An unusual presentation of extra-follicular variant of adenomatoid odontogenic tumor: A rare case report

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Abstract

Adenomatoid odontogenic tumor (AOT) is a benign, relatively rare and distinct odontogenic tumor that is exclusively derived from odontogenic epithelium. Although being odontogenic in origin, the presence of the so-called “duct-like structures” is a unique microscopic feature that gives the lesion a glandular, i.e. adenomatoid appearance. AOT affects young individuals mainly in the second decade with a female predominance, occurring most commonly in the anterior maxilla and is usually associated with an impacted canine. Here, we are presenting a rare case report of an AOT in the mandible in relation to 43, 44, and 45 in 30-year-old male patient. The tumor was a well-circumscribed intraosseous lesion without an embedded tooth. Microscopic examination showed the characteristic histological features of AOT along with calcified epithelial odontogenic tumor-like areas.

Key words: Adenomatoid odontogenic tumor, calcified epithelial odontogenic tumor, odontogenic tumor, rosette like

Introduction

Adenomatoid odontogenic tumor is a relatively uncommon, distinct, slow growing, odontogenic, benign epithelial neoplasm that was first described by Steensland in 1905.[1] Dreibladt, in 1907, described it as a pseudoadoenameloblastoma.[2] In 1948, Stafne considered it a distinct entity and labeled it as “an epithelial tumor”. [3] Thoma in 1955 described it as an adenoameloblastoma. Philipson and Birn proposed the name adenomatoid odontogenic tumor in 1969 and suggested that it should not be regarded as a variant of ameloblastoma because of its different behavior. This term was adopted in the World Health Organization (WHO) classification in 1971.[4] Marx and Stern coined the term, “adenomatoid odontogenic cyst” in 2003.[5] According to the second edition of the WHO journal “Histological Typing of Odontogenic Tumors”, AOT is defined as “A tumor of odontogenic epithelium with duct-like structures and varying degrees of inductive change in the connective tissue. The tumor may be partly cystic, and in some cases the solid lesion may be present only as masses in the wall of a large cyst”. [6]

It is an uncommon odontogenic tumor with frequency of 2.2-7.1%. [7] AOT usually affects young patients, mostly during their second decade of life. Females are more commonly affected than males, with a male:female ratio of 1:2.[7] This tumor occurs more frequently in the anterior part of the jaws with 76% developing anterior to cuspid in the maxilla. It has been reported that in 74% of the cases, this tumor was associated with an impacted tooth.[8]

There are three variants of AOT: follicular, extra-follicular, and peripheral. The follicular type is a central intra-bony lesion associated with an unerupted tooth, which accounts for about 70% of all cases. The extra-follicular type is also an intraosseous lesion, but unrelated to an unerupted tooth and represents 25% of all AOTs. The peripheral type is a rare form that arises in the gingival tissue, and very few well-documented cases are reported.[9,10] All three variants have the same histological aspect and clinical behavior.[11]

As the histogenesis of AOT is still uncertain, there has long been a debate as to whether it represents anomalous hamartomatous growth or is a true benign neoplasm.[8]

The tumor may be cystic in its presentation or solid areas in some lesions may present as nodules in the capsule of a large cyst. While AOT is reported as a tumor in the histological sign out, the notion that it represents a hamartomatous malformation adds a new dimension to its assorted histological architecture.[12]
In this unique case report, we have emphasized the diverse histoarchitectural features and the unusual site of occurrence of AOT.

Case Report

A 30-year-old male patient visited the department of Oral Pathology and Microbiology, MGM Dental College and Hospital with the chief complaint of a swelling in the lower right front region since one year. He had a history of trauma at the same region one year back, and since then he noticed the swelling in that region that gradually increased in size. The patient had no relevant medical or dental history except pain on palpation and mastication.

An extra-oral examination revealed a facial asymmetry with solitary, diffuse swelling of approximately 2 × 3 cm in size present on the mandibular right side. The swelling extended from the right angle of the mouth to approximately 3 cm posteriorly [Figure 1]. The swelling was round to oval in shape, the overlying skin was not fixed to underline swelling and appears to be normal, and no draining sinus was present. On palpation, the swelling was bony hard with well-defined borders, non-tender, non-fluctuant, non-compressible, non-pulsatile, and afebrile.

Intraoral examination revealed a diffuse solitary swelling in relation to 43, 44, and 45 obliterating the mucobuccal fold, measuring about 2 × 3 cm in size with apparently normal overlying mucosa [Figure 2]. On palpation, swelling was diffuse, firm in consistency, slightly tender, non-pulsatile, non-fluctuant, and non-compressible and showed no evidence of discharge on digital pressure. Hard tissue status revealed carious 46, grade I mobility in 31, 41, and over-retained 73. No lymphadenopathy was present. Orthopantomogram showed a well-defined radiolucency in periapical region of 42 extending up to the mesial aspect of 47, and no evidence of calcifications was observed. Root resorption of 43 and 44 was noted. It also showed horizontally impacted 33 [Figure 3]. Lateral cephalograph and PA view were performed [Figure 4] which also showed the well-defined radiolucent areas.

Fine-needle aspiration cytology was performed. The aspirated fluid was reddish in color and microscopically non-diagnostic and inconclusive [Figure 5].

A differential clinical diagnosis of an aneurysmal bone cyst, odontogenic cyst, and odontogenic tumor was considered.

The patient was referred to the department of oral surgery for treatment. Surgical enucleation of the lesion was carried out under local anesthesia.

Histopathology revealed the presence of solid nodules of varying sizes composed of cuboidal or columnar epithelial cells forming nests and rosette-like structures [Figure 6]. Between the epithelial cells of the nodules and in the center of the rosette-like structures, eosinophilic amorphous materials (tumor droplets) as well as calcified bodies were present [Figures 7 and 8].

Spindle-shaped or polygonal, closely opposed epithelial cells filled in the spaces between the epithelial nodules. Focal areas showed tubular and duct-like pattern of arrangement of cells. The duct-like spaces were lined by a single row of low columnar epithelial cells [Figure 9]. The lumen in some duct-like structures contained a variable amount of eosinophilic material. Some areas of the section showed cuboidal to columnar cells forming the convoluted cords [Figure 10]. Some areas also showed an unusual group of cells that appeared polyhedral, giving a squamatoid appearance but lacking inter cellular bridges [Figure 11]. The scanty connective tissue stroma was loosely structured containing thin-walled congested blood vessels, a hyaline dysplastic dentinoid material, and irregular calcified bodies [Figure 12]. Some islands showed pools of amorphous amyloid-like material and globular masses of calcified substances. The amyloid-like substances...
showed positivity for Congo red staining [Figure 13]. There are some areas that appeared clear but these showed the negative staining for Periodic Acid Schiff stain [Figure 14].

Considering all these histopathological features, a diagnosis of adenomatoid odontogenic tumor was made.

**Discussion**

AOT comprises approximately 2.2-7.1% of all odontogenic tumors,[9] ranking behind odontoma, ameloblastoma, periapical cemental dysplasia, and myxoma.[11] The origin of AOT is controversial. However, evidence also exists that the tumor could be derived from epithelial remnants of the dental lamina complex system.[13,14]
This case report illustrates the characteristic clinical and radiographic features of the extra-follicular variant of AOT at an unusual site (the mandible). Usually, AOT is prevalent in the second decade of life, and the female gender is affected twice as often as the male gender, contrary to our case in which...
it occurred in a 30-year-old male patient. There are reports of AOT presenting in infants and also in individuals in eighth decade of life. The common site is maxillary anterior region and is usually associated with an impacted tooth, mostly the canine with displacement of adjacent teeth. In our case, AOT occurred in the mandibular anterior region extending posteriorly up to the molars having no association with an impacted tooth. Our case followed the biological trend of the common intraosseous location.

Tumors are usually in the dimensions of 1.5-3 cm, although larger lesions have been reported in the literature. Moreover, the presence of an intact capsule in most of the cases further reinforces the benign nature of AOT. It could be associated with a tooth anomaly, displacement of teeth, buccal plate perforation, hypothesis, and root resorption. In our case, the patient clinically showed a swelling of approximately 2 x 3 cm in size with buccal cortical plate expansion.

Radiographically, the intraosseous AOT has distinct features. It usually appears as a pericoronal well-circumscribed unilocular radiolucency or radiopaque-radiolucent mixed lesion with well-defined corticated or sclerotic border, usually surrounding an unerupted tooth, and may contain multiple minute variable-shaped calcifications or radiopaque foci, which may appear like a “cluster of small pebbles”. These calcified deposits are seen in approximately 78% of the lesions. Rarely the lesion manifests with no radiopaque component, as seen in our case. A distinct radio-opaque border of the unilocular radiolucent is a characteristic radiographic manifestation of AOT but multilocular variants have also been reported giving credence to the occurrence of multiple AOT. It is usually associated with the displacement of teeth and may contain fine calcifications. In our case, a well-defined unilocular radioluency surrounded by radio-opaque border with root resorption of 43 and 44 was present with no evidence of calcifications.

WHO has described the histological features of the tumor as “A tumor of odontogenic epithelium with duct-like structures and with varying degree of inductive changes in the connective tissue. The tumor may be partly cystic and in some cases the solid lesion may be present only as masses in the wall of a large cyst”. At low magnification, the most striking pattern observed is that of multi-sized solid nodules of cuboidal or columnar epithelial cells forming nests or rosette-like structures. Moreover, eosinophilic uncalcified amorphous material called “tumor droplets” can be found in these structures. Calcification was seen in the form of irregular masses, leisegang rings, spheroidal, and globular forms. Spindle-shaped or polygonal, closely opposed epithelial cells with dark eosinophilic cytoplasm, and round hyperchromatic nuclei fill in the spaces between the epithelial nodules. Conspicuous within the cellular areas are structures of tubular or duct-like appearance, the duct-like spaces are lined by a single row of low columnar epithelial cells, the nuclei of which are polarized away from the luminal surface. The lumen may be empty or contain a variable amount of eosinophilic material or cellular debris. Our case showed all the histopathological features of the classic AOT described in literature. Also seen in some areas were clear cells negative for PAS and mucicarmine staining. Some islands showed pools of amorphous amyloid-like material and globular masses of calcified substances. The amyloid-like substances showed positivity for Congo red staining. Some areas showed sheets of squamous-like cells in which intercellular bridges were not prominent. However, the presence of calcifying epithelial odontogenic tumor (CEOT)-like foci within an AOT does not influence its biological behavior or growth potential. CEOT-like areas occurring in AOTs should be considered a normal feature within the continuous histomorphologic spectrum of AOT. In our case, histopathology reveals the features of adenomatoid odontogenic tumor along with the foci of calcified epithelial odontogenic tumor. Lastly, all published cases of the CEOT/AOT variant show a biological behavior identical to that of AOT, i.e. a truly benign odontogenic lesion.

Conservative surgical enucleation or curettage is the treatment of choice with only rare recurrence (0.2%) Surgical enucleation of the lesion was performed under the local anesthesia in the department of Oral and Maxillofacial surgery. The patient is under a regular follow-up since last the one year and has no recurrence till the date.

Conclusion

It should be emphasized that extra-follicular variant of AOT with CEOT-like areas is very rare. Only careful diagnosis and adequate interpretation of clinical and radiographic findings may be helpful in arriving at a correct diagnosis; however, the treatment plan remains the same.

References

Kelgandre, et al.: Extra-follicular adenomatoid odontogenic tumor


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