Case Report

Maxillary ameloblastic carcinoma: A diagnostic conundrum

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Abstract

Ameloblastic carcinoma (AC) is a rare malignant epithelial proliferation that is associated with an ameloblastoma or histologically resembles an ameloblastoma. It is considered to be an aggressive neoplasm that is locally invasive and spread to regional lymph nodes or distant sites. It requires aggressive surgical treatment, and regular follow-up, therefore, differs from ameloblastoma. Sometimes, ameloblastomas exhibit a mild-to-moderate degree of cytological atypia; hence, in such cases, a correlation should be established between the clinical, radiological and histopathological findings, thus detecting the aggressiveness of the tumor. Here, we present the case report of a 52-year-old male patient diagnosed as AC based on histopathological and immunohistochemical findings.

Keywords: Ameloblastic carcinoma, ameloblastoma, cytological atypia, immunohistochemistry

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INTRODUCTION

Odontogenic neoplasm derives from epithelial and mesenchymal remnants of the tooth germ that are classified into benign and malignant tumors. The malignant odontogenic neoplasm is extremely challenging to study due to their rarity of occurrence. Most of what we know regarding these malignant neoplasms is derived from either few case reports or small case series. The limited number of cases makes it difficult to establish standardized diagnostic criteria and tumor clinical characterization.[1-3]

The term ameloblastic carcinoma (AC) was introduced by Elzay in the year 1982.[4] AC is one such rare odontogenic malignancy accounting for 1.5%–2.0% of all malignant odontogenic tumors. It has features of ameloblastoma intermingled with features of carcinoma, regardless of whether it has metastasized, i.e., ameloblastoma in which there is histological malignant transformation.[5,6] The term malignant ameloblastoma is confined to those ameloblastomas that metastasize despite an apparently typical benign histology in both the primary and metastatic lesions.[7] The incidence ratio of AC to malignant ameloblastoma was found to be 2:1.[8]

Most of the ACs appear to be of the primary type.[9] Only six cases of secondary AC have been reported.[10] Clinically, AC is aggressive, locally invasive and causes distant metastasis.[9] Here, in this article, we report a rare case of AC of the maxilla with clinical, radiological and histopathological features along with the Immunohistochemical (IHC) findings.

CASE REPORT

A 52-year-old male patient presented with the chief complaint of pain and swelling in the upper right anterior

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region for the past 8 months. The patient gave a history of trauma in the same region, after which the swelling had developed and gradually increased in size. The patient was prescribed with analgesic and antibiotic by a local physician, but there was no relief in the symptoms, and hence referred to the institute. Extraoral examination revealed a diffuse swelling of size 4.5 cm × 3.5 cm present on the right side of the face (maxilla), anteroposteriorly 1 cm behind the ala of the nose to the lobe of the ear and superoposteriorly occupying themed region of the face. The swelling had evolved over a period of 8 months. The swelling was firm in consistency and painful, and no local rise of temperature was found [Figure 1].

On intraoral examination, erythematous growth was seen on the right side of the maxilla extending from 14 to 17 [Figure 2].

Radiographic examination revealed ill-defined radiolucency and significant bone loss on the right side of the maxilla. Destruction of the lower orbital margin can also be seen. Root resorption of 16 was seen [Figure 3].

Based on the clinical examination and radiographic finding, a provisional diagnosis of ameloblastoma was considered. Differential diagnoses of odontogenic keratocyst, primary intraosseous carcinoma and metastatic neoplasm were considered.

After obtaining an informed consent, an incisional biopsy was performed to arrive at a definitive diagnosis.

Microscopic examination of hematoxylin and eosin stained histopathological revealed the presence of odontogenic epithelium in various patterns, predominantly in follicular pattern. Sheets like arrangement of the odontogenic epithelial cells were also seen with hyperchromatic nuclei and scanty cytoplasm. Few of the follicles showed stellate reticulum-like cells. There was evidence of increase in cellular atypia and mitotic activity. Connective tissue stroma was fibrocellular [Figure 4].

Based on histopathological examination, a diagnosis of AC was established. To further confirm the diagnosis, immunohistochemistry was performed. The sections were stained with CK19 and were found to be positive [Figure 5].

Thus, histopathology and immunohistochemistry confirmed the diagnosis of AC.

DISCUSSION

AC is a rare odontogenic malignancy that has histologic features of both ameloblastoma and carcinoma. In the World Health Organization (WHO) classification, published in 2005, AC was defined as a rare odontogenic malignancy that combines the histological features of ameloblastoma with cytological atypia even in the absence of metastases.

Malignant epithelial odontogenic tumors included in the 2017 WHO classification are primary intraosseous carcinoma, sclerosing odontogenic carcinoma, clear cell odontogenic carcinoma and ghost cell odontogenic carcinoma.[11] AC has been classified into two types: primary and secondary. The former develops *de novo*, and the latter develops by malignant transformation of a preexisting benign ameloblastoma.[12,13]

The mean age of occurrence has been found to be 53.5 years, but the age range for AC has been found to be

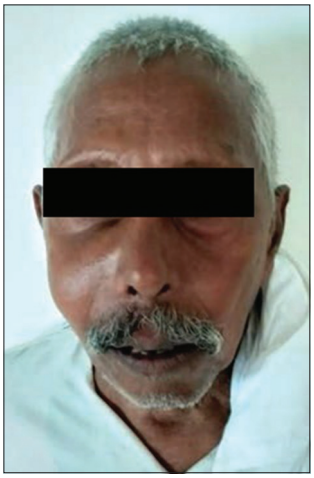

Figure 1: Clinical picture showing a diffuse extraoral swelling of size 4.5 cm × 3.5 cm present on the right side of the face (maxilla)

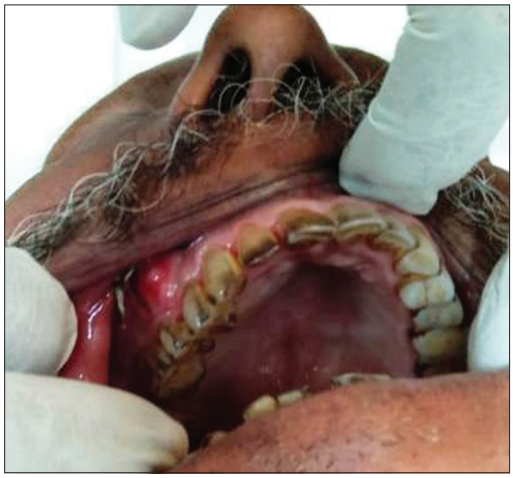

Figure 2: Intraoral picture showing erythematous growth on the right side of the maxilla extending from 14 to 17

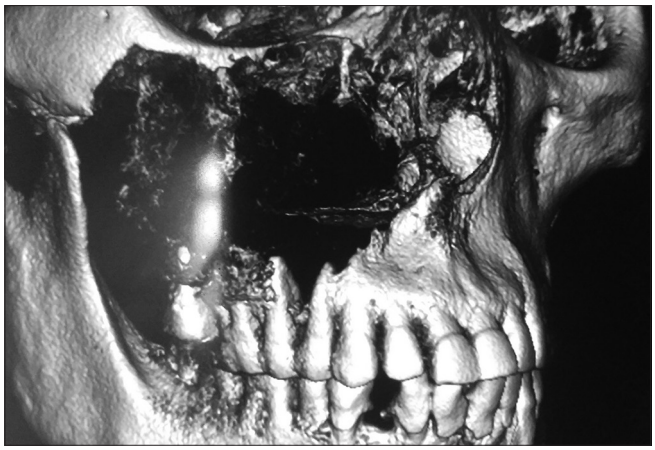

Figure 3: Radiograph revealing ill-defined radiolucency and significant bone loss on the right side of the maxilla

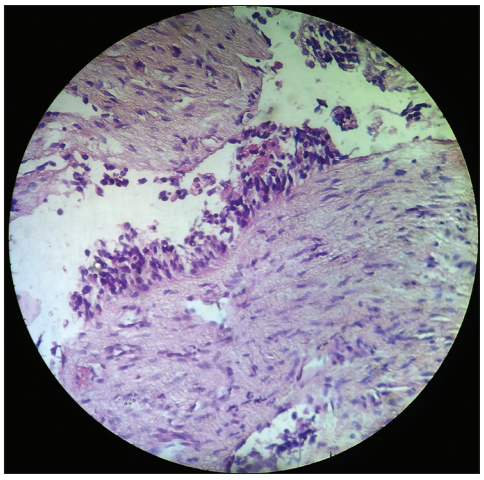

Figure 4: Odontogenic epithelium in follicular pattern, few of the follicles showing stellate reticulum-like cells, along with increase in cellular atypia and mitotic activity

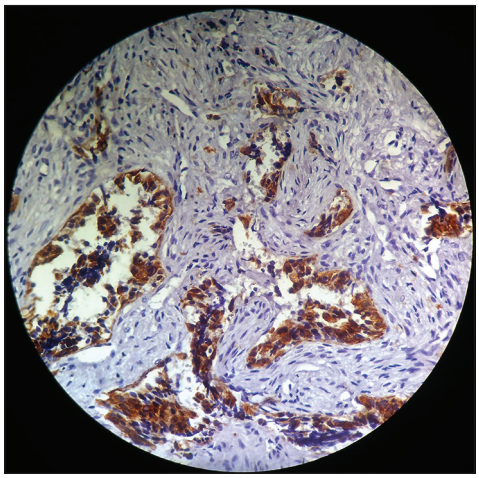

Figure 5: CK19 was found to be positive

51–84 years.[14] A male predominance has been reported, and similar to ameloblastoma, the mandibular posterior region is the common site associated with AC.[15] In this case report, the patient presented with swelling in the maxillary posterior region. Uzawa *et al*. presented a case report of maxillary AC.[12]

AC is the malignant counterpart of ameloblastoma, and BRAF V600E mutation has been found which is identical to those in other ameloblastic neoplasms.[11]

The clinical symptoms of AC are similar to ameloblastoma except few other features seen in AC are the aggressive and painful growth is more aggressive, painful, trismus, dysphonia and perforation of cortical plates is observed in AC.[11]

The radiographic appearance is usually similar to that of ameloblastoma except for the presence of focal radio-opacities which reflects dystrophic calcifications. The case described in this report also showed similar radiological features with destruction of the cortical plate. In this case, we found complete destruction of the cortical plates but no radio-opacities. Corio *et al*. reported that dystrophic calcification is not specific to ACs but can be seen in the desmoplastic variant of ameloblastoma.[15]

Histopathologically, AC is similar to ameloblastoma except for it will exhibit cytological atypia is seen in both primary and metastatic ACs. However, the classical features of ameloblastoma including reverse polarity and peripheral palisading are lost. So pleomorphism, altered nuclear–cytoplasmic ratio, abnormal mitoses and vascular or nerve invasion are considered to be important features for the diagnosis. The presence of necrosis may be helpful. The mitotic rate is usually increased, but the increase in mitotic activity alone is not valuable.[16,17]

Few of the histopathological differential diagnoses included were primary intraosseous carcinoma, basaloid squamous cell carcinoma, mucoepidermoid carcinoma and acanthomatous ameloblastoma.

The various IHC markers for AC are CK19, Ki-67, MMP-2 and MMP-9. In this case, we conducted IHC with CK19 on the sections and found it to be positive.

There are four points in the clinical criteria that can be helpful for the diagnosis of AC such as rapid growth, tendency to perforate the cortex, pain and paresthesia that are distinct from their benign counterpart.[17]

The treatment for AC is radical surgical resection along with chemotherapy and radiotherapy as an adjunct.

CONCLUSION

Ameloblastic carcinoma is a rare malignant odontogenic tumor exhibiting histologic atypical changes and hence should be differentiated from its benign counterpart ameloblastoma. Ameloblastic carcinoma should be included in the differential diagnosis of those cases where the patient is presenting with rapid growth, pain and perforation of the cortex.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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