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Carta al editor

Odontogenic ghost cell carcinoma in the maxilla: A case report

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ABSTRACT

Introduction: Odontogenic Ghost Cell Carcinoma (occc) is an extremely rare malignant epithelial tumour. So far there is little understanding of its aetiology and behaviour. The treatment basis is complete excision of the lesion, with continued adjuvant radiotherapy or chemotherapy, which is still controversial. **Objective:** To increase insight into the possible differential diagnosis of maxillary tumours. Even though its incidence is low, it is essential to become familiar with the characteristics of odontogenic ghost cells. **Case presentation:** A 29-year-old woman presented with a progressively growing mass. The CT scan showed an expansive lesion with destruction of the ascending branch of the right mandible. The lesion was excised with pathological analysis compatible with Ghost Cell Carcinoma. The tumour was positively diagnosed as an odontogenic

ghost cell carcinoma. **Conclusion:** The OGCC represents a rare malignant tumour and must be considered in oral expansive enlargements, especially in the maxillary bones.

Keywords: Odontogenic Ghost Cell Carcinoma, Maxilla, cancer.

INTRODUCTION

Odontogenic Ghost Cell Carcinoma (OGCC) represents a rare malignant epithelial tumour mostly occurring in the maxilla[1,2]; its aetiology, physiopathology, and behaviour are poorly understood. Its diagnosis is possible only with pathologic examination, and the imagenologic appearance is different in every case reported[2]. Wide surgical excision represents the main treatment, thus avoiding the recurrences observed in these tumours, and because there is unclear effectiveness in adjuvant chemotherapy or radiotherapy.

CLINICAL CASE PRESENTATION

In July 2018, a healthy 29-year-old woman attended the Oncologic Centre with a history of mandibular pain on eating and a progressing mass in the right maxilla. The physical examination revealed right preauricular mass, trismus, and lower lip paraesthesia.

The Computed tomography (CT) multiplanar reformation and three-dimensional image reconstruction show an expansive bone tumour with soft tissue component, with an average density of 53 Hounsfield units (HU), homogeneous evidence after the administration of intravenous contrast, loss of the fatty plane of the adjacent elements to the pterygoid, the masseter muscles and the anterior border of the parotid gland infiltration. The tumour destroys the ascending ramus, the mandibular condyle, and the mandibular notch (Figure 1). There was no clinical or radiologic evidence of cervical adenopathy. Histological examination (Figure 2. A-F) showed sheets and nests of odontogenic epithelium containing numerous ghost cells showing focal calcification. Based on these findings, the tumour was diagnosed as an odontogenic ghost cell carcinoma. The patient underwent a total excision and immediate mandibular intraoperative reconstruction. Postoperative period was uneventful, and no adjunctive radiotherapy was recommended.

DISCUSSION

Most odontogenic tumours (OT) are benign ang malignancies are exceedingly rare. The OGCC is considered a malignant variant of the calcifying odontogenic tumour (COT)[1,2] described for the first time in 1985 by Ikemura *et al*.,[3] and is also called a malignant COT, aggressive epithelial ghost cell odontogenic tumour, and dentinogenic ghost cell ameloblastoma.

Etiology remains unclear, some are diagnosed after multiple recurrences of COT[4], while others develop *de novo*. In the present case, the tumour seems to have developed *de novo*. Less than 30 cases have been reported in medical literature. Most tumours are present in females, Asians and young adults[5]. After a brief comparison with the literature, age and sex

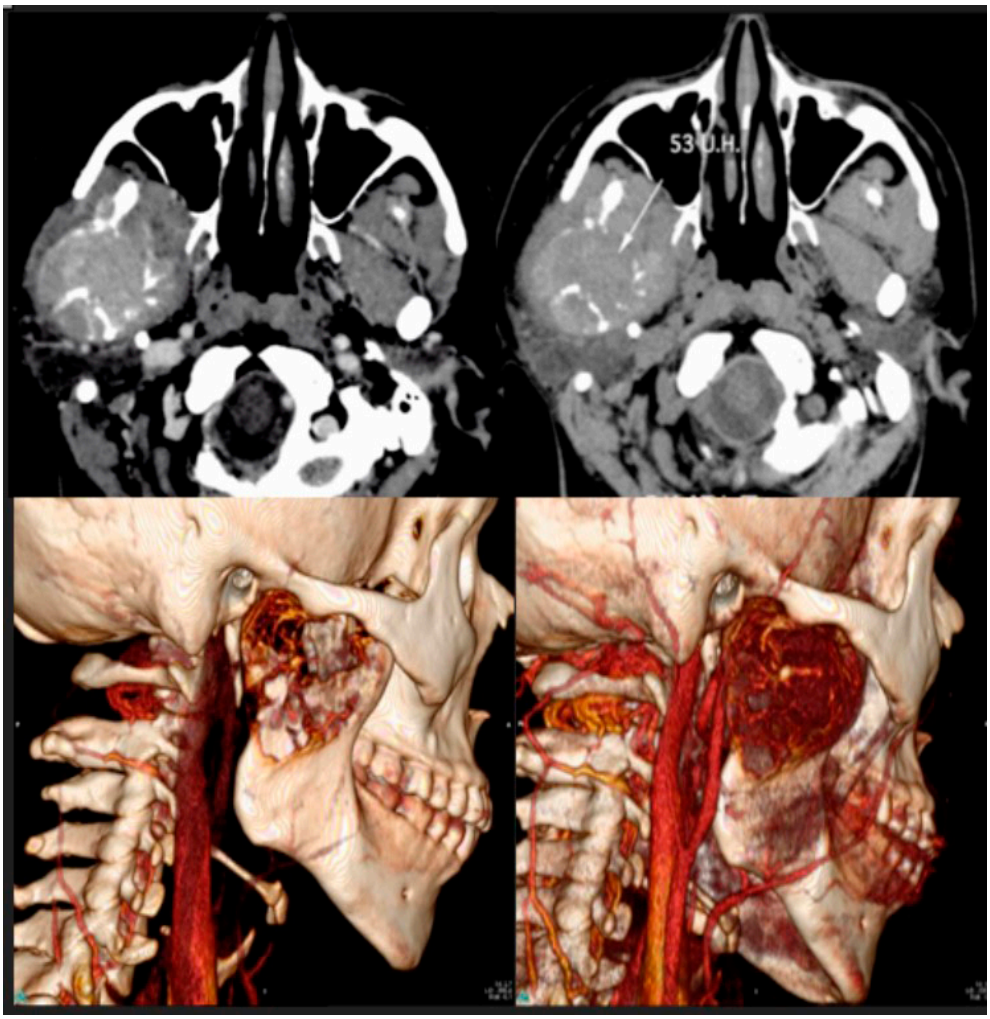

Figure 1. Tomographic images of Odontogenic Ghost Cell Carcinoma in the Maxilla. *Upper*: axial section. *Lower*: 3D reconstruction.

were compatible with our patient. Seventy-two percent of the cases reported in the literature involved the maxilla and the most common site is the posterior maxilla[6], but in this case the tumour was located in the right mandibula.

The clinical features of OGCC are unspecific. A painful swelling of the jaws with local paraesthesia is the most frequent symptom, and expansion of the mandible or maxilla may be noted, as in this clinical case. The OGCC presents a spectrum of growth patterns, slowly growing locally to an aggressive tumour, to an extremely aggressive and rapidly growing neoplasm[7]. Distant metastases are uncommon[7]. Radiographically, OGCCs have been described as purely radiolucent or mixed radiolucent-radiopaque lesions. Conventional radiographs show a large, poorly defined osteolytic lesion of the mandible with several foci of increased radiopacity within it.

Histologically, OGCC is 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, and proliferating odontogenic epithelium with characteristic small groups or large masses of "ghost cells." These ghost cells are pale eosinophilic, plump, polygonal, keratinized epithelial cells that have lost their nuclei.

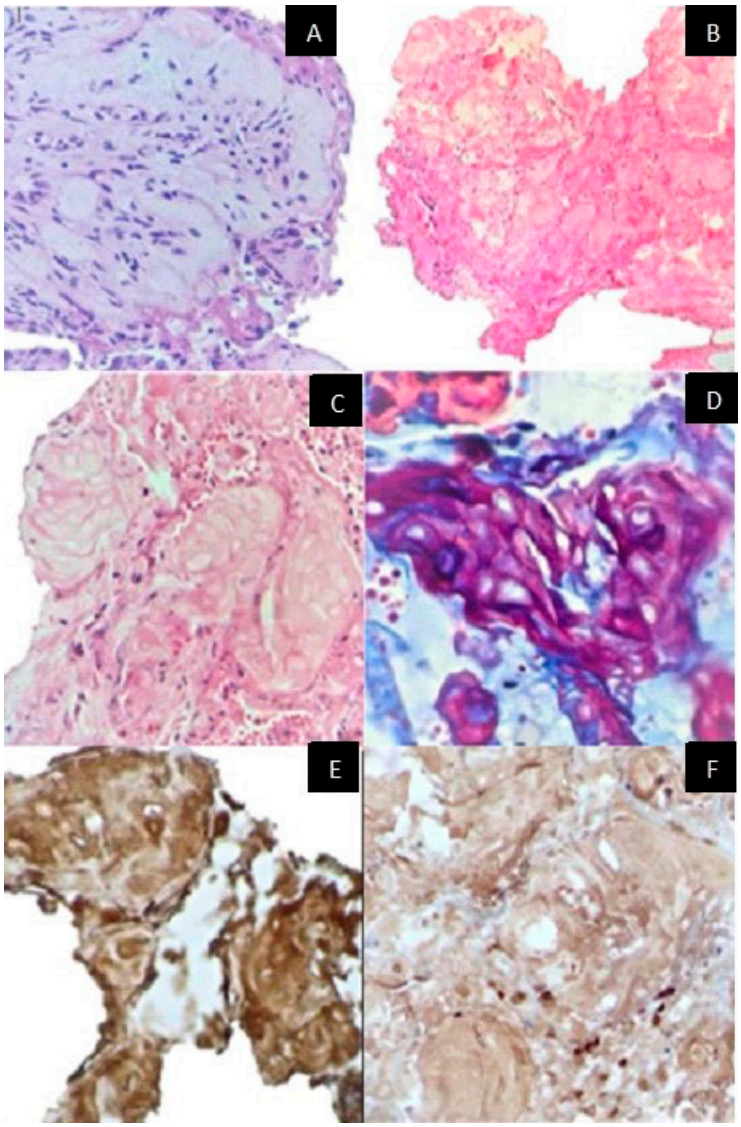

Figure 2. Histopathological images. A. Odontogenic neoplasia with digitiform projections without nuclear atypia. B-C. Ghost cells: neoplastic cells lacking nuclei with abundant eosinophilic cytoplasm. D. Ghost cells Masson stain. E. Positive cytokeratin AE1-AE3. F. Positive epithelial membrane antigen (EMA).

They contain a distinct intracytoplasmic keratin that preserves the outline of the cell and the corresponding previous site of the nuclei[6]. The immunohistochemical findings are positive for epithelial markers, such as cytokeratin 5/14 and Ki-67, and sometimes are positive for epithelial membrane antigen, neuron- specific enolase and p53[6]. The differential diagnosis includes craniopharyngioma, odontoma, pilomatricoma and ameloblastic fibroodontoma.

The recommended treatment for OGCC has been wide surgical excision. For the best surgical plan, the imagenologic evaluation must suffice for an adequate evaluation of the tumoral extension. The 3D CT reconstruction remains as an important instrument to avoid a second surgical intervention for positive tumoral margins[2], suggesting that all patients must be evaluated with this equipment. The use of postoperative radiation therapy with or without adjuvant chemotherapy is controversial[7].

Because of the unpredictable biologic behaviour of this type of tumour, careful, long-term follow-up is highly recommended after therapy. The overall 5-year survival rate of the first 16 reported cases is 73%[8].

CONCLUSION

The OGCC represents a rare malignant tumour and must be considered when dealing with oral expansive masses, especially in maxillary bones.

CONFLICT OF INTEREST.

The authors declare no conflict of interest.

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