The Journal of Laryngology and Otology
October 1995, Vol. **109**, pp. 1019–1022

Ameloblastic carcinoma of the maxilla

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Abstract
Ameloblastic carcinoma is an unusual tumour. There have been a total of 34 cases of ameloblastic carcinoma in the English literature to date. Of these only 11 cases have occurred in the maxilla. The authors report the 12th such case. The histological classification for odontogenic carcinoma has been debated for many years and recently revised, thus differentiating between malignant ameloblastoma and ameloblastic carcinoma. The authors review the current literature regarding diagnosis and treatment of this unusual lesion, and support the use of the term malignant ameloblastoma for the tumours that metastasize in spite of their benign histological appearance, whereas, the ameloblastic carcinoma is referred to as the primary tumour with malignant transformation, regardless of its metastatic potential.

Key words: Maxillary sinus neoplasms; Carcinomas, ameloblastic

Introduction

Ameloblastomas are rare, odontogenic tumours: approximately 80 per cent are found to occur in the mandible and the remaining 20 per cent in the maxilla. It is theorized that the tumour originates from the epithelial components of the embryogenic tooth that has arrested development prior to the stage of enamel formation. Carcinomas arising from ameloblastomas have been given many names including malignant ameloblastoma, ameloblastic carcinoma, metastatic ameloblastoma and primary interalveolar epidermoid carcinoma.

Malignant ameloblastomas have been classified by the World Health Organization (WHO) (see Pindborg *et al.*, 1972) and included with odontogenic carcinomas. According to their definition malignant ameloblastomas represent tumours that, in spite of the cytologically benign appearance of the lesions, metastasize while both the primary and the metastatic lesion retain their benign histological appearance (Table I). In 1982, a classification was proposed by Elzay (1982) of primary intraosseous carcinomas (Table II). He introduced the term ameloblastic carcinoma which unlike malignant ameloblastoma is poorly differentiated: the tumour exhibits features of an ameloblastoma and a squamous cell carcinoma. Slootweg and Muller (1984) expanded the definition of ameloblastic carcinoma to include the lesions that combine features of ameloblastoma with less differentiated areas (Table III). They went on to describe 11 cases of amelobastoma in the literature with anaplastic transformation exhibiting squamous cell carcinoma in their epithelial components. We report the 12th case of ameloblastic carcinoma in the maxilla.

TABLE I
WORLD HEALTH ORGANIZATION CLASSIFICATION

Odontogenic carcinoma
(A) Malignant ameloblastoma
(B) Primary intraosseous carcinoma
(C) Other carcinoma arising from odontogenic epithelium including those arising from odontogenic cysts

TABLE II
CLASSIFICATION OF ELZAY (1982)

Primary intraosseous carcinoma
Type 1 Arising ex-odontogenic cyst
Type 2 Arising ex-ameloblastoma
(A) Well differentiated – malignant ameloblastoma
(B) Poorly differentiated – ameloblastic carcinoma
Type 3 Arising *de novo*
(A) Non-keratinizing
(B) Keratinizing

Case report

An 82-year-old, white, female without any past history of smoking or alcohol exposure presented to her family physician with trismus and bleeding from the gums. The patient was referred for evaluation of a right buccal lesion extending into both the soft and hard palates. The biopsy of the lesion was interpreted as a squamous cell carcinoma. The patient was referred to us for further evaluation and treatment. An MRI study revealed a slightly lobulated, irregular, mass immediately lateral to the inferior aspect of

TABLE III
CLASSIFICATION OF SLOOTWEG AND MULLER (1984)

Type 1 Primary intraosseous 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

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Accepted for publication: 27 May 1995.

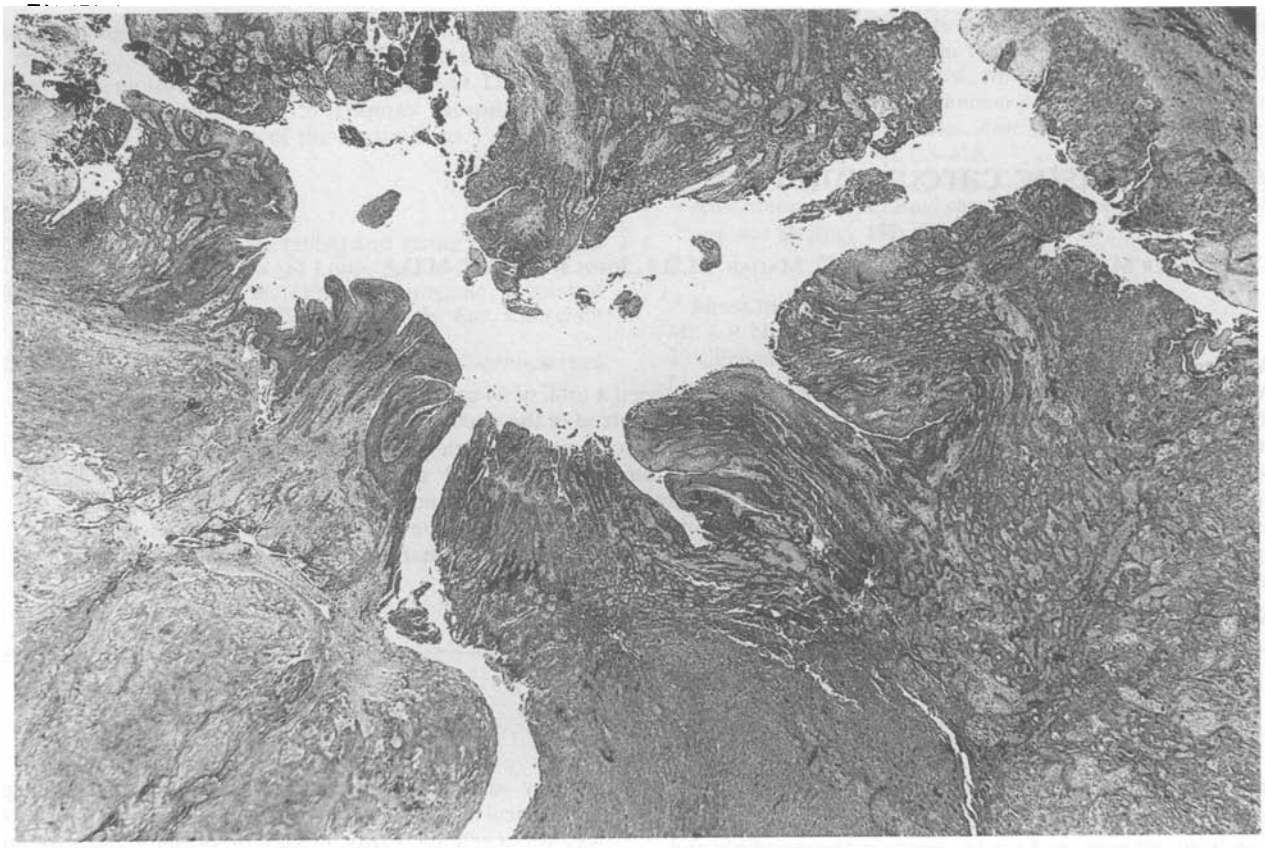

FIG. 1
Centrally cystic and ameloblastic tumour. (H&E; ×4).

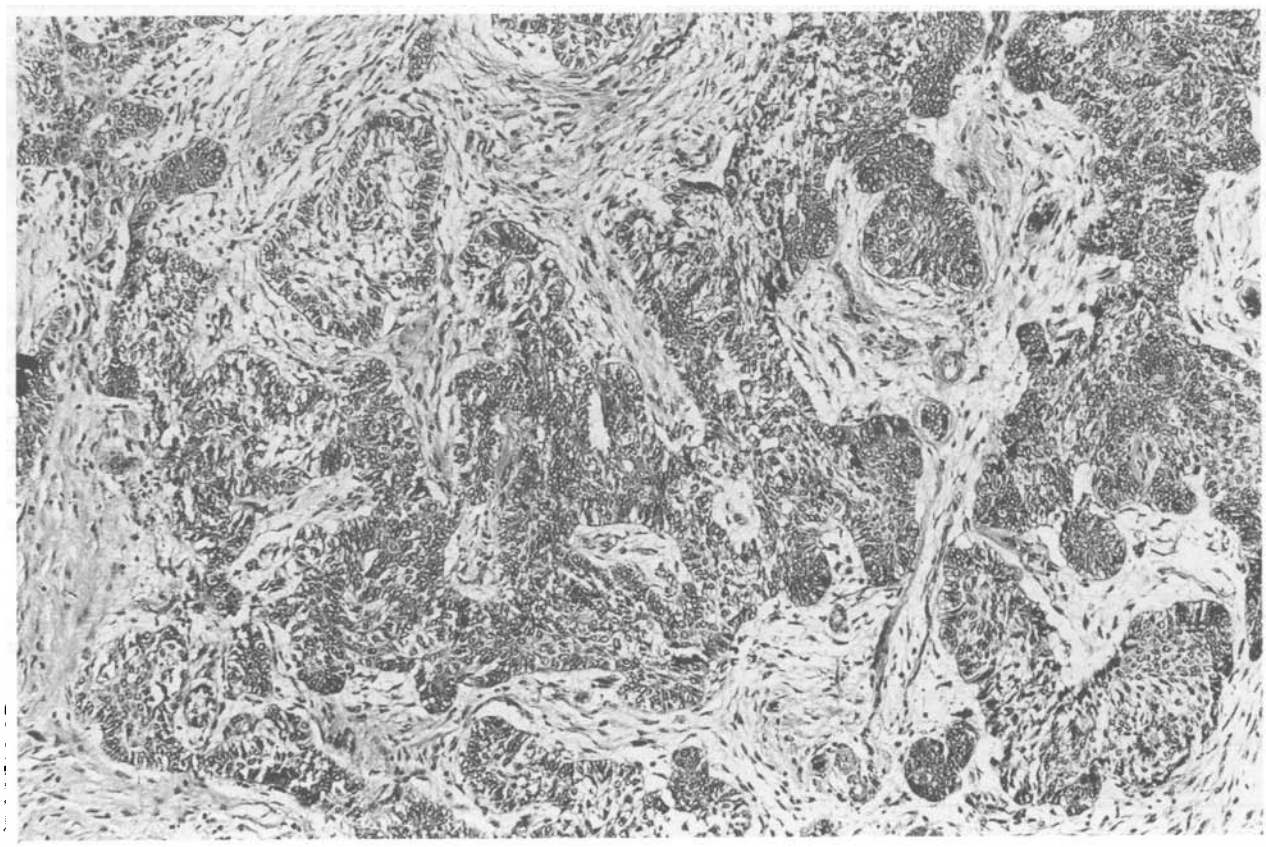

FIG. 2
Tumour with features of classical ameloblastoma. (H&E; ×10).

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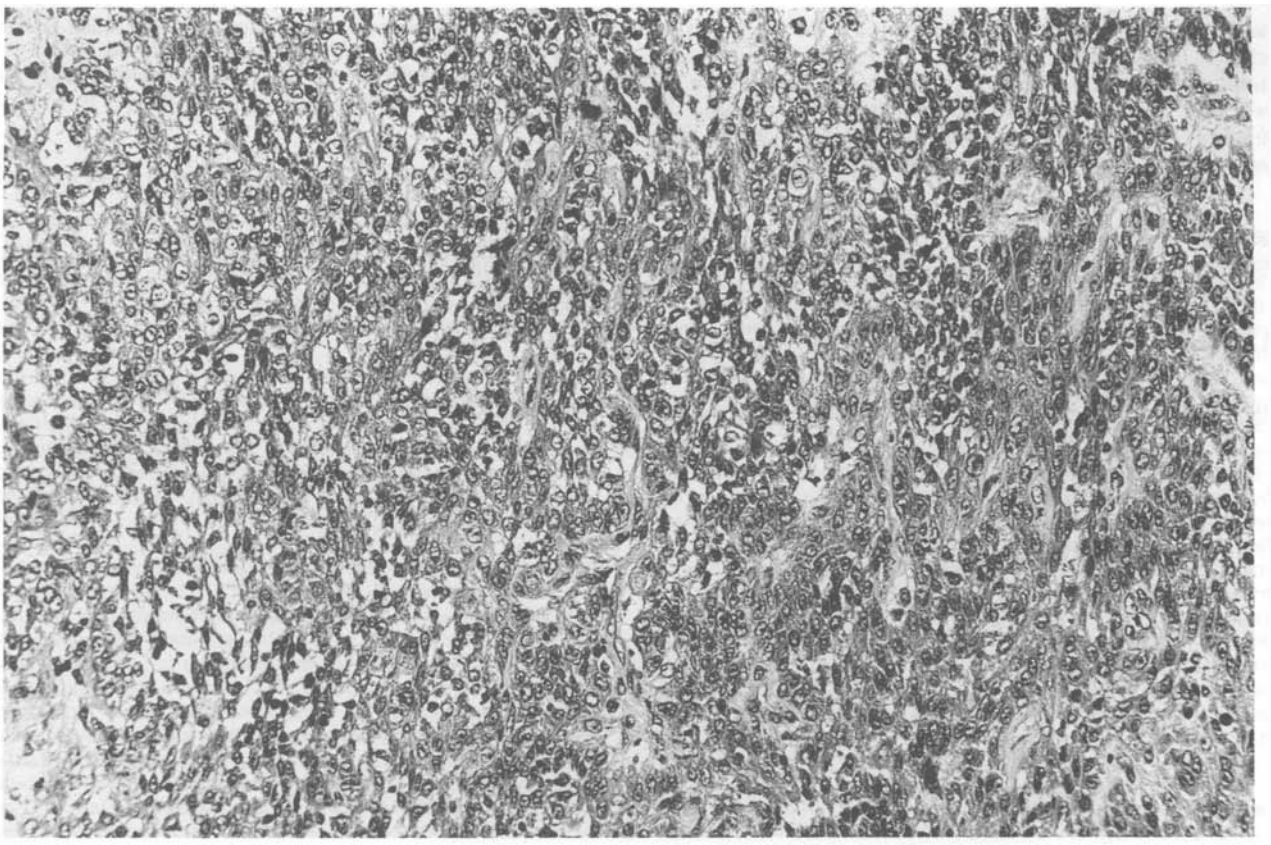

FIG. 3
Anaplastic undifferentiated areas within the tumour (H&E; ×10).

the right maxillary sinus. The mass appeared to be invading the inferolateral wall of the maxillary sinus. No other lymph nodes were detected on the MRI study.

The patient subsequently underwent partial maxillectomy and excision of the buccal lesion. The tumour was found to be in the form of a nodule measuring 3.5 × 1.4 × 1.2 cm. The cut surface of the tumour was grey/white and smooth with a central cystic area. Microscopically, the tumour showed features of a typical amelobastoma with a peripheral, palisading arrangement and acanthomatous differentiation (Figure 1). The central cystic area was partially lined with epithelium displaying squamous differentiation (Figure 2). In other areas, the neoplastic cells infiltrated between the minor salivary glands and dense fibrous stroma as broad sheets and cords. The neoplastic cells demonstrated indistinct cellular margins and round to oval vesicular nuclei with marked nuclear pleomorphism. The presence of occasional mitotic figures was noted. There was loss of the peripheral palisading pattern (Figure 3). These features were felt to indicate malignant transformation within an ameloblastoma. There was no evidence of bone invasion in the decalcified sections, indicating the peripheral odontogenic nature of this neoplasm.

Discussion

Ameloblastic carcinoma of the maxilla either peripheral, or central, is a very rare lesion. There have been only 11 reported cases to date of ameloblastic carcinoma (Chee *et al.*, 1990). The most common site is the mandible, involving the posterior portion more frequently. The common clinical signs and symptoms include swelling, pain, trismus and dysphonia. The age range appears to vary widely with a reported mean age of 30 years. No sex predilection is reported. Histologically, these tumours show ameloblastoma displaying features of malignancy, including nuclear pleomorphism, high nuclear cytoplasmic ratio and increased mitotic activity. The primary intra-alveolar epidermoid carcinoma and metastatic tumours from other sites including breast, lung, gastrointestinal tract and salivary glands should be included in the differential diagnosis.

From the literature review, it is apparent that the WHO classification of odontogenic carcinoma should be further revised. The term malignant ameloblastoma, in this classification is referred to as a neoplasm with the features of an ameloblastoma in the primary and metastatic growth. The diagnosis of malignant ameloblastoma can be made only retrospectively after the tumour has metastasized. Tumours with features of ameloblastoma and histological evidence of malignancy cannot be properly classified. Several authors in the literature have introduced the term ameloblastic carcinoma for the ameloblastic tumour with histological features of malignancy, regardless of evidence of metastasis. The malignant amelobastoma is referred to as an ameloblastic tumour that metastasizes despite benign histological features in both primary and metastatic growth. The classification proposed by Elzay (1982) (Table II) also emphasizes the histogenesis of these tumours. Slootweg and Muller (1984) in addition, include the ameloblastic carcinomas arising from odentogenic cysts in their classification, making it a more complete classification. The introduction of the term ameloblastic carcinoma allows the separation of histologically aggressive ameloblastic tumours with metastatic potential.

The treatment of ameloblastic carcinoma remains somewhat undefined. At this juncture complete excision

https://doi.org/10.1017/S0022215100131925 Published online by Cambridge University Press

with negative surgical margins is felt to be the treatment of choice. The response of a well differentiated ameloblastoma to radiotherapy is felt to be poor. The question of whether an ameloblastic carcinoma would respond is unclear. Changes to the present classification scheme and apropriate follow-up will hopefully resolve this issue.

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