PHILIPPINE JOURNAL OF OTOLARYNGOLOGY-HEAD AND NECK SURGERY VOL. 39 NO. 2 JULY – DECEMBER 2024

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Adenoid Cystic Carcinoma of the External Ear Canal: A Case Report

ABSTRACT

Objective: To present a rare case of Adenoid Cystic Carcinoma of the external ear canal in a 73-year-old man, including its diagnosis and surgical management.

Methods:

Design: Case Report
Setting: Tertiary Government Training Hospital
Patient: One

Results: A 73-year-old man who consulted due to decreased hearing on the right ear had a 4-year history of gradually enlarging mass occupying the right external auditory canal. Wide en bloc resection of the external auditory canal mass and superficial parotidectomy with facial nerve preservation was performed with final histopathology results of adenoid cystic carcinoma. Follow-up at 2 years has shown no signs or symptoms or recurrence.

Conclusion: Adenoid cystic carcinoma of the external ear canal is a very rare disease that can present similarly to other otologic infections. Early identification of symptoms, performance of necessary imaging, and timely biopsy are key to reducing misdiagnosis and improving the survival rate. Complete resection of the tumor with adequate margins is the recommended treatment due to its high rates for recurrence.

Keywords: *adenoid cystic carcinoma; ear neoplasm; ear neoplasm surgery; external auditory canal tumor; rare tumor*

Adenoid cystic carcinoma (ACC) is a type of malignant epithelial tumor with biphasic features of ductal and myoepithelial differentiation.[1] These tumors may occur as primary carcinomas of the major and minor salivary glands and account for 10% of their cases.[2] They may develop in any site where exocrine glands exist in the facial region.[3] Among the major salivary glands, the parotid glands are the most commonly affected.[1] Other unusual locations include the external auditory canal, lacrimal glands, breast, sinonasal tract, nasopharynx, oropharynx and trachea.[4] Only about 1% of adenoid cystic carcinomas account for those tumors seen in the head and neck region.[2,3] We present the case of a man who presented with a right external auditory canal mass diagnosed as adenoid cystic carcinoma that was later confirmed postoperatively.

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Philipp J Otolaryngol Head Neck Surg 2024; 39 (2): 41-44

CASE REPORT

A 73-year-old man consulted for decreased hearing on the right and a right external auditory canal nodule for the last 4 years. He noted gradual progression in size when he would palpate it by inserting a cotton bud in his external ear meatus. He denied experiencing any otalgia, otorrhea, bleeding or vertigo.

One month prior to consultation, he noted yellowish foul-smelling discharge and hearing loss. He consulted a private physician who started otic drops that did not provide resolution of the symptoms. Persistence of the symptoms prompted consult at our institution. Otoscopy revealed a fleshy, well circumscribed mass extending up to the opening of the external auditory canal (EAC) with minimal yellowish discharge. *(Figure 1)* No facial weakness, cervical lymphadenopathy or any neck mass were found.

Pure tone audiometry revealed severe conductive hearing loss in the right ear and normal hearing in the left. *(Figure 2)* A contrast enhanced temporal bone computed tomography (CECT) scan revealed a 2 x 1.9 cm mixed density soft tissue focus in the right external auditory canal. There were no bony erosive changes noted in the epitympanum and hypotympanum as well as in Prussak's space. The bony ossicles were unremarkable with no extension of the mass in the mesotympanum. *(Figure 3)* The lesion was confined to the EAC and was well encapsulated. The tumor did not invade the parotid gland and there were no extensions or connections noted to the parotid gland inferiorly or adjacent bony structures. An incision biopsy revealed mature squamous epithelium with presence of neoplastic cells in the fibrous stroma, forming glands with cribriform formation and basophilic mucoid material while some showed tubular pattern. The glands were composed of ductal and myoepithelial cells with hyperchromatic nuclei and clear cytoplasm, and appeared to infiltrate the surrounding fibrous stroma. These histopathologic findings were consistent with adenoid cystic carcinoma. *(Figure 4)* A preoperative chest x-ray revealed a short linear density in the left lower lobe which could be subsegmental atelectasis or pulmonary fibrosis, with no active pulmonary infiltrates or pulmonary mass noted.

A wide en bloc resection *(Figure 5)* of the external auditory canal mass included sleeve resection of the entire cartilaginous portion of the canal extending to the conchal bowl cartilage and skin within a 1.0-1.5 cm radius from the tumor as well as the tragal cartilage and the adherent skin posterior to it. Medially, this included the circumferential canal wall skin from the bony canal, around 5-10 mm posteriorly and 5 mm anteriorly. To provide an even wider margin, the posterior bony canal was carried down further and posteriorly up to 1 cm from its lateral border. A superficial parotidectomy with preservation of the facial nerve was also performed.

Intraoperatively, the mass had no attachment anteriorly and inferiorly but was noted to be attached supero-posteriorly with a wide base. Frozen section analysis of the margins (posterior canal wall skin, superior canal wall skin, anterior canal wall skin and posterior conchal bowl) were all negative for any neoplastic or malignant cells.

The final histopathologic evaluation demonstrated infiltration of the epithelium by clusters of neoplastic cells in cribriform, tubular and solid sheets. Numerous lymphoplasmacytic cells were likewise seen. The neoplastic cells were composed of ductal and myoepithelial cells while the cribriform pattern was composed predominantly of myoepithelial cells with myxoid globules. There was local perineural invasion, but no lymphovascular invasion was seen. This was signed out as adenoid cystic carcinoma. Histopathologic examination of the superficial parotid was negative for any tumor. There were no symptoms or signs of recurrence at 2 years post operative follow-up.

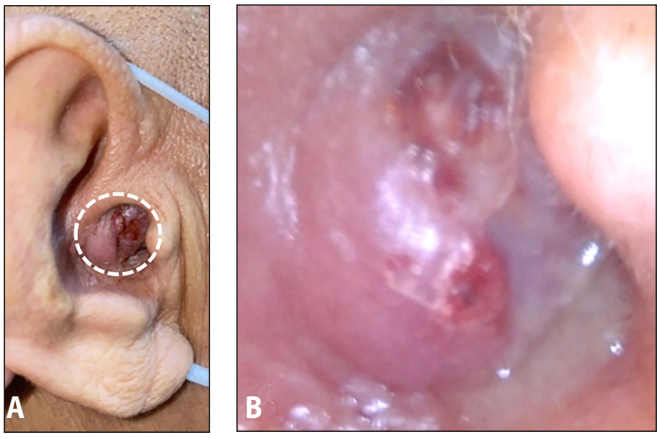

Figure 1. Physical examination photographs showing **A.** the fleshy mass protruding from the external auditory meatus (dotted circle); **B.** extending up to the external auditory canal

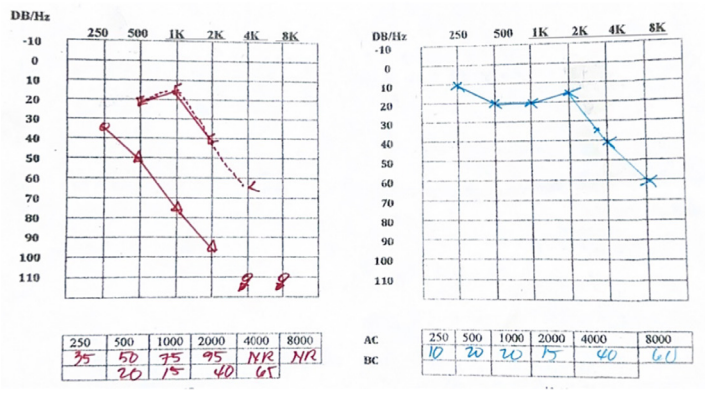

Figure 2. Pre-operative Pure Tone Audiometry (PTA) results showing right severe conductive hearing loss at 0.25-8KHz frequencies (with an air-bone gap of 48dB) and normal hearing on the left ear, based on the average of the threshold of 500Hz, 1000Hz, and 2000Hz. Both audiograms showed slopping towards the right, indicating bilateral high frequency loss

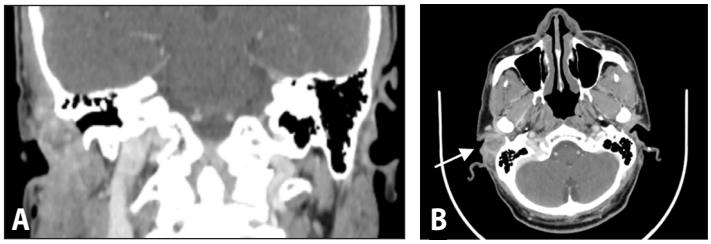

Figure 3. Contrast enhanced temporal bone computed tomography scans **A.** Coronal cut at the level of the external auditory canal; and **B.** Axial cut at the level of the maxillary sinus showing a 2 x 1.9 cm mixed density soft tissue focus (white arrow) in the right external auditory canal

DISCUSSION

External ear canal tumors are rare and exhibit variable histological and clinical presentations. They may present as primary tumors or as an extension of tumors from adjacent structures. The most common primary benign EAC masses are osteomas and exostoses.[1] On the other hand, squamous cell carcinoma (SCC) followed by basal cell carcinoma (BCC) and adenoid cystic carcinoma (AdCC) comprise the most common primary malignant rumors of the EAC.[5]

Primary carcinomas of the EAC are extremely rare malignant tumors.[6] Extension into the temporal bone, skull base or even intracranially can be seen in these type of tumors. These tumors are described to be locally aggressive and have a high degree of lymphatic spread.[7] The majority of EAC tumors are SCCs comprising 80%.[8] Adenoid cystic carcinomas arising in the EAC are infrequently encountered and account for only 5% of EAC tumors.[9]

External auditory canal carcinomas often present similarly to any benign or inflammatory condition of the ear, contributing to the difficulty in their diagnosis. The most common complaint of patients with carcinoma of the EAC is intermittent otalgia.[6] Patients may also present with unilateral severe or dull aching constant ear pain of prolonged duration, ear discharge, reduced hearing and a palpable mass in the external ear.[10] With this type of clinical picture, early diagnosis of EAC carcinomas is often missed. They may be misdiagnosed as chronic otitis media, external otitis and benign neoplasms of the external ear canal.[11]

In the case of our patient, his chief complaint was hearing loss. This was corroborated by audiometry that showed mixed hearing loss in the affected ear. Being elderly, he already showed signs of age-related sensorineural hearing loss demonstrated bilaterally in the audiogram. However, there was an additional conductive component in the right ear that may be explained by the presence of the mass and discharge in the ear canal.

Although the catalyst for his consult was the hearing loss, he also complained of a slow-growing palpable mass for 4 years prior to consultation. It appeared that the mass was not large enough to occlude the ear canal and only upon development of discharge and subsequent hearing loss was he eventually disturbed, prompting medical consult.

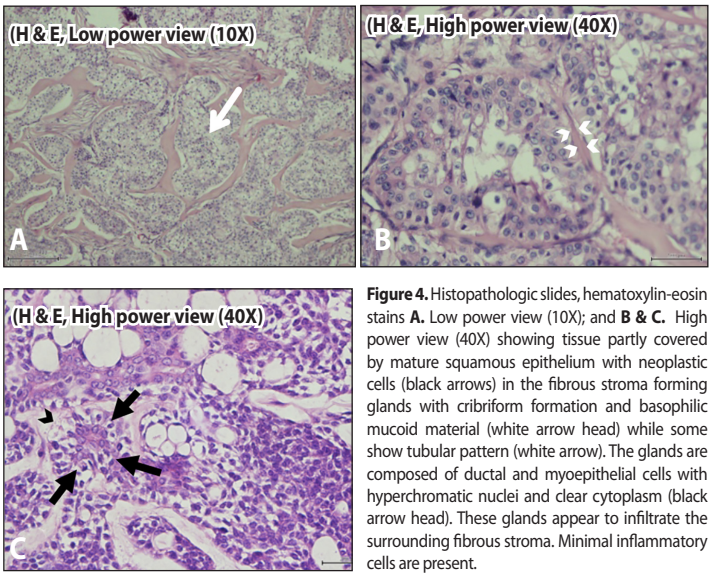

Figure 4. Histopathologic slides, hematoxylin-eosin stains **A.** Low power view (10X); and **B & C.** High power view (40X) showing tissue partly covered by mature squamous epithelium with neoplastic cells (black arrows) in the fibrous stroma forming glands with cribriform formation and basophilic mucoid material (white arrow head) while some show tubular pattern (white arrow). The glands are composed of ductal and myoepithelial cells with hyperchromatic nuclei and clear cytoplasm (black arrow head). These glands appear to infiltrate the surrounding fibrous stroma. Minimal inflammatory cells are present.

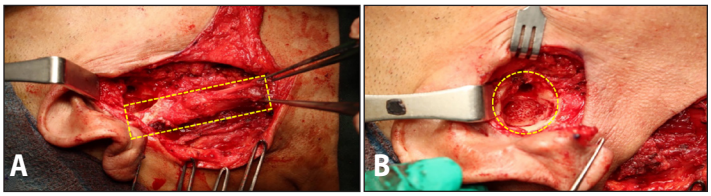

Figure 5. Wide resection of the external auditory canal mass included **A.** sleeve resection of the entire cartilaginous portion of the canal extending to the conchal bowl cartilage and skin (dotted rectangle); and **B.** Post resection margins included the posterior canal wall skin, superior canal wall skin, anterior canal wall skin and posterior conchal bowl (dotted circle)

The natural history of AdCC of the EAC is characterized by an indolent clinical course which usually leads to a late diagnosis.[9] Similar to cases documented in the available literature,[10,12] our patient was initially misdiagnosed as having a more common inflammatory otologic condition. The chronic use of otic drops which afforded no change in the size of the EAC mass further contributed to difficulty in diagnosing the patient due to delayed management. Moreover, the necessity of performing a biopsy in such cases is low among physicians.[12] In cases of EAC tumors, the significant overlap between the presentation and clinical features of benign and malignant tumors makes early and accurate diagnosis through histopathologic examination important for their proper treatment.[11]

The decision for surgical management is complicated as there are no available treatment guidelines as of yet. Aggressive tumor resection which may or may not be accompanied by radiotherapy and chemotherapy is the current recommendation, the extent of which is best guided by the severity of the disease.[9]

Currently, there is no generally accepted staging of this lesion. Although the University of Pittsburgh staging system for external

auditory meatus carcinoma is widely used for squamous cell carcinoma, its application has not been fully established for adenoid cystic carcinoma of the EAC.[13] However, Nyrop and Grøntved opined that the Pittsburgh staging system may also be useful for non-SCC.[6] Using the Pittsburgh staging system, our case may be classified as low grade T1N0M0. As such, treatment warrants complete surgical excision of the mass including the surrounding tissues such as the cartilaginous EAC.

A further question in this case is the treatment of the parotid gland because of its close histological association with the EAC. Although its origin remains controversial, it has been suggested that AdCC of the EAC may have originated from ceruminous glands on the EAC, sweat glands or ectopic salivary glands.[12] On the other hand, some cases may arise from the adjacent parotid salivary gland as it has been hypothesized that a parotid tumor may invade its surrounding fissures and extend to the EAC.[14] These fissures of Santorini can be found in the cartilaginous portion of the EAC and can provide a path for invasion in either direction between the EAC and the parotid gland.[15] Although the CT scan of the patient showed no bony erosions or extensions to the adjacent parotid gland, our decision to perform a superficial parotidectomy was based on recommendations of representative literature.[13,15] Jiang, *et al.* recommended that patients diagnosed with early disease should undergo superficial parotidectomy at the same time as tumor resection because studies had shown that there was a lack in the recurrence of the disease when it is removed simultaneously.[12] Hence, our patient also underwent superficial parotidectomy on the right with facial nerve preservation.

Frozen sections were also obtained to ensure margins were clear of tumor due to the high risk of repeat local recurrence of the tumor.[10] Adjunctive radiotherapy on the other hand, is only recommended for tumors staged T2 or higher or with osseous, cartilaginous and soft tissue invasion, which were not demonstrated in our patient.[16] Adenoid cystic carcinoma of the EAC is associated with perineural and bone invasion and a high risk of direct intracranial extension.[7] Recurrence may be observed several years after surgery, and mortality has been mainly attributed to lung metastasis.[7] Our patient is currently two years post-operative and currently has no signs of recurrence. This regular follow-up will continue for the next three more years as he will be monitored for recurrence through meticulous clinical and radiologic evaluation.

In summary, adenoid cystic carcinoma of the external ear canal is a very rare disease that can present similarly to other otologic infections. Early identification of symptoms, performance of necessary imaging, and timely biopsy are key to reducing misdiagnosis and improving the survival rate. Complete resection of the tumor with adequate margins is the recommended treatment due to its high rates for recurrence.

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