

Calcifying Cystic Odontogenic Tumor with Compound Odontoma

Sunil Sidana, Kartik Poonja, Jyotsna Galinde, LS Poonja

ABSTRACT

Calcifying cystic odontogenic tumor (CCOT) previously known as calcifying odontogenic cyst (COC) is a relatively rare lesion. It was first reported as a separate pathologic entity by Gorlin et al in 1962 as a likely analog of the cutaneous calcifying epithelioma of Malherbe or pilomatricoma. Because of the diverse clinicohistologic features and the various neoplastic potential, there have been disagreements on the terminology as well as whether to classify CCOTs as a cyst or a neoplasm. CCOT is frequently associated with other lesions, such as odontoma, ameloblastoma and ameloblastic fibroma, and the most common of these is the CCOT-associated odontoma (CCOTaO). CCOTaO tends to occur in the anterior portion of upper jaw and occurs in younger age group as compared to other types. We present a case of CCOTaO in the posterior mandible of a 23-year-old male.

Keywords: Calcifying cystic odontogenic tumor, Calcifying odontogenic cyst, Calcifying cystic odontogenic tumor-associated odontoma.

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INTRODUCTION

Calcifying cystic odontogenic tumor (CCOT) previously known as the calcifying odontogenic cyst (COC) was first reported as a separate pathologic entity by Gorlin et al in 1962.¹ It is a rare lesion and accounts for only 2% of all odontogenic tumors.^{2,3}

Owing to its variable histology, clinical behavior and association with dysregulated beta-catenin signaling⁴ confusion persists to this day, including whether or not it is a reactive or developmental cyst or a neoplasm.⁵ Recently, when the World Health Organization (WHO) updated its classification of odontogenic tumors, the name of the COC was changed to CCOT to emphasize the neoplastic nature of a lesion previously categorized as an odontogenic cyst.⁶ The CCOT often occurs in association with or contains areas histologically similar to a variety of odontogenic tumors, such as complex and compound odontoma, ameloblastoma, ameloblastic fibroma, ameloblastic fibro-odontoma, odontoameloblastoma, CEOT and AOT.⁷ Among these, the odontoma is most commonly found in association with

CCOT (CCOTaO). Their association has been reported as 20 to 24%.⁷ CCOTaO tends to occur in the anterior portion of upper jaw and occurs in younger age group as compared to other types. We present a case of CCOTaO in the posterior mandible of a 23-year-old male.

CASE REPORT

A 23-year-old male presented to our department with a chief complaint of painless swelling in the left mandibular posterior region of 1 month's duration. The swelling gradually increased to its present size. On examination, extraorally there was asymmetry of the left lower face. Intraoral palpation revealed a nontender buccal bony hard swelling (Fig. 1). There was no lower lip numbness.

The orthopantomogram revealed a well-defined radiolucent lesion extending from the left mandibular second premolar to the ramus of the mandible anteroposteriorly. Superioinferiorly, it extended from crestal bone to the inferior border of the mandible. Resorption of distal root of left mandibular first molar and both roots of left mandibular second molar were seen. Left mandibular third molar was impacted and along with inferior alveolar neurovascular bundle was seen displaced to the inferior border of the mandible. Multiple small radiopacities were seen distal to second molar (Fig. 2). The computed tomograph showed expansion of buccal and lingual cortical plate. No discontinuity of bone could be seen along the inferior border or lingually (Fig. 3).

An incisional biopsy was taken. The histopathological findings revealed an epithelial lining with a basal layer



Fig. 1: Intraoral photograph of patient with buccal expansion in the left posterior molar region

consisting of columnar cells with hyperchromatic polarized nuclei. The epithelium was of variable thickness. Above the basal layers were seen two to three layers of stellate reticulum like cells. Masses of ghost cells were seen in the epithelial lining and in the fibrous capsule (Fig. 4). Variable amounts of dentinoid and calcified material were also evident. Histological diagnosis of CCOT was made. Under general anesthesia, through intraoral approach, the lesion was enucleated (Figs 5 and 6). Lower left second premolar, first and second molars and impacted third molar were extracted. Hemostasis was achieved. The inferior alveolar neurovascular bundle was identified lying on the lower border of mandible and saved. Closure was done using 3-0 vicryl. The specimen was sent for histopathology.

Histopathologically, the lining was again confirmed to be of CCOT. The multiple radiopacities which were seen on the radiograph were histologically found to consist of

multiple denticles with enamel, dentin and pulp arranged in a tooth-like pattern (Fig. 7). A final diagnosis of calcifying cystic odontogenic tumor with compound odontoma was made. Postoperatively, there was dehiscence of the suture line. An acrylic plug was given to keep the opening patent and regular irrigation and follow-up was done. At 1 year follow-up, there is good bony healing with no evidence of recurrence (Fig. 8).

DISCUSSION

COC was first described by Gorlin et al as COC in 1962. WHO reclassified it as a tumor in 1992. It was renamed as CCOT in 2005. Due to the diverse clinicohistologic features and the various neoplastic potential, there have been disagreements on the terminology as well as whether to classify CCOTs as a cyst or a neoplasm. Adding to the confusion is the finding that the CCOT has been associated



Fig. 2: OPG shows a well-defined radiolucent lesion extending from the left second premolar to the ramus of the mandible. Root resorption of first and second molar is seen with third molar displaced to the lower border. Also seen are multiple small radiopaque masses in the ramus of the mandible

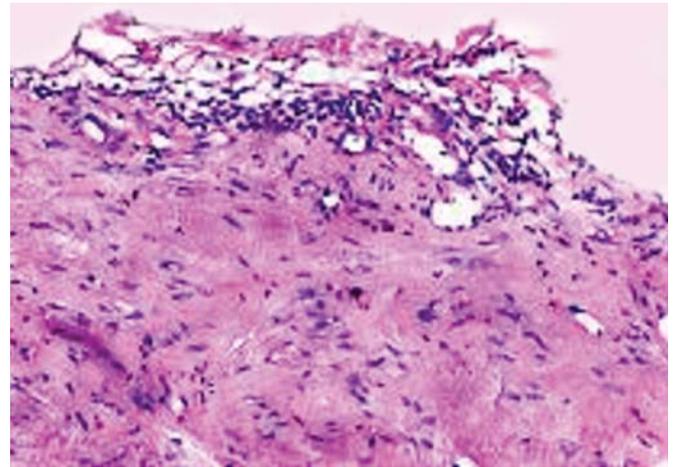


Fig. 4: Low power photomicrograph showing epithelial lining with columnar basal cells, stellate reticulum like cells and scattered ghost cells



Fig. 3: Axial CT scan showing bony expansion with multiple small radiopaque masses in the ramus of the mandible



Fig. 5: Part of the enucleated lining and extracted tooth

with dysregulated Wnt/-catenin signaling.⁴ Wnt is a glycoprotein ligand for frizzled, a receptor that, when activated, promotes translocation of catenin to the nucleus where it activates the transcription factor T-cell factor/lymphoid enhancer factor and a number of other Wnt target genes that favor cell-cycle progression. The constitutive activation of catenin/TCF-mediated transcription is known



Fig. 6: The enucleated denticles

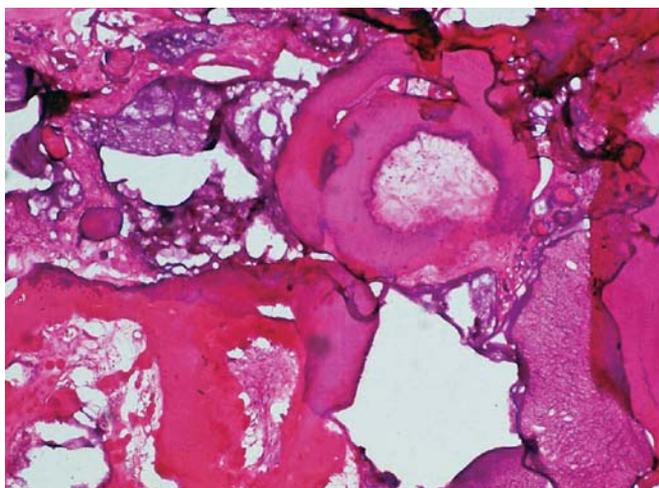


Fig. 7: High power photomicrograph showing formation of enamel matrix around two denticles containing dentin and pulp. Ghost cells seen in the vicinity



Fig. 8: Postoperative OPG at 1 year follow-up showing good bony healing

to play a role in human oncogenesis and may be important in the development of CCOTs as well. It is therefore possible that the CCOT is a 'cystic neoplasm,' a belief reflected in the change of the name to CCOT and a new classification from odontogenic cyst to odontogenic tumor by the WHO.⁶

The evidence suggesting neoplastic nature of the CCOT is as follows:

1. The CCOT is not always cystic and approximately 13% (Praetorius et al)⁸ and 17% (Mcgowan and Browne) of CCOTs have been reported as solid lesions. In some cases, microcyst formation is also found as seen in cases of ameloblastoma. Therefore, Fejerskov and Krogh have interpreted the lesion as a tumor or hamartoma with a marked tendency for cystic degeneration.
2. The CCOT shows histologic features similar to other odontogenic tumors. In some cases, the epithelium shows resemblance to ameloblastoma.
3. The biologic behavior of the CCOT is similar to odontogenic tumors. For instance, it shows root resorption in 13 to 50% of CCOTs. Marked resorption is also seen in some cases. These features are seldom found in a cyst. Erosion of bony cortices and a moderate amount of bone destruction are also occasionally seen in CCOT. Taking the non-neoplastic type of the lesion in mind many names have been proposed, calcifying ghost cell odontogenic tumor, cystic calcifying odontogenic tumor and dentinogenic ghost cell tumor although the last name is restricted to the noncystic type.

Moreover, CCOT is frequently associated with other lesions, such as odontoma, ameloblastoma and ameloblastic fibroma, and the most common of these is the CCOTaO.^{7,9} Gold was the first to describe CCOT with compound odontoma in 1963. It is well known that odontomas are not infrequently associated with CCOT. Their association has been reported as 20 to 24%, whereas some have reported a higher frequency of association. A consensus still lacks on the classification of CCOTaO either as a separate type of CCOT, i.e. combined odontogenic lesion with some proliferative potential or as a subtype of nonproliferative simple unicystic type of CCOT.⁷ Hirshberg analyzed 52 cases of CCOTaO and proposed CCOTaO to be regarded as a separate entity and suggested the term 'odontocalcifying odontogenic cyst', due to the unique histologic features and its female predilection with the predominant distribution pattern to the maxilla.⁷

Several possibilities have been suggested for pathogenesis of CCOTaO:

1. Coincidental juxtaposition of CCOT and odontoma.
2. CCOT develops secondarily from the odontogenic epithelium that participates in the formation of odontoma.

3. Odontoma can develop secondarily from the odontogenic epithelium of CCOT since the epithelium has the potential for mesenchymal induction.

However, it is generally accepted that CCOTaO occurs in a significantly younger age group compared to other types.¹⁰ The frequency of 'enveloped' teeth is reported to approximate 32%, and a lesion involving an impacted tooth may simulate a dentigerous cyst when viewed on radiographs. Iida et al in 2004 reported three COCs associated with odontoma and all of these were also associated with impacted teeth.² Our case also showed an association with an impacted tooth. In previous cases, reported the CCOTaO has a tendency to occur in the anterior portion of the upper jaw. Our case becomes unique because of its occurrence in the posterior segment of the mandible.

Radiographically, CCOT generally appears as a unilocular lesion with a well-defined margin and contains calcifications. The frequency of the multilocular form has been noted as 5% and the presence of calcification, which is an important radiographic feature in the interpretation of CCOT, is detected in about half of all CCOTs. CT findings characteristic of CCOT include calcifications along the outline of the bony cavity and have been reported previously.⁸

The differences between CCOT and CCOTaO (CCOT associated with odontome) are as follows:

1. A female predominance (2:1) was noted in cases of COCaO compared with an even distribution in the simple CCOT.
2. The mean age discovery was 16 years compared with 34.3 years in simple CCOT.
3. More cases of CCOTaO are found in maxilla (61.5%) in anterior region (75%), while CCOT shows equal predilection for both jaws but also seen in the anterior region.
4. The differences in the radiographic features of the two entities is because of presence of the odontoma component in CCOTaO.

The 90% of CCOTaOs show mixed radiolucent-radioopaque lesion or a pure radioopaque mass. The simple CCOT presents as a radiolucent lesion with only minute radiopacities representing dentinoid and dystrophic calcifications.

5. Histologically, the difference between the two clinical entities is the presence of tooth-like structures as in case of CCOTaO.⁵

The COCaO should be treated conservatively by surgical enucleation because recurrences are very uncommon.¹⁰ The lack of recurrence is dependent on complete excision.

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ABOUT THE AUTHORS

Sunil Sidana (Corresponding Author)

Reader, Department of Oral and Maxillofacial Surgery, MGM Dental College and Hospital, Navi Mumbai, Maharashtra, India
e-mail: drsunilsidana@gmail.com

Kartik Poonja

Lecturer, Department of Oral and Maxillofacial Surgery, MGM Dental College and Hospital, Navi Mumbai, Maharashtra, India

Jyotsna Galinde

Professor and Head, Department of Oral and Maxillofacial Surgery MGM Dental College and Hospital, Navi Mumbai, Maharashtra, India

LS Poonja

Professor, Department of Oral Pathology, MGM Dental College and Hospital, Navi Mumbai, Maharashtra, India