SOFT TISSUE SARCOMA

Dr Neeraj Jain

Professor Radiation Oncology

Sri Guru Ram Das University of Health Sciences, Amritsar

NATURAL HISTORY

- Heterogenous group of solid tumours of mesenchymal origin
- Accounts 1% of all adult malignancies
- Accounts 15% of paediatric malignancies
- Arise from Muscle, Connective Tissue, Supportive Tissue, Vascular Tissue
- Highly invasive locally and high propensity for local recurrence
- Metastasis is via bloodstream and less commonly via lymphatics

ETIOLOGY/RISK FACTORS

- Arsenic
- Vinyl Chloride
- Ionising Radiation
- Pesticides
- Fluoridated Water

ETIOLOGY/RISK FACTORS

- Neurofibromatosis Type 1 or Von Recklinghausen Disease

High incidence of Neurofibromas

- Gardner Syndrome

High incidence of Desmoid Tumour

- Li Fraumeni Syndrome

High Incidence of STS

Von Recklinghausen Disease

- Neurofibromatosis Type 1
- 1 in 3000 births
- 50% have family history
- Presents as multiple hyperpigmented skin nodules
- Malignant peripheral Nerve sheath tumour risk is high

Li Fraumeni Syndrome

- 5 in 20000 births
- Caused by mutation in TP53 Gene
- Can cause STS , osteosarcoma, Breast Cancer, GBM, Medulloblastoma, Choroid Plexus Tumor, AML, Adrenocortical Ca, Neuroblastoma

Gardner Syndrome

- Rare genetic disorder
- 5q21band on chromosome 5
- Can cause Desmoid tumours, Osteomas, Fibromas
- Present as colon polyps. May become Ca Colon

Clinical Presentation

- Painless Mass over extremity
- Abdominal discomfort
- Abdominal swelling
- Cough and Breathlessness

Diagnostic Workup

- Biochemistry
- Imaging of Site

CECT

MRI

PET CT

- Chest Imaging
- Biopsy and IHC

ADIPOCYTIC TUMOURS

Benign
Lipoma and lipomatosis
Lipomatosis of nerve
Lipoblastoma and lipoblastomatosis
Angiolipoma
Myolipoma of soft parts
Chondroid lipoma
Spindle cell/pleomorphic lipoma
Atypical spindle cell/pleomorphic atypical lipomatous tumor
Hibernoma
Intermediate (locally aggressive)
Atypical lipomatous tumor
Malignant adipocytic tumours
Well differentiated liposarcoma: lipoma-like, sclerosing, inflammatory
Dedifferentiated liposarcoma
Myxoid liposarcoma
Pleomorphic liposarcoma
Myxoid pleomorphic liposarcoma

Fibro/Myofibroblastic Tumours

Benign
Nodular fasciitis
Proliferative fasciitis and proliferative myositis
Myositis ossificans and fibro-osseous pseudotumor of digits
Ischaemic fasciitis
Elastofibroma
Fibrous hamartoma of infancy
Fibromatosis colli
Juvenile hyaline fibromatosis
Inclusion body fibromatosis
Fibroma of tendon sheath
Desmoplastic fibroblastoma
Myofibroblastoma
Mammary-type myofibroblastoma
Calcifying aponeurotic fibroma
EWSR1-SMAD3-positive fibroblastic tumour (emerging)
Angiomyofibroblastoma
Cellular angiofibroma
Angiofibroma NOS
Nuchal fibroma
Acral fibromyxoma
Gardner fibroma

Fibro/Myofibroblastic Tumours

Intermediate (locally aggressive)
Palmar/plantar-type fibromatosis
Desmoid-type fibromatosis
Lipofibromatosis
Giant cell fibroblastoma
Dermatofibrosarcoma protuberans
Intermediate (rarely metastasising)
Dermatofibrosarcoma protuberans, fibrosarcomatous
Solitary fibrous tumour
Inflammatory myofibroblastic tumour
Low-grade myofibroblastic sarcoma
Superficial CD34-positive fibroblastic tumour
Myxoinflammatory fibroblastic sarcoma
Infantile fibrosarcoma

Fibro/Myofibroblastic Tumours

Malignant
Solitary fibrous tumour, malignant
Fibrosarcoma NOS
Myxofibrosarcoma
Low grade fibromyxoid sarcoma
Sclerosing epithelioid fibrosarcoma

Fibrohistiocytic Tumours

Benign
Tenosynovial giant cell tumour
Deep benign fibrous histiocytoma
Intermediate (rarely metastasising)
Plexiform fibrohistiocytic tumour
Giant cell tumour of soft parts NOS
Malignant
Malignant tenosynovial giant cell tumour

Vascular Tumors

Benign
Synovial haemangioma
Intramuscular haemangioma
Arteriovenous malformation/haemangioma
Venous haemangioma
Anastomosing haemangioma
Epithelioid haemangioma
Lymphangioma and lymphangiomatosis
Acquired tufted haemangioma

Vascular Tumors

Intermediate (locally aggressive)
Kaposiform haemangioendothelioma
Retiform haemangioendothelioma
Papillary intralymphatic angioendothelioma
Composite haemangioendothelioma
Kaposi sarcoma
Pseudomyogenic haemangioendothelioma
Malignant
Epithelioid haemangioendothelioma
Angiosarcoma

Pericytic(Perivascular) Tumours

Benign and intermediate
Glomus tumour NOS
Myopericytoma, including myofibroma
Angioleiomyoma
Malignant
Glomus tumour, malignant

Smooth Muscle Tumour

Benign
Leiomyoma
Intermediate
Smooth muscle tumour of uncertain malignant potential
EBV-associated smooth muscle tumour
Malignant
Inflammatory leiomyosarcoma
Leiomyosarcoma

Skeletal Muscle Tumours

Benign
Rhabdomyoma
Malignant
Embryonal rhabdomyosarcoma
Alveolar rhabdomyosarcoma
Pleomorphic rhabdomyosarcoma
Spindle cell / sclerosing rhabdomyosarcoma
Ectomesenchymoma

GIST

Benign
MicroGIST
Malignant
Gastrointestinal stromal tumors

Periphral Nerve Sheath Tumour

Benign
Schwannoma
Neurofibroma
Perineurioma
Granular cell tumour
Nerve sheath myxoma
Solitary circumscribed neuroma
Meningioma
Hybrid nerve sheath tumour
Malignant
Malignant peripheral nerve sheath tumour
Melanotic malignant nerve sheath tumour
Granular cell tumour, malignant
Perineurioma, malignant

Tumour of Uncertain Differentiation

Benign
Myxoma (cellular myxoma)
Deep (aggressive) angiomyxoma
Pleomorphic hyalinising angiectatic tumour
Phosphaturic mesenchymal tumour
Perivascular epithelioid tumour, benign
Angiomyolipoma
Intermediate (locally aggressive)
Haemosiderotic fibrolipomatous tumour
Angiomyolipoma, epithelioid
Intermediate (rarely metastasising)
Atypical fibroxanthoma
Angiomatoid fibrous histiocytoma
Ossifying fibromyxoid tumour
Myoepithelioma

Tumour of Uncertain Differentiation

Malignant
Phosphaturic mesenchymal tumour, malignant
NTRK-rearranged spindle cell neoplasm (emerging)
Synovial sarcoma
Epithelioid sarcoma: proximal and classic variant
Alveolar soft part sarcoma
Clear cell sarcoma
Extraskeletal myxoid chondrosarcoma
Desmoplastic small round cell tumour
Rhabdoid tumour
Perivascular epithelioid tumour, malignant
Intimal sarcoma
Ossifying fibromyxoid tumour, malignant
Myoepithelial carcinoma
Undifferentiated sarcoma
Spindle cell sarcoma, undifferentiated
Pleomorphic sarcoma, undifferentiated
Round cell sarcoma, undifferentiated

Staging AJCC 8TH

- **A) Trunk and extremity and B) Retroperitoneum**
- **Definition of primary tumor (T)**
- T Category T Criteria
- TX Primary tumor cannot be assessed
- T0 No evidence of primary tumor
- T1 Tumor 5 cm or less in greatest dimension
- T2 Tumor more than 5 cm and ≤10 cm in greatest dimension
- T3 Tumor more than 10 cm and ≤15 cm in greatest dimension
- T4 Tumor more than 15 cm in greatest dimension

Staging AJCC 8TH

- **C) Head and Neck**
- **Definition of primary tumor (T)**
- T Category T Criteria
- TX Primary tumor cannot be assessed
- T1 Tumor ≤2 cm
- T2 Tumor >2 cm to ≤4 cm
- T3 Tumor >4 cm
- T4 Tumor with invasion of adjoining structures
- T4a Tumor with orbital invasion, skull base/dural invasion, invasion of central compartment viscera, involvement of facial skeleton or involvement of pterygoid muscles
- T4b Tumor with brain parenchymal invasion, carotid artery encasement, prevertebral muscle invasion or central nervous system involvement via perineural spread

Staging AJCC 8TH

- **Abdomen and thoracic visceral organs**
- **Definition of primary tumor (T)**
- T Category T Criteria
- TX Primary tumor cannot be assessed
- T1 Organ confined
- T2 Tumor extension into tissue beyond organ
- T2a Invades serosa or visceral peritoneum
- T2b Extension beyond serosa (mesentery)
- T3 Invades another organ
- T4 Multifocal involvement
- T4a Multifocal (2 sites)
- T4b Multifocal (3–5 sites)
- T4c Multifocal (>5 sites)

Staging AJCC 8TH

- **Definition of regional lymph node (N)**
- N Category N Criteria
- N0 No regional lymph node metastasis or unknown lymph node status
- N1 Regional lymph node metastasis
- **Definition of distant metastasis (M)**
- M Category M Criteria
- M0 No distant metastasis
- M1 Distant metastasis

Staging AJCC 8TH

- G G Definition
- GX Grade cannot be assessed
- G1 Total differentiation, mitotic count and necrosis score of 2 or 3
- G2 Total differentiation, mitotic count and necrosis score of 4 or 5
- G3 Total differentiation, mitotic count and necrosis score of 6, 7 or 8

Prognostic Factors

- OVERALL SURVIVAL

Grade, size, stage, surgical margins, distant metastasis, age, sex, performance status, and haemoglobin value were significant for overall survival.

- LOCAL RECURRENCE

Histology, grade, stage, and surgical margins were significant for local recurrence.

- DISTANT RECURRENCE

Histology, grade, stage, and surgical margins were significant for local recurrence.

AJCC Prognostic Stage Grouping For Extremity and Trunk

Stage	Primary tumor (T)	Regional lymph node (N)	Distant metastasis (M)	Histologic grade (G)
IA	T1	N0	M0	G1, GX
IB	T2, T3, T4	N0	M0	G1, GX
II	T1	N0	M0	G2, G3
IIIA	T2	N0	M0	G2, G3
IIIB	T3, T4	N0	M0	G2, G3
IV	Any T	N1	M0	Any G
	Any T	Any N	M1	Any G


Thank You