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Malignant mesenchymoma of the retropharyngeal space

H. K. LEONG, F.R.C.S.,* C. Y. W. KWAN, F.R.C.P.A.,† R. E. STANLEY, F.R.C.S.,‡ S. TOW, M.B.B.S.,** (Singapore)

Abstract
Malignant mesenchymoma is a very rare head and neck tumour. To date only 15 cases have been reported in world literature and all in children under 16 years of age. We present here a case of a 40-year-old man with malignant mesenchymoma of the retropharyngeal space. The clinical picture is that of progressive dysphagia, voice change, snoring and dyspnoea. CT scan showed a soft tissue space-occupying lesion of the retropharyngeal space which enhanced very well with intravenous contrast. The tumour was excised *in toto* and the patient given post-operative radiotherapy. Histopathology showed two unrelated differentiated tissue types (bone and fat) in addition to the fibrosarcomatous element thus satisfying Stout's criteria (Stout, 1948) for a diagnosis of malignant mesenchymoma.

Key words: Mesenchymoma, malignant; Neck

Introduction

Malignant mesenchymoma is a name given to an intriguing but small group of malignant soft tissue tumours that do not fit into any other tumour category and are characterized by the presence of two or more different tissue components in the same neoplasm. Stout (1948) further defined this sarcoma as a malignant tumour in which there are two or more types of unrelated, undifferentiated tissues in addition to the fibrosarcomatous elements.

After a thorough review of world literature in English, we only found 15 cases of malignant mesenchymoma arising in the head and neck. Of these, six cases arose in the neck and all in children below 16 years of age.

This report describes a case of malignant mesenchymoma in a 40-year-old man presenting as a space-occupying retropharyngeal tumour. This is the first reported case of malignant mesenchymoma of the head and neck in Singapore.

Case report

A 40-year-old man who complained of progressive dysphagia, dyspnoea, snoring and voice change for two months was seen in the Department of Otolaryngology, Singapore General Hospital on 7 March 1992. He was noted to have a 'hot potato' voice on presentation.

Clinical examination of his pharynx showed the posterior oropharyngeal wall to be markedly swollen with congested overlying mucosa. The swelling significantly narrowed the oropharyngeal lumen and rendered mirror examination of the larynx impossible. A flexible nasolaryngoscope was able to bypass the swelling, revealing a normal larynx with normal mobile folds. There was no cervical lymphadenopathy. Complete ENT examination showed no other abnormalities.

An initial plain lateral X-ray of the neck showed a markedly widened retropharyngeal soft tissue shadow between C2 and C6.

He was admitted for observation in view of the danger of airway obstruction. Laboratory data including complete blood count, white cell differential count, liver function tests and urinanalysis were within normal limits. Chest X-ray showed minimal fibrotic changes of the left lung apex characteristic of old pulmonary tuberculosis with no other abnormality.

CT scan of his neck (Figure 1) showed a large mass in the retropharyngeal region measuring approximately 4.5 cm in the widest transverse diameter. It appeared hypodense to isodense compared with adjacent soft tissue and contained areas of calcification. There was no involvement of the adjacent vertebrae of the neurovascular bundles on either side but the tumour enhanced very well with intravenous contrast. This raised the possibility of a very vascular lesion and angiography was therefore arranged to identify the feeding vessels with a view to embolization. However, no obvious feeding artery was identified at angiography from the common carotid or vertebral arteries on either side (Figure 2).

The patient underwent excision of the retropharyngeal tumour *via* an oblique neck incision (Figure 3). At operation, a well circumscribed tumour measuring 5 cm in diameter was enucleated from the retropharyngeal space without difficulty and with minimal blood loss (Figure 4).

On sectioning, the tumour had a firm homogeneous beige-coloured appearance. Light microscopy showed a circumscribed but nonencapsulated soft tissue tumour with mixed histological features. Most of the tumour was composed of plump fusiform cells resembling primitive mesenchyme, arranged in distinct whorls and short fascicles. These cells had pale vacuolated cytoplasm and vesicular nuclei. In some areas the tumour showed fibroblast-like cells with a storiform pattern (Figure 5). Mitotic figures were common and the mitotic index was about 6/10 high power field in the most active areas. There were small areas of lipoblastic and lipocytic differentiation (Figure 6). Benign bone and osteoid formation within the other two component types were present (Figure 7). There was no evidence of mucin secretion or epithelial differentiation. Immunostains revealed positivity for vimentin and focal positivity for S100 protein but the tumour was negative for AE1 and 3, CAM 5.2. desmin and factor VIII. A diagnosis of malignant mesenchymoma was made based on the presence of a predominant fibrosarcomatous component

From the Department of Otolaryngology*, National University Hospital, Mount Elizabeth Laboratories† and the Departments of Otolaryngology‡ and Pathology**, Singapore General Hospital.
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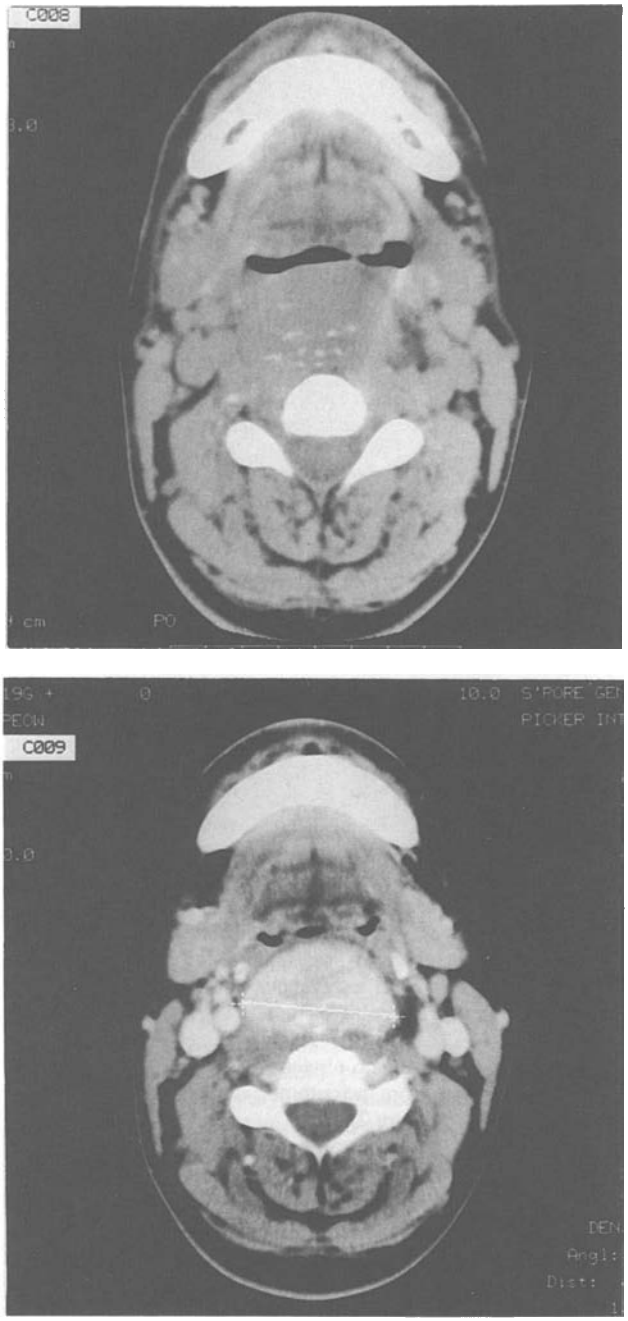

FIG. 1

CT scan demonstrating tumour: (Top) with areas of calcification in unenhanced scan; (Bottom) strong enhancement with intravenous contrast.

together with two unrelated differentiated tissue types which in this case consist of bone/osteoid and lipocytic/lipoblastic differentiation.

Post-operatively, the patient made an uneventful recovery. He underwent external beam radiation therapy of 55 Gy over six weeks. At six months after excision there was no clinical evidence of recurrence.

Discussion

Stout (1948) first coined the term malignant mesenchymoma and defined it as a 'malignant tumour showing two or more unrelated, differentiated tissue types in addition to the fibrosarcomatous element'. He applied this term to a wide variety of neoplasms and subsequent to his initial report of eight cases in 1948: he had collected 335 cases by 1959. In the experience of

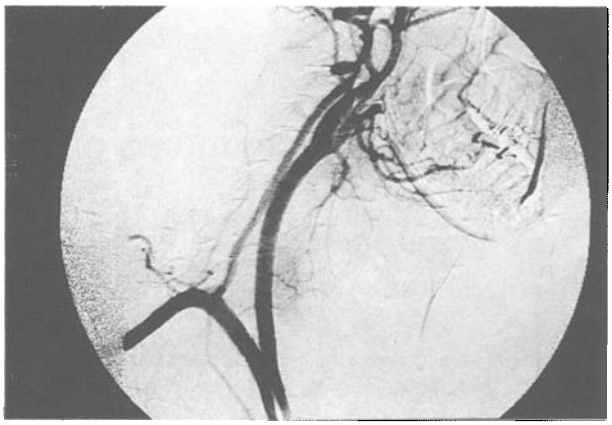

FIG. 2

Digital subtraction angiogram with dye injected into brachiocephalic trunk.

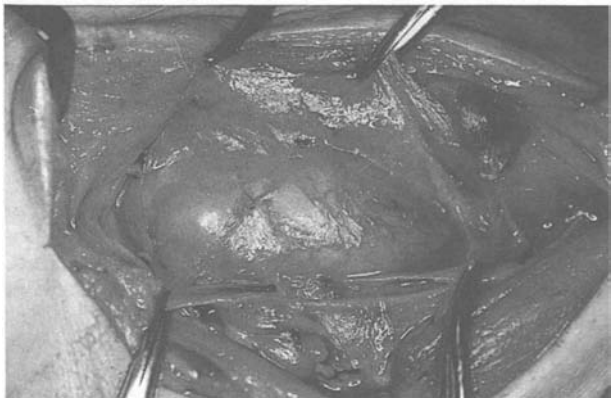

FIG. 3

Tumour in retropharyngeal space as approached from the right neck.

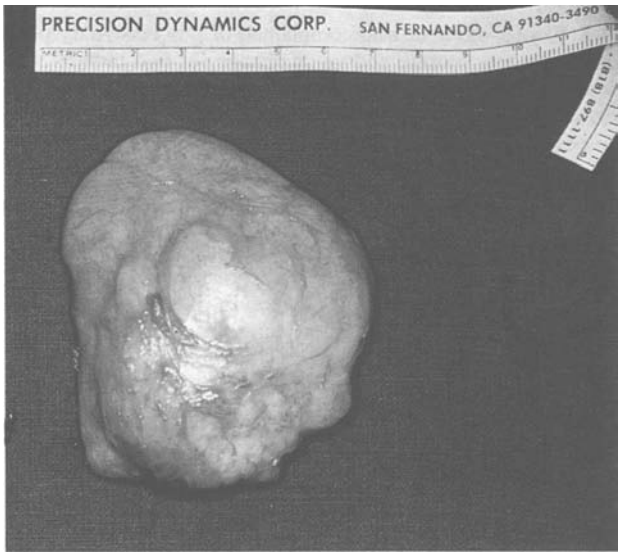

FIG. 4

The excised specimen.

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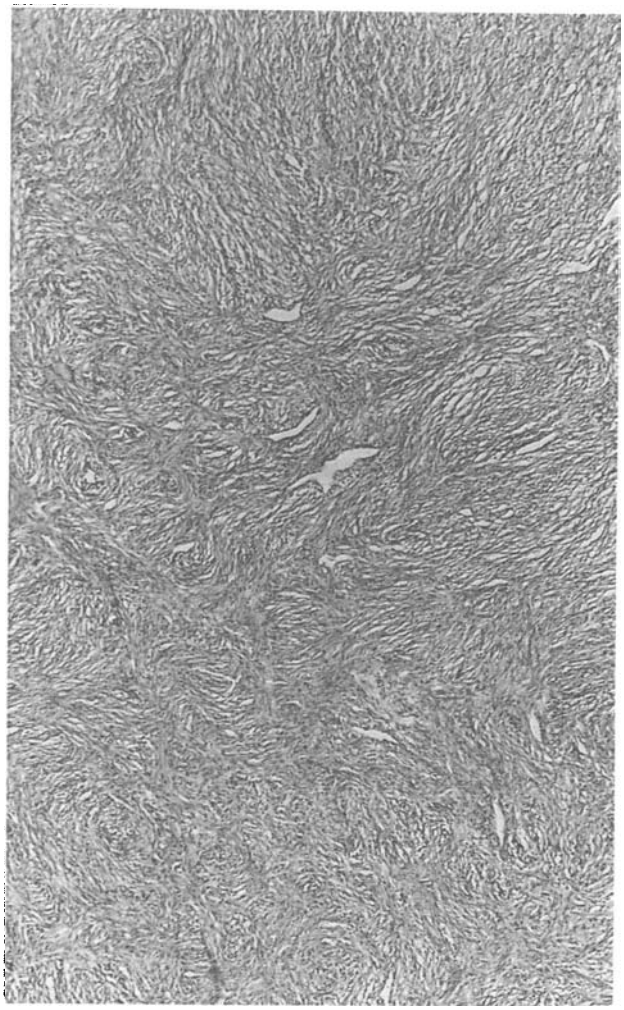

FIG. 5
Photomicrograph showing an area of differentiation with a storiform arrangement. (H & E; × 40).

others, this is a rare tumour and there is little information about it in the literature (Enzinger and Weiss, 1988).

The majority of these tumours occur in patients over 55 years of age and only a few affect children and young adults (Enzinger and Weiss, 1988).

The vast majority of malignant mesenchymomas arise in the retroperitoneum and thigh and only rarely are they encountered in the head and neck region. Altogether six other cases of this tumour were reported in the neck, all in children younger than 16 years of age (Stout, 1948; Nash and Stout, 1961; Mayer *et al.*, 1974; Senoo *et al.*, 1983), and none of these as a midline retropharyngeal mass. Other sites reported in the head and neck region are shown in Table I. According to these references, the malignant elements of the tumour were most frequently rhabdomyosarcoma, haemangiopericytoma and leiomyosarcoma. This is the first reported case of a malignant mesenchymoma presenting as a retropharyngeal tumour. Its radiological feature of enhancement with contrast on CT scan, with no vascularity on angiography was also first noted in a malignant mesenchymoma.

Histological examination of our case showed two unrelated differentiated tissue types in addition to the prominent fibrosarcomatous element. Firstly, small areas of lipoblastic and lipocytic differentiation were found. Secondly, there was benign bone and osteoid formation and these showed up as areas of calcification on the CT scan. The tumour presented in this report therefore adequately fulfils the criteria of malignant mesenchymoma which Stout (1948) first established. A differential diagnosis of liposarcoma, composite type was considered, in of the

TABLE I
MALIGNANT MESENCHYMOMA OF THE HEAD AND NECK REGION OTHER THAN THE NECK

Site	No. of cases	Author
Nasal cavity/nasopharynx	3	Nash and Stout (1961)
Face	1	Ashbell *et al.* (1972)
Oral cavity	2	O'Day *et al.* (1964)
Larynx	1	Kawashima *et al.* (1990)
Unknown	2	Gerbaulet *et al.* (1985)

presence of lipocytic and lipoblastic cells. The bone/osteoid formation may then be taken as a metaplastic phenomenon. However, fat stains were unsatisfactory and unequivocal positive-staining was not demonstrated. Considering the predominance of the fibrosarcomatous component and other features discussed above, we favoured the diagnosis of a malignant mesenchymoma.

The histogenesis of malignant mesenchymoma is uncertain. It has been postulated to arise from primitive mesenchymal elements which differentiated along multiple cells lines. Prognosis is usually determined by the predominant component and hence survival rates vary from case to case. It is usually best in tumours with a predominantly liposarcomatous element and worst in those with a predominant rhabdomyosarcomatous component. Mode of treatment is selected based on the predominant and

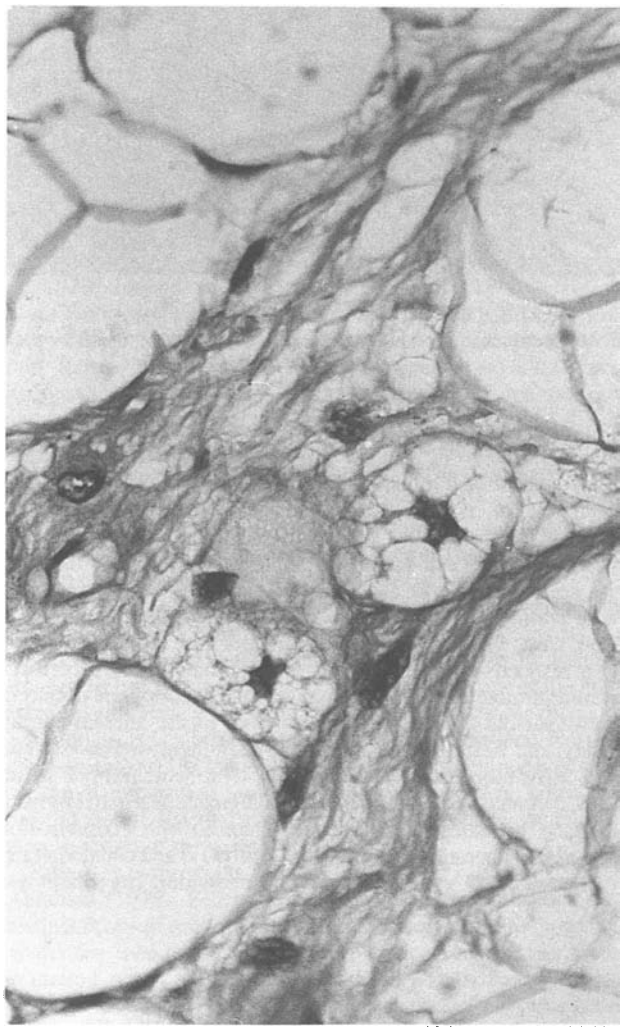

FIG. 6
Photomicrograph showing lipocytic and lipoblastic component of the tumour. (H & E; × 400).

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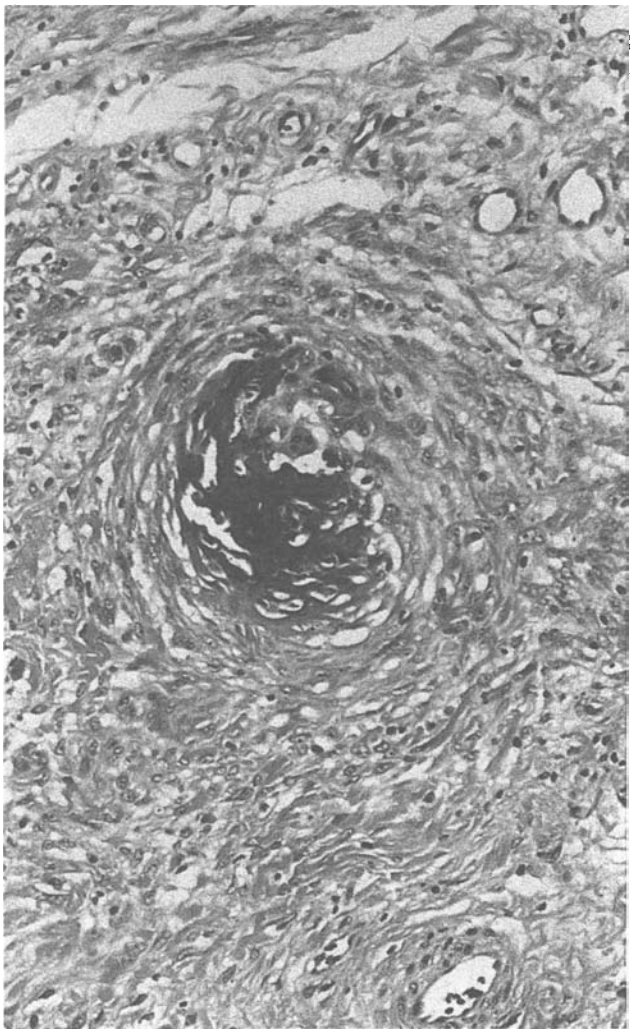

FIG. 7

Photomicrograph showing metaplastic bone formation with fibrosis. (H & E; × 100).

most poorly-differentiated component (Enzinger and Weiss, 1988). In our case, the treatment of choice is complete surgical removal. In view of the known risk of local recurrence and metastasis, combined modality treatment with radiotherapy and/or chemotherapy is often used. Nash and Stout (1961) treated two cases arising in the neck by simple excision alone. The first case was a reticulum cell sarcoma with haemangiopericytoma while the second case was a congenital malignant mesenchymoma with leiomyosarcoma and haemangiopericytoma components. Both cases were alive at eight years and nine months after surgery respectively. The authors also in the same report recorded a similar tumour (leiomyosarcoma and haemangiopericytoma) in a 14-year-old boy, which was adherent to the internal carotid artery, and treated with surgery and radiotherapy. The boy was still alive more than 13 years after treatment. Ashbell *et al.* (1972) treated a case of congenital malignant mesenchymoma of the face (liposarcoma, rhabdomyosarcoma) with a combination of surgical operation, irradiation (4 650 cGy) and chemotherapy (actinomycin D, vincristin, cyclophosphamide). The patient was alive six months later. Myer *et al.* (1974) combined surgery and chemotherapy (vincristin, cyclophosphamide, actinomycin D) in the management of a case of malignant mesenchymoma of the neck (chondrosarcoma, rhabdomyosarcoma). The chemotherapy was repeated after 12 months. The child showed no evidence of recurrence 44 months after initial diagnosis. Gerbaulet *et al.* (1985) used radiation therapy (Iridium 192 in interstitial implants) in addition to the surgery and chemotherapy protocol used by Mayer *et al.* (1974). Their two cases were still alive and disease-free two and 5.5 years respectively after treatment. O'Day *et al.* (1964) however treated two cases of soft palate and inferior labial fold malignant mesenchymoma (osteogenic sarcoma with osteoid production and liposarcoma) with surgery and irradiation. These two patients died after 10 and 20 months respectively.

In view of the experience reported in these cases and the fact that it is impossible to obtain a wide margin of excision in the retropharyngeal space, we decided that post-operative radiation therapy should be carried out to reduce the chance of recurrence in our patient.

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Address for correspondence:
H. K. Leong,
Senior Lecturer,
Department of Otolaryngology,
National University Hospital,
5 Lower Kent Ridge Road,
Singapore 0511.