CASE REPORT

Dentinogenic ghost cell tumor: A variant of Gorlin's cyst

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ABSTRACT
Calcifying odontogenic cyst (COC) was described as a distinct entity for the first time by Gorlin and his associates in 1962. Dentinogenic ghost cell tumor (DGCT) was described by Praetorius *et al.* in 1981 as a neoplastic variety of 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 male patient is being reported.
Key words: Calcifying odontogenic cyst, dentinogenic ghost cell tumor, Gorlin's cyst, odontogenic ghost cell tumor

INTRODUCTION

Calcifying odontogenic cyst (COC) is an entity well known to clinicians and pathologists. Controversies and confusions still prevail regarding the different subtypes of the lesion. One such less-known variant is the neoplastic type of COC which is called dentinogenic ghost cell tumor (DGCT). There is paucity in the number of DGCT cases in the literature, with only 16 cases available. DGCT can exhibit either a benign or a malignant form or can undergo malignant transformation.

CASE HISTORY

A 40-year-old male patient visited the Department of Oral Medicine and Radiology with a chief complaint of a swelling in the lower front tooth region since 4 months. The history revealed that the swelling had started insidiously, not preceded by trauma, which steadily increased in size since its onset. Patient had experienced mild and continuous pain in it of 20 days duration, and it was not associated with discharge of any sort. Medical, surgical, dental, family, and personal histories were not noteworthy. General physical examination revealed no abnormalities.

Extraoral examination disclosed a solitary, diffuse swelling over the mandibular symphysis, perceptible on the right side, oval in shape, measuring 3 × 2 cm in size, extending 0.5 cm below the vermillion and 3 cm above the inferior mandibular border, with no secondary changes or local rise of temperature. It was mildly tender and hard on palpation [Figure 1]. No regional lymphadenopathy was evident.

Intraoral examination revealed a solitary, diffuse, oval swelling in the mandibular labial sulcus, measuring 2 × 3 cm, extending from the tooth 31 up to the tooth 43; and mucosa over it was found to be normal. It was hard in consistency except at the inferior portion on the labial side, where decortication was evident [Figure 2]. Lingual cortical expansion was appreciated in the region of interest [Figure 3]. The teeth 31, 42, and 43 were grade I mobile; and 41 was grade III mobile. Thermal vitality test performed on 31, 41, 42, and 43 produced positive response. Aspiration yielded no fluid.

Clinical differential diagnosis included an ameloblastoma, central giant cell granuloma, adenomatoid odontogenic tumor, and pindborg tumor. Routine hematological investigations revealed normal values.

Intraoral periapical radiograph in the region of interest revealed a well-defined mixed radiolucent-radiopaque lesion measuring approximately 1.5 × 1 cm in size, extending from the mesial margin of the root of 31 up to the mesial margin of 43, with an ill-defined radiopacity 1 cm below the apices of the teeth. External root resorption of 31, 41, and 42 was evident [Figure 4].

Cross-sectional mandibular occlusal radiograph showed bicortical expansion, and the anterior mandibular occlusal radiograph showed a well-defined mixed radiolucent-radiopaque lesion with a wider area of involvement [Figure 5].

Panoramic radiograph revealed a well-defined mixed radiolucency, roughly 2.5 × 1.5 cm in size, inferiorly lined by a sclerotic margin, with an ill-defined radiopacity within. External root resorption of 31, 41, and 42 was evident, as well as superior displacement of the teeth [Figure 6].

The radiographic differential diagnosis incorporated a Gorlin's cyst, a pindborg tumor, an odonto-ameloblastoma, and ameloblastic-fibro-odontome.

The lesion was enucleated by raising a mucoperiosteal flap.

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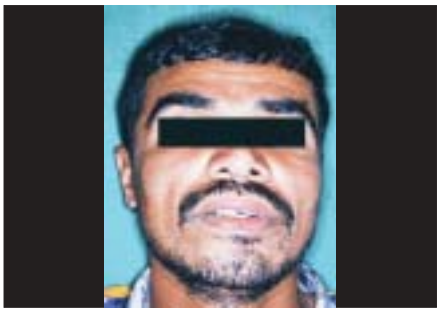

Figure 1: Extraoral photograph showing a diffuse swelling on the right side of symphysis of the mandible

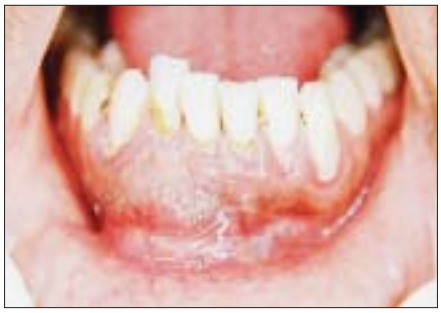

Figure 2: Intraoral photograph showing the swelling in the mandibular labial vestibule on the right side

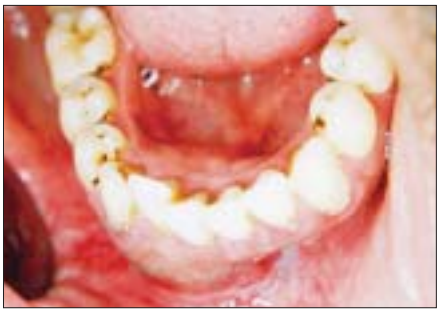

Figure 3: Intraoral photograph showing lingual expansion

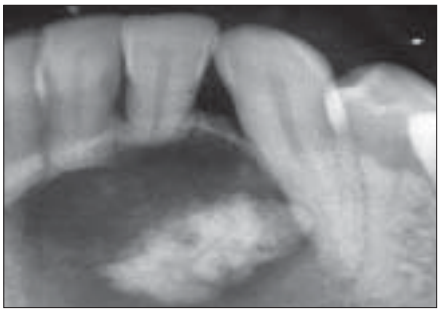

Figure 4: Intraoral periapical radiograph showing the mixed radiolucent-radiopaque lesion

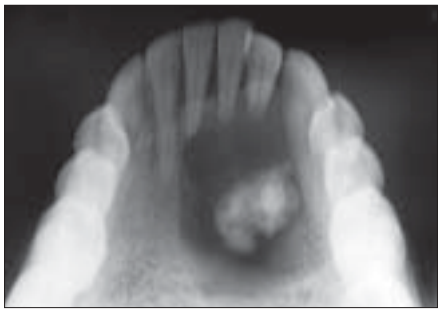

Figure 5: Anterior occlusal radiograph showing the mixed radiolucent-radiopaque lesion

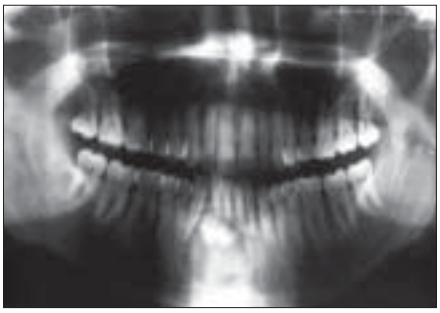

Figure 6: Panoramic radiograph showing the mass and displacements of the anterior teeth

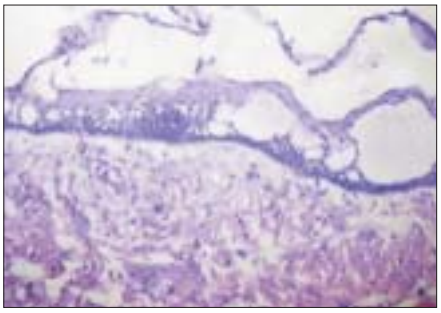

Figure 7: Photomicrograph showing odontogenic epithelial lining with ghost cells (H and E, 10×)

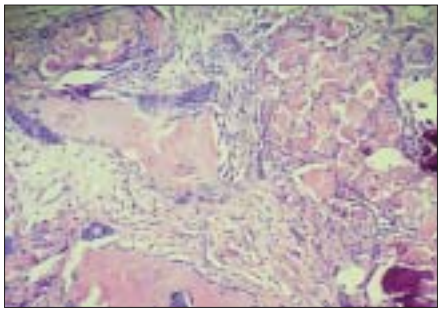

Figure 8: Photomicrograph showing large areas of ghost cell and dentinoid (H and E, 40×)

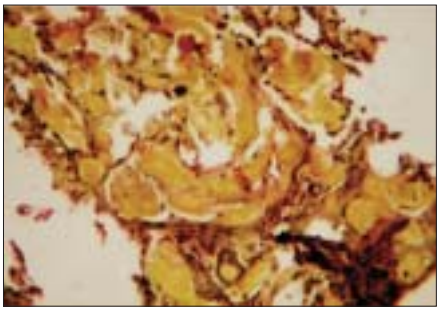

Figure 9: Photomicrograph showing yellow staining ghost cells with red staining dentinoid (Van-Geison stain)

The H and E - stained sections showed epithelium having tall columnar cells with hyperchromatic nuclei, along with loosely cohesive stellate reticulum - like cells. Eosinophilic cells with distinct outline characteristic of ghost cells were evident. Large areas of eosinophilic globules suggestive of dentinoid were seen throughout the section [Figures 7, 8]. The histopathological impression was that of a dentinogenic ghost cell tumor. To confirm the diagnosis, Van-Geison staining was done, which showed yellow-staining ghost cells with red-staining dentinoid [Figure 9]. No recurrence of the lesion has been observed 14 months after the treatment.

DISCUSSION

Calcifying odontogenic cyst (COC) was described as a distinct entity for the first time by Gorlin and his associates in 1962.[1] Since its first description, a number of cases have been reported, and it is now an entity well known to clinicians and pathologists. Controversies and confusions are still prevailing regarding the different subtypes of the lesion ever since it was identified. Two main types of COC are the cystic- and the solid-tumor type.[2] COCs have been called by different names by different investigators. Keratinizing and calcifying odontogenic cyst, calcifying ghost cell odontogenic cyst, cystic calcifying odontogenic tumor, and dentinogenic ghost cell tumor are a few of the terminologies that have been applied.

Dentinogenic ghost cell tumor (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.*[3]

DGCT is an extremely rare odontogenic tumor and exists both as a central and a peripheral type. COCs account for only 1% to 2% of all odontogenic cysts, and only 2% to 14% of them are DGCTs.[4] According to the available literature on central DGCTs, only 16 cases have been reported.[4,5]

Average age for the development of DGCT is 46.5 years. A slight propensity for development in males has been noted, with a ratio of 5:3.[4-6] A mandibular preference is noted for the central tumor,[4,5] and the present case occurred in the mandibular anterior region.

Not enough case reports are available to note the clinical presentation; nevertheless, DGCTs present as swellings causing bicortical expansion of the involved bone with smooth surfaces, with no mucosal ulcerations or regional lymphadenopathy.[4-6]

DGCTs on panoramic radiographs show a relatively well-defined radiolucent-radiopaque lesion of considerable size with either unilocular or multilocular presentation. Occlusal radiographs show a bicortical expansion. CT of the lesion reveals a soft tissue density mass with foci of calcifications.[4,5]

H and E sections of the solid type of the lesion show a lining consisting of a proliferative epithelium with numerous ghost cells having a tendency to develop foreign body granulomas.[7] The proliferative epithelium and the ghost cells are interspersed with abundant material called 'dentinoid,' and hence the lesion is collectively called a dentinogenic ghost cell tumor.[1] Under van-Geison staining, the ghost cells appear yellow; and the dentinoid, red.[8]

DGCT can be either benign or malignant, depending on the histopathological features. Malignant DGCTs can show aggressive clinical behavior and can metastasize.[1,9] Malignant transformation of a benign DGCT has also been reported.[10] The case being reported is a benign form of the tumor.

Treatment for central DGCT is surgical resection, which can be a segmental resection or an *en block* excision depending on the site and extent.[1] In the present case, enucleation of the tumor mass was done.

Local recurrences can be present in COCs in general and DGCTs in particular. Central DGCTs have been found to have a high rate of recurrences after resection.[5] Recurrent cases have occurred over 5 to 8 years following initial treatment.[4] The present case is under follow-up, and it has been 14 months after the treatment and no recurrence has been observed.

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