

# Calcifying Cystic Odontogenic Tumor: Review with Discussion

Dr. Ritika Jindal BDS, Dr. Ravikiran Ongole MDS, Dr. Junaid Ahmed MDS, Dr. Cenna Denny< MDS  
Manipal University, India

## Abstract

**Aim:** We report a case of calcifying cystic odontogenic tumor affecting maxilla in a 48 year old male patient presented with painless extensive intraoral swelling with unilocular radiolucency and marked resorption of roots on radiographs.

**Summary:** Calcifying cystic odontogenic tumor (CCOT) is rare developmental odontogenic pathology. The first description of lesion was given in 1962. Various terminologies and classifications have been proposed for description of the lesion. CCOT has extraosseous and intraosseous variants. Clinically it usually presents as a slow growing painless swelling of maxilla or mandible. It commonly involves anterior region of jaws, shows no gender predilection. Radiographically the lesion has variable appearance. The most common presentation is a well defined unilocular radiolucency associated with irregular calcification. Presence of ghost cells with proliferative odontogenic epithelium is the characteristic feature of the lesion. Surgical enucleation is the treatment of choice.

*Index terms: biopsy, dentistry, radiography, tumor*

## INTRODUCTION

Calcifying cystic odontogenic tumor first described by Gorlin in 1962, is a rare lesion representing 2% of all odontogenic pathological changes in the jaw. WHO in 2005, designated it as a tumor and renamed it as "Calcifying cystic odontogenic tumor" (CCOT).

CCOT is believed to be developmental in origin, derived from odontogenic epithelial remnants. It can occur at any age, with maximum incidence in the 2<sup>nd</sup> decade of life. It can occur centrally or peripherally, usually presenting as a painless swelling of jaw, equally affecting maxilla and mandible and more commonly involving anterior region than posterior. Prognosis of the lesion is good with few cases of recurrence have been reported following surgical enucleation.

## CASE

A 48 year old male patient presented with a swelling on the left side of his face for the past 1 week. On extraoral examination patient had noticeable facial asymmetry with a diffuse, firm, mildly tender swelling on left middle third of the face. Intraorally, obliteration of buccal vestibule was seen in maxillary left posterior region and palatally the swelling was extending to the midline (Figure 1). On palpation, swelling was firm with slightly fluctuant area in the centre; there was expansion of cortical plates. Teeth in relation 23, 24, 25, 26 to swelling were mobile and gingival recession was present with respect to 23. Electric pulp testing revealed no response in relation to 22, 23 and delayed response in 24, 25, 26, 27. A provisional diagnosis of periapical cyst was made.

Panoramic radiograph revealed a unilocular radiolucency with well defined borders with respect to 23, 24, 25, 26, 27 and root resorption with respect to 24, 26 (Figure 2). Intraoral periapical radiographs with respect to 22, 23, 24, 25, 26 revealed unilocular radiolucency with well defined borders in relation to apices of these teeth and marked root resorption in relation to 23, 24, 26 (Figure 3, 4). Lateral maxillary occlusal radiograph revealed multilocular radiolucency on palatal and buccal aspect with respect to 23, 24, 25, 26 and expansion and thinning of cortical plates (Figure 5). Paranasal sinus view revealed a dome shaped radiopacity in left maxillary sinus (Figure 6).

Around 4 ml of blood tinged fluid was aspirated from the swelling which revealed inflammatory cells in a hemorrhagic background.

Incisional biopsy revealed cystic lumen lined by non keratinized stratified epithelium which exhibited palisading of basal cells with reversal of polarity. Overlying cell layers showed stellate reticulum like morphology. Aggregates of ghost cells were seen in epithelial lining. Capsular stroma showed the presence of dentinoid.

Based on these findings a final diagnosis of calcifying cystic odontogenic tumor was given. The lesion was surgically enucleated along with extraction of the teeth involved. No recurrence of the lesion was noticed on 11 months follow up.

## REVIEW

Calcifying cystic odontogenic tumor (CCOT), as defined by WHO, is a benign cystic neoplasm of odontogenic origin, characterized by an ameloblastoma-like epithelium with ghost cells that may calcify.<sup>1</sup>

The structure was first described by Gorlin et al in 1962 and was therefore called Gorlin cyst. There are two variants of lesion namely cyst and neoplasm. The basis for existence of lesion as cyst or neoplasm leads to various hypothesis which were based on either monistic concept or dualistic concept. According to monistic concept all variants are neoplastic, with tendency to undergo cystic change. Dualistic concept postulates that the lesion exists as two separate entities cyst and neoplasm. Various classifications of CCOT were proposed (Praetorius et al 1981, Hong et al 1991, Buchner 1991, Toida 1998, WHO 2005, Ledesma Montes et al 2008) based on monistic and dualistic concepts.<sup>2</sup>

CCOT is an extremely rare lesion accounting for 2% of all odontogenic pathologies. Tomich reported incidence of CCOT as less than two cases per year and recorded a total of 51 cases in 34 years.<sup>3</sup> Praterious suggested that the calcifying odontogenic cyst is a unicystic process which develops from reduced enamel epithelium or remnants of odontogenic epithelium in the follicle, gingival tissue or bone. Beta catenin gene mutations and beta catenin over expression, aberration in Wnt signaling pathway have been identified as characteristics of CCOT leading to disturbance in cell proliferation in odontogenic epithelium.<sup>4,5</sup>

## CLINICAL FEATURES

Clinically CCOT may present centrally or peripherally. Incidence of central variant is more than peripheral. In 2006, Buchner et al reported that the peripheral CCOTs account for 26% of all CCOTs.<sup>6</sup> CCOT can occur at any age, most reported cases shows maximum incidence in the 2<sup>nd</sup> decade of life, while some support bimodal age distribution with maximum incidence in 2<sup>nd</sup> and 7<sup>th</sup> decade.<sup>7</sup> CCOT can equally affect males and females.

It also has an equal predilection for both maxilla and mandible. Buchner reported an increased maxillary involvement in Asians. It commonly affects the anterior segment of jaws, with maximum propensity for incisor-canine region. The central, intraosseous variant usually presents as a slow growing painless swelling of jaws, unless secondarily infected (Table1). Most of the cases are asymptomatic, may sometimes complain of epistaxis and headache in case of maxillary involvement.<sup>8</sup> Extraosseous variant presents as pink to reddish, well circumscribed, smooth surfaced elevated masses measuring up to 4cm in diameter with no distinctive clinical features.<sup>1</sup> Peripheral CCOTs occur more commonly in mandible, anterior region and more often in females than males.<sup>6</sup> In 2011, Resende et al studied the clinicopathological profile of peripheral CCOTs from 1962 to 2010; he found out that the maximum number of cases present as painless swellings affecting the anterior segment of jaws.<sup>9</sup>

## RADIOGRAPHIC FEATURES

Radiographic features of CCOT are variable. It may appear as unilocular radiolucency with well defined or ill defined margins or multilocular radiolucency in 5-13% of cases. Generally appears as unilocular lesion with a well defined margin. Radiopaque structures within the lesion either irregular calcifications or tooth like densities are present in about one third to one half of the cases. Teeth divergence and root resorption are common findings.<sup>10</sup> It may produce expansion of cortical plates. Most cases vary from 2 to 4 cm in greatest diameter but lesion as large as 12 cm have been reported.

In early stages of development, it appears as a radiolucent lesion. In later stages it develops calcifications with a mixed radiolucent – radiopaque appearance. The radiopacity can be seen as flecks of salt and pepper, fluffy cloudy pattern or crescent – shaped pattern.<sup>3</sup>

Radiographic differentials of CCOT in early stages include odontogenic keratocyst, ameloblastoma and dentigerous cyst. In later stages adenomatoid odontogenic tumor, partially mineralized odontome, calcifying epithelial odontogenic tumor, ameloblastic fibro-odontoma should be considered in the radiographic differential diagnosis.

Extraosseous CCOTs may show saucerization and sometimes displacement of adjacent teeth.<sup>1</sup>

## HISTOPATHOLOGICAL FEATURES

The lesion shows characteristic histopathological features. The cyst lining shows proliferation and may resembles ameloblastoma, columnar cells over which are stellate and spindled shaped cells are present similar to stellate reticulum. Some cells undergo ghost cell keratinization. It may be associated with odontoma. The presence of ghost cells within proliferative odontogenic epithelium is the essential characteristic for the diagnosis.

## TREATMENT

The prognosis of CCOT is good, only few cases of recurrence have been reported after simple enucleation. When CCOT is associated with some other recognized odontogenic tumor, the treatment and prognosis are likely to be same as the associated tumor. In our review two cases of transformation into ghost cell odontogenic carcinoma have been reported following surgical enucleation (Table 1).

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**Dr. Ritika Jindal** B.D.S. (First Author), Post Graduate Student, Oral Medicine and Radiology, Manipal College of Dental Sciences, Mangalore, Manipal University, India



**Dr. Ravikiran Ongole** B.D.S., M.D.S. (Corresponding Author), Professor, Oral Medicine and Radiology, Manipal College of Dental Sciences, Mangalore, Manipal University, India. Corresponding Author: [oralcare@gmail.com](mailto:oralcare@gmail.com), +91 9448430387



**Dr. Junaid Ahmed** B.D.S., M.D.S., Professor and Head, Oral Medicine and Radiology, Manipal College of Dental Sciences, Mangalore, Manipal University, India



**Dr. Ceena Denny E.** B.D.S., M.D.S., Associate Professor, Oral Medicine and Radiology, Manipal College of Dental Sciences, Mangalore, Manipal University, India



Figure 1: Intraoral photograph showing palatal extent of swelling with respect to 24, 25, and 26.



Figure 2: Cropped panoramic radiograph revealing well defined unilocular radiolucency with respect to 23, 24, 25, 26, and 27. Root resorption is evident in relation to 24, 26.



Figure 3: Intraoral periapical radiograph with respect to 22, 23, 24, 25 showing well defined unilocular radiolucency with external root resorption evident with respect to 23, 24.



Figure 4: Intraoral periapical radiograph with respect to 25, 26, and 27 showing well defined radiolucency with external root resorption in 26 extending to coronal third of root.



Figure 5: Lateral maxillary occlusal radiograph with multilocular radiolucency with respect to 23, 24, 25 and 26 with expansion of buccal cortical plate.

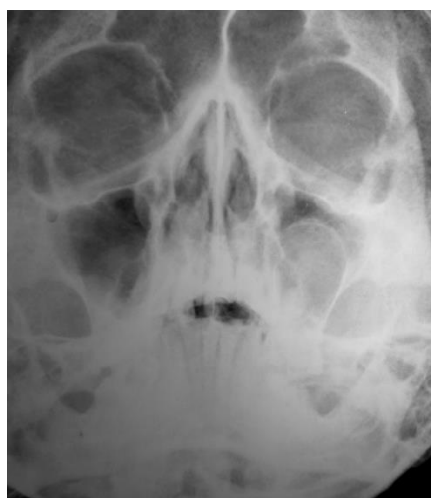


Figure 6: Cropped paranasal sinus view revealing dome shaped radiopacity in left maxillary sinus.

Table 1: Review of literature

Author	Age and Sex	Site	Clinical presentation	Radiographic finding	Diagnosis	Follow up
Santoshi Murakami et al, 2003	26/ F	Md molar region	Painful swelling	Cystic radiolucency with impacted third molar	COC	Description not given
Buch RS et al, 2003	11/ F	Md premolar region	Asymptomatic swelling with expansion of cortical plates	Radiolucency around crown of retained 84	Adenomatoid odontogenic tumor in COC	Description not given
Gallana Alvarez S et al, 2005	19/ M	Mx canine region	Asymptomatic tumefaction at canine fossa with unerupted canine	Well defined unilocular radiolucency with radiopaque mass	COC associated with complex odontome	No recurrence on 28 months follow up
Reyes D et al, 2007	5/ F	Mx	Asymptomatic swelling	Osteolysis with expansion of bony tables	Odontogenic calcificant cystic tumor	No recurrence on 1yr follow up
Reyes D et al, 2007	35/ F	Mx	Asymptomatic swelling with cortical expansion	Unilocular radiolucency with diffuse calcification with unerupted canine	Odontogenic calcificant cystic tumor	No recurrence on 1yr follow up
Han PP et al, 2007	15/ M	Md canine premolar region	Asymptomatic	Unilocular radiolucency with distinct sclerotic margins containing radiopaque mass	Pigmented CCOT with compound odontome	Description not given
Mala Kamboj et al, 2007	58/ F	Md canine to angle region	Pain and swelling with expansion of cortical plates	Multilocular radiolucency	Ameloblastoma tous COC	No recurrence
Ximena Zornosa et al, 2010	38/ M	Mx anterior region	Firm expansile swelling	Unilocular radiolucency with indistinct opacities in center	CCOT	Description not given
Patricia Devilliers et al, 2010	17/ M	Mx canine premolar region	Swelling associated with nasal obstruction and proptosis of eye	Unilocular radiolucency	CCOT associated with tumors such as keratocystic odontogenic tumor and orthokeratinized odontogenic cyst	Description not given
S.M. Balaji et al, 2012	43/ M	Mx incisor-canine region	Non tender hard bulge over alveolar ridge	Radiolucency	COC with Adenomatoid odontogenic tumor like features	No recurrence on 6 month follow up
Archana Sonone et al, 2011	23/ F	Mx anterior region	Non tender bony hard swelling	Unilocular radiolucency	Calcifying ghost cell	Description not given

				with radiopaque structures	odontogenic cyst	
Chindosomhat-jaroem et al,2012	13/F	Mx anterior region	Swelling of upper lip with vestibular swelling, unerupted lateral incisor and canine	Well defined expansile lesion with internal calcification with impacted canine	CCOT with odontoma, supernumerary tooth and a dentigerous cyst with impacted canine	No recurrence on 2 yr follow up
Arashiyama et al,2012	86/M	Md incisor-molar region	Painless swelling	Radiolucency	CCOT	On 18 year follow up transformed into ghost cell odontogenic carcinoma
Zhi-Yu et al,2012	51/M	Mx	Tender soft swelling	Radiolucency	CCOT	On 1 year follow up transformed into ghost cell odontogenic carcinoma
Thina Raren et al,2012	89/F	Mx	Bony hard swelling with tenderness	Radiolucency	Calcifying ghost cell odontogenic cyst	No recurrence on 1 yr follow up
Harkanwalpreet singh et al,2013	24/F	Md	Asymptomatic swelling	Well defined radiolucency with sclerotic margins with specks of calcification	Ameloblastoma tous calcifying ghost cell odontogenic cyst	No recurrence on 2 yr follow up

Age is given in years; Sex: M- Male, F- Female; Abbreviations used: COC- calcifying odontogenic cyst; CCOT- calcifying cystic odontogenic tumor; Mx- Maxilla; Md- Mandible