The Journal of Laryngology and Otology
August 1994, Vol. **108**, pp. 679–681

Myxoma of the paranasal sinuses

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Abstract
Myxomas of the paranasal sinuses are rare but well described in the literature. They may be related to dental malformations or missing teeth, but may also occur without any such abnormalities. Their local aggressiveness and ability to erode bone should not be underestimated, and they should be totally removed whenever possible. A case of myxoma of the sinuses is described, in which recurrence followed several local removals. The patient was only cured by radical maxillectomy. The operation is described, and the literature on the subject reviewed.

Key words: Maxilla; Myxoma, Nose; Paranasal sinuses

Introduction

Myxomas of the paranasal sinuses are well-described tumours (Metha *et al.*, 1974; Canalis *et al.*, 1976; Fu and Perzin, 1977; Harris *et al.*, 1977; Perzin *et al.*, 1982), and may also occur within the soft tissues of the face, (Faccini and Williams, 1973; Mirejovsky and Konecny, 1978) and of the eyelid (Daicker, 1979) as well as the nasal bone (Kopke *et al.*, 1992).

Myxomas appear to occur in the nasal cavities of horses (Rahko *et al.*, 1972; House *et al.*, 1976). Myxomas are also reported in the nasopharynx by Sinha *et al.*, (1978) and in the head and neck area in children by Smith *et al.* (1977).

One of the problems with myxomas is making the histological diagnosis and the differential diagnosis, from Schwann cell neoplasms and myxomas, fibroblastic tumours, fibrous histiocytomas, and fibro-osseus lesions, may be difficult, according to Perzin *et al.* (1982).

Case report

A 47-year-old, Caucasian female presented with left-sided facial pain. This problem had started in 1970 when a polyp was removed from her left antrum and diagnosed as a benign growth. Following recurrence of the tumour in the nose, the lesion was diagnosed as a myxoma.

Physical examination of the patient revealed few changes except for a small oro-antral fistula on the left side. There was no trismus and no evidence of tumour in the nasopharynx. Examination of CT scans revealed a mass in the left maxillary antrum (Figure 1). Involvement of the lateral and posterior wall of the antrum with erosion of the pterygoid plates on the left side was noted. The pterygoid musculature did not appear to be invaded to any significant degree. There was no involvement of the orbit or the inferior orbital plate. The anterior portion of the alveolus did not appear to be involved so that sparing of the incisor and canine teeth was feasible.

The patient was operated on in February, 1986. Surgery consisted of a left total maxillectomy. This operation was performed through a left-sided Weber-Fergusson incision. The incision was made through the lip and along the left side of the nose below the left eye. The anterior teeth up to the left canine were spared with a cut being made through the tooth socket of the fourth left upper premolar tooth. The rest of the osteotomy was made down the midline of the hard palate. The whole of the soft palate was spared. Because the tumour was invading the posterior antrum a very lateral approach was made by removing the coronoid process of the ramus of the mandible. In this way the pterygoid muscles were dissected away without disturbing the pterygoid plates. The pterygoid plates were osteotomized away from their origin from the base of the skull. The floor of the orbit was included by making osteotomy cuts through the orbital rim and the lacrimal bone and lamina paparacea medially and laterally through the frontal process of the maxilla up to the inferior orbital fissure. In this way an *en bloc* excision was achieved. Intraoperative frozen section was performed to confirm that there was no residual tumour in the bed of the removed specimen. A previously made prosthesis was fitted by the prosthodontist while the patient was still in the operating room.

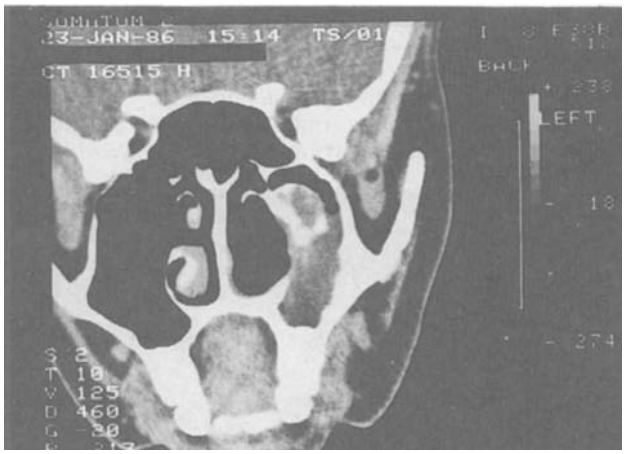

FIG. 1

A coronal CT scan through the nose and paranasal sinuses taken preoperatively. Note loss of the lateral wall of the nose, and tumour involvement of the lateral antral wall, with bone erosion.

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Accepted for publication: 2 September 1993.

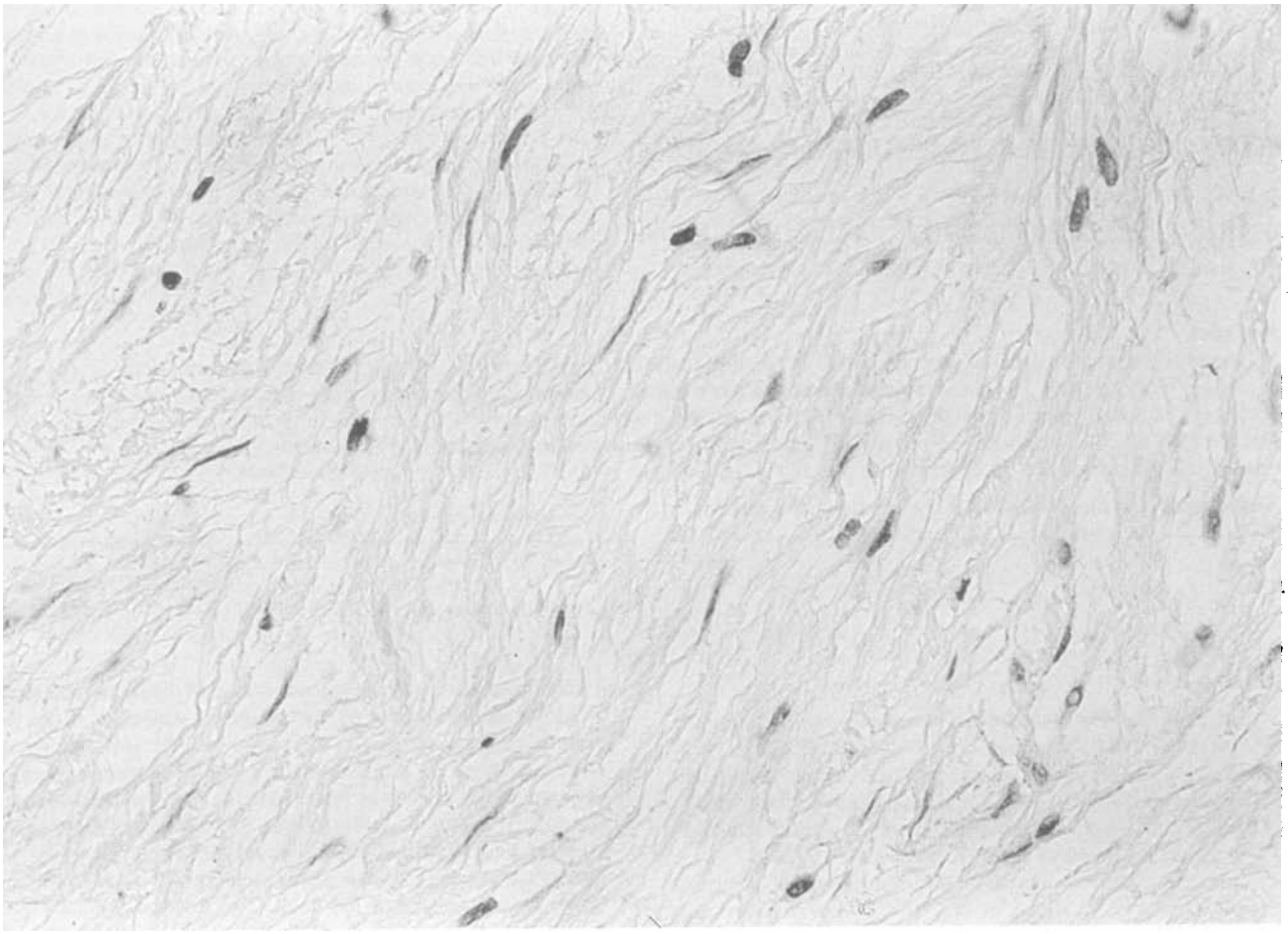

FIG. 2

A hypocellular proliferation characterized by small, bland, spindle, cells embedded in a myxomatous stroma. (H & E; × 312).

Grossly the tumour had a glistening grey-white cut surface with a gelatinous consistency. Histologically there was a myxomatous background surrounding a hypocellular proliferation of small stellate or spindle-shaped cells with regular nuclei and a variable amount of cytoplasm. Odontogenic epithelial islands were not present. Atypia or mitoses were not seen, though the tumour was locally infiltrative. Review of the previously removed tumour tissue showed similar features and the pathological diagnosis was of a myxoma (Figure 2).

This patient made an uneventful recovery except that there was some ectropion of the lower eyelid of the left eye which was subsequently dealt with by composite mucosal and cartilage grafts taken from the nasal septum. A post-operative CT scan was made a year later showing no evidence of recurrence (Figure 3). There was no clinical sign of recurrence of this tumour after seven years.

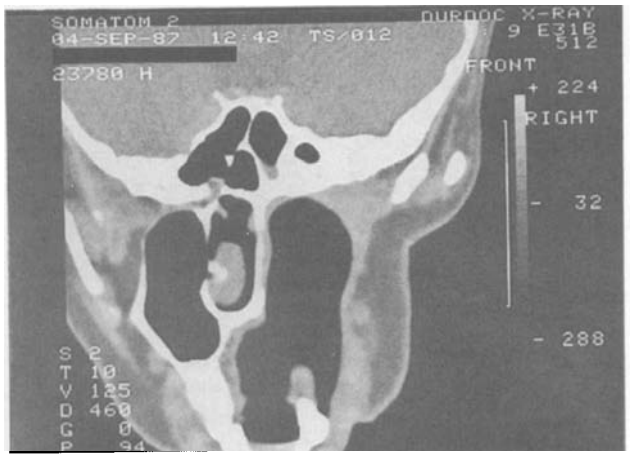

FIG. 3

A coronal CT scan taken 19 months later through a similar region, showing the total maxillectomy cavity, free of any residual tumour.

Discussion

In a review of cases received by the Registry of Otorhinolaryngologic Pathology of the Armed Forces Institute of Pathology, Heffner (1983) found nine sinunasal myxomas in a five-year period from 1976 to 1981. The patients ranged in age from two to 15 years. Out of 256 non-epithelial tumours involving the nasal cavity and paranasal sinuses and nasopharynx, Fu and Perzin (1977) found only six to be myxomas. Harrison and Lund (1993) in their series found only one myxoma in 638 neoplasms of the upper jaw. In the Netherlands Central Pathology Registry (National Automated Pathology Archive, PALGA) only one myxoma was found out of a total of 1053 tumours of the nose and sinuses over the years 1990, 1991 and 1992. These are therefore extremely rare tumours in this area.

Myxomas are mesenchymal tumours of uncertain histogenesis which may arise from dental anlage tissue (White *et al.*, 1975). This is demonstrated by the fact that myxomas of the jaws may be related to unerupted or missing teeth. Their origin from odontogenic mesenchyme is supported by a recent immunohistochemical and ultrastructural study (Moshiri *et al.*, 1992). These authors suggest that fibroblasts comprising the tooth germ undergo modification when giving rise to odontogenic myxoma. Histologically odontogenic myxomas are similar to soft tissue myxomas arising elsewhere in the body except for the presence

https://doi.org/10.1017/S0022215100127811 Published online by Cambridge University Press

of islands of odontogenic epithelium. The latter is not however necessary for the diagnosis and was present in only one out of nine cases in a series (White *et al.*, 1975). Myxomas do also develop from the sinunasal tract and facial bones, apparently from non-odontogenic mesenchyme (Sinha *et al.*, 1978; Kopke *et al.*, 1992).

The tumour cells are small, spindle-shaped or stellate, embedded in a prominent myxoid background which contains fine collagen fibrils. The pattern is mostly hypocellular. Nuclear atypia and mitoses are rare; vascular invasion has not been reported and a malignant counterpart has not been described. The tumour does however show local infiltration which explains why incomplete removal leads to local recurrence.

Ultrastructurally, the tumours examined by Moshiri *et al.* (1992) showed spindle-shaped cells surrounded by amorphous material representing the abundant ground substance seen by light microscopy. Immunohistochemically, the tumour cells were positive for vimentin, a pan-mesenchymal marker, and also for actin (Moshiri *et al.*, 1992). The histological differential diagnosis may be difficult and even impossible from biopsy material. Many mesenchymal tumours have a myxoid background and hypocellular areas. Among the benign and malignant tumours to be considered, chondromyxoid fibroma, neurofibroma, and myxoid chondrosarcoma are important. Nontumorous lesions also need to be eliminated, such as inflammatory polyp with stromal cell atypia.

It has been said that a myxoma of the antrum is essentially a benign lesion. However, in this patient the lesion recurred several times after local removals, and was only cured by radical excision. This was similar to the experience of other authors. Fu and Perzin (1977) found three patients who had been treated using limited local excisions: the tumour persisted and recurred in all three. These authors believe that myxomas in this area should be widely resected to prevent recurrence. Several of the myxomas of the head and neck have been described in children by Sinha *et al.* (1978) and these authors also found the best treatment to be total excision. In cases which were inadequately excised recurrence occurred. These authors have therefore suggested that in the paediatric patient excision should be done by frozen section control but that this should be as conservative as possible so as to preserve vital structures in these young patients. Moshiri *et al.* (1992) describe the radiological features of odontogenic myxomas as being uni- or multilocular radiolucent lesions exhibiting a delicate trabeculation. Expansion of the cortex and root resorption of teeth may be present as well as the so-called 'sun ray', 'honey-comb' and 'soap bubble' appearance.

Conclusions

Myxomas of the nose and sinuses are extremely rare. They probably account for no more than 0.5 per cent of all paranasal sinus and nasal tumours. This diagnosis should be suspected in sinusnasal tumours in children, and particularly when teeth are missing. This tumour is even more unusual in adults, and clinical features may mimic those of carcinoma of the sinuses. Their biological behaviour is aggressive and wide excision is the treatment of choice.

Acknowledgements

This patient was referred by Dr A. Hooper, of Durban, South Africa, and was treated at the Park Lane Clinic, Johannesburg, South Africa.

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