FEATURED GRAND ROUNDS

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

A Case Report on Maxillary Ameloblastic Fibroma in a 55-Year-Old Man

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Keywords: *odontogenic tumors; ameloblastic fibroma; ameloblastic fibrosarcoma; maxillectomy*

Ameloblastic Fibroma (AF) accounts for only 2% of all odontogenic tumors.[1] It has been shown to be more common in males than females.[2] A malignant transformation of this tumor is known as ameloblastic fibrosarcoma. To the best of our knowledge, there are no published reports on AF in the Philippines, with only one report of its malignant counterpart, ameloblastic fibrosarcoma.[3] We present a rare case of AF with an unusual presentation and discuss its rarity, pathogenesis, histologic features and management.

CASE REPORT

A 55-year-old Filipino man consulted at our tertiary government hospital for a one-year history of a gradually enlarging maxillary alveolar ridge mass extending to the hard palate. Upon first noticing the mass a year prior, he consulted with a local dentist and an intraoral examination revealed a smooth swelling on the right maxillary alveolar ridge between the first and second maxillary premolar, approximately measuring 2 cm x 2 cm x 1 cm. The mass was noted to extend to the hard palate. *(Figure 1A)* He was advised consultation with an ENT surgeon for further evaluation and management but he did not comply.

In the interim, the mass gradually enlarged. Upon first consultation at ITRMC, intraoral examination revealed a 6 cm x 4 cm x 2 cm soft, non-friable, ulcerative mass with irregular borders on the right maxillary alveolar ridge extending to the hard palate. *(Figure 1B)* The mass was malodorous, easily bled on manipulation and had violaceous areas. A punch biopsy revealed AF. Past medical history was unremarkable. A contrast-enhanced computed tomography (CECT) scan of the paranasal sinuses (PNS) revealed a 6.7 cm x 8.7 cm x 6.7 cm expansile, lobulated, mixed attenuating, heterogeneously enhancing mass in the midface. The bulk of the mass involved the right maxilla but extended to involve the entire hard palate and most of the left maxillary sinus. *(Figure 2)* The lesion was associated with osteolytic destruction and abutted the floor of the right orbit and bilateral zygomaticomaxillary buttresses. The inferior and posterior portions of the nasal septum were also eroded. *(Figure 3)* Given the clinical and radiographic features, odontogenic myxoma and ameloblastoma were considered as differential diagnoses as they may present as a painless, slow-growing, multiloculated mass with bony expansions.[4,5] Oral cavity squamous cell carcinoma was also considered. The mass was excised via a right subtotal maxillectomy and left inferior maxillectomy using a Weber-Ferguson approach *(Figure 4)* and a temporary surgical obturator was fitted. Post-operative recovery was unremarkable and the patient was discharged after five days. Once the surgical site healed he was fitted with an acrylic

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Presented at the Annual Research Forum for Case Reports 2024, held at the Ilocos Training and Regional Medical Center, San Fernando, La Union. February 21, 2024.

surgical obturator. Final histopathological report was AF, compatible with the pre-operative biopsy result. *(Figure 5)* On follow up three months post-surgery, the patient was able to eat and speak with ease and no tumor recurrence was observed. One year and three weeks later, there was still no recurrence noted on inspection of the surgical defect, his speech was hypernasal but understandable, and he had no difficulty communicating and eating.

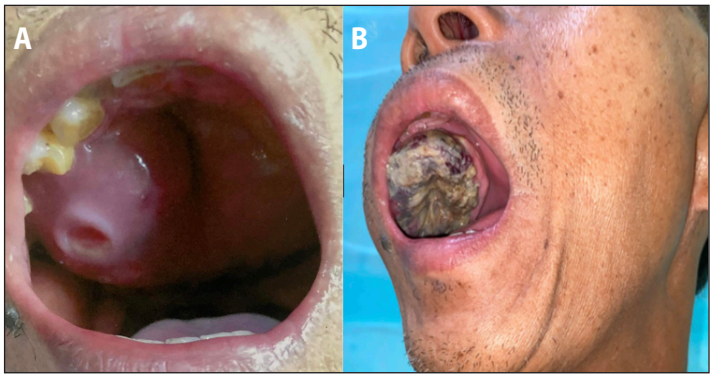

Figure 1. Oral cavity mass **A.** on initial consultation with dentist; and **B.** upon consultation at ITRMC after 1 year of gradual enlargement

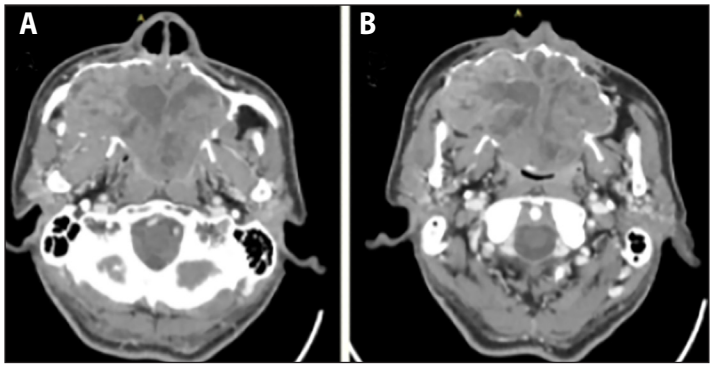

Figure 2. Axial cuts of the PNS CECT showing **A.** involvement of the right and left maxillary sinuses; and **B.** obliteration of the hard palate

DISCUSSION

Odontogenic tumors are a rare group of heterogeneous tumors which are derived from epithelial, ectomesenchymal and mesenchymal elements of the dental apparatus.[6] The World Health Organization defines AF as a "neoplasm composed of proliferating odontogenic epithelium embedded in cellular ectomesenchymal tissues that resembles the dental papilla and epithelial strands and varying degrees of inductive changes and dental hard tissue formation."[7] According to Reichart and Philipsen, as cited by Lalitha *et al.*, it has a slightly higher male predilection (male to female ratio: 1.4:1) and a mean age of occurrence of 14.8 years (range: 0.5 to 62 years).[2]

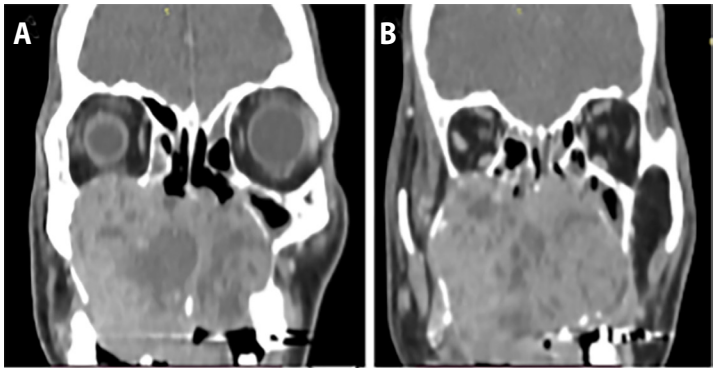

Figure 3. Coronal cuts of the PNS CECT showing **A.** partial erosion of the right orbital floor and abutment of bilateral zygomaticomaxillary buttresses; and **B.** involvement of the posterior nasal septum

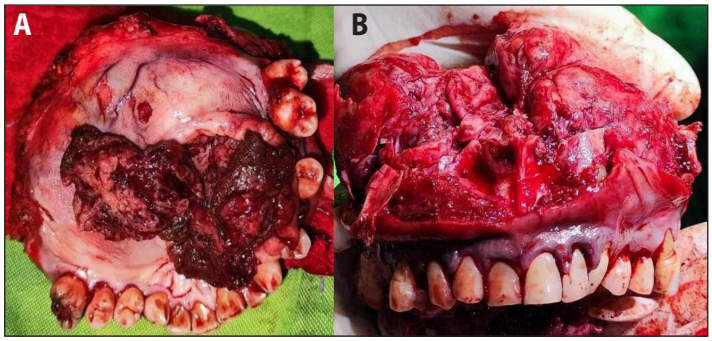

Figure 4. Gross specimen, **A.** Inferior view showing extrusion of the mass through the hard palate; and **B.** Front view showing superior projection of the mass through the hard palate, into the maxillary sinuses and nasal cavity

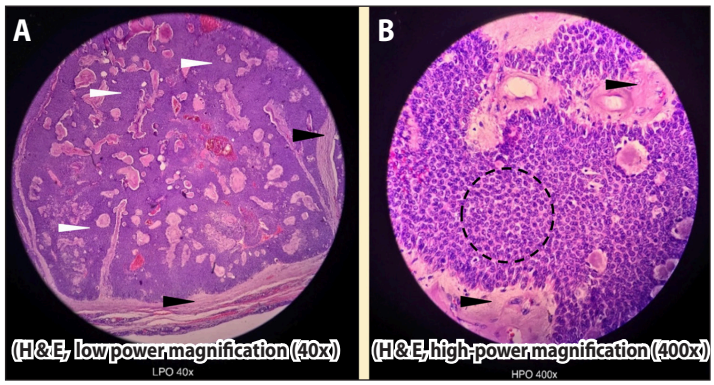

Figure 5. Histopathologic sections, Hematoxylin – Eosin **A.** low power magnification (40x) showing sheets of monotonous columnar and cuboidal with hyperchromatic nuclei (white arrowheads) and a myxoid connective tissue stroma (black arrowheads); and **B.** high-power magnification (400x) showing several layers of ameloblastic epithelial cells (black dotted circle) embedded in a myxoid connective tissue stroma (black arrowheads), with few to no mitotic figures and mild atypia

In approximately 75% of cases, it has been shown to be associated with an impacted tooth.[8] Ameloblastic Fibroma is usually well-encapsulated, slowly-growing, and commonly located in the posterior mandible. Bilateral presentation is exceedingly rare. Patients may also present with a hard swelling but intraoral ulceration, pain, tenderness, or drainage may also be observed.[9] In the present case, our patient presented with soft mass with ulceration and tenderness, however

his tumor was located in the maxilla and was not associated with an impacted tooth. This case of AF was, therefore, unusual in presentation.

The pathogenesis of AF remains unclear.[8] It is theorized that the primordial enamel organ gives rise to the odontogenic epithelium of AF while the dental papilla gives rise to its stromal component. This behavior is analogous to the similarities seen in hamartomas and true neoplasms.[9] It is believed that the tall columnar ameloblast-like cells in the epithelial component are too primeval to influence the cells of the ectomesenchyme, and even less is known about their interactions. It is also not clear why induction of odontoblastic differentiation is lacking in AF.[8] Very little is known about the genetic influences in AF. Radiographically, AF are commonly located near the crest of the alveolar processor in a follicular relationship with an unerupted tooth or they may arise in an area where a tooth failed to erupt.[2] It may appear as a unilocular lesion or a multilocular lesion with smooth, well-demarcated and corticated borders.[10] Differential diagnoses include benign lesions like ameloblastomas and myxomas but malignancies should also be considered.

Histologically, a proliferation of odontogenic epithelium in the background of primitive mesenchymal connective tissue is seen in AF. Ameloblastic Fibroma has a myxomatous appearance due to the spindled and angular cells in scant collagen. Its epithelium exhibits nests, buds, and cords of cuboidal or columnar cells and has a central portion that resembles stellate reticulum. Abundant mitosis is not characteristic of AF, such that when present malignant entities should be entertained.[2]

There is no consensus regarding the definitive approach to its surgical management. A more radical approach may be preferred due to AF's recurrence rate and malignant transformation rate, which may be as high as 33.3% and 11.4%, respectively.[11] Possible surgical management involves enucleation, excision and curettage, or marsupialization and curettage, segmental resection and the use of iliac crest graft.[9,10] For our patient, a radical approach via a right subtotal maxillectomy and a left inferior maxillectomy was done mainly due to the size and extent of the tumor. It can be said that management is dictated by patient age, extent and spread of the lesion, and histopathological findings. For younger patients, the goals of treatment are to remove the tumor and decrease the chances of recurrence while preserving adjacent vital structures.

The recurrence rate of AF is unclear since not all cases have a long term follow up. In a study by Pitak-Arnnop, AF had an approximately 70% recurrence rate at 10 years after operation.[9] Also, malignant transformation of AF has been reported in recurrent AF or after multiple surgeries.[9] According to Chen *et al.*, 14 out of 41 recurrent AF cases developed malignant changes and the estimated 10-year malignant transformation rate was 25%.[11] Reichart and Philipsen argue that careful surgical enucleation and close follow up should be preferred over aggressive initial management because of the innocuous behavior of AF.[12] Regardless of its benignity, a thorough histopathological assessment is warranted in recurrent cases of AF in order to rule out malignant transformation.[10]

Though benign in nature, AF has a potential to be malignant and has a high recurrence rate. Because of the relatively few number of cases reported, its exact incidence, genetic features, and pathophysiology are still unclear. Despite being a benign lesion, AF requires long term follow up post-operatively, due to its recurrence rate and potential for malignant transformation.

In this report, we described an unusual presentation of an already rare tumor. As seen in our patient, AF can present in the older age group, involve the maxilla, and not be associated with an unerupted tooth. Our case also demonstrates that it can present as a sizable mass requiring radical surgery yet still remain benign. Despite being rare, AF should therefore still be considered in the differential diagnosis of masses in the jaw and alveolus.

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