OPEN ACCESS TEXTBOOK OF GENERAL SURGERY

SOFT TISSUE NEOPLASM **D M Dent, E Panieri**

INCIDENCE AND PATHOGENESIS

Soft tissue neoplasms may be benign, locally aggressive, or malignant metastasizing sarcomas.
Soft tissue masses are at least 100 times more common than malignant soft tissue sarcomas
The evaluation and investigations are determined by the need to exclude or confirm the presence of the rarer, but malignant sarcomas.

LIPOMA

These are soft, fatty neoplasms which most typically occur in the subcutaneous tissues. They have an indolent history, and will not cause local symptoms, other than mild discomfort.

They are completely benign neoplasms which will not mutate into malignant sarcoma if untreated.

Management is conservative unless they are large or unsightly, in which case a local excision is curative.

Given that benign, it can be difficult to determine which soft tissue masses warrant further evaluation. The following patients meet criteria for urgent referral with a soft tissue lesion:

- Soft tissue mass > 5 cm (golf ball size or larger)
- Painful lump
- Lump that is increasing in size
- A lump of any size that is deep to the muscle fascia
- Recurrence of a lump after previous excision

DESMOID TUMOURS / DEEP FIBROMATOSIS

Desmoid tumors, also referred to as aggressive or deep fibromatosis, are not sarcomas but they represent neoplasms of fibroblastic tissue that lack the ability to metastasize. However, desmoid tumors have a propensity for local recurrence, even after complete resection, and they have the capacity to cause local morbidity and death in rare cases.

Desmoid tumors typically arise in the extremity in sporadic cases, or in the abdominal wall in association with pregnancy (where they usually occur post-partum) or arise in the mesenteric root in the setting of familial adenomatous polyposis (FAP). Mesenteric desmoids are those with the highest degree of mortality

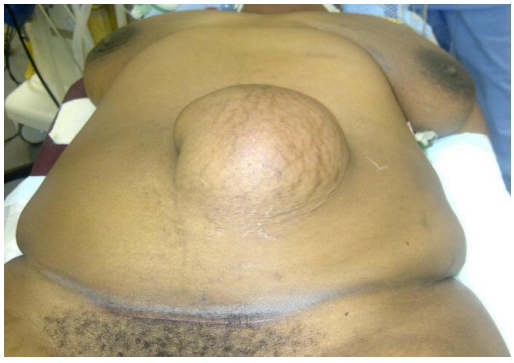
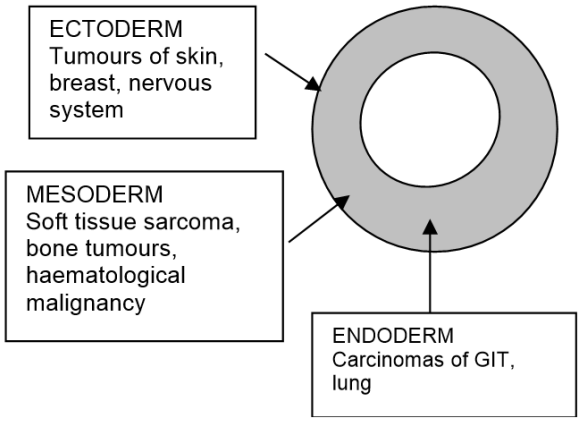


Fig1- *Desmoid tumour protruding through the anterior abdominal wall*

SARCOMA

sarcomas are rare tumours, which comprise about 1% of all malignancies, and are thus equivalent in incidence to that of Hodgkins Disease and malignancy of the tongue and testis. Their incidence is 1/100,00 in adults, being slightly more common in children. There are no sex or racial differences.



They arise from mesodermal tissues (muscle, fascia, fat, and synovium), which make up 50% of the body weight.

In most cases the cause is unknown. There is a genetic association in certain rare disorders such as neurofibromatosis (where neurofibrosarcomas may develop in 15% of cases) and Gardners Syndrome (where Desmoid tumours may develop). Certain acquired conditions may develop sarcomas. The association with HIV infection, and the numerous animal models relating animal sarcomas, oncogene expression and viruses hint at a viral origin.

Sarcoma associations

- multiple neurofibromatosis
- Gardners Syndrome
- Li-Fraumeni Syndrome
- lymphoedema
- irradiation
- HIV infection (Kaposi Sarcoma)

Sites of development

Approximately 80 percent of sarcomas originate from soft tissue and the rest from bone. The frequency of occurrence relates to the sites of the bulk of soft somatic tissue. Thus, most tumours are located in the buttock and upper thigh, the shoulder girdle or retro-peritoneum.

Thigh, buttock, and groin	46%
Upper extremity	13%
Torso	18%
Retroperitoneum	13%
Head & neck	9%

Table 1- *incidence of sacrcoma in realtion to anatomical site*

Pathological Classification

There are exhaustive pathological classifications of sarcomas.

The most common soft tissue sarcoma subtypes are undifferentiated soft tissue sarcoma (sarcoma NOS),liposarcoma, leiomyosarcoma, synovial sarcoma, and malignant peripheral nerve sheath tumor (MPNST).

In most cases the knowledge of the precise histological type is not important: the diagnosis of type is seldom made clinically (except in the case of neuro-fibromatosis), there is a great variation between pathologists in the diagnosis of the material and many sarcomas are unclassifiable.

Most importantly, it is the *grade* and *stage* that determine treatment, rather than the *type.* However, particularly in children rhabdomyosarcomas are susceptible to chemotherapy, and a preoperative diagnosis is important.

Grades of sarcomas

low grade*:* little cellular atypia, few mitoses, no tumour necrosis

intermediate grade*:* atypia, numerous mitoses, little or no tumour necrosis

high grade*:* necrosis in addition to atypia and frequent mitotic figures.

Clinical features

The majority of cases present as a painless mass. In many patients these have been misdiagnosed as “pulled muscles” or “haematomas”.

In clinical terms, if the body is regarded as having three levels (skin, subcutaneous tissue, deep structures), soft tissue sarcomas arise, and are fixed to deep tissues.

In rarer cases pain may be a presenting feature where there is compression of nerves, joints or muscle; retroperitoneal liposarcomas may present as large intra-abdominal masses.

The size of tumour at presentation would appear to be related to the body awareness of the importance of the site of origin, (and is probably related, in this to awareness of the homunculus in the brain): patients present at an early stage with lesions of the hand and face, less frequently with lesions of the limb, and at a late stage with lesions of the back and buttock.

Clinical presentation

- painless mass in limb, trunk or retro-peritoneum
- pain or immobility of joint
- paraneoplastic phenomena (eg hypoglycaemia)

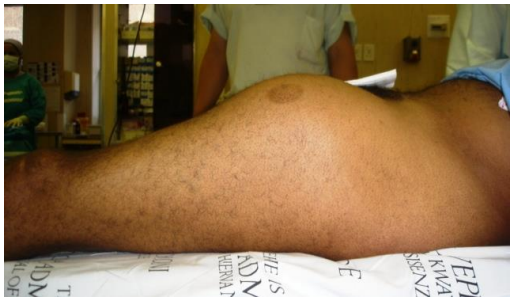


Fig 2- *large mass in the proximal anterior thigh of a patient*

The differential diagnosis is limited, and all soft tissue masses must be regarded as sarcomas until otherwise disproven by investigation. It is common for patients to notice a lump after minor trauma, and to misinterpret it as a"haematoma" or a "bruise"

Differential diagnosis

- haematoma
- ruptured muscle
- benign soft somatic tumour (neurofibroma, lipoma)
- cold abscess
- false aneurysm
- incarcerated hernia

Natural history

Soft tissue sarcomas tend to compress and invade locally, without regard to "anatomical compartments". Metastases are characteristically haematogenous, to the lung, brain, liver and other organs. Unlike carcinomas which have early lymph node dissemination, sarcomas have nodal spread in only 5% of cases. Metastatic spread is frequent on first diagnosis, and relentless local recurrence is a feature of inadequate excision.

Management

Referral

These tumours are rare, and special expertise is required in their management. General practitioners and general surgeons have limited experience of them, and specialist referral is essential, in order that appropriate diagnosis and assessment may be made, and combined expertise utilised.

Specialist clinics should offer the combined services of dedicated surgeons, oncologists, pathologists, orthopaedic surgeons and prosthetists.

Diagnosis and staging

Aspiration cytology may be helpful in excluding haematomas and abscesses, but is inadequate in obtaining the finite diagnosis of a sarcoma.

The diagnosis can only be made by obtaining a generous amount of tissue for **histological evaluation**. This is best done by the surgical team responsible for care of the patient, and

for practical purposes a patient with a suspected sarcoma must not have biopsies attempts at primary care level.

TruCut core biopsy is useful in making the diagnosis of a sarcoma, but is inadequate for determining the grade; it is, nonetheless, the preferred method of diagnosis. On certain occasions *incision or excision biopsy* is used, particularly for small (<5cm lesions), but the route must be able to incorporate the final excisional area should the lesion be a sarcoma.

Staging is performed by a *CT scan* of the area in question, together with that of the liver and lungs (a CT of the lungs may reveal metastases not shown on a radiograph). A *liver enzyme profile* is performed to determine hepatic metastases. A *MRI scan* provides excellent imaging of soft tissue sarcomas. Rarely an arteriogram may be performed to determine the source of the feeding vessels.

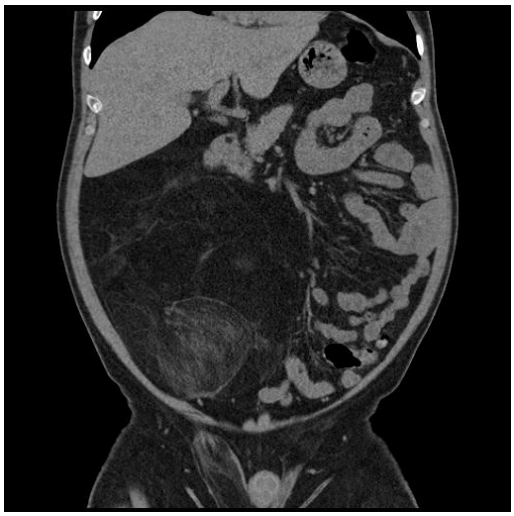


Fig 3- *CTscan showing a massive liposarcoma occupying the right side of the abdomen*

Combined assessment and planning

Patients are best managed by a focused multi-disciplinary team comprising of an oncology surgeon, orthopaedic surgeon, pathologist, oncologist and prosthetist is ideal. Extensive patient counseling is essential.

Staging

Extensive pre-treatment staging is undertaken in order to determine appropriate therapy, to provide prognosis and to compare results. The importance of *grade* in the assessment of soft tissue sarcomas is emphasized in the incorporation of grade into the staging systems. These detailed staging systems are not necessary for the student or practitioner to comprehend, and are given for interest.

TNM staging of soft tissue sarcomas

- **G**(rade) G1 (low), G2 (moderate), G3 (high)
- **T**(umour) 1 (<5cms), 2 (>5cms)
- **N**(ode) N0, N1 (5%)
- **M**(etastases) M0, M1

AJC (American Joint Committee)

- **stage 1**: G1, T1 or T2, N0, M0
- **stage 2**: G2, T2 or T2, N0, M0
- **stage 3**: G3, T1 or T2, N0, M0
- **stage 4**: G1-3, T1-2, N0-1, M0-1

Treatment

Surgery

The fundamental treatment choice of *curative* surgery (no metastases, low grade and small size), or *palliative* surgery must be made. The only cures are found with early disease which has been completely excised.

With *curative* surgery, the lesion must be excisable with clear margins. This would imply wide local excision (without the tumour being transgressed during the surgery), or limb amputation. Curative surgery is frequently quite destructive as entire muscle groups or compartments are removed to achieve a negative margin. Amputation is considered if the excision could not encompass the

tumour, or the residual limb would be without function.

With *palliative* surgery (an incurable lesion, with metastases or extensive local disease) the aim is to provide a comfortable residual life. Local resections (which may leave residual disease) are marginal (local excision of the mass), or intracapsular (de-bulking of the tumour).

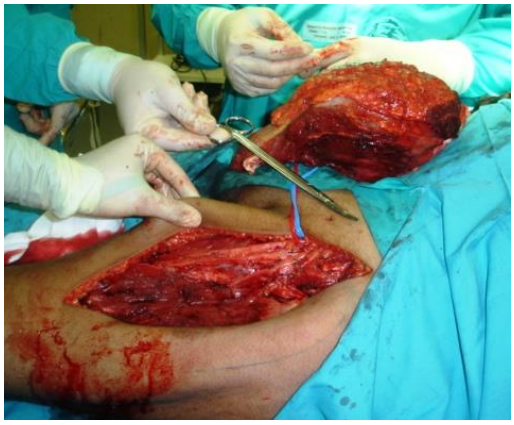


Fig 4- *Surgical image showing the specimen and the surgical field defect*

Radiotherapy

Two approaches may be used: external beam radiotherapy to the excised tumour bed or brachytherapy, where plastic tubes are introduced into the excised tumour bed at the time of surgery, and subsequent radio-active Irridium wires rail-roaded through the tubes post operatively, to provide high dose radiotherapy. Evidence suggests that external beam therapy remains the treatment of choice.

Chemotherapy

Chemotherapeutic regimens are used as *adjuvant* therapy (where the lesion has been excised, but metastatic recurrence is feared), or as palliation. The evidence of absolute benefit for chemotherapy is limited, and cases are best managed by units with expertise in this regard. Some regimens used are: CYVADIC (cyclo-phosphamide, vincristine, doxorubicin, dicarbizine), or cysplatin. The toxicity of chemotherapy for sarcomas is high.

Management summary

- **low grade lesions, <5cms, no spread**
  wide excision alone
- **low grade lesions, >5cms, no spread**
  wide excision + radiotherapy
- **high grade lesions**
  wide excision + radiotherapy + chemotherapy
- **disseminated disease**
  quality of life issues.
  ± ablative surgery, ± chemotherapy, ± node dissection