* 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 fibrohistiocytic tumors
* Describe morphological changes of benign and malignant fibrohistiocytic tumors
* Discuss benign and malignant smooth muscle tumors
* 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, **vimentin-staining**
* **Fibrohistiocytic tumors:**
* **defined as neoplastic tumors contain cellular elements that resemble both fibroblasts and histiocytes (macrophages)**
* **The phenotype:fibroblasts& fibrohistiocytic 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 fibrohistiocytic tumor:**

**2-Dermatofibrosarcoma protuberans (DFSP)**

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

* **Benign Fibrohistiocytic Tumors**

**FIBROUS HISTIOCYTOMA (DERMATOFIBROMA)**

* **Site: Common lesion-dermis and subcutis.**
* **Age: presents in mid-adult 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 histiocyte-like 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?**

* **Dermatofibrosarcoma protuberans**

**\* 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?**

* **Malignant fibrous histiocytoma (MFH)**
* **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.**
* **How common**: **most common type of** soft tissue sarcoma of adults. F>M
* **Currently classified as variants of Fibrosarcoma (myxofibrosarcoma, pleomorphic fibrosarcoma)**
* **Site: occur in thigh, retroperitoneum and upper limbs, bone, muscles, cartilage**
* **Associated with 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**

**The phenotype of the 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
* **smooth muscle tumors classification**
* **I. Benign**
* **1- LEIOMYOMAS (Benign)**
* **\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_**
* **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, pseduo- caspulated.
* **Microscopic**
* **Fascicles of SMspindle cells that tend to intersect each other at right angles. “storiform pattern”**
* **The tumor cells have blunt-ended, elongated nuclei and show minimal or no atypia.**
* **Few mitotic figures (<5 per 10 hpf)**
* **No necrosis or frequent mitoses.**
* **LEIOMYOSARCOMA- Malignant SMT**

**\* Malignant SMT, considered as 10% to 20% of soft-tissue sarcomas**

\* Bulky, invasive solid masses into adjacent structure

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

**Commonest sites:**

**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**

**Morphology:**

**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**

**1- Adequate clinical history, 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)