The Journal of Laryngology and Otology
August 1991, Vol. **105**, pp. 676–677

Extraskeletal myxoid chondrosarcoma of the maxillary sinus

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Abstract
A case of extraskeletal myxoid chondrosarcoma of the maxillary sinus in a 45-year-old man is reported and discussed. This is the first report of this tumour in this site.

Introduction

Extraskeletal myxoid chondrosarcoma was formally delineated as an entity by Enzinger and Shiraki in 1972, although other cases not so designated had been described previously (Stout and Verner, 1953; Goldenburg, *et al.*, 1967).

It is a rare but a distinctive type of chondrosarcoma with special clinicopathological features (Enzinger and Shiraki 1972; Fu and Kay, 1974). In 1979 Batsakis reported 'To my knowledge, an extraskeletal myxoid chondrosarcoma has not been reported in the head and neck'.

In addition to many isolated reports, Enzinger and Shiraki in 1972 and Fletcher *et al.*, in 1986 reported and studied 34 and nine cases respectively of extraskeletal myxoid chondrosarcoma. None of these were in the head and neck.

Case report

A 45-year-old Caucasian man presented in February 1989 with a one week history of numbness over the left maxillary area and left upper anterior teeth, and a one month history of intermit-

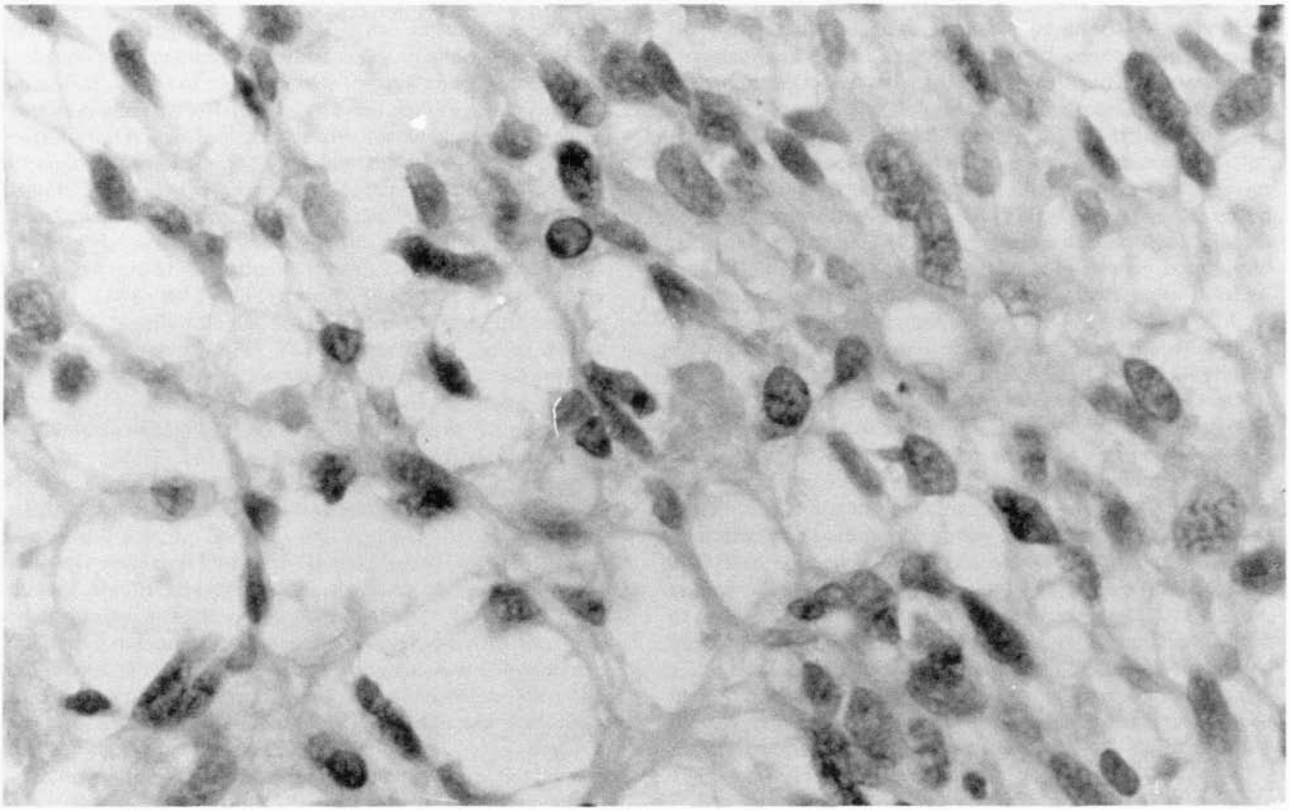

FIG. 1

The tumour is composed of an infiltrate of tumour cells with ovoid and round nuclei, with abundant eosinophilic cytoplasm set in a loose myxoid background. No intercellular mucin was demonstrated.
The extracellular myxoid material maintained staining with Alcian Blue up to an 0.5 mmol concentration of $MgCl_2$.

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Accepted for publication: 27 March 1991.

tent left epistaxis. The only other history of note was trauma of the face five years previously.

Examination of the nose showed a blood clot and a polypoid swelling protruding from the left middle meatus. The remainder of the clinical examination was normal. Plain X-ray of the sinuses showed an opaque left maxillary sinus without obvious bony erosion. C.T. scan showed a soft tissue mass filling the left maxillary and ethmoid sinuses. There was bony erosion of the medial wall of the maxillary antrum.

Examination of the nose under general anaesthetic revealed erosion of the lateral nasal wall above and below the interior turbinate. The left ethmoid sinus was full of blood clot and the left maxillary sinus was replaced by tumour. A biopsy was taken through an intranasal antrostomy.

The preliminary histopathological report confirmed the presnce of a tumour of connective tissue origin and further material was sought.

A left maxillectomy was then performed with preservation of the eye. At operation, it was confirmed that the tumour was confined to the maxillary sinus and this was completely removed. The final histological diagnosis was that of an extraskeletal myxoid chondrosarcoma. An 18 month follow-up has so far showed no evidence of local recurrence or distant metastasis.

Pathology

Macroscopic Appearances

The tumour was well circumscribed and measured 4.5 × 3.5 ×1.5 cms. The anterior wall of the maxillary sinus appeared to be eroded by the tumour. On sectioning, the mass showed a myxoid appearance.

Microscopic Appearances

The tumour was composed of an infiltrate of ovoid and round cells with vesicular nuclei and eosinophilic cytoplasm set in a myxoid stroma with a prominent vascular network (Fig. 1). Mild pleomorphism was present and mitoses were few in number. Intercellular mucin was not evident. The extracellular myxoid material maintained staining with Alcian Blue up to 0.5 mmol concentration of $MgCl_2$ (Fig. 1).

Immunocytochemical staining confirmed the mesenchymal nature of the tumour by showing reactivity with the mesenchymal marker vimentin. The tumour cells did not react with antibodies to the epithelial markers CAM 5.2 and epithelial membrane antigen, to the lymphoid marker, common leucocyte antigen, to the endothelial marker factor VIII related antigen, or to the muscle markers desmin, myoglobin or muscle specific actin.

On the basis of the morphological features, the absence of lipoblasts and the Alcian Blue staining reactions, a diagnosis of extraskeletal myxoid chondrosarcoma was made. Electron microscopy was performed but did not reveal any differentiating features, in particular no epithelial intercellular junctions or myofilaments or lipoblasts were seen.

Discussion

Extraskeletal myxoid chondrosarcoma is regarded as a malignant tumour showing chondroid differentiation which occurs in extraskeletal sites. It is an uncommon tumour which may easily be misdiagnosed as the myxoid variant of some other sarcoma, for example, liposarcoma or malignant fibrous histiocytoma or a mesenchymal chondrosarcoma (Bloch *et al.*, 1979). Avoidance of such an error becomes important when one realises that this tumour generally carries a comparatively good prognosis, having a five year survival of 85 per cent or more (Enzinger and Shiraki, 1972).

The tumour most commonly afflicts patients older than 35 years, and only a few cases have been encountered in children and adolescents. Males are more commonly affected than females. More than two thirds of the tumours occur in the extremities, especially the thigh and popliteal fossa.

The aetiology of this tumour is unknown but like other chondrosarcomas previous accidental or surgical trauma to the nose, facial skeleton or exposure to inhaled hydrocarbons may be significant (Coates *et al.*, 1977; El Ghazali, 1983). Five years prior to presentation the patient in this report had trauma to the facial bones.

Macroscopically, the tumour may appear quite haemorrhagic. Occasionally, haemorrhage may be so severe that the tumour is mistaken for a haematoma (Enzinger and Weiss, 1983). In this case the ethmoid sinus was filled with blood clot.

Although the clinical behaviour of this tumour varies from case to case, it is in general a relatively slow growing tumour that, nonetheless may recur and may even metastasize. The rate of recurrence and metastasis is related to the degree of cellularity and the relative amount of myxoid material (Enzinger and Weiss, 1983). Therefore, the least cellular and most myxoid tumours carry the best prognosis. There is no reliable data available as to its radiosensitivity, but it seems advisable to treat the more cellular examples with adjunctive radiotherapy and chemotherapy. In our case due to the scarcity of mitortic figures and the hypocellularity, post-operative radiotherapy was not planned.

Acknowledgements

The authors wish to thank the Audio-Visual Department of the Royal College of Surgeons for their photographic assistance and Ms. Saundra Dalton for typing the manuscript.

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Key words: Paranasal sinuses; Chondrosarcoma