Calcifying Epithelial Odontogenic Cyst: A rare case

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Abstract:
Calcifying odontogenic cyst (COC) is a very rare developmental odontogenic cyst. It was first described as a separate entity by Gorlin et al in 1962. COC has been categorized into 2 basic groups: cystic and neoplastic. It is characterized by the presence of 'ghost' epithelial cells. In this case report, we present a case of calcifying epithelial odontogenic cyst in the mandibular left region in a 19 year old male patient. The diagnosis was confirmed by clinical, radiographic and histological co-relation.

Keywords: Calcifying odontogenic cyst, Gorlin's cyst, Ghost cells

Introduction

The calcifying epithelial odontogenic cyst (CEOC) was first described by Gorlin et al who were impressed by the significant presence of the so-called "ghost cells". [1] Over the years since its first description, it has become clear that the calcifying epithelial odontogenic cyst (CEOC) has a number of variants, including features of a benign odontogenic tumor. There has been a complete re-evaluation of this lesion by many authors. One major conclusion of Praetorius et al. about this lesion was that, it comprised two entities: a cyst and a neoplasm. [2]

Case report:

A 19 year old male patient presented with a chief complaint of a slow growing swelling in the mandibular left canine premolar region since 6 months with no associated pain or paraesthesia. Swelling was initially small for the first 2 months and then increased to its present size.

Clinical examination revealed noticeable facial asymmetry caused by swelling in the lower left side of the face. Swelling was approximately 5cm×4cm in size extending superiorly from angle of mouth to the lobe of the left ear, inferiorly from inferior border of mandible to the submandibular region, anteriorly 1cm away from the midline and posteriorly till the ramus of the mandible. (Fig.1)

Intraoral examination revealed an ill-defined solitary expansile oval shaped swelling measuring 3cm×2cm in diameter involving buccal and lingual cortices and extending antero-posteriorly from 41 to 47. Supero-inferiorly it extended from the level of the attached gingiva obliterating the buccal and lingual vestibules. The mucosa over the lesion appeared normal. The swelling was non-tender, non-reducible, non-compressible in constituency. Grade 1 tooth mobility was seen with respect to 44,45,46,47. Provisional diagnosis of Ameloblastoma was given. OPG revealed well defined multilocular radiolucency extending from 33 till well into the ramus beyond 47 (Fig.2).

Microscopically, the histopathological report revealed cystic lining of stratified squamous epithelium with intramural ameloblastomatous proliferations. The basal layer showed tall columnar cells with hyperchromatic nuclei facing away from the basement membrane. The basal layer showed tall columnar cells with hyperchromatic nuclei facing away from the basement membrane. Certain focal areas showed eosinophilic balloon shaped elliptical epithelial cells with faint outline of nuclei resembling 'ghost cells'. Connective tissue capsule showed parallelly arranged collagen fibres. Juxta epithelial hyalinization was also seen in focal areas. Few chronic inflammatory cells with extravasated RBC’s were also seen. These findings were consistent with the diagnosis of "Calcifying Epithelial Odontogenic Cyst". (Fig. 3)
Discussion

The CEOC is usually a non-aggressive cystic lesion lined by odontogenic epithelium that resembles that of the ameloblastoma, but with characteristic ghost cell keratinization. It is a developmental odontogenic cyst, and its occurrence constitutes about 0.3-0.8% of all odontogenic cysts. [3]

Calcifying epithelial odontogenic cysts are thought to be a unicystic process, which develop from the reduced enamel epithelium or remnants of odontogenic epithelium in the follicle, gingival tissue or bone. The epithelial lining of a CEOC appears to have the ability to induce the formation of dental tissues in the adjacent connective tissue wall, and may thus be associated with other odontogenic tumours. [4]

The age of these patients may range from 5 to 92 years, with a peak incidence in the second decade of life. [5] Other authors however state a bimodal age distribution, with a second peak in the 6th-7th decade of life. [3,5] The lesion has no sex predilection and is equally distributed between the maxilla and mandible.

Calcifying epithelial odontogenic cyst commonly occurs anterior to the first molar region in which 75% of cases are in the incisor-canine region or inter-canine region, usually crossing the midline in the mandible, which is however a rare feature in the maxilla. Clinically, the lesion usually presents as an asymptomatic swelling causing a hard bony expansion of the jaw. [5] Early lesions are usually detected following a routine radiographic examination, and they are often associated with an unerupted tooth. The peripheral variant occurs on the gingiva as a non-specific well-circumscribed sessile or pedunculated mass with a smooth surface, which may resemble a gingival fibroma, gingival cyst or peripheral giant cell granuloma. [6]

Radiographically, majority of the lesions present in an unilocular form with a well-defined corticated margins while 5-13% of cases are multilocular. [4] The internal structure may vary in appearance - it may be completely radiolucent, but is usually mixed (radiolucent-radiopaque), and may have scattered irregular sized calcification producing a variable range of opacities (salt & pepper type of pattern), or may even show large solid amorphous masses. [5] Central CEOC has been reported to be associated with odontoma in 24-35% of cases, usually a canine. [6] Unilocular lesions may mimic dentigerous, radicular or residual cysts while multilocular lesions may resemble ameloblastoma or odontogenic keratocysts. [3]
Our case appeared as a multilocular radiolucency with expansion of buccal and lingual cortical plates resembling an ameloblastoma radiographically.

Over the years, various authors have attempted to classify and group the lesion owing to its unique character. Praetorius (1981) proposed a classification for grouping CEOC as Type I (cystic type) & Type II (neoplastic type, dentinogenic ghost cell tumour). He further subdivided the cystic patient (Type I) into 3 different types:

a) Simple unicystic type
b) Odontome-producing type, and
c) Ameloblastomatous proliferating type.[7]

Toida (1998) proposed a classification in which he called the cystic variant as calcifying ghost cell odontogenic cyst (CGCOC) and used the term calcifying ghost cell odontogenic tumor (CGCOT) for the neoplastic variant, as given below [7,8]

1. Cyst: CGCOC
2. Neoplasm:
   a. Benign- CGCOT
   b. Solid variant- Solid CGCOT
   B. Malignant- Malignant CGCOT
3. Combined lesion: Each of the categories above associated with the following lesions:
   a. Odontoma
   b. Ameloblastoma
   c. Other odontogenic lesions.

The histological features of a classical CEOC include a cystic cavity with a fibrous capsule, with a lining of odontogenic epithelium that is 6-8 cells thick. The basal layer is made up of ameloblast-like columnar or cuboidal cells with hyperchromatic nuclei polarized away from the basement membrane. The overlying loosely arranged epithelial layer shows similarity to the stellate reticulum of the enamel organ. The typical microscopic characteristic of this lesion is the presence of variable amounts of aberrant epithelial cells, devoid of nuclei, which are eosinophilic and retain their basic cell outline and are called "ghost cells". These ghost cells may undergo calcification and lose their cellular outline to form sheet-like areas of calcified keratin. The nature of ghost cell change is controversial. It may be due to the effect of coagulative necrosis and dystrophic calcification or may be a form of normal or abnormal keratinization of the odontogenic epithelium. Ghost cells are not unique to CEOC, but are also seen in odontoma, ameloblastoma, craniopharyngioma, and other odontogenic tumours.[6] The present case showed all the typical features of CEOC, with the presence of a cystic epithelial lining with ameloblast-like cells, ghost cells, dystrophic calcification and irregular areas of dysplastic dentin.

The CEOC is treated conservatively by surgical enucleation and recurrences are very uncommon (recurrences depend on the completeness of cyst removal). The malignant transformation of a pre-existing benign CEOC can occur but is extremely uncommon.[9] The CEOC may also be associated with other odontogenic tumours such as adenomatoid odontogenic tumour, ameloblastoma, ameloblastic fibro-odontoma and ameloblastic fibroma where wider excision may be required.[10]

Conclusion
The CEOC is an uncommon lesion which may show characteristics of both a cyst and a solid neoplasm. Its clinical and radiographic features may mimic other odontogenic cysts/tumours, and a definitive diagnosis can only be made histologically. The present case showed non-specific clinico-radiographic findings, which resembled an ameloblastoma. Our case can be classified as Type (c) (Ameloblastomatous proliferating type) of calcifying epithelial odontogenic cyst (according to the classification proposed by Praetorius).

References

Conflict of interest: Nil
Source of fund: Self