.

For many years, surgery has been the primary modality of therapy for malignancies of the parotid gland, combined with neck node dissection (Witten, Hybert and Hansen, 1990).

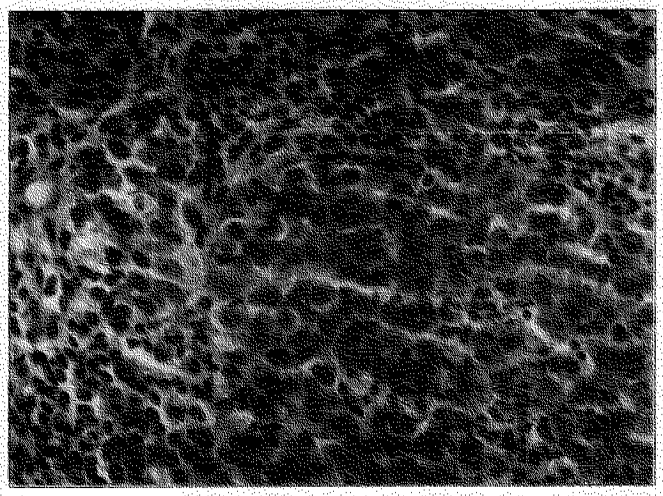

Figure 2c: Many mitotic are seen. Cells do not show any evidence of differentiation.

Therefore, whether the mass is metastaticanode or primarily a parotid lesion most experts agree that surgery is indicated. Supplementation with post operative irradiation is likewise recommended depending on the histopathologic report.

Considering the management options for this patient, surgery was planned. The possibility of a metastatic lesion was, however, still entertained so panendoscopy, as a diagnostic procedure was performed prior to the actual surgery. However, no definite mass was seen in the larynx, nasopharynx, oropharynx, hypopharynx, esophagus and bronchus. Punch biopsy done on the nasopharynx, (R) tonsil, and (R) base of tongue revealed chronic inflammation (results obtained with the final surgical pathology).

Surgical intervention consisted of a radical neck dissection with wide excision of the upper lateral neck mass. Intraoperatively the mass was noted to originate from the parotid gland.

Total parotidectomy with facial nerve sacrifice was done. The neck tumor was removed en bloc with the RND specimen. The mass measured 6 x 8 cm with a portion of infrauricular skin undergoing ulceration. The skin defect was reconstructed with a pectoralis major myocutaneous flap. A static sling was applied using nylon sutures (Fig. 4).

The final histophatologic result revealed mucoepidermoid carcinoma, high grade, parotid gland.

Positive for tumor: specimen labelled posterior triangle node

Negative for tumor: all 10 cervical lymph nodes specimens labelled nasopharynx, tonsil, base of tongue

In retrospect, therefore, it is wise to consider a periauricular mass as a parotid tumor and work to-

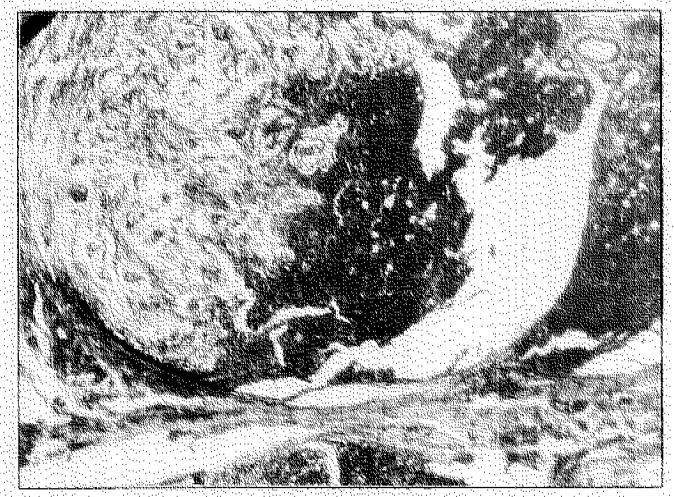

Figure 3a: This section shows normal parotid tissue adjacent to tumor

Figure 3b: Higher magnification shows cells with round to oval basophilic nuclei.

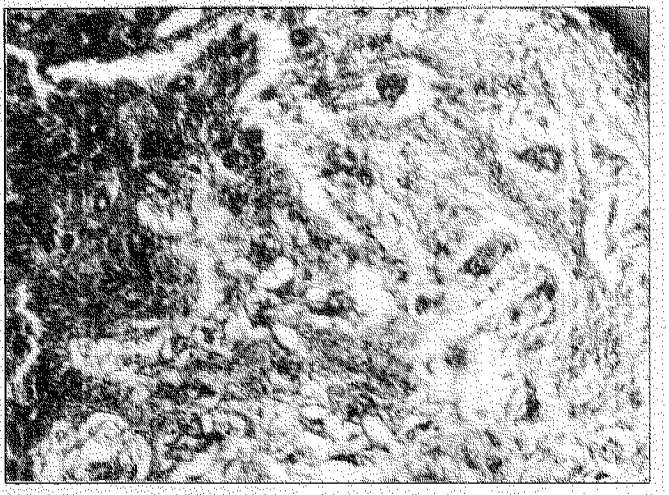

Figure 3c: This section at the border of the tumor shows nest of indifferentiated tumor cells infiltrating the parotid gland.

Figure 3d: This section shows intervening bands of fibrocollagenous tissue between the nests of tumor cells.

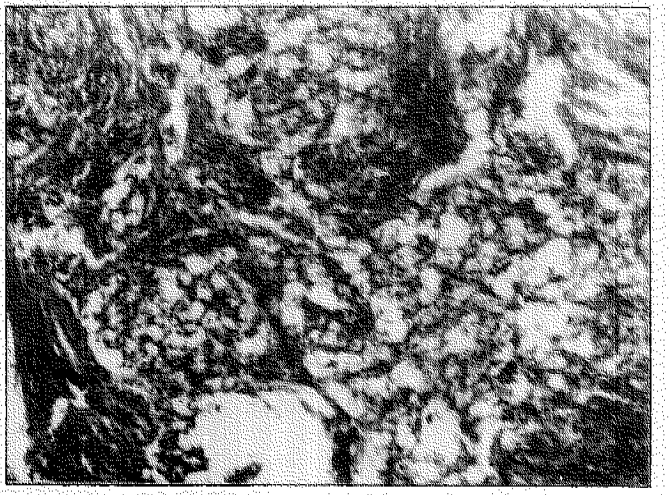

Figure 3e: This section shows predominance of mucus cells. At right lower corner, nest of epidermoid cells is seen.

wards this diagnosis. The possibility of NPCA, however may also be likely and important steps have to be undertaken (ie. NP biopsy) to ensure that this is not the diagnosis. Surgical intervention should be weighed carefully and a good clinical eye is necessary if this option is contemplated.

The name mucoepidermoid CA was given to salivary gland lesions in 1965 by Stewart, et al, when they split the neoplasms off from the broader category of mixed tumors. They choose the designation to emphasize the two main histological features of these tumors. The term is actually a contraction of mixed epidermoid and mucus secreting cell patterns. Clear cells and abundant lymphoid stromal components are also relatively common. The parotid gland is most commonly involved by the neoplasm (Batsakis, 1979).

Mucoepidermoid carcinomas may occur at any age with the highest incidence in patients who are between their fourth and fifth decades of life. There is a slight female predominance.

Clinical presentation varies. Some tumors may manifest as fixed to the surrounding tissues and therefore be non-mobile masses. In some patients, a facial paralysis is also a possible morbid event associated with the neoplasm.

There is a wide variation in the cellular morphology of individual mucoepidermoid carcinomas. Basically it is composed of three cell types: mucus secreting, epidermoid, and intermediate (Batsakis and Luna, 1990).

Foote and Frazel separate mucoepidermoid carcinomas into three grades of malignancy: low grade, high grade and intermediate. High grade mucoepidermoid carcinomas manifest considerable anaplasia. Three main histologic patterns are found. 1) The bulk of the lesion is squamous cell in character and resembles squamous cell carcinoma of other sites. 2) The carcinoma is composed of an abundance of cellular mucus and an equally prominent squamous cell distribution. 3) Carcinomas in which all cell lines and differentiation are present but in the peripheral zones there are anaplasia and frequent mitoses (Batsakis, 1979).

Mucoepidermoid carcinomas are said to be prone to recurrences. Frazell reported that 15% of low grade and 60% of high grade carcinomas recur. Stevenson and Hazard reported that 75% of the carcinomas recur. When recurrences appear, they usually do so during the first postoperative year (Batsakis, 1979).

Most authors consider the mucoepidermoid carcinoma to be of relatively low radiosensitivity. Complete regression as a result of radiation therapy is probably not achievable. Therefore, treatment is primary surgical removal, with the extent of surgery governed by the location of the in the gland, the presence or absence of palpable regional lymph nodes and other histological appearance of the tumor (Thorvaldson, Bearhs, Woolner and Simons, 1970).

The main dilemma in the case presented was whether to think of a parotid malignancy, which was based on the clinical picture, or to consider a metastatic lesion with an unknown primary, based on the biopsy. The recommendation is to consider the clinical picture very well because biopsy reports may mislead rather than enlighten us. For one, the various patterns of mucoepidermoid CA may be unfamiliar to the pathologist. Secondly, the biopsy specimens may not be adequate even if a wedge biopsy has already been done. The core of the tumor which may show distinct mucoepidermoid CA patterns, may not have been reached by the biopsy surgeon.

The presence of a biopsy report of metastatic CA, in fact, created the diagnostic problems in this case. Had the report revealed mucoepidermoid CA, initially, there would have been no question as to diagnosis & management.

Surgical intervention in this case was both wise & timely. As has been pointed out, most mucoepidermoid CAS are radio resistant. The question to do radical neck dissection is based on the surgeon's decisions. In this case, the presence of skin involvement on top of a huge parotid malignancy was enough to warrant such a procedure. Secondly, it was necessary to facilitate the use of the pectoralis major myocutaneous flap which was important to close the large defect created by the wide surgical excision.

CONCLUSION:

While it is ideal to extensively investigate and arrive at a more definitive histologic diagnosis preoperatively. It may be more important for a clinician to exercise good clinical judgement and practicality to approach the problem of neck mass.

Several points are emphasized from this case:

1. A histopatologic report should always be correlated with the clinical picture.
2. In the face of an unresolved diagnostic dilemma and limited diagnostic clues, the clinician should arrive at a practical therapeutic management that should save the patients time and resources.
3. The basic tool of a clinician, careful history taking and detailed physical examination, should not be overlooked and must be correlated well with the diagnostic work-up.

   Keen diagnostic decision, a good knowledge of differential diagnosis and a practical as well as a logical approach, are essential in order to arrive at a justified management of an upper lateral neck mass.

TABLE I: The common neck masses

Category	Neoplastic	Congenital/Devt'l	Inflammatory
Metastastic	Unknown Primary Epidermoid CA	Sebaceous Cyst Branchial Cleft Cysts, first and second	Lymphadenopathy Bacterial Viral Granulomatous
	Primary H & N Epidermoid CA or Melanoma Adenocarcinoma	Thyroglossal Duct Cysts Lymphangioma Hemangioma	Tuberculosis
Primary	Thyroid tumor Lymphoma Salivary tumor Lipoma Angioma Carotid Body Tumor Rhabdomyosarcoma	Dermoid Cysts Ectopic Thyroid Laryngocoele Thymic Cysts	Cat-scratch Sarcoidal Fungal Sialadenitis Parotid Submaxillary Congen. Cysts

(From Cummings, Otolaryngology - Head and Neck Surgery Volume II, 1986)

(FIGURE 4) POST-OPERATIVE PICTURE OF THE PATIENT.

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THE PHILIPPINE JOURNAL OF OTOLARYNGOLOGY HEAD AND NECK SURGERY

All manuscripts and other editorial matter should be addressed to Joselito C. Jamir, M.D., Editor-in-Chief, The Philippine Journal of Otolaryngology- Head and Neck Surgery, Department of Otolaryngology, UP-PGH Center, Taft Avenue, Manila.

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Head & Neck- An Area of Co-existence

The specialty of otorhinolaryngology, since its separation from its ever seeing twin Ophthalmology, has grown by leaps and bounds. Through the years, much development has occurred in the specialty in the form of new technology as exemplified by the fiberoptic scopes, the functional endoscopes, and the operating microscopes. These provided the practicing specialists new and more precise armamentarium in handling the problems and conditions within its traditional area of responsibility.

Growth in the specialty took on another dimension when its scope of study increased to include the neck and all the attendant conditions and diseases that may be present in this area. This happened because of several factors, namely: it is but a natural extension of events and interest; the first pure ENT men in this country (the so-called Heroic Nine) are all trained abroad where Head & Neck is really a part of and the proper domain of the Otolaryngologist; an adequate training program in Head & Neck Surgery has been incorporated in the residency program for ENT; other specialties has been so engrossed in other fields of endeavor that they have simply neglected Head & Neck and which the ENT men have been more than willing to fill-in the void.

But things and perceptions have changed and the situation has to be corrected, so says some of our colleagues in the medical profession. What is de facto the realm of the ENT man can not be made de jure even with the passage of time. The ENT man's claim to the area is void ab initio and, therefore, the request for recognition can not be granted. And that, apparently, should bring an end to the discussion. And yet, like an aphthous ulcer, the issue has become a recurrent sore that has strained relationships with our surgical brothers.

Is there really a need for such a conflict? Are the stakes high enough to divide the medical profession? Is it worth pursuing for the sake of academic gains? Or professional? Or monetary? I dare say no to all these questions. Not one specialty has the right to exclude his brother physicians from catering to the medical needs of his fellowmen. There simply are just to many cases around to satisfy all parties concerned. Besides, the patient has the right to choose among the different health professionals as the ultimate provider of services.

In these times of detente, crumbling of restraining walls, end of the cold war, and abolition of trade barriers, it seems that the medical profession is lagging behind the changing times. Head & Neck could be an area of peaceful co-existence among surgical specialists and the formation of the Philippine Council of Head & Neck Surgery to be composed of qualified practicioners is but the first step towards reconciliation and cooperation among men of goodwill sharing common interests and responsibilities.

JOSELITO C. JAMIR, MD
Editor-in-chief