

CASE REPORT

Intraosseous calcifying cystic odontogenic tumor

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ABSTRACT

The calcifying odontogenic cyst was first reported by Gorlin *et al.* in 1962. It had been classified as a neoplasm related to the odontogenic apparatus because of its histological complexity and morphological diversity until it was renamed as a calcifying cystic odontogenic tumor by the WHO, in 2005. Here we describe a case of mandibular calcifying cystic odontogenic tumor in a 75-year-old male, which was present since five years, with a history of occurrence after the extraction of teeth in the involved region. The lesion was surgically removed and a histopathological examination revealed a cystic tumor with predominance of ghost cells and some amount of dentinoid tissue.

Key words: Calcifying cystic odontogenic tumor, ghost cells, odontogenic tumor

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INTRODUCTION

The calcifying cystic odontogenic tumor (CCOT) was described by Gorlin *et al.* in 1962, as a distinct entity derived from odontogenic epithelial remnants, showing various clinical and histological features.^[1] It is an extremely rare lesion representing 2% of all odontogenic pathological changes in the jaw.^[2] The CCOT usually arises intraosseously, but it may also occur extraosseously, with about equal frequency in the mandible and maxilla. The age of the patients may range from 5 to 92 years,^[3] with peak incidence in the second and sixth decade of life.^[4]

The unique histopathological features include a fibrous wall and a lining of odontogenic epithelium with columnar or cuboidal basal cells resembling ameloblasts. Stellate reticulum - like cells overlie the basal cell layer with the presence of 'ghost cells', which may occasionally become calcified, situated in the cyst lining or in the adjacent fibrous tissue. Hyalinized eosinophilic material, suggestive of immature or dysplastic dentin, is present, and is closely associated with the epithelial lining.^[1]

This report describes a case of CCOT that occurred in the mandible of a 75-year-old male, with no reports of recurrence since its complete removal in September 2007.

CASE REPORT

A 75-year-old male was referred to M M College of Dental Sciences and Research for a swelling on left side of the mandible. The patient first noticed the swelling five years back when he underwent uneventful extraction of 33, 34. Initially the swelling was minimal, but it had grown slowly with time to the present size. The swelling was painless throughout its course.

Clinical examination disclosed a 4 × 2 cm, ovoid firm swelling

on the residual ridge in the region of 32 to 34 [Figure 1]. The swelling was noncompressible and immobile with an intact overlying mucosa. There was no associated lymphadenopathy. Radiographically a poorly demarcated unilocular radiolucency with radiopaque foci in the region of 33, 34 was seen [Figure 2]. Expansion of the cortical plates in relation to the lesional site was evident.

The lesion was excised under local anesthesia. After fixation in buffered formalin the tissue was processed and stained with H and E stain.

The H and E stained section showed a cystic cavity lined by odontogenic epithelium of varying thickness (1–3 cell layers). The basal cells were cuboidal to columnar in shape with their nuclei pushed away from the basement membrane resembling ameloblasts. The outer layers of the cells were composed of loosely arranged angular cells resembling stellate reticulum, which in some areas were eosinophilic, ballooned, and keratinized forming ghost cells [Figure 3]. The nuclei of these cells were pushed to the periphery and eventually disappeared as the cells enlarged. Individual cells became confluent with the adjacent ballooning cells and gradually lost their cell boundaries, making large sheets of amorphous, acellular eosinophilic material, filling the cystic lumen. Some ghost cells were also seen in the underlying connective tissue, which was loose and vascular, with hemorrhagic areas along with some variable-sized islands of odontogenic epithelium. Irregular masses of hyalinized acellular calcified material (dentinoid) were also observed in the connective tissue, in relation to both epithelial lining and masses of ghost cells [Figure 4, Figure 5].

Based on these features the lesion was diagnosed intraosseous calcifying cyst odontogenic tumor. Healing was uneventful and in the six-month follow-up there were no signs of recurrence.



Figure 1: Swelling of the left anterior residual ridge

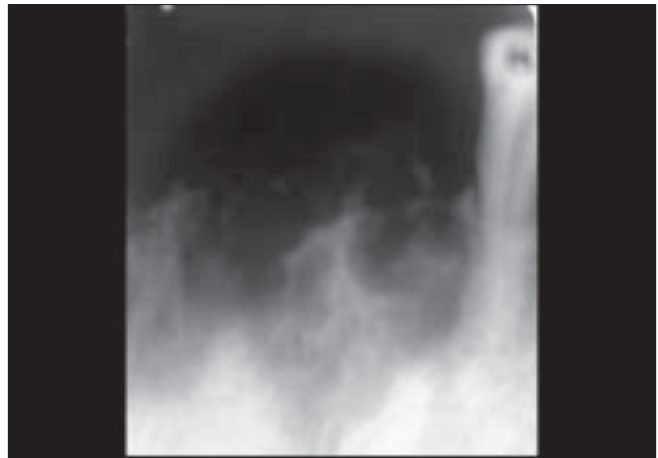


Figure 2: Diffuse unilocular radiolucency with radio-opaque foci in the region of 33, 34

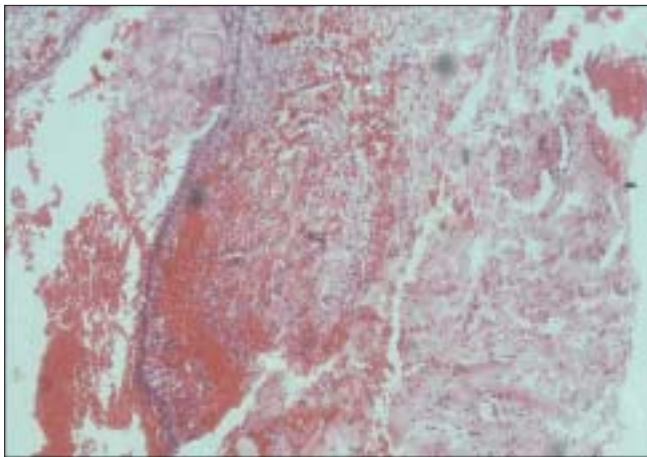


Figure 3: Photomicrograph showing odontogenic cyst lining with ghost cell proliferation (H and E, 10x)

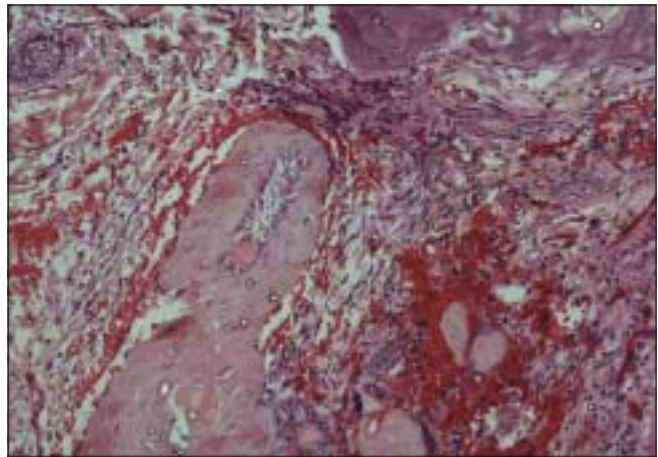


Figure 4: Photomicrograph showing dentinoid material and odontogenic islands (H and E, 20x)

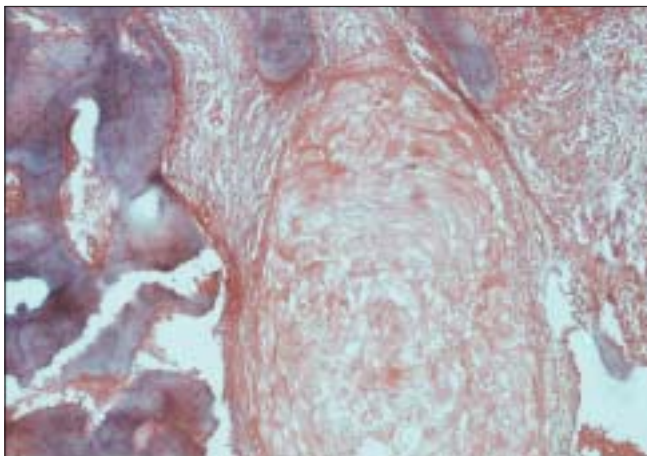


Figure 5: Photomicrograph showing sheets of ghost cells lined by epithelium adjacent to bone (H and E, 10x)

DISCUSSION

In 1971, calcifying odontogenic cyst was described as a nonneoplastic cystic lesion, but in 1992 the WHO classified

this lesion with odontogenic tumors because of its histological complexity and diversity. In 2005, the WHO renamed it as a calcifying cystic odontogenic tumor.^[5]

Clinically, CCOT corresponds to 2% of the odontogenic tumors^[2] with the age range between 5 to 92 years. Several articles have reported that a greater incidence takes place in the second decade, but other authors have noticed a bimodal distribution with a second peak of incidence in the sixth and seventh decade of life.^[3] In the present case the patient was in his eighth decade of life.

The location of CCOT can be intraosseous or extraosseous. Both central and peripheral forms occur with equal frequency in the maxilla and mandible with a majority of lesions being located in the incisor-canine area.^[4] The reported lesion was of the central variety and was present in the lower left mandibular region.

The central CCOT occurs as an asymptomatic hard swelling in the jaws, which produces expansion of bone, whereas, the peripheral variant presents as a nonspecific well-circumscribed

sessile or pedunculated mass, with a smooth surface.^[5] The presented case, similar to other reports in the literature, was an asymptomatic hard swelling present since the past five years.

Radiologically the lesion presents as a unilocular or multilocular, well-defined radiolucency. Radiopaque structures within the lesion, either irregular calcification or tooth-like densities are present in about one-third to half of the cases. The roots of the adjacent teeth are displaced or resorbed from the expansion of the lesion. The cortical bone is thinned, expanded or perforated.^[4] Radiographically the present case shows a diffuse unilocular radiolucency with some radio-opaque foci in the region of 33 and 34. Expansion of the buccal and lingual cortical plates is evident.

Although histological criteria have been established for the diagnosis of the CCOT, its pathogenesis is still speculative. Freedman *et al.* proposed that the neoplastic cell originated from a well-differentiated ameloblast, and its neural crest origin confers to this cell a pluripotential capacity to undergo terminal differentiation. Starting from the postulate that ameloblasts are stem cells, terminal differentiation is not necessarily required to originate the CCOT neoplastic cells.^[6]

Praetorius *et al.* and Buchner *et al.* believe COC cystic epithelium originates from the reduced enamel organ, from islands of odontogenic epithelium within the tooth follicle or from the remnants of odontogenic epithelium in the bone or gingival tissue.^[7] In the reported case, we believe the neoplastic epithelium arose from the remnant of odontogenic epithelium present in the bone.

The presence of the so-called ghost cells, which are assumed to represent an abnormal form of keratinization, is the most conspicuous feature of the lesion. However their mere presence does not justify a diagnosis of CCOT, as other lesions also show their formation. Hence diagnosis of CCOT should only be made for a lesion in which the formation of ghost cells takes place in a typical epithelial cyst lining, presenting a basal layer of cuboidal or short cylindrical cells and an overlying layer consisting of cells that bear resemblance to stellate reticulum-like cells.^[7]

The formation of dentinoid or osteoid, which is frequently described as being present in connection with masses of ghost cells is another characteristic finding of the lesion. Gorlin *et al.* considered the appearance of dentinoid material in CCOT to represent an inflammatory response of the body tissue toward masses of ghost cells.^[8] Abrams and Howell further stated that masses of ghost cells induce granulation tissue to lay down juxtaepithelial osteoid which may calcify.^[9] Contrary to these interceptions Sauk stated that the juxtaepithelial osteoid and dentinoid are frequently found in areas free of either granulation tissue or ghost cells and postulated that it might be an inductive phenomenon.^[10] To date it remains to be clarified as to whether this material represents a true inductive effect or merely a

metaplastic change in the connective tissue. Nevertheless in the present case these dentinoid-like regions were observed both in association with epithelium and ghost cells.

Due to the clinical characteristics of this lesion, the differential diagnosis of CCOT must be done with regard to the ameloblastoma, dentigerous cyst, and keratocysts. In more advanced stages it could be confused with calcifying epithelial odontogenic tumor, fibro odontoma, adenomatoid odontogenic tumor and partially mineralized odontoma.^[3] Histopathologically the differential includes ameloblastic fibro odontoma, complex and compound odontoma, ameloblastoma, and carcinomas.^[3]

In relation to the treatment, it must be conservative by means of enucleation or local resection. Recurrence of CCOT is rare, only eight cases of recurrences have been reported in English literature.^[2] The lack of recurrence depends on the complete excision. In the reported case complete excision of the tumor was performed. No complications have been reported till date. However, the case must be followed further to prevent any recurrence.

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