Long term survival in non-encapsulated primary liposarcoma of the mediastinum

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Abstract
An elderly woman presented with dyspnoea secondary to extensive mediastinal invasion by a primary well differentiated liposarcoma of the mediastinum. Five years after partial resection and postoperative radiotherapy (45 Gy) she is alive and well. To our knowledge, long term survival has not previously been reported for a non-encapsulated mediastinal liposarcoma treated by incomplete resection and subsequent radiation therapy.

(Thorax 1993;48:1276–1277)

Primary liposarcomas are relatively uncommon neoplasms, occurring most commonly in the lower extremities and retroperitoneum.1 We are aware of only 60 patients with primary liposarcoma in the mediastinum.2-5 Only five of these patients, each with a well encapsulated tumour, have been reported as free of disease after surgical excision.3,5 Whenever possible, complete surgical excision is the recommended mode of treatment.3,5

We present a patient with a primary mediastinal liposarcoma which was characterised by extensive local invasion. Although only partial resection was possible, after postoperative radiotherapy the patient is alive and well five years later.

Case report
A 75 year old previously healthy woman presented in May 1987 with a 10 month history of gradually progressive dyspnoea on exercise. Respiratory rate was 22/minute. Physical examination was otherwise normal, and there was no evidence of superior vena cava obstruction. The results of routine blood tests, electrocardiography, and barium swallow were normal. Chest radiography revealed a large anterior mediastinal mass mimicking a very large heart and also widening the superior mediastinum to the right of the trachea. Computed tomographic scanning (fig 1) showed the mass was of predominantly fatty attenuation. Arterial blood gas (room air) pH was 7.42, Pco2 5.2 kPa, and Po2 8.4 kPa. Forced expired volume in one second (FEV1) was 0.8 litres (40% predicted), forced vital capacity (FVC) was 0.8 litres (30% predicted), and FEV1/FVC ratio was 96%.

A median sternotomy and bilateral anterior thoracotomy were performed in July 1987. A large anterior mediastinal fatty tumour was found containing numerous hard foci, each measuring 1–3 cm in diameter. Gross invasion of the pericardium and left atrium was evident. The tumour encased the left phrenic nerve. Foci of invasion of the right lung, aortic arch, aortopulmonary window, and left pulmonary artery were also present. Approximately one quarter of the total tumour mass (450 g) was resected from the anterior mediastinum. Additional resection was considered impossible because of invasion of adjacent structures. The extent of the mass appeared unchanged on postoperative chest radiographs.

On gross pathological examination the tumour was soft, fatty and contained ill defined regions of firmness. Microscopic examination revealed well differentiated liposarcoma. In many regions, large fat cells showed only minor variations in size and nuclear atypia. In other regions, lipoblasts with irregular hyperchromatic nuclei and cytoplasmic lipid droplets were seen (fig 2). Foci that felt firm on gross examination corresponded to areas of atypical spindle cells amidst fibrosis on histopathological examination. Special stains for mucin (mucicarmine, periodic acid schiff (PAS), and PAS with diastase) were negative. Immunoperoxidase stains for the epithelial markers cytokeratin and epithelial membrane antigen were negative. An S-100 stain was positive both in the

Figure 1 Anterior mediastinal mass in a 75 year old woman. Computed tomographic scan showing a broad mass of adipose attenuation laced throughout by ill defined strands of soft tissue attenuation. The mass displaces the ascending aorta posteriorly.
identified easily by its low attenuation levels (–70 to –130 Hounsfield units). On
computed tomographic scanning attenuation of a liposarcoma varies from predominandy that
of normal fat to soft tissue levels. Of the computed
tomographic appearance correlates closely with the gross and microscopic
anatomical findings. Poorly differentiated
tumours, which tend to be quite cellular, have
mean attenuation levels (15 to 25 Hounsfield
units) approaching that of other solid
tumours. Magnetic resonance imaging is
also accurate in identifying fatty tumours; however,
it is less anatomically precise than
computed tomographic scanning.

Liposarcomas are classified histologically
into four main types: (1) well differentiated;
(2) myxoid; (3) round cell; and (4) pleomorphic. For
the lower extremities and
retrouteritoneum the five year survival for
patients with well differentiated and myxoid
liposarcomas is about 80%, whereas for
round cell and pleomorphic types it is around
20%. For the mediastinum no long term sur-
vivor to our knowledge has had other than a
well differentiated, encapsulated tumour.

Radiotherapy has generally produced poor
results in treating liposarcoma of the medi-
astinum compared with liposarcoma in other
sites. Large doses of radiation (approxi-
nately 90 Gy) are believed to be necessary to
attain remission, but the risk of mediastinal
fibrosis precludes giving such high doses to
patients with mediastinal liposarcoma. The
dose of radiation used in the mediastinum has
been variable but is generally less than 60 Gy.
We are unaware of any previous report of
prolonged survival in a patient with medi-
astinal liposarcoma treated with radiation.

To our knowledge this is the first report of
a documented long term survivor with a non-
capsulated, diffusely infiltrating mediastinal
liposarcoma. We suggest that treatment with
subtotal resection of the tumour (‘debulk-
ing’) followed by radiation therapy be con-
sidered for this entity.

Discussion

The spectrum of mediastinal liposarcoma
varies from well differentiated and encapsu-
lated to pleomorphic and non-encapsulated.
The reported age range varies from nine
months to 77 years, but most patients with
this disease are over 40 years.

Liposarcomas have been described in all
mediastinal compartments, most commonly
in the posterior mediastinum. These
tumours can attain massive sizes and weigh
as much as 7 kg. Common presenting symp-
toms include chest pain, dyspnoea, wheezing,
cough, and weight loss, but patients may be
asymptomatic. Obstruction of the superior
vena cava has been described in association
with early mortality.

The appearance of the chest computed
tomographic scan may be diagnostic. Fat is
normal in the mediastinum and can be

well differentiated area and the less differenti-
ated spindle cell area, consistent with the
diagnosis of liposarcoma.

The patient was treated with 45 Gy ex-
ternal beam radiation therapy to the anterior
mediastinum from July to September 1987.
Dyspnoea increased and a new dry cough
developed. Chest computed tomographic
scanning in October 1987 revealed bilateral
patchy pulmonary opacities. Prednisone (30
mg/day) was instituted for presumed radia-
tion pneumonitis and symptoms improved.
Over the next three months, as the dose of
prednisone was tapered, she reported increasing
injection dyspnoea which again responded to pred-
nisone. She was then maintained on 5 mg
prednisone every other day. Four years later,
in July 1991, FVC was 1.06 litres (39% pre-
dicted and 28% greater than her preoperative
value). The patient continued to do well five
years after initial presentation, with mild
dyspnoea on exertion as her only complaint.
The radiographic appearance of the mass
remained unchanged.

Figure 2  Well differentiated liposarcoma. Atypical hyperchromatic nuclei are abundant. Rare lipoblasts (arrow) are seen (inset). Haemotoxylin and eosin. Magnification × 100
(inset × 400).

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Thorax 1993 48: 1276-1277
doi: 10.1136/thx.48.12.1276

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