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Retropharyngeal myxoma: a case report

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Abstract
Myxomas are rare tumours which may occur in the head and neck region. About 60 cases have been reported in the literature so far. We describe a retropharyngeal myxoma in this paper.

Key words: Head and neck neoplasms; Myxoma

Introduction

Myxomas are rare connective tissue tumours of unclear histogenesis, occurring in the heart, jaw bones and soft tissues of the body but infrequently in the soft tissues of the head and neck. Tse and Vander (1985) in an extensive review of the literature found only 42 such cases, tabulated them according to location, and added one of their own.

The palate is the most common site followed by the parotid. Sixteen more soft tissue myxomas of the head and neck have been reported since 1985. We describe a case of retropharyngeal myxoma and were unable to find another in the head and neck region in the literature.

Case report

A 68-year-old Caucasian man was seen in the ENT outpatient clinic at Sandwell District General Hospital, in July 1986, complaining of an odd sensation in his throat and some difficulty in swallowing of three months' duration. He also had a history of right-sided nasal obstruction of one year's duration. He was hypertensive and had had a cerebrovascular accident four years prior to presentation. His blood pressure was under control with antihypertensive medication.

Examination revealed bilateral nasal polypi and a bulging posterior pharyngeal wall swelling, more to the left side, with an intact overlying mucosa. Posterior rhinoscopy showed extension of the swelling into the nasopharynx. The swelling extended inferiorly into the hypopharynx, partially obscuring the left vocal fold on indirect laryngoscopy. Both folds were, however, normally mobile. There was no cranial nerve deficit. A lateral neck radiograph revealed a smooth, rounded, soft tissue dense mass in the retropharyngeal region, extending from the second to the fifth vertebrae (Figure 1). Computed tomography (CT) showed a non-enhancing soft tissue mass occupying the retropharyngeal space on the left side. There was no clarification or necrosis in the tumour. Surrounding bones were not eroded. Among the differential diagnoses, the possibility of neurofibroma was considered (Hussain *et al.*, 1989).

The patient was taken to the operating theatre for a nasopharyngoscopy and exploration of the neck mass. Nasopharyngeal examination confirmed the presence of intact mucosa over the tumour, which was seen to cross the midline extending partially to the right. The mass was approached in the neck through a transverse incision on the left lateral aspect, dissecting medial to the great vessels. This revealed a soft gelatinous tumour (encephaloid-like) behind the pharynx, deep red in colour. Dissection around the tumour was easy on all sides except posteriorly where it seemed to be adherent to the vertebral column. It was felt safer at this point to defer removal of the swelling until further radiological tests ruled out any connection with the spine. It is mandatory with such a midline retropharyngeal lesion to rule out any spinal connection (Singh and Pahor, 1977; Pahor,

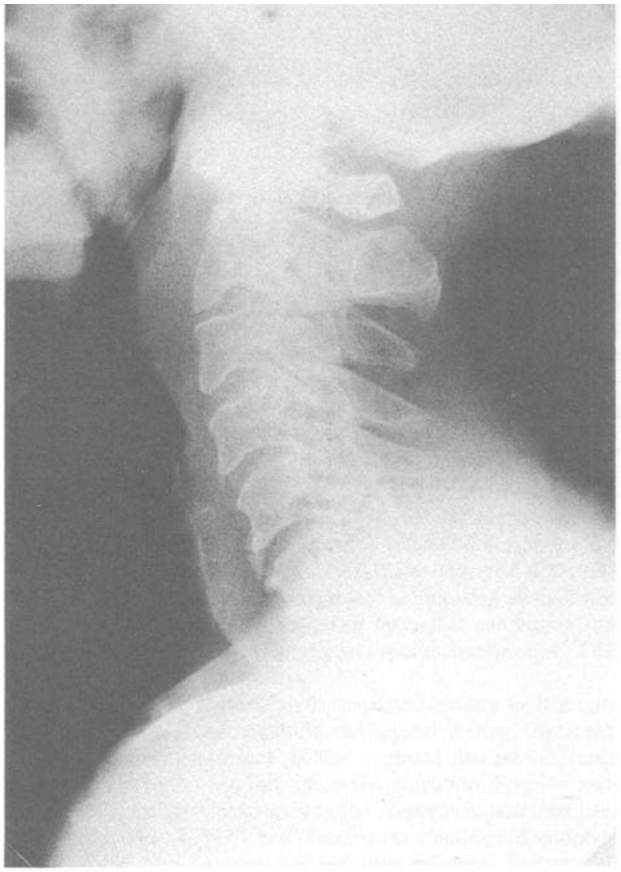

FIG. 1
Lateral plain X-ray of the neck showing retropharyngeal mass.

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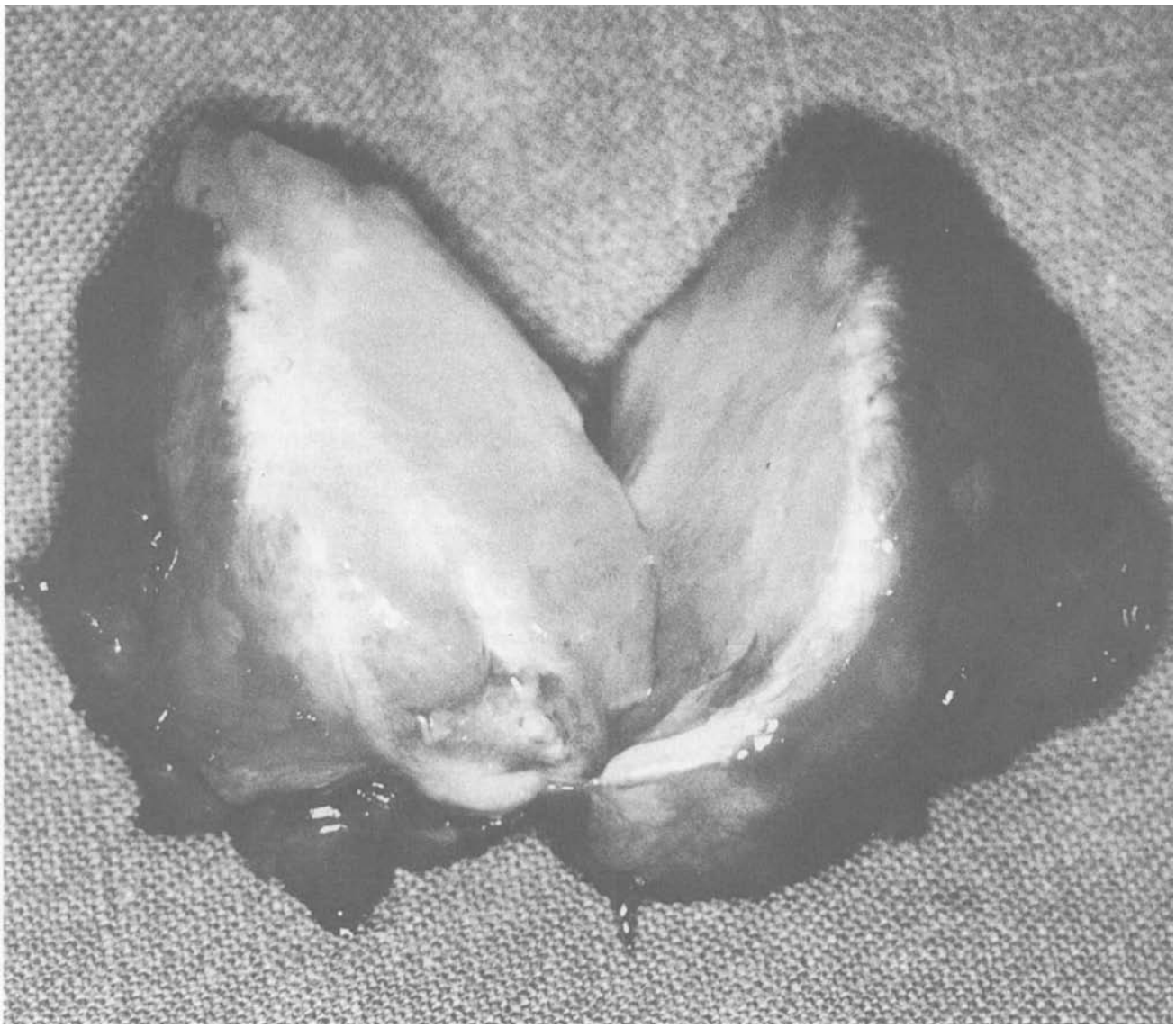

FIG. 2

Retropharyngeal mass – bisected showing apparent capsule.

1980). A part of the tumour was excised for biopsy and the wound closed with drainage. On sectioning, the lesion was found to consist predominantly of myxomatous areas that stained for mucopolysaccharide, with intervening fibrous tissue. Mitoses were absent and vascularity was minimal. Computed tomography was repeated to rule out any connection posteriorly with the spine. A fortnight after the first surgery, the neck was re-explored through a collar incision and an 8 × 3 cm mass excised in toto by blunt finger dissection. There was no cranial nerve deficit following the operation. Although the tumour appeared encapsulated in some areas (Figure 2), in most parts no capsule was seen on histological examination.

A nasal polypectomy was carried out a year later. The tumour has not recurred during the four-year follow-up period.

Discussion

The term myxoma was first introduced in 1871 by Rudolph Virchow (1821–1902), to describe tumours whose histological appearance was similar to the mucinous tissue of the umbilical cord (Canalis *et al.*, 1976). Many soft tissue sarcomas may develop areas of myxoid degeneration leading to an erroneous diagnosis of myxoma. This difficulty was first recognized by Stout (1948) who elaborated clear guidelines for the diagnosis of myxomas.

Myxomas of the head and neck region are rare. Canalis *et al.* (1976) encountered only 10 such lesions over a 20-year period at the UCLA centre for the Health Sciences. Tse and Vander (1985), found only 42 cases of soft tissue myxomas of the head and neck region reported in literature. Of these, 10 were in the palate and 6 in the parotid. Since this report, we have found 16 more cases in the literature. A revised table citing all soft tissue myxomas reported to date is presented here, including our case (Table I).

Although five tumours have been reported in the subcutaneous tissue of the lateral neck and three in the oropharynx there is no mention of a myxoma in the retropharyngeal space.

On gross examination the lesion was found to be slimy, greyish, nodular, avascular in appearance and only partly encapsulated. The consistency of the tumour varied slightly depending upon the proportion of fibrosis. On microscopy the tumour consisted predominantly of an abundant mucoid or myxoid intercellular matrix in which there were stellate or spindle-shaped cells and haphazardly directed reticulin fibres. The mucoid material stained positively with alcian blue mucicarmine and colloidal iron stains could be removed by prior treatment of the sections with hyaluronidase (Enzinger and Weiss, 1988). The tumour was poorly circumscribed lacking a capsule in most parts and tended to infiltrate the surrounding soft tissues or bone.

Myxomas have been described as neoplasms of the primitive

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TABLE I
MYXOMAS OF THE HEAD AND NECK

Location	No. of cases
Palate	10
Parotid	6
Scalp	3
Cheek	3
Lateral neck	5
Lip	4
Alveolar ridge	2
Oral	5
Oropharynx	3
Larynx	3
Face	2
Masseter	2
Chin, tongue, conjunctiva, buccal mucosa, interdental gingiva, retromolar pad, genio-hyoid muscle, eyelids, external auditory canal, laryngo-pharynx, cornea	1 of each
Retropharyngeal (present case)	1
Total	60

mesenchyme. Wirth *et al.* (1971), proposed that a basic error in tissue metabolism resulted in these tumours. Enzinger (1965) suggested that the stellate myxoma cells were actually altered fibroblasts secreting mucoid material instead of collagen. Ultrastructural examination of the tumour cells revealed an abundant, rough, endoplastic reticulum and cytoplasmic fibrillar material characteristic of fibroblasts (Harrison, 1973). A viral origin has also been suggested (Glazunov and Puckhov, 1960) based on the finding of nuclear and cytoplasmic inclusion bodies in certain intramuscular myxomas. Even a traumatic origin has been proposed (Enzinger, 1965; Whitman *et al.*, 1971) and Batsakis (1979) doubted the true neoplastic nature of the tumour.

It is important not to confuse myxoma with myxoid degeneration in soft tissue sarcomas and neurofibromas. Soft tissue sarcomas have abundant mitotic activity, rich vascularity and infiltration by inflammatory and mast cells. All of these are absent in myxomas, whose cells have a benign and bland appearance. Neurofibromas have neural elements such as Schwann cells and nerve cells. Also to be distinguished from myxomas are focal mucinous degenerations of the skin or soft tissues e.g. ganglions and follicular mucinosis, which lack the reticulin fibre network seen in myxomas (Tse and Vander, 1985).

From the management point of view, these tumours are best considered as benign but locally aggressive. Lack of encapsulation and tumour infiltration into neighbouring tissues are regularly seen, leading to recurrence of the tumour if merely enucleation is carried out. A conservative surgical resection with a margin of healthy surrounding tissue is the optimal management. Recurrences, if they do occur, are seen mostly in the first two years. Radiation has been used to control these lesions but has never been found to be satisfactory (Dutz and Stout, 1961; Attie *et al.*, 1966).

In the case presented here enucleation was the treatment of choice because of the locality of the tumour. The patient was followed up for four years and no recurrence was seen. We recommend that in cases where a retropharyngeal mass is encountered, thorough radiological investigations, to exclude a spinal connection, should be carried out and the differential diagnosis of a myxoma considered.

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