CASE REPORT

# Calcifying Odontogenic Cyst: A Brief Case-Based Insight

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## Abstract

Gorlin initially elucidated the Calcifying Odontogenic Cysts (COC), in 1962. It is an uncommon benign odontogenic cyst that arises from dental lamina residues. These encompass a spectrum, including cystic type, comprising the majority of intraosseous COCs, while neoplastic variant is less common. Certain other odontogenic tumors like Adenomatoid Odontogenic Tumors (AOT), Odontomes or Ameloblastomas might be observed in conjunction with COCs.

A male individual, aged 13 years, came to Oral Pathology OPD of our college, having a problem associated with an uncomfortable swelling involving the upper jaw. Intraorally, a painless and expansile swelling of left maxilla causing slight expansion of palatal cortical plates was evident.

OPG showed presence of a relatively large, unilocular radiolucency, containing fine fleck like radiopacities, having fairly corticated margins. The hematological assays of the patient were within normal limits. Hematoxylin and eosin staining revealed cystic odontogenic epithelial lining, resembling ameloblastomatous features, with characteristic accumulation of ghost cells and dispersed foci of dystrophic calcification. Our case was managed through enucleation.

COCs affect adults during third decades, usually involving the maxillary incisor-cuspid regions. These are characterized by asymptomatic painless swellings, whilst pain, bony expansion, tooth displacement, and root resorption occasionally might be noted. CTNNB1 mutation is a defining feature of COCs. Herein we describe COC in a 13 years old male, with relevant clinico-pathological diagnostic aspect.


**Key words:** Calcifying, Cystic, Odontogenic cyst, Ghost cell, Enucleation

## Introduction

The Calcifying odontogenic cyst (COC), also known as the Gorlin cyst, was first elucidated by Gorlin in 1962.[1] It is an uncommon odontogenic cyst, which exhibits ameloblastoma mimicking cystic epithelium, possessing characteristic localized ghost cell aggregations, which sometimes undergo dystrophic calcification. COCs encompass a wide spectrum, including cystic type, comprising the majority of intraosseous COCs, while solid or neoplastic variant is less common. COCs belong to a spectrum of ghost cell odontogenic entites, including dentinogenic ghost cell tumor (DGCT), ghost cell odontogenic carcinoma (GCOC). They may be accompanied by other odontogenic neoplams, like odontomas, ameloblastomas or adenomatoid odontogenic tumors.[2,3]

COCs comprise lesser than 1% amongst the odontogenic cystic elements, and are quite infrequent. It tends to affect both males and females, with almost equal distribution, during 3rd decades of life. Although, it can involve both maxilla and mandible, maxilla is slightly more affected, with the maxillary incisor-canine region bearing an increased propensity. The COCs are generally asymptomatic swellings, but occasionally these tend to exhibit cortical expansion or swelling, pain, unerupted, displaced or mobile teeth. Radiographically, they exhibit a fairly delineated and corticated, usually unilocular or occasionally multi-loculated radiolucent zone, which tend to contain variable quantity of radiopaque masses. Sometimes, a non-erupted tooth, mostly canine, might be evident, in association with the radiolucency.[2,4,5,6]

COCs tend to originate from cell remnants of dental lamina.[6]

COCs are managed through enucleation, and recurrences are rare. However, when these are associated with other entities like odontomes or ameloblastomas, conservative surgical removal is the modus operandi. Herein, we present

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the occurence of COC in involving a young individual, aged 13 years, possesing relevent clinical-radiological-pathological diagnostic aspects.[2,6]

## Case report

A 13 years old male visited the Oral Pathology OPD at our institution with the complaint of an uncomfortable swelling involving upper jaw. The hematological assays of the patient were within normal limits and the individual did not have any deleterious oral habits. The family and medical histories were non contributory. Intraoral examination revealed an expansile swelling of left maxilla, extending from the palatal aspects of 22 to 28 regions, causing slight palatal expansion (Fig 1). OPG showed the presence of a relatively large, unilocular radiolucency, containing fine flecklike radiopacities, having fairly corticated margins, along with resorption of roots of 22,23,24,25 and displacements of 26 and 27 respectively (Fig 2). The subject underwent surgical enucleation of the cystic contents, after obtaining informed consent. The enucleated contents were subjected to hematoxylin and eosin staining, which revealed cystic odontogenic epithelial lining of 5-9 layers, resembling ameloblastomatous features, showing basal columnar pallisading pre ameloblast like cells and above them stellate reticulum like areas were evident. The defining feature was the presence of aggregates of eosinophilic, amorphous ghost cells within the lining. Dystrophic calcifications were observed.(Fig 3a and 3b) The underlying fibrocollagenous capsule revealed numerous ameloblastomatous proliferations. (Fig 3c) Based on overall clinico-radiologic-histopathologic features, our diagnostic findings leaned towards COC alongwith ameloblastomatous proliferations.

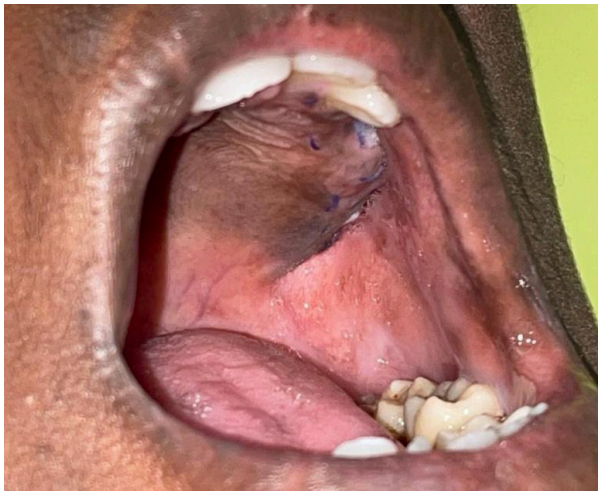

Fig 1- a painless and expansile swelling of left maxilla causing slight expansion of palatal cortical plates, extending from the palatal aspects of 22 to 28 regions

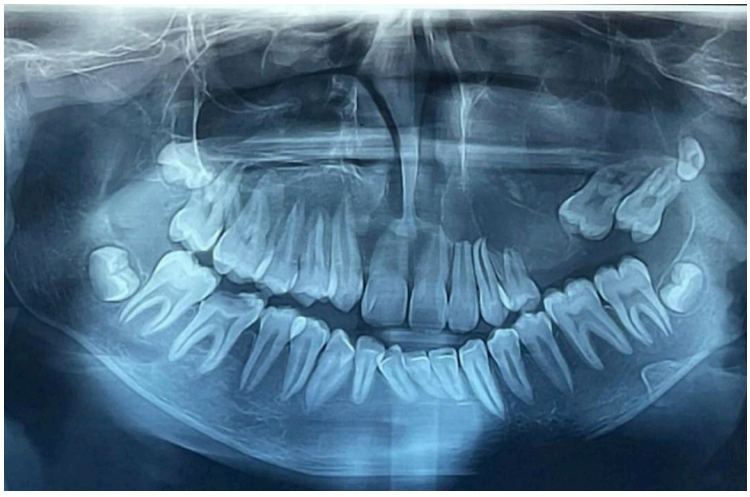

**Fig 2-** OPG showed presence of a relatively large, unilocular radiolucency, containing fine flecklike radiopacities, having fairly corticated margins, along with resorption of roots of 22,23,24,25 and displacements of 26 and 27 respectively.

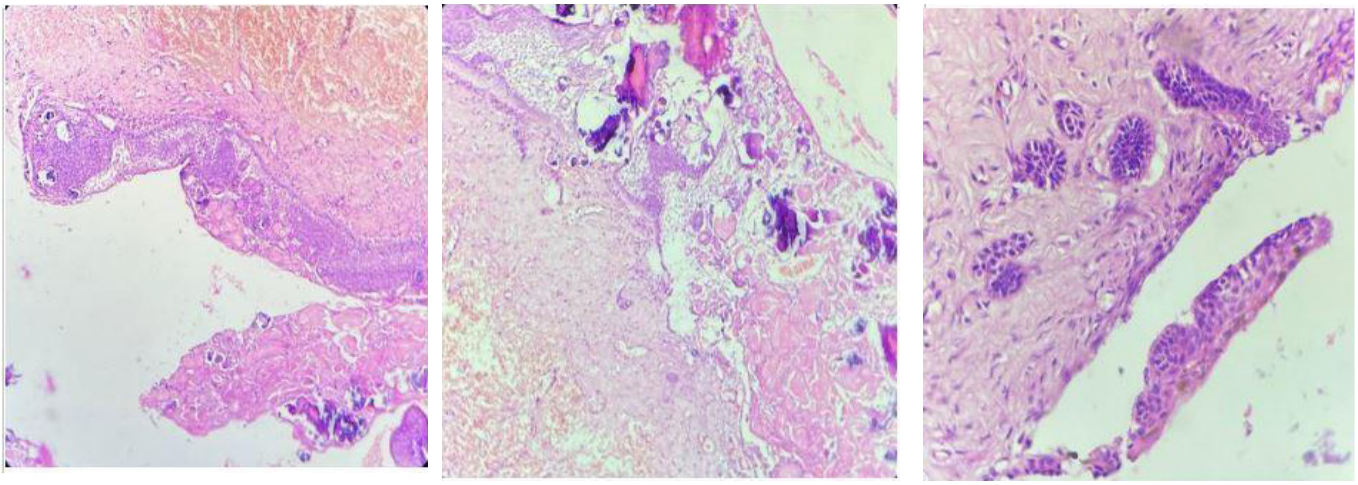

**Fig 3a and 3b-** Hematoxylin and eosin staining (10x magnification) revealed cystic odontogenic epithelial lining of 5-9 layers, resembling ameloblastomatous features, showing basal columnar pallisading pre ameloblast like cells and above them stellate reticulum like areas were evident. The defining feature was the presence of aggregates of eosinophilic, amorphous aggregations of the ghost cells within the lining. Dystrophic calcifications were observed (Fig 3a and 3b) **3c-**The underlying fibrocollagenous capsule revealed numerous ameloblastomatous proliferations.

## Discussion

COCs are infrequent odontogenic cysts, possesing locally aggresive clinical nature.[7] They are benign cysts belonging to an odontogenic lineage, signified by the presence of an ameloblastomaous epithelial lining, comprised of ghost cells which might undergo calcification. It was first described by Gorlin et al in 1962, who were fascinated by the evidence of ghost cells within the cysts, and subsequent histopathologic similarity with the pilomatrixoma of skin. COCs represent a range of lesions having 'odontogenic epithelium'along with characteristic 'ghost cell' features- wherein the ghost cells undergo calcification. The term calcifying signifies the ability of the ghost cells to exhibit dystrophic calcification.[2] The present WHO classification categorizes COCs as odontogenic cysts with variable spectrum (Table 1) 2 Most of them are intraosseous, although extraosseous or peripheral COCs have been reported. 6 Peripheral COCs have been studied in detail by Chranovic and Gomez. These usually affect the mandible more than the maxilla, evident during 6th -8th decades of life, and present as pinkish red, smooth, papular, circumscribed masses involving gingiva or alveolar mucosa. These might resemble gingival fibromas or pyogenic granulomas.[8]

COCs originate from the dental lamina remnants, observed in the dental follicle, bone or periodontal tissues ( cell rest or Malassez or Serres). Some researchers opine that these entites might arise from the reduced enamel epithelium. The capability of the COCs to influence the genesis of hard tissues like dentinoid and relationship with the other odontogenic tumors such as odontomas, ameloblastomas, ameloblastic fibromas or adenomatoid odontogenic tumors point towards a hamartomatous nature of the entity. COCs are associated with mutations of beta catenin (CTNNB1) involving Wnt signalling pathway. LEF1 positivity (transcription factor of Wnt signalling pathway) is also evident in a lot of cases.[4,9]

COCs generally tend to occur during the 3rd decade of life, with the highest incidence typically seen in the second decade of life. But COCs can affect individuals across a wide age range, ranging from 5-92 years. COCs associated with odontomes generally occurs in younger persons.[10] Our subject was a 13 years old. Mostly they range between 2-4 cm in largest diameter, and generally larger than radicular cysts or periapical granulomas.11 There is no clear gender preference, but, a few studies have noted a mild skewing towards males.[2] In our case, the individual was a male. Intraossoeus COCs can involve both maxilla and mandible, with maxilla having an increased propensity for involvement. Mostly the maxillary incisor-canine region gets affected, followed by the posterior mandibular region. They can present as asymptomatic swellings, which are detected through radiographic examinations. Additionally, pain, unerupted teeth, tooth displacement, mobility or divergence might be noted in some instances. Expansion and distortion of the buccal or palatal cortical plates might be noted.[4]

Our case showed a painless and expansile swelling of maxilla causing slight expansion of palatal cortical plates, extending from the palatal aspects of 22 to 28 regions.

Radiologically, usually unilocular or rarely a multilocular radiolucency, possesing well-delineated corticated margins, or infrequently scalloped margins, might be evident, with many cases containing variable degrees of radiopaque material- in form of irregular fleck like calcifications or dense toothlike radiodensities-representing foci of dystrophic calcifications. Approximately, a third of the radiolucent zones exhibit an association with a non-erupted tooth, most commonly the maxillary cuspid. Alongside, expansion of cortical plates, tooth displacement, resorptive changes of the roots and displacement involving regional tooth roots might be noted. 12,13 Our case showed the presence of a relatively large, unilocular radiolucency, containing fine flecklike radiopacities, having fairly corticated margins, along with resorption of roots of 22,23,24,25 and displacements of 26 and 27 respectively.

Surgical enucleation is the treatment of choice, which was also done in our case.[6]

Grossly, COCs are usually unicystic entities, with focal luminal proliferations, and no solid zones.[4]

Histopathologically COCs are distinctive cystic lesions. The cystic odontogenic epithelial lining is 4-10 layers in thickness, and resembles an unicystic ameloblastomatous lining. The basal layer exhibit columar or cuboidal pallisaded cells, with nuclei polarized away from basement membrane. The supra

**Key Features**

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| **Calcifying odontogenic cyst**<br>➢ **Simple cyst** | Typical cystic lesion, ameloblatomatous lining with ghost cells,induces occasional variants with proliferating epithelium |
| ➢ **Cyst associated with odontoma** | Cyst associated with an odontoma in the wall |
| ➢ **Dentinogenic ghost cell tumor** | Solid lesion with ameloblastomatous epithelium, ghost cells and dentinoid |
| ➢ **Ghost cell odontogenic carcinoma** | Malignant lesion, ameloblastoma like epithelium, ghost cells and dentinoid |

**TABLE 1** Present WHO classification of Calcfying odontogenic cyst

basal layers of cells mimicks stelate reticulum like areas. The most distinguishing characteristics is identification of focal conglomerations of ghost cells; involving cystic lining or projecting towards the lumen or occasionaly protruding towards the fibrous wall of COCs. The ghost cells are transformed epithelial cells- pale, amorphous, balloned or elliptical cells with distinct pinkish eosinophilic cystoplasm, wherein cellular silhouette is preserved but the nuclear outline is lost. These ghost cells often undergo dystrophic calcification- thus giving the appearance of basophilic aggregates. Various hypotheses try to explain the genesis of ghost cells- including coagulative necrosis, cellular degenerations, accumulations of enamel proteins, aberrant keratinization of odontogenic epithelium etc. Melanin pigmentation sometimes very infrequently might be found within the cystic lining or ghost cells. Dentinoid matrix might be evident adjacent to epithelial lining, as a result of an inductive effect of cystic lining exerted to subjacent stromal capsule. The ghost cells projecting into the cyst wall- can produce a foreign body giant cell reaction alongside occasional dystrophic calcification. The cystic capsule might contain rests of odontogenic epithelium, satellite cysts, or ameloblastomatous differentiation with juxtaepithelial dentinoid, or odontome like areas. [2,4,5,7]

In our case, the enucleated contents were subjected to hematoxylin and eosin staining, which revealed cystic odontogenic epithelial lining of 5-9 layers, resembling ameloblastomatous features, showing basal columnar pallisading pre ameloblast like cells, and above them regions simulating stellate reticulum were evident. A notable characteristic was existence of aggregates of eosinophilic, amorphous aggregations of the ghost cells within the lining. Dystrophic calcifications were observed. The underlying fibrocollagenous capsule revealed numerous ameloblastomatous proliferations.

Immunohistochemical evaluations have been performed with respect to the the basal cells pertaining to cystic lining epithelium, the suprabasal cells, and the ghost cells. They demonstrate moderately high levels of amelogenin and Cytokeratin[6]. The basal and suprabasal layers of cells demonstrated an upregulation of Cytokeratin 19, but not the ghost cells. The ghost cells exhibit accumulation of amelogenin and hard keratins in their cytoplasm.[14]

The malignant equivalent of COCs are Ghost cell odontogenic carcinomas (GCOC). These are rare, and demonstrate cytonuclear pleomorphism, and mitotic activity with invasion of surrounding tissues.[15]

COCs need to be differentiated from lesions such as odontogenic keratocyst, dentigerous cysts, unicystic ameloblastoma, adenomatoid odontogenic tumors, ameloblastic fibroma, calcifying epithelial odontogenic tumor and myxomas.[2,13]

COCs are managed through enucleation and recurrences are rare. Larger cysts demand marsupialization followed by enucleation. Whilst those associated with odontomes or ameloblastomas are treated by surgical excisions.[2,6]

COCs are unique, enigmatic entities possesing a wide spectrum of manifestation, ranging from simple cystic forms to solid entities, association with oodontogenic tumors and even a malignant counterpart. Ghost cells are an integral component of these pathologies. Proper and thorough understanding of these entities, regarding the pathogenesis of ghost cells, and further future research studies encompassing histopathological, genetic and immunohistochemical aspects of these unique lesions will help in our elucidation and understanding of this novel pathologies.

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