SOFT TISSUE TUMOR

Advances in Imaging, diagnosis, surgical technique, instrumentation, adjuvant therapy and basic knowledge of tumor biology have decreased disease free survival in a soft tissue sarcoma from 20% to 70%.

The myth that a small lesions are always benign is not true. About 1/3 sarcoma present like a benign tumor. The main stay of treatment of a soft tissue sarcoma is wide en bloc surgical resection with a pre or post operative radiation. This allows a local control 90%.

<table>
<thead>
<tr>
<th>Tissue Origin</th>
<th>Benign</th>
<th>Malignant</th>
<th>Reactive</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fibrous</td>
<td>Fibroma</td>
<td>Fibrosarcoma</td>
<td>Dupuytrens contracture</td>
</tr>
<tr>
<td>Fibrohistiocytic</td>
<td>Histiocytoma</td>
<td>MFH</td>
<td></td>
</tr>
<tr>
<td>Fat cells</td>
<td>Lipoma/ Angiolipoma</td>
<td>Liposarcoma</td>
<td>Hibernoma</td>
</tr>
<tr>
<td>Striated muscle</td>
<td>Rhabdomyoma</td>
<td>Rabdomyosarcoma</td>
<td></td>
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<tr>
<td>Smooth muscle</td>
<td>Leimyoma</td>
<td>Leiomyosarcoma</td>
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<tr>
<td>Blood vessel</td>
<td>Hemangioma</td>
<td>Haemangiosarcoma</td>
<td>Hemangioendothelioma</td>
</tr>
<tr>
<td></td>
<td>Glomus</td>
<td>Malignant haemangiopericytoma</td>
<td></td>
</tr>
<tr>
<td>Lymph</td>
<td>Lymphangioma</td>
<td>Lymphangiosarcoma</td>
<td></td>
</tr>
<tr>
<td>Synovial tissue</td>
<td>Giant cell</td>
<td>Synovial sarcoma</td>
<td>Synovial chondromatosis</td>
</tr>
<tr>
<td></td>
<td>Tumor of tendon sheath</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>PVNS</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Peripheral nerves</td>
<td>Neurilemmoma</td>
<td>Neurofibrosarcoma</td>
<td>Mortons Neuroma</td>
</tr>
<tr>
<td>Unknown cells</td>
<td>Ewings tumor</td>
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</tbody>
</table>

Post or Pre op radiation: no difference in prognosis. Logically, pre-operative radiotherapy shrinks tumor and enables en block excision. (FDG) positron emission tomography (PET) scans for detecting the response to chemotherapy
**Clinical feature**

1. Onset and development of swelling
2. Any associated pain. How long?
3. History of trauma
4. Single or multiple swelling:
   - Multiple lipoma and neurofibroma
5. Clinical: Describe
   - Consistency
   - Mobility
   - Cystic or not
   - Relation to muscle, nerve, bone
6. Previous medical history of Carcinoma. 0 Secondaries in the soft tissue
7. Size
   - < 5 cm in its greatest dimension is unlikely to be malignant
   - > 5 cm at least a 20% chance of being a soft tissue sarcoma.
   Exception to this rule: Epithelioid Sarcoma, Synovial sarcoma, Clear cell sarcoma may present as a small tumors
8. Superficial or deep?
   - Superficial lesions are more likely to be benign
9. Consistency on physical examination
   - Soft tissue sarcomas tend to be firm and not very painful
10. Cystic or solid [ultrasound]. Most cystic lesions are benign. Lesion is solid either a benign or malignant neoplasm.
11. Adenopathy: Rhabdomyosarcoma, Ewing’s sarcoma, Synovial sarcoma
12. Calcification
   - Vascular: Phleboliths
   - Synovial sarcoma: Fine calcification
   - Liposarcoma
   - Myxoid chondrosarcoma
   - Soft tissue Osteosarcoma
   - Myositis ossificans: central lucency
13. MRI

15. Chest X ray, CT scan of chest, for metastases

**MRI**

Good soft tissue definition

Relation to Neurovascular bundle

T1; T2; Fast spin echo (FSE), Gad (Cystic Vs solid):

30% indicate specific diagnosis with MRI eg., Lipoma and Hemangioma

**Interpretation**

<table>
<thead>
<tr>
<th>Tumor</th>
<th>Tissue type</th>
<th>T1</th>
<th>T2</th>
<th>Pattern</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ganglion/synovial cyst</td>
<td>Fluid</td>
<td>Low</td>
<td>High</td>
<td>Homogenous</td>
</tr>
<tr>
<td>Lipoma</td>
<td>Fat</td>
<td>High</td>
<td>Intermediate</td>
<td>Dark on fat suppression</td>
</tr>
<tr>
<td>PVNS</td>
<td>Hemosiderin</td>
<td>Low</td>
<td>Low</td>
<td>Dark on fat suppressed</td>
</tr>
<tr>
<td>Desmoid</td>
<td>Fibrous</td>
<td>Low</td>
<td>Low</td>
<td>Dark nodular</td>
</tr>
<tr>
<td>Hemangioma</td>
<td></td>
<td>High</td>
<td>High</td>
<td>Serpinginous</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>High fat</td>
</tr>
<tr>
<td>Soft tissue sarcoma</td>
<td></td>
<td>Low</td>
<td>High</td>
<td>Perilesional oedema</td>
</tr>
</tbody>
</table>
Ultrasound

Helpful: Cystic lesion around the joint
  Pseudoaneurysms

Adv: Readily available/ low cost
  US guided biopsy

Stain

Cytokeratin: differentiate Sarcoma from carcinoma.

Desmin in Smooth and striated muscle.

S 100 in Neural and cartilage lesions

Cytogenetic analysis: Needs fresh unfixed specimen

Genetics

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<tr>
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<tbody>
<tr>
<td><strong>RB gene</strong></td>
<td><strong>Osteosarcoma</strong></td>
</tr>
<tr>
<td><strong>Ewings Sarcoma</strong></td>
<td>t11-22 t(11:22)(q24;q12)</td>
</tr>
<tr>
<td><strong>Synovial Sarcoma</strong></td>
<td>t X-18 t(X;18)(11;q11)</td>
</tr>
<tr>
<td><strong>Myxoid Liposarcoma</strong></td>
<td>t 12-16</td>
</tr>
<tr>
<td><strong>Fibrous Histiocytoma</strong></td>
<td>[MFH] Abnormal Chr 19</td>
</tr>
</tbody>
</table>
Treatment

Margin (Benign)
Wide margin (2-4 cm of normal tissue) common
Radical (Whole compartment) for recurrence

Sarcoma: Marginal 30% recurrence; Wide margin 15% and radical 5%
In 85% of sarcomas, the limb salvage is possible
Problem with radical: Morbidity

Post operative assessment

At 3 months Post operative, a MRI scan and then 6 monthly to see any recurrence.

CT lungs every 3 months
**Radiotherapy**

Should be combined with wide resection

General dose: 6300 cGy extremely effective for microscopic disease

Advantage: sterilize reactive zone and enable to remove the tumor while sparing critical structures.

Brachy therapy tubes useful when the plane of surgical dissection is close to critical NV structures. This will allow high doses of radiation to be delivered.

*When margin is –ve: X ray Rx = 100% effective in controlling recurrence and when margin is +ve, X ray Rx = 50% effective.*

**Synovial Sarcoma**

10% of soft tissue sarcoma

Rare; III or IV decade

Around the knee; Lower limb (foot, ankle and knee) commonly involved

Usually extrasynovial and extracapsular

Higher incidence of metastasis to the Lymph node (25%)

X ray: 25% speck of calcification

Treat with wide excision and adjuvant radiotherapy

Chemotherapy

5 yr survival 50%
LIPOSARCOMA

Malignant tumors with differentiation towards fatty tissue.

Types: WDLS, Myxoid, Pleomorphic, Round cell.

Range from low grade to high grade sarcoma. Low grade liposarcomas can be difficult to differentiate from a benign lipoma. Low grade liposarcomas treated with wide local excision or observation is controversial as they do not metastasize

High grade liposarcomas treated with wide local excision and radiotherapy

50% are Myxoid Liposarcoma [commonest liposarcoma]. Retroperitoneal Liposarcoma has a worst prognosis than extremity.

WDL [Well differentiated liposarcoma]

Lipoma like liposarcoma. Low grade I

Cytogenetically: 12Q13-15 amplification

Great overlap with benign lipoma clinically

All deep lesions: need MRI

Lipoma: Homogenous, high in T1, no enhancement

Liposarcoma: Heterogenicity, nodularity and thick nodular septa

Controversy: Excision Vs observation

I. Heterogenous deep fatty tumor
   [no fatty signal]
   Low T1 and high T2 [could be myxoid, round or pleomorphic or dedifferentiated lipoma.
   Should be dealt in the tumor center
   Need en-bloc

II. WDL and Lipoma
   Difficult to differentiate
   MRI may be helpful
Discuss with tumor centre
When unsuspected and excised and cytogenetics.
Observe: there is incidence of recurrence and 10% of recurrence are dedifferentiate LS

III. Benign Lipoma on MRI
Observe or marginal excision

RHABDOMYOSARCOMA
The most common sarcoma in young patients
Highly malignant
Grows rapidly
Spindle cells in parallel bundles, multinucleated giant cells and racquet shaped cells,
cross striations within the tumor cells (rhabdomyoblasts)
Sensitive to multi-agent chemotherapy
Treat with preoperative chemotherapy, followed by wide surgical excision and radiotherapy

FIBROSARCOMA
Age: 30-50years
1/3: Secondary to Chronic osteomyelitis, Fibrous dysplasia, irradiation, bone infarct [all these can cause MFH]
Microscopic: Fibroblast and collagen arranged in a “Herring bone pattern”
Well differentiated: 5 year survival is 60%
and 30% in poorly differentiated
Treatment is wide local excision.
If > 5cm add radiation therapy,
preop/postop/periop
MALIGNANT FIBROUS HISTIOCYTOMA [MFH]

Commonest soft tissue sarcoma in patients over 50 years

Micro: Storiform or pin wheel pattern

Spindle cells and histiocytes

After liposarcoma, MFH is common

Minimal periosteal reaction

Cortical destruction