Soft Tissue Sarcomas

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- The large majority of soft tissue tumours are benign.
- Malignant mesenchymal tumours are less than 1% of all malignant tumours.
- Multidisciplinary approach.

History:

- Personal history.
- Medical history.
- Mass, change in size of the mass, pain, limb dysfunction, neurologic symptoms, duration of presence of the mass and other symptoms.

Physical examination:

- Tumour size, location, depth.
- Peripheral neurovascular examination.
- Inspection of overlying skin may reveal characteristic skin changes e.g. neurofibromatosis, Kaposi's sarcoma, arteriovenous malformations.

Physical examination:

- Transillumination → cystic nature.
- Auscultation of a bruit → aneurysm.
- Tinel's sign → peripheral nerve tumour.

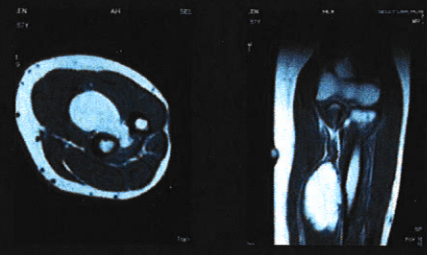
Imaging:

- To define tumour size, location, and extent.
- **Plain radiographs:**
  - Soft tissue shadow.
  - Calcification:
    - Phleboliths in haemangiomas.
    - Calcium deposition in necrotic tissue in STS: synovial sarcomas, liposarcomas and epithelioid sarcomas.
    - Calcified loose bodies in synovial chondromatosis.
  - Ossification: peripheral in myositis ossificans and central in soft tissue osteosarcoma.
  - Reactive changes in the adjacent bone due to external pressure.

Imaging:

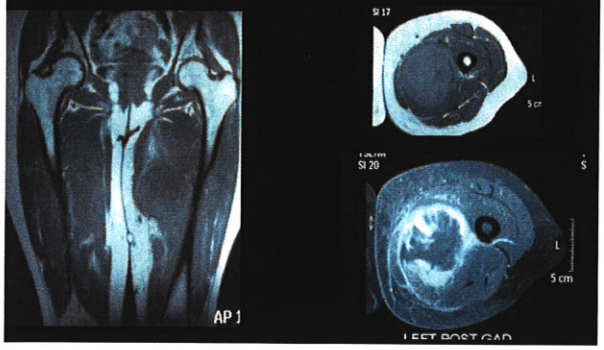
- **Ultrasound:**
  - Differentiation of solid from cystic masses.
  - Ultrasound-guided biopsies.
- **MRI:**
  - Defines the soft tissue extent of the tumour and involvement of adjacent neurovascular structures.
  - Specific diagnoses are possible with MRI (cysts, lipomas, haemorrhage, haematoma, vascular malformations and haemangiomas, PVNS).

Atypical lipoma in the forearm



- Intermediate/locally aggressive. 40-45% of all liposarcomas.
- Adipocytes of different size, nuclear atypia and fibrous septae.
- Deep to the deep fascia or retroperitoneal.
- MDM2 immunopositivity.
- High risk of LR but no malignant potential unless dedifferentiation occurs.

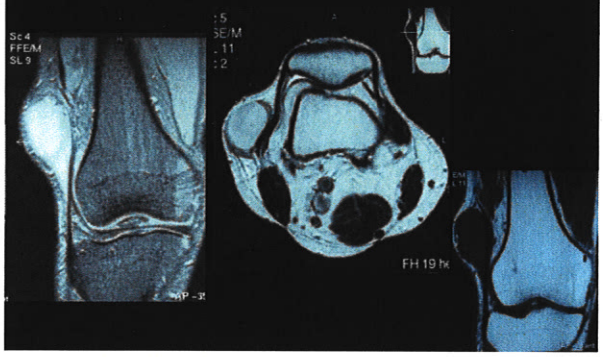
Pleomorphic Sarcoma in the adductor compartment



Myxofibrosarcoma:

- One of the most common sarcomas in elderly patients (6th-8th decade). Mostly in limbs. 50% dermal/subcutaneous.
- Subcutaneous very infiltrative.
- Repeated local recurrence in 50-60%. Progression to higher grade with recurrences.
- Metastasis in 20-35% of cases (pulmonary, osseous, lymphatic). Overall 5-year survival 60-70%.

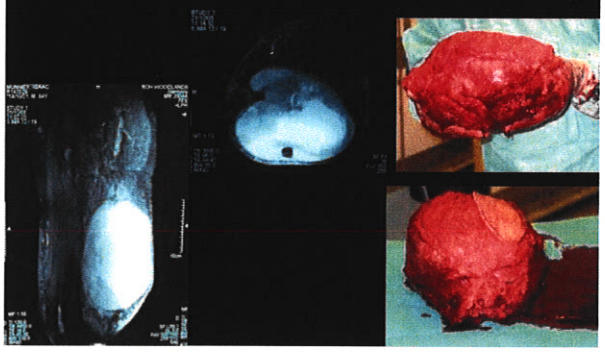
Myxofibrosarcoma in the thigh



Myxoid liposarcoma:

- Primitive mesenchymal cells + lipoblasts in myxoid stroma.
- 10% of all STS and 1/3 of all liposarcomas. Deep, large, painless mass. Two thirds in the thigh. Young adults.
- Hypercellular or round cell morphology (>5%) is associated with poorer prognosis.
- Unusual metastatic pattern.
- Preoperative Radiotherapy advisable.

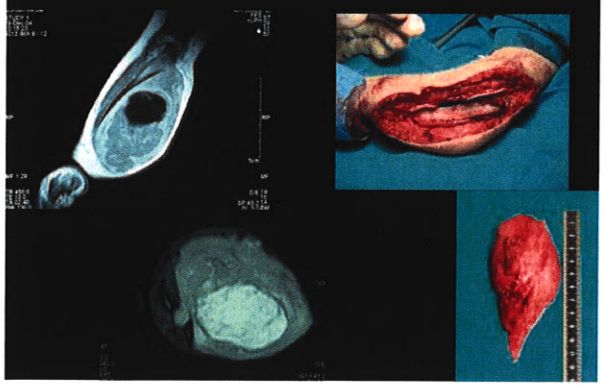
Myxoid liposarcoma in the hamstring compartment



Rhabdomysarcoma:

- Most common STS in children and adolescents.
- **Embryonal:** Most common, <10years, head and neck, genitourinary, sporadic or inherited mutations. Better prognosis than the alveolar.
- **Alveolar:** 20%, any age esp. adolescents, extremities, chromosomal translocations between chromosomes 2 and 13 (1 and 13).
- **Pleomorphic:** Adults, men, lower extremity, poor prognosis.

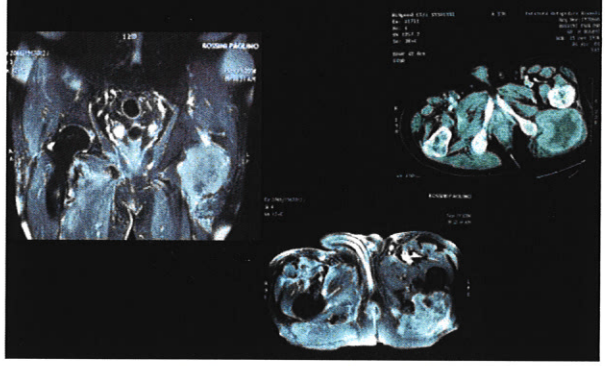
Rhabdomyosarcoma in the forearm



Synovial Sarcoma:

- Spindle cell tumours with epithelial component.
- 5-10% of all STS, young adults (10-40 years). Slowly growing mass, long history. Unrelated to synovium. Juxtaarticular. Deep tissues of extremities.
- Minute <1cm lesions in hands and feet.
- Chromosomal translocation (18 and X) in >90%.
- Biphasic (epithelial and spindle cell) or monophasic. Calcifications in 20-35%.

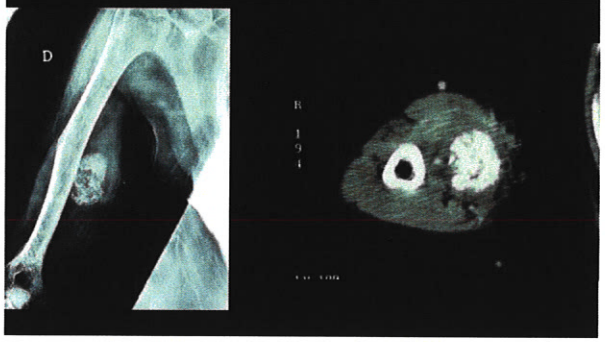
Synovial sarcoma of the gluteus maximus



Epithelioid Sarcoma:

- Predominant epithelioid morphology, men (2:1), adolescents and young adults. Rare <1%.
- Commonly superficial, flexor surface of hand and forearm. Firm, slowly growing nodules or plaque like lesions.
- High grade, aggressive, tend to propagate along fascial planes. Local recurrence 35%.
- Early metastasis(lymphatic and haematogenous). Multiple local recurrences common.

Epithelioid sarcoma in the arm

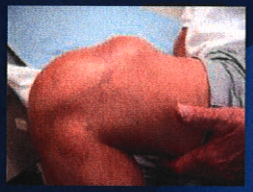


Presentation

- Any lump presenting with the following should be considered malignant until proved otherwise:
  - > 5cm
  - Increasing in size
  - Deep to the deep fascia
  - Pain
  - Any recurrence of a previously excised lump

How worrying are these features?

Features	Risk of malignancy
None	Nil
1	17%
2	43%
3	78%
4	86%



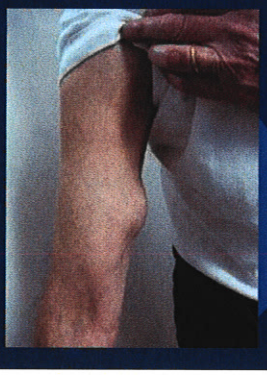
An Example

Concerned?

Size ~ 4cm
Deep
Painful
Increasing in size

i.e. 3 out of 4 +ve
Likely to be a sarcoma

...and it was



What lumps can be safely excised without biopsy first?

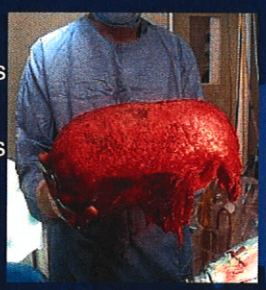
- < 3cm
- Subcutaneous
- Well defined

If in doubt DON'T cut it out - investigate it first!

How good are we at early detection?

Mean sizes:
STS 11 cms

Breast cancer 2.5cms

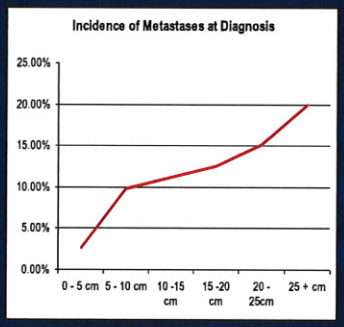


Is early diagnosis important?

YES !!

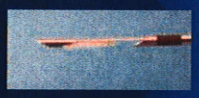
- Prognosis related to size for all sarcomas.
- Patients like to be diagnosed early.
- Delays add to difficulties in treatment.
- Litigation may follow delays.

Does size matter?

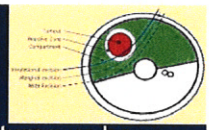


Biopsy:

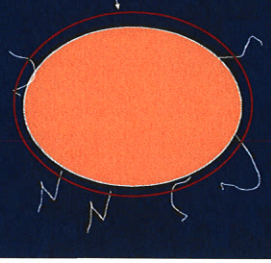
- Open vs trucut
- Trucut is the best.
- Aim NOT to contaminate normal tissue.
- Aim to get a representative sample of tumour.
- Use imaging to hit the target.



Treatment of STS:



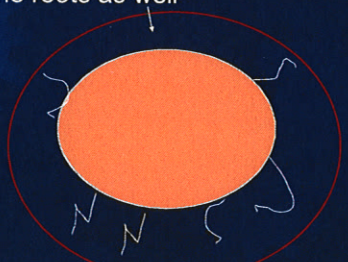
- Wide surgical excision – tumours have "roots" – shelling them out is not enough



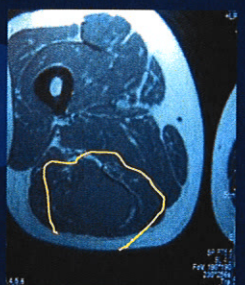
The "roots" will be left behind...

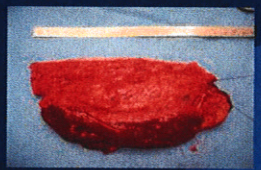
Proper Treatment

- Wide surgical excision – aim to remove the roots as well



What is an adequate margin?



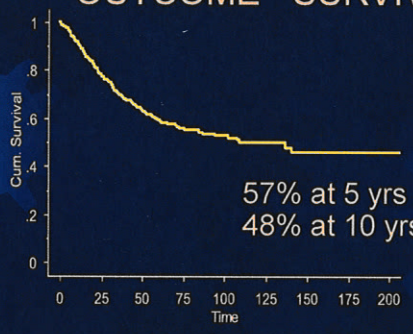


Limiting factor is sciatic nerve .....

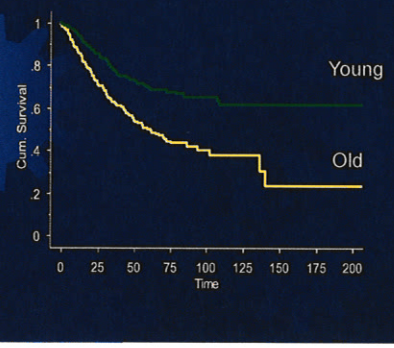
Local recurrence

- Related to
  - Grade of tumour
  - Margins of Excision
- Wide margins – 5%
- Inadequate margins - ?30%
  - Can be reduced by R/T - 15%

OUTCOME - SURVIVAL



Age – older or younger than 50

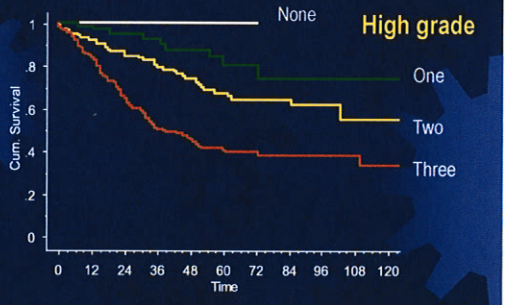


SIGNIFICANT FOR OVERALL SURVIVAL..

- Grade (high/intermediate)
- Depth (deep)
- Size (>5cm)
- Age (>50)
- Local Recurrence (+)

ALL
p >0.0001

Survival by risk factors



Local control rates:

Compartmental/amputation	100%
Wide local excision	92%
Marginal excision + RT	90%
Marginal excision alone	63%

Risk 13x higher for marginal v compartmental and 3X higher if tumour >10cm diameter

Scandinavian Sarcoma Group 1989

Unplanned excision of STS

- The inadvertent excision of a lump which turns out to be a sarcoma.
- Surgeon invariably optimist and will say "it's all out".
- Pathologist more honest "margins involved".
- What do you do now?

The effect of an unplanned excision of a soft-tissue sarcoma on prognosis

C. R. Chandrasekar, H. Wafa, R. J. Grimer, S. R. Carter, R. M. Tillman, A. Abudu

From The Royal Orthopaedic Hospital Oncology Service, Birmingham, England

We investigated whether our policy of routine re-excision of the tumour bed after an unplanned excision of a soft-tissue sarcoma was justified.

Between April 1982 and December 2005, 2201 patients were referred to our hospital with the diagnosis of soft-tissue sarcoma, of whom 402 (18%) had undergone an unplanned excision elsewhere. A total of 363 (16.5%) were included in this study. Each patient was routinely restaged and the original histology was reviewed. Re-excision was undertaken in 316 (87%). We analysed the patient, tumour and treatment factors in relation to local control, metastasis and overall survival.

Residual tumour was found in 188 patients (59%). There was thus no residual disease in 128 patients of whom 10% (13) went on to develop a local recurrence. In 149 patients (47%), the re-excision specimen contained residual tumour, but it had been widely excised. Local recurrence occurred in 30 of these patients (20%). In 39 patients (12%), residual tumour was present in a marginal resection specimen. Of these, 46% (18) developed a local recurrence. A final positive margin in a high-grade tumour had a 60% risk of local recurrence even with post-operative radiotherapy.

Metastases developed in 24% (86). The overall survival was 77% at five years. Survival was related to the grade of the tumour and the finding of residual tumour at the time of re-excision.

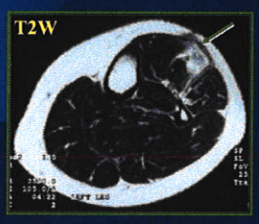
We concluded that our policy of routine re-excision after unplanned excision of soft-tissue sarcoma was justified in view of the high risk of finding residual tumour.

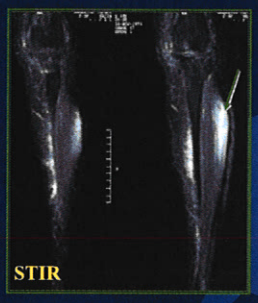
What to do?

- Restage with MRI → unreliable in excluding residual disease.
- If MRI +ve for residual tumour
  - Correct in 98% of cases
- If MRI –ve for residual tumour
  - Correct in 60%
  - i.e. WRONG in 40%
- So, we always recommend a wide re-excision of the area + drain track.
- 60% will have residual tumour.

A TRUE POSITIVE SCAN

Residual Soft Tissue Sarcoma <1cm



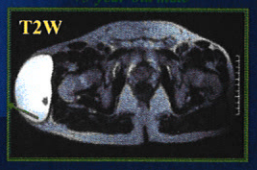


44-year-old male

FALSE POSITIVE

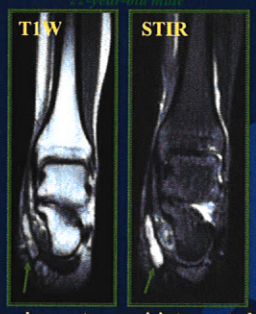
No Residual Tumour

23-year-old male



- small clot within seroma misinterpreted as residual tumour

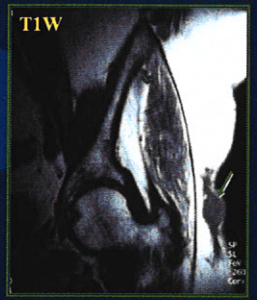
22-year-old male

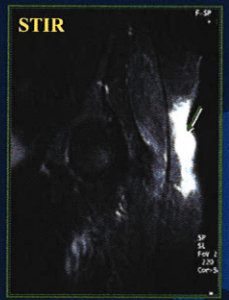


- haematoma misinterpreted as tumour

FALSE NEGATIVE

Residual Tumour (>1cm)





- tumour mass misinterpreted as seroma in scar

46-year-old female

CONCLUSION

- Most lumps and bumps are benign.
- Excision biopsy can be performed if the lump is superficial, < 3cm, and well-defined.
- If a lump has any of the four worrying features – be SUSPICIOUS:
  - > 5cm.
  - Deep.
  - Painful.
  - Increasing in size.

CONCLUSION

- Surgical treatment of STS should aim to achieve wide resection margins.
- Surgical re-excision +/- RT is the standard treatment after unplanned excision of soft tissue sarcomas.
- Prognosis is related to grade, size and depth.

Thank You