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World
Journal of Clinical Cancer Research

ISSN 2831-8064

Case Report

Rare Maxillary Neoplasm With Odontogenic Differentiation And Malignant Behavior: A Case Report.

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Abstract

Ghost Cell Odontogenic Carcinoma (GCOC) is a rare malignant neoplasm of odontogenic epithelium, characterized by aggressive behavior and distinctive histological features, including ghost cells and infiltrative growth. This study reports the case of an 80-year-old male patient who presented with right facial swelling and nasal obstruction. Computed tomography revealed an expansive, heterogeneous mass with cystic areas and coarse calcifications, causing bone destruction in the right maxillary region. Histopathological analysis showed odontogenic epithelial tissue, ghost cells, inflammatory infiltrate, and dentin deposits. Immunohistochemical studies confirmed an epithelioid neoplasm with atypia, mitotic figures (6/10 HPF), and diffuse positivity for cytokeratin, supporting the diagnosis of GCOC. Radiotherapy was initiated but discontinued after six sessions due to clinical deterioration, culminating in the patient's death. This case underscores the diagnostic and therapeutic challenges of GCOC, reinforcing its aggressive nature, the role of immunohistochemistry in differentiating it from dentinogenic ghost cell tumor, and the importance of early diagnosis for improved prognosis.

Keywords : *Ghost cell odontogenic carcinoma, Ghost Cells, Odontogenic tumors.*

INTRODUCTION

Ghost cell odontogenic carcinoma (GCOC) is an uncommon malignant neoplasm of odontogenic epithelium, first described in 1985 (1). This lesion belongs to a group known as Ghost Cell Odontogenic Lesions, which are microscopically identified by the presence of ghost cells. In 2005, the World Health Organization classified these lesions into three categories: calcifying cystic odontogenic tumors with cystic morphology; dentinogenic ghost cell tumor (DGCT) with solid morphology; and GCOC, described as aggressive malignant neoplasms with metastatic potential (2,3). Radiographically, GCOC lacks pathognomonic features. The lesion may present as radiolucent, radiopaque, or mixed depending on the degree of calcification, and may appear unilocular or multilocular with either well-defined or poorly demarcated margins. Associated findings may include root resorption, displacement of adjacent teeth, and the presence of impacted teeth (4). According to the World Health Organization, GCOC typically exhibits histological features including prominent mitotic activity, nuclear atypia, cellular pleomorphism, clusters of ghost cells, necrosis, and demonstrates an infiltrative growth pattern with aggressive behavior (3). DGCT and GCOC represent a spectrum of ghost cell odontogenic neoplasms

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Received: 22-Sep-2025, Manuscript No. WJCCR-5146 **; Editor Assigned:** 24-Sep-2025 **; Reviewed:** 10-Oct-2025, QC No. WJCCR-5146 **; Published:** 27-Oct-2025, **DOI:** 10.52338/wjccr.2025.5146
Citation: Antonione Santos Bezerra Pinto. Rare maxillary neoplasm with odontogenic differentiation and malignant behavior: a case report. World Journal of Clinical Cancer Research. 2025 October; 13(1). doi: 10.52338/wjccr.2025.5146.
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that share numerous characteristics and require careful histopathological evaluation, considering their overlap with other odontogenic tumors and the potential for benign lesions to undergo malignant transformation (5). Immunohistochemical findings are valuable for distinguishing between benign and malignant tumors as well as for identifying aggressive behavior. Proliferation markers such as Ki-67 are commonly employed to support the malignant nature of the tumor (6). The primary recommended treatment for this lesion is surgical resection with clear margins (7). This report presents a case of GCOC, highlighting the diagnostic challenges, therapeutic approaches, and clinical implications of this rare lesion.

CASE REPORT

An 80-year-old male patient presented to the clinic complaining of right facial swelling and nasal obstruction. Clinical examination revealed a friable, non-ulcerated, non-bleeding, and painless lesion in the right maxillary region. Computed tomography (CT) scans of the face were performed in sagittal, axial, and coronal planes with 5 mm slice thickness and 5 mm intervals **(Figure 1)(Figure 2)**.

Figure 1. Computed tomography images. (A and B) Sagittal and coronal sections showing an expansive lesion invading the maxillary sinus on the right side of the maxilla. (C) Axial section demonstrating lesion expansion crossing the midline.

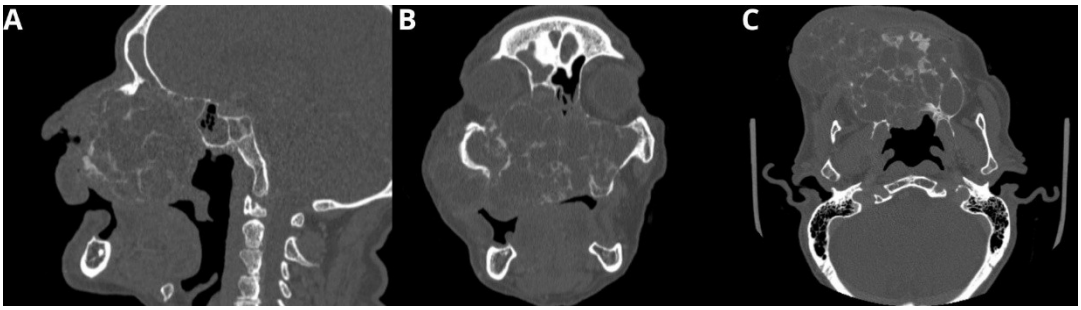
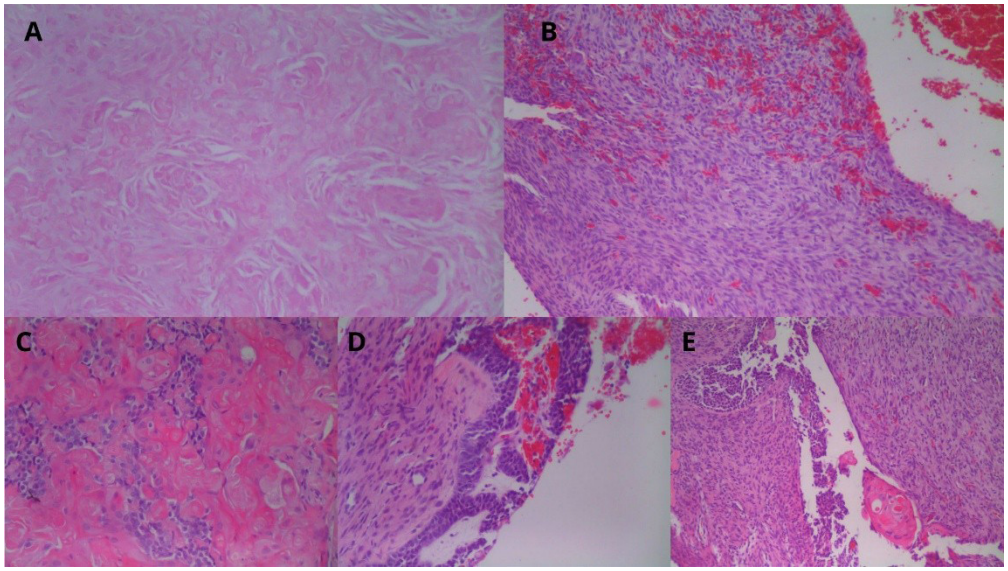


Figure 2. Three-dimensional reconstruction images of the lesion.



The imaging revealed an expansive, infiltrative-appearing, heterogeneous mass with lobulated contours and areas of soft tissue attenuation. Multiple cystic areas with internal septations and coarse calcifications were observed, centered in the right median/paramedian maxillary region. The lesion caused destruction of adjacent bony structures, making precise measurement difficult according to the study protocol. The lesion extended at least 11.3 × 6.2 × 6.4 cm (lateral-lateral × anteroposterior × craniocaudal). Initial diagnostic hypotheses included myxoma, ossifying fibroma, and ameloblastoma. Histological sections showed tissue fragments partially lined by odontogenic epithelial cells and occasionally squamous cells (**Figure 3**). Numerous eosinophilic ghost cells, mixed inflammatory infiltrate, and hypercellular fusiform stroma were observed, along with areas of dentin deposition. These histological findings were consistent with the differential diagnosis of DGCT with uncertain biological behavior, and immunohistochemical study was recommended for further evaluation.

Figure 3. Hematoxylin and eosin (H&E) stained histological image, 40x magnification. (A-E) Epithelial proliferation with ghost cells characterized by absent nuclei, eosinophilic cytoplasm, and preserved cellular outlines.



Immunohistochemical analysis revealed an epithelioid neoplasm with atypical cells arranged in solid patterns with mitotic figures (6/10 HPF) amidst numerous keratinizing foci and calcifications. The morphological findings, combined with diffuse positivity for cytokeratin, were suggestive of GCOC (**Table 1**).

Table 1. Immunohistochemistry results.

Antibody	Clone	Results
AE1/AE3 (CKPAN)	AE1/AE3	Positive
P63	4A4	Positive

Given the extensive stage of the lesion and the patient's advanced age, surgical treatment was ruled out, and radiotherapy was selected as the therapeutic approach. However, after six sessions, treatment was discontinued due to significant clinical deterioration. The patient was referred for home palliative care, where he subsequently died.

DISCUSSION

GCOC is an aggressive malignant neoplasm (1). This report describes a case illustrating the clinical, radiographic, and histopathological implications of this lesion, highlighting the associated diagnostic and therapeutic challenges.

First described in 1985 by Ikumra et al. as a documented case report, GCOC is known by several names: malignant COC, carcinoma arising in COC, aggressive/malignant epithelial ghost cell odontogenic tumor, dentinogenic ghost cell ameloblastoma, and malignant calcifying ghost cell odontogenic tumor (8). GCOC is the most recent designation added by WHO (3). It occurs more frequently in males, with peak incidence in the fourth decade of life (9). Clinically, GCOC may present as a slow-growing tumor with jaw swelling, paresthesia, ulceration, pain, tooth mobility, and root resorption or displacement in the affected area, along with possible invasion into surrounding soft tissue (10).

Radiographically, it typically appears as a poorly defined radiolucent lesion or mixed radiolucent/radiopaque lesion, with opacity caused by dentinoid formation or ghost cell mineralization (11). Histologically, it demonstrates rounded epithelial islands in a fibrous stroma, with epithelial cells that are typically small and round with hyperchromatic nuclei or large with vesicular nuclei. Numerous mitotic figures are observed, and ghost cells are found in varying numbers, either isolated or in clusters (12). Studies have shown that immunohistochemical biomarkers Ki-67 and MMP-9, which are related to tumor proliferation and invasion, are positively expressed in GCOC, proving useful for diagnosis, particularly in the differential diagnosis from DGCT (13).

In the present case, the patient presented with an extensive lesion in the right maxillary region, with bone destruction and involvement of adjacent structures. Diagnosis was confirmed through histopathological and immunohistochemical studies, revealing ghost cells and cytokeratin positivity. Radiotherapy was discontinued due to clinical deterioration, ultimately resulting in the patient's death. This case underscores the aggressive nature of GCOC and the need for early diagnosis and multidisciplinary management.

CONCLUSION

This case report describes a rare instance of GCOC, a lesion with unpredictable prognosis due to its high recurrence rate, wide variety of growth patterns, and extremely limited number of reported cases in the literature. Given its aggressive

behavior and unpredictability, long-term patient follow-up is strongly recommended.

Author Contributions

Antonione Santos Bezerra Pinto: Study conception and design, data collection, data analysis and interpretation, final approval of the manuscript.
Jean de Pinho Mendes: Manuscript writing and critical review.
Alan Leandro Carvalho de Farias: Data collection, data analysis and interpretation.
João Fernando Araújo Lages: Manuscript writing and critical review.
João Marcus Vaz Soares Castro: Manuscript writing and critical review.

Funding

This study received no financial support.

Conflict of Interest

The authors declare no conflicts of interest.

Acknowledgments

Not applicable

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