OPEN ACCESS TEXTBOOK OF GENERAL SURGERY

SOFT TISSUE SARCOMA

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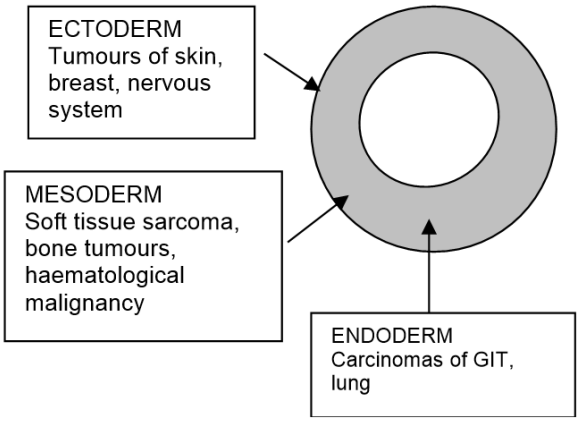
INCIDENCE AND PATHOGENESIS

Soft tissue 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
- lymphoedema
- irradiation
- HIV infection (Kaposi Sarcoma)

Sites of development

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.

Classification

There are exhaustive classifications of sarcomas. 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.*

Table 1: 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. Many patients have undergone treatment for “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 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)

The differential diagnosis is limited, and all soft tissue masses must be regarded as sarcomas until otherwise disproven by investigation.

Differential diagnosis

- haematoma
- ruptured muscle
- benign soft somatic tumour (neurofibroma, lipoma)
- abscess

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.

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

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. *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.

Combined assessment and planning

A combined assessment with a general surgeon, orthopaedic surgeon (where indicated), pathologist, oncologist and prosthetist is ideal. Extensive patient counseling is essential.

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. 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).

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. Some regimens used are: CYVADIC (cyclo-phosphamide, vincristine, doxorubicin, dicarbizine), or cysplatin.

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