Case Report
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Gorlin Cyst of Maxilla: A Rare Case Report

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Abstract

Calcifying odontogenic cyst was first described by Gorlin, so the name, Gorlin cyst. Calcifying Odontogenic Cyst (COC) is rare in occurrence and usually asymptomatic in nature. It is an uncommon benign odontogenic neoplasm which may show characteristics of both a cyst and solid neoplasm. Its clinical and radiographic features may mimic other odontogenic cysts or tumors, and a definitive diagnosis can only be arrived on histological examination. We report a classic case of calcifying odontogenic cyst in a 50 year old patient.

Keywords: Calcifying Odontogenic Cyst, Gorlin Cyst, Odontogenic Cyst

1. Introduction

Calcifying odontogenic cyst is a rare developmental odontogenic cyst, represent about 1% of all odontogenic cysts[1]. It was first described by Gorlin in 1962 and hence known as Gorlin cyst. It has clinical and histological features of a cyst and a solid tumour. In 2005, the World Health Organization (WHO) classified COC as a neoplasm and named Calcifying Cystic Odontogenic Tumour (CCOT) for benign cystic type, the Dentinogenic Ghost Cell Tumour (DGCT) for the benign solid type lesions, and the malignant one as ghost cell odontogenic carcinoma[2]. It was reclassified as a cystic lesion again by the WHO in 2017.

It presents as a well-circumscribed, solid or cystic lesion derived from odontogenic epithelium, which originates from reduced enamel epithelium or remnants of odontogenic epithelium in the follicle, gingival tissue or bone, contains "ghost cells" and spherical calcifications. It can be identified as a slow growing painless lesion with equal incidence in maxilla and mandible, with the majority affecting the region anterior to molar teeth (incisor/canine region)[3]. It may also show bimodal age distribution, first peak at the second decade and the second peak at the sixth to the seventh decade with no particular sex predilection[4].

Radiographically, Gorlin cyst may be seen as a unilocular or multilocular radiolucent lesion with either well-circumscribed or poorly defined margins[5]. Histopathologically, COC shows a lining of odontogenic epithelium with either columnar or cuboidal basal cells resembling ameloblasts with a fibrous wall. Stellate reticulum-like cells over the basal cell layer and ghost cells, which may be calcified, are also present in the lining of the cyst[5]. The preferred treatment of COC is surgical enucleation.

We report a case of COC in a 50-year-old male with emphasis on its clinical and radiographic features, histopathological characteristics and surgical management.

2. Case Report

A 50-year-old male reported to our Out-Patient Department with a history of swelling over the alveolus corresponding to right maxillary anteriors for two months with intermittent pain. Extra orally, a diffuse swelling on the right anterior maxilla of size 2.5 cm × 2 cm in greatest dimension from the midline of the upper lip to the right corner of the mouth with obliteration of the nasolabial fold was present (Figure 1). Intra orally, buccal vestibule was obliterated in relation to 11,12,13 and 12,13,14 was not

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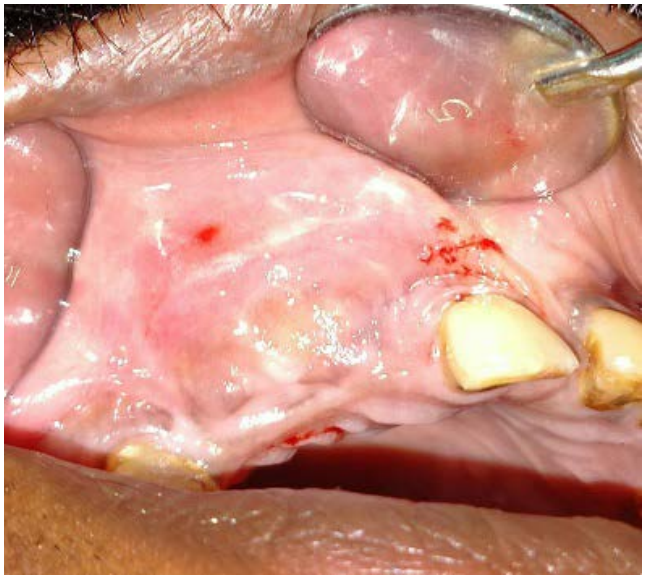


Figure 1. Intra oral swelling over anterior maxilla.

present clinically. The mucosa over the lesion appeared normal. On palpation, the swelling showed varying consistency from firm to soft whereas some areas showed "egg shell crackling" representing the possibility of buccal cortical plate perforation. A straw-coloured fluid was obtained through fine needle aspiration cytology.

Upper anterior occlusal radiographic view showed ill-defined radiolucency with mild root resorption of 11. Ortho-Pantamo-Graph (OPG) presented with similar findings with no impacted teeth and apparently normal bilateral maxillary sinus (Figure 2 and 3). Aspiration of the cystic content yielded a straw-coloured fluid (Figure 4). A differential diagnosis of Residual cyst, Adenomatoid Odontogenic Tumour and Calcifying Odontogenic Cyst were made.

Complete surgical enucleation of the cyst was done under local anaesthesia and specimen was sent for histopathologic examination (Figure 5a and b). Histopathologic examination revealed cyst of odontogenic

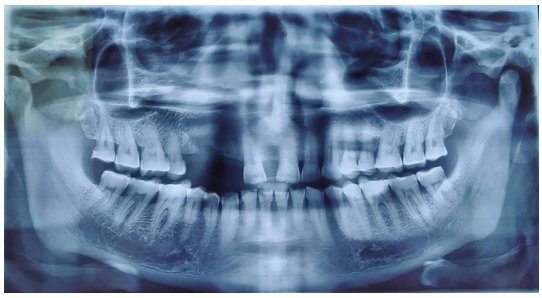


Figure 2. OPG showing ill-defined radiolucent lesion.

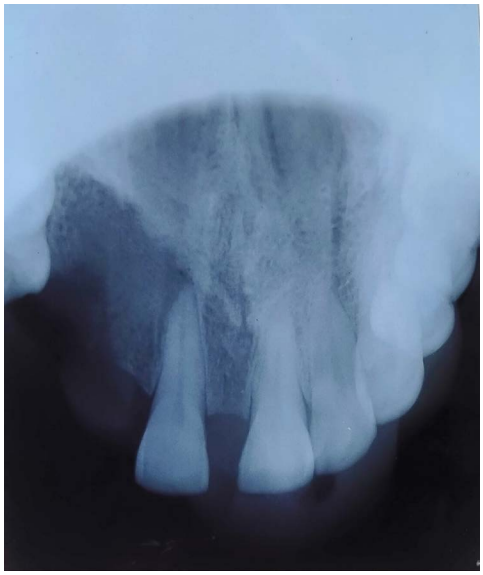


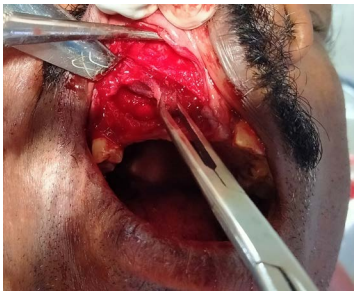
Figure 3. Anterior occlusal view showing radiolucent lesion with mild root resorption.

Figure 4. Positive fine needle aspiration with straw coloured fluid.

lining epithelium with loosely cohesive stellate and spindle cells, numerous ghost cells and few calcification. Von Gieson special staining confirmed the presence of faint yellow coloured ghost cells and dark pink coloured areas of dentinoid material (Figure 6). Definitive diagnosis of COC was made after histopathologic examination.

3. Discussion

Calcifying Odontogenic Cyst is a heterogeneous group of odontogenic lesions that represent diverse clinicopathological features. This distinct pathological entity was first described by Gorlin[6] in 1962. He pointed out the histological resemblance of COC to cutaneous calcifying epithelioma of Malherbe[6] and described the relationship between COC and calcifying epithelial



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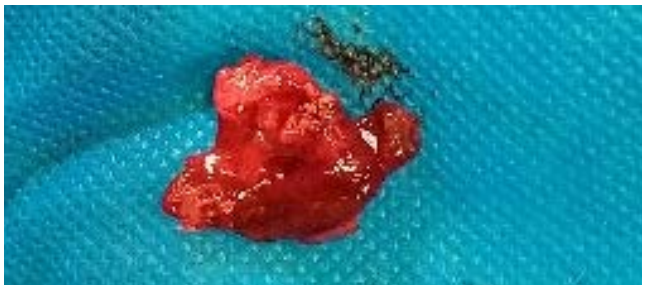

b

Figure 5. a. showing intraoperative picture, and b. Post enucleation specimen sent for Histopathology.

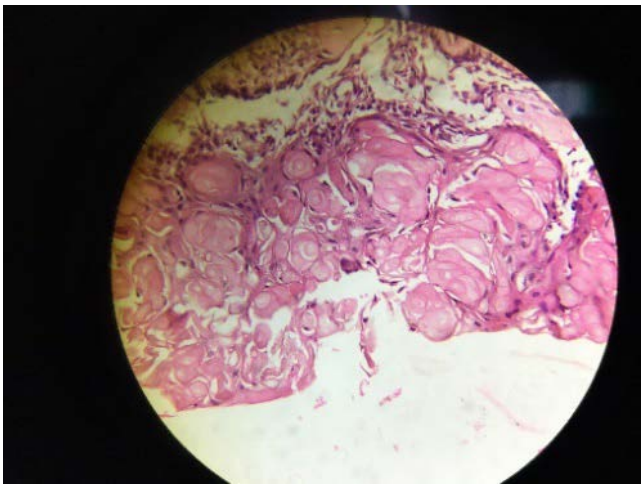

Figure 6. Showing eosinophilic dentinoid material adjacent to sheet of ghost cells.

odontogenic tumour[7] in 1963. In the past, COC was considered as a variant of ameloblastoma or a sort of odontoma. As per 2005 WHO guidelines, the term COC was introduced as a basic subtype of Odontogenic ghost cell lesions. In 2017 it was reclassified as cystic lesion again by WHO.

WHO in 1992 defined COC as a cystic lesion in which the epithelial lining shows a well-defined basal layers of columnar cells, an overlying thick layer of stellate reticulum like cells and masses of 'ghost' epithelial cell that may be in the epithelial lining or in the fibrous capsule[8]. In 2005 edition, it was defined as "A locally invasive neoplasm characterized by ameloblastoma-like islands of epithelial cells in a mature connective tissue stroma. Aberrant keratinization can be found in the form of ghost cells associated with varying amounts of dysplastic dentin[9]. The other names include Gorlin's cyst, Keratinizing ameloblastoma, and Melanotic ameloblastic odontoma[10] (Table 1).

Table 1. Classifications

Author	Classification
Praetorius *et al.*[11]	Type 1 – Cystic a) Simple unicystic b) Odontoma producing type c) Ameloblastomatous proliferating type Type 2 – Neoplastic type – Dentinogenic ghost cell tumour
Buchner[12]	Central Calcifying odontogenic cyst Peripheral Calcifying odontogenic cyst
Toida[13]	Cyst Neoplasm a) Benign Calcifying ghost cell odontogenic tumour b) Malignant - Dentinogenic ghost cell tumour

According to the study by Devildos in Indiana university of dentistry, they concluded that a Maxillofacial Surgeon on an average is probably going to see only one or two cases of COC during his/her career, accounting for 1% of all odontogenic cysts with equal incidence in maxilla and mandible, 75% of cases reported in canine and premolar area[14]. Although COC is most prevalent in second decade, there are cases reported in various age groups from 1 year to 80 years[11]. It shows negligible gender and racial predilection.

Most of the cases are asymptomatic which are diagnosed during routine radiographic examinations. It may present as painless expansile lesion when present in the jaws. Rarely COC leads to cortical perforation. Sometimes displacement of adjacent teeth can be seen[15]. It can also be found in association with odontoma or impacted tooth[16]. COC can be seen peripherally over the gingiva as painless swellings or nodules which are

identified as nonspecific well-circumscribed sessile or pedunculated mass with a smooth surface.

On a radiograph, it can be seen as an area of well-defined or ill-defined radiolucency intermingled with areas of opacification due to the focal areas of calcifications[14]. Occasionally COC is also associated with impacted tooth[17]. A definitive diagnosis of COC can be made only by histological means as it mimics other odontogenic cysts clinically and radiologically. Cystic lining shows proliferation and within the epithelium, cells undergo characteristic ghost cell keratinisation. The extent of cell proliferation can be used as a potential indicator of behavior, treatment response, and recurrence[18].

When dentinoid material is found in abundance in juxtaposition to the proliferative lining epithelium or adjacent to a sheet of ghost cells, the lesion is solid and named as dentinogenic ghost cell tumor. The presence of ghost cells within the proliferative odontogenic epithelium is pathognomonic. Ghost cell is identified as an enlarged, ballooned, ovoid or elongated, ellipsoid somatic cell with eosinophilic cytoplasm but without a nucleus and shadow appearance. Hence these cells are also called shadow cells or translucent cells[19]. Takata and coworkers[20] reported that the ghost cells express enamel-related proteins. Badger and Gardner[21] found that both calcifying odontogenic cyst and the craniopharyngioma have similar immunoreactivity to low and high molecular weight cytokeratins and involucrin, leading to the fact that COC is the oral counterpart of craniopharyngioma. These ghost cells may be accentuated in routine paraffin sections by the van Gieson, Goldner, or Ayoub-Shklar histochemical stains helpful in the solid lesions in which dentinoid material is seen in abundance while the ghost cells are less conspicuous. Correlating the clinical and radiographic findings, the differentials include Calcifying epithelial odontogenic tumour, Adenomatoid odontogenic tumour, Ameloblastic fibro-odontoma, Dentigerous or other odontogenic cysts[22].

According to the histopathological findings, Ghost cell keratinisation can be observed in Odontomas, Ameloblastomas, Ameloblastic fibro-odontomas and Ameloblasticodontomas. Peripheral lesions may resemble Gingival fibroma, Gingival cyst or Peripheral giant cell granuloma.The recommended treatment option for COC is conservative surgical enucleation[6]. The risk of recurrence is low (3%–11%) but requires a regular radio-clinical follow-up of six months after the procedure and further once in a year for five years to evaluate for complete re-ossification. Recurrence is seen commonly in neoplastic lesions such as dentinogenic ghost cell tumours. Peripheral lesions are usually managed by conservative excision whereas radical excision of the lesion is preferred when neoplastic component of COC predominates[23].

4. Conclusion

Calcifying odontogenic cyst is a rare odontogenic cyst which is mostly asymptomatic. Various authors have classified the entity according to its histological findings. Presence of ghost cell is one of the characteristic features of Calcifying odontogenic cyst. Complete excision is the treatment of choice. Even though the recurrence rate is less, regular follow up is recommended.

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Conflict of Interest: Nil

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