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%.

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

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. ? 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 ossificance: central lucency

13. MRI

14. Blood: FBC, ESR, CRP, LFT, E&U

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

T1 low and high T2	Most sarcoma
Low Tl and T2	Desmoid tumor, extensive scar tissue, PVNS
High T1 and T2	Hemangioma

Tumor	Tissue type	T1	T2	Pattern
Ganglion/synovial cyst	Fluid	Low	High	Homogenous
Lipoma	Fat	High	Intermediate	Dark on fat suppression
PVNS	Hemosiderin	Low	Low	Dark on fat suppressed
Desmoid	Fibrous	Low	Low	Dark nodular
Hemangioma		High	High	Serpinginous High fat
Soft tissue sarcoma		Low	High	Perilesional oedema

Ultrasound

Helpful: Cystic lesion around the joint

Pseudoaneurysms

Adv: Readily available/ low cost

US guided biopsy

Stain

Cytokeratin: differentiate Sarcoma from carcinoma.

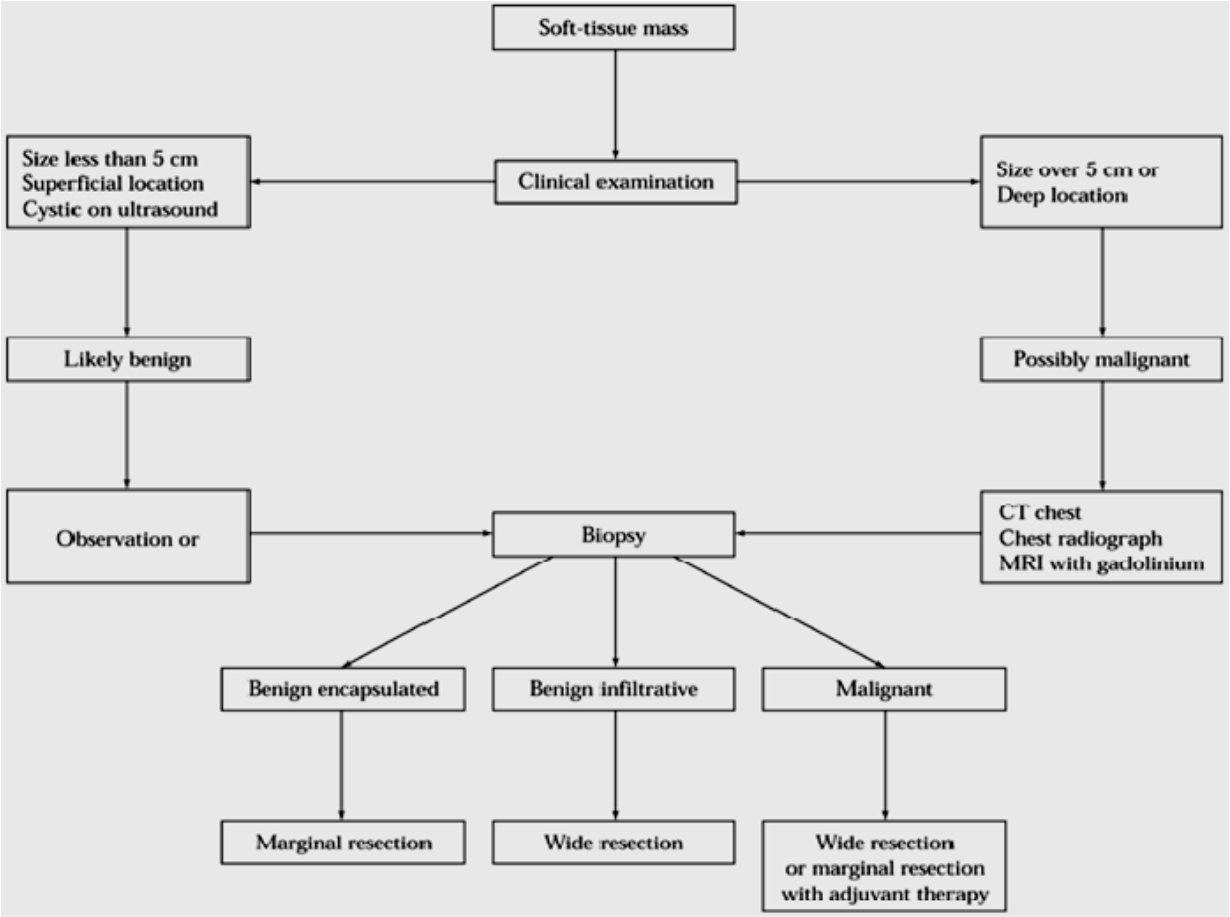
Desmin in Smooth and striated muscle.

S 100 in Neural and cartilage lesions

Cytogenetic analysis: Needs fresh unfixed specimen

Genetics

RB gene	Osteosarcoma
Ewings Sarcoma	t11-22 t(11:22)(q24:q12)
Synovial Sarcoma	t X-18 t(X;18)(11;q11)
Myxoid Liposarcoma	t 12-16
Fibrous Histiocytoma [MFH]	Abnormal Chr 19



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

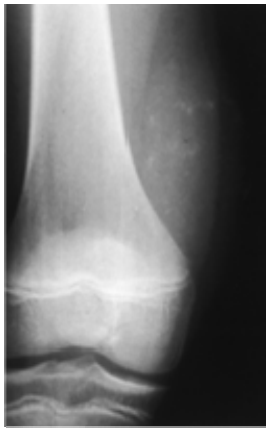
Higher incidence of metastasis to the Lymph node (25%]

X ray: 25% speck of calcification

Treat with wide excision and adjuvant radiotherapy

Chemotherapy

5 yr survival 50%



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Micro

Biphasic: Spindle and columnar cells.

Columnar cells forming gland like spaces in which cytokeratin and epithelial membrane can be demonstrated.

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

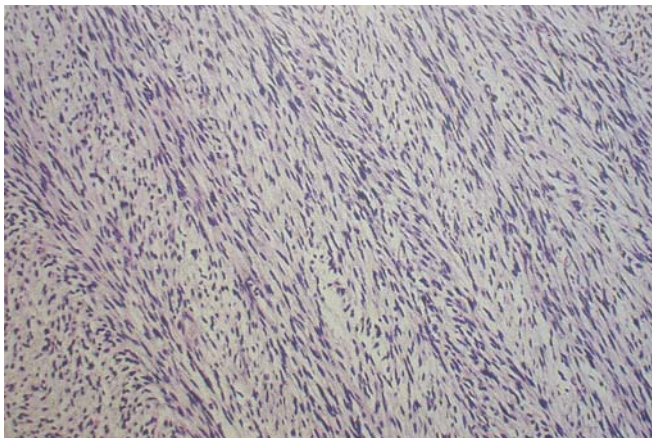
Age: 30-50years

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