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# Central Calcifying Cystic Odontogenic Tumor Of Mandible – A Case Report

[1] Rajasekhar Gali — [1] Associate Professor
[2] Madan Mohan Reddy — [2] Reader
[3] Vandana Raghunath — [3] Professor
[4] Sajan Anand — [4]Post Graduate Student

[1,2,4] Department of Oral and Maxillofacial Surgery, Narayana Dental College, Nellore, Andhra Pradesh, India.
[3] Department of Oral and Maxillofacial Pathology, Narayana Dental College, Nellore, Andhra Pradesh, India.

**ABSTRACT:**. The calcifying cystic odontogenic tumor (CCOT) is a rare benign odontogenic cystic neoplasm. The terminological conundrum regarding its categorization as a cyst or a tumor has been resolved after the latest WHO classification (2005) that has labelled it as a tumor. The diversity in its morphological patterns, clinical behaviour, histologic complexity and prognosis has led to the practise of multiple management strategies with variable results. This report describes a case of CCOT presenting as a large cystic lesion in theanterior mandible that was managed by enucleation and curettage with a postoperative one year recurrence free follow up. A brief review of literature pertaining to various management strategies of CCOT in comparison with dentinogenic ghost cell tumor (DGCT) is also presented.

**KEYWORDS: Calcifying cystic odontogenic tumor, Calcifying odontogenic cyst, Cystic neoplasm**

## INTRODUCTION

The calcifying odontogenic cyst (COC) as a distinct clinical entity was first described by Gorlin et al. in 1962 as a rare developmental odontogenic cyst. [1]This lesion is known to exhibit diverse range of clinical, morphological and histo -pathologic features. This has led to several terminologies, classifications being proposed and various treatment modalities being tried and tested with variable results.[1,2,3] Putting an end to this confusion, the world health organization in 2005 has proposed a revised classification, based on morphological presentation and histopathological features, in to cystic or solid variants.The cystic variant was called calcifying cystic odontogenic tumor (CCOT), instead of COC and the solid variant was called dentinogenic ghost cell tumor (DGCOT).[4] The CCOT is a rare benign odontogenic cystic neoplasm that constitutes to about from 0.37% to 2.1% of all odontogenic tumors [5, 6].

### Case Report:

**History and Examination:** A 60 year old male patient was referred to the department of oral and maxillofacial surgery with a 6 month duration asymptomatic swelling in the chin region. On examination a 6 X 5 cm swelling was seen in the chin region with normal appearing overlying skin. Intra oral examination revealed a swelling measuring 5 x 6 cm causing labial vestibular obliteration extending from distal aspect of left mandibular second premolar to right mandibular canine region **(Fig.1).** The overlying mucosa was normal in appearance. The teeth in the region of the swelling were grossly attrited anddisplaced by the swelling but were firm and not decayed. On palpation the swelling was non-tender, bony hard in consistency except in left first premolar region where it was soft and compressible suggesting cortical perforation. There were no neurosensory disturbances in the surrounding region.

**Fig.1. Swelling in the anterior region of mandible causing vestibular Obliteration and displacement of teeth**

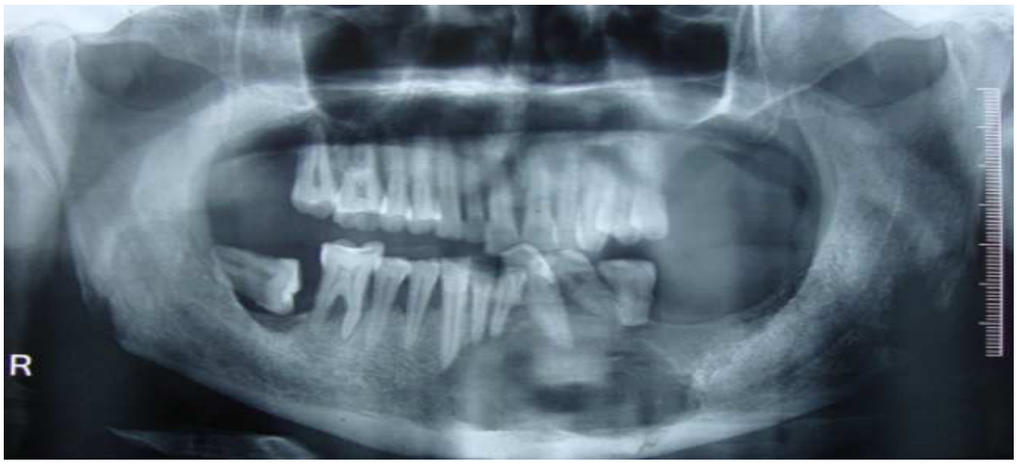


**Fig.2. Orthopantomograph demonstrating unilocular mixed lesion radiolucency with radio-opacity with sclerotic border causing root resorption and displacement of teeth along with signs of cortical perforation.**

**Radiographic Features:** Orthopantomograph revealed a unilocular mixed lesion - radioluscency with homogenous radio-opacities, with a sclerotic border located in the anterior region of mandible extending from right lower canine to an area 1cm distal to left lower 2nd premolar. Resorption of the roots and displacement of the teeth were observed **(Fig.2).**

**Aspiration and Incisional Biopsy:** Aspiration of the lesion yielded a clear straw coloured fluid. Electrical pulp vitality testing of all the involved lower anterior teeth showed them to be non-vital. Incisional biopsy of the lesion was performed that demonstrated the following histopathological features.

**Histopathological Features:** Histopathological examination revealed areas of cystic degeneration, Scattered irregular acellular dentinoid material, Numerous pale eosinophilic ghost cells, Multinucleated osteoclasts resorbing dentinoid, Solid areas of odontogenic epithelial cells **(Fig.3-A, Fig. 3-B, Fig. 3-C, Fig. 3-D).** Correlating the radiological, clinical and histopathological features a diagnosis of calcifying cystic odontogenic tumor of mandible was made.

**Treatment:** The lesion was treated by enucleation and curettage under localanethesia. A gingival crevicular incision was made in the mandibular anterior region extending from right lower second premolar to left lower second premolar. A mucoperiosteal flap was raised to expose the lesion that was enucleated followed by extraction all the lower anterior teeth from right canine to 2nd premolar **(Fig.4-A and Fig.4-B).** A sharp volkman's curette was used to curette the bony defect to remove remnants of the lesion and sharp bony margins smoothened by a vulcanite bur. Hemostatsis was achieved and the wound was closed with absorbable sutures. Post - operative period was uneventful. The patient was last seen 1year after surgery with no evidence ofrecurrence and is still under regular follow up **(Fig.5).**

## Discussion:

The calcifying cystic odontogenic tumor (CCOT) was previously known as calcifying odontogenic cyst (COC) or Gorlin cyst. This is a rare developmental odontogenic cystic lesion that encompasses a wide spectrum of clinical behaviour and histopathological features including cystic, solid and rarelymalignant variants [7, 8.] As a result of this diversity, different classification schemes and nomenclature for the lesion and its variants have been proposed. The world health organization in 2005 has proposed a revised classification, based on morphological presentation and histopathological features, in to cystic or solid variants. The cystic variant was called calcifying cystic odontogenic tumor (CCOT), instead of COC and the solid variant was called Dentinogenic ghost cell tumor (DGCOT)[4].

CCOT generally occurs in 2nd or 3rd decades of life and does not show any gender predilection. But the present case shows that it can occur in the sixth decade of life also. It can affect maxilla or mandible equally with specific predilection to the anterior region of the jaws.[9] The CCOT is believed to arise from odontogenic epithelial remnants trapped within the bones of the maxilla and mandible or gingival tissues [10], this explains the reason behind the occureence of CCOTs centrally (intraosseous) or peripherally (extraosseous)[7,9]. It usually presents as a painless, slow growing swelling that can cause cortical expansion and perforation in advanced stages.

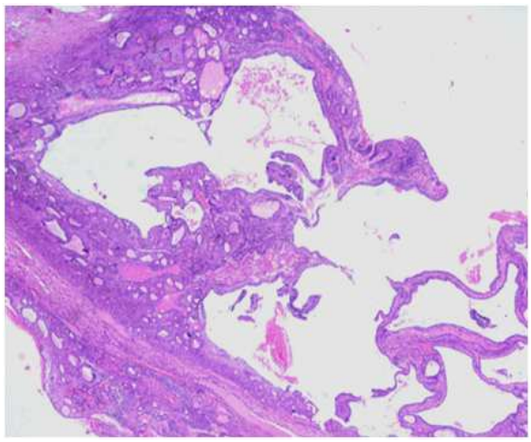

**Fig.3.A. Histopathology picture showing Solid areas of odontogenic epithelial cells arranged in strands along with areas of cystic degeneration**

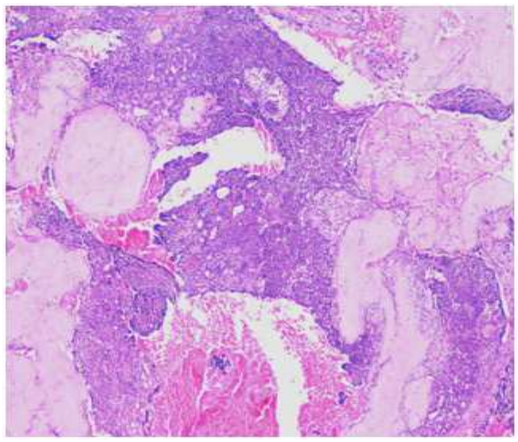

**Fig.3.B. Histopathology picture showing Hyper cellular solid arrangement of odontogenic epithelial cells with Scattered irregular acelluar dentinoid material and Numerous ghost cells**

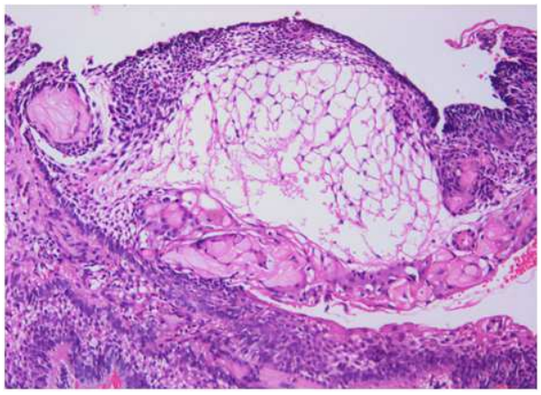

**Fig.3.C. Histopathology picture showing Proliferating ameloblastomatous strands, Central stellate reticulum like cells, numerous pale eosinophilic ghost cells**

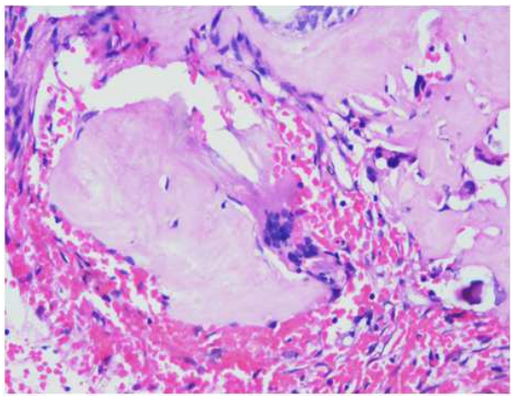

**Fig.3.D. Histopathology picture showing multi nucleated osteoclasts resorbing dentinoid**

Radiographic features include a variety of presentations but unilocular radioluscency with scattered radio-opacities lined by a sclerotic border is the commonest type seen. When CCOT is associated with an odontome, mixed lesions that are predominantly radio-opaque are observed9. Differential diagnosis of CCOT includes dentigerous cyst, adenomatoid odontogenic tumor, ameloblastic fibro-odontoma, and calcifying epithelial odontogenic tumor. Definitive diagnosis can be made histologically only.[9]

The histologic picture of CCOT can be very varied. The epithelial lining of CCOT has characteristic odontogenic/ ameloblastic features. The most remarkable feature is the presence of ghost cells and aberrant calcified tissue resembling dentine. Occasionally odontomes also can be associated with the lesion. The treatment of CCOT depends up on site and size of the lesion and histological pattern and includes simple enucleation and curettage, which means enucleation followed by removal of a 1 to 2 mm. layer of bone around the periphery of the cystic cavity with a sharp curette or a bone bur, to eliminate the epithelial offshoots that can lead to a recurrent lesion. Though occasional recurrence and malignant transformation have been reported, these are rare as far as CCOT is concerned. Buchner highlighted the importance of the long term follow up to 8yrs for CCOT lesions after treatment [9].

**Fig.4A. Intra-operative view: Enucleation of the lesion**

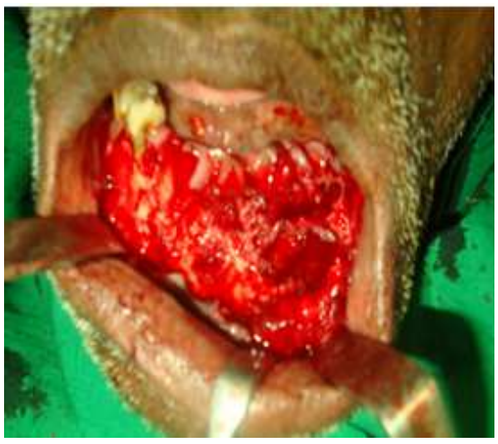

**Fig.4B. Intra-operative view:Post enucleation surgical defect of mandible**

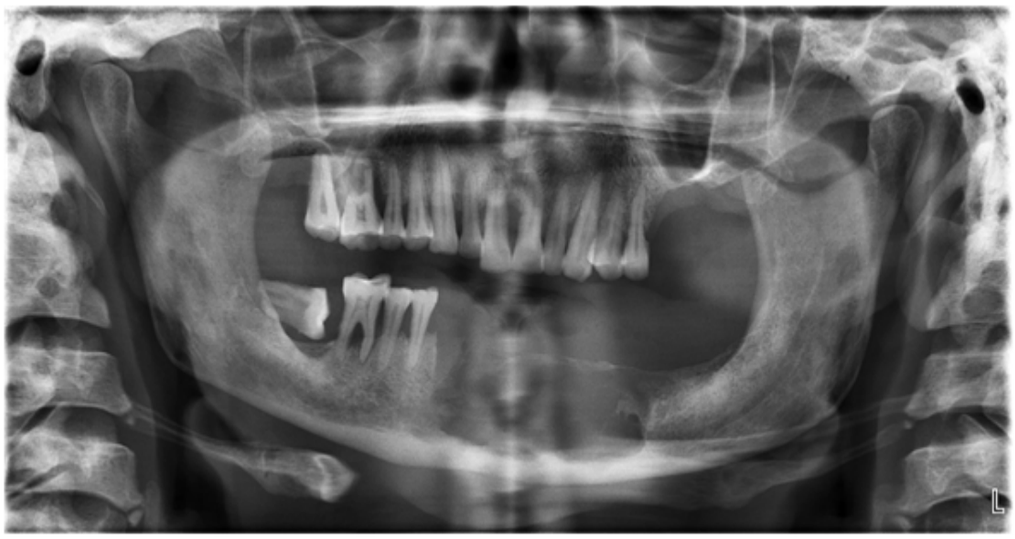

**Fig.5.one year Post-operative orthopantomograph showing no signs of recurrence of lesion.**

The need for distinguishing clinically and histologically between the solid variant (DGCT) and cystic variant (CCOT) is of paramaount importance as DGCT behaves aggressively and is prone to recur in complete contrast to CCOT. It warrants an aggressive surgical approach resection with or without continuity defect with a clearance margins of 0.5 mm similar to that of ameloblastoma [9].

## CONCLUSION

The present case report demonstrates that CCOT can occur in the sixth decade of life also and can mimic a peri apical cyst being associated with attrited and non-vital teeth. The lesion can cause significant amount of bone destruction causing cortical expansion and erosion if left unrecognized and untreated for a long time. The present case report contributes to the existing literature on this rare cystic neoplasm highlights the need for adding CCOT as one the differential diagnoses for mixed lesions (radiolucent and opaque) lesions affecting anterior mandible.

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**Corresponding Author**

Dr.Raja Sekhar. Gali, MDS, DNB ,
Associate Professor,
Department of Oral
and
Maxillofacial Surgery,
Narayana Dental College and hospital,
Nellore, Andhra Pradesh, India. – 524003
Phone No: 91-9490142971.
E-mail: rajmaxfacs@gmail.com