#### Soft tissue Tumors II

#### Lecture 36: Soft tissue tumors II

At the end of session the student should be able to:

 Discuss benign and malignant <u>fibrohistiocytic</u> <u>tumors</u>

- Describe morphological changes of benign and malignant fibrohistiocytic tumors
- Discuss benign and malignant <u>smooth</u> <u>muscle</u> <u>tumors</u>
- Describe morphological changes of benign and malignant smooth muscle tumors

Suggested Ref: Robbins Basic Pathology 8th edition 832 – 836 

## **Fibrohistiocytic Tumors**

- A definition of the fibroblast tumour is required a range of cellular differentiation, consists of spindle-cell morphology, vimentinstaining
- Fibrohistiocytic tumors:
- defined as neoplastic tumors contain cellular

elements that resemble both <u>fibroblasts</u> and <u>histiocytes</u> (macrophages)

 The phenotype:<u>fibroblasts</u>& <u>fibrohistiocytic</u> should be viewed as descriptive in nature and not one that connotes the cell of origin.

## Fibrohistiocytic Tumor classification

- I. Benign fibrohistiocytic tumor:
- 1-Benign Fibrous histiocytoma (Dermatofibroma)

 II. Malignant <u>fibrohistiocytic tumor:</u>
 2-Dermatofibrosarcoma protuberans (DFSP)

#### 3- Malignant fibrous histiocytoma (MFH)

## Benign

## Fibrohistiocytic Tumors

#### FIBROUS HISTIOCYTOMA (DERMATOFIBROMA)

- Site: Common lesiondermis and subcutis.
- Age: presents in midadult life (F>M).
- Clinically: It is painless and slow growing.
- Morphologic features:
- Gross: firm, small mobile nodule, skin intact.
- Circumscribed- sharp border between tumor & subcutis

- Basophilia is due to increased cellularity
- Storiform pattern arranged foam cells, fibroblasts and histiocytelike cells; foam cells are somewhat specific for this lesion.(positive Vimentin, Factor XIIIa, CD34)

### CASE- 1

Skin nodule over right thigh in a 38 year old woman, excisional biopsy done with safe margin?

#### <u>Dermatofibrosarcoma</u> <u>protuberans</u>

- \* Site: Common lesion- in dermis
- Age: usually adults 20-40 years
- Clinically: Low grade malignancy, Locally aggressive, low rate of metastasis,
- Morphologic features:
- Gross: firm, papulo-nodular skin lesion.
- Un-circumscribed- locally aggressive, entrappment.

- Tight Storiform pattern, radiating& infiltrating subcutaneous fat. Hemorrhage and necrosis are rare
- Special stains(Positive Vimentin, Negative for CD34)

CASE 2 Left chest wall recurrent nodule slowly growing for 15 years with sudden recent growth in a 43 year old woman, with another papulonodular lesion on the thigh, chest lump is excised with safe

## margin and sent for histopathology? <u>Malignant fibrous</u> <u>histiocytoma (MFH)</u>

- Groups of soft-tissue tumors, previously diagnosed as MFH, composed of considerable cellular pleomorphic sarcoma with prominent osteoclast-like giant cells-DIAGNOSIS BY EXCLSUSION- origin has been debated.
- <u>How common</u>: most common type of soft tissue sarcoma of adults. F>M
- <u>Currently</u> classified as variants of Fibrosarcoma

(myxofibrosarcoma, pleomorphic fibrosarcoma)

- <u>Site:</u> occur in thigh, retroperitoneum and upper limbs, bone, muscles, cartilage
- <u>Associated with</u> radiation therapy or surgical scars
- Malignant fibrous histiocytoma MFH
- Microscopic features: Characterized by
- 1- Non-circumscribed, uncapsulated highly cellular,
- 2- "tight" storiform pattern.

## 3- considerable cytologic pleomorphism with Presence of bizarre multinucleate cells, mitoses <u>The phenotype of the</u> neoplastic cell: 1- SMA stain: Negative

- 2- Desmin: Negative
- 3- CD34: Negative
- 4- Positive CD68, S100 and S100
- Case 3
- 62 year old woman
- Large subcutanous mass on anterior aspect of right lower leg

## <u>I. Benign</u> 1- LEIOMYOMAS (Benign)

- tumors classification
- smooth muscle

## II. Malignant

#### 2- LEIOMYOSARCOMA

## I. LEIOMYOMAS (Benign) morphology

- benign SMT, They develop in 77% of women.
- Clinically: depending on (number, size, and location) may cause a variety of symptoms including infertility.
- Gross: Solitary "uni-focal" or multiple.
- Size > variable , whorled, firm cut surface, and they are usually not necrotic or hemorrhagic, pseduocaspulated.

### Microscopic

- Fascicles of SMspindle cells that tend to intersect each other at right angles. "storiform pattern"
- The tumor cells have bluntended, elongated nuclei and show minimal or no atypia.
- Few mitotic figures (<5 per 10 hpf)</li>
- No necrosis or frequent mitoses.

### LEIOMYOSARCOMA-Malignant SMT

- \* Malignant SMT, considered as 10% to 20% of softtissue sarcomas
- \* Bulky, invasive solid masses into adjacent structure

## Age: occur in adults and afflict women > men.

### <u>Commonest sites:</u>

- 1) Retroperitoneum.
- 2) Deep soft tissues of the extremities.
- 3) Uterus
- 4) Blood vessels 5) Superficial dermis.
- Clinical outcome: depend on
  - the size& site.
- (Prognosis of cutaneous tumor better> retroperitoneum )
  - LEIOMYOSARCOMA-Malignant SMT
- <u>Morphology</u>:
- a) Gross: Size- large and bulky, infiltraive
- b) Microscopic: consist of

- Malignant spindle cells with cigar-shaped pleomorphic nuclei arranged in interweaving fascicles.
- Brisk mitoses
- Necrosis- coagulative
- c)Immunohistochemical: stain positive with antibodies to smooth muscle actin and desmin.
- summary
- I) Fibrohistiocytic tumor:
- A- Dermatofibroma
   B- MFH
- II) Smooth muscle tumor:
- A- Leiomyoma

# B- Leiomyosarcoma Excision of the tumour

- Based on features noticed in next slide
- How to differentiated between these sections:
- a) MFH?
- b) Leiomyosarcoma?

## Spindle cell sarcomas <u>Adequate clinical history</u>, past hx, pre-operative

image and operation findings.

2- Previous biopsy report.

#### 3- Histopathology diagnosis-

- Study gross appearance.(Consistency, color,...

- Study of cells shape and pattern(differen.)

- Presence of mitoses, necrosis.

Presence of

multinucleated giant cells,

Heterologus material,
 Vascular invasion ,

Nerve invasion • Bone invasion.

## 4- Immunohistochemical stain& Ancillary techniques Basic Panel of Immuno markers for Spindle Cell Tumours Vimentin (all mesenchymal tumour-SMA, MFH, etc

- Cytokeratin/EMA-(epithelial markers)
- S100P- (Smooth muscle tumour- origin)

- Desmin (Smooth muscle tumour- origin)
- SMA (Smooth muscle tumour- origin
- CD34 (benign fibrohistiocytic tumour)
- **Ckit** "CD99"– (GIST)