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**Muthu Ponnusamy Sumathy**  
Associate Professor, Department  
of Oral Pathology, Tamil Nadu  
Government Dental College and  
Hospital, Chennai, Tamil Nadu,  
India

**Subramanian Kuzhali**  
Senior Assistant Professor,  
Department of Oral Pathology,  
Tamil Nadu Government Dental  
College and Hospital, Chennai,  
Tamil Nadu, India

**Viswanathan Harishnath**  
Senior Assistant Professor,  
Department of Oral Pathology,  
Tamil Nadu Government Dental  
College and Hospital, Chennai,  
Tamil Nadu, India

**Santiago Gnanadeepam**  
Associate Professor, Department  
of Oral Pathology, Tamil Nadu  
Government Dental College and  
Hospital, Chennai, Tamil Nadu,  
India

**Packiaraj Selvajothi**  
Assistant Professor, Department  
of Oral Pathology, Tamil Nadu  
Government Dental College and  
Hospital, Chennai, Tamil Nadu,  
India

**Sivaraj Ramyadevi**  
Assistant Professor, Department  
of Oral and maxillofacial  
surgery, Tamil Nadu  
Government Dental College and  
Hospital, Chennai, Tamil Nadu,  
India

**Corresponding Author:**  
**Subramanian Kuzhali**  
Senior Assistant Professor,  
Department of Oral Pathology,  
Tamil Nadu Government Dental  
College and Hospital, Chennai,  
Tamil Nadu, India

## **Cystic lesion in the maxilla: A case report of calcifying odontogenic cyst with review of literature**

**Muthu Ponnusamy Sumathy, Subramanian Kuzhali, Viswanathan Harishnath, Santiago Gnanadeepam, Packiaraj Selvajothi and Sivaraj Ramyadevi**

### **Abstract**

Calcifying odontogenic cyst (COC) is a developmental odontogenic cyst which was first described as a separate entity by Gorlin *et al.* in 1962. COC characterizing as a cyst and neoplasm accounts for only about 1% of all odontogenic cysts. This unique lesion occurs mostly as intraosseous and rarely as a peripheral variant. Due to its diversified histological presentations and spectrum of variants the pathogenesis and the biological behavior needs to be understood. In this article, we present a case of COC in the maxilla of a 14 year old female with brief discussion.

**Keywords:** Calcifying odontogenic cyst, cystic lesion, mixed radiolucent - radiopaque lesion

### **1. Introduction**

Head and neck region is known for its diverse pathologies owing to its complicated structural anatomy. The existence of odontogenic epithelial remnants contributes to the development of various odontogenic cysts and tumors in the jaw region. Most of the odontogenic cysts are usually benign and asymptomatic in the early stages. When unattended, they cause localized destruction of tissues and become painful when infected. Calcifying odontogenic cyst (COC) is an uncommon odontogenic cyst with varied clinical presentation ranging from a simple cyst to a solid neoplasm and may have association with other odontogenic lesions often posing problems in classification and assessment of its clinical behavior. <sup>[1]</sup> Radiologically, COC most often present as a well-defined radiolucent lesion with irregular radiopacities <sup>[2]</sup>, frequently imitating various lesions of both odontogenic and non-odontogenic origin. Final diagnosis is accomplished only after a comprehensive histopathological evaluation.

### **2. Case Report**

A fourteen year old female patient reported to the Tamil Nadu Government Dental College and Hospital, Chennai with a complaint of swelling in the right upper front tooth region with associated pain and discharge for a duration of six and 4 months respectively. Swelling was insidious in onset and has gradually attained the present size of approximately 3 x 2cms obliterating the right nasolabial fold. The swelling was diffuse in nature and tender on palpation (Fig: 1). Intraorally, a swelling of size 3 x 2 cms was seen obliterating the buccal vestibule in relation to the maxillary right quadrant. The swelling extended from the distal aspect of the right central incisor to mesial aspect of the right first premolar. Facial asymmetry was evident. The overlying mucosa was intact imparting a bluish hue revealing its cystic nature which was later confirmed by aspiration of a straw colored fluid from the swelling. Intraoral examination revealed the presence of a palatally displaced and partially erupted right permanent maxillary canine, retained right deciduous maxillary canine and a buccally displaced rotated maxillary right first premolar (Fig: 2).

Intraoral periapical and maxillary occlusal radiographs revealed a well-defined unilocular radiolucent lesion beneath the retained deciduous canine (Fig: 3). In addition to the divergence of the roots of the involved teeth, the presence of radiopaque foci within the corticated radiolucent lesion was evident in the panoramic view (Fig 4).

After analyzing the clinical and radiological findings i.e., mixed radiolucent-radiopaque lesion in the anterior maxillary region of a young female, the following differential diagnoses namely adenomatoid odontogenic tumour, ameloblastic fibro odontoma, calcifying odontogenic cyst,

calcifying epithelial odontogenic tumour and ossifying fibroma were considered.

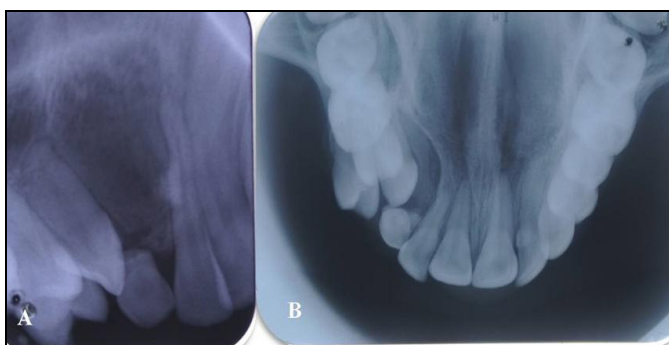
Under local anaesthesia, an incision was made in the buccal vestibule and a bone window was created. The lesion was exposed, removed in toto (Fig 5) and submitted for histopathological examination. On gross examination, the specimen was grayish white in color, soft to firm in consistency measuring 2 x 1 cms. The inner surface of the specimen had numerous surface projections enclosed by an outer leathery cyst wall (Fig: 6). The entire specimen was processed and sections were examined after proper hematoxylin and eosin staining. On examining the sections, we observed a cystic lesion with an odontogenic epithelial lining and fibrous connective tissue wall. The lining odontogenic epithelium was characterized by tall columnar basal cells with hyperchromatic palisaded nuclei polarized away from the basement membrane. The superficial part of the epithelium showed stellate reticulum like cells, calcifications and eosinophilic masses resembling ghost cells which were devoid of nucleus but with well-defined cell outlines (Fig:7).



**Fig 1 A, B:** Extraoral photographs showing a diffuse swelling obliterating the nasolabial fold in the maxillary anterior region (R).



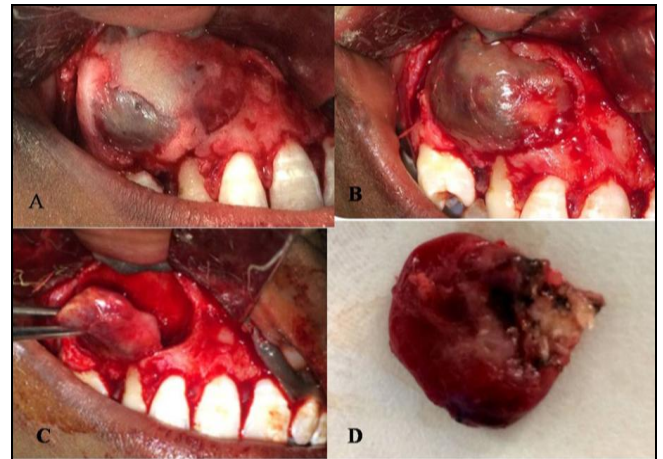
**Fig 2 A, B:** Intraoral photograph reveals a swelling of size 3 x 2 cms extending from the distal aspect of right maxillary central incisor to the mesial aspect of the right first premolar.



**Fig 3 A, B:** IOPA and maxillary occlusal radiographs reveal a well-defined unilocular radiolucent lesion beneath the retained deciduous canine.



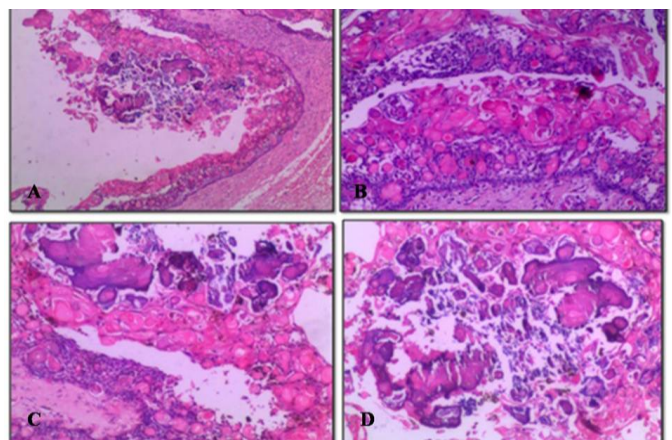
**Fig 4:** The panoramic view reveals presence of radiopaque foci within the radiolucent lesion



**Fig 5 A, B C, D:** Under local anaesthesia, an incision was made in the buccal vestibule. A bone window was created, Lesion was removed in toto.



**Fig 6 A, B:** Gross specimen. The inner surface of the specimen had numerous surface projections enclosed by an outer leathery cyst wall.



**Fig 7 A B, C, D:** At lower magnification, a fibrous cyst wall lined by odontogenic epithelium is seen. At higher magnification, the basal cells are columnar with hyperchromatic, palisaded nuclei resembling ameloblasts. The superficial part of the epithelium showed stellate reticulum like cells, calcifications and eosinophilic masses resembling ghost cells.

### 3. Discussion

Despite the fact that the first description of calcifying odontogenic cyst was made in the German literature as early as in 1932 by Rywkind, it was initially assumed to be a variant of cholesteatoma [3]. Many case reports were documented in the next three decades highlighting the presence of unusual keratinization or the 'ghost cell' formation, however under various terms namely atypical ameloblastoma, a type of odontoma and odontogenic tumor of epithelial and mesenchymal origin. In 1962, Gorlin made an audacious attempt claiming that calcifying odontogenic cyst as a distinct clinicopathological entity, which is now eponymically termed as "Gorlin cyst". Mislead by the abnormal keratinization of both the odontogenic and the metrical cells, Gorlin considered calcifying odontogenic cyst was an oral analogue of pilomatricoma, a well-recognized skin lesion, which was later proved to be incorrect. In fact calcifying odontogenic cyst is related to craniopharyngioma microscopically, a rare intracranial neoplasm which in turn histogenetically related to Rathke's pouch [3], an invagination of the superior aspect of the primitive oral cavity that gives rise to the anterior lobe of the pituitary gland. Concurrently in 1963, Gold *et al.* published a report of four cases and named the lesion as 'keratinizing and calcifying odontogenic cyst.' Consequent to these landmark publications, calcifying odontogenic cyst acquired the attention of many researchers [1].

Although COC was recognized by WHO as a non-neoplastic cystic lesion in 1971, the accumulating information has led to a dilemma whether COC is truly a cyst or a neoplasm that is cystic [4]. Based on the dualistic concept, in 2005 WHO classification COC was renamed as *Calcifying cystic odontogenic tumour* and was placed under benign cystic neoplastic category, whereas the solid variant was included as a separate entity and termed as *dentinogenic ghost cell tumour* (DGCT). [5]

Studies conducted by (Hong *et al.* & Ledesma-Montes *et al.*) revealed that majority of the ghost cell lesions are simple cysts with a benign course which may arise either alone or associated with odontomas and rarely recur. Hence, in the latest edition of the WHO classification (2017), the simple cyst form is again renamed as calcifying odontogenic cyst. [6]

COC is a rare lesion which accounts for about 0.3 - 8% of all the odontogenic lesions. [7] COC arise from the reduced enamel epithelium or odontogenic epithelial remnants (cell rest of Serres) [8] present within the bone and the gingiva, former producing the intraosseous forms and the latter giving rise to the peripheral variants. The more common intraosseous forms (70%) clinically present as a painless hard swelling affect the maxilla and mandible with equal frequency along with the site predilection for incisor – canine region [9]. The maxillary lesions are more frequent in the canine – premolar region, while some of the mandibular lesions may cross the midline. Majority of the cases are cystic (86-98%), while the solid form comprises only 2-16% of the cases [9]. Occasionally, lesions are asymptomatic and may be detected only during the radiographic evaluation of an impacted tooth. The peripheral lesions arise as a pink to red, sessile or pedunculated soft tissue swelling of the gingival [9, 10].

COC affects both the gender with equal frequency and is most common in the second decade of life. Some studies show a bimodal [11, 12] occurrence affecting the individuals in the sixth decade, particularly the extra osseous forms.

The intra osseous COCs have a variable radio graphical presentation ranging from a well-defined unilocular or

occasionally a multilocular radiolucent lesion to a mixed radiopaque – radiolucent lesion within which the irregular calcified masses of variable sizes producing a salt and pepper effect, often mimicking various odontogenic and fibrous lesions [2]. Root resorption and divergence are common radiographic findings. In addition, one-third of the cases are associated with impacted teeth, particularly canine [13].

As defined in the WHO classification of 1992 [10], COC is 'a cystic lesion in which the epithelial lining shows a well-defined basal layer of columnar cells, an overlying layer that is often many cells thick and that may resemble stellate reticulum, and masses of "ghost" epithelial cell that may be in the epithelial lining or in the fibrous capsule. The "ghost" epithelial cells may become calcified. Dysplastic dentine may be laid down adjacent to the basal layer of the epithelium, and in some instances the cyst is associated with an area of more extensive dental hard tissue formation resembling that of a complex or compound odontoma.'

Ghost cells are enlarged, ballooned, ovoid or elongated elliptoid cells within the epithelial lining. Though these eosinophilic cells have well-defined cell outlines, they may fuse to form amorphous sheets, which further undergo dystrophic calcification. Nuclei are either absent or only the nuclear outline is retained. The presence of ghost cells in COC is characteristic but not pathognomonic, as they are occasionally present in other lesions like odontoma, ontoameloblastoma, ameloblastoma, ameloblastic fibrodontoma, and clear cell odontogenic carcinoma. The ghost cell formation may occur as a result of coagulative necrosis and dystrophic calcification or due to the abnormal keratinization of the odontogenic epithelium. [10]

Some researchers believe that the atypical dentin/ dentinoid is formed as a result of inductive ability of the odontogenic epithelium on the underlying mesenchymal tissue, while another school of thought says that it might have originated from the ghost cells. [9, 14]

A thorough histopathological examination of the surgical specimen becomes mandatory, as COC is known for its association with other odontogenic lesions like odontoma (22-47% cases), adenomatoid odontogenic tumour and ameloblastoma [15].

### 4. Conclusion

COCs are treated by simple enucleation and curettage, unless associated with other odontogenic lesions like ameloblastoma, in which case a wider surgical excision is needed. The recurrence rate of COC is less than 5%. Although rare, the malignant transformation has been reported in recurrent and long standing cases. [8] Therefore, long-term post-operative follow-up of the patient is essential.

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