Case Report

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Dentistry Section

Cystic Extrafollicular Adenomatoid Odontogenic Tumour: A Case Report and Pictorial Update

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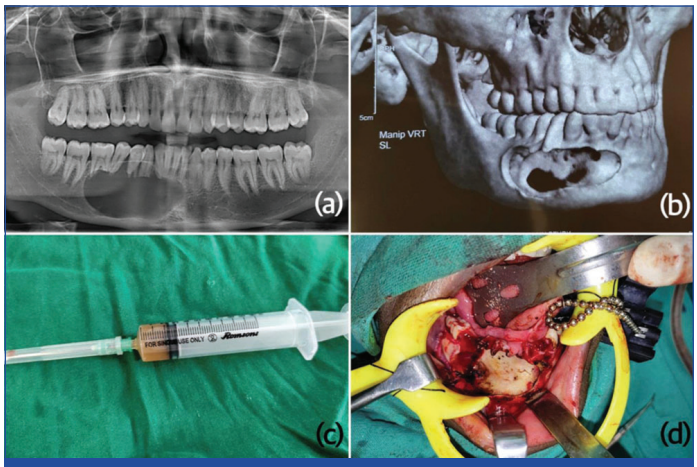
ABSTRACT

Adenomatoid Odontogenic Tumour (AOT) constitutes about 5% of all odontogenic tumours and is most commonly seen in young females, in association with impacted maxillary canines in a Dentigerous Cyst (DC) like relationship. Extrafollicular AOT is an uncommon variant seen unassociated with impacted teeth. Rare cystic presentation is reported with most cases seen arising from or in association with DC. Here, the authors present a case of extrafollicular AOT in a 29-year-old Dravidian male, who reported with a chief complaint of swelling of lower anterior jaw region. Radiographically, a well-defined radiolucent lesion was noted extending from mesial aspect of the roots of right mandibular second molar, crossing the midline to involve the body of mandible till mesial aspect of lower left canine. There was evidence of cortical perforation. The lesion was excised and sent for histopathological evaluation. Histopathologically, the lesion was diagnosed as cystic AOT-extrafollicular variant. There was adequate healing and no signs of recurrence or residual disease were noted 18 months after the surgery. The present case is a unique presentation of cystic extrafollicular AOT with pictorial demonstration and detailed explanation of its pathogenesis.

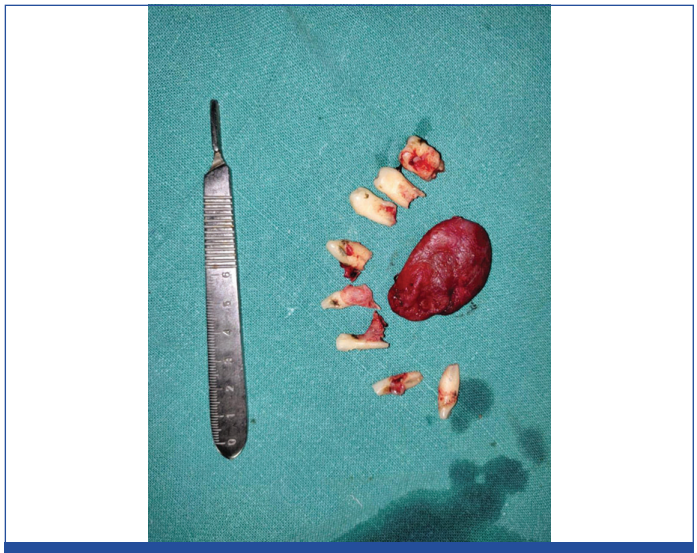
Keywords: Dentigerous cyst, Orthopantomogram, Protein

CASE REPORT

A 29-year-old Dravid Indian male reported with a chief complaint of swelling of lower anterior jaw region for one and a half years. The swelling was slow in onset with no associated pain or pus discharge. On intraoral examination, an ill-defined bony hard swelling was noted centered to the right anterior body of the mandible crossing the midline with buccal and lingual cortical expansion and obliteration of buccal sulcus. The swelling was non-tender, non-fluctuant, non-pulsatile and non-compressible with no associated bleeding or pus discharge. All the teeth were vital. The Orthopantomogram (OPG) showed a well-defined radiolucent lesion extending from mesial aspect of the roots of 47, crossing the midline to involve the body of the mandible till mesial aspect of 33 [Table/Fig-1a]. There was buccal and cortical plate perforation and concomitant root resorption. The findings were confirmed by Computed Tomography (CT) [Table/Fig-1b]. The lesion was provisionally diagnosed as developmental odontogenic cyst, however considering the bony expansion and root resorption the existence of odontogenic neoplasm could not be completely ruled out. Two mL of straw-coloured watery fluid was aspirated [Table/Fig-1c]; however, the cytological features were non contributory to the diagnosis. A total protein of 6.7 g per 100 mL was estimated. As the protein content was >4 g/100 mL, a non keratinising cystic lesion was considered and the lesion was thus enucleated under local anaesthesia and sent for histopathological examination [Table/Fig-1d,2].

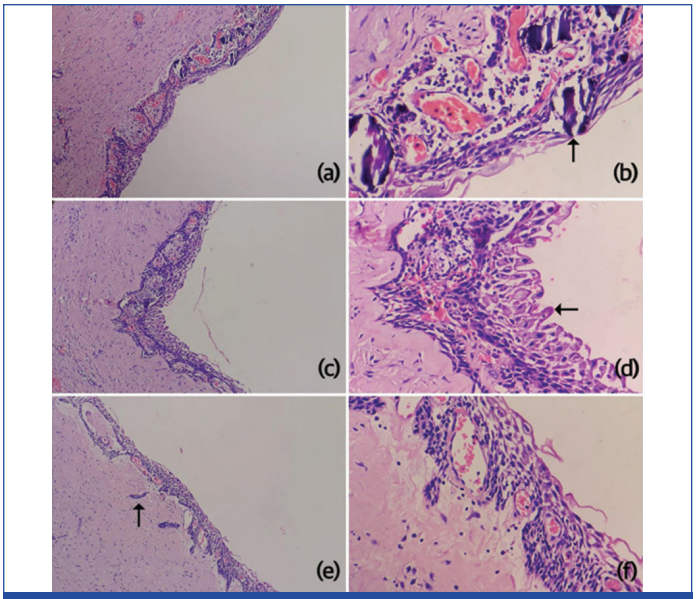


[Table/Fig-1]: Orthopantomogram (OPG) and Computed Tomography (CT) showing a,b) A well-defined unilocular radiolucency in relation to 46-33 region with cortical perforation and concomitant root resorption; c) Straw-coloured aspirate; d) Intraoperative presentation.



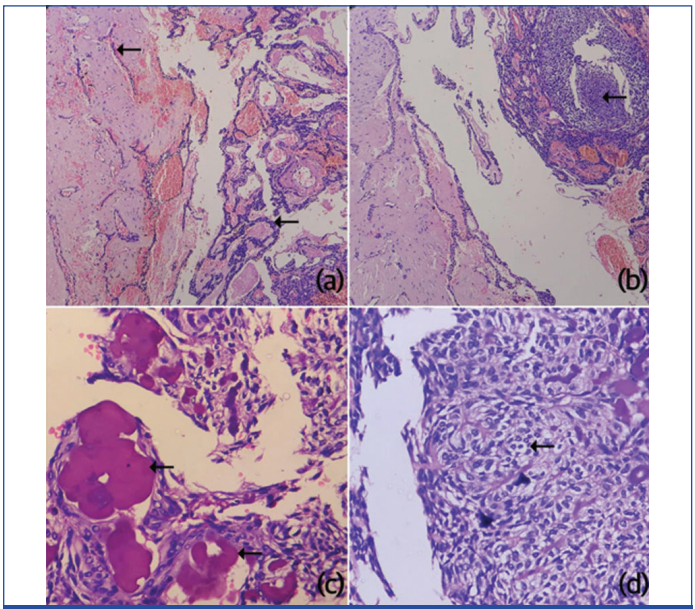
[Table/Fig-2]: Gross appearance of the lesion, a large cystic bag which was excised in toto.

Histopathological examination showed tissue which predominantly appeared cystic in architecture. The cystic lining was stratified squamous and lacked the classical features of any of the named odontogenic cysts and tumours such as Dentigerous Cyst (DC), Odontogenic Keratocyst (OKC), Calcifying Odontogenic Cyst (COC), Glandular Odontogenic Cyst (GOC) or unicystic ameloblastoma. The lining was predominantly non-keratinised, 4-6 layered thick with superficial loosening of cells and heaped up appearance of surface cells [Table/Fig-3a-d]. There was evidence of proliferation of thin strands and bud like extensions from the lining into the wall [Table/Fig-3e,f]. Basophilic calcifications were noted close to the lining [Table/Fig-3b]. At areas, the cyst wall showed hyalinisation/inductive changes particularly sub-epithelially and around the strands [Table/



[Table/Fig-3]: a,c,e) Photomicrographs showing variegated epithelial lining (H&E, 40X); b) Basophilic calcifications, d) Heaping of surface cells and f) Basal budding (H&E, 400X).

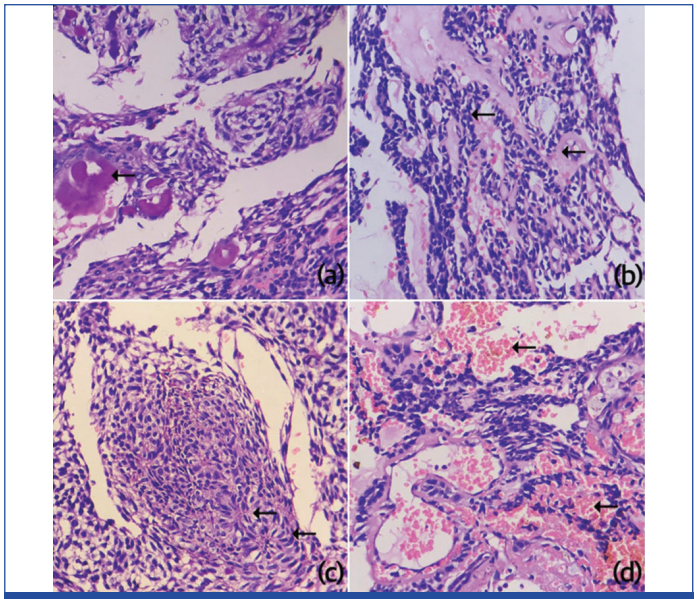
Fig-4a,b]. The other areas consisted of odontogenic epithelial cells arranged in plexus and solid nodules [Table/Fig-4c,d]. The cells were cuboidal/spindle/polygonal in shape with scanty eosinophilic to clear cytoplasm and deeply basophilic monomorphous round to oval nuclei. At areas, the tumour cells were arranged in pseudo rosettes and double convoluted structures with hyaline ring [Table/Fig-5]. Tumour/hyaline droplets were also seen at many areas [Table/Fig-4c,d,5a]. Ayoub-Shklar and periodic acid Schiff staining did not reveal ghost cells and mucous cells respectively. A final diagnosis extrafollicular cystic AOT was rendered. OPG taken on the seventh month after enucleation showed adequate healing and regeneration of lingual plate [Table/Fig-6]. The patient is under regular follow-up for past 18 months.



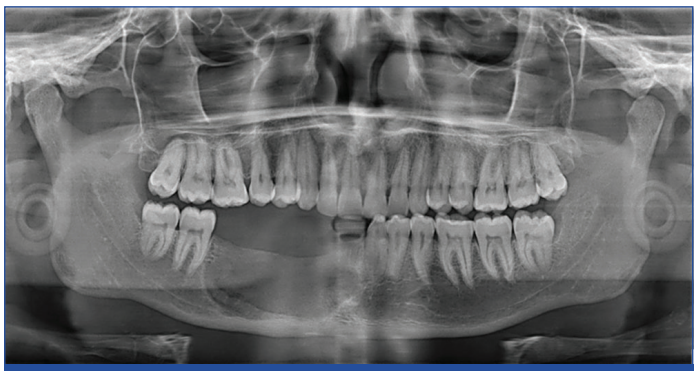
[Table/Fig-4]: Photomicrographs showing a,b) Proliferating odontogenic strands and solid tumour (H&E, 40X); c) Tumour droplets (H&E, 400X) and d) Solid nodules of clear and cuboidal tumour cells (H&E, 400X).

DISCUSSION

Odontogenic tumours are heterogenous pathologies derived from dental apparatus [1,2]. AOT, also known as 'two-third tumour' is an uncommon epithelial odontogenic tumour accounting for less than 5% of all odontogenic tumours, most commonly seen in association with impacted maxillary canines; the follicular type [3]. This variant radiographically resembles DC, and in fact, around 77% of follicular AOTs are diagnosed as DC initially [4].



[Table/Fig-5]: Photomicrographs showing a) Tumour droplets; (b) Plexiform arrangement of basaloid cells with inductive changes; c) Pseudo-rosettes and d) Extensive haemorrhage among tumour cells (H&E, 400X).



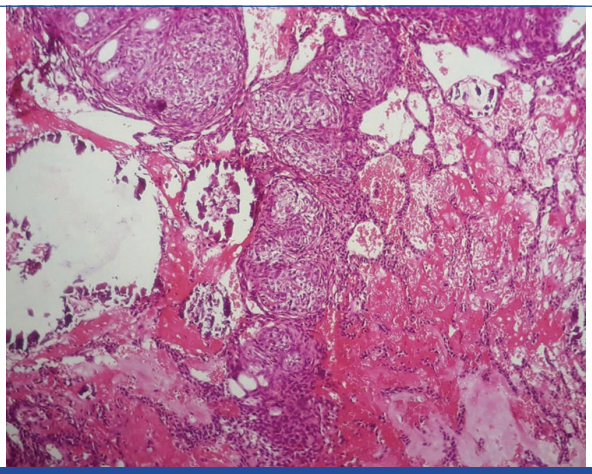
[Table/Fig-6]: Orthopantomogram (OPG) taken at 7th month follow-up with adequate healing and regeneration of cortical plates.

In one of the earliest literature reference, Harbitz F reported "cystic Adamantoma" in the year 1915 which was most probably the cystic presentation of AOT [4]. Extra-follicular subtypes are comparatively rare, seen unassociated with impacted teeth, and may be seen above, in between, superimposed to or unrelated to the roots of the teeth. The nature of AOT has been a never-ending debate. While some authorities consider it as a true neoplasm of odontogenic origin, others consider it as hamartoma with their own solid reasons [5]. Regardless, as of now, it is considered as a benign neoplasm and is classified under benign epithelial category, even in the latest World Health Organisation (WHO) classification of head and neck tumours [6].

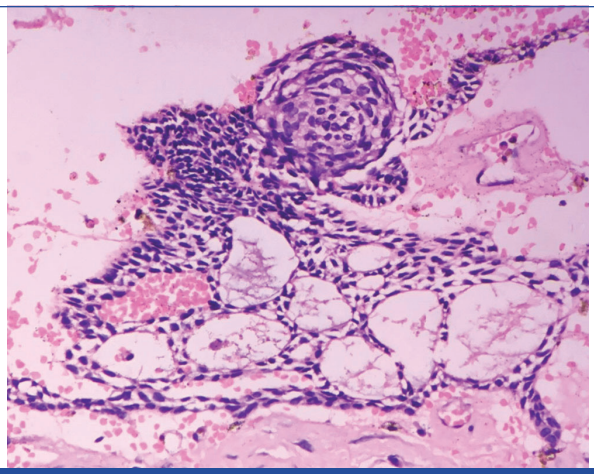
Cystic degeneration in solid tumours is not an uncommon histological feature and is commonly seen in many epithelial and mesenchymal tumours such as Pindborg tumour, cemento-ossifying fibroma and schwannoma to name a few [6-10]. Extensive degeneration is not unusual in ameloblastoma also. Literature search revealed 30 reported cases of cystic AOT; DC is the most commonly associated cyst [11-13]. The aim of the present paper is to report a unique case of cystic variant of AOT with special emphasis on the current understanding.

Gadewar DR and Srikant N described AOT as 'an intra-cystic proliferation of polygonal and spindle cells'. The term 'Adenomatoid Odontogenic Cyst (AOC)' was put forward by Marx and Stern [12]. These authors opined that lesion was cystic and the solid nature of AOT was due to intraluminal proliferation of the tumour cells which filled up the cystic space. Further, there are reported cases of AOT that arise either from the lining of DC or Calcifying Odontogenic Cyst (COC); unspecified cystic lining in many cases.

Cystic component is an indispensable histological feature of AOT [Table/Fig-7,8]; presumably can happen due to pooling of mucoid stroma resulting from the rupture of thin lattice like pattern (mainly seen at the periphery of the tumour close to the capsule) or development of AOT within or adjacent to a pre-existing odontogenic cyst, most commonly DC [5]. In the present case, we support the former possibility. As in multiple serial sections, DC like epithelial lining was not noted, neither the lesion was associated with any impacted teeth. The lesion was seen extra-follicularly apical to root apices of mandibular teeth ruling out the origin from pre-existing DC. COC, Glandular Odontogenic Cyst (GOC), Odontogenic Keratocyst (OKC) and unicystic ameloblastoma were also excluded histopathologically.In available literature, description of cystic AOT pertains mainly to AOT seen in association with DC [12,13]. DC develops by accumulation of fluid between the Reduced Enamel Epithelium (REE) and the unerupted tooth crown, and separation of DC from hyperplastic follicle is many a time very subjective [14]. The REE is attached to the Cemento-Enamel Junction (CEJ) in contrast to the AOT where the attachment is noted on the radicular portion raising another query whether all the previous reported cases with DC like lining were transformation of DC to AOT or just represents the basaloid cells that form the plexiform pattern. We support the latter hypothesis as in multiple serial sections we also demonstrated bud-like or strand-like extensions into the adjacent stroma.



[Table/Fig-7]: Photomicrograph of conventional AOT showing cystic degeneration with mucoid to flocculent material in cystic spaces (H&E, 100X).



[Table/Fig-8]: Photomicrograph showing breakdown of peripheral plexus of basaloid spindle cells with extensive haemorrhage (H&E, 400X).

In contrast to classical AOT, cystic variants were found to be statistically significant larger in size [13], which is explained in the subsequent section. Generally, AOT has an indolent course with no sign of recurrence or persistent growth even after incomplete surgical removal. In one of the earlier analytical reports, Philipsen HP et al., reported a protein content of 5.2 g/100 mL and 7 g/100 mL in one case each of follicular type and extra-follicular type (of AOT) respectively [15]. These protein content values were within the range provided for the cystic protein content of non-keratinising odontogenic cysts (5-11 g/100 mL) in contrast to lower range in keratinising cysts (<3.5 g/100 mL) [15]. The protein content in the present case was close to the extrafollicular AOT reported previously [16]. The values have further been correlated with the nature and direction of expansion (owing to the osmotic pressure), serum osmolarity, lymphatic drainage, nature of globulins and the internal hydrostatic pressure of the cysts. In OKCs, the bony destruction happen in anteroposterior direction at the expense of marrow space while other cysts cause buccal and lingual cortical expansion akin to the present case. This has been widely described in the literature. Definitely, the role of cyst wall cannot be overlooked which through various collagenolytic enzymes, prostaglandins, interleukins, and many more, dictates the clinical behaviour of any lesion. It was demonstrated that in contrary to collagen in the connective tissue wall of radicular cyst and DC, the stroma of AOT consists of poorly packed or pathologic collagen which resembled the connective tissue of OKC wall [17]. The extensive expansion and root resorption in the present case, unlike conventional solid AOT with minimal cystic component could thus be due to the cystic degeneration of the solid component with further expansion owing to Toller's theory and unique connective tissue components of the cyst wall [16].

It is therefore more logical to consider the cystic degeneration as secondary phenomenon. Such lesions may behave aggressively as compared to solid variants and may be regarded as cystic variant of AOT rather than AOC. Furthermore, these lesions should be delineated from the AOT arising in the wall of odontogenic cysts. Cystic variants of AOT are documented in reported cases of follicular and extraosseous AOT. The extra-follicular nature of the lesion in the present case thus supports the existence of cystic variant of AOT.

CONCLUSION(S)

Although rare, cystic variant of AOT should be considered in the differential diagnosis of radiolucencies of the anterior mandible. Further, this variant deserves delineation from AOT arising from the wall of DCs. The authors believe that cystic degeneration is a secondary phenomenon. Conservative enucleation is the treatment of choice.

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