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Odontoma-associated calcifying cystic odontogenic tumour: Report of a case

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ABSTRACT

The calcifying cystic odontogenic tumour (CCOT) is an uncommon benign neoplasm of odontogenic origin, which was first described by Gorlin. It has been shown to have extensive diversity in its clinical and histopathological features and its biological behaviour. Most of these cases present with cystic characteristics, but a few are of the solid type.

Cystic variant of CCOT associated with odontoma is considered as rare variant. A case of calcifying cystic odontogenic tumour associated with odontoma is reported in an 11-year-old female patient. In this report, the lesion was very large extending from the midline up to the right tuberosity region and impeded the eruption of five permanent teeth. Through this article, the radiographic and histopathological features of this entity are discussed and the literature is reviewed.

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1. Introduction

Calcifying odontogenic cyst (COC) or Gorlin's cyst is an uncommon benign odontogenic lesion. It was first described by Rywkind[1] in 1932 as a variant of the cholesteatoma, while Maitland[2] in 1947 regarded it as a type of ameloblastoma. It was first distinguished as a separate entity by Gorlin et al. in 1962 who labelled it as the calcifying odontogenic cyst.[3,4] The World Health Organization (WHO) in 2005 has defined the entity as a benign cystic neoplasm of odontogenic origin, characterized by an ameloblastoma-like epithelium with ghost cells that may calcify and suggested the preferred terminology as Calcifying cystic odontogenic tumour (CCOT).[5]

Praetorius et al. have suggested an odontogenic origin of this entity. It may develop in the dental follicle, gingival tissue or bone, from remnants of either odontogenic epithelium or reduced enamel epithelium.[6] The etiopathogenesis and molecular pathology of these tumours is, however, still controversial. According to Shear, CCOT associated with features of odontogenic tumours either develop these secondarily, or they may themselves be secondary phenomena in

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pre-existing odontogenic tumours.[7] According to Abrams and Howell, this may be attributed to the multi-potentiality of the odontogenic epithelium.[8] CCOT in association with an odontoma results from the processes of differentiation and degeneration of the odontogenic epithelium. The dysplastic dentine formation in CCOT results from induction changes in the mesenchymal tissue by the odontogenic epithelium.

CCOT includes various subtypes that show diversity in clinical and histopathological features as well as in biological behaviour. While most cases present cystic characteristics, few are of the solid type (15%) and its rare malignant transformation is well documented.[5,9] The CCOT is considered as a rare lesion and comprises of 0.37–2.1% of all odontogenic tumours. Cystic variant of CCOT associated with odontoma is further considered as a rare variant and accounts for only 20% of these lesions.[10]

We report a case of cystic variant of CCOT associated with a complex odontoma in an 11-year-old female patient in the right maxillary region. In this report, this lesion was very large extending from the midline up to the right tuberosity region and impeded the eruption of five permanent teeth.

2. Case history

An 11-year-old female reported with the complaint of intermittent dull pain in right anterior maxillary region since 3 years. Around three months back, the patient noticed a gradually increasing swelling in the same area. The patient also complained of missing upper right anterior teeth. No history of trauma or exfoliation of teeth was present. There was no history of paresthesia or nasal blockage. The medical and family history of the patient was insignificant.



Fig. 1 – Extraoral photograph of the patient showing diffuse swelling on right side.



Fig. 2 – Intraoral photograph showing swelling and missing teeth in upper anterior region.

There was facial asymmetry noted extra-orally. An ovoid, firm swelling of about 5 cm × 5 cm in size with diffuse margins was present on the right side of the face. It was non-tender, non-fluctuant and the overlying skin appeared normal. It extended supero-inferiorly from right infra-orbital margin to the ala tragus line and antero-posteriorly from a line along the right ala of nose to a line along the outer canthus of eye. There was no evidence of trismus or lymphadenopathy (Fig. 1).

Intra-orally, there was a swelling with buccal and palatal expansion extending in the right maxillary posterior region. There was absence of permanent upper right central incisor, lateral incisor and canine. The deciduous upper right lateral incisor and canine were retained. Grade I mobility was noted in upper right deciduous molars. Palpation revealed a soft-to-firm consistency with perforation of labial cortex in the region of the swelling. The swelling was non-fluctuant, non-tender and the overlying mucosa appeared normal (Fig. 2). The clinical examination was suggestive of dentigerous cyst or a benign tumour like adenomatoid odonogenic tumour (AOT) as the probable diagnosis.

The intra-oral periapical radiograph showed retained deciduous lateral incisor, canine and molars (Fig. 3). Upper right permanent lateral incisor was impacted and surrounded by pericoronal radiolucency within the lumen of which radiopaque masses were seen. The degree of radiopacity of these masses resembled that of a tooth. Panoramic radiograph demonstrated the complete extent of the lesion extending from the midline of the maxilla to the tuberosity region posteriorly (Fig. 4). A large radiolucency of about 10 cm, with well-defined corticated borders, was seen on the panoramic radiograph in the right maxillary region in close proximity to the orbit. Right maxillary canine not visualized on the intra-oral views was found to be displaced close to the tuberosity

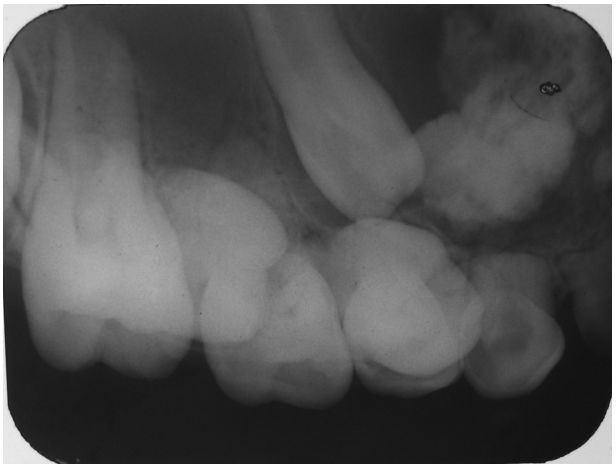


Fig. 3 – Periapical radiograph showing impacted lateral incisor with a pericoronal radiolucency and radiopaque masses within.

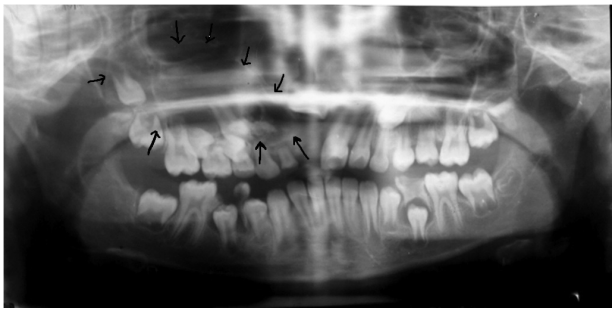


Fig. 4 – Panoramic radiograph showing the large cystic lesion on right side of maxilla.

region. The right central incisor was also impacted and pushed apically. The occipitomental view showed haziness in the right maxillary sinus and the walls of the sinus were intact.

The radiographic appearance was suggestive of a dentigerous cyst with some associated odontoma-like lesion. Adenomatoid odontogenic tumour, calcifying epithelial odontogenic tumour, calcifying cystic odontogenic tumour and ameloblastic fibro-odontoma were considered in the differential diagnosis. The presence of straw coloured fluid with cholesterol crystals on aspiration was indicative of a cystic lesion.

CT (Computed tomography) showed the lesion with well-defined hyperostotic borders and loose hyperdense masses within the lumen resembling teeth (Fig. 5). Enucleation of the lesion was done and sent for histopathological examination. Haematoxylin and eosin (H & E) stained sections showed the presence a cystic epithelial lining with areas of fibrous tissue. There was presence of ameloblastoma-like epithelium in the basal layer with eosinophilic ghost cells and multiple foci of calcifications (Fig. 6). Strands and cord of proliferating ameloblastomatous cells were not present in the cystic lining, thus ruling out dentinogenic ghost cell tumour (DGCT). These features were indicative of cystic variant of CCOT associated with a complex odontoma.



Fig. 5 – Axial CT showing the extent of the lesion.

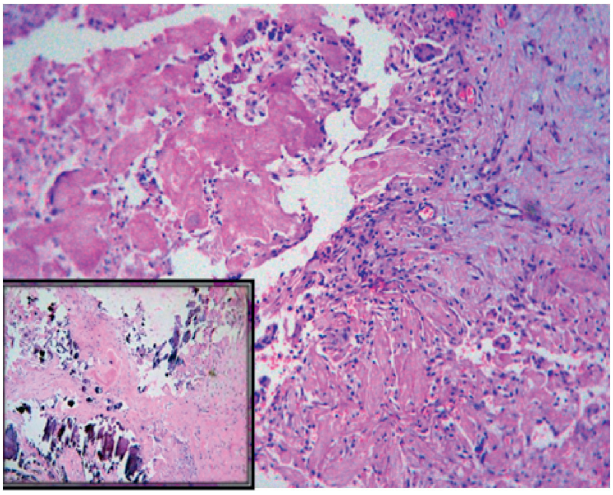


Fig. 6 – Histopathological examination with H& E staining (10X) showing innumerable ghost cells. Inset shows areas of calcifications.

3. Discussion

There has been an extended debate about the biologic behaviour of these lesions and many nomenclatures and classifications have been published towards this effect. Although various classifications have been discussed to distinguish the cystic form from solid variants, none has been universally accepted.[5,6]

Both monistic and dualistic concepts have been proposed for this entity. According to the monistic concept, all COCs are neoplastic in nature although the majority are cystic in architecture and appear to be non-neoplastic. Sauk proposed

that this entity should be considered like a tumour rather than a cyst and that the osteoid and dentinoid formed are derived by the developing epithelium.[11] In order to eliminate the problems of previous classifications, Toida has proposed a simple and basic classification based on ''dualistic'' concept. This concept regards CCOT as a cyst as well as a neoplasm.[12]

In the histologic classification of odontogenic tumours by WHO, COC was categorized into three types, one cystic type, the calcifying cystic odontogenic tumour (CCOT), and the two solid types, the Dentinogenic ghost cell tumour (DGCT) and Ghost cell odontogenic carcinoma (GCOC).[5] The WHO has classified CCOT as a neoplasm.[10]

CCOT mainly occurs as an intra-osseous lesion but the peripheral (extra-osseous) lesion has also been reported. The extra-osseous CCOT is a localized sessile or pedunculated gingival mass and comprises 13–25% of the CCOTs.[10] Both central and peripheral forms occur with equal frequency in the maxilla and mandible.[9] Buchner found an almost equal gender distribution.[13]

Praetorius et al. reported a bimodal age distribution, with peaks in the second and sixth decades.[6] The odontoma-associated type occurred at an earlier age (average age 17 years), while the neoplastic variant was more common in older patients. This case is of an 11-year-old female patient with multiple unerupted teeth involving the permanent dentition.

The central CCOT presents as an asymptomatic hard swelling of the jaws, which produces expansion rather than erosion of bone. Pain indicates secondary infection. In our case, the patient initially experienced a swelling and later on it was associated with dull pain. Early lesions are usually detected following routine radiographic examination and they are often associated with an unerupted tooth.[10] It has been noted that the lesions usually appear anterior to the first molar region and about 65% of CCOT's are reported to occur in the incisor/canine region. Hirschberg et al. have concluded that the upper jaw was affected in 61.5% and the anterior region of the jaw in 75% of the reported cases.[14] In this case, the lesion was detected late and had involved the entire right maxilla and was associated with displacement of five unerupted permanent teeth. Lesions present in the maxilla may present with nasal stiffness, epistaxis and headache.[15] Our patient did not give history of any such symptom.

Radiographically, CCOT has been found to have a varied radiographic presentation ranging from a unilocular radiolucency to a multilocular one. Moreover, it may be associated with odontoma and may or may not show calcifications.[4] Root resorption or divergence of adjacent teeth may be present. The association with the apices of teeth reveals an incidence of root resorption in 75–77% of cases.[5,6] There was no associated root resorption noted in our patient. CCOT may have an association with an impacted tooth, which occurs in approximately one-third of the cases. These features further suggest the diverse radiographic presentation of CCOT.

Diagnosis of this lesion especially in the maxillary region becomes more challenging using only conventional images. CT evaluation made the radiographic examination in this present case more comprehensible. CT gave additional three-dimensional information about the anatomy of the lesion and its relationship with adjacent structures. CT can also characterize a lesion as cystic or solid by virtue of Hounsfield attenuation values.[4] This case present was challenging from diagnostic as well as with the treatment point of view.

In the case presented here, the patient's age and site were in favour of the diagnosis. A positive aspiration further helped in the diagnosis of a cystic lesion. The Hounsfield units of the lesion on CT examination were also suggestive of a cystic lesion. Presence of odontome-like bodies within the lesion helped to shortlist the probable differential diagnosis for the lesion. The lesion was large enough to involve the entire right maxillary quadrant impeding the eruption of five permanent teeth.

The definitive diagnosis of CCOT is made histologically, due to the lesion's lack of characteristic clinical and radiologic features, as well as its variable biological behaviour. The most notable features of this pathologic entity are the eosinophilic ghost cells within the epithelium, with a tendency to calcify. The basal cells are cuboidal and resemble stellate reticulum of an ameloblastoma. There may be association of this entity with certain odontogenic tumours, including the odontoma and ameloblastoma occasionally.[3–5]

Treatment of the cystic variety of CCOT is usually conservative and consists of enucleation followed by removal of a 1- to 2-mm layer of bone around the periphery of the cystic cavity with a sharp curette or a bone bur so as to remove any epithelial remnants and prevent recurrence.[15] The prognosis of CCOT is considered to be good and only occasional recurrences have been reported. In this case enucleation of the lesion with proper curettage was done to prevent any recurrence. The patient was on a regular follow-up to check for any recurrence.

4. Conclusion

This rare entity of odontogenic origin has been a topic of debate regarding its classification as a cyst or tumour. The CCOT lacks pathognomonic clinical, radiological and CT features and shows variable biological behaviour. It should be included in the differential diagnosis of cystic lesions that present anterior to the first molar teeth. The definitive diagnosis and treatment of this lesion remains dependent on histological evaluation.

Conflicts of interest

The authors have none to declare.

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