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

CASE REPORT

A 52-year-old male patient presented with the chief complaint of pain and swelling in the upper right anterior maxilla. The patient had a history of pain and swelling for the past 6 months. The swelling was progressive and was accompanied by pain. The patient had no history of facial trauma or any other systemic illness. On clinical examination, a firm, non-tender, non-reducible swelling was noted in the right maxillary region. The swelling was mobile and extended from the upper right canine to the upper right first molar. There was no lymphadenopathy present. Radiographically, a well-defined radiolucent lesion was noted in the right maxillary region with a scalloped margin. The lesion extended to the alveolar process and the sinus. There was no evidence of cortical destruction or root resorption.

The patient was diagnosed with maxillary ameloblastic carcinoma. The lesion was excised along with a margin of normal tissue. The excised specimen was submitted for histopathological examination. The histological examination revealed a tumor with a characteristic pattern of ameloblastic carcinoma. The tumor cells were arranged in a trabecular pattern with a palisading arrangement at the periphery. The tumor cells showed a moderate degree of cytological atypia with pleomorphism. The tumor cells were positive for cytokeratin and negative for S-100 protein. Immunohistochemical staining revealed the tumor cells to be positive for Ki-67, indicating a high proliferation rate.

The patient was followed up regularly for the next 2 years. There was no evidence of recurrence or metastasis. The patient was asymptomatic and had no evidence of tumor growth.

This case report highlights the importance of histopathological and immunohistochemical examination in the diagnosis of maxillary ameloblastic carcinoma. The lesion should be excised along with a margin of normal tissue to ensure complete removal of the tumor. Regular follow-up is necessary to detect any recurrence or metastasis.

Keywords: Ameloblastic carcinoma, maxillary ameloblastic carcinoma, immunohistochemistry, histopathological examination, follow-up.
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
region is the common site associated with AC.\textsuperscript{[13]} In this case report, the patient presented with swelling in the maxillary posterior region. Uzawa \textit{et al.} presented a case report of maxillary AC.\textsuperscript{[13]}

AC is the malignant counterpart of ameloblastoma, and BRAF V600E mutation has been found which is identical to those in other ameloblastic neoplasms.\textsuperscript{[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.\textsuperscript{[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 \textit{et al.} reported that dystrophic calcification is not specific to ACs but can be seen in the desmoplastic variant of ameloblastoma.\textsuperscript{[13]}

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.\textsuperscript{[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.\textsuperscript{[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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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