Case Report

Dentinogenic ghost cell tumor – A case report and review of literature

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Abstract

Dentinogenic ghost cell tumor (DGCT) was first described by Praetorius *et al.* in 1981 and now believed to be the tumor counterpart of the calcifying odontogenic cyst (COC). DGCT is an extremely rare odontogenic tumor and accounts for only 2% to 14% of all COCs. A case of DGCT in a 40-year-old female patient is being reported.

Keywords: Calcifying odontogenic cyst, dentinogenic ghost cell tumor, Gorlin's cyst, odontogenic ghost cell tumor

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INTRODUCTION

Calcifying odontogenic cyst (COC) is an entity well known to clinicians and pathologists. The COC was first described by Gorlin and his colleagues in 1962, as a separate entity of odontogenic origin.[1] Controversies and confusions still prevail regarding the different subtypes of the lesion. As all lesions are not cystic and the biological behavior is often not compatible with a cyst, there is a controversy as to whether COC is a cyst or a tumor.[2] Two organizing principles of classification of COCs have been put forward: monistic and dualistic.[3] The monistic concept, best exemplified by the World Health Organization classification,[4] postulates that all COCs are neoplastic in nature, even though the majority are cystic in architecture and appear to be nonneoplastic. In contrast, the dualistic concept, favored by most researchers,[5-7] proposes that COCs contain two different entities, a cyst and a neoplasm. The cystic lesions are termed as "calcifying cystic odontogenic tumors" and the neoplastic entity as a "dentinogenic ghost cell tumor" (DGCT).[1]

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There is paucity in the number of DGCT cases in the literature. The purpose of this article is to report a case of DGCT in a 40-year-old female, in the anterior region of the mandible.

CASE REPORT

A 40-year-old female patient reported with a chief complaint of a swelling in the front region of the lower jaw for 2 years. The history revealed that the swelling had started following the extraction of teeth, which gradually increased in size since its onset. The patient had experienced mild and continuous pain for 20 days and was not associated with discharge of any sort. Medical, surgical, dental, family and personal histories were not noteworthy. Extraoral examination disclosed a solitary, diffuse swelling of size approximately 4 cm × 5 cm in the lower anterior region of the jaw extending superoinferiorly from the right corner of the mouth to lower border of the mandible and extending into the submental region. Anteroposteriorly,

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it extended from right to left parasymphyseal region. On palpation, swelling was firm to hard in consistency and nontender on palpation. No regional lymphadenopathy was evident.

Intraoral examination [Figure 1] revealed swelling of size approximately 4 cm × 4 cm extending from distal surface of 33 to mesial surface of 43 labially and from distal surface of 34–46 lingually. Labial vestibule and lingual vestibule were obliterated and involved floor of the mouth. Buccal and lingual cortical plate expansion was seen. The swelling was hard in consistency labially and soft to firm in consistency lingually and tender on palpation. Clinical differential diagnosis included an ameloblastoma, central giant cell granuloma, adenomatoid odontogenic tumor and Pindborg tumor. Routine hematological investigations revealed normal values. Orthopantomograph (OPG) [Figure 2] revealed a multilocular radiolucency extending from 36 to 47. The lower border of mandible did not appear to be intact.

The hematoxylin- and eosin-stained sections showed numerous islands of odontogenic epithelium with tall columnar ameloblast-like cells with hyperchromatic nuclei. Large areas of eosinophilic globules suggestive of dentinoid [Figure 3] and ghost cells [Figure 4] were evident throughout the section. The histopathological impression was that of a DGCT.

DISCUSSION

COC was described as a distinct entity for the first time by Gorlin and his associates in 1962. COC constitutes 1% to 2% of all odontogenic tumors in which 88.5% are cystic and the remaining 11.5% are solid tumors.[7,8]

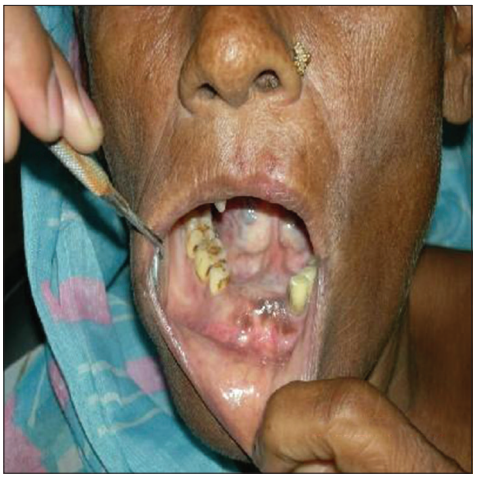

Figure 1: Intraoral view showing swelling obliterating labial vestibule and lingual vestibule

DGCT as a terminology was first proposed by Praetorius *et al.* in 1981 for the neoplastic variety of COC, i.e., the Type 2 of COC. DGCT has also been termed as odontogenic ghost cell tumor by Colmenero *et al.*[7] DGCT is an extremely rare odontogenic tumor and exists both as a central and a peripheral type. According to the available literature on central DGCTs, only 16 cases have been reported.[8,9] The average age for the presentation of this lesion is 50 years (range: 17–72 years), with slight male predilection.[7] Tumor occurs in the maxilla and the mandible with equal frequency,[7] with canine to first molar region the most often the affected site.[10] Patients are usually without symptoms, although with a few complaints of pain or discomfort. The present lesion was seen in a 45-year-old female, in the anterior region of the mandible.

DGCTs on panoramic radiographs show radiolucency with scattered radio-opaque calcifications with either unilocular or multilocular presentation. Occlusal radiographs show a bicortical expansion.[8,9] Root resorption or an impacted tooth in relation to the tumor mass is also noted in some cases.[7]

The present lesion on OPG revealed multilocular radiolucency with indistinct lower border of the mandible.

Histologically, DGCTs are composed primarily of ameloblastoma-like areas and odontogenic epithelial islands with varying amounts of ghost cells showing keratinization and calcification.[11]

The most important histologic feature of DGCT that distinguishes it from conventional ameloblastoma and other odontogenic tumors is the presence of ghost cells

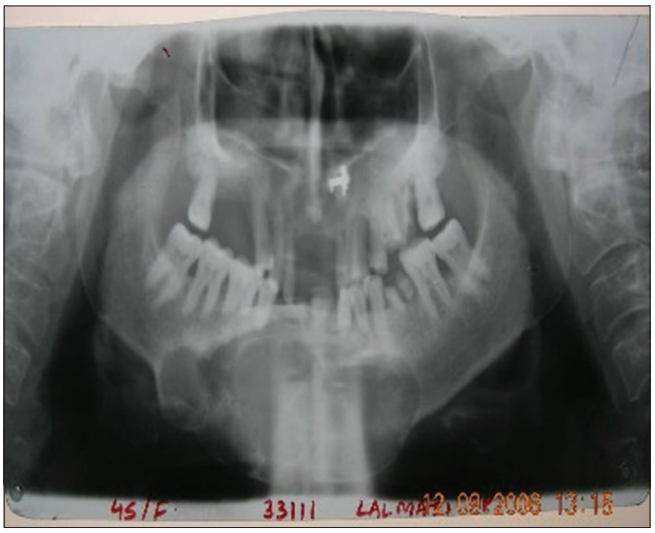

Figure 2: Orthopantomograph showing multilocular radiolucency extending from 36 to 47. The lower border of the mandible does not appear to be intact

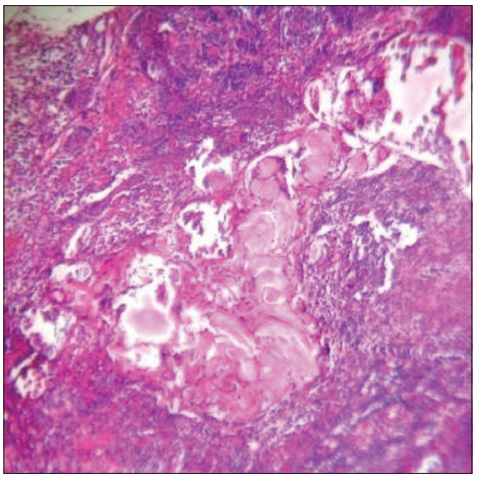

Figure 3: Eosinophilic dentinoid-like material interspersed in proliferating sheets of odontogenic epithelium (H&E stain, ×10)

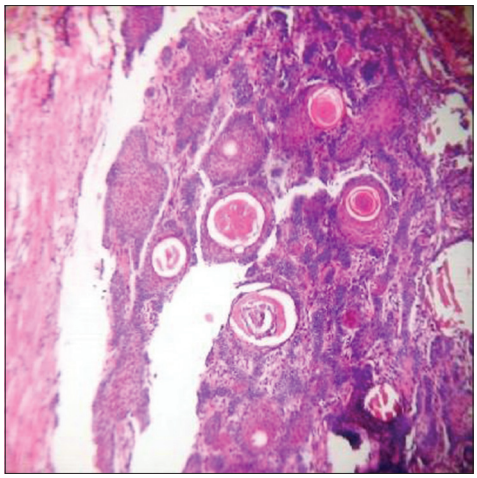

Figure 4: Ghost cells (H&E stain, ×10)

and dentinoid substances.[5] Ghost cells are believed to be transformed odontogenic epithelial cells, the mechanism of which is still unclear.[5,11] Although the presence of ghost cells is a defining feature for the diagnosis of DGCT, these cells can also be observed in other tumors, such as pilomatricoma, craniopharyngioma, odontoma and ameloblastic fibro-odontoma.[5,11] The nature of the dentinoid substance found in DGCT is unknown. It is amorphous eosinophilic material containing widely separated cell bodies. It lacks the tubular structure of normal dentin and appears as an irregular mass within the connective tissue adjacent to the proliferation of odontogenic epithelium.[5]

DGCT can be either benign or malignant, depending on the histopathological features.[6,12] Malignant transformation of a benign DGCT has also been reported.[13] The case being reported is a benign form of the DGCT.

Initially, enucleation was the primary treatment for central DGCT, but local recurrence was noted. Hence, at present, a more radical approach is accepted. The present lesion was treated with segmental mandibulectomy followed by rib grafting. The patient has been under observation for 9 months, and no recurrence has been noted till now.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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