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CASE REPORT

Odontogenic ghost cell carcinoma: A rarity encountered

Dominic Augustine[1], Maya Ramesh[2], S. Murali[2], B. Sekar[2], George Paul[3]

[1]Department of Oral and Maxillofacial Pathology, Faculty of Dental Sciences, MS Ramaiah University of Applied Sciences, Bengaluru, Karnataka, India, [2]Department of Oral and Maxillofacial Pathology, Vinayaka Missions Sankarachariyar Dental College & Hospital, Ariyanoor, Salem, Tamil Nadu, India, [3]Oral and Maxillofacial Surgeon, Paulose Dental Clinic, Fairlands, Salem, Tamilnadu, India

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Correspondence
Dr. Dominic Augustine, Department of Oral and Maxillofacial Pathology, Faculty of Dental Sciences, MS Ramaiah University of Applied Sciences, Bengaluru - 560 054, Karnataka, India. Phone: +91-9886226214, Email: dominic2germain@gmail.com

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Abstract

Odontogenic ghost cell carcinoma (OGCC) is a rare neoplastic variant of calcifying odontogenic cyst, with aggressive clinical characteristics. A swelling in the jaws along with irregular destruction of the adjacent bone and local paresthesia are common symptoms. Microscopically, islands of varying size and anucleate cell clusters with homogenous, pale eosinophilic cytoplasm called ghost cells, admixed with sheets of tumor is seen. We present a rare case of OGCC occurring in the mandible of a 70-year-old male. This report carries a new message since prognosis is poor for OGCC but in the present case there was no evidence of recurrence after a 2-year follow-up.

Introduction

The calcifying odontogenic cyst (COC), classified as an odontogenic tumor by the World Health Organization, was first identified as an entity by Gorlin *et al*. in 1962.[1]

In 1981, Praetorius *et al*.[2] identified four different histological patterns of COC and classified them as Type 1A (simple unicystic), Type 1B (odontome-producing), Type 1C (ameloblastomatous proliferating), and Type 2 (dentinogenic ghost cell tumor). In their opinion, Type 2 shares many of the histological features of the cystic variants; however, the solid growth pattern of the Type 2 suggests that its classification as a neoplasm is more appropriate.

Ellis and Shmookler[2] used the term epithelial odontogenic ghost cell tumor (EOGCT) for the neoplastic variant of COC. Although the terminology is inconsistent, ghost cells are clearly the most distinctive histological feature of this tumor.

The first-documented case of a malignancy arising in a COC to appear in the English language literature was reported by Ikemura *et al*.[3] in 1985. Until now, only 26 odontogenic ghost cell carcinomas (OGCCs) have been reported in the English-language literature.[4]

Gender distribution of all the reported lesions of OGCC showed a male predilection of 3.4:1. This is different from its benign counterpart, which has been reported as equally common in both men and women.[4]

The paucity of cases makes the understanding of these peculiar lesions difficult and makes this case necessary to document.

Case Report

A 70-year-old male visited the dental clinic with a chief complaint of a swelling in the right posterior mandible since 3 months. His medical history was noncontributory. No lymph nodes were palpable. Intraoral examination revealed a large ulcerated lesion [Figure 1] in the right posterior mandible causing expansion of the buccal and lingual cortical plates. The ulcer was present on the edentulous alveolus extending from the right mandibular first premolar to the ramus area. The lesion was firm to palpate, nontender, immobile, and no surface discharge was observed.

Orthopantomogram revealed a large lytic radiolucent lesion involving the right posterior mandible measuring 3 cm × 3 cm causing erosion of the cortical bone. Approximation of the mandibular canal was seen. Teeth numbers 45-48 were missing [Figure 2].

A provisional diagnosis of intra-alveolar malignancy was arrived at.

An incisional biopsy was done and on microscopic examination at ×10 low power view large sheets of atypical cells

in the form of islands invading the connective tissue was seen [Figure 3a]. The islands had peripheral palisading of the basal cells with areas of stellate reticulum and cystic degeneration resembling odontogenic islands [Figure 3b].

At higher magnification, the center of the islands had large pale eosinophilic cells with keratinization called ghost cells [Figure 4a]. The atypical epithelial cells showed features of pleomorphism, hyperchromatism, vesicular nuclei, altered nuclear cytoplasmic ratio, and mitotic along with apoptotic bodies [Figure 4b].

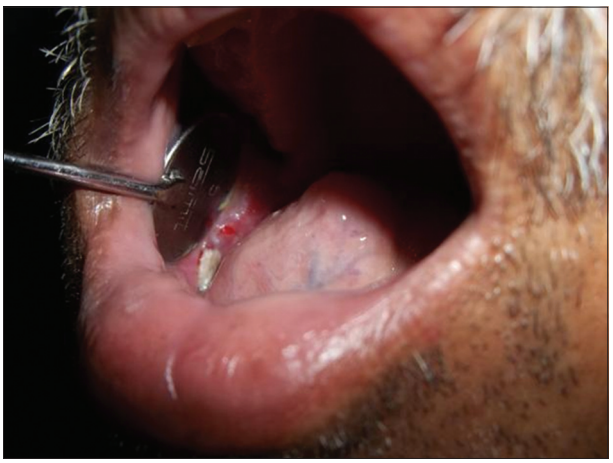

Figure 1: An ulcerated lesion of the right posterior mandible in a 70-year-old male

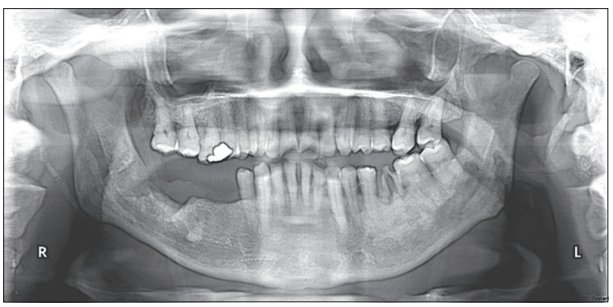

Figure 2: Orthopantomogram showing a large lytic lesion involving the right posterior mandible with erosion of the cortex

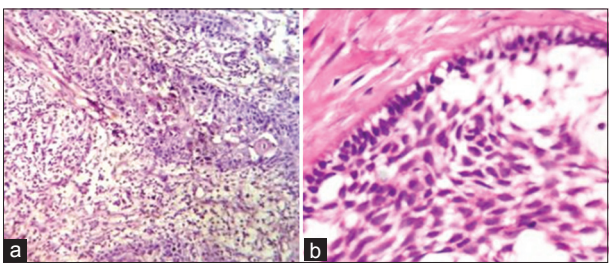

Figure 3: (a) Histopathologic section showing large sheets of atypical cells in the form of islands invading the connective tissue (H and E, ×100). (b) Section shows peripheral palisading of the basal cells with areas of stellate reticulum and cystic degeneration resembling odontogenic islands (H and E, ×400)

A final diagnosis of OGCC was concluded.

A hemimandibulectomy was performed, and the patient was referred for adjuvant radiotherapy. Patient is under regular follow-up for 2 years and has shown no evidence of recurrent disease [Figure 5].

Discussion

OGCC is a rare malignant odontogenic tumor with features of COC. Clinically, it could occur as a de novo tumor or from a long-standing COC.

It is of interest that previous cases of OGCC are often described as multiple recurrences of COC or long-term persistent swelling followed by the onset of rapid, painful swelling before a definitive diagnosis of OGCC. This might be perceived as evidence of a transformation of a long-standing or recurrent benign process into a malignant one. In the present case, the tumor seems to have developed *de novo*.[5]

In a study conducted by Cheng *et al.*,[6] of the 22 OGCC reported, 15 (68.2%) were in the maxilla and 7 (31.8%) in

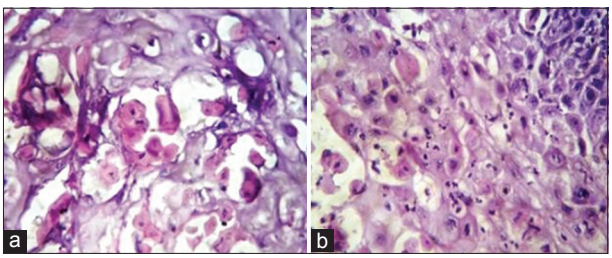

Figure 4: (a) Histopathologic section showing large pale eosinophilic cells with keratinization called ghost cells (H and E, ×400). (b) The atypical epithelial cells showing features of pleomorphism, hyperchromatism, vesicular nuclei, altered nuclear cytoplasmic ratio, apoptotic bodies, and mitotic figures (H and E, ×400)

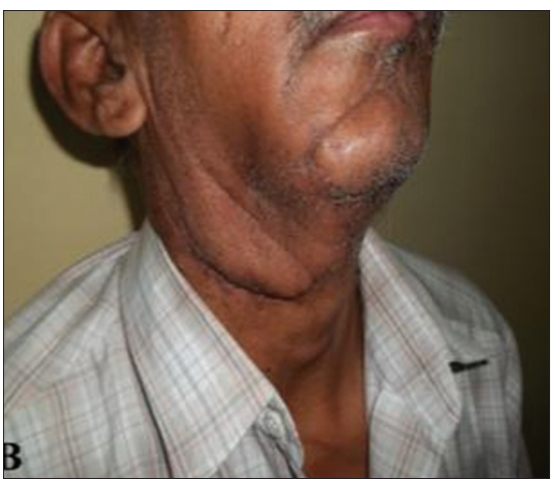

Figure 5: Postoperative picture of the patient

the mandible. There was a clear predilection of OGCC for the maxilla (ratio 2.1:1), differing from the site distribution of the benign COC, which occurs equally in both the maxilla and mandible. In the maxilla, OGCC was not restricted to only one region such as incisor–canine, premolar and molar region, but rather involved an extensive region. Of the 13 cases in the maxilla where anatomical location is provided, eight were located in the incisor–canine, premolar and molar region; three in incisor–canine, premolar region; and two in premolar–molar region. Twelve cases showed destruction of the maxillary sinus with obliteration. The lesions in the maxilla did not cross the midline. On the other hand, in the mandible, of the six cases where anatomic location was provided, three were restricted to the molar region and three crossed the midline but were located between bilateral premolar regions of mandible. Our case involved the posterior mandible of a male patient.

The OGCC usually appears as a painful swelling in the mandible or maxilla with bony destruction and with paresthesia being a frequent finding.

It may cause expansion of the mandible or maxilla. In our case, pain was not a major symptom but osseous destruction was present.

OGCC occur predominantly in middle-aged and young adults, usually within an age range of 13-72 years.[7] It is difficult to make a diagnosis of OGCC based on radiographic features alone. OGCC shows radiographic features of a malignant tumor and is not specific.

Other possible diagnoses of malignant tumors include osteosarcoma and malignant ameloblastoma. A diagnosis of OGCC was possible only after the resected specimen was examined histologically.[8]

The histological diagnostic criteria of OGCC are an epithelial lining showing a basal layer of columnar cells, a layer of cells resembling the stellate reticulum of the enamel organ, and masses of ghost cells that may be calcified or not, accompanied by atypical epithelial cell foci presenting mitosis, keratin pearls, necrosis and other malignant features.[9] All these features were observable in the present case.

Recurrence is common after initial operation. Therefore, OGCC shows both clinical and radiographic features of a malignant tumor with high recurrence. No recurrence has been observed in our patient after a 2-year follow-up.

Conclusion

The neoplastic variant of COC has various designations, and its malignant counterpart has been reported as aggressive EOGCT or OGCC. We have presented a very rare case of OGCC that has shown good prognosis. This necessitates the documentation of this case.

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