Case Report

Dentinogenic ghost-cell tumor of the maxilla: A case report and review of literature

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Abstract

Dentinogenic ghost-cell tumor (DGCT) is a rare, odontogenic neoplasm which is considered to be a solid variant of the calcifying odontogenic cyst (COC) with locally aggressive behavior and is characterized by ameloblastoma-like epithelial islands, ghost cells and dentinoid. It accounts for only 2%–14% of all COCs. Herewith, we report the case of DGCT in a 40-year-old male patient with clinical presentation as swelling on the right side of the face.

Keywords: Dentinogenic ghost-cell tumor, dentinoid, ghost cell, odontogenic tumor

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INTRODUCTION

The dentinogenic ghost-cell tumor (DGCT) is an odontogenic neoplasm, which is uncommon. It is deemed to be a solid variant of the calcifying odontogenic cyst (COC). COC was first recognized as a distinct entity by Gorlin *et al.*[1] and hence the eponym of Gorlin cyst. Praetorius *et al.*[2] classified them into two distinct types: the cystic type (Type I) and the solid type (Type II). The solid variant of COC (Type II) is rare and is designated as DGCT, with only 2%–14% of COCs presenting as solid tumors, which are considered to be DGCTs.[2]

DGCT is categorized by ameloblastomatous odontogenic epithelium, presence of ghost cells and dentinoid material. It is predominantly seen in middle-aged persons and can present either as a central or a peripheral lesion. Excisional biopsy is routinely performed on suspected cases.[2] Herewith, we report the case of DGCT in a 40-year-old male patient with clinical presentation as a swelling on the right side of the face.

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Review of literature

"Odontogenic ghost-cell lesions (OGCL)" or "ghost-cell odontogenic tumors," are characterized by their most characteristic microscopic feature, that is, the presence of ghost cells. They were projected beneath the 2005 United Nations Organization pointers,[2] within which DGCTs are classified beneath the benign solid tumors.[3]

Due to its varied histological appearances, many terms are employed by completely different authors to explain this lesion such as DGCT,[4] calcifying ghost-cell odontogenic tumor,[5] keratinizing ameloblastoma,[6] cystic calcifying odontogenic tumor,[7] peripheral odontogenic tumor with ghost cells,[8] dentinoameloblastoma,[9] ameloblastic

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dentinoma,[10] DGCT[11] and odontogenic ghost-cell tumor.[12]

DGCT is considered to be the rarest condition among OGCL, accounting for <1% of all odontogenic tumors.[13] The term "ghost cells" was introduced by Thoma and Goldman[14] in 1946. In 1933, Masaki described the first-ever case of DGCT, and in 1953, Husted and Pindborg[15] in Copenhagen reported a case of DGCT showing recurrence.

As per the systematic review by de Arruda *et al*.[13] and peers, DGCT is most commonly seen in the lower jaw, primarily seen anteriorly. It is more commonly seen in males than that of females, with a maximum incidence in the fifth decade of life. They mentioned that the intraosseous DGCT shows infiltrative growth pattern and recurrence even after resective procedure.[16]

CASE REPORT

In the present case, a 40-year-old Indian male visiting a private dental facility presented with a 6-month history of a swelling on the right side of his face. On extraoral examination, the base of the left ala was slightly raised. There was, however, no change in color/texture or erythema over the overlying skin or upper lip [Figure 1]. Lymph nodes were nonpalpable on physical examination.

The intraoral examination demonstrated a gingival swelling around the right second premolar and first molar of the maxilla, obliterating the buccal vestibule. Intraorally, the swelling was around 3 cm in diameter, hard and nonulcerated, smooth and exhibited redness and tenderness on palpation. The right second premolar and first molar were extruded with slight mobility. Intraorally, an ovoid mass was visible on the attached gingiva between the right second premolar and first molar of the maxilla. It had a reddish-pink color similar to the normal surrounding gingiva [Figure 2]. On palpation, it was firm and had a "Ping-Pong ball" feel.

Written consent was obtained from the patient, and the lesion was excised under local anesthesia and sent for histopathological examination. On gross examination, it was found to be a soft-tissue specimen, which was firm in consistency but with friable edges and cream to white to brown in color. It was a well-encapsulated lesion. It measured 2.5 cm × 2.2 cm × 1.5 cm in size, irregularly shaped and had a smooth surface [Figure 3].

Differential diagnosis

Based on the clinical assessment, the lesion was a maxillary swelling presenting for a short duration. A wide range of differential diagnosis could include inflammatory lesions such as osteomyelitis, nonodontogenic malignancies such as chondrosarcoma, osteosarcoma and metastatic tumors as well as odontogenic malignancies such as ameloblastic fibroodontosarcoma-dentinosarcoma or ghost-cell odontogenic carcinoma.[17]

In addition, benign nonodontogenic tumors such as cemento-ossifying fibroma or benign odontogenic tumors containing hard tissues, namely calcifying epithelial odontogenic tumor (CEOT), calcifying cystic odontogenic tumor, adenomatoid odontogenic tumor (AOT) and DGCT can also be considered in the differential diagnosis.[17]

The clinical description indicated a rapidly enlarging lesion with no mucosal ulceration. Although the malignancy of odontogenic origin containing dental hard tissue is a possible diagnosis, discrepancies in the clinical and radiological description do not allow the exclusion of locally aggressive benign nonodontogenic tumors and

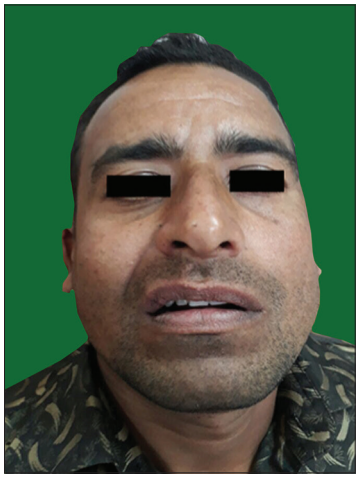

Figure 1: Extraoral swelling on the right side of the face

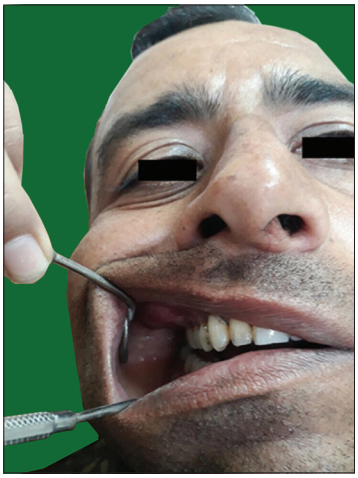

Figure 2: Intraoral swelling seen adjacent to 15, 16

odontogenic tumors containing hard tissues such as CEOT and DGCT.

Although AOT predominantly occurs in the anterior maxilla, clinical description of the aggressive lesion described in the present case did not fit with the recognized behavior of AOT, which is nonaggressive.[17] Moreover, as malignancy of odontogenic origin containing dental hard tissue is very rare, with only a few cases documented in literature, a locally aggressive but benign odontogenic tumor containing dental hard tissue is the most likely diagnosis.[18]

The absence of associated signs such as lymphadenopathy, fever, leukocytosis and predisposing conditions such as trauma, Paget's disease, diabetes as well as the occurrence of the lesion in the maxilla, which has a good blood supply exclude the possibility of chronic suppurativeosteomylelitis.[17]

Osteosarcomas and metastatic malignancies commonly occur in the mandible as well as the age of the patient could also be used to exclude the above-mentioned lesions.[17]

Diagnosis and histopathology

The patient was initially treated with a conservative surgery under the provisional diagnosis of an odontogenic tumor. The entire specimen was sent for routine processing in the histopathology laboratory. Microscopically, on viewing the hematoxylin and eosin-stained sections for this case, the specimen revealed odontogenic epithelium lining with a prominent basal layer consisting of palisaded columnar cells and hyperchromatic nuclei polarized away from the basement membrane [Figure 4]. Numerous ghost cells could be appreciated throughout the section [Figure 5]. Homogenous eosinophilic area simulating dentinoid could be seen in close proximity to the odontogenic epithelial lining and the ghost cells [Figure 6].

Van Gieson special stain was carried out to examine the nature of the dentinoid-like material [Figure 7]. The characteristic microscopic features, the confirmation of dentioid-like material and ghost cells by special stain contributed to the diagnosis of DGCT.

DISCUSSION

The World Health Organization (WHO) panel of experts on odontogenic tumors has defined DGCT as "locally invasive neoplasm characterized by ameloblastoma-like islands of epithelial cells in a mature connective tissue stroma. Aberrant keratinization may be found in the form of ghost cells in association with varying amounts of dysplastic dentin."[2] It occurs at an average age of 50 years with a slight male predilection and equal frequency of involvement of maxilla and mandible in canine to the first molar region as a predominant site. Calcification, root resorption and association of impacted tooth are observed radiographically.

The etiology of this rare lesion is still unknown, but it has been suggested that the missense mutation in β-catenin in the wingless integrated pathway plays a crucial role in the development of DGCT. The treatment is conservative enucleation, but local recurrence was noted.[19]

In this article, the authors present the case of a 40-year-old male diagnosed with DGCT, a neoplastic form of COC,

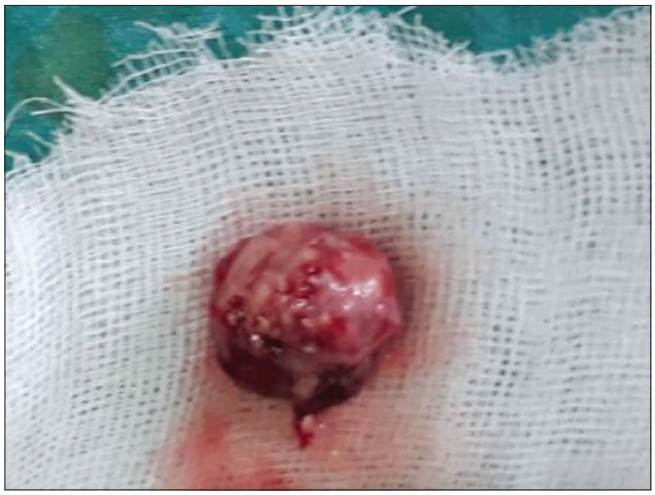

Figure 3: The gross appearance of the excised lesion. Grossly, submitted specimen was soft tissue, firm in consistency but with friable edges, cream to white to brown in color, irregularly shaped, measuring 2.5 cm × 2.2 cm × 1.5 cm

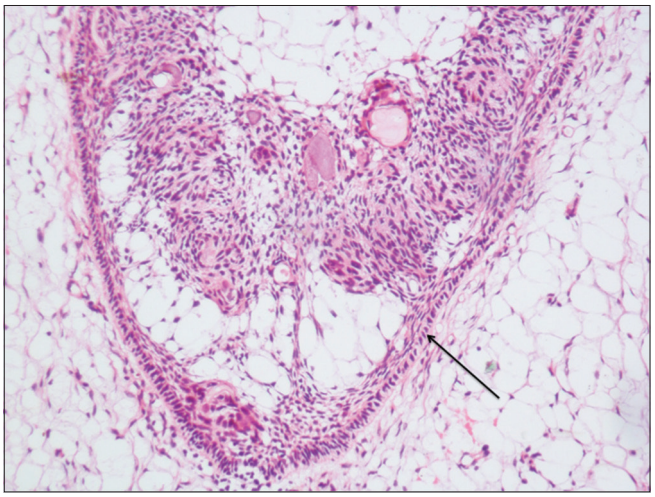

Figure 4: H and E-stained photomicrograph revealing odontogenic epithelium lining with a prominent basal layer consisting of palisaded columnar cells and hyperchromatic nuclei polarized away from the basement membrane (arrow), at × 40

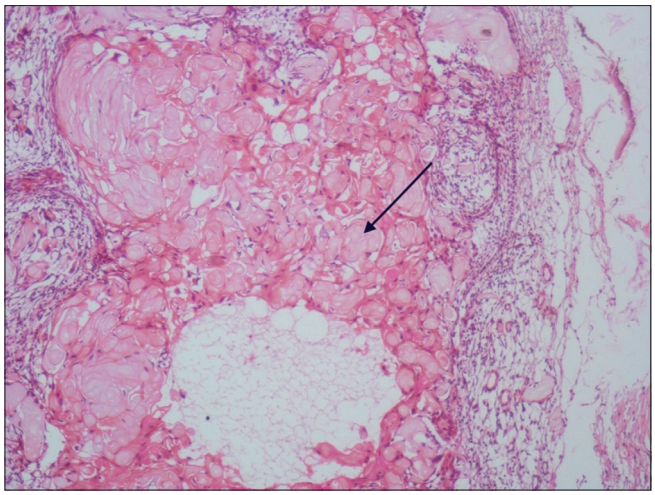

Figure 5: H and E-stained photomicrograph showing numerous ghost cells (arrow), at ×40

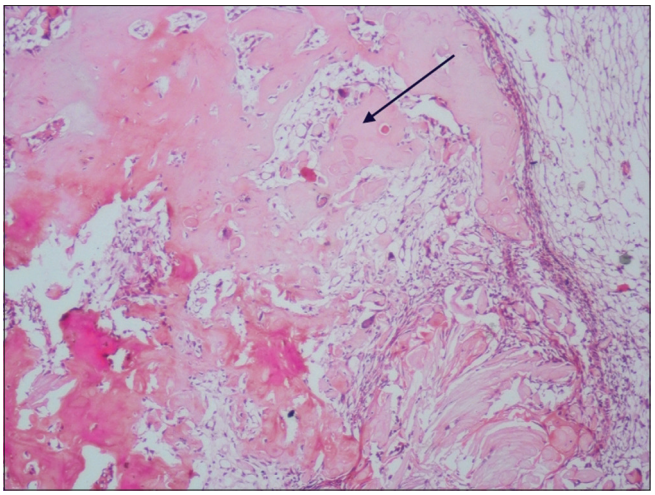

Figure 6: H and E-stained photomicrograph showing homogeneous eosinophilic area simulating dentinoid (arrow) seen in close proximity to the odontogenic epithelial lining and the ghost cells), at × 40

due to its characteristic histologic features, numerous ghost cells and dentinoid material.

COC constitutes 1%–2% of all odontogenic lesions occurring in the oral cavity. Of this, 88% of COC shows cystic nature, whereas 12% are solid in nature.[19] In 2005, the WHO has renamed COC based on its proliferative qualities as calcifying cystic odontogenic tumor to the cystic type of COC and neoplastic variant as DGCT to the solid form of COC. DGCT is a neoplastic counterpart of COC described and renamed by Praetorius and Ledesma-Montes in 1981.[3] It affects both the jaws with a slightly higher ratio in the anterior region. Clinically, the lesion is asymptomatic but causes noticeable swelling with the asymmetry of the face, which, in turn, depends on the size of the lesion. Mixed features of radiolucent destruction and radiopaque flecks were observed with the wide incidence of 2nd–7th decade of life.

Histopathologically, sheets and rounded islands of odontogenic epithelial cells seen in a mature connective tissue characterize the central and peripheral DGCT. The epithelium of the tumor islands resembles that of ameloblastoma. Mitosis is not seen. Minor cysts may form in the epithelial islands.[2]

A characteristic feature of DGCT is the transformation of the epithelial cells into ghost cells, which are keratinized and appear swollen and ellipsoidal. They present with a loss of nuclei and preservation of basic cellular outlines, are resistant to resorption, and have the potential to calcify. They are presumed to be derived either from the transformation of epithelial cells, metaplastic transformation of odontogenic epithelium, squamous metaplasia with secondary calcification due to degeneration of epithelial cells, ischemia or as a result of apoptotic process.[3] Several researchers have proposed that ghost cells represent an abnormality or incomplete keratinization process or are in an advanced stage of keratinization. Bafna *et al.* suggested that ghost cells may be representative of abortive enamel matrix formation in the odontogenic epithelium.[3]

Although ghost cells are a basic prerequisite for the diagnosis of the DGCT, it must be stressed that the presence of ghost cells alone is not pathognomonic since they can also be identified in other neoplasms such as odontomas, ameloblastomas and ameloblastic fibro-odontomas. The latter tumor can be eliminated from the histopathological differential diagnosis by the presence of a cellular primitive ectomesenchyme resembling dental papilla.[1]

Based on the study of H and E-stained paraffin sections, under light and electron microscope, Donath *et al.*, in 1979, were of the opinion that dentinoid material is not a product of mesodermal cells but is representative of a hard type of keratin similar to that found in nails. Praetorius *et al.* also suggested the material is of mesodermal origin based on the following findings:[2]

- Dentinoid will stain with connective tissue stains such as Van Gieson, Heidenhain, Goldner and Masson, like collagen
- Dentinoid is usually not found in the luminal proliferations, unless there is a disintegration of the basement membrane with the outgrowth of connective tissue between the epithelial ghost cells.

The present case discusses the occurrence of DGCT, and it is in accordance with the literature on the tumor. DGCT is seen

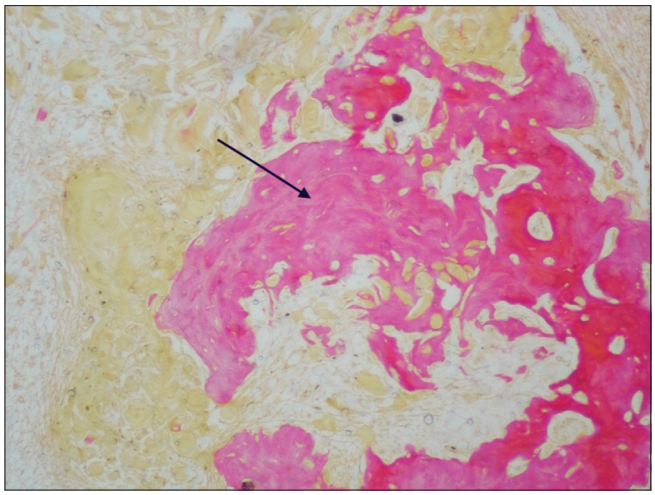

Figure 7: Photomicrograph showing dentinoid-like material (arrow) stained with Van Gieson stain, at × 40

most commonly in the 2–7th and can present in both upper and lower jaws, which is in accordance with our patient, who is 40 years of age and presented with a swelling in the maxilla.

CONCLUSION

It is important to study DGCT clinically, macroscopically, radiographically and histopathologically. Malignant transformation of this lesion can occur to its more aggressive counterpart, odontogenic ghost cell carcinoma and hence, regular follow-up of diagnosed cases is imperative. The purpose of this article is to report a case on a rare entity so that professionals globally can understand the biological behavior of these lesions better which will enable us to reach an effective diagnosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for 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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Nil.

Conflicts of interest

There are no conflicts of interest.

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