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Calcifying Cystic Odontogenic Tumor: A Diagnostic Challenge

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Authors' contributions

This work was carried out in collaboration between all authors. Author AKN designed the report and performed case analysis. Author AD wrote the manuscript, did patient management and literature research. Author SA managed the case. Author AA did histopathological reporting. Author AK performed case analysis. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

The calcifying odontogenic cyst was first described by Gorlin et al in 1962 and has been referred to as "Gorlin's cyst" or "Calcifying ghost cell odontogenic cyst". The lesion has cystic as well as tumor-like elements and was termed "calcifying cystic odontogenic tumor" (CCOT) by the World Health Organization (WHO) in 2005. The CCOT is benign; more commonly affects the anterior aspect of jaws and is without gender dominance. Knowledge of its clinical, radiographic, and pathological features is necessary to arrive at a proper diagnosis and management because it resembles other forms of pathology. Diagnosis relies mainly on radiographic features and the histologic presence of ghost cells within the epithelium. We report here a classic case of a CCOT in a 28 year old female.

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Keywords: Ghost cells; odontogenic cyst; odontogenic tumor; advanced imaging; calcification masses.

1. INTRODUCTION

Odontogenic tumors comprise a range of pathologic disorders that include malignant and benign neoplasms. The majority of odontogenic tumors are benign; however, some, such as the ameloblastoma, demonstrate aggressive and locally infiltrative behaviour [1]. Ameloblastoma (from the early English word *amel*, meaning enamel + the Greek word *blastos*, meaning germ) [2] is a benign tumor of odontogenic epithelium (ameloblasts) origin much more commonly appearing in the lower jaw than the upper jaw [3]. “Ghost cells”, described, by Gorlin, are enlarged eosinophillic cells without nuclei within the epithelium of the CCOT and believed to be an oral representation of the dermal calcifying epithelioma of Malherbe. It was classified as SNOMED code 930/0 and renamed in 2005 as “The calcifying cystic odontogenic tumor” in the WHO publication, Histological Typing of Odontogenic Tumors [4,5]. The CCOT represents less than 2% of all the odontogenic cysts and tumors making the CCOT an uncommon entity [5].

From its original determination it has always been believed to be a benign lesion and containing an ameloblastic epithelium lining with a varied amount of ghost cells that may calcify gradually over time and give a characteristic appearance of a mixed radiolucent-radiopaque lesion on radiographs [6]. Males and females are equally affected with no race predeliction. It has an intraosseous and extraosseous form with the intraosseous form being more frequent representing about 70-80% of the cases reported [7].

The CCOT usually presents as an asymptomatic, slow growing swelling, that may involve either the maxilla or mandible and most commonly found in the anterior region of the jaws (incisor/canine region) appearing during third and fourth decades of life [7]. Radio-graphically, it presents as a well circumscribed radiolucency containing multiple radio-dense foci suggesting calcification and associated with an impacted tooth or odontoma [8].

2. CASE REPORT

Our patient was a 28-year-old female who presented to the department of oral medicine & radiology with a chief-complaint of a painless swelling and missing teeth in the left anterior maxilla (Fig. 1). Her past medical history was non-contributory and her past dental treatment included an uneventful extraction of one tooth three years ago. She was aware of the swelling but did not give a history of associated symptoms such as pain, bleeding, or a purulent discharge from the affected area.

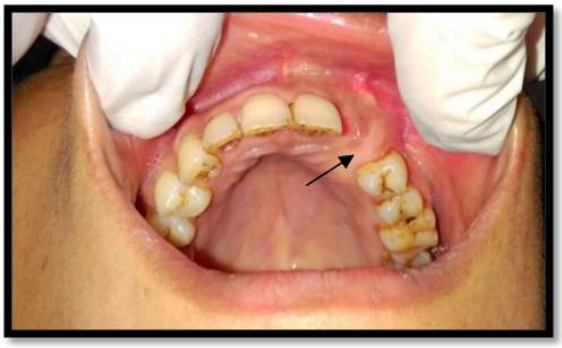

Fig. 1. Intra-oral view of edentulous area and swelling

On physical examination a diffuse asymptomatic swelling was noted in the left upper labial vestibule extending to the region of 22 and 23. The mucosa over the swelling appeared pale in color, with slight areas of erythema in the depths of alveolar mucosa with respect to areas 22 and 23. The swelling appeared to be diffuse, firm, and non-tender to palpation and with no signs of bleeding, compressibility, or pulsatality. A provisional diagnosis of an adenomatoid odontogenic tumor or residual cyst was made because of the missing tooth. Also, based on all the presenting clinical features our differential diagnosis included: dentigerous cyst, calcifying cystic odontogenic tumor, calcifying epithelial odontogenic tumor and complex odontoma. Radiographs were made to include: periapical views, an orthopantomogram (OPG), and computerized tomography (CT) imaging.

2.1 Radiographic Analysis

Radiographic imaging of the anterior left maxilla revealed a well defined radiolucent lesion approximately 2 cm x 3 cm in diameter surrounded by a radiopaque border in the regions of 22 and 23. Multiple irregular radio-opaque masses were present within the

circumscribed lesion at the periapical region of 24. OPG revealed a well-circumscribed radiolucency of approximately 2x3 cm in diameter containing multiple diffuse radio-opaque areas of calcifications in the form of clusters adjacent to the radiolucent lesion (Fig. 2). An impacted tooth #23 was seen above the osteolytic lesion. Coronal, sagittal and axial sections of CT images revealed a hypodense mass with antero-posterior and latero-medial expansion of the cortices containing irregular shaped radio-dense masses arranged in clusters in the anterior region of the maxilla (Fig. 3). Panoramic and CT images revealed a unilocular radiolucency with thick corticated margins at the lateral aspect of 25 and 26 suspected to be a lateral periodontal cyst.

2.2 Treatment and Follow Up

Patient was treated under general anesthesia in a hospital setup. Mucoperiosteal flap was raised between 22 and 25 and the friable cystic lesion was completely enucleated along with complete removal of radio-opaque masses along with the impacted tooth and lateral periodontal cyst. The specimen was sent for histo-pathological examination. Patient attended regular follow-ups and after 6 months no recurrence was seen with the lesion.

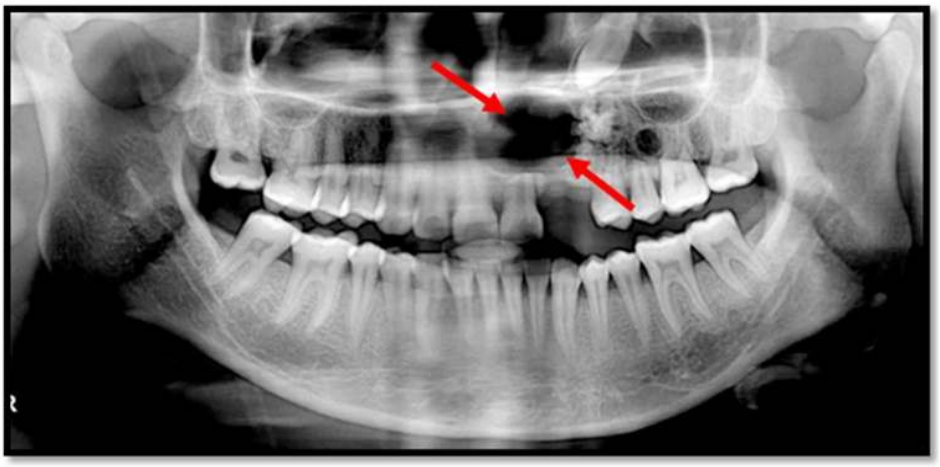

Fig. 2. OPG image showing well circumscribed radio-lucency and radio-opaque clusters

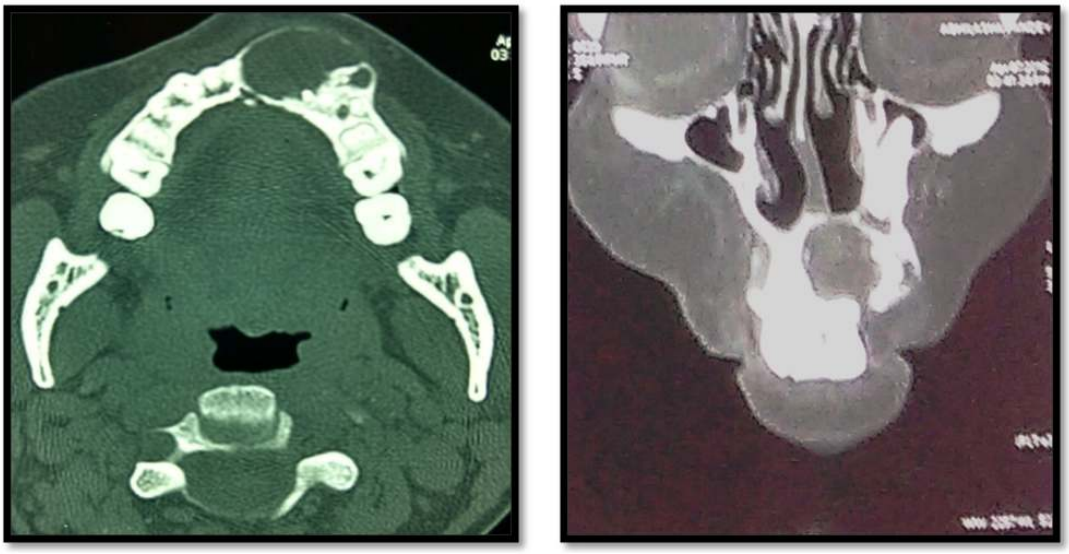

Fig. 3. CT scans showing a hypodense mass

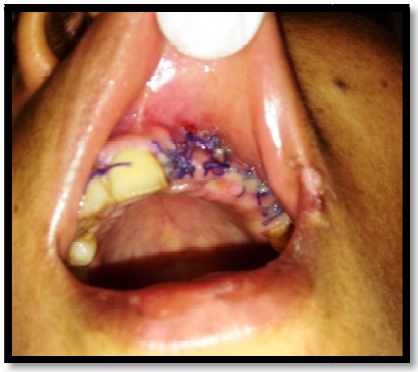

Fig. 4. Post-operative intraoral picture 1 week after the surgery

Microscopic examination revealed a cystic space lined by odontogenic epithelium consisting of basal cuboidal to columnar cells and superficial stellate reticulum like cells. Ghost cells and concentric calcification masses within the epithelium and in the lumen were also noted. The cystic wall is fibrocellular with few blood vessel, numerous odontogenic rests and cortical bone evident (Fig. 5).

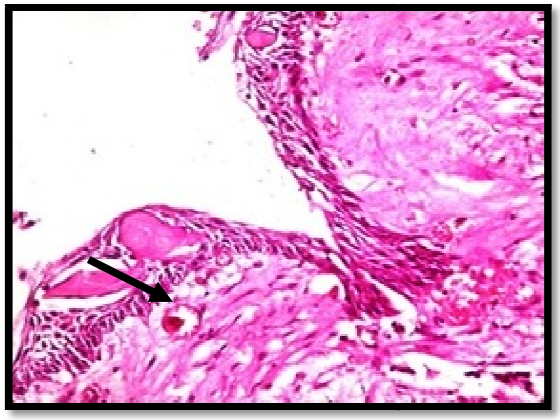

Fig. 5. Photomicrograph showing presence of ghost cells and calcification masses within the epithelium (H and E, 40x)

3. DISCUSSION

Few authors have stated that the initial lesions of CCOT are solid tumors and cyst formation is just a secondary phenomenon [9]. A definitive diagnosis of calcifying odontogenic cyst can be reliably made on the basis of a histological examination. The presence of ghost cells within the lesion is a characteristic histologic feature of this lesion. During the cascade of its development ghost cells undergo dysplastic calcification with accumulation of osteoid or dentinoid like material as an inductive effect on the epithelium and hence appears as flecks of radiopaque masses within the radiolucent cavity, suggesting a mature state of the lesion [9]. With regard to the very small number of recurrences, only eight cases of recurrences have been documented in English literature [10].

Since it was first described, the CCOT has been a topic of discussion regarding the true nature of the condition mainly because of its dual structure consisting of neoplastic and cystic elements. Few authors take sides with a dualistic approach; either cyst or a neoplasm and some prefer to have a monistic approach; that CCOT is a tumor with a marked tendency toward cystic architecture [11] (cited properly).

On radiographs, the lesion appears to be either unilocular or multilocular and as a well-defined radiolucency that may contain small irregular calcified bodies of varying sizes. These scattered irregular-sized calcifications produce a variable range of opacities and are referred to as “salt & pepper type of patterns”. In 50% of the cases the lesions are associated with tooth like densities (odontoma-like formations) and one third of the cases are associated with an un-erupted tooth, most often the canine [12]. An abnormal form of keratinization in the form of the ghost cells is the most distinguishing feature of a CCOT. However, there presence does not confirm the diagnosis as other lesions show similar presentation. Hence, a diagnosis of CCOT should only be made for a lesion in which the formation of ghost cells takes place in a typical epithelial cyst lining, presenting a basal layer of cuboidal or short cylindrical cells and an overlying layer consisting of cells that bear resemblance to stellate reticulum-like cells [13]. As the lesion more often arises in the tooth bearing areas of the jaws or gingiva, CCOTs are commonly arise associated with a tooth, as seen in this case [14].

Our present case classically revealed a radiolucent unilocular lesion with scattered radiopaque masses of calcification in the upper anterior region of the maxilla and was associated with a cystic cavity and an impacted tooth. The CCOT was once considered to be a calcifying epithelioma of Malherbe due to the similarities present between these two entities. They have a similar site of occurrence that primarily involves the head and neck region and both demonstrate foci of calcification. CCOT is differentiated from

its counterpart in its origin from cells of odontogenic epithelium while calcifying epithelioma of Malherbe is mainly found in skin and is a benign neoplasm arising from hair molecule hair matrix cells [15].

4. CONCLUSION

CCOT is a rare condition with diverse clinical, histological and radiographic forms. Since its inception it has been a diagnostic challenge and has given rise to various terminologies and classifications to determine the true nature of origin. Its diagnosis can be confusing due to other conditions with resembling clinical features. Advanced imaging techniques such as the CT scan along with more traditional radiographic techniques, biopsy and careful microscopic examination will lead to accurate diagnosis and treatment of a CCOT.

CONSENT AND ETHICAL APPROVAL

All authors here by declare that an informed consent was taken by the patient as well as his guardians to report this case for scientific purposes prior to publication only.

All the ethical guidelines were carefully followed.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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