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Case Report

Maxillary Ameloblastic Fibroma: Two Case Reports of a Rare Tumor

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ABSTRACT

Ameloblastic fibroma is a rare benign tumor usually affects the first two decades of life. The neoplasm is more predominant in mandibular molar-premolar region and rarely affects the maxilla. In this report, we present a couple of Ameloblastic fibroma cases, affecting boys at their 1st decade. The lesions were presented as swellings of their maxilla, which is atypical location. Radiographic images showed well-defined radiolucency containing areas of radio-opacities and impacted teeth. Differential diagnosis was established as cystic/neoplastic conditions. The lesions were incised and histopathologically diagnosed as Ameloblastic fibroma, since they were composed of immature odontogenic mesenchymal and epithelial cells showing different characteristic features.

Introduction

Ameloblastic fibroma (AF) is a true benign tumor composed of neoplastic proliferation of odontogenic mesenchyme and epithelium. It constitutes 1.5-6.5% of all odontogenic tumors, can occur either in the mandible or maxilla, but over 80% of tumors occurs in the posterior mandible; the ratio of mandibular to maxillary localization is 3.3: 1 (1, 2). However, there have been reported cases arising peripherally (extra osseous), and in the maxillary sinus (3).The mean patient age is 14.9 years, range from 7 weeks to 57 years, mostly (about 80%) occur in patients younger than 22 years (i.e. before the end of odontogenesis), and the male-to-female ratio is 1.4:1 (4). Clinically, the tumor presents as asymptomatic slowly growing swelling and when large enough it may lead to facial deformity, teeth malalignments and failure of teeth eruption, or it may be found incidentally on radiographic imaging (5).

The radiographic features are variable, ranging from a well-circumscribed small radiolucent (RL) unilocular lesion to a more expansive multiloculated appearance seen in larger tumors (6). Borders of the lesion are well defined with sclerotic margins (5).

Histopathologically, AF is a biphasic tumor of soft tissue made up of odontogenic ectomesenchyme resembling tooth-related structures such as dental papilla, and epithelial strands and nests similar to the dental lamina and enamel organ but without dental

hard tissues. The stromal component features imparts a myxomatous appearance (2). Excessive cellularity of AF is thought to be a result of overproduction of the basal lamina during tooth formation, but without odontogenic differentiation (i.e. no inductive events) (7). Because AF is often associated with non-erupted teeth, it may initially be interpreted as a dentigerous cyst and ameloblastoma (8). Others stated that ameloblastoma, odontogenic myxoma, odontogenic keratocyst (OKC) and central giant cell lesion should be considered in the differential diagnosis (9). Ossifying fibroma, extra-follicular variant of adenomatoid odontogenic tumor, calcifying epithelial odontogenic cyst/tumor were also considered as hypotheses of diagnosis (6). Microscopic differential diagnoses include odontogenic myxoma and odontogenic fibroma (10).

Surgical conservative treatment with excision followed by curettage seems to be the most appropriate therapeutic option and follow-up should be for a long period of at least 3-5 years to enable detection of possible recurrence or development of ameloblastic fibrosarcoma, which is the malignant counterpart (11, 12).

Our goals in this report were to record two adulthood maxillary AF cases and bring attention to the differential diagnosis of this particular tumor that would affect the type of treatment. Moreover, we discuss it`s histological characteristics that might be confused with other odontogenic lesions.

Clinical and radiographical description of the first case

A two and a half years old male child, presented by his mother with a facial swelling of left posterior mid side of one year duration. She claimed that the condition was asymptomatic and his general health was good. Intraoral examination revealed a smooth, 2*6 cm swelling of the posterolateral vault part covered by pinkish mucosa. On palpation, the lesion was solid and the regional lymph nodes were normal. The 2nd deciduous molar was missing. On the antero-posterior plane, the swelling extends from the distal surface of a carious upper first deciduous molar to the tuberosity, and medio-laterally from the median raphe to involve the remaining alveolar process (Figures 1- a and b). The patient was sent to magnetic resonance imaging.

The MRI images showed a 33*27 mm heterogeneous ill-defined lesion filling the left maxillary sinus and affecting the regional alveolar process, an impacted tooth crown was involved in the lesion (Figure 1- c, d, and e). Provisional diagnoses were established as dentigerous cyst, ameloblastoma, and rhabdomyosarcoma. Therefore, excisional biopsy was performed.

Clinical and radiographical description of the second case

A healthy male child of 19 months age was presented with a growing mass in the right upper mid face of eight months duration. The lesion was lifting the upper lip, stretching the right nostril, and a slightly cutaneous erythema was observed under the right eye (Figure 2- a). There were no abnormal habits or previous treatments, and no history of any medical diseases according to the parents. Intraoral examination revealed a solid swelling that was noted to be hard on palpation, not compressible, non-fluctuant covered by normal colored mucosa. The patient was sent to computed tomographic (CT) scan.

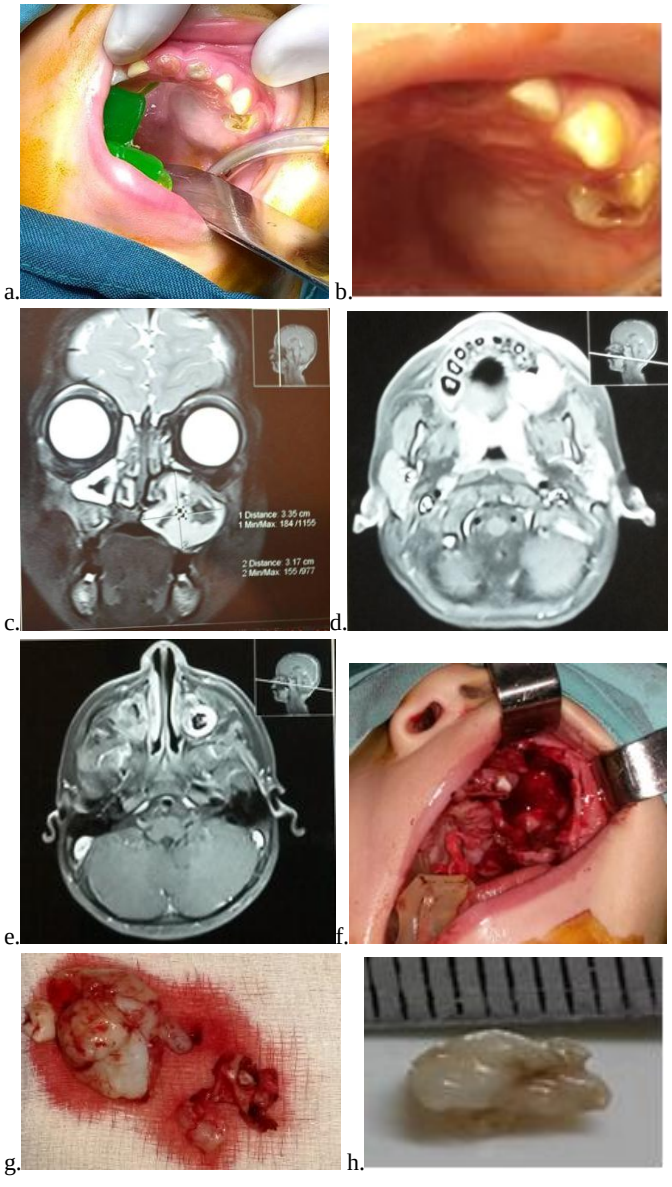


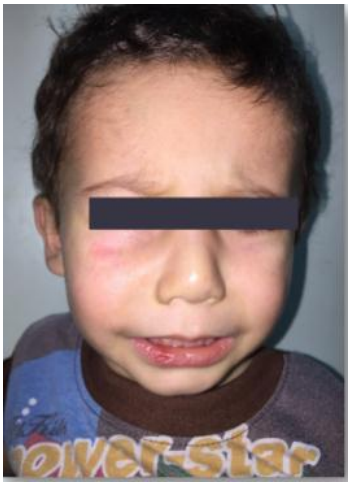
Figure1: Case 1 clinical, radiographical and gross pictures.
a- Intra-oral view of the palatal lesion.
b- A closer view of the palatal lesion (Note; that the second deciduous molar is missing).
c- Coronal MRI view reveals that the lesion involves the left maxillary sinus. Expansive lesion containing multiple radiopaque foci.
d- Axial MRI view at the level of the maxillary alveolus shows a high-intensity mass on the left side.
e- Axial MRI view at the level of the maxillary sinus shows a high-intensity mass on the left maxillary sinus. A well-defined, radiolucent, unilocular, expansile mass is seen without bony erosion. Scattered radiopaque calcifications (arrows) are also present.
f- The oral approach and antrostomy performed to excise the lesion.
g- Gross mass of the lesion following surgical enucleation.
h- Gross pathological specimen.

CT images showed a crown of an impacted tooth in a well-defined mixed RL and radio-opaque (RO) lesion surrounded by a clear sclerotic margin, filling the right maxillary sinus region. The expanded lesion caused compression on the sinus plates in all directions and partially obliterating the lower and middle region of left nasal cavity (Figure 2- b, c, and d). The surgeon`s provisional diagnoses were established as dentigerous cyst, KOC, and ameloblastoma. Therefore, excisional biopsy was performed to remove the lesion (Figures 2- e).

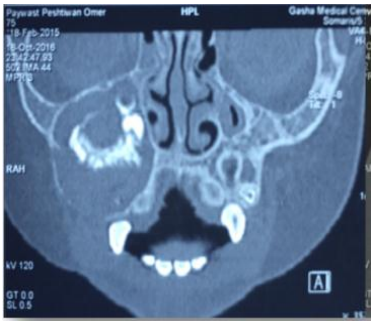
Grossly, multiple excised white or white and brown soft tissue fragments, having a firm consistency with folded granular surfaces, and an impacted crown was included. All were immediately fixed and imbedded. The largest one measured 2.5* 1 cm (Figure 2- f).

Histopathological features of the reported cases

Microscopic sections of both cases (Figure 3) showed soft tissue masses consisting of (1) dominant fibroblast-rich stroma (spindle, oval and stellate shapes) with collagen, very similar to primitive dental papilla cells. (2) Little proliferating odontogenic epithelium that was scattered as separate or interconnecting thin cords, buds, nests and islands. As for the cords and buds, they appeared to be formed of double or triple layered small hyperchromatic odontogenic epithelial cells that contain little amount of cytoplasm. While the nests and islands showed peripheral columnar folded cells with reverse polarity similar to the inner enamel epithelium surrounding loosely arranged central cells resembling stellate reticular cells (Vickers-Gorlin change). In addition, some of the epithelial components were surrounded by hyalinized connective tissue, and some others showed mild cystic degeneration. No dental hard tissue, no mitotic figures, or cellular atypia were observed. Teeth follicles also were seen on the periphery of both cases. Therefore, the final diagnosis was AF.



a.



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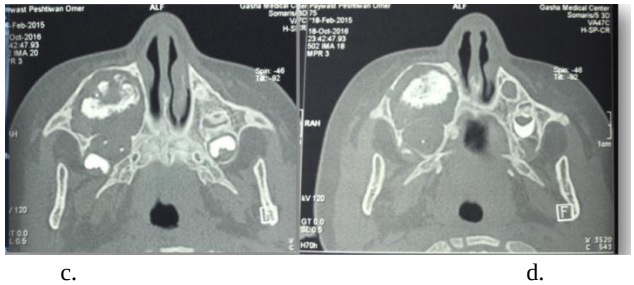

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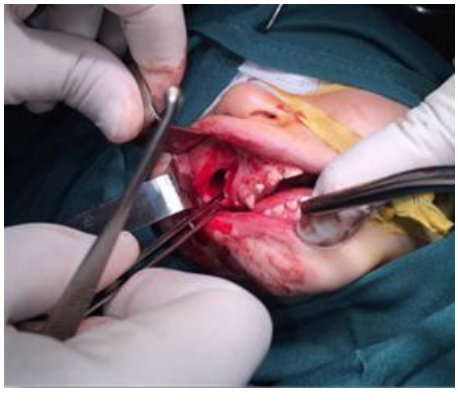
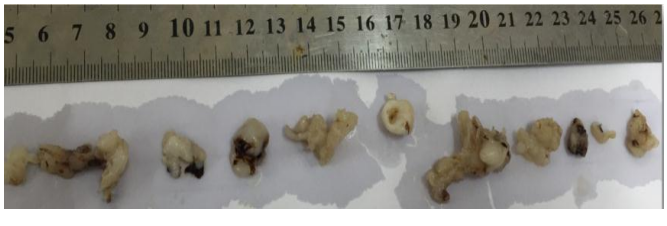


e.



f.

Figure 2: Case 2 clinical, radiographical, and gross pictures.
a- Extra-oral view of the maxillary swelling.
b- Coronal CT scan showing a well-defined expansile rounded hyper-dense mass in maxillary sinus region.
c- Axial CT scan showing a well-defined opaque mass of left maxillary sinus and deviation of the nasal cavity
d- Axial CT scan showing right maxillary sinus opacification, with cortical bone expansion and destruction, extending into the right nasal cavity. The 1st deciduous molar has been displaced and supra-erupted.
e- The oral approach and antrostomy performed to excise the lesion.
f- Multiple excised white or white and brown soft tissue fragments

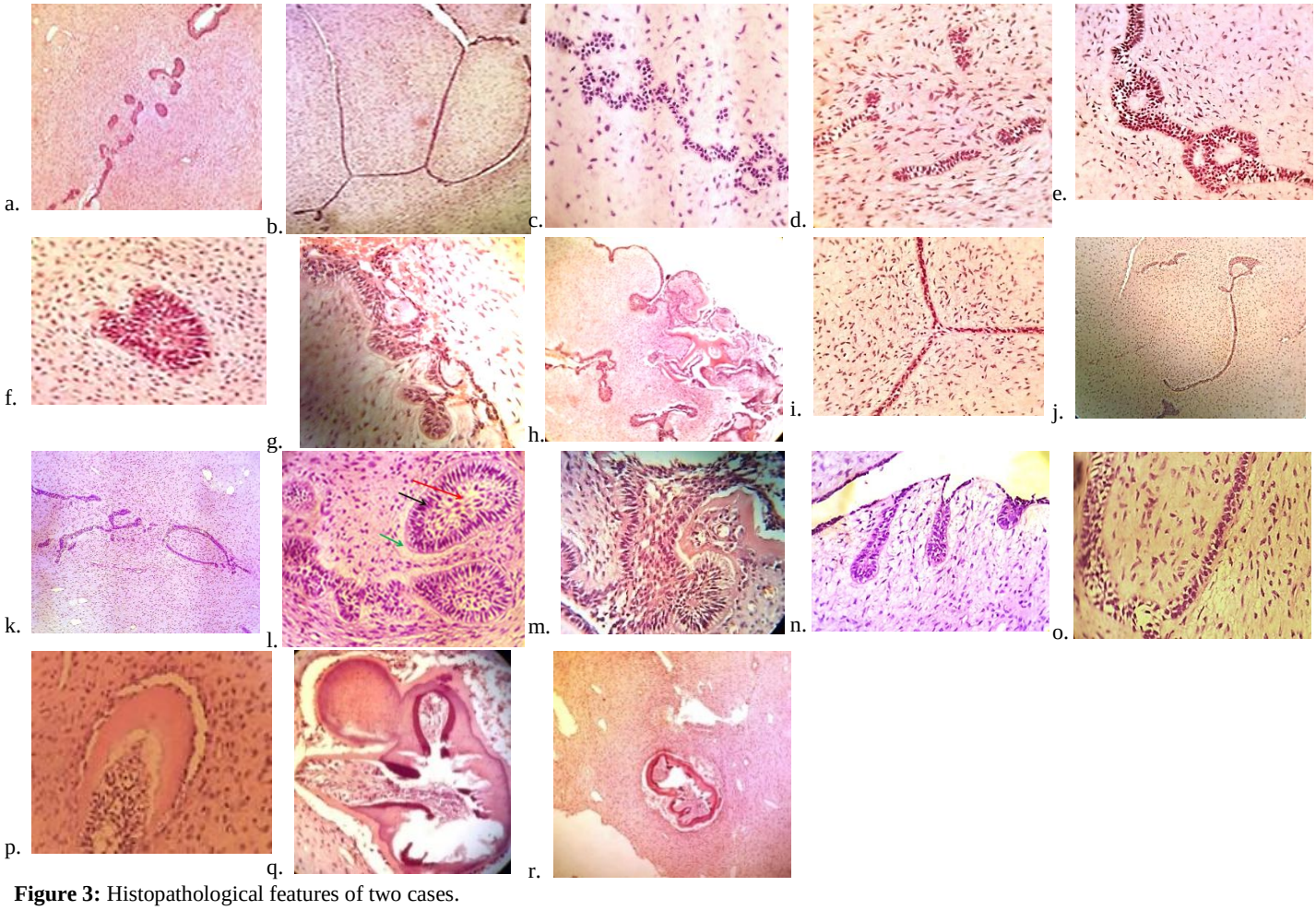


Figure 3: Histopathological features of two cases.

a- Low-power view shows nests, buds and cords of odontogenic epithelial cells in a fibroblastic stroma.

b- Low-power view shows thin strands of odontogenic epithelial cells in a fibroblastic stroma (intra-canalicular pattern), long narrow anastomosing cords.

c, d- High-power view of the double layered odontogenic epithelium appears as narrow cords embedded in a cellular connective tissue stroma.

e- Hyperchromatic triple layered odontogenic epithelium appears as interconnecting nests.

f- Epithelial island with peripheral tall columnar cells showing reverse polarity of nucleus simulating ameloblasts and central loosely arranged cells simulating stellate reticulum embedded in a primitive mesenchymal background.

g- Hyaline-like material surrounding islands, cords, and buds of ameloblastic epithelium embedded in a cellular connective tissue stroma.

h- Epithelial cords with numerous buds and foci of hyalinization and hemorrhage.

i- Interconnecting epithelial strand in fibroblastic connective tissue. These strips were formed of two layers of small cells.

J, k- Low-power views of thin cords and small islands of proliferating epithelium.

l- High-power of the epithelial islands made of a central stellate reticulum-like cells (red arrow) and peripheral columnar ameloblast-like cells with palisading hyperchromatic nuclei (black arrow), surrounded with a clear thin layer representing cell free zone (Vickers-Gorlin change) (7).

m- Epithelial islands with cystic degeneration.

n- Buds of double-layered odontogenic epithelium in the fibroblastic stroma, drop-like islands.

o- Narrow strand of odontogenic epithelium in uniform cells of myxoid stroma, lacking the atypia.

p- Homogenous epithelial hyalinization

q, r- Cross sections in teeth follicles.

Discussion

Depending on the clinical descriptions of AF in the literature, this tumor often appears in the form of an asymptomatic bulge in the jawbones (predominantly lower jaw) and when it is large in size it leads to facial deformity (1). Infrequently it involves the maxilla (13), and the maxillary sinus (14). Males are more prone to the tumor than females and it occurs exclusively in the first and second decades of life (2, 9).

According to the 4th edition of WHO head and neck tumors, AF may be incidentally found on radiographic imaging, and close to 75% of all cases, an impacted tooth is associated with the lesion. A mixed radiographic image is very unusual, but a well-defined unilocular or multilocular RL, with a mean size of 4.0 cm are more popular (2). The borders of the lesion are well-defined with sclerotic margins, there may be cortical expansion in a buccolingual plane but this may be misinterpreted on a 2D image (15), and therefore, in our 1st case we considered MRI images where soft tissue invasion was suspected, while in the 2nd case CT images make sense to assess tumor extent and invasion. However, these clinical and radiographic features may be common with other types of jaw tumors and cysts may deviate the surgeons to a list of diseases in the differential diagnosis. Regarding rhabdomyosarcoma, the most common type of soft-tissue cancer in children, children can develop it at any age, but most cases are in kids between 2 and 6 years, where the first sign might be a lump or swelling affecting head and neck region, including the nasal and oral cavities, as well as the orbit and middle ear (16). In addition, OKC can occur anywhere in the jaw, although commonly seen in the posterior mandible. Radiographically, most OKCs are unilocular when presented at the periapical region (2).

AF is often confused with ameloblastoma and dentigerous cyst due to the presence of impacted teeth and it is not unusual to find these diseases early in life especially the unicystic ameloblastoma (17). The odontogenic myxoma most frequently occurs in second or third decades of life, and has a slight female predilection (9). The exact diagnosis of AF is only histopathologically established (18).

Grossly, the surgical specimen of ameloblastic fibroma is characteristically loose, slippery or gelatinous in nature (18). Alternatively, it appears as firm, lobular soft tissue mass with a smooth surface, may be accompanied with a tooth (7), or it may have a rubbery consistency with a white to yellowish cut surface (15).

Whitson et al. (8) mentioned that AF could have many different histopathological characteristics. It consists of strands and islands of epithelial cells somewhat resembling the arrangement seen in ameloblastoma and the intervening tissue, however, it is a richly cellular connective tissue that resembles the dental papilla of the developing tooth. While in the other pattern, the epithelial cells form small discrete islands that resemble the follicular stage of the developing enamel organ. These show peripheral columnar cells, which surround a mass of loosely arranged epithelial cells that resemble stellate reticulum (19). The epithelial component may be remnant of dental papilla or Rests of Malassez. The center of the epithelial component may contain stellate reticulum (6). The periphery of the epithelial component is often lined by basally palisaded nuclei, occasional cases may even show reverse polarity of the epithelium or an intra-canalicular pattern and the mesenchymal component typically has bland plump to ovoid-spindled cells in a myxo-hyaline matrix (20). Multiple fragments of richly cellular mesenchymal tissue contain round drop-like islands and long narrow anastomosing cords of odontogenic epithelium. The Presence of tooth/teeth follicles indicates that the tumor begins at different points in a person's lifetime (21). In contrast to the follicular type of ameloblastoma, these follicular islands in the AF seldom demonstrate micro cyst formation (18). Lesions with an identical histology can show a neoplastic as well as a hamartomata's behavior (4).

Mitoses should not be a feature of ameloblastic fibroma; nevertheless, the presence of mitosis should expand the differential diagnosis to include malignant entities (21). The presence of Vickers and Gorlin criteria in pathologic interpretation constitutes evidence of neoplasia i.e. nuclear hyperchromatism, nuclear palisading with reverse polarization, and cytoplasmic vacuolization with intercellular spacing, all observed together but without cellular atypia (4), as in our cases.

Ameloblastic fibro-odontoma is identical to the AF but the calcifying element consists of foci of enamel and dentin matrix present in close relationship to the epithelial structures. The more calcified lesions show mature dental structures in the form of rudimentary small teeth or conglomerate masses of enamel and dentin (22, 23).

Surgical conservative treatment with close clinical follow-up seems to be the most appropriate therapeutic option. Although it is rare, both included tissues (epithelial and mesenchymal) in the tumor have a potential for recurrence and malignant transformation (11).

Conclusion

AF is an odontogenic benign tumor, expansile but it is not tending to spread. Generally, most of the affected patients are children. The clinical and radiographic signs of this tumor are very similar to those of other jaw tumors. Therefore, we confirm that the correct diagnosis is by extensive histological examination, especially in rare and unfamiliar cases, to differentiate it from similar diseases. The treatment of choice is surgical excision. The recurrence and malignant transformation are rare. Hence, evaluation with long-term follow up is necessary.

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Conflict of Interest

Authors declare no conflict of interest.

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