

# Ghost cell odontogenic carcinoma of the mandible: case report and review of literature

## Case Report

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## ABSTRACT

**Introduction** Ghost cell odontogenic carcinoma is described as an exceptionally rare malignant odontogenic tumor. The most characteristic histological feature is the presence of ghost cells; however the presence of ghost cells is not a specific feature. This article reports a mandibular case and documents its clinicopathological features, histological and radiological images and finally treatment received. Case report A 19-year-old female patient consulted for painful left cheek swelling, gradually increasing in volume. A 8 cm × 3 cm expansive mass was examined in the mandible, Physical examination revealed absence of molar contact on the left side with deviation of the lower dental arch on the contralateral side. Radiological assessment revealed a large, multi-compartmental osteolytic lesion of the left mandible, reaching upwards to the coronoid process and the temporal condyle. Histological examination of the surgical piece supported the diagnosis of ghost cell odontogenic carcinoma. Additional resection of the tumorous bone boundaries was performed. Then the decision of the multidisciplinary consultation meeting was to supplement the care with external radiotherapy. **Discussion** Ghost cell odontogenic carcinoma is very rare. The patient whose observation we report is particularly young. Clinical and radiological signs are not specific. The diagnosis is histological. The aggressive and recurrent nature requires radical wide surgery and close surveillance.

**Key Words:** odontogenic carcinoma, Ghost cell, mandible .

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## INTRODUCTION

Ghost cell odontogenic carcinoma is described as an exceptionally rare malignant odontogenic tumor that was first documented by Ikemura and al in 1985. <sup>[1]</sup> .

In addition, odontogenic ghost cell carcinoma is an extremely rare malignant odontogenic epithelial tumor with features resembling calcifying odontogenic cysts that can occur as a newly tumor or from dentinogenic ghost cell tumor after multiple recurrences or from calcifying odontogenic cystic tumor. <sup>[2]</sup>.

The most characteristic histological feature is the presence of ghost cells, however the presence of ghost cells is not a specific feature, they could be seen in others tumors. Histologically ghost cells are epithelial cells that have lost their nuclei leaving only a faint out-line of the original nuclei. <sup>[3]</sup>.

## OBSERVATION

It's about a 19-year-old female patient consulted

in our department with a chief complaint of painful left cheek swelling, his medical and family history did not reveal any relevant information.

The anamnesis reveals the installation 3 years ago of a swelling of the left cheek, gradually increasing in volume despite symptomatic treatment with paracetamol and Amoxiciline-Clavulanic acid. No clinical symptoms or lesions prior to this episode was reported. All of this evolving in a context of conservation of the general condition.

The physical examination objectified poor oral hygiene, generalized caries. A 8 cm × 3 cm expansive mass was examined in the mandible, and the swelling extended from the first premolar to the posterior border of the ramus of the mandible. On palpation, the swelling was solid without fluctuation. The mandibular teeth were affected with malposition of teeth N ° 35, 36, 37. The overlying mucosa was normal. The overlying skin was smooth and normal. No neurological deficit was observed.

Panoramic dental x-ray done on 252020/08/ (Figure 1) highlighted a large, multi-compartmental osteolytic lesion of the left mandible, reaching upwards to the coronoid process and the temporal condyle. It comes in intimate contact with molar and premolar teeth with an included aspect of tooth 38.



CT of the Facial Massif (Figure 2) done on 252020/08/ objectified: a voluminous osteolytic mass involving the entire left mandible, measuring about 80mm long axis, comprising fleshy areas and other fluid and multiple partitions, blowing the bony cortices and coming into intimate contact with teeth 35, 36, 37 and 38 lysing their alveolar edge. It pushes back muscle structures and adjacent soft parts without invading them.



Based on clinical findings, a provisional diagnosis of ameloblastoma was done. But the diagnosis of an odontogenic tumor has been mentioned once the results of the scanner were obtained.

On 202020/10/, the patient underwent a left parasymphiseal interrupting left hemimandibulectomy bearing the tumor as a single piece with placement of a scalloped mandibular reconstruction plate (Figure 3).



Histopathological examination of the surgical specimen showed an odontogenic carcinoma whose histological appearance is compatible with ghost cell odontogenic carcinoma and the bone boundaries were tumorous.

Histologically, it is a tumor proliferation infiltrating pre-existing bone tissue arranged in clumps of varying size and in spans. In places, tumor cells are basaloid in appearance, rounded, with an enlarged nucleus, heterogeneous chromatin, and low basophilic cytoplasm. Certain massifs are bordered at the periphery by cylindrical cells of palisade arrangement provided with a nucleus projected towards the apical pole. At the center of these massifs, cellularity is looser. Several clusters of eosinophilic ghost cells are often seen in the center of clumps.

The patient was resumed on 07/12/2020, thus benefiting from a complementary excision of the tumorous bone boundaries with a margin of 1 cm, removing the both premolars. And the anatomopathological examination of the sample confirmed healthy internal boundaries.

This case was discussed in a multidisciplinary meeting on 01/14/2021 and the decision was to complete the treatment with external radiotherapy.

## DISCUSSION

The term « ghost cell odontogenic carcinoma » is the most recent and has been adopted in the latest classification of the World Health Organization [4, 6].

Ghost cell odontogenic carcinoma is also known by several names: calcified malignant odontogenic cyst, malignant ghost cell epithelial odontogenic tumor, carcinoma developed on calcified odontogenic cyst and calcified odontogenic malignant ghost cell tumor.

This tumor is exceptional. This predominance is also observed for calcified odontogenic cyst [5, 7]. Men are twice as affected as women and the age ranges from 13 to 72 years with a peak in the fourth decade [4, 7].

Ghost cell odontogenic carcinoma can develop newly or in a known calcified odontogenic cyst [7, 8].

Multiple recurrences of calcified odontogenic cyst have been observed prior to diagnosis of ghost cell odontogenic carcinoma, suggesting secondary malignant transformation after a long evolution [7]. Few cases have reported the simultaneous existence of a calcified odontogenic cyst and ghost cell odontogenic carcinoma [8, 9].

The clinical manifestations are not specific. They associate maxillary or mandibular bone swelling with sometimes jugale extension and local paresthesias [4, 7].

The seat is both maxillary and mandibular with no particular location [5, 7]. Macroscopically, the lesion is cystic, well circumscribed, with a solid part with a rough surface. It is rarely entirely solid [6].

The radiological appearance is that of an osteolytic image, with blurred boundaries, sometimes containing radiopaque material. A tooth displacement is often observed more rarely a rhizolysis [5, 7].

The diagnosis is histological, based on the presence of malignant odontogenic epithelial cells associated with a benign calcified odontogenic cyst [4, 7]. The stroma is fibrous. Cellular pleomorphism is prominent and the figures of mitosis are numerous. The malignant contingent can be separated or mixed with the benign contingent. Ghost cells vary in number and can be isolated or grouped into clusters [6, 7]. These are keratinized, polygonal, eosinophilic, pale and nucleus-free epithelial cells. They are thought to result from abnormal terminal differentiation of keratinocytes or from apoptosis of poorly differentiated odontogenic cells [4, 10].

Foci of calcification, necrosis and dysplastic dentin can also be seen [4, 7]. Ghost cells are not specific for ghost cell odontogenic carcinoma. They are also seen in pilomatrixoma, craniopharyngioma, odontoma and ameloblastic fibro-odontoma.

In immunohistochemistry, malignant cells and ghost cells express epithelial markers, cytokeratin and EMA. Positivity for NSE and the p53 protein has been reported. They do not express CD34, CEA, vimentin and the S100 protein [7].

The treatment of ghost cell odontogenic carcinoma is not well codified. The evolution is unpredictable and relapses are frequent [4, 7]. Wide surgical excision is recommended. Adjuvant radiotherapy with or without chemotherapy is still controversial [8, 11].

## CONCLUSION

Ghost cell carcinoma is a very rare odontogenic carcinoma. All reported cases have demonstrated malignant histological features such as cell pleomorphism, ghost cells, mitosis and necrosis in association with the odontogenic epithelium. Some cases are characterized by aggressive behavior and others by relatively indolent growth like our case report. A multidisciplinary decision including the opinion of an expired pathologist is essential in determining the appropriate treatment to ensure optimal outcome, although wide excision with clean margins and long-term monitoring are strongly recommended. However, more studies are needed to determine if adjuvant therapy is necessary.

## CONFLICT OF INTEREST

This clinical study was self-funded by the author, with no conflict of interest.

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