Case Report

Dentinogenic ghost cell tumor: a case report

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ABSTRACT

Dentinogenic ghost cell tumor is a rare odontogenic neoplasm that was initially considered to be a solid variant of Calcifying Odontogenic Cyst with locally aggressive behavior. Dentinogenic ghost cell tumor comprises less than 0.5% of all odontogenic tumors and characterized histologically by an ameloblastomatous epithelium with an area of ghost cell formation and a varying amount of dentinoid. Herein we report a case of intraosseous Dentinogenic ghost cell tumor in a 32 years old male patient with a clinical presentation of bony hard swelling in the left mandibular region. The diagnosis was made on the basis of histopathological findings and histochemical analysis.

INTRODUCTION

Dentinogenic ghost cell tumor (DGCT) is an extremely rare neoplasm accounting for less than 0.5% of all odontogenic tumors. It was initially described as a solid variant of Calcifying Odontogenic Cyst (COC).¹ DGCT is a benign but locally infiltrating neoplasm of odontogenic epithelium. It has biphasic morphology, consisting of a predominant ameloblastomatous proliferation and less prominent component of basaloid to stellate reticulum cells. The tumor characteristically contains aberrant keratinization, with a variable number of ghost cells and material morphologically resembling dentinoid or osteodentin.² DGCT, occurs centrally in the jaw and peripherally in the gingival or alveolar mucosa.¹

# CASE REPORT

A 32-years male patient presented with complains of swelling in the left lower back region of the jaw for 5 years. The swelling was gradually increasing in size and was not associated with pain. The patient had a habit of smoking 5-7...
cigarettes per day for 10 years. On extraoral examination, gross facial asymmetry was noted with noticeable swelling on the left lower back region of the jaw. Lymph nodes were not palpable.

On intraoral examination, bony hard swelling of about 3cm x 2cm in size was present on the left side of mandible extending from 41 to distal aspect of 34. There was obliteration of the labial vestibule (fig. 1). No visible changes in overlying mucosa were appreciated and the associated teeth were vital. On radiographic examination computed tomography revealed ill defined multilocular lesion having mixed radiopacity and radiolucency extending from 44 to 34 (fig. 2A). Expansion of labial cortical plate with areas showing perforation of the labial cortical plate at a different level was present. No expansion of the lingual cortical plate was noted (fig. 2A and 2B).

Based on the clinical and radiographic findings differential diagnosis of Central ossifying fibroma and Calcifying epithelial odontogenic tumor were given. An incisional biopsy of the lesion was performed. Histopathological examination of the tissue revealed areas of ameloblastomatous epithelium with stellate reticulum like cells. Aggregates of ghost cells with eosinophilic cytoplasm were appreciated scattered within epithelium (fig 3A). Large areas of dentinoid with no tubular structures having few cells were also noted. Aggregates of ghost cells were also seen entrapped within the dentinoid like areas (fig. 3B). For the confirmation of the diagnosis Masson trichrome stain was performed in which dentinoid like areas appeared blue and ghost cells appeared red which further confirmed the nature of the ghost cells and dentinoid like areas (fig. 3C).

Figure 1: Intraoral picture showing swelling on left side of mandible obliterating the labial vestibule.

Figure 2: Computed tomography scan (A) axial view showing mixed radiopaque and radiolucent lesion with ill defined border (B) 3D reconstruction showing lesion causing expansion and perforation of the labial cortical plate.

Figure 3: Photomicrograph (A) showing ghost cells within odontogenic epithelium. (B) Aggregates of ghost cells embedded within the dentinoid like areas (HE stain, x400). (C) Numerous ghost cells (red) in irregular islands of tumor epithelium and Dentinoid (blue) formed in stroma (Masson trichrome stain, x400).
Although the microscopic description of DGCT is very obvious, it can be confused with ameloblastoma, COC and Ghost cell Odontogenic Carcinoma (GCOC). A diagnosis of a COC was excluded in the present case because of the very high amount of histologically confirmed dentinoid which accounted for the solid structure of a DGCT, in contrast to a cystic structure of a COC. The presence of dysplastic dentin and a large number of ghost cells seen in the present case differentiated it histologically from ameloblastoma. The absence of mitotic activity ruled out the diagnosis of GCOC.

Based on the histopathological features, findings of histochemistry, the final diagnosis of Intraosseous DGCT was given. The patient was treated with segmental resection followed by reconstruction.

**DISCUSSION**

Calcifying Odontogenic Cyst (COC) constitutes 1 to 2% of all Odontogenic tumors in which 88.5% are cystic and the remaining 11.5% are solid tumors. As all the lesions are not cystic, it is still controversial whether COC is a cyst or a neoplasm. Based on this dualistic concept, in 2005 WHO histological classification, all the cystic lesions were termed as Calcifying Cystic Odontogenic tumor (CCOT) and neoplastic entity as DGCT. In 2017, WHO histological classification of head and neck tumor again reclassified CCOT as COC.

DGCT is the rarest of the ghost cell lesions accounting for less than 3% of all the ghost cell lesions. The tumor is twice more common in men than in women. The reported patient age range is 11-79 years with a mean of 39.7 years. The frequently affected sites for intraosseous DGCT are posterior maxilla and mandible with a slight predilection for mandible. Patients are usually asymptomatic, although with a few complain of pain or discomfort. The peripheral lesion seems to be less aggressive than its central counterpart with no recurrences reported after excision. Radiographically, DGCT appears as a radiolucent, radiopaque or mixed lesion depending on the amount of calcification. Lesions can be unilocular or multilocular with either well defined or ill demarcated margins. In the present case lesion was ill defined multilocular lesion with mixed radiodensity (fig. 2A). Root resorption, displacement of adjacent teeth and presence of impacted teeth have also been reported.

The central DGCT are likely to arise from the proliferation of remnants of the dental lamina. The tumor is mainly composed of ameloblastoma like areas of odontogenic epithelial islands with varying numbers of ghost cells showing keratinization and calcification which was also seen in the present case. Ghost cells are thought to be transformed odontogenic epithelial cells, the mechanism of whose transformation remains unknown. In the light microscope, they appear as enlarged, ovoid, or elliptoid epithelial cells, which are eosinophilic, usually with well defined cell outlines but may be blurred giving them a fused appearance. Sometimes ghost cells may contain nuclear remnants in various stages of degeneration. Histochemically, they are positive for keratin giving a yellow fluorescence with Rhodamine B.

The formation of dentinoid or osteoid material, which is frequently described as being found in connection with masses of ghost cells, is another characteristic finding of the lesion. Dentinoid like areas appear blue and ghost cells appear red in Masson trichrome. Similar finding was present in our case. Van Gieson stains are also frequently used for identifying ghost cells in which ghost cells appear yellow and dentinoid like areas appear pink to red color. Treatment of the intraosseous DGCT requires wide marginal resection. Recurrence potential of intraosseous lesions appears to be similar to that for conventional ameloblastoma.

**CONCLUSION**

DGCT is a rare odontogenic tumor with distinctive histological features and aggressive biological behavior. Differentiating this lesion from other odontogenic lesions histologically is important for the appropriate management.

**Conflict of Interest:** None

**REFERENCES**


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