■ ***Case Report***

Calcifying cystic odontogenic tumor- a unique developmental lesion arising in mandible

S Vasudev[1], V P Singh[2], R Jhawar[1], D K Roy[2]
[1]D.A.P.M.R.V.Dental College, Bangalore, India; [2]College of Dental Surgery, BPKIHS Dharan, Nepal

Abstract

Calcifying cystic odontogenic tumor (CCOT) is a unique developmental lesion arising from the odontogenic epithelial remnants present in the jaw bones. It is a very rare lesion having a distinctive histopathologic appearance characterized by the presence of ghost cells with considerable amount of histopathologic diversity.

The case report describes a rare and unusually large presentation of CCOT in an 18 year old female. The patient presented with a large swelling in the mandibular' anterior region. After clinical, radiographic investigations, an incisional biopsy was performed and a diagnosis of CCOT was made. The cyst was enucleated with complete removal of cystic lining. The patient has recovered well and has been kept under observation.

Keywords: calcifying cystic odontogenic tumor; calcifying odontogenic cyst

Introduction

Calcifying cystic odontogenic tomor (CCOT) is a rare entity accounting for less than 2% of all odontogenic tumor.[1,2,3] It was originally described by Gorlin, Pindborg, Praetorius-Clausen and Vickers in 1962 as Calcifying odontogenic cyst (COC).[4] The lesion since its discovery was found to exhibit cystic, benign tumor like and rare malignant characteristics.[5] A wide range of histopathological appearances were also noted.

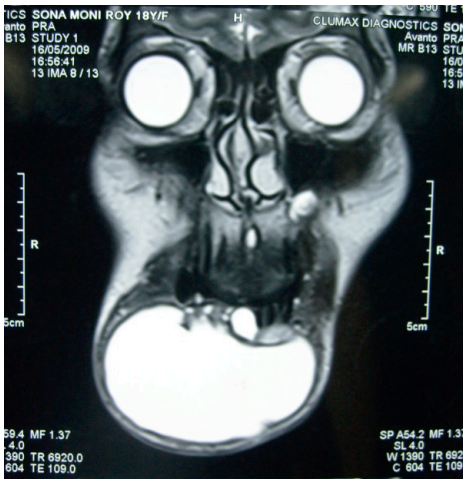
Thus its diverse clinical behavior and histopathologic characteristics prompted the usage of several terminologies and classifications. Gold in 1963 named the lesion as 'Keratinizing calcifying odontogenic cyst. Fejerskov and Krogh in 1972 called it 'Calcifying ghost cell odontogenic tumor' whereas Freedman et al in 1975 suggested the name 'Calcifying cystic odontogenic tumor (CCOT).Praetorius et al. in 1981 divided the lesion into two groups, 1) cystic and 2) neoplastic considering the different histologic patterns in them. The cystic form was further divided into (1a) simple unicystic, (lb) odontoma producing, and (1c) ameloblastomatous proliferating. The neoplastic counterpart was named 'Dentinogenic ghost cell tumor (DGCT)'. Subsequently, WHO in 2005 renamed COC as CCOT and the term DGCT was retained. A third malignant counterpart was found and the name 'Ghost cell odontogenic carcinoma (GCOC)'was proposed for the same. This classification has been widely accepted and is currently in use. Among the several forms of CCOT, the simple unicystic form of CCOT is most commonly encountered followed by the odontome associated CCOT.[6] This case reports one such rare and unusually large case of CCOT

Case report

An 18 year old female patient reported to the department of Oral and Maxillofacial surgery with chief complaint of a painless swelling in the mandibular anterior region for past 6 months. The swelling was insidious in onset and rapidly progressed to a larger extent. Mobility of the teeth in the same region was noted**.** Patient gave a history of consultation in another dental hospital one month prior where the mobile teeth were extracted followed by incisional biopsy and MRI of the lesion which were suggestive of a keratinizing odontogenic tumor. (Figure 1)

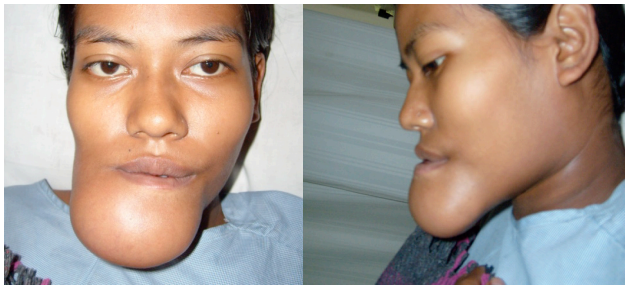
Address for correspondence:
Dr Sunil Vasudev, Professor
Department of Oral & Maxillofacial Surgery
D.A.P.M.R.V.Dental College, Bangalore (India)
Email: varundc@gmail.com

Figure 1: A coronal, Tl weighted MRI image showing large hypointense lesion arising from the mandible involving the symphysis and bilateral bodies measuring 5x8 cm



On extraoral examination, a marked facial asymmetry was seen due to a large swelling measuring 5x8 centimeters in the chin region extending laterally to involve the right lower third of face. The swelling extended around 5 cm inferiorly beneath the lower lip and a line joining the right angle of mouth to the tragus of ear .Antero-posterior it extended from left angle of the mouth to the border of ramus of the mandible. Skin over the swelling appeared stretched, the swelling was non tender and bony hard in consistency (Figure 2).

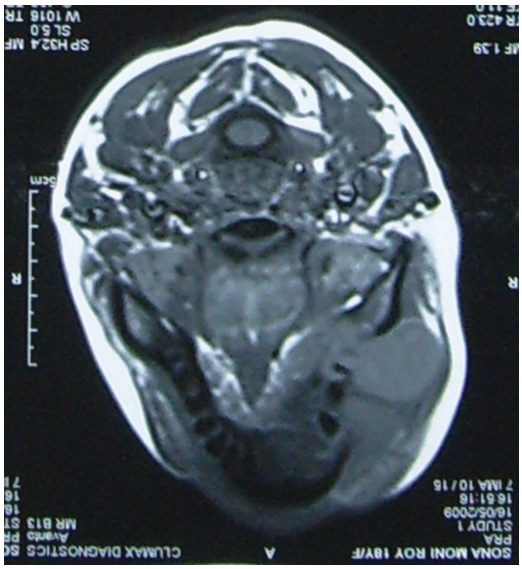
Figure 2: A large swelling measuring about 5x8 centimeters is seen in the right lower third of the race



Intraorally 41, 42 and 43 were missing. A bony hard swelling involving the alveolus with respect to 41, 42 and 43 was noted with marked expansion of the buccal cortical plate extending from 45 to 33 region which was further confirmed by MRI (Figure 3). The lingual cortical plate was expanded with respect to 41, 42 and 43 region.

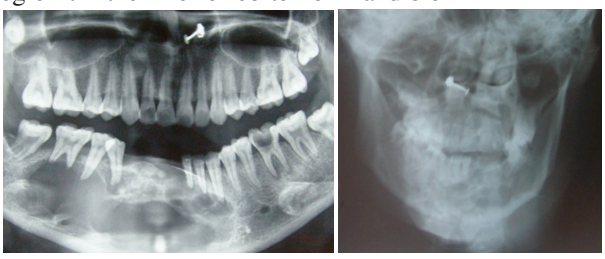
On electric pulp testing 35, 34, 33, 32, 31, 44, 45 gave a positive response. The Panoramic and

Figure 3: Expansion of buccal cortical plate seen extending from 45 to 33 region seen



Posteroanterior view of the skull showed a well defined multilocular radiolucency with scalloped margins involving the body of the mandible in the symphyseal right parasymphyseal region (Figure 4). Resorption of the roots of 44, 45 and 46 was also observed.

Figure 4: Radiographic examination shows a well defined multilocular radiolucency extending from 46 to 33 mediolateraly and superinferiorly, it extends a few centimeters below the alveolar crest in relation to 41,42,43 region till the inferior cortex of mandible



An incisional biopsy was performed and the histopathologic report revealed cystic areas with ghost cells undergoing calcification in the epithelial lining, features suggestive of calcifying cystic odontogenic tumor (CCOT) of the simple unicystic variety (Figure5).

The cyst was enucleated with complete removal of its lining along with the extraction of 44, 45 and 46 under general anesthesia preserving the lower border of mandible [Figure 6 (a,b,c)]. The Patient was under antibiotic coverage for 5 days in hospital and was discharged after that**.** The patient's recovery was satisfactory and the patient has been kept under observation and regular follow-up (Figure 7).

Figure 5: Histopathologic picture showing the presence of cystic areas with ghost cells undergoing calcification in the epithelial lining

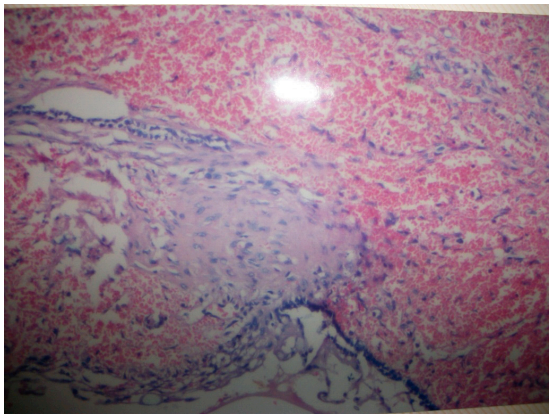


Figure 6(a,b,c): Intraoperative pictures showing the exposure of cyst lining and enucleation of cyst

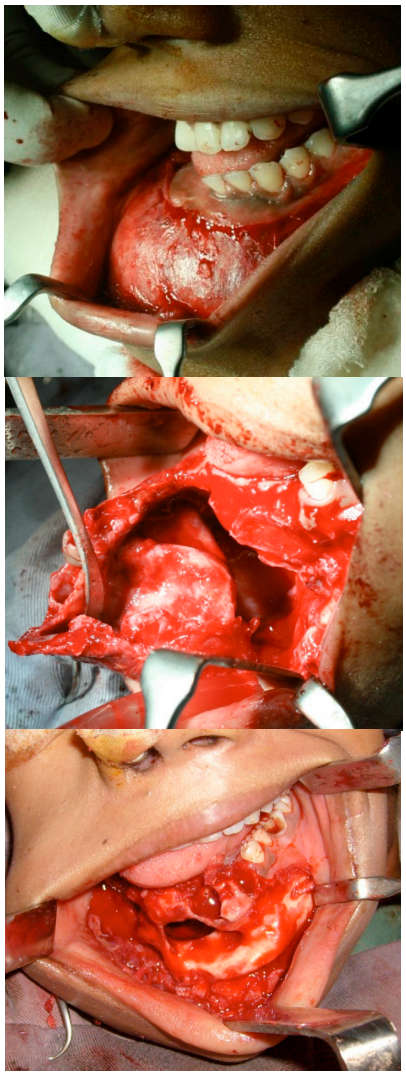
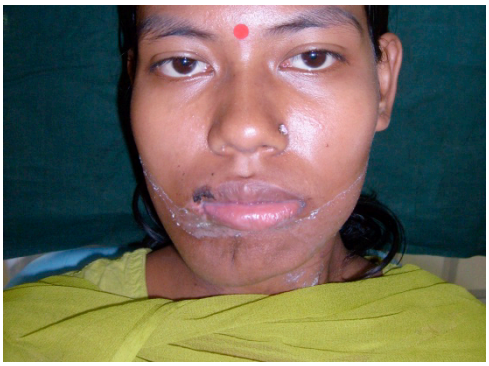


Figure-7: Post- operative photograph during followup



Discussion

CCOT is thought to originate from the remnants of odontogenic epithelium entrapped maxilla / mandible or gingival tissue giving rise to intraosseous or extraosseous lesion respectively.[7] However some authors suggest that CCOT may also originate from enamel organ or from islands of odontogenic epithelium within the tooth follicle.[2,8] The intraosseous form is more common with a prevalence rate of 70%.[9] Most of the studies do not demonstrate any gender predilection;[2,7,9] however one study has found a male predilection for unicystic CCOT.[10] The case reported here is of a female which is extremely rare.

The lesion has been found to occur over a wide age range of 1 to 82 years, the simple unicystic variant is found in the 2nd and 3rd decade. Bimodal peak of occurrence of the lesion in the 2nd and 7th decade has also been reported. This probably correlates with the age of occurrence of the intraosseous and extraosseous variants respectively.[2,6,9,11] The lesion is equally distributed between the maxilla and mandible,[7,9]although few studies suggests a preponderance in the maxilla especially for the simple unicystic variant . The anterior region is commonly involved followed by the premolar and molar area. Mandibular lesions may cross the midline which is a rare feature in the maxilla.[6,9,10] The lesion occurs in the tooth bearing area of the jaw either in a periapical, lateral periodontal or periocornal relationship of teeth. Patients usually report of an asymptomatic slowly growing swelling with a duration ranging from 6 months to 25 years. The size of the lesion usually ranges from 1 to 4 cm.[12,13]

The present case occurred in an 18 year old and was in a periapical to the mandibular anterior teeth which is in accordance with the previous reports. It demonstrated a rapid increase in the size of the

swelling with associated mobility of teeth and was found to be much larger measuring 5x8 cms which are in contrary to what has been reported. Expansion of the labial or buccal cortical plate invariably occurs usually sparing the lingual cortical plate.[9,11] The reported case here is unusual to what has been published in literature since lingual cortical expansion was noted. The enlarging lesion can occasionally perforate the cortical plate and displacement of adjacent teeth can also be seen.

The histopathological features of a classic calcifying cystic odontogenic tumor include the presence of cystic lumen with a fibrous capsule and a lining of odontogenic epithelium; The basal layer is well defined and made up of ameloblast-like columnar cells, an overlying layer often many cells thick that may resemble the stellate reticulum of the enamel organ is seen. The characterestic feature of this lesion is the presence of masses of 'ghost cells' which may be located within the epithelial lining or in the fibrous capsule.[2,6,9] These are epithelial cells devoid of nuclei which are eosinophilic and retain their basic cell outline. Ghost cells may form due to coagulation necrosis followed by dystrophic calcification or it may be a form of normal or abnormal keratinization of the odontogenic epithelium.[13] Occasionally ghost cells can evoke a foreign body reaction when they are present close to the connective tissue due to the disintegration of the basal layer and subsequent growth of granulation tissue.[9] Variable quantities of dentinoid or dentin like material are laid down adjacent to the epithelial lining and sometimes dental hard tissues resembling an odontome are found.[6] The lesion has to be differentiated from other lesions which may also show ghost cells namely ameloblastic fibroodontoma, odontoma, and carcinomas.[11]

Radiographically, a majority of lesions appear unilocular [6-9] with well-defined margins while in 5-13% of the cases they are multilocular.[9] Scattered irregular sized calcifications producing a mixed radioopaque radiolucent lesion may also be encountered, which may coalese later and give an appearance of tooth like densities within the lesion.[8] Simple unicystic CCOT occur more frequently as unilocular (60.9%) and well defined (95.2%) lesions.[6] Displacement and resorption of roots of teeth associated with the lesion may be seen.[2,7] In the present case the radiolucency was well defined and multilocular. The multilocularity in the present case could probably be related to the large size of the lesion.

A CCOT in the pericoronal location can in its earlier stages resemble a dentigerous cyst or ameloblastoma. In later phases when radiopaque foci are evident, it resembles adenomatoid odontogenic tumour, partially mineralized odontoma, calcifying epithelial odontogenic tumour, ameloblastic fibroodontoma.[2,9] Unilocular lesions in periapical location may mimic radicular or residual cysts, while multilocular lesions may resemble ameloblastomas or odontogenic keratocysts.[8]

The treatment of this cystic lesion involves enucleation with a long-term follow-up.[2,7,9] Recurrence though depends on completeness of cyst removal and has rarely been reported. Prognosis is good in case of simple unicystic CCOT and less certain for the other forms. In the case reported here, the CCOT involved the mandibular anterior region and radiographically presented as a well defined multilocular radiolucent lesion which is in accordance with literature. However, the fact that the lesion had achieved an enormous size in a short span of 6 months with expansion of buccal and lingual cortices and mobility of teeth gave a misleading picture of an aggressive neoplastic lesion. Therefore CCOT should be considered in the differential diagnosis of expansile aggressive lesions occurring in the anterior mandible.

References

1. Swan RH, Houston GD, Moore SP. Peripheral calcifying odontogenic cyst (Gorlin cyst). J Periodontal 1985; 56:340-3.
2. Manveen JK, Subramanyam RV, Simmerpreet SV, Ramandeep NS Calcifying Cystic. Odontogenic Tumour Mimicking as a Residual Cyst. J Clinical and Diagnostic Research. August 2010; 4(4): 2979-2983.
3. Kler S, Palaskar S, Shetty VP, Bhushan A. Intraosseous Calcifying cystic odontogenic tumor. J oral and maxillo facial pathology. Jan-Jun 2009;13(1):27-29.
4. Gorlin RJ, Pindborg JJ, Odon T Clausen FP, Vickers RA. The calcifying odontogenic cyst: a possible analogue of the cutaneous calcifying epithelioma of Malherbe. Oral Surg Oral Med Oral Pathol. 1962; 15:1235 - 1243.
5. Toida M. So-called calcifying odontogenic cyst: review and discussion on the tenninology and classification. J Oral Pathol Med. 1998; 27:49 - 52.
6. Montes CL et al. International collaborative study

on ghost cell odontogenic tumours: calcifying cystic odontogenic tumour, dentinogenic ghost cell tumour and ghost cell odontogenic carcinoma. J Oral Pathol Med. 2008; 37: 302-308.

7. Zomosa X, Muller S. Calcifying Cystic Odontogenic Tumor. Head and Neck Pathol 2010, DOl 10.1007/ sl2105-010-0197-z.
8. Erasmus JH, Thompson IOC, Rensburg LTV, Westhuijzen AJV. Central calcifying odontogenic cyst A review of the literature and the role of advanced imaging techniques. DentomaxiUofacial Radiology .1998; 27:30- 35
9. Rajkumar K, Kamal K, Sathish MR, Leena S. Calcifying odontogenic cyst. J oral maxillofacial pathology. 2004; 8 (2): 99-103.
10. Regezi A Joseph, Sciubba J James. Cysts of the oral region. In: Oral Pathology Clinical Pathologic correlations. 3rd edition. W. B. Saunders Company. 304-305.
11. Reyes D, Viilanueva J, Espinosa S, Comejo M. Odontogenic calcificant cystic tumor: A report of two clinical cases. Med. oral patol. oral cir.bucal 2007;12: E 126-129.
12. Norman K.Wood and Paul W Gaoz. Pericoronal radiolucencies. In : Norman K. Wood and Iris M. Kuc, editor. Oral and Maxillofacial lesions. edition. USA Mosby. 279-285.
13. Neville W Brad. Damn D Douglas. Odontogenic cyst & tumour. In: Allen tvl Caul, Bouquout E Jerry .editor. Oral & Maxillafacial Pathology, 4th ed. Ii-Saunders Company,Philadelphia. 506-509.