

CASE REPORT

Calcifying Cystic Odontogenic Tumor with Ameloblastoma: A Case Report with Review of the Literature

¹Deepa Jose, ²Karishma Desai, ³Deepa R Mane, ⁴Alka Kale

ABSTRACT

Calcifying cystic odontogenic tumor (CCOT) is a rare odontogenic entity that exhibits a diverse array of clinical, radiographic, and histopathologic features. It accounts for less than 2% of all odontogenic tumors. Praetorius et al have classified CCOT associated with other odontogenic tumors as type II variant. So far, about 31 cases of this rare variant have been reported in the literature. In this report, a rare case of CCOT with ameloblastoma in a 38-year-old male, involving the left mandibular molar region with its associated radiological and clinical diagnosis, is presented. All clinical, radiographic, and histological features of the case were analyzed in comparison to those reported in the literature. Due to the heterogeneous presentation, there has always been confusion about the nature of CCOT as a cyst, neoplasm, or hamartoma. The occurrence of an ameloblastoma with CCOT could suggest a possibility of it being a neoplasm.

Keywords: Cyst, Odontogenic, Tumor.

How to cite this article: Jose D, Desai K, Mane DR, Kale A. Calcifying Cystic Odontogenic Tumor with Ameloblastoma: A Case Report with Review of the Literature. *Int J Prev Clin Dent Res* 2016;3(1):38-41.

Source of support: Nil

Conflict of interest: None

INTRODUCTION

Cysts of the jaws are broadly classified as odontogenic or nonodontogenic. Calcifying cystic odontogenic tumor represents 1 such rare (2%) heterogeneous group of odontogenic developmental lesions.¹ It was first described by Gorlin in 1962 and hence the eponym Gorlin cyst.¹ Fejerskov and Krogh (1972) used the term "calcifying ghost cell odontogenic tumor" and Freedman et al (1975) referred to it as "cystic calcifying odontogenic tumor (CCOT)."²

Ever since the description of calcifying odontogenic cyst (COC) as a specific odontogenic lesion in 1962,

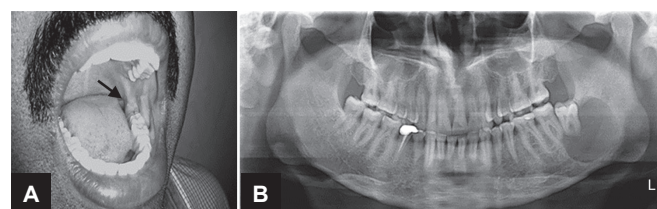
controversies existed regarding the relationship between non-neoplastic cystic lesions and solid tumor masses. The so-called COC shows great diversity in the spectrum of clinical behavior and histopathological features, including cystic, solid (neoplastic), and aggressive (malignant) variants, because of which different categorization and nomenclature for the lesion have been proposed. In the 2005 WHO classification, these tumors were reclassified into 3 categories: calcifying cystic odontogenic tumor, a benign cystic neoplasm; dentinogenic ghost cell tumor (DGCT); and ghost cell odontogenic carcinoma (GCOC), which is the malignant variant of CCOT or DGCT.³

In the present case report, we emphasize the rarity of the lesion and highlight various histopathological features of current CCOT types from other subtypes.

CASE REPORT

A 38-year-old male patient was reported to KLE Dental Hospital with a complaint of pain in the lower left posterior region of the jaw since two and a half months. This was accompanied with pus discharge from the 38 region, for which he visited a local dentist, and symptoms subsided after taking medication. The patient also had a history of diabetes since 2 years.

On extraoral examination no visible swelling was noted. Intraorally, a nondiscernable swelling in relation to partially impacted 38 was seen (Fig. 1A). Orthopantomograph (OPG) showed a well-circumscribed radiolucency surrounding the root of 38, extending till the lower border of the mandible superior-inferiorly, and anteroposteriorly from the distal root of 37 to 2 cm posteriorly into the mandibular ramus area (Fig. 1B). It also showed flecks of calcification near the apex of the distal root of 38. Based on clinical and radiographic



Figs 1A and B: (A) Photomicrograph of the intraoral examination of lesion, and (B) Photomicrograph showing orthopantomograph, which revealed radiolucency in relation to 38

^{1,2}Postgraduate Student, ³Reader, ⁴Professor and Dean

¹⁻⁴Department of Oral Pathology and Microbiology, KLE VK Institute of Dental Sciences, KLE University, Belgaum Karnataka, India

Corresponding Author: Karishma Desai, Postgraduate Student, Department of Oral Pathology and Microbiology, KLE VK Institute of Dental Sciences, KLE University, Nehru Nagar Belgaum, Karnataka, India, Phone: +919920833545, e-mail: drdesaikarishma@gmail.com

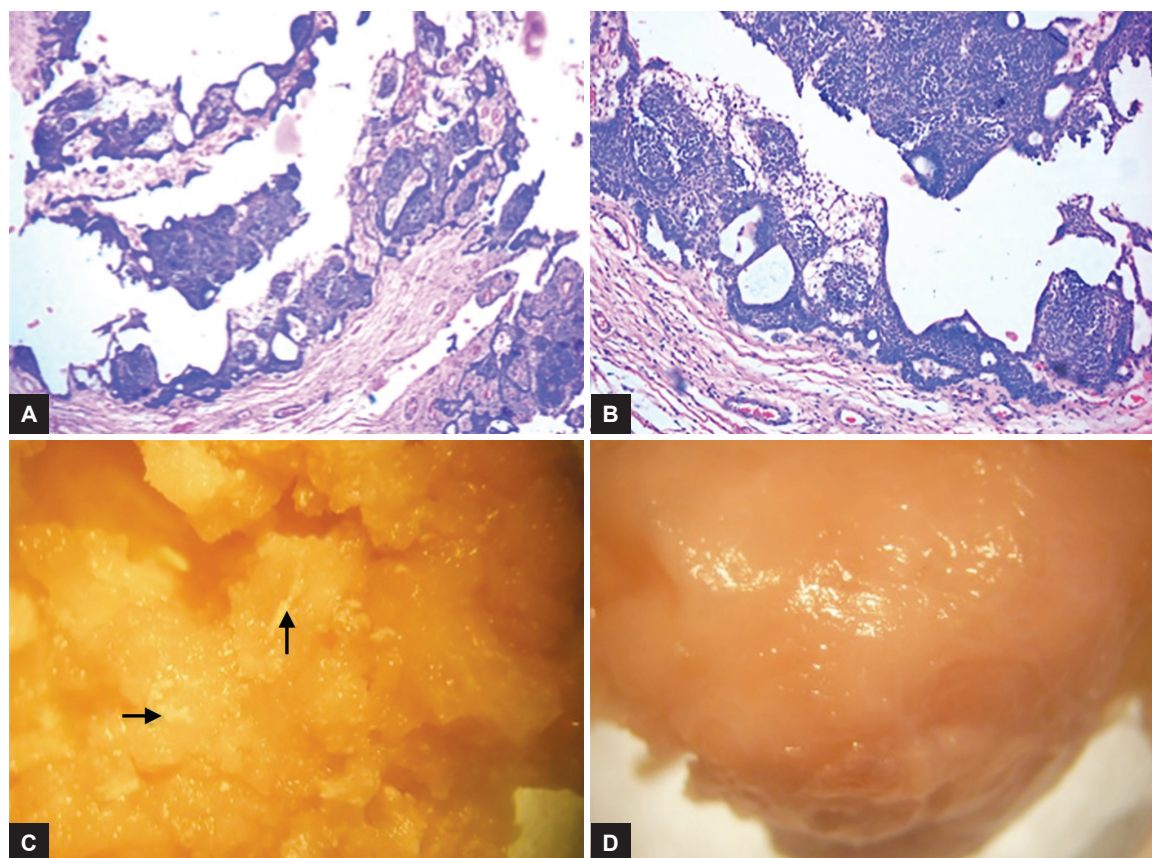
findings, a differential diagnosis of the dentigerous cyst, unicystic ameloblastoma, odontogenic keratocyst, and calcifying epithelial odontogenic tumor was made. Incisional biopsy was advised and specimen submitted for histopathological examination.

Microscopically, hematoxylin and eosin-stained section showed a parallel arrangement of connective tissue capsule with odontogenic epithelial lining enclosing a large cystic cavity. The epithelial lining shows both intraluminal and intramural proliferations. Under higher magnification, epithelial lining comprised hyperchromatic the basal cell layer and the superficial layer of stellate reticulum-like cells. Some areas of follicular ameloblastoma-like cell proliferations were seen. Pale homogenous eosinophilic areas showing cellular outlines with lack of nuclear and cytoplasmic details suggestive of ghost cells were seen. The connective tissue capsule was made up of loosely arranged collagen fibers (Figs 2A and B). Based on these features, a diagnosis of the calcifying epithelium odontogenic cyst with ameloblastomatous proliferations was given.

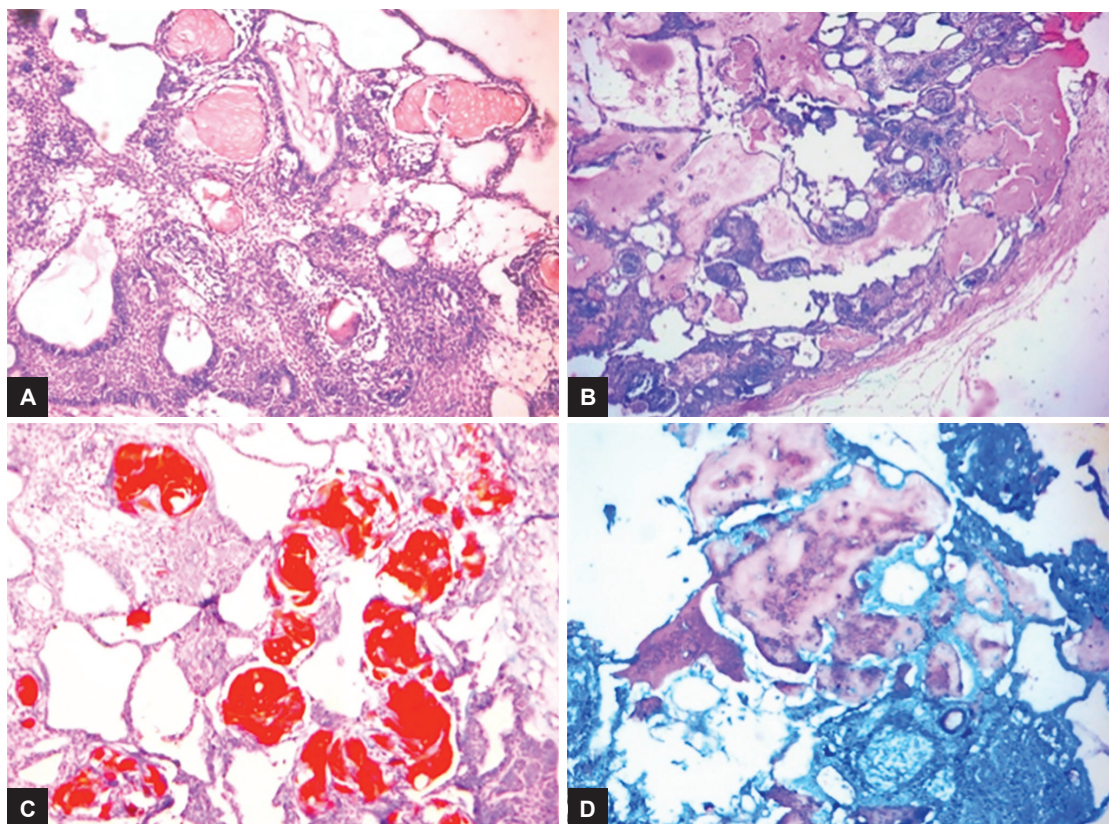
In accordance with this, an excisional biopsy was performed along with extraction of 37 and 38 and was sent for histopathological examination. On evaluation of the gross specimen under a stereomicroscope, the external

surface of the capsule with smooth and regular contours was appreciated (Figs 2C and D). The cut section of the specimen showed irregular proliferations of soft tissues interspersed with a few white specks.

Histopathologically, an odontogenic epithelium showing mural proliferations along with a connective tissue capsule as seen in incisional biopsy was noted. Along with this some cells showed predominantly plexiform ameloblastoma-like arrangement. A few follicular arrangement patterns could also be noted. Numerous ghost cells were seen interspersed between the proliferating epithelium. Surrounding this, stellate reticulum-like cells were noted and a prominent hyperchromatic basal cell layer was seen. Juxtaepithelially, areas of connective tissue showed evidence of a dysplastic dentin/dentinoid-like structure (Figs 3A and B). For confirmation of these findings, special histochemical staining techniques using the Ayoub-Shklar stain, methylene blue acid fuchsin, and congo red stain were performed. Ayoub-Shklar stained the aberrant keratin of the ghost cells in the epithelium brick red, while methylene blue acid fuchsin stained the calcified dentinoid material pink in blue background (Figs 3C and D). Congo red was negative, ruling out the presence of any amyloid-like substance. Based on these contributory findings, a



Figs 2A to D: Photomicrograph showing microscopic picture of (A and B) incisional biopsy (hematoxylin and eosin 4×, 10×); (C and D) stereomicroscopic image, (C) cut surface revealing irregular proliferations of soft tissue with few white specks, (D) smooth external surface of capsule with regular contours



Figs 3A to D: Photomicrograph showing microscopic picture of (A and B) excisional biopsy (hematoxylin and eosin 10 \times , 10 \times), (C) Ayoub-Shklar special stain for ghost cells (10 \times), and (D) methylene blue acid fuchsin for dentinoid areas (10 \times)

final diagnosis of calcifying odontogenic cyst – type ID (calcifying odontogenic cyst with unicystic, plexiform ameloblastomatous proliferation of epithelium) as per Riechart's classification was given.

DISCUSSION

Calcifying cystic odontogenic tumor is an uncommon benign lesion, which forms around 2% of the odontogenic tumor since its discovery by Gorlin et al in 1962. Although named and defined as a cyst, there is no agreement in the literature regarding its classification as a cyst or a neoplasm, since some examples of CCOT show areas suggestive of neoplasia. The CCOTs that occur in relation to odontogenic tumors like odontomas, ameloblastoma, ameloblastic fibroma, etc. behave more aggressively and warrant a radical or extensive surgical intervention.⁴ Buchner (1991) suggested that if CCOT was associated with ameloblastoma, its behavior and prognosis would be that of an ameloblastoma, not CCOT.⁵

Literature survey suggests that so far only about 31 cases of ameloblastomatous CCOT have been reported. It is hard to compare the details with findings from previous reports as this is a rare lesion and different authors used different histologic criteria to categorize it. Earlier reports reveal that these lesions present in the 3rd to 4th

decade of life without any gender predilection.⁶ The age of occurrence in the 3rd decade was correlating with our present case; however, a bimodal age distribution was recorded by Praetorius et al.⁷

Calcifying cystic odontogenic tumor presents as a painless slow-growing lesion showing an anterior predilection with an equal distribution in the maxilla and mandible. The present case also reported with a painless swelling, which was associated with an impacted 3rd molar tooth. According to Hoffman et al,⁸ central lesion constitutes 78.5% cases, and 21.5% are observed as peripheral lesion. The development of CCOT is dependent on remnants of reduced enamel epithelium, odontogenic epithelium, and basal cells of oral mucosal epithelium, which constitutes the source of the lesion.⁷

Radiographically, CCOT is a mixed lesion, with unilocular or multilocular radiolucent area, and varying amount of radiopaque material.⁹ Our case also presented as a unilocular radiolucent lesion with some radioopaque flecks. McGowan and Browne⁹ found that the presence of mineralization is apparently more frequent in histologic examination compared to radiographic analysis. Our case seems to support this conclusion, as it had very low detectable calcified bodies on radiographic evaluation.

Histological findings support the features of CCOT documented with that of the literature.⁶ Microscopic



Fig. 4: Postoperative orthopantomograph with bone formation in the area of surgical excision

examination showed cystic epithelium having luminal and mural proliferation consisting of cuboidal cells with hyperchromatic nuclei and subnuclear vacuolization. Clusters and sheets of pale eosinophilic cells with faint nuclear outline suggestive of ghost cells were also evident. Sheets of eosinophilic area with entrapped cells suggestive of dentinoid areas were also seen. To categorize CCOT, Riechart's histological classification was adopted.¹⁰ Various special stains, such as Ayoub-Shklar, which shows reaction for keratin, demonstrated positive reaction for ghost cells; methylene blue acid fuchsin was done to differentiate the calcification, and dense stroma revealed a strong positive stain for calcified areas. The present case radiographically mimics dentigerous cyst in respect to its typical location and association with impacted 3rd molar; however, histologically it presents with a picture of ameloblastomatous CCOT. Although treatment of CCOT is conservative, surgical enucleation and its recurrence is rare, but a long-term follow-up is considered mandatory. In our case, based on the clinical, radiographic, and histological features, treatment opted was surgical enucleation. The patient is on regular follow-up and after a period of 1 year, we observed normal healing and with no signs of recurrence (Fig. 4).

CONCLUSION

Calcifying odontogenic cyst with ameloblastoma is a rare entity and has a wide array of histomorphologic presentation, and is hence still a diagnostic dilemma.

However, its association with ameloblastoma suggests it to be a neoplasm. A comprehensive analysis of clinical, radiographic, and histopathological features is needed to delineate an appropriate diagnosis and treatment plan of these lesions. The current case radiographically simulated a quiescent image of dentigerous cyst; however, histologically it was characterized as an ameloblastomatous COC. These lesions, although associated with ameloblastoma, are usually treated by enucleation and the recurrence is dependent on the complete enucleation.

REFERENCES

1. Ramaraju D, Ramlal P, Sushma A, Harisha S, Deepthi K, Reddy J. Ameloblastomatous Gorlin's cyst – a rare case report. *J Res Adv Dent* 2013;2(2):42-46.
2. Vasudev S, Singh VP, Jhawar S, Roy K. Calcifying cystic odontogenic tumor – a unique developmental lesion arising in mandible. *Health Renaissance* 2012;10(1):64-68.
3. Jing W, Xuan M, Lin Y, Wu L, Liu L, Zheng X, Tang W, Qiao J, Tian W. Odontogenic tumours: a retrospective study of 1642 cases in a Chinese population. *Int J Oral Maxillofac Surg* 2007 Jan;36(1):20-25.
4. Toida M. So-called calcifying odontogenic cyst: review and discussion on the terminology and classification. *J Oral Pathol Med* 1998 Feb;27(2):49-52.
5. Buchner A. The central (intraosseous) calcifying odontogenic cyst: an analysis of 215 cases. *J Oral Maxillofac Surg* 1991 Apr;49(4):330-339.
6. Yoshida M, Kumamoto H, Ooya K, Mayanagi H. Histopathological and immunohistochemical analysis of calcifying odontogenic cysts. *J Oral Pathol Med* 2001 Nov;30(10):582-588.
7. Praetorius, F.; Ledesma-Montes, C. Calcifying cystic odontogenic tumor. Barnes, EL.; Everson, JW.; Reichart, P.; Sidransky, D., editors. *Pathology and genetics of head and neck tumours. World Health Organization Classification of Tumors*. Lyon (France): IARC Press; 2005. p. 313.
8. Hoffman, S.; Jacoway, JR.; Krolls, SO. Intraosseous and parosteal tumors of the jaws. *Atlas of tumor pathology*. Washington (DC): Armed Forces Institute of Pathology; 1987. p. 30-34.
9. McGowan RH, Browne RM. The calcifying odontogenic cyst: a problem of preoperative diagnosis. *Br J Oral Surg* 1982 Sep;20(3):203-212.
10. Reichart, PA.; Philipsen, HP. *Odontogenic tumors and allied lesions*. 1st ed. London: Quintessence Publishing Co. Ltd; 2004.