

CALCIFYING CYSTIC ODONTOGENIC TUMOR- A RARE CASE REPORT

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Abstracts

Calcifying cystic odontogenic tumours (CCOTs) account for around 2% of all odontogenic tumours. The term "Calcifying Ghost Cell Odontogenic Cyst" (CGCOC) is also commonly used to describe this condition. It was first recognized as a distinct entity by Gorlin et al. in 1962. It has its roots in the odontogenic stage of development. This unusual lesion can resemble either a solid tumour or a cyst. Although extra-osseous or peripheral forms of the disease are possible, intra-osseous lesions are much more common [1]. There have been several changes in the terminologies used to describe this condition and various classifications have been made by several authors since its first description. In 2005, the World Health Organization(WHO) reclassified the growth as a calcifying cystic odontogenic tumour (CCOT). This report describes a case of CCOT with an impacted mandibular 3rd molar.

Keywords: Calcifying cystic odontogenic tumor, ghost cells, Gorlin cyst, Marsupialization

Introduction

Calcifying Cystic Odontogenic Tumour (CCOT) or Calcifying Ghost Cell Odontogenic Cyst (CGCOC) is a developing odontogenic cyst. Gorlin et al. were the first to categorise it as a distinct entity in 1962.[1-2] The

WHO currently classifies CCOT as a benign cystic neoplasm of an odontogenic origin. It presents with an ameloblastoma-like epithelium and ghost cells which may have the tendency to calcify.[3-4]

This condition is distinct from the Pindborg tumor (CEOT - Calcifying Epithelial Odontogenic Tumour). It has been considered to have an odontogenic origin due to the following factors: 1) It's association with a developing or unerupted tooth; 2) Histological sections indicating the presence of a basal cell layer composed of ameloblastic cells and the formation or induction of dentinoid material; and 3) the lesion's development in close proximity to dental structures. Some of the reported literature indicate that this neoplasm could have origin in the dental follicle at the top of an unerupted tooth, which includes dental lamina remnants or reduced enamel epithelium (REE).[2-3]

Calcifying epithelial odontogenic cyst (CEOC) is most commonly seen as a central intra-osseous lesion, while a peripheral extra-osseous site in soft tissue is unusual. CCOT frequently appears as a painless, slow-growing tumour spread uniformly over the maxilla and mandible, with a predilection for the front half (incisor/canine region). Because the lesion lacks differentiating clinical or radiological signs and has variable biological activity, a definite histological diagnosis is done. [1-7]

The objective of presenting this case report is to demonstrate the efficacy of marsupialization in a large CCOT located on the right mandibular angle region extending on to the ramus and the subcondylar region. In this specific case, excision as a single step surgery would jeopardize the continuity of lower border of mandible and would require a reconstruction plate and also would cause neurosensory deficit of inferior alveolar nerve.

Case-report

A 28-year-old female patient visited the Oral Medicine and Radiology department at Dayananda Sagar College of Dental Sciences complaining of edema on the right side of the lower jaw for two months. The patient revealed a history of edema that had been present for the past two months. It was sudden in onset and gradually progressed to the present size. On extra-oral examination, there was no noticeable facial asymmetry. However, on palpatory examination, a diffuse edema was evident on the right side of the mandibular body. The swelling was hard in consistency, non-tender on touch and the skin over the swelling resembled the surrounding normal skin.

On intra-oral examination, a diffuse edema was present in relation to right mandibular posterior tooth region obliterating the buccal vestibule extending from 45-48 region. Tooth 48 was missing and Grade I mobility of 47 was noted. The swelling was found to be firm in consistency with a smooth surface and non-tender.



Figure 1- Diagnostic OPG revealed a radiolucent lesion on right angle region of the mandible.

A diagnostic OPG was taken in order to assess the lesion radiographically. The OPG revealed an impacted 48 present within a large radiolucent lesion involving the mandibular body from the 47 region, mandibular angle and the ramus on the right side (Figure 1).

Histopathological examination was done via an Incisional biopsy. It was reported as “Lining epithelium of varying thickness with a relatively flat connective tissue interface. The wall of the cyst is uninfamed with parallel arrangement of collagen fibers. The epithelium shows hyper-chromatic cuboidal cells as the basal layer and superficial stellate reticulum like cells with some whorled islands showing ghost cell like changes. In some areas, only basal layer of the epithelium is seen. Histopathology is suggestive of a Gorlin cyst”.

Based on the investigations, a treatment plan was devised -Initial marsupialization followed by enucleation after 3 months. An enucleation procedure has the risk of causing a fracture of the mandibular border (lower). Hence, a two stage ‘Marsupialisation – Enucleation’ treatment was planned.



Figure 2- After Marsupialization of the lesion

Marsupialisation of the cyst (figure-2) was done under LA through an intraoral incision extending from 47 region to the ascending border of the ramus of mandible. The impacted 48 was removed and inferior alveolar neurovascular bundle was preserved. Medicated pack was placed in the defect and the patient was followed up periodically.



Figure 3: one month postoperative intraoral photograph

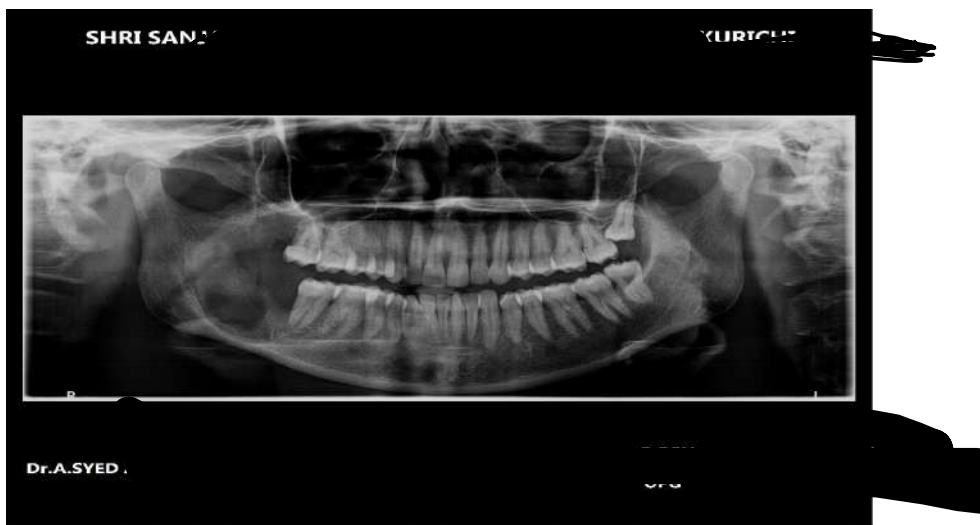


Figure 4: one month postoperative OPG



Figure-5: Three-month postoperative OPG

Figure 3 and 4 depict the clinical scenario and radiographic appearance 1 month after marsupialization. 3 months postoperative OPG (figure-5) reveals new bone formation in the mandibular body, angle and ramus region and the patient presented with no neurosensory deficit. Following this, enucleation of the cyst was done under LA and the patient was followed up periodically.

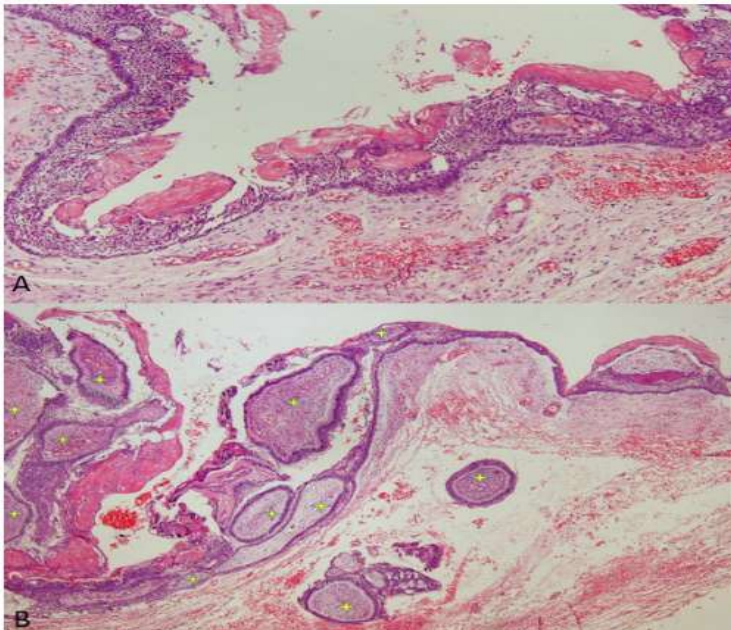
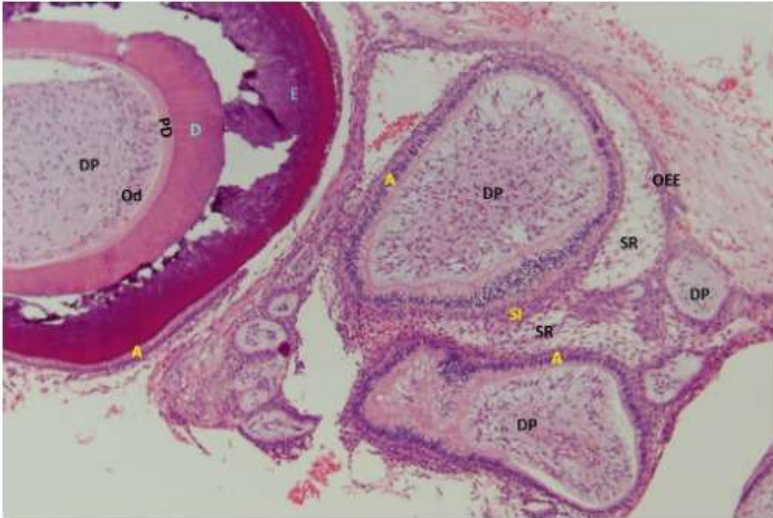


Figure 6 - Histopathologic feature of the epithelial lining of COC with prominent ghost cells ($\times 40$) (A). Sections of tooth buds in the epithelium and also in the cyst wall ($\times 100$)



(B). Sections of numerous tooth buds in different stages. Dental papilla (DP); Stellate reticulum (SR); Stratum Intermedium (SI); Outer enamel epithelium (OEE); Ameloblasts (A); Odontoblasts (Od); Enamel (E); Tubular dentin (D) ($\times 200$)

Discussion

Although CCOT has a cystic form, its epithelial cells may display hallmarks of ameloblastoma and other locally invasive neoplasms. Because of the heterogeneity in its classification, it is unclear whether the present lesion can be considered as a single entity. As a result, its clinical and histological features, as well as its interaction with other neoplasms, are considered in order to define it more clearly.[1,2,7,8]

The standard protocol which is also the currently accepted protocol for treating CCOT involves surgical excision of the lesion as a single step surgery. In our case, owing to the anatomy of the mandible and proximity or involvement of adjacent structures, we chose a 2-step procedure of 'Marsupialization – Enucleation'. This is similar to the principle of surgical management of unicystic ameloblastoma.[1,2,8,9] We performed the first step of Marsupialization through an intra-oral approach. Marsupialization is advantageous as it retards the growth of the lesion and promotes its regression by reducing the intra-cystic pressure. there are no reported contraindications for enucleation. One of the common complaints received from patients is foul odour from the region even after following excellent dental hygiene and irrigating the open cavity with saline solution.[2,11-13] This can be managed by increasing the frequency of oral hygiene practices. Following marsupialization, the lesion may take years to regress. A thorough follow-up after decompression is required for monitoring the regression rate and to determine when to perform the 2nd stage enucleation procedure. Thorough histopathological examination of the entire specimen is a must to rule out any accompanying neoplasm.[2]

In our case, Gorlin's cyst is found in the posterior part of the mandible which is quite rare as the lesion is more commonly to occur in mandibular body anterior to the first molar. One month postoperative intraoral photograph (figure 3) shows marked soft tissue healing and three months postoperative OPG (figure-5) shows

marked new bone formation and also the patient presented with no neurosensory deficit.

Conclusion

CCOT sometimes has been found with the presence of associated neoplasms in the histological examination. The possibility of associated neoplasms gives rise to several diagnostic controversies. It is also the cause for different schools of thought regarding therapeutic approaches for it.

Surgical marsupialisation of a large-sized and non-solid CCOT can be an effective therapeutic option to allow regression of the tumor which can be followed by enucleation as a second stage surgery under local anesthesia. CCOT has the possibility of long term recurrence. Hence, a long-term clinical and radiological follow-up is necessary to catch it early and avoid extensive future surgeries.

Conflict of Interest: None.

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