

Calcifying Epithelial Odontogenic Cyst: A Review

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Abstract: *Gorlin Cyst or calcifying odontogenic cyst (COC) is an uncommon developmental odontogenic cyst and is a very rare lesion. This lesion has so many variations that it has got various terminologies according to their characteristic features by different scientists. The main aim of writing this article is to give a review of the Gorlin Cyst and classify it and give its clinical representation, and also describe other similar lesions to differentiate it from Gorlin cyst, and then have a brief idea about the various treatment modalities for it. And also we will see the various forms of CEOC and the association of CEOC with other lesions. Also, the prognosis of this lesion is good because it has a recurrence chance which occurs after approximately 5 - 6 years after surgery but this is also rare.*

Keywords: Gorlin Cyst, Ghost Cell, Dentinogenic Ghost Cell Tumor (DGCT), Calcifying Epithelial Odontogenic Cyst (CEOC)

1. Introduction

Firstly the Calcifying Epithelial Odontogenic Cyst was described as a different type of entity in 1962 by Gorlin et al and so given the name "Gorlin cyst", and accounts for 1 - 2% of all odontogenic tumors and 0.3 - 0.8% of all odontogenic cyst. Gorlin cyst is also classified as a benign odontogenic tumor by Kramer et al. Two entities are related to the Gorlin cyst a cyst and a neoplasm which was proposed by Praetorius et al (1). In various cases, it is seen that the majority of the tumors of the jaws mimic the various features of the cysts so one should always understand and know the classification of the cysts so as to identify cysts and differentiate between cysts and tumors (2). This calcifying odontogenic cyst condition was also described in 1932 by Rywkind in German literature (3, 4, 5). These are solid or cystic lesion that is well circumscribed and rare they are formed by the odontogenic epithelium that gives the appearance of follicular ameloblastoma but contains cells that do not have any nucleus which is known as ghost cells and these lesions have calcifications that are spherical in shape (6). CEOC is an uncommon and unusual lesion that may show characteristics of both solid neoplasm and a cyst and represents a heterogeneous group of lesions that demonstrate considerable histopathologic diversity and variable clinical behavior. These are classified under developmental or inflammatory origin. Embryonic Epithelial rests are not found in the skeletal bone and this is the reason why only skeletal bone is affected by the epithelium - lined cysts. It can also occur in the jaws where these are lined by the epithelium. These cysts are not always well defined in some cases the margins may be absent as in the case of enlarging epithelial cysts. These calcifying odontogenic cysts in a more descriptive way can also be named calcifying and keratinizing odontogenic cysts and this description is given by Gold. And so this condition is also termed melanoticameloblasticodontoma or it may be referred to as keratinizing odontogenic cysts. The scientists who found the presence of ghost cells in the calcifying odontogenic cysts were Fejeskov and Krough. And so they named this

calcifying epithelial odontogenic cyst the ghost cell odontogenic tumor. (7). The CEOC can also occur with any other tumors that are odontogenic in nature, and the commonest type of these lesions are ODONTOMA, which occurs in about 24% of the cases and also is associated with impacted tooth most commonly canine in about 10 - 32% of the cases. (5, 8)

2. Discussion

CEOC has a unique process that is unicystic which develops from the remnants of odontogenic epithelium that are present in various structures like follicles, bone, or gingival tissues or can develop from the reduced enamel epithelium. These epithelium linings have the ability to form the tissues in the connective tissues that are adjacent to these epithelium, and so are associated with other odontogenic tumors. CEOC can be both central lesion or peripheral localization but central lesions are most commonly occurring, whereas peripheral localizations that occur in the soft tissue also known as gingival lesions are rare but occur in about 13 - 25% of the cases (8, 9). When the CEOC occurs as an intra - osseous lesion then it is referred to as an "odontocalcifying odontogenic cyst", a name given by Hirshberg et al. The most common example of CEOC being an intra - osseous lesion is Odontoma. These odontomas develop as secondary lesions from the CEOC's lining epithelium (5). Clinically, the site prediction in CEOC is equal in both the jaws and most commonly in the incisor and canine region in 65% of the cases, having no gender prediction (10).

CEOC Variant	Age	Mean Age
CEOC	30 - 40 years	33 years
CEOC with ODONTOMA	Younger adults	17 years
NEOPLASTIC	Old age	

These are asymptomatic swelling having the bony hard expansion of the mandibular or maxillary jaw. Usually, CEOC is asymptomatic and commonly detected by

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incidental radiography. Usually, these lesions are associated with an impacted tooth which gives the differential diagnosis of a dentigerous cyst, calcifying epithelial odontogenic tumor, and adenomatoid odontogenic tumor as these lesions are also associated with an impacted tooth. These can also mimic the gingival fibroma, gingival cyst, or peripheral giant cell granuloma as these present well - circumscribed sessile pedunculated mass with a smooth surface that occurs in gingiva (8). Radiographically, CEOC is unilocular, well - defined radiolucency occasionally multilocular in 5 - 13% of the cases. One - third of the radiolucent lesion occurs in association with an unerupted tooth, commonly canine. In one - third to half of the cases, the lesions are radio - opaque in nature (10). These lesions are mixed radiolucent and radio - opaque having salt and pepper like patterns as these have irregular - sized calcifications that produce a range of opacities. In some cases and studies, it is shown that these calcifying odontogenic cysts as bimodal in origin which means these cysts may occur in two generations as in the second and the seventh decade of life. The size of these calcifying cysts varies in a lot of cases, the size may be small or as big as it can also cover the entire lesion (2). In radiographic features, if the CEOC is unilocular, it gives the differential diagnosis of a dentigerous cyst or residual or radicular cysts and if the lesion is multilocular it mimics ameloblastoma or odontogenic keratocyst (8). The size of the lesion is between 2 - 4 cm but can be as large as 12 cm. Histopathologically, there is the presence of a variable number of ghost cells, which are devoid of the nucleus, in the epithelial component which is the most characteristic feature of CEOC. Ghost cells can also be found in ameloblastoma, odontoma, and other odontogenic tumors. Odontogenic epithelial lining is 6 - 8 cells thick (10). In

1971 CEOC was designated as a non - neoplastic lesion by WHO, but they decided to classify CEOC as an odontogenic benign tumor (3). Then in 1992, CEOCs has been classified as neoplasms and tumors but most CEOCs were non - neoplastic and this classification was again given by WHO (5). Praetorius in 1981 further classified CEOC for their heterogeneous groups. In this classification, Praetorius's motive was to separate these cystic lesions from the neoplasms such as solid lesions (1).

1) Type 1 (Cystic type)

- Simple Unicystic Type
- Odontoma - producing Type
- Ameloblastoma Proliferating Type

2) Type 2 (Neoplastic Type)

- Dentinogenic ghost cell tumor, or
- Epithelial odontogenic ghost cell tumor (7).

When CEOC is associated with the teeth apex then there is a high risk of root resorption (3). Adenomatoid odontogenic tumor, Dentigerous cyst, and Calcifying epithelial odontogenic tumor are the differential diagnosis of CEOCs. (9). CEOC has a basal layer that is well defined and the layer is thick containing many cells and this is the reason that it also resembles the stellate reticulum of the enamel organ. Also, those ghost cells are present in the lining epithelium of the cyst or it may be present in a capsule which further becomes fibrous so - called fibrous capsule. These ghost cells also after some time become calcified due to calcifications and also the dentin next to the ghost cells becomes dysplastic and these ghost cells and dentin are laid down just next to their basal cell layer. (1).

Features	CEOC	CEOT	AOT	Dentigerous Cyst
Age	30 - 40 years	8 - 92 years	2 - 19 years	20 - 40 years
Sex	Female>Male	Male>Female	Female>Male	Equal in both
Site	Equal in maxilla and mandible, anterior to the first molar, most commonly incisor, and canine region	Mandible: Maxilla =2: 1 In premolar to molar areas	Maxilla>Mandible Anterior portion of maxilla	Maxilla - 33% Mandible - 67% Most commonly angle of the mandible
Clinical Presentation	Slow - growing, painless, swelling	Painless slow - growing bony expansion that is hard on palpation	Slight painful	Painless swelling and pain arise when secondarily infected
Radiographic Presentation	Cyst - like radiolucency is usually associated with impacted teeth	Driven snow appearance with mixed radiolucent and radio - opaque lesions frequently associated with impacted teeth	Well - circumscribed radiolucency is usually associated with an impacted tooth having snowflakes - like calcifications	Unilocular or multilocular well - defined radiolucency is always associated with the impacted tooth

Radiographically, these lesions also present as driven snow appearance and may present as a honeycomb pattern. This driven snow pattern of these cysts is due to the alignment of the radiopaque flecks around the neck of the involved tooth; this appearance can be seen in mixed radiopaque and radiolucent lesions. And the honeycomb pattern is due to the compartments of the smaller size that tends to be uniform in their size as in the honey - bees comb, this pattern occurs in the radiolucent multilocular lesions (11). The histological finding is the only appropriate and definitive diagnosis for the CEOC because these conditions lack characteristics in radiologic and clinical features as well as this condition shows variable biological behavior. As earlier said that the ghost cells are the characteristic histological feature of

CEOC, by the time these cells undergo calcification and they form a firm sheet of calcified keratin. These ghost cells are also seen in odontoma, ameloblastoma, and other odontogenic tumors (12). Recurrences also occur in these calcifying cystic lesions and these recurrences occur approximately after 5 - 6 or more years of surgery (13). In these conditions, the expansion of the jaw bone occurs be it buccal or lingual but most commonly lingual cortical plate expansion occurs in these types of lesions. And also most commonly seen in periodontal areas of the dentition and the periapical regions as their lesion are strongly related to the teeth (14).

3. Treatment

Treatment of CEOC is conservative in nature and the treatment is done with complete surgical enucleation of the cyst and by curettage of the total lesion. The treatment is based on the size and location of the lesion. Enucleation stands for the surgical removal of mass without cutting or dissecting the lesion and on the other hand curettage stands for the scooping out the tissue lining of the lesion. If the lesion is associated with the tooth then that tooth first should undergo root canal treatment and then the enucleation or the curettage of the lesion should be done. If there is a need of extraction of the tooth as in many cases the extraction of the involved impacted tooth is to be done. For combined lesions like CEOCs with odontoma or ameloblastoma, odontogenic tumors are treated according to the aggressiveness of the lesion (15) and the prognosis is good with only a few recurrences in patients with CEOC. The prognosis of CEOC associated with other odontogenic tumors is the same as for the associated tumor (10).

Figure Legends:



Figure 1



Figure 2



Figure 3

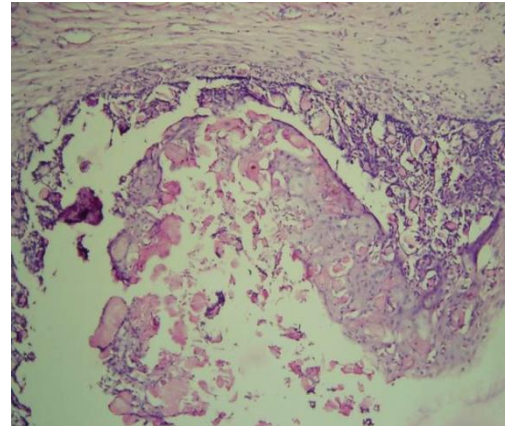


Figure 4

The H & E stained section revealed a cystic lesion having an odontogenic epithelial lining associated with a fibrovascular connective tissue capsule. The epithelium is of variable thickness. The basal layer is lined by columnar cells with the nucleus towards the base of the cell and is arranged in a palisaded manner. Stellate reticulum - like cells are seen above the basal layer. Numerous ghost cells with variation in size which appears as pale, eosinophilic epithelial cell with pyknotic nuclei are seen. Adjacent to the ghost cells, irregular basophilic masses are suggestive of dystrophic calcification is also seen. The connective tissue shows numerous endothelial lined blood vessels, mixed inflammatory cell infiltrate, and predominantly lymphocytes are seen. Within the capsule, some areas show eosinophilic masses suggestive of retinoid - like material beneath the epithelial lining. Histopathologic features were suggestive of calcifying epithelial odontogenic cysts.

4. Conclusion

This presentation was made in favor of Calcifying Epithelial Odontogenic Cysts so as to study the incidence and look forward to the history of this condition and to also differentiate between the calcifying epithelial odontogenic cyst and the tumors of the jaws which in fact correlate with each other. This presentation was made to look at how this condition occurs clinically, radiographically, and histologically. Also, this presentation briefly explains this condition with the various classifications given by WHO and many more scientists that are mentioned above in this presentation. This also gives information to the viewers about how to identify these conditions and how to proceed with the treatment to get better results and a good prognosis for the well - being of the patients. CEOCs in fact COC and many other terminologies given for this single condition, is a very uncommon or rare condition. This condition mimics the features of odontogenic tumors. And also CEOC is very strongly related to the impacted teeth and its dental follicle so one should always check the histopathological findings of the follicle also.

Conflict of Interest: Nil

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