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Autores / Authors:

jean della giustina, Peterson Bilhar, Hildebrando Massahiro Nagai, Juliana Lucena Schussel

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ODONTOGENIC GHOST CELL TUMOR: CLINICAL MANAGEMENT OF A RARE MAXILLARY NEOPLASM

TUMOR DE CÉLULAS FANTASMA ODONTOGÉNICO: TRATAMIENTO CLÍNICO DE UNA NEOPLASIA MAXILAR POCO FRECUENTE

Jean Carlos Della Giustina¹, Peterson Billhar², Hildebrando Nagai² y Juliana Lucena Schussel³

¹Oral and Maxillofacial Surgeon. Department of Oral & Maxillofacial Surgery, Hospital Uopecan (Cascavel Cancer Hospital), Cascavel, Brazil. ²Head and Neck Surgeon. Department Head and Neck Surgery, Hospital Uopecan (Cascavel Cancer Hospital), Cascavel, Brazil. ³Professor & Oral and Maxillofacial Pathologist. Department of Stomatology, Federal University of Paraná (UFPR), Curitiba, Brazil

CORRESPONDENCE:

Jean Carlos Della Giustina
jcdgiustina@yahoo.com.br

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ABSTRACT

The odontogenic ghost cell tumor is an extremely rare tumor, representing less than 0.4 % of odontogenic tumors, and is categorized into two forms: extraosseous (peripheral) and intraosseous (central). The central variant exhibits aggressive and infiltrative behavior, affecting the alveolar regions of the maxilla and mandible, causing swelling and possessing a high recurrence potential. This article presents a clinical case of a 30-year-old woman with a large tumor in the maxilla causing facial deformity, treated with maxillectomy and microsurgery. The final diagnosis was established after clinical, radiographic, histopathological, and immunohistochemical evaluations.

Keywords: Odontogenic tumor, odontogenic ghost cell tumor, microsurgery.

RESUMEN

El tumor odontogénico de células fantasma es un tumor extremadamente raro, que representa menos del 0,4 % de los tumores odontogénicos, y se clasifica en dos formas: extraóseo (periférico) e intraóseo (central). La variante central presenta un comportamiento agresivo e infiltrativo, afecta a las regiones alveolares del maxilar y la mandíbula, provoca inflamación y tiene un alto potencial de recurrencia. Este artículo presenta el caso clínico de una mujer de 30 años con un tumor grande en el maxilar que le provocaba deformidad facial, tratado con maxilectomía y microcirugía. El diagnóstico definitivo se estableció tras evaluaciones clínicas, radiográficas, histopatológicas e inmunohistoquímicas.

Palabras clave: Tumor odontogénico, tumor odontogénico de células fantasmas, microcirugía.

INTRODUCTION

The odontogenic ghost cell tumor (OGCT) is a rare benign neoplasm, accounting for less than 0.4 % of all odontogenic tumors. According to the 5th edition of the WHO Classification of Odontogenic Cysts and Tumors (2022), OGCT is defined as a benign mixed epithelial and mesenchymal tumor that is locally aggressive. Histologically, it is characterized by strands and islands of epithelial cells resembling ameloblastoma infiltrating mature connective tissue. These cells exhibit keratinization in the form of ghost cells, some of which undergo calcification, along with varying degrees of dysplastic dentin production¹.

OGCT may present locally as a central (intraosseous) variant, which displays aggressive and infiltrative behavior and carries a high recurrence rate following resection. In contrast, the peripheral variant tends to exhibit milder and less aggressive behavior².

The current literature includes limited reports on this pathology. To date, only 57 cases (39 central and 18 peripheral) have been documented and classified under the 2017 WHO criteria for odontogenic tumors³. This rarity is further emphasized by other studies; a PubMed search for OGCT reports featuring imaging findings- restricted to English-language publications from 2017 onward- identified only 15 reports comprising 16 cases⁴.

This study aims to present a clinical case involving a 30-year-old female patient diagnosed with this rare pathological entity, describing its clinical, radiological, histopathological, and immunohistochemical characteristics, along with the adopted treatment approach.

CASE REPORT

A 30-year-old female patient presented to the Oral and Maxillofacial Surgery Department at Uopeccan Hospital (Cascavel Cancer Hospital) with complaints of nasal obstruction, extraoral and intraoral swelling, and obliteration of the vestibular fold in the posterior left maxilla, with a six-month evolution (Figure 1). Imaging studies revealed a well-defined radiolucent lesion in the left maxilla, extending from teeth 22 to 27, associated with root resorption and invasion of the maxillary sinus (Figure 2). Exploratory aspiration yielded negative results, and the patient subsequently underwent an intraosseous incisional biopsy.

Histopathological analysis revealed epithelial islands and strands ameloblastoma-like embedded in mature connective tissue, Areas with keratinization forming ghost cells were observed, characterized by their eosinophilic appearance, absence of nuclei, and distinct outlines (Figure 3). Additionally, the tumor exhibited varying amounts of dysplastic dentin-like material. Immunohistochemical analysis demonstrated immunopositivity for β -catenin and calretinin, which, together with the histopathological features, confirmed the diagnosis of odontogenic ghost cell tumor (OGCT).

The patient was treated with a novel rehabilitative approach involving a left hemimaxillectomy, followed by reconstruction using a free iliac crest bone graft and a

microsurgical radial forearm flap to close the intraoral communication (Figure 4). She has been under follow-up for two years, with no signs of recurrence, and shows successful consolidation of the bone graft and vitality of the microvascularized flap.

DISCUSSION

The intraosseous OGCT exhibits locally invasive behavior and can occur across a broad age range (12-75 years), with a mean age of 40 years⁴. In contrast, extraosseous variants show limited growth potential and typically appear in the sixth decade of life, with an age range of 10-92 years. OGCT affects males more frequently, with similar rates of occurrence in both the mandible and maxilla⁵.

OGCT most commonly affect the canine-to-first molar region and usually present clinically as painless bony swelling, although some patients report mild numbness or discomfort⁶. In the present case, the patient exhibited significant yet asymptomatic intraoral and extraoral swelling, involving an extensive region (teeth 22-27) reflective of the tumor's aggressive and infiltrative nature.

Radiographic features of OGCT are variable and may present as radiolucent, radiopaque, or mixed lesions depending on the degree of calcification⁷. The lesion may be unilocular or multilocular, with well-defined or ill-defined margins. Adjacent teeth may show signs of displacement, root resorption, or impaction³. In this case, panoramic imaging revealed a well-defined, unilocular radiolucent lesion with root resorption but without evidence of impacted teeth or calcifications.

To confirm the diagnosis, immunohistochemistry testing was performed and revealed β -catenin positivity. This finding is consistent with the literature, which has shown that mutations in *CTNNB1*, the gene that encodes β -catenin, are involved in the formation of ghost cells and are commonly observed in OGCT⁸.

Due to its aggressive and infiltrative nature, intraosseous OGCT is associated with high recurrence rates- up to 71 %- particularly following conservative treatments such as enucleation or local excision. Even more extensive surgical interventions, including segmental mandibulectomy and partial maxillectomy, can be followed by recurrence within five years⁸. Malignant transformation into odontogenic ghost cell carcinoma has

also been reported⁹. Therefore, conservative approaches carry a have higher risk of recurrence, and aggressive surgical management is recommended to reduce the risk of relapse⁹.

The radial forearm flap, developed in 1978 by Dr. Yang Goufan, is a versatile reconstructive option for addressing large midface and palate defects. This chimeric flap can be designed as fasciocutaneous or osteocutaneous and is widely used for closing large oroantral communications, especially in patients undergoing maxillectomy of cleft palate corrections. The flap features a long vascular pedicle (~15 cm), which facilitates microvascular anastomosis with the cervical vessels, and its pliable, thin structure allows for excellent adaptation to the palatal region. In the present case, the radial forearm flap was chosen for reconstruction and provided excellent functional and aesthetic outcomes¹⁰.

Given the tumor's size and aggressive behavior, we opted for an extensive surgical approach via left hemimaxillectomy to minimize the risk of recurrence. To reduce postoperative sequelae, immediate reconstruction was performed using a mixed surgical technique that combined a free iliac crest bone graft with a microvascularized radial forearm flap. The radial flap was used to vascularize the bone graft and to reconstruct the intraoral soft tissue defect. To enhance the interface between the iliac crest graft and the soft tissue flap, miniplates were employed as anchoring points, allowing sutures between the plates and the microvascularized graft to generate superior traction. In our experience, this innovative approach proved effective in addressing extensive bone and soft tissue defects of the maxilla.

CONCLUSION

Intraosseous OGCT is a rare odontogenic neoplasm with aggressive and infiltrative behavior, and few cases have been reported in the literature. Early clinical, radiographic, and histopathological diagnosis is essential. Given its high recurrence potential, aggressive surgical intervention with safety margins of up to 1 cm is recommended to prevent recurrence and possible malignant transformation. Long-term clinical, radiographic, and histopathological follow-up is crucial. This case

highlights a novel therapeutic approach for maxillary intraosseous OGCT and may contribute to the development of standardized treatment protocols in the future.

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Figure 1. Clinical examination revealed a firm, rippled swelling extending from the left anterior maxillary region to the molar area, with obliteration of the vestibular sulcus.



Figure 2. Panoramic radiograph showing a relatively well-defined, homogeneous, round radiolucent lesion occupying the left maxillary sinus, extending from the root apices of teeth 22 to 27, with evident root resorption.

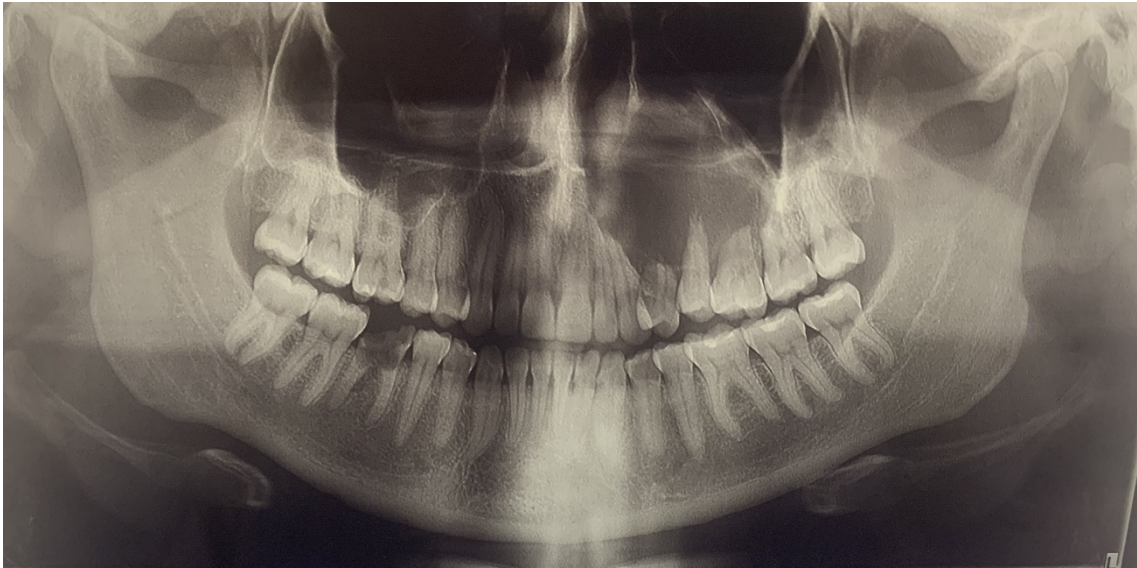


Figure 3. Histopathological and surgical findings. A: H.E. staining showing cystic and solid areas; the solid areas are composed of basaloid odontogenic cells separated by eosinophilic stroma; B: Eosinophilic matrix containing clear “phantom” areas and clusters of cells transiting into ghost cells; C: Surgical specimen - occlusal view; D: Surgical specimen - superior view.

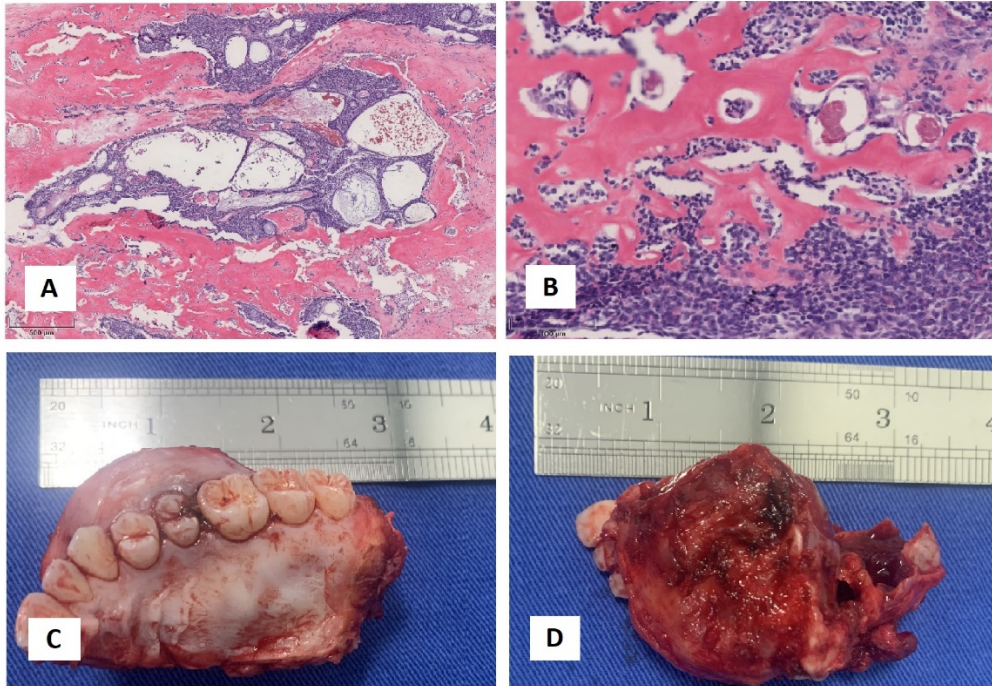


Figure 4. Reconstructive surgical procedure and follow-up. A: Harvested. Iliac crest graft used reconstruction of the left maxilla; B: Microsurgical radial forearm soft tissue flap with completed vascular anastomoses; C: Intraoral view of the radial forearm flap at 2-year follow-up, showing satisfactory integration; D: Panoramic radiograph at 2-year follow-up showing graft consolidation and absence of disease recurrence.

