Case Report

Peripheral Dentinogenic Ghost Cell Tumor of the Gingiva

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Background: *A dentinogenic ghost cell tumor is a locally invasive neoplasm that is characterized by ameloblastoma-like islands of epithelial cells in a mature connective tissue stroma.*

Methods: *A 43-year-old male patient presented a well-circumscribed sessile, exophytic mass of the gingiva with a diameter of 2 cm located in the canine area of the right maxilla. The lesion was enucleated.*

Results: *The lesion showed odontogenic epithelium, ghost cells, dentinoid material, and giant cells. The final microscopic diagnosis was a dentinogenic ghost cell tumor.*

Conclusions: *A dentinogenic ghost cell tumor is an extremely rare tumor, and only a few cases have been reported in the English literature. The peripheral, extraosseous lesion can be easily confused with other gingival lesions such as reactive or inflammatory lesions or other peripheral odontogenic tumors. The clinical appearance of all of these lesions is similar; therefore, the definitive diagnosis depends on histology, and a biopsy with a microscopic examination is mandatory.* J Periodontol 2007;78:1635-1638.

KEY WORDS

Calcifying odontogenic cyst; odontogenic tumors.

A dentinogenic ghost cell tumor (DGCT) is a locally invasive neoplasm that is characterized by ameloblastoma-like islands of epithelial cells in a mature connective tissue stroma.[1] An aberrant keratinization is present in the form of ghost cells and varying amounts of dysplastic dentin.[1,2] In previous classifications of odontogenic tumors, DGCT was considered a solid variant of calcifying odontogenic cyst.[2-9] DGCT occurs mainly as an intraosseous lesion and less commonly as an extraosseous mass.[2,10-12] The extraosseous variant has a predilection for the anterior portion of the jaws,[1] and it appears as a sessile, sometimes pedunculated, exophytic nodule located in the gingival or alveolar mucosa.[1,10-12]

DGCT is an extremely rare tumor, and only a few cases have been reported in the English literature. Extraosseous, peripheral variants of DGCT can be easily confused with other gingival lesions, such as other peripheral odontogenic tumors.[13-19] Moreover, because all of these lesions are rare, a histopathological assessment is necessary for an accurate diagnosis.

The aim of the present case report was a clinical, histologic, and immunohistochemical evaluation of an extraosseous DGCT of the gingiva.

CASE REPORT

In February 2000, a 43-year-old male was referred to the Outpatient Department of the Dental School of the University of Chieti-Pescara for a well-circumscribed, sessile, exophytic mass of the gingiva with a diameter of 2 cm located in the canine area of the right maxilla (Fig. 1). A residual root of the right first premolar was present. The first clinical impression was of a radicular cyst; however, no bone lesions were present on the periapical radiograph (Fig. 2). The lesion was covered by normal, healthy mucosa and was soft to palpation and painless. The patient underwent an excisional biopsy with a complete enucleation of the lesion. Histologically, most of the lesion was composed of ghost cells and an odontogenic ameloblastomatous epithelium (Fig. 3); moreover, there was continuity between the basal layer of the mucosal epithelium and islands of odontogenic epithelium, dysplastic dentin, and

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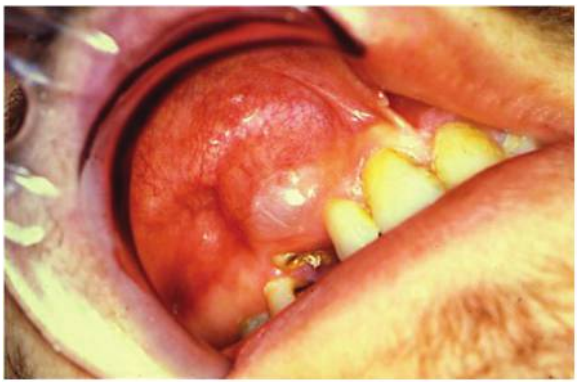

Figure 1.
Clinical appearance of the lesion at diagnosis.

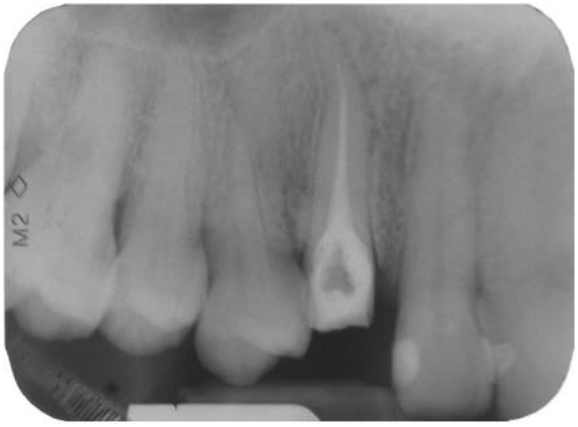

Figure 2.
Periapical radiograph shows the absence of bone lesions.

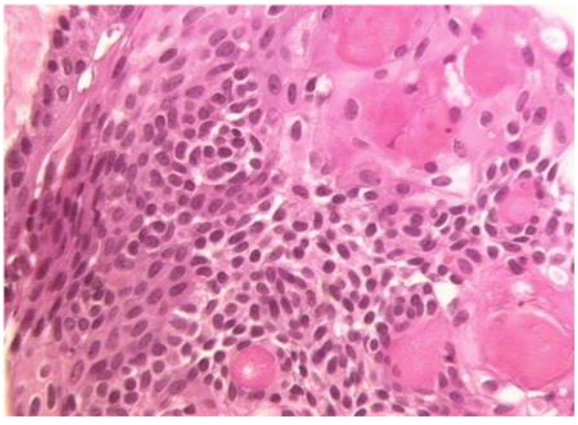

Figure 3.
Nests of odontogenic epithelium containing ghost cells in different stages of development (hematoxylin and eosin; original magnification ×400).

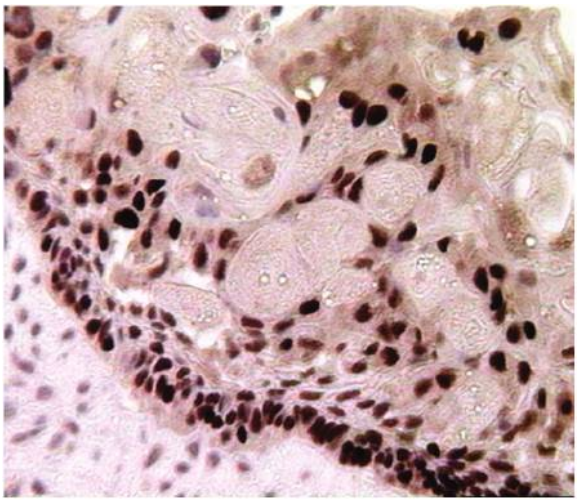

Figure 4.
Strong positivity of the cells of the odontogenic epithelium for MIB-1, whereas the ghost cells were completely negative (original magnification ×400).

ghost cells. The epithelium was formed by peripheral columnar or cuboidal cells and by central cells similar to those of the stellate reticulum. Nests of odontogenic epithelium and dysplastic dentin were present in the connective tissue of the gingiva. It was possible to observe islands of odontogenic epithelium trapped inside the dysplastic dentin and nests of ghost cells with odontogenic epithelium. Mitoses were absent. Immunohistochemistry for cytokeratins AE1/AE3 showed a strong but non-homogeneous positivity of the odontogenic epithelium, whereas the ghost cells were always strongly positive, immunohistochemistry for B-cell leukemia/lymphoma 2 (Bcl-2) showed a strong positivity of the odontogenic epithelium, and immunohistochemistry for p53 showed the presence of positive cells in the odontogenic epithelium and the dentinoid material only rarely. Immunohistochemical staining for a monoclonal antibody against the recombinant part of the Ki-67 antigen (MIB-1) showed a strong nuclear positivity of the cells of the odontogenic epithelium (Fig. 4). Ghost cells, giant cells, and dentinoid material were completely negative for MIB-1, Bcl-2, and p53. The definitive diagnosis was extraosseous DGCT. No recurrence was present after a 6-year follow-up.

DISCUSSION

There has been controversy about the nomenclature of DGCT.[20-31] This lesion has been termed an odontogenic ghost cell tumor, calcifying ghost cell odontogenic tumor, odontogenic ghost cell tumor, epithelial odontogenic ghost cell tumor, and dentinoameloblastoma.[20-31] The term "odontogenic ghost cell carcinoma" should be used to designate a malignant

tumor with the potential to metastasize,[24,26] and "odontogenic ghost cell ameloblastoma" should be used for the rare cases in which there is enamel deposition as well as dentin.[8] Gunhan et al.[21] preferred to use "epithelial odontogenic ghost cell tumor" because the epithelial odontogenic ghost cells were the main component of the tumor. Scott and Wood[22] described a case with an ameloblastomatous proliferation and masses of poorly differentiated or basaloid cells. They suggested that these elements might point to a neoplasm, which is a subgroup of ameloblastoma rather than a variant of calcifying odontogenic cyst (COC), and they suggested the term "dentinogenic ghost cell ameloblastoma." Malignant transformation into an odontogenic ghost cell carcinoma has been described.[24-26,31] Colmenero et al.[28] reviewed the literature and found that the neoplastic form of COC could be divided into two subgroups: 1) presence of an infiltrative pattern with invasion of stroma, mucosa, and bone and a high rate of local recurrences; and 2) presence of an infiltrative mass with cytological features of malignant transformation, a more aggressive local behavior, and, occasionally, distant metastases. In a histologic and immunohistochemical evaluation of a case of DGCT, there was a strong positivity of the cells of the odontogenic epithelium for Bcl-2 and MIB-1, whereas only a rare positivity for p53 was observed.[32] The ghost cells, giant cells, and dentinoid material were completely negative.[32] It was concluded that the cells which expressed Bcl-2 and MIB-1 probably represented the portion of the tumor that proliferated and that could undergo malignant transformation.[32] Some peripheral DGCTs are located entirely within the lamina propria, whereas others are in continuity with the gingival surface stratified squamous epithelium.[2] Peripheral tumors either show no radiographic alterations, as in the present case, or a saucerization of the cortical bone (in ~20% of cases).[1,2] Generally, peripheral tumors are 1.5 to 2.0 cm in size.[2] They present as sessile or pedunculated exophytic nodules of the gingiva;[2] they may be hard or soft and friable.[2] Two features distinguish DGCTs from ameloblastoma and other odontogenic tumors: ghost cells and dentinoid material.[2,33,34] Ghost cells are needed for the diagnosis of DGCT, but they are neither unique to nor pathognomonic of these lesions.[2,34] Ghost cells also are found in pilomatricoma and craniopharyngioma and may be present in some odontomas and ameloblastic fibro-odontomas.[2]

Peripheral odontogenic tumors are reported only rarely in the literature.[13,34] Ide et al.[15] reported 30 peripheral odontogenic tumors out of 39,660 specimens evaluated. The most frequently reported are peripheral odontogenic fibroma, peripheral ameloblastoma, peripheral calcifying epithelial odontogenic tumor, peripheral ameloblastic fibroma, peripheral squamous odontogenic tumor, and peripheral odontoma.[13] These tumors are located in the gingival or alveolar mucosa, and they present as a swollen area or an exophytic mass covered by healthy, normal-appearing mucosa.[13,34] Typically, the clinical preoperative diagnosis is that of a reactive or inflammatory lesion (peripheral giant cell granuloma, pyogenic granuloma, epulis, gingivitis, parulis, or irritation fibroma).[13] The clinical appearance of all of these lesions is similar; therefore, the definitive diagnosis requires histology, and a biopsy with microscopic examination is mandatory.[13]

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