CASE REPORT

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Extra-follicular Variant of Adenomatoid Odontogenic Tumor: A Diagnostic Enigma

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Abstract

Adenomatoid odontogenic tumor (AOT) is an uncommon, slow-growing, noninvasive odontogenic tumor mostly in the anterior maxilla with three well-recognized clinico-pathological variants: follicular, extra-follicular, and peripheral. Extra-follicular variant presents as a well-defined, unilocular, radiolucency in between, above, or superimposed on the roots of an erupted tooth. A 19 years female reported with the chief complaint of a loose tooth in the right front region of the upper jaw for 6 months, associated with firm swelling without pain or discharge. On orthopantomogram and cone-beam computed tomography, the lesion appeared as a single, localized, well-defined, roughly oval unilocular radiolucency with flecks of radiopacity integrating radicular and cervical third of 13. Complete surgical enucleation followed by histopathological examination revealed the lesion as AOT.

The extra-follicular AOT can cause diagnostic dilemmas and is often misdiagnosed as an odontogenic cyst.

Keywords: Adenomatoid odontogenic tumor; Cone-beam computed tomography; Extra-follicular; Swelling.

Declarations

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Adenomatoid odontogenic tumor (AOT) is a rare, benign and slow-growing well-defined tumor, representing 3-7% of all odontogenic tumors [1]. It may arise from epithelium of enamel organ and contain connective tissue elements which have been interpreted as dentin or enamel-like materials. Some consider AOTs to be benign and noninvasive neoplasms whereas others describe them as hamartomas [2].

Depending on its location and origin from the tooth, AOT has been classified as three different clinico-pathologic variants - follicular, extra-follicular, and peripheral type [3, 4]. Follicular variant (about 70%) is diagnosed earlier in life than extra-follicular and is associated with a crown or root of impacted permanent or supernumerary tooth displaying radiographic features of a well-circumscribed, unilocular radiolucent lesion [5, 6]. The extra-follicular type is a central lesion located in between, above or over the roots of an erupted tooth [4, 6, 7].The peripheral type is attached to the gingival structures with thinning or resorption of the cortical cortex and in some cases, tooth displacement without root resorption can be noted [6, 7].

Regarding management, conservative surgical excision is the treatment of choice as the tumor is well encapsulated, not locally invasive, and easily separable from the bone. The reported recurrence rate is approximately 0.2% [2]. We present the case of extra-follicular AOT as it can act as a diagnostic dilemma and is often misdiagnosed as an odontogenic cyst.

CASE

A 19 years female reported to the department with the chief complaint of a loose tooth in the right front region of the upper jaw for 6 months which was progressive in nature and was associated with swelling. It was not associated with pain and no other relevant history was present. On examination, 13 was grade 3 mobile and associated with single, localized, well-defined swelling, roughly oval in shape, approximately 3 cm × 2 cm in size. The swelling extended from the distal surface of 11 to the mid-buccal portion of 14 antero-posteriorly and crest of marginal gingiva to the depth of labial vestibule with respect to 13 supero-inferiorly with a smooth overlying surface, surrounding mucosa appeared normal **(Fig. 1)**. On palpation, the swelling was firm, non-tender and there was no discharge present. Oral hygiene appeared good and none of the teeth was carious. A provisional diagnosis of AOT with differential diagnosis of dentigerous cyst, odontogenic keratocyst, and ameloblastoma were made.

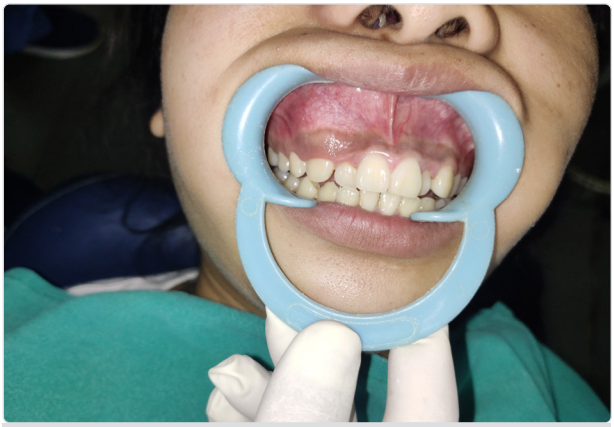

Figure 1: Initial presentation with swelling over maxillary right front region with respect to 12, 13 and 14.

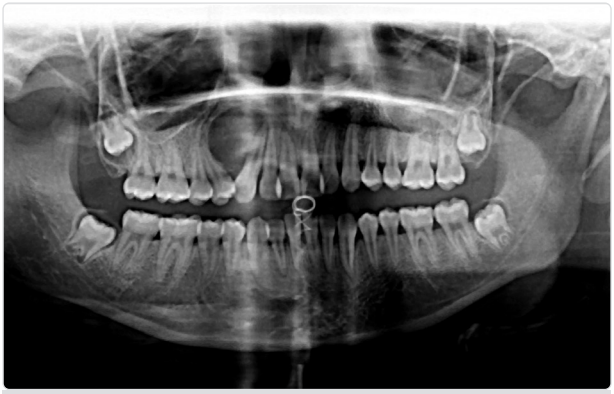

Figure 2: Orthopantomogram showing a single, well-defined, inverted pear-shaped radiolucency with foci of calcification (right maxilla)

The panoramic radiograph revealed a single, localized, well-defined, roughly oval, and unilocular homogenous radiolucency with flecks of radiopacity interspersed within radiolucency. The radiolucency seemed to integrate the radicular and cervical third of 13 **(Fig. 2)**.

The cone beam computed tomography (CBCT) revealed a single, localized, well-defined, roughly circular, homogenous radiolucency with a corticated outline extending between the periapical area of 12 and 14 antero-posteriorly. Inferiorly, the radiolucency extended from the crest of the alveolar bone between 12, 13, and 13, 14; superiorly extending about 9 mm from the floor of nasal fossa in 12, 13 region and 3.5 mm from the floor of maxillary sinus in 14 region. The buccal and palatal cortical plate was expanded and thinned out in 12, 13 and 14 region. Multiple foci of calcifications were present within the lesion. The root of 13 was displaced

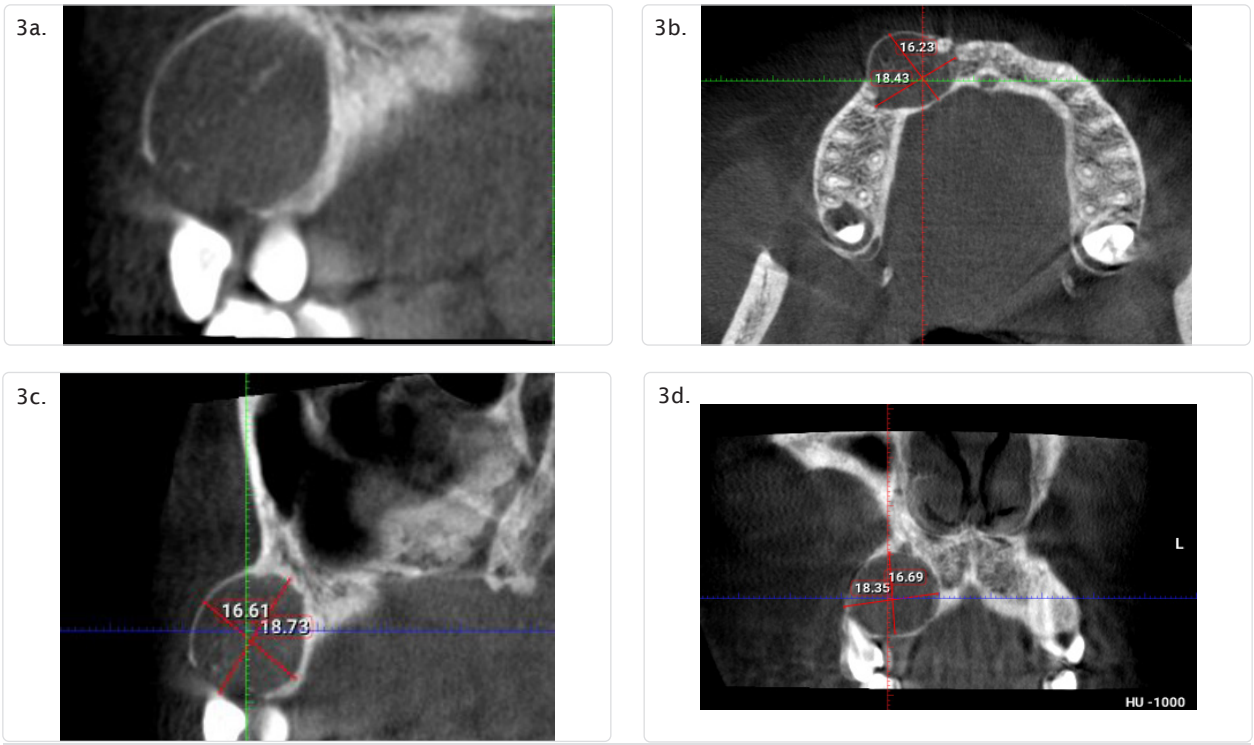

Figure 3: CBCT images showing maximum size of the lesion on different planes– (a) sagittal section showing foci of calcifications (b) axial section (c) sagittal section (d) coronal section.

mesially and that of 14 was displaced distally. The epicenter of the lesion appeared to be between the roots of 13 and 14. The maximum dimension of the lesion was about 18 × 16 mm in axial, coronal, and sagittal section **(Fig. 3)**.

The lesion was enucleated completely under local anesthesia and the tissue was sent for the histo-pathological examination which on gross examination revealed a soft, brown cystic mass measuring 22 mm × 18 mm × 7 mm in maximum dimension. Microscopically, cystic lining of odontogenic epithelium, two-three layers with localized area of tumor cells and foci of calcification intraluminally was seen along with fibrocellular capsule and inflammatory infiltrates predominantly of lymphocytes and plasma cells. Tumor cells were polygonal in shape with scanty cytoplasm and arranged in a sheet and whorled pattern. Numerous duct-like structures lined by cuboidal and tall columnar cells were also evident **(Fig. 4)**. Combining clinical picture, radiographic presentation, and histo-pathological presentation, the lesion was finally diagnosed as an extra-follicular AOT. The patient was followed up after two months. Clinically, there was no swelling present and the OPG showed a decrease in the size of the lesion **(Fig. 5)**.

DISCUSSION

First described in 1905, AOT has been described as pseudo-adenoameloblastoma and cystic adamantoma [1, 8, 9]. In 1948, Stafne et al. described it as a distinct entity while others believed it was a variant of ameloblastoma [8]. In 1971, the World Health Organization (WHO) adopted the name 'adenomatoid odontogenic tumor'. Subsequently, in 2003, the name 'adenomatoid odontogenic cyst' was coined [8]. It is also commonly known as 'two-third tumor,' as 2/3rd of cases occur in the maxilla, 2/3rd in young females, 2/3rd are associated with impacted teeth, and 2/3rd of the affected teeth are canines [10]. Various other terms used to describe this tumor are ameloblastic adenomatoid tumor, adamantinoma, epithelioma adamantinum, adenoameloblastoma, and teratomatous odontoma [8]. AOT is a non-invasive, benign odontogenic lesion, most commonly seen in the anterior maxilla of young females with a ratio of 1.9:1, similar to our case. An impacted maxillary canine is most commonly associated with AOT but in our case, the canine has erupted [8]. As seen in our case, the tumors are usually small, asymptomatic with the dimensions of 1.5 to 3 cm [7].

Radiographically, unilocular radiolucency

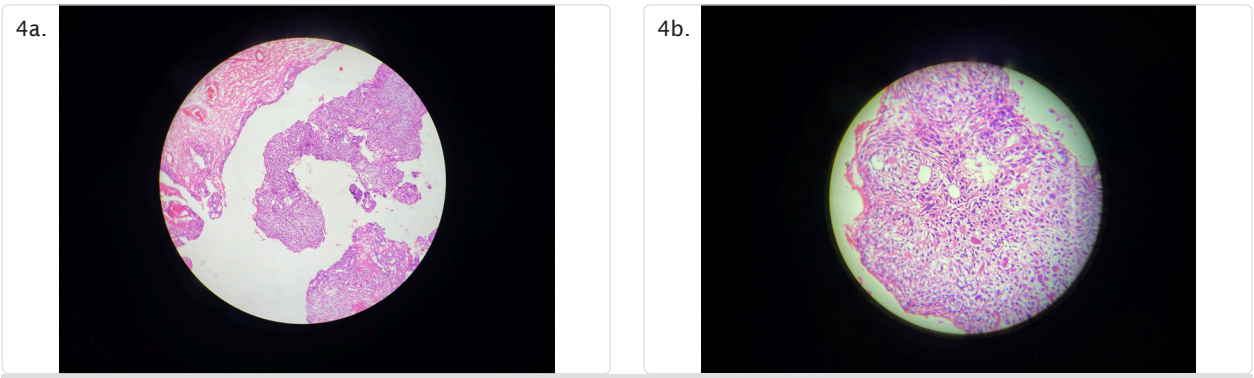

Figure 4: Histopathological presentation under (a) 4X and (b) 10X magnification showing tumor cells with scanty cytoplasm, arranged in sheet/ whorled pattern along with numerous duct-like structures and hyaline rings and drops.

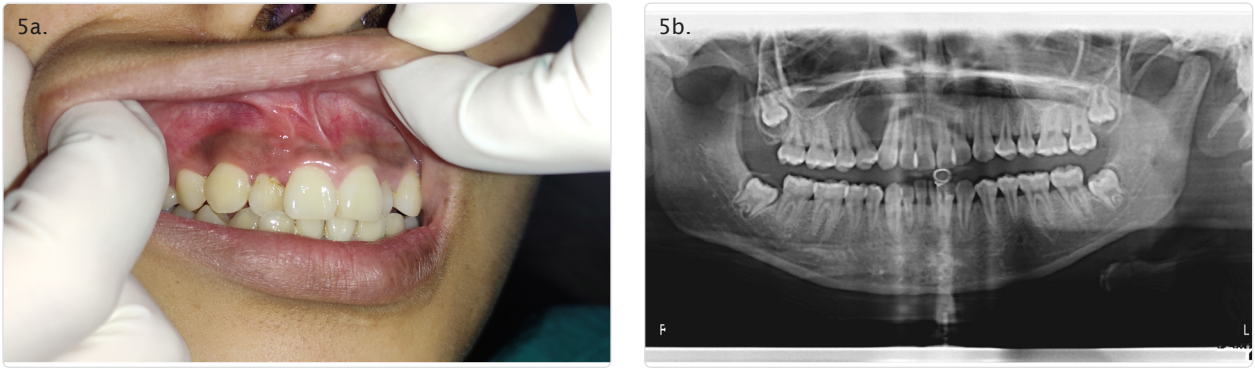

Figure 5: Follow up at 2 months: (a) clinical picture (b) OPG.

surrounded by a distinct radiopaque border is a feature of AOT [7]. It is usually associated with the displacement of teeth without root resorption. Approximately 78% of the lesions are associated with multiple, minute different shaped calcifications or radiopaque foci, appearing like a cluster of small pebbles [7]. Differentiating AOT from other lesions similar to it (e.g., calcifying cystic odontogenic tumors dentigerous cyst, unicystic ameloblastoma, and odontogenic keratocyst) via radiographs may be difficult. The ability of radiographs to show the radiopaque foci within a lesion is pertinent for the diagnosis of AOT. CBCT is beneficial modality in the case of small opacification or superimposed area in the anterior region, to demonstrate the detailed internal structures of lesions including radiopaque calcified foci [3].

Identical histological features are shown by all three variants of AOT [8]. The WHO has described the histological features of the tumor as a tumor of odontogenic epithelium with duct-like structures and with varying degrees of inductive change in the connective tissue. AOT is usually surrounded by a well-developed connective tissue capsule. The tumor cells are spindle-shaped or polygonal, forming sheets and whorled masses in a scant connective tissue stroma along with foci of calcification and characteristic duct-like structures lined by a single row of columnar cells, the nuclei of which are polarized away from the central lumen [10]. A similar histopathological picture was evident in our case.

Findings of our case were similar to other common presentations of AOT such as displacement of tooth without root resorption, presence of calcifications or radiopaque foci and spindle or polygonal-shaped tumor cells arranged in sheets or whorled mass with scanty cytoplasm as seen microscopically. Conservative complete surgical enucleation or curettage is the treatment of choice [5].

CONCLUSION

AOT in itself is not common; extra-follicular is rarer compared to other variants of AOT. Being aware of epidemiology, clinic-radiographic features, and histopathology, one can head to a definitive diagnosis even in the case of rare disorders, and thus the proper management.

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