

# Calcifying Odontogenic Cyst and Dentinogenic Ghost Cell Tumour – A Review

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## Abstract

*Calcifying odontogenic cyst (COC) is also known as “Gorlin cyst,” first described by Gorlin in 1962 which is a rare developmental odontogenic lesion. The calcifying odontogenic cyst (COC) is a heterogeneous lesion existing either as cystic or solid variant. This lesion has different terminologies and is classified into various subtypes. The World Health Organization classified (2005) COC as a neoplasm and used the term calcifying cystic odontogenic tumor for benign cystic type, the dentinogenic ghost cell tumor for the benign solid type lesions and the malignant type as ghost cell odontogenic carcinoma. The dentinogenic ghost-cell tumor (DGCT), a solid variant of the COC, is an uncommon odontogenic neoplasm. This article discusses the various terminologies, clinical features, classification, histopathology, and treatment of COC.*

**Keywords:** *Calcifying odontogenic cyst, ghost cell, odontogenic cyst, Dentinogenic ghost cell tumor*

## Introduction

**Calcifying odontogenic cyst (COC)** is a benign odontogenic tumor mainly a cystic type which affects the anterior areas of the jaws. It is mainly seen in people in their second or in third decades but can be seen at any age. In one-third of cases, an impacted tooth is involved along with unilocular radiolucency seen in radiographs. Histologically many cells are seen as "ghost cells", enlarged eosinophilic epithelial cells without nuclei. A solid variant of COC is called Dentinogenic ghost cell tumor which is an uncommon type of odontogenic neoplasm.

## Classification<sup>8</sup>

Suggested classification of odontogenic ghost cell lesion

|         |  |
|---------|--|
| Group 1 | Simple cysts<br>Calcifying odontogenic cyst (coc)  |
| Group 2 | Benign neoplasm of calcifying cystic odontogenic tumours (CCOT). The following combination may be<br><br>a.CCOT associated with an odontome<br>b.CCOT associated with adenomatoidodontogenic tumor<br>c.CCOT associated with ameloblastoma<br>d.CCOT associated with ameloblastic fibroma<br>e.CCOT associated with ameloblastic fibro-odontoma<br>f.CCOT associated with odonto-ameloblastoma<br>g.CCOT associated with odontogenic myxofibroma |
| Group 3 | Solid benign odontogenic neoplasm with dentinoid formation<br>Dentinogenic ghost cell tumor  |

|            |  |
|------------|--|
| Group<br>4 | Malignant odontogenic neoplasm with similar feature of Dentinogenic ghost cell tumor<br>Ghost cell odontogenic carcinoma |
|------------|--|

### Terminologies<sup>1,2,3</sup>

| Terminologies                                   | Author Proposed            |
|---|----------------------------|
| Calcifying odontogenic cyst (COC)               | Gorlin et al. (1962)       |
| Keratinizing calcifying odontogenic cyst (KCOC) | Gold et. al. (1963)        |
| Keratinizing ameloblastoma (KA)                 | Bhaskar (1965)             |
| Calcifying ghost cell odontogenic tumor (CGOT)  | Fejerskov and Krogh (1972) |
| Cystic calcifying odontogenic tumor (CCOT)      | Freedom et al (1975)       |
| Dentinogenic ghost cell tumor (DGCT)            | Praetorius et al. (1981)   |
| Epithelial odontogenic ghost cell tumor (EOGCT) | Eliis and shmooker (1986)  |
| Calcifying ghost cell odontogenic cyst (CGCOC)  | Toida (1998)               |
| Odontogenic ghost cell tumor (OGCT)             | Colmenero et al. (1990)    |
| Odontogenic ghost cell ameloblastoma (OGCA)     | Shear (1994)               |
| Odontocalcifying odontogenic tumor (OOT)        | Wirshberg et. al. (1994)   |
| Calcifying cystic odontogenic tumor (CCOT)      | WHO classification (2005)  |

### Clinical features

Swelling is the main complaint and pain is rare in most cases. Intraosseous lesions produce a hard-bony expansion and sometimes fairly extensive. Lingual expansion may also be observed in some cases. Occasionally, the calcifying odontogenic cyst perforates the cortical plate and may extend into the underlying soft tissues<sup>1</sup>. In a few cases, displacement of the teeth has been present. Some cases may be completely symptomless and have been diagnosed during radiological examination. Extraosseous lesions tend to be pink to red, circumscribed elevated masses have a measurement of 4 cm in diameter. Recurrence rate for COC is rare.<sup>4</sup>

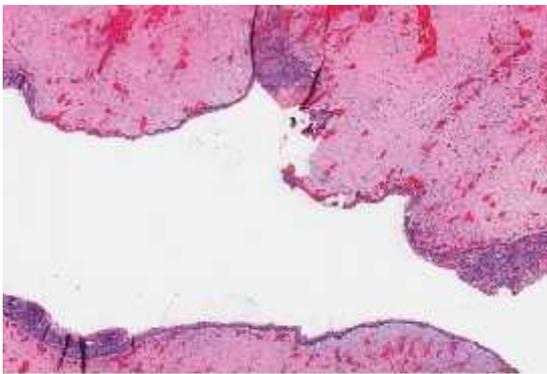


Fig 1 Swelling obliterating buccal vestibule in  
Calcifying odontogenic cyst

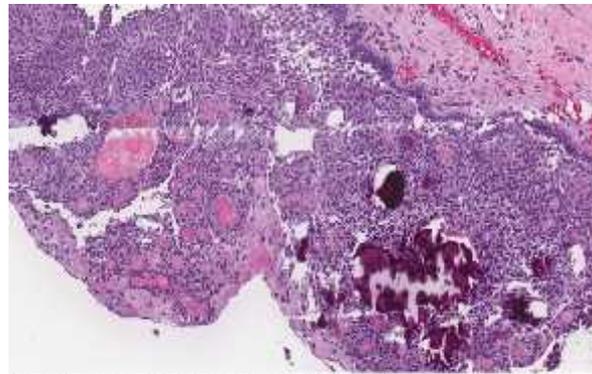
## Pathophysiology

The COC was a unicystic process mainly developed from reduced enamel epithelium or remnants of odontogenic epithelium, gingival tissue, or bone. Dentinoid or an odontome, may be found in the cyst wall, induced by the lining epithelium. The classic COC is frequently a unilocular lesion, but multicystic lesions sometimes be reported. The epithelial lining has odontogenic features with a prominent basal layer consisting of palisade arrangement of columnar or cuboidal cells with hyperchromatic nuclei usually polarized away from the basement membrane.<sup>5</sup> The epithelium may be 6–8 cells thickness. Budding from the basal layer into the underlying connective tissue cells and epithelial proliferations into the lumen are frequently visible. Melanin deposits may be present in the epithelial linings. The most striking feature of the COC is the presence of ghost cells. The ghost cells are usually found in groups, particularly in the thicker part of the epithelial lining.<sup>6</sup>

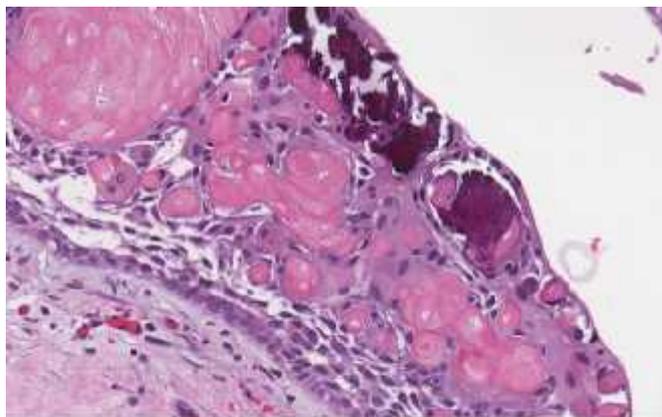
The spinous cells may be widely separated by intercellular edema, and the epithelium around the ghost cells is convoluted. The ghost cells are enlarged, ovoid, ballooned or elongated epithelial cells. They are mainly eosinophilic and although the cell outlines are well defined. The ghost cells represent an abnormal keratinization pattern and have calcification.<sup>5,6,7</sup>



Ca lcifying odontogenic cyst, 3x



Ca lcifying odontogenic cyst epithelium with calcifications and ghost cells, 10x



Ca lcifying odontogenic cyst epithelium with ghost cells, 20x

## Radiographic feature

Radiographically, these COC occur as an intra-osseous lesion appear to have well-demarcated margins, but in some cases, irregular poorly defined margins can also be seen. They are usually unilocular, but few have multilocular lesions. Denser opacities are present if the cyst is associated with a complex odontome. Irregular calcified bodies of varying size and opacity seen in the radiolucent lesion.<sup>8,9</sup> Frequent findings in radiographs are displacement of teeth along with root resorption of adjacent teeth. Local expansion may occur and perforation of the cortical plate is radiologically demonstrable. The extraosseous lesions show mainly a localized superficial bone resorption, or a saucer-shaped radiolucency's and sometimes with adjacent teeth displacement.<sup>8,9,10</sup>

### **Differential diagnosis**

In Central jaw lesion with basal palisading of epithelium the differential diagnosis may be COC, Dentinogenic ghost cell tumor/carcinoma, COC in combination with second tumor, Ameloblastoma, Keratocystic odontogenic tumor and Odontogenic cyst or tumor. If Ghost cells identified from unknown anatomic location the differential diagnosis may be COC, Ameloblastic fibro-odontoma, Pilomatrixoma, Dentinogenic ghost cell tumor, Ghost cell odontogenic carcinoma, Odontoma, and Craniopharyngioma.<sup>11</sup>

### **Treatment**

Treatment for COC is surgical excision, if it is associated with another odontogenic tumor wider excision is indicated. In the presence of complex odontoma conservative removal will be adequate. After treatment of COC the recurrence rate is rare.<sup>8,11</sup>

### **Conclusion**

Calcifying odontogenic cysts may mimic various odontogenic and nonodontogenic lesions and make the diagnosis difficult. Understanding of specific clinical and histological features of various odontogenic and non-odontogenic lesions are important in order to provide accurate diagnosis and treatment.

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