

GHOST CELL ODONTOGENIC CARCINOMA: A CASE REPORT OF AN EXCEPTIONAL ODONTOGENIC TUMOR

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ABSTRACT

Ghost cell odontogenic carcinoma is an unusual histological entity. it corresponds to a malignant odontogenic tumor. We report the case of a mandibular ghost cell odontogenic carcinoma, occurring in a 19-year-old woman. The diagnosis was made by histology after a hemi mandibulectomy. Evolution was favorable. Clinical and radiological signs of Ghost cell odontogenic carcinoma are not specific. The diagnosis is histological. Evolution is unpredictable. Wide Surgical resection is recommended. Conclusion: This case report is an opportunity to discuss the epidemiological, clinical and pathological features of this exceptional tumor. More case reports are needed for further understanding and characterization of this tumor.

Keywords: Ghost cell carcinoma; Mandible; Odontogenic tumor.

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INTRODUCTION:

The first case of Ghost cell odontogenic carcinoma (GCOC) was described by Ikemura in 1985 as a malignant calcifying odontogenic tumor [1]. This tumor may appear de novo or arise from a pre-existing calcifying cystic odontogenic tumor (CCOT) or dentinogenic ghost cell tumor (DGCT) [2]. It is exceedingly rare and only approximately 50 cases have been reported, with more than half occurring in Asian patients [3]. The following is a report case of GCOC in the mandible. We discuss epidemiological, clinical, and pathological features of this exceptional tumor.

CASE REPORT

A 19-year-old woman with no pathological history, presented with left cheek swelling that has been progressing for 3 years and gradually increasing in volume. The clinical examination objectified a hard left cheek swelling without other abnormalities. A CT scan was performed and showed a tumor located

in the vertical branch of the left mandible, suspected of ameloblastoma. A hemimandibulectomy was performed. On macroscopic examination, the mandible was deformed by a tumor measuring 8 cm (**Figure 1**). Histological examination of the specimen showed poorly limited infiltrative growth proliferation, arranged in nodules, islands and strands in a fibrous stroma. The margin showed evidence of tumor invasion into the mandible alveolar bone. Tumor cells were round, of middle size, basaloid with enlarged nuclei, heterogeneous chromatin, and basophilic cytoplasm. Some massive showed peripheral palisading with reverse polarization. Several clusters of ghost cells were observed. These ghost cells have a polygonal shape with eosinophilic cytoplasm without visible nucleus. Mitotic figures were numerous (**Figures 2-5**). Based on the study of cytonuclear atypia's, mitoses, infiltrative growth pattern, and the presence of ghost cells, the diagnosis of ghost cell odontogenic carcinoma was made. An adjuvant treatment by radiotherapy was indicated.

The extension workup did not reveal any metastatic lesion. The evolution was favorable without recurrence.

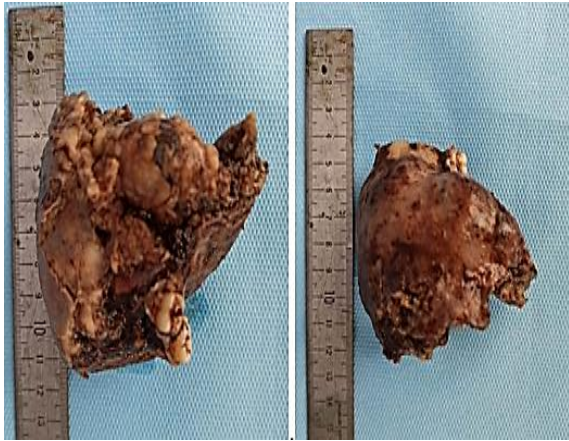


Figure1: Macroscopic appearance.

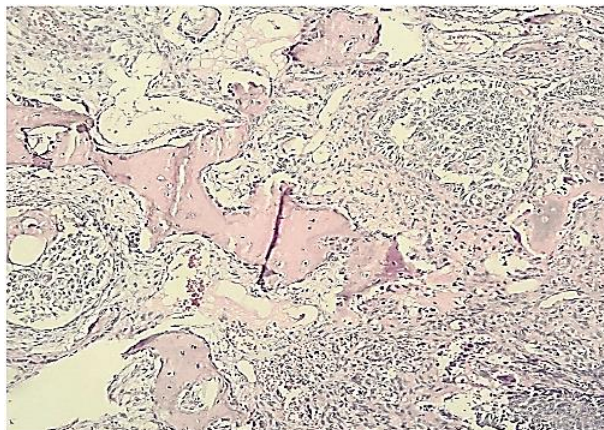


Figure 2: Poorly limited proliferation infiltrating bone (Haematoxylin & Eosin x100)

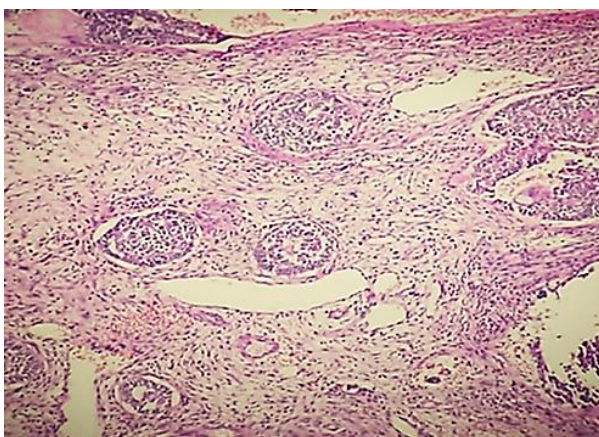


Figure.3: Tumor cells arranged in nodules and islands (Haematoxylin and Eosin x100)

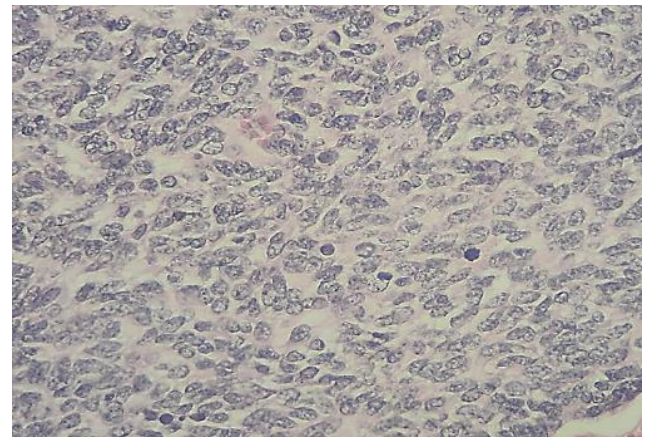


Figure 4: atypia & mitosis (Haematoxylin & Eosin x400)

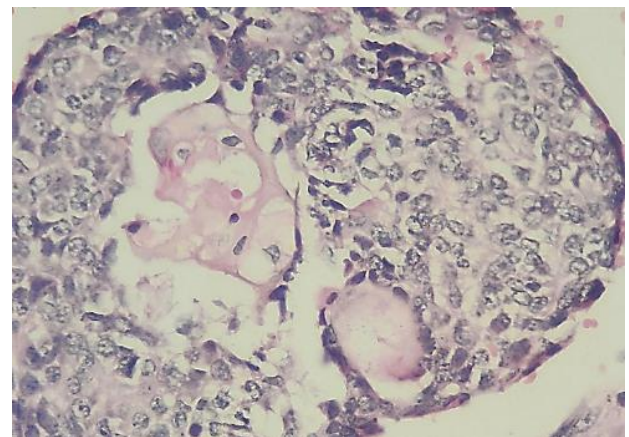


Figure 5: clusters of ghost cells within nodules (Haematoxylin & Eosin x400)

DISCUSSION

GCOC, also known as calcifying ghost cell odontogenic carcinoma or malignant epithelial odontogenic ghost cell tumor [4], is an exceptional entity [5]. Almost 50 cases have been reported with more than half occurring in Asian patients [14]. The man is four times more affected than the woman and the age is between 11 and 79 years old with a peak in the fourth decade [5]. Our patient was young.

It can be located in the maxillary or the mandible. The maxillary is the most commonly involved site [4]. The clinical symptoms are not specific. It may be manifested by slow growing swelling of the jaw, pain, ulceration, loosening of teeth, nerve signs, root resorption, and sometimes soft tissue invasion [3]. In radiological exploration, mixed multilocular radiolucent-radiopaque lesions with or without clear borders are more common than radioactive lesions [6]. Tooth impaction, displacement or resorption of tooth roots could be reported; infiltration of structures around the tumor or bone destructions are also commonly described [7,8].

The diagnosis of GCOC is histological. It's based on the presence of cytological evidence of malignancy, including mitotic activity; pleomorphism and hyperchromatism; necrosis and an infiltrative growth pattern associated with ghost cell keratinization, dentinoid formation, or presence of a DGCT or a precursor of calcifying odontogenic cyst. The malignant cells are arranged in strands and islands inside a hyalinized or fibrous stroma. The cells are uniform, small basaloid cells with round dark nuclei in most cases, but can be larger with pleomorphic vesicular nuclei [4]. GCOC is histologically characterized by small clusters or large masses of keratin-rich ghost cells. These ghost cells are anucleate cells with homogeneous, pale eosinophilic cytoplasm. They contain spherical masses of keratin that preserve the outline of the cell and correspond to the former site of the nucleus [9]. Ghost cells are not specific for GCOC. They can be observed in pilomatrixoma, craniopharyngioma, odontoma, and ameloblastic fibro-odontoma [10]. In immunohistochemistry, malignant cells, and cells phantoms express the epithelial markers, cytokeratin, and EMA. Positivity for NSE and the p53 protein was reported. They do not express CD34, ACE, vimentin and the S100 protein [10]. The treatment of COCF is not well codified. Wide Surgical resection is recommended. Adjuvant radiotherapy with or without chemotherapy is still controversial [11, 12]. The prognosis is unpredictable due to the wide variety of growth patterns reported with the tumors ranging from slow-growing, locally invasive carcinomas to highly aggressive and rapidly growing tumors with local recurrence and metastasis [13].

CONCLUSION

Ghost cell odontogenic carcinoma is an extremely rare tumor. Its diagnosis is made by histological examination. More case reports are needed for further understanding and characterization of this tumor.

No Competing Interest to Declare.

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