

Liposarcoma

Definition

Primary malignant tumour arising from fat.

Epidemiology

Age 40-60

Males > females

Site: thigh and retroperitoneum. Rare in bone.

Pathogenesis

Myxoid liposarcoma has a specific cytogenetic marker; reciprocal translocation of chromosome 2 with 13. Pleomorphic liposarcoma does not.

It is controversial whether or not a lipoma can undergo malignant transformation; if this does occur it is rare.

The tumours arise from primitive mesenchymal cells rather than mature lipocytes which accounts for its rarity in subcutaneous tissues and fatty bone marrow.

Clinical presentation

Slow-growing, deep-seated ill-defined mass that is seldom tender or painful. Because of the lack of complaints and because it is commonly found in areas with plenty of room to expand, it can reach a huge size (e.g. Mr. McGrath with a 40cm long tumour in his thigh which weighed 7 kg).

Radiology

Plain XR

The radiodensity approaches that of adjacent muscle so liposarcomas appear just as soft tissue swellings.

Calcification is rare and implies synovial sarcoma.

The rare liposarcoma arising in bone has a classical destructive permeative appearance with no special features.

Bone scan

An unusual feature is the high intensity on bone phase which may be due to the high amount of ionised calcium in the lesion.

CT

The lesion is less radiodense on CT than the surrounding muscle.

Contrast causes enhancement.

MRI

Generic changes of sarcomas, viz low to medium intensity on T1 and high intensity on T2.

Lobulation is a fairly constant feature.

Pathology

Gross

The mass often appears to be deceptively well encapsulated and can easily be shelled out from the surrounding tissues.

It appears lobulated.

The cut surface of a myxoid liposarcoma shows a soft myxomatous tissue, which sometimes can resemble fat, but more usually is composed of immature lipoblasts with a tissue that appears richly vascular, soft and gelatinous.

Pleomorphic liposarcomas appear infiltrative and fatty, and have areas of cystic degeneration and haemorrhage.

Satellites about the periphery of the mass are often seen about both types of liposarcoma

Histology

1. Myxoid
 - a. Sheets of lipocytes and or lipoblasts interspersed in a myxomatous amorphous matrix. Relatively hypocellular.
 - b. Coursing through this tissue are myriads of fine branching capillaries which give the appearance of a detailed roadmap.
2. Pleomorphic liposarcoma
 - a. Composed of tissue that is much more cellular than myxoid liposarcoma
 - b. The tissue lacks the branching capillaries.
 - c. There are large, bizarre lipoblasts which resemble the bizarre cells of rhabdomyosarcoma and MFH. If not for the obvious fat cells, this may make distinguishing between them difficult.

Special stains

If diagnosis is difficult due to a lack of differentiation, then staining with Oil red O may be useful.

EM

EM may reveal intracytoplasmic pools of lipid.

Immunohistochemistry

Liposarcoma stains positively only against the S-100 antigen.

This is not particularly useful because sarcoma of cartilaginous, neural and muscle origin also stain S-100 positive.

Treatment

Surgery

Low-grade stage 1 lesions require either a wide marginal, or a marginal excision following satisfactory response to preoperative radiotherapy.

Stage II lesions require either radical margins or wide marginal excision following radiotherapy.

Radiotherapy

Used extensively preoperatively to facilitate limb salvage resection if the functional results will be better than with more aggressive surgery alone.

Chemotherapy

Used for treatment of Stage III disease and by some prophylactically for stage II-B disease. There is insufficient experience to know how effective this is.

Prognosis

5-year survival rates for adequately treated myxoid liposarcoma are around 90%
For pleomorphic liposarcoma the figure is around 50%

A peculiar feature, almost unique to liposarcoma, is the high rate of a second lesion in the retroperitoneal region after control of an initial lesion in the lower extremity.