

Calcifying Odontogenic Cyst: Case Report and Review of Literature

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ABSTRACT

The calcifying odontogenic cyst (COC) was first described by Gorlin et al. in 1962. Calcifying odontogenic cyst is comparatively rare in occurrence, constituting about 0.37% to 2.1% of all odontogenic tumors. The most notable features of this pathologic entity are histopathological features which include a cystic lining demonstrating characteristic “Ghost” epithelial cells with a propensity to calcify. In addition, the COC may be associated with other recognized odontogenic tumors, most commonly odontomas. There are variants of COC according to clinical, histopathological, and radiological characteristics. Therefore, a proper categorization of the cases is needed for better understanding of the pathogenesis of each variant. Here, we report a classic case of calcifying odontogenic cyst occurring in a 15 year old girl in right maxillary region.

KEYWORDS: Calcifying Odontogenic Cyst, Gorlin, Ghost Cell

INTRODUCTION

True bone cysts are often encountered in facial bones due to the presence of embryonic epithelial rests in these bones. Most of them are odontogenic in origin. The calcifying odontogenic cyst is an uncommon benign cyst of odontogenic origin, characterized by an ameloblastoma-like epithelium with ghost cells that may calcify, first described by Gorlin et al in 1962.¹⁻³ It is extensively diverse in terms of its clinical presentation, histopathological features and biological behavior. Majority of the cases present with cystic characteristics, few are of the solid type (15%), and rare occurrence of malignant transformation.³⁻⁵ The calcifying odontogenic cyst (COC) is a rare example of a developmental odontogenic cyst, its occurrence constituting about 0.37% to 2.1% of all odontogenic tumors.⁶ It was classified as SNOMED code 930/0, in the World Health Organization’s publication

histological Typing of Odontogenic Tumors.⁷ There has been a complete re-evaluation of this lesion by many authors. Praetorius et al. concluded that this lesion comprised of two entities: a cyst and a neoplasm.⁸ But it is its variable histology and clinical behavior that has raised the question of whether or not it is a cyst or a true neoplasm.

CASE REPORT

A 15 year old girl reported to our outpatient clinic with the complaint of swelling in upper right side of the jaw that had been present for approximately 2 years. On evaluation, there was an asymmetry involving the right mid-face region (figure 1). Swelling was approximately 6 cm × 4 cm in size, extending supero-inferiorly from 1 cm below infra orbital rim to angle of the mouth and antero-posteriorly from right ala of the nose to about 3 cm in front of the tragus.

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Intra oral examination (Figure 2) revealed buccal cortical expansion extending front 11 to 16 regions. The mucosa over the lesion was intact. On palpation the Swelling was cystic and non-tender. Dental examination showed distal tipping of teeth 12 and mesial tipping of 13. Aspirate was found to be straw colored fluid(Figure 3).



Figure 1

Figure 2

Figure 1: Extra Oral View, Figure 2: Intra Oral View

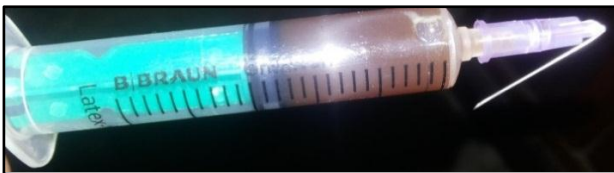


Figure 3: Straw Colored Aspirate.

Radiographic examination (Figure 4) disclosed a unilocular well-circumscribed round radiolucency extending from 11 to 16 regions, with irregular minute radiopaque specks within it.



Figure 4. Pre-Operative OPG

Based on clinical and radiological findings, a provisional diagnosis diagnosis of calcifying odontogenic cyst was considered. The adenomatoid odontogenic tumor was not included in differential diagnosis because of the lack of teeth inclusion. The operation was performed under general anaesthesia by enucleation of the lesion, in agreement with the



Figure 5. Intra Operative- Crevicular Incision And Site Exposure



Figure 6. Defect After Enucleation of Cyst



Figure 7. Whitehead's Varnish Dressing Packed And Wound Closure



Figure 8. Excised Cystic Specimen Sent For HPE

principle of clinical method for treating small cystic lesions of jaws. Intraoral crevicular incision was given from distal of 21 to mesial of 17 and cystic lesion was exposed. Enucleation was performed and

defect was packed with whitehead's varnish dressing, wound closure was done (Figure 5-8).

The enucleated specimen was cystic, entire specimen was sent for histopathological evaluation, and it was revealed as calcifying odontogenic cyst. On histopathological examination, a well-defined cystic lesion is found with a fibrous capsule. The basal cells of the epithelial lining are similar to ameloblasts (Figure 9). There is presence of variable numbers of ghost cells within the epithelial component. These eosinophilic ghost cells are altered epithelial cells that are characterized by the loss of nuclei with preservation of the basic cell outline. Masses of ghost cells fuse to form large sheets of amorphous, acellular material. Areas of an eosinophilic matrix material) that are considered dysplastic dentin (dentinoid) is present adjacent to the epithelial component (Figure 10). Calcification within the ghost cells is seen, appears as fine basophilic granules (Figure 11). This is believed to be the result of an inductive effect by the odontogenic epithelium on the adjacent mesenchymal tissue.

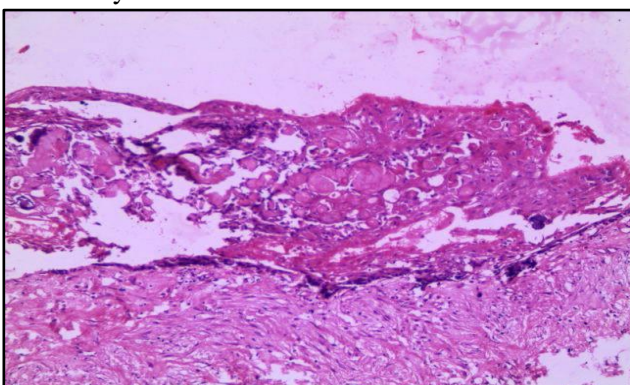


Figure 9. The Cyst Lining Shows Ameloblastoma Like Epithelial Cells, With A Columnar Basal Layer. Large Eosinophilic Ghost Cells Are Present Within the Epithelial Lining.

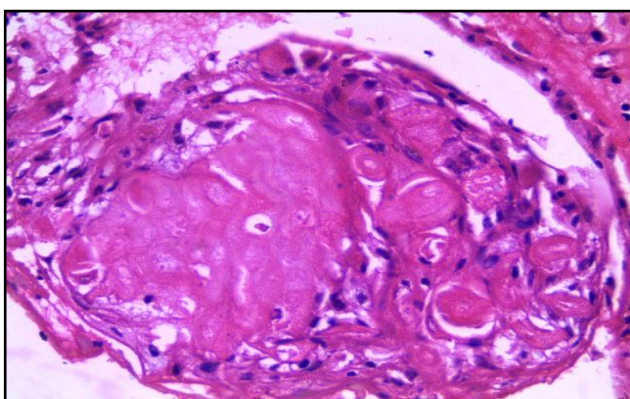


Figure 10. Eosinophilic Dentinoid Material Is Present Adjacent To Sheet Of Ghost Cells.

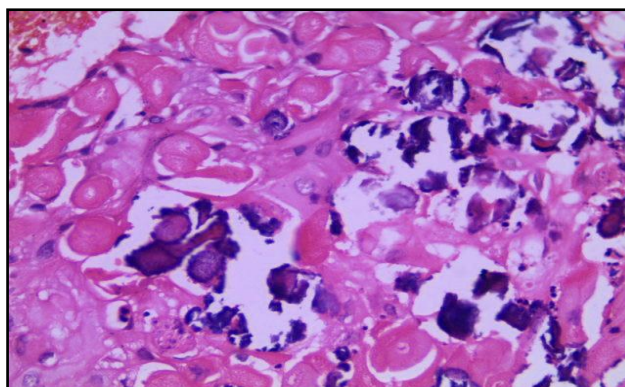


Figure 11: Basophilic Granules (Calcification) Appearing In Some Ghost Cells.

The white head varnish pack was removed after 2 weeks and primary closure was done. Healing after surgery was uneventful and there was no recurrence after 6 months (Figure 12).



Figure 12. Post-Operative Intraoral View After 6 Months

DISCUSSION AND REVIEW OF LITERATURE

Gorlin and colleagues identified file COC as a distinct pathological entity in 1962 although according to Altini and Farman, a similar occurring condition had previously been mentioned in German literature in 1932 by Rywkind.⁹ It was earlier thought to be an oral presentation of dermal calcifying epithelioma of Malherbe.^{10,11,12} The COC has also been reported under a variety of other designations including keratinizing cyst¹³, keratinizing cyst and calcifying odontogenic cyst (KOC)¹³, calcifying ghost cell odontogenic tumor¹⁴, dentinogenic ghost cell odontogenic tumour, epithelial odontogenic ghost cell tumour, ghost cell cyst, calcifying ghost cell odontogenic tumour, and dentino-ameloblastoma by various authors.¹⁵ The controversy and confusion have existed regarding relationship between non-neoplastic, cystic lesions and solid tumor masses

that shares the cellular and histomorphologic features described by authors.¹⁶ In 1971, the WHO described COC as a “non-neoplastic” cystic lesion; choosing it to be classified as a benign odontogenic tumor. In 1992, the World Health Organization (WHO) classified COC as a neoplasm rather than a cyst but confirmed most of the cases are non-neoplastic. In view of this duality, many different terminologies have been applied to cystic and solid COC variants, but calcifying odontogenic cyst is the preferred term.¹⁷

Different terminologies for COC are reviewed in Table 1.

| | |
|--------------------------|--------------------------------------------------|
| Gorlin et al. 1962 | Calcifying odontogenic cyst |
| Gold 1963 | Keratinizing calcifying odontogenic cyst (KCOC) |
| Fejerskov and Krogh 1972 | Calcifying ghost cell odontogenic tumor (CGCOT) |
| Freedman et al. 1975 | Cystic calcifying odontogenic tumor (COCT) |
| Praetorius et al. 1981 | Dentinogenic ghost cell tumor (DGCT)* |
| Ellis and Shmookler 1986 | Epithelial odontogenic ghost cell tumor (EOGCT)* |
| Colmenero et al. 1990 | Odontogenic ghost cell tumor (OGCT)* |

*These terms are used restrictedly for the solid neoplastic variant of COC.

Li and Yu (2003) have drawn attention to the dilemma regarding the nature of these ‘ghost cell lesions’ as cysts, neoplasms and even malignancies. They have therefore proposed what they have called a more concise terminology and classification based on the likely differences in biological behaviour of the COC and its related lesions. They divided the lesions into three groups – cysts, benign tumours and malignant tumours – and suggested that the term COC should be used specifically to designate the unicystic lesions with or without an associated odontoma.

These fulfilled the diagnostic histological criteria described in the 1992 WHO classification and were best classified as developmental odontogenic cysts. As such, they believed that this group should retain the term COC. They have argued that the reason for

inclusion of odontoma-associated COCs in this group is that these odontomas are hamartomatous and unlikely to modify their behaviour. They believed that other ghost cell lesions identified as benign or malignant tumours, should be classified separately and named accordingly.

Praetorius (2006), who was one of the authors of the first description of the lesion (Gorlin *et al.*, 1962) and who has had extensive experience of the range and variations of ghost cell lesions of the jaws in general, disagrees in some respects with the classification proposed by Li and Yu (2003). He argues that the COC is not just a developmental cyst like the dentigerous cyst because it often forms islands of epithelium and dentinoid in the wall; while in some of them, an odontoma forms in the wall.³²

Several classifications of CGOC subtypes have been proposed, but most of them have limitations in separating cystic and neoplastic variant.

Classification of COC by Reichart.¹⁵

(1) Non neoplastic (simple cystic) variant (CGCOC^a):

- with non-proliferative epithelial lining,
- with non-proliferative (or proliferative) epithelial lining associated with odontomas^b,
- with proliferative epithelial lining,
- with unicystic, plexiform ameloblastomatous proliferation of epithelial lining^c.

(2) Neoplastic variant:

(A) benign type (CGCOT^d):

(a) cystic subtype (cystic CGCOT)

- SMA ex epithelial cyst lining^e,

(b) solid subtype (solid CGCOT)

- Peripheral ameloblastoma-like^f
- SMA-like^g,

(B) malignant type (malignant CGCOT or OGCC^h):

(a) cystic subtype (b) solid subtype.

(a. Calcifying ghost cell odontogenic cyst. b. Also

classified as compound (or complex) cystic ghost cell odontomas. c.Does not completely fulfill the histopathologic criteria of early ameloblastoma as suggested by Vickers and Gorlin. d.Calcifying ghost cell odontogenic tumor. e.With histopathologic features of early ameloblastoma as suggested by Vickers and Gorlin. f.Resembling a peripheral ameloblastoma, hence denoted as peripheral epithelial odontogenic ghost cells tumor. g.Often called central epithelial odontogenic ghost cell tumor. h.Odontogenic ghost cell carcinoma.)

Recently, when the World Health Organization (WHO) in 2005 updated its classification of odontogenic tumors, the name of the COC revised universally to the calcifying cystic odontogenic tumor (CCOT) to emphasize the neoplastic nature of a lesion previously categorized as an odontogenic cyst.^{18,19} But in general owing to its duality, cystic and solid COC variants, calcifying odontogenic cyst is the preferred term.¹⁷

COC is a rare developmental cyst. Tomich reviewed about 34 years for odontogenic tumors and cyst and found—less than two cases per year! It follows that the average oral and maxillofacial surgeon is likely to see only a case or two during his/her professional career.⁷

The cells responsible for the calcifying odontogenic cyst are dental lamina rests (rests of Serres) within either the soft tissue or bone. Therefore, calcifying odontogenic cysts are cysts of primordial origin and are not associated with the crown of an impacted tooth.²⁰ It most often occurs as a central (intra-osseous) lesion, whereas peripheral (extra-osseous) localization in the soft tissue is rare.

The COC normally appears as a painless, slow-growing tumor, which affects the maxilla and mandible equally, showing a strong predilection for the anterior segment (incisor/canine area).It does not show any gender predilection generally affecting the young adults in the third to fourth decade of life.⁵ In our case report, the age of female patient was 15 years and occurred at right side of maxilla. Presence of pain generally indicates secondary infection.

Radiographically, the COC is usually a mixed lesion, with radiolucent area, may present with a uni or multilocular appearance, containing different

amounts of radiopaque material.^{21, 22} McGowan and Browne²³, in 1982, found that the presence of mineralization was approximately twice as frequent in microscopic examination compared to radiographic analysis. The present case had almost no or very low radiographically detectable calcified bodies/flecks in the lesion. In our case report, radiographic examination disclosed a solitary unilocular well-circumscribed round radiolucency with minute calcified flecks within it.

The definitive diagnosis of COC can be made more appropriately only histologically, due to the lesion's lack of characteristic clinical and radiological features, as well as its variable biological behaviour.

Histological features of a classic calcifying odontogenic cyst are characteristic and present few diagnostic problems. The microscopical feature of a classical COC includes a fibrous capsule with a lining of odontogenic epithelium. The basal layer is made up of ameloblast-like columnar or cuboidal cells of 4-10 cell thickness over lined by a loosely arranged epithelial cells hearing similarity to stellate reticulum of the enamel organ.^{24,25} Also present are a number of epithelial cells devoid of any nuclei, which are eosinophilic with their basic cell outline retained (ghost cells). Sometimes these ghost cells may undergo calcification and lose their cellular outline. When this happens they form firm sheet like area, of calcified keratin.^{24,26,27} Ghost cells may be due to the effect of coagulative necrosis and dystrophic calcification or it may be a form of normal or abnormal keratinization of the odontogenic epithelium. Ghost cells are not unique to COC, but are also seen in odontoma, ameloblastoma, craniopharyngioma, and other odontogenic tumours^{22,25,28} and can undergo calcification, which is believed to be dystrophic in nature.²⁹⁻³¹ The ability to induce dental hard tissue formation appears to be a property of epithelial cell lining of the COC .

The malignant transformation of a pre-existing benign COC could occur, yet is extremely uncommon.

The COC is treated by surgical enucleation unless it is associated with another odontogenic tumour, in

which case wider excision may be required. In the presence of a complex odontome, conservative removal will still be adequate.

An ameloblastoma or one of its variants with foci of ghost cells must be treated as would be an ameloblastoma without ghost cells. Although classic uncomplicated cases of COCs may grow to a large size, reported recurrences are rare

CONCLUSION

COC is a unique lesion which is rare in its occurrence, possessing both cystic and neoplastic potential and showing considerable number of variants clinically, radiographically, and histopathologically. Further studies based on its different variants are required for a better understanding and may aid in its classification.

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