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CASE REPORT

Maxillary Ameloblastic Carcinoma

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ABSTRACT

Odontogenic carcinomas of the maxilla are classified as malignant ameloblastoma, ameloblastic carcinoma, or primary intraosseous carcinoma. The term ameloblastic carcinoma is used to describe those ameloblastomas in which there is histological evidence of malignancy in the primary, recurrent, or metastatic tumour. The maxilla is an unusual site for ameloblastoma. This report is of a patient with an ameloblastoma of the maxilla with an unusual multiphasic histological pattern. Treatment consisted of right subtotal maxillectomy and ipsilateral modified radical neck dissection, free-flap reconstruction, followed by postoperative adjuvant external beam radiation. A literature review describing clinical and histological presentation of this rare tumour is presented.

Key Words: *Ameloblastoma; Maxilla; Radiotherapy*

INTRODUCTION

Ameloblastoma is a benign odontogenic tumour, although it is locally aggressive. Ameloblastoma is believed to originate from the remnants of dental epithelium. This tumour accounts for 1% of all cysts and tumours of the jaw and is the second most common odontogenic tumour after odontoma. Approximately 80% of ameloblastomas occur in the mandible and 20% occur in the maxilla.[1] The malignant variant of ameloblastoma has been the source of discussion for many years, and the classification of the subtypes has been vague. 'Primary ameloblastic carcinoma' has been classified recently by the World Health Organization (WHO) as a tumour that demonstrates the morphological features of ameloblastoma with atypia, regardless of the presence or absence of metastasis.[2] This report is of a patient with a rare ameloblastic carcinoma of the right maxilla, with regional metastasis to the ipsilateral cervical lymph nodes.

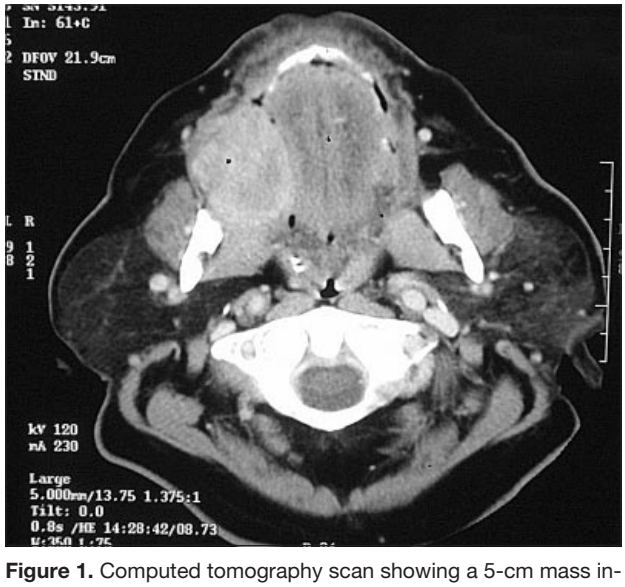

Figure 1. Computed tomography scan showing a 5-cm mass involving the right maxillary alveolus, extending into the lateral aspect of the hard palate.

CASE REPORT

A 73-year-old Caucasian man presented to his dentist in 2005 with a white exophytic lesion in the right upper gingiva, that had been gradually growing for the past 2 years, preventing him from wearing his upper denture set. He was referred to an oral and maxillofacial surgeon, who performed an incisional biopsy of the mass. Histopathological studies confirmed a diagnosis of spindle-cell carcinoma. He was referred to the head and neck oncology service for treatment.

Clinical examination revealed an edentulous maxilla with a 4- to 5-cm exophytic mass, extending from the

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right canine prominence anteriorly to the right maxillary tuberosity posteriorly. The mass invaded the lateral aspect of the hard palate. Neck examination revealed an enlarged right submandibular lymph node. His past medical history was significant for prostate cancer that was treated with radical radiotherapy and lupron-based antiandrogen hormone injections.

An infused computed tomography (CT) scan of the head and neck showed a 4-cm destructive mass involving the right maxillary alveolus, extending to the inferior portion of the maxillary sinus and into the pterygopalatine fissure. The bulk of the tumour was located in the oral cavity (Figure 1). CT scan also showed a 2-cm metastatic right submandibular node. Chest radiograph and full blood count were unremarkable.

The patient was diagnosed with spindle-cell carcinoma of the right upper alveolus, staged as T4N1M0 according to the American Joint Committee on Cancer staging system.[3] The recommended treatment was right subtotal maxillectomy and ipsilateral modified radical neck dissection, followed by adjuvant external beam radiation.

The patient underwent general anaesthesia with an orotracheal tube, and surgical tracheostomy was performed to maintain the airway. Right modified neck dissection was performed at levels I, II, III, IV, and V, with preservation of the spinal accessory nerve, internal jugular vein, and sternocleidomastoid muscle. The primary tumour was then approached using a Weber-Ferguson incision, through which a subtotal maxillectomy was performed. A split-thickness skin graft harvested from the right thigh was used to cover the defect. Iodoform gauze was packed in the defect, and a prefabricated obturator was placed and secured in position. Frozen-section assessment showed clear margins.

Intraoperatively, the patient developed ST segment elevation associated with hypotension, which required vasopressor support. His immediate postoperative electrocardiogram showed evidence of acute inferior myocardial infarction that required an emergent angiogram with a right coronary artery stent. The remainder of the postoperative course was uneventful. The pathology report showed the mass to be an ameloblastic carcinoma of the maxilla, with cytological features of malignancy, as well as increased nuclear to cytoplasmic ratio, nuclear hyperchromatism, mitotic figures, and necrosis in the tumour consistent with ameloblastic carcinoma of the maxilla (Figure 2). One neck node was positive for metastasis.

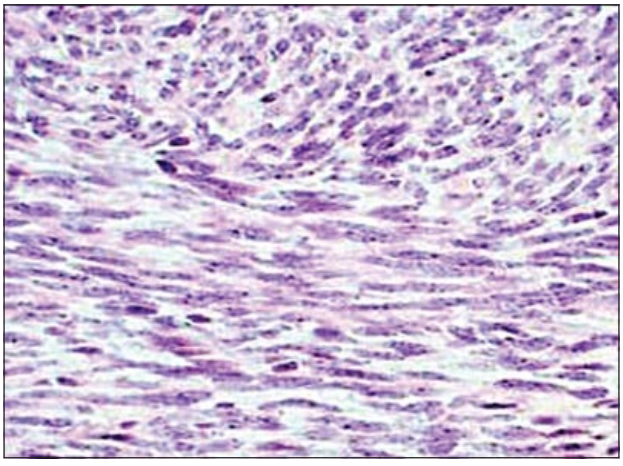

Figure 2. Pathology specimen showing the pattern of ameloblastoma and features of atypia, including increased nuclear to cytoplasmic ratio, mitotic figures, and necrosis (haematoxylin and eosin stain; original magnification, x 10).

The patient received postoperative adjuvant external beam radiation in view of the locally advanced primary tumour at presentation. The dose was 66 Gy in 33 fractions using a 6 MV photon machine targeting the tumour bed and ipsilateral neck. Clinical and radiological follow-up for 8 years has not shown evidence of disease.

DISCUSSION

In 1972, the WHO published a classification of odontogenic carcinomas, which recognised malignant ameloblastoma, primary intraosseous carcinoma, and other cancers arising from odontogenic epithelium.[4] However, there was no provision for ameloblastic carcinoma and similar lesions in this classification. Elzay modified the classification, but many pathologists did not agree with the proposed subgrouping.[5] The term 'ameloblastic carcinoma' was first introduced by Shafer et al in 1983 to describe a malignant epithelial odontogenic tumour that histologically retains the features of ameloblastic differentiation, and exhibits cytological features of malignancy in the primary or recurrent tumour.[6]

Slootweg and Muller stated that ameloblastoma may show features of malignancy other than metastasis, and suggested a new classification in which the type 2B category contained ameloblastic carcinoma, arising de novo, ex ameloblastoma, or ex odontogenic cyst.[7]

In 2005, the WHO revised the classification of odontogenic carcinoma and described primary ameloblastic carcinoma as an odontogenic malignancy that combined

the histological features of ameloblastoma with cytological atypia with or without metastasis.[2] Despite these classifications, the diagnosis, histological features, treatment, and prognosis remains challenging to clinicians. There is a lack of pathological features to distinguish ameloblastoma with no malignant/metastatic potential from malignant histology that may metastasise. Often, pathologists report ameloblastoma as a primary intraosseous carcinoma. The initial diagnosis of spindle-cell carcinoma for this patient shows the challenge faced by pathologists when reviewing tumours with multiple characteristics such as ameloblastic carcinoma.

The treatment of and prognosis for ameloblastic carcinoma is unclear in the literature due to the rarity of this tumour 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.[8] For this patient, surgical excision with neck dissection followed by radiotherapy was planned according to the initial diagnosis of spindle-cell carcinoma, and the treatment plan was not changed after the final diagnosis of ameloblastic carcinoma.

As ameloblastic carcinomas are rare, there is no consensus for their treatment. Despite the lack of adequate clinical data, surgery followed by radiotherapy seems to be the treatment of choice.[9] Preoperative radiotherapy has been suggested to decrease the tumour size and may be used to treat some rapidly growing tumours before radical surgery.[10] At the University of Florida, 3 patients with ameloblastic carcinoma were treated with surgery and postoperative radiation, to a total mean dose of 68 Gy. All patients had local control after 24 months.[11]

There are limited data for the efficacy of radiation. Robinson reported a series of 18 patients treated with radiation alone, 72% of whom had a recurrence.[12] Recently published studies highlight that radiation can induce regression but not cure.[13] The role of chemotherapy is not yet proven.[14]

Currently most clinicians treat ameloblastic carcinoma in the same manner as other oral cavity carcinomas, with surgery and postoperative radiation. Indications for adjuvant radiation include close or positive margins, nodal involvement, extracapsular extension, and stage T3 or T4 tumours. Documented reports with meaningful follow-up are rare. Meticulous follow-up is essential because recurrence and metastasis to the lungs and regional lymph nodes have been reported in the literature.[15] Further reporting of ameloblastic carcinoma is encouraged.

Ameloblastic carcinoma is an aggressive odontogenic tumour 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 2 entities when the pathological diagnosis is not certain.

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