

Short reports

Liposarcoma of the mediastinum: report of two cases and review of the literature

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Liposarcoma is a rare tumour accounting for only 15-20% of sarcomas. They usually originate in the interfascial planes of the legs or retroperitoneum.¹ Even rarer is mediastinal liposarcoma of which only 53 cases have been reported.²⁻⁴ This paper concerns two cases seen at the Brompton Hospital in April 1980.

Case reports

CASE 1

A 40-year-old man presented with an enlarging mole on his anterior chest. The chest radiograph revealed a large lobulated mass to the right of the mediastinum. His only symptoms were tiredness for two months and facial swelling upon severe exertion. On examination he looked well and had no signs of superior vena caval obstruction. There was a 1.5 cm flaking mole on his anterior chest. At his right apex there was dullness to percussion and decreased breath sounds.

The skin lesion was completely excised and was found to be a superficial spreading melanoma. Anterior mediastinotomy via the second right costal cartilage revealed a lobulated fatty extrapleural mass that pushed the superior vena cava and azygos vein to the anterior chest wall. Frozen section could not distinguish between lipoma and liposarcoma.

Ten days later a well-differentiated myxoid liposarcoma weighing 1.6 kg was removed via a right thoracotomy (fig 1). The superior vena cava was encased by the fatty tumour which was removed piecemeal. The superior vena cava was narrow but patent at the conclusion of the procedure. On the evening of the operation the patient developed venous obstruction and was treated with warfarin. Over the course of two weeks the venous distension and upper extremity oedema subsided.

At follow-up six weeks later the caval obstruction had resolved. At 10 weeks a CT scan showed possible residual tumour in the region of the innominate vein and the right border of the trachea. He is now undergoing radiotherapy to that area.

CASE 2

A 43-year-old man presented with a two-month history of non-productive cough. On examination he had non-pulsatile jugular venous distension without upper limb oedema and dullness to percussion over the right anterior

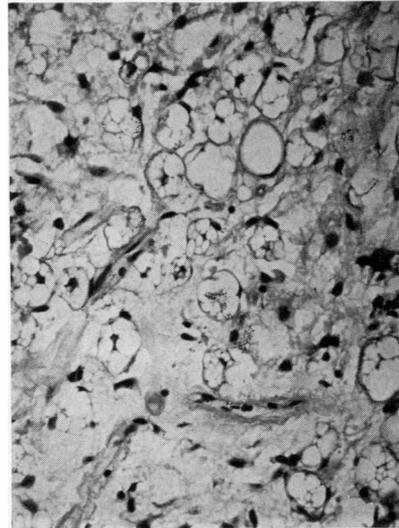


Fig 1 Well-differentiated liposarcoma consisting of vacuolated lipoblasts in a myxoid stroma. Haematoxylin and eosin, original magnification $\times 380$.

chest. A chest radiograph showed a large anterior mediastinal mass extending laterally to both hila. At bronchoscopy the carina was widened.

Right anterior mediastinotomy revealed a fatty lobulated mass. The pathological diagnosis was well differentiated myxoid liposarcoma. Two weeks later the bulk of the tumour was removed piecemeal through a vertical median sternotomy but some tumour remained at the right cardiophrenic angle. The superior vena cava and innominate vein were freed of tumour, and this caused a marked fall in venous pressure. The tumour weighed 1.4 kg and had areas of undifferentiated highly anaplastic cellularity (fig 2). Five weeks later there was bilateral hilar shadowing suggestive of recurrence. The patient was referred for radiotherapy but despite this had rapid regrowth of the tumour in both chest cavities and out through the sternal wound. He died eight weeks after the operation.

Discussion

The age range of patients with mediastinal liposarcoma is

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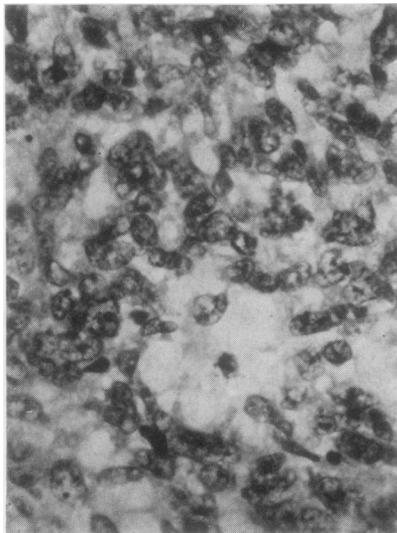


Fig 2 Anaplastic sarcoma with multinucleated giant cells which in a few other areas showed lipoblastic differentiation. Haematoxylin and eosin, original magnification $\times 900$.

from 14 to 77 years (mean 45 years). They usually present with dyspnoea, wheezing, chest pain, cough, or weight loss, but may be asymptomatic.⁴ Our first patient was discovered on routine chest radiography but the second had symptoms and signs of local compression. Mediastinal liposarcomas often achieve great size and weights of 7 kg have been reported.

Radiographically the density of liposarcoma is similar to fat. Using a CT scanner, Mendes distinguished between lipoma and liposarcoma, showing the EMI density of the

tumour to be midway between fat and water.³ It may be impossible to differentiate between lipoma and liposarcoma on a frozen section and so definitive histology may have to wait until after tumour excision.

There are two clinical groups of mediastinal liposarcoma. Microscopic examination of tissue from the first group reveals embryonal lipoblasts and mature fat cells in a myxoid stroma with few mitoses (fig 1). There may be a pseudo capsule which often contains tumour; these tend to grow locally. The four longest disease-free survivors previously reported (3 to 17 years after excision) had this structure.⁴ Case one represents this variety. The second type has a very malignant appearance with highly anaplastic cells, bizarre giant cells and many mitoses without mature fat cells (fig 2). The longest reported survivor in this group is two years. Often death comes sooner, as in case two, as a result of liver or lung metastasis or local recurrence.⁴

Chemotherapy has been of no value in treatment, and although radiotherapy without excision has been used in a few patients, no tumour regression has been seen. Mediastinal liposarcomas lack discrete boundaries, making total excision difficult, but removal of the bulk of the tumour can give symptomatic relief. The primary treatment in both clinical varieties should be as complete a resection as possible followed by radiotherapy.

Late recurrences of well-differentiated tumours may be resected again with benefit in some cases.

References

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