

Odontoameloblastoma with Calcifying Cystic Odontogenic Tumor of Mandible - Report of a Rare Case

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Abstract: *Odontoameloblastoma is a very rare Odontogenic tumor with few reported cases. Its association with calcifying cystic Odontogenic tumor is rarer with only three reported cases in the English language literature according to the author's knowledge. The terminology and classification of these tumors are very controversial. The presence of few reported cases and lack of strict criteria makes the diagnosis of these lesions very challenging. We report a rare case of Odontoameloblastoma associated with calcifying cystic Odontogenic tumor that occurred in the right posterior mandible of a 14-year-old girl.*

Keywords: Odontoameloblastoma, Calcifying cystic Odontogenic tumor, Ghost cells

1. Introduction

Odontoameloblastoma is a very rare odontogenic tumor with less than 50 reported cases of which many have been reclassified as different entities¹. It was first reported by Thoma et al in 1944. It has features of both ameloblastoma and odontoma^{1,2}. An outstanding feature of this tumor is the presence of sheets of typical ameloblastoma of one or another recognised types usually basal cell, follicular or plexiform which is intermingled with dental tissues with varying degrees of maturity as seen in Odontomes^{1,2,4}. In the present case this solid tumor was associated with Calcifying cystic odontogenic tumor.

Calcifying cystic odontogenic tumor (CCOT) is the new name for Calcifying Odontogenic Cyst (COC) [2] which was first reported by Gorlin et al in 1962. Many controversies exist about its nomenclature and classification⁴. It is known for its association with various odontogenic tumors^{3,4}. It accounts for 1% of all odontogenic tumors. Its association with Odontoameloblastoma is very rare. According to the author's knowledge only three cases have been reported in the English language literature.

We present a case of an Odontoameloblastoma associated with calcifying cystic odontogenic tumor which showed unusual histopathological features simulating various other ghost cell lesions and discuss its differential diagnosis. The steps taken to establish the final diagnosis are also described.

2. Case Report

A female patient aged 14 years reported with a chief complaint of swelling in the right side of the face since 7 years. Six month's later swelling and pain started. Pain was moderate & intermittent. Patient reported of salt watery discharge since 3 years, pus discharge since 8 months and difficulty in mastication & reduced mouth opening since 2 months. Past history was not contributory.

On examination the patient was moderately built and was alert and attentive. Extra orally a diffuse swelling was present on the right side of the face over the mandible. On palpation swelling was tender and hard in consistency. The mouth Opening was 2 cms. Intra orally the right lower

buccal vestibule was obliterated by the swelling, the second and third molars on the right side were missing and the first molar was lingually inclined with grade II mobility. A sinus tract with pus discharge was noted in the retromolar area.

Radiographic examination included Occlusal view, PA view and OPG (fig 1). OPG revealed a radio opacity surrounded by a radiolucent rim occupying the posterior part on the right side of the mandible extending from the second premolar anteriorly to the anterior part of the ramus posteriorly; with displacement of the second premolar and first molar roots. Alveolar bone around the first molar was completely resorbed. The lesion was associated with an impacted tooth which was displaced to the lower border of the mandible. Both Occlusal and PA views showed cortical plate expansions.

A provisional diagnosis of calcifying epithelial odontogenic tumor was made and the differential diagnosis of Odontoma and calcifying odontogenic cyst were considered. The lesion was completely excised and the specimen was sent for histopathological examination.

Specimen on grossing revealed hard tissue bits attached to soft tissues which on radiographic examination showed calcified masses. The tissue was subjected to surface decalcification before sectioning.

On histopathological examination initial sections revealed numerous globular basophilic calcifications in a mature fibrous connective tissue stroma. (Fig 2) Eosinophilic calcifications surrounded by basophilic rimming (simulating liesgang rings) were also seen (fig 3). Deeper sections revealed a variety of cells and tissues in complex distribution including, typical ameloblastoma like follicles which showed cystic spaces, palisaded ameloblast like cells, and stellate reticulum like cells. (The ameloblast like cells followed the Vikers and Gorlin criteria which include polarised palisaded basal cells showing hyperchromatism and cytoplasmic vacuolisation)⁵ (fig 4); Cords, rosettes and nests of Odontogenic cells (fig 5); dentinoid associated with Odontogenic cells (fig 6); Complex odontome like areas consisting of enamel spaces, dentinoid and small areas of primitive ectomesenchyme mimicking dental papilla numerous ghost cells associated with dentinoid which were

confirmed by Van Gieson staining where the ghost cells were stained yellow and the dentinoid was stained red in colour (fig 7) were seen in the mature stroma. A cystic lining consisting of a few layers of odontogenic cells associated with dentinoid and ghost cells was also visualised (Fig 8).

The clinical diagnosis of this lesion was calcifying epithelial Odontogenic tumor but this was ruled out due to the lack of sheets of epithelial cells and negative staining with congo red for amyloid. Based on the microscopic features the diagnostic hypothesis considered were Odontoameloblastoma, Calcifying Cystic Odontogenic tumor (CCOT), Dentinogenic ghost cell tumor (DGCT) and Ameloblastic fibro odontoma which show presence of dentinoid and ghost cells. To arrive at a definitive diagnosis the clinical, radiological and histopathological features of our case were compared with that of all the four lesions as shown in tables 1 and 2.

3. Discussion

According to table 1 all clinical and radiological features of the present case matched with those of odontoameloblastoma and dentinogenic ghost cell tumor.

The presence of varied histopathological features (table 2) made the diagnosis very challenging. Presence of enamel spaces and cystic lining ruled out DGCT and unequivocal ameloblastic follicles and mature stroma ruled out ameloblastic fibro odontoma²⁻⁴. Odontoameloblastoma matched with all the features of the present case except for the presence of cystic lining²⁻⁴. Hence hybrid lesion of odontoameloblastoma with a cystic lesion either dentigerous cyst or CCOT was considered.

Presence of only a few layers of odontogenic cells in the cystic lining and association with an impacted tooth favored dentigerous cyst³, but the presence of dentinoid and ghost cells associated with the lining ruled out dentigerous cyst and favoured CCOT though the cystic lining did not show classical ameloblastoma like epithelium. The lack of classical ameloblastoma like epithelium in the lining may occur in non proliferative type of COC (CCOT) as classified by Hong et al and Riechart and Philipsen⁴, thus a final diagnosis of Odontoameloblastoma associated with CCOT was made.⁶⁻⁸

The incidence of odontoameloblastoma is very rare. According to Masqueda Taylor et al only 14 cases reported in literature met the WHO histological and clinical criteria for odontoameloblastoma⁹. Dean and White have questioned the existence of these lesions as it is based on a few case reports and supporting documentation is marginal and not confirmatory¹⁰. This lesion is unusual in that relatively undifferentiated neoplastic tissue is associated with highly differentiated tissue both of which show recurrence after inadequate removal¹¹.

CCOT is reported to be associated with other Odontogenic lesions including Adenomatoid Odontogenic tumor (AOT), ameloblastoma, odontoma, ameloblastic fibro odontoma and odontoameloblastoma. These are not included in the WHO 2005 classification, but are included in the suggested

classification of Pretorius (2006)⁴ and international collaborative study on ghost cells of (2008)¹². Classifications of by Li and Yu^{4,12}, Hong⁴, Buchner⁴ and Toida⁴ have also included association of Calcifying Odontogenic Cysts with other Odontogenic tumors.

Ide et al have stated that combined lesions of CCOT represent only variation in the inductive capacity of odontogenic epithelium rather than true hybrid nature¹³. They have also questioned the clinical significance of these combined lesions¹³. According to Li and Yu¹², when combined lesions of CCOT are present they are named according to the associated tumor and behave accordingly. The potential for odontoameloblastoma to recur is well known with 3 cases out of 14 showing recurrence^{1,9}. The designation of the present lesion as Odontoameloblastoma associated with CCOT will better alert the surgeon about the behaviour of this tumor. The lesion should be followed for at least five years. The present case has not shown any recurrence after 1 year of follow up.

4. Conclusion

Diagnosis of Odontogenic tumors by clinical and radiographic features alone is not possible and careful histopathological assessment of specimen with deeper sections and correlation with clinical and radiographical features are necessary for arriving at the right diagnosis, which helps the surgeon in correctly treating and following the case. Odontogenic tumors with ghost cells present with varied histopathological features. Lack of literature on exact features of these lesions and controversies in the classification of CCOT and terminology complicate the diagnosis. Documentation of more Odontogenic lesions with ghost cells will help in resolving the controversies and help to formulate strict criteria for definitive diagnosis.

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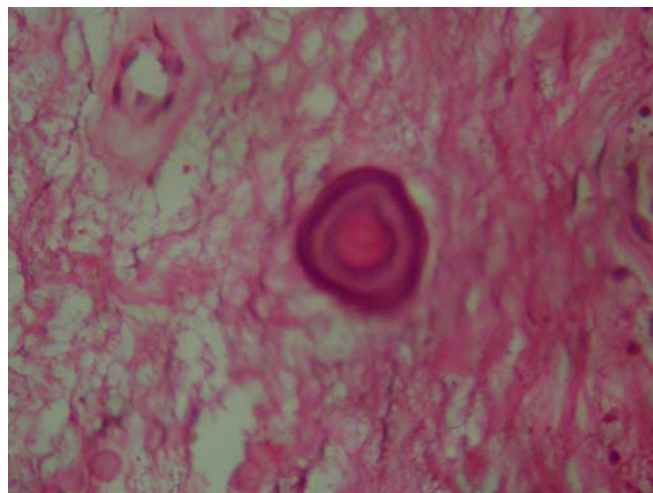


Figure 3: Calcifications simulating liesgang rings (Original magnification 40X).

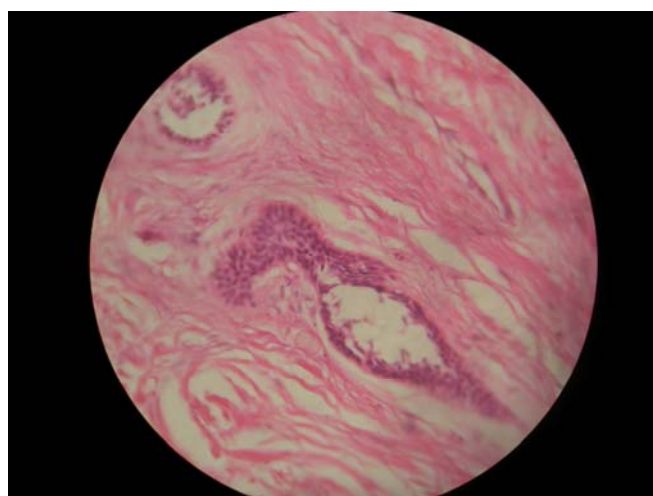


Figure 4: Unequivocal ameloblastoma with follicles showing palisading of basal cells, cystic spaces and stellate reticulum like cells are seen (Original magnification 40X).

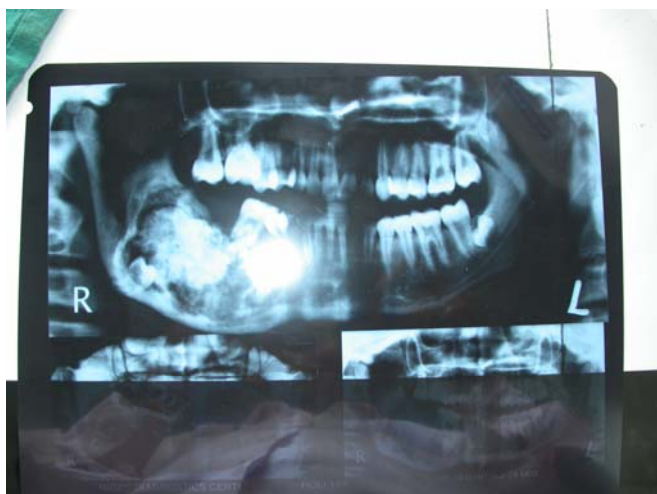


Figure 1: OPG--Mixed lesion with radio opacity surrounded by radiolucent rim, associated with an impacted tooth and causing displacement of second premolar and first molar roots on the right side.

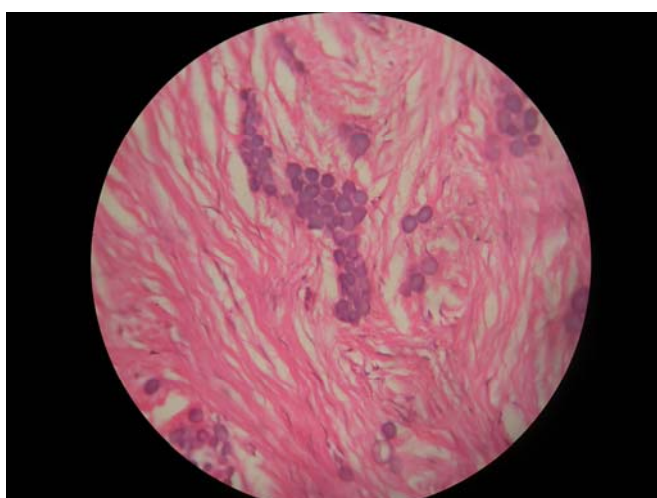


Figure 2: Numerous basophilic globular calcifications in mature fibrous connective tissue stroma . (Original magnification 10X)

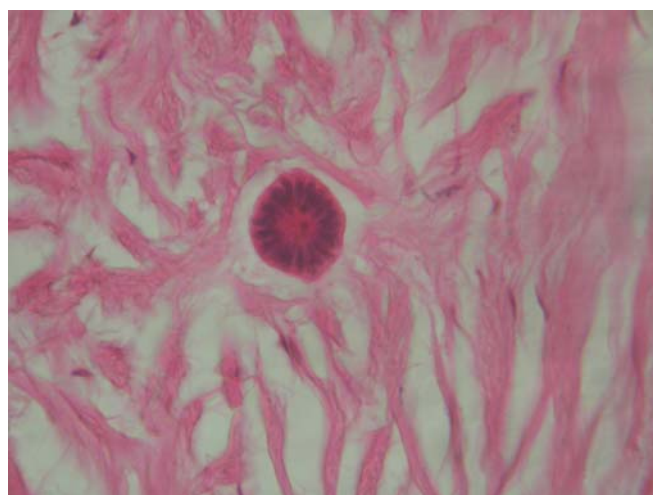


Figure 5: Rosette of odontogenic cells (Original magnification 40X).

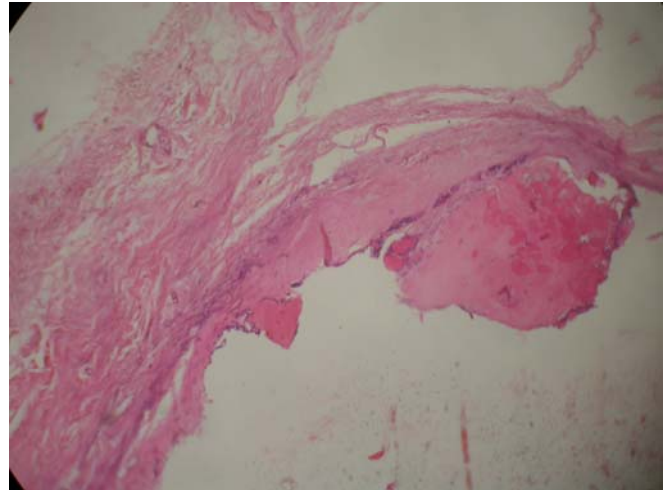
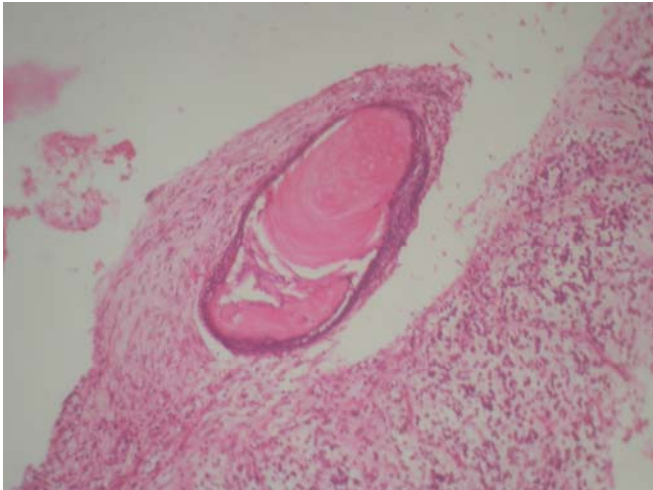


Figure 8: Cystic lining showing 2-3 layers of odontogenic cells associated with dentinoid and ghost cells characteristically lack classical ameloblastoma like epithelium H & E (Original magnification 10X)

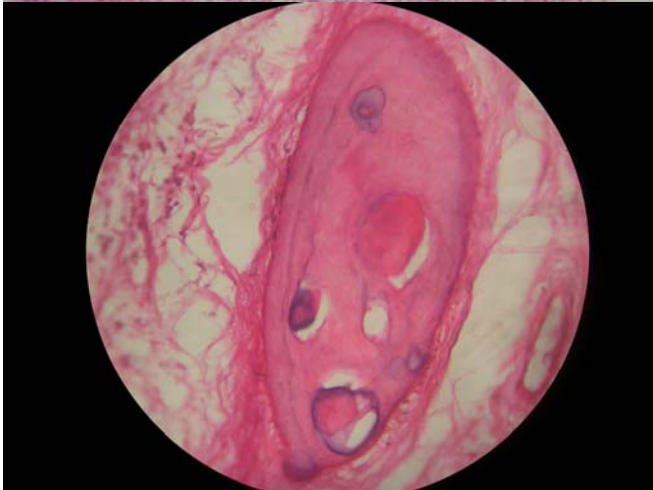


Figure 6: Areas of Dentinoid formation(Original magnification 40X).

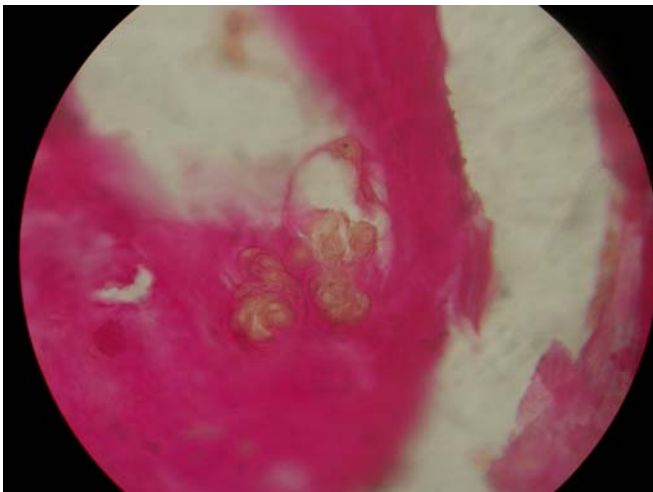


Figure 7: Ghost cells stained yellow and Dentinoid stained red Van Gieson (Original magnification 40X).