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Case Report

Large ameloblastic carcinoma: A rare case with management

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ABSTRACT

The ameloblastic carcinoma is a rare malignant counterpart of the ameloblastoma. Ameloblastic carcinoma is a malignant lesion with characteristic histologic features and behavior that dictates a more aggressive surgical approach than that of a simple ameloblastoma. However, reliable evidence of its biologic activity is currently unavailable due to the scarcity of well-documented cases. We present a rare case of a large ameloblastic carcinoma occurring in the mandible of 44-year-old female highlighting its unique histopathologic features and aggressive behavior treated by radical surgery and reconstruction. Involvement of left ramus to right ramus of the mandible makes this case unusual.

Key Words: Ameloblastic carcinoma, malignant counterpart, mandible

INTRODUCTION

In 1983, Shafer[1] introduced the term ameloblastic carcinoma to describe ameloblastomas in which there had been histologic malignant transformation.

Cases of ameloblastic carcinomas are rare and limited details on prevalence, incidence or relative frequency are presently available. Compared to the metastasizing, malignant ameloblastoma; however, the ameloblastic carcinomas seem to be more common (2:1).[2]

Ameloblastic carcinoma occurs in a wide range of age groups, but the mean age of 30.1 years is in agreement with that reported for ameloblastomas. There is no apparent sex predilection. The most commonly involved area is the posterior portion of the mandible. Involvement of the maxilla by ameloblastic carcinoma seems to be less frequent than that of the mandible. The most common sign described has been swelling; although, others include associated pain, rapid growth, trismus and dysphonia.[3]

Carcinomas derived from ameloblastomas have been designated by a variety of terms, including malignant ameloblastoma, ameloblastic carcinoma, metastatic ameloblastoma, and primary intra-alveolar epidermoid carcinoma.[4]

In 1971, the World Health Organization,[5] published its classification of odontogenic carcinomas recognizing the following subtypes:

- Malignant ameloblastoma
- Primary intra-osseous carcinoma
- Other carcinomas arising from odontogenic epithelium, including those arising from odontogenic cysts.

In this classification, "malignant ameloblastoma" refers to a neoplasm in which typical histologic features of ameloblastoma are seen in the primary tumor located in the jaw as well as in any associated metastatic deposits.

In 1984, Slootweg and Müller,[6] further emphasized that ameloblastomas may exhibit malignant features other than metastasis and suggested a modified classification system for malignant tumors with

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features of ameloblastoma, based on characteristics of malignancy:

Type 1: Primary Intraosseuos Carcinoma ex odontogenic cyst

Type 2: (a) Malignant ameloblastoma (b) Ameloblastic carcinoma, arising *de novo*, ex ameloblastoma or ex odontogenic cyst

Type 3: Primary Intraosseous Carcinoma arising *de novo* (a) Non-keratinizing (b) Keratinizing.

The treatment of and prognosis for ameloblastic carcinoma is unclear in the literature due to the rarity of this tumor and the lack of well-documented patients. Surgical excision, with or without adjuvant radiotherapy, seems to be required for local control. Surgery is the optimal treatment; although, the best approach remains controversial.

CASE REPORT

A 44-year-old female patient presented to the Outpatient department with a chief complaint of swelling in the lower jaw since 6 months. There was difficulty in speech, mastication, and deglutition. There was no associated pain. She had no contributing medical history.

On extra oral examination a large well-defined swelling was noticed in the mandibular anterior region crossing the midline causing facial asymmetry [Figure 1a].

The swelling extended below the inferior border of the mandible and the skin over the swelling was stretched and smooth. She had difficulty in opening the mouth. No lymph nodes were palpable.

Intraoral examination showed a large swelling, which extended completely into the floor of the mouth and completely obliterating the lingual and buccal vestibules mediolaterally [Figure 1b].

The lingual frenum was pushed back. Anteriorly, it extended from the labial sulcus to the ramus posteriorly. There was no surface discharge present, the mucosa over the swelling was normal and its color was same as that of the normal tissue. All the mandibular incisors were missing. Generalized extrinsic stains were present.

On palpation the inspectory findings were confirmed. The swelling was bony hard, non-tender, and immobile. None of the teeth present showed mobility.

Routine blood and urine examination was normal. Fasting and post-prandial sugar levels were also normal.

Computed tomography showed a large multilocular osteolytic lesion extending from 37 to 47 region crossing the midline, destruction of both cortices and pathologic fracture was seen [Figure 2a]. Soap bubble and honeycomb patterns were appreciable. A multi-centric growth pattern was seen showing a permeative type of destruction [Figure 2b].

A chest radiograph was taken to rule out any primaries in the lung.

A provisional diagnosis of ameloblastoma was established. A differential diagnosis of odontogenic keratocyst was made.

An incisional biopsy was performed under local anesthesia and microscopic examination revealed odontogenic islands infiltrating the connective tissue, the peripheral tall columnar cells showed proliferation

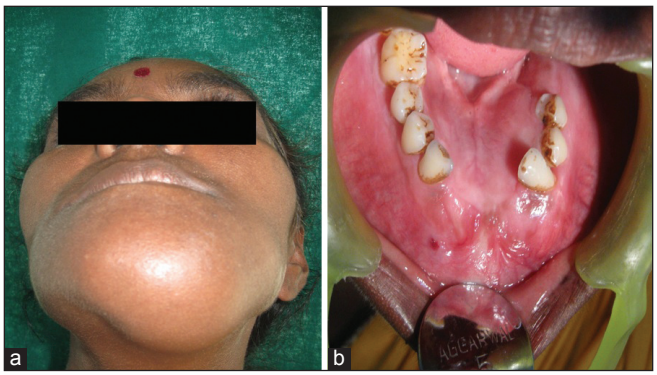

Figure 1: (a) Large well-defined swelling involving the mandibular anterior region crossing the midline causing facial asymmetry, (b) large swelling extending completely into the floor of the mouth and completely obliterating the lingual and buccal vestibules

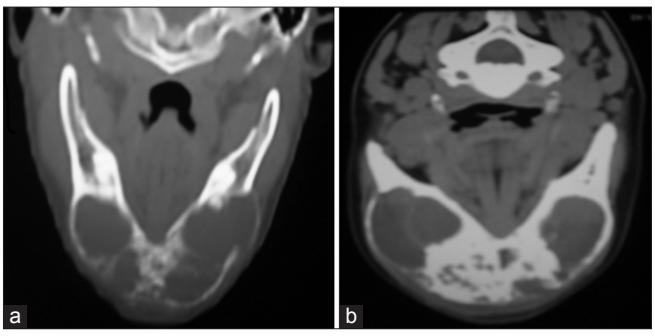

Figure 2: (a) Computed tomography (CT) showing a large multilocular osteolytic lesion extending from 37 to 47 region crossing the midline, destruction of both cortices and pathologic fracture are seen, (b) CT revealing a multi-centric growth pattern and showing a permeative type of destruction

and peripheral palisading of basal cells with reverse polarity of the nucleus. Stellate reticulum was scanty [Figure 3a].

High power view showed cells with atypical features of pleomorphism, hyperchromatism, altered nuclear-cytoplasmic ratio, and mitotic figures [Figure 3b].

A final diagnosis of ameloblastic carcinoma was established.

A total mandibulectomy was carried out under general anesthesia [Figure 4a]. An immediate reconstruction was done [Figure 4b].

DISCUSSION

Malignant epithelial odontogenic tumor, which includes the malignant ameloblastoma, ameloblastic carcinoma, primary intraosseous squamous cell carcinoma, clear cell odontogenic tumor, and malignant epithelial ghost cell tumor are very rare.

Elzay,[7] Slootweg and Müller[6] used the term ameloblastic carcinoma to convey the presence of cytologic features of malignancy. The degree of differentiation in epithelial neoplasms is usually considered to be significant in predicting biologic behavior of metastasis. The main difference between Elzay's and Slootweg and Müller's schemes relates to the minor point of histogenesis. According to these authors, the term ameloblastic carcinoma should be used to designate lesions that exhibit histologic features of both ameloblastoma and carcinoma.

The tumor may metastasize and histologic features of malignancy may be found in the primary tumor, the metastases or both. Our case did not show any evidence of metastasis. The term malignant ameloblastoma should be confined to those ameloblastomas that metastasize despite an apparently typical benign histology in both the primary and the metastatic lesions. The incidence of ameloblastic carcinoma is greater than that of malignant ameloblastoma by a 2:1 ratio.[8]

The neoplasm may be derived from a number of different sources such as those of odontogenic origin, including ameloblastoma, odontogenic cysts or epithelial odontogenic rests as well as entrapped salivary gland epithelium or epithelium entrapped along embryonic fusion sites.[8]

The consensus now is to use the term ameloblastic carcinoma for those tumors with the histological evidence of malignancy in the primary, recurrent, or metastatic tumor regardless of whether there is metastasis or not although malignant ameloblastoma is reserved for metastasizing ameloblastomas, which exhibit benign histological features both in the primary and metastatic lesion.[9]

The characterization of carcinoma arising centrally within the mandible and the maxilla is an uncommon, but complex problem. The first step in the staging process must be the exclusion of metastasis or invasion of bone by tumor from adjacent soft-tissue or paranasal sinus.

Carcinomas in the jaws metastasizing from primary locations such as the lung, the breast and the gastrointestinal tract may mimic ameloblastic carcinoma and must always be ruled out clinically before a diagnosis is made.[10]

The clinical and radiological picture most commonly resembles ameloblastoma, but ameloblastic carcinoma can be suspected if there is a sudden increase in the size of the swelling, pain, trismus, paresthesia and numbness or if there is any of foci of calcification as these features are unusual in ameloblastoma. In

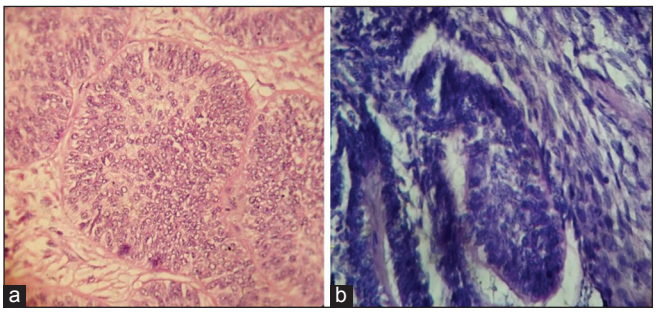

Figure 3: (a) H and E, ×10 low power view showing an odontogenic island with peripheral tall columnar cells showing proliferation and peripheral palisading of basal cells with reverse polarity of the nucleus, (b) H and E, ×40 low high power view showing cells with atypical features of pleomorphism, hyperchromatism, altered nuclear-cytoplasmic ratio, and mitotic figures

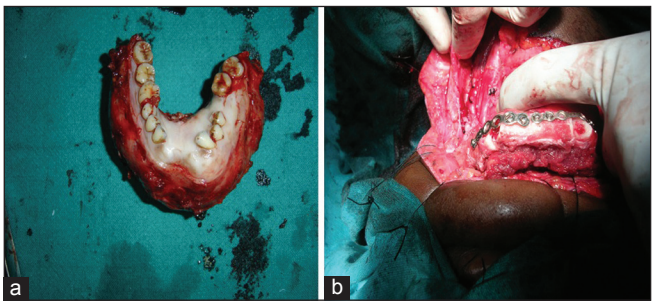

Figure 4: (a) The excised gross specimen, (b) mandibular reconstruction carried out

the present case, the swelling was large involving a considerable portion of the mandible extending from the left ramus to the right ramus.

The mitotic features in ameloblastic carcinoma have been considered as significant by few authors especially when ameloblastic carcinoma is arising de novo, wherein mitosis is higher.

In the present case, there was no evidence of regional or distant metastasis, but there was histological evidence of typical ameloblastic cells and anaplastic cells in the same tumor. In addition, there was cellular pleomorphism and nuclear hyperchromatism with occasional mitoses in the same tumor.

Primary intra-alveolar epidermoid carcinoma must be considered in the differential diagnosis of ameloblastic carcinoma. Although, the primary intra-alveolar carcinoma and the ameloblastic carcinoma exhibit some clinical differences, their histologic features are similar enough to suggest a histogenetic relation. It is possible, then, that the primary intra-alveolar carcinoma may represent simply a less differentiated, usually non-keratinizing form of ameloblastic carcinoma, both lesions being derived from odontogenic remnants.

Our case occurred in a 44-year-old female with involvement of both sides of the mandible.

Thus, the term ameloblastic carcinoma can be applied to our case, which showed focal histologic evidence of malignant disease including cytologic atypia and mitoses with indisputable features of classic ameloblastoma.

The treatment of and prognosis for ameloblastic carcinoma is unclear in the literature due to the rarity of this tumor and the lack of well-documented patients. Surgical excision, with or without adjuvant radiotherapy, seems to be required for local control. Surgery is the optimal treatment; although, the best approach remains controversial. For this patient, surgical excision (total mandibulectomy) was carried out.

CONCLUSION

We have presented a rare case of a large ameloblastic carcinoma in a 44-year-old female, which involved a large considerable part of the mandible crossing the midline making this case unusual, which was treated by radical surgery and immediate jaw reconstruction.

It has been suggested that the high rate of recurrence is due to its mode of growth and surgical mismanagement rather that any inherent malignant properties and metastases are "exceedingly rare."[11]

Ameloblastic carcinoma is an aggressive odontogenic tumor that requires aggressive surgical treatment. The clinical and biological differences between conventional ameloblastoma and ameloblastic carcinoma are significant and can be useful to distinguish between the two entities when the pathological diagnosis is not certain.

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