MSS

Musculoskeletal System

Doctor 2019 | Medicine | JU

NO.

Pathology

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Grammatical correction

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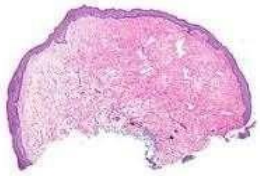
Fibromas and Fibrosarcomas

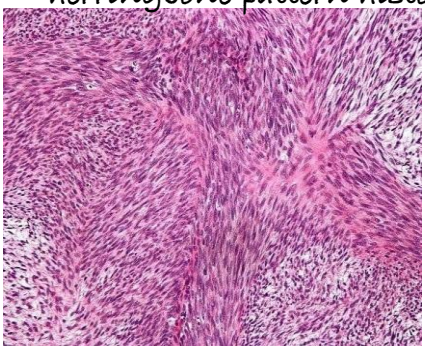
Tumors that arise from fibroblasts:



- **Fibromas**:
  - benign proliferation of fibroblasts
  - very common cutaneous and subcutaneous tumor
  - usually occurs in the skin and subcutaneous tissue
  - under the microscope they are planned benign appearing fibroblasts (spindle cells which has fibroblast appearance and immunostim chemical features)

Skin tag , neoplastic

herringbone pattern histology



- **Fibrosarcoma**:
  - malignant counterpart of fibromas.
  - superficial cutaneous tumors of fibroblasts.
  - under the microscope they are cellular, storiform pattern (The name "**storiform**" in Latin means (woven), which is a histopathologic sign consists of spindle cells with elongated nuclei radiating from a center point) with increased mitosis.

'' They aren't bad if we treated them very early , not common metastasize or necrosis or common to happen ,we see them more in children

- When we are not sure of what is the cells of origin we apply couple of stains, skeletal muscle marker, smooth muscle marker, the fibroblast muscle markers and the adipocytes marker. If we have fibroma or fibrosarcoma we will have positive fibroblast markers and the remnant will be negative (Tumor markers have traditionally been proteins or other substances that are made by both normal and cancer cells but at higher amounts by cancer cells. These can be found in the blood, urine, stool, tumors, or other tissues or bodily fluids of some patients with cancer**).**

Superficial fibromatoses

- ✓ Group of syndromes or diseases in which we will have proliferation of fibroblasts (Tumor-like conditions)- **Fibromatoses syndromes** Cdiseases
- ✓ There are 2 major types of fibromatoses syndromes, **superficial fibromatoses** (which we will explain) and the **deep fibromatoses** (will be explained later)

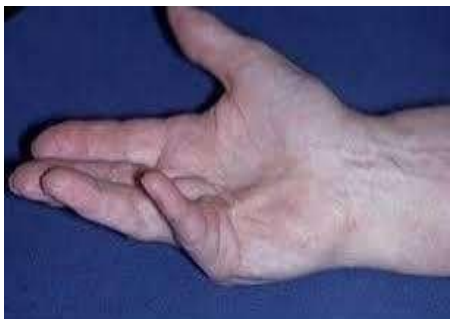
- **Superficial fibromatoses**:
  - ✓ Superficial fibromatoses occur in cutaneous and subcutaneous area close to the skin, you can see them and examine them with your hands.
  - ✓ They are infiltrative lesions but they are benign (they do not metastasize)

 Morbidity due to infiltration

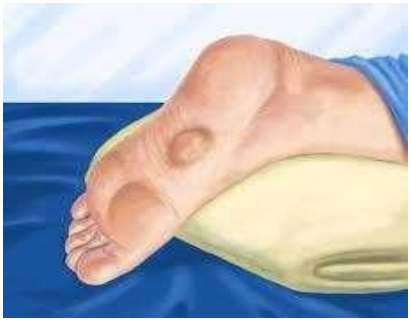
They look bland (normal)
No necrosis or hemorrhage
or increased mitosis

- ✓ Hereditary (may run in the family)
- ✓ Has a negative impact on local function
- ✓ There are three major forms:

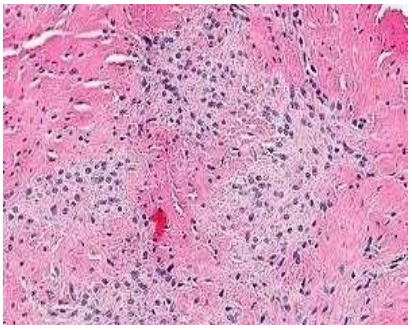
- **Palmar fibromatoses** (**DUPUYTREN CONTRACTION**) occur in the palms of the hands -the palmar fascia in the hand- (any figure can be involved and they impart the function of the affected finger, difficulty in flexing and extending, and they cause pain)
- **Planter fibromatoses:** usually occur in the sole of the foot, they will cause pain, impart on the walking and cause issues when wearing shoes
- **Penile fibromatosis** (**PEYRONITE DISEASE**)**:** occur in the dorsolateral aspect of the penis, cause painful erection and difficult to treat



Palmar fibromatoses



Planter fibromatoses



Penile fibromatosis

DEEP FIBROMATOSES (DESMOID TUMOR)

Desmoid reaction : increase TGF-B >increase collagen > hard mass >>> Desmond tumor

- ✓ Occur deep inside the tissue, invisible usually
- ✓ Deep infiltrative but bland fibroblastic proliferation; doesn't metastasize but recur
- ✓ 20-30 years, females more common
- ✓ Abdominal wall, mesentery and limbs
- ✓ they have characteristic mutations in ***CTNNB1*** (responsible for the production of fusion protein called **β-catenin** and when we suspect deep fibromatoses we can utilize this protein because we have special immunohistochemical stain for this protein) or ***APC***- *Adenomatous polyposis coli*- genes leading to increased **Wnt** signaling

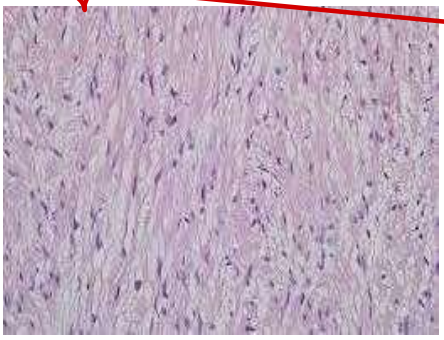
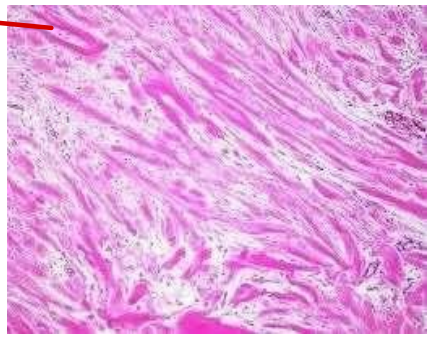
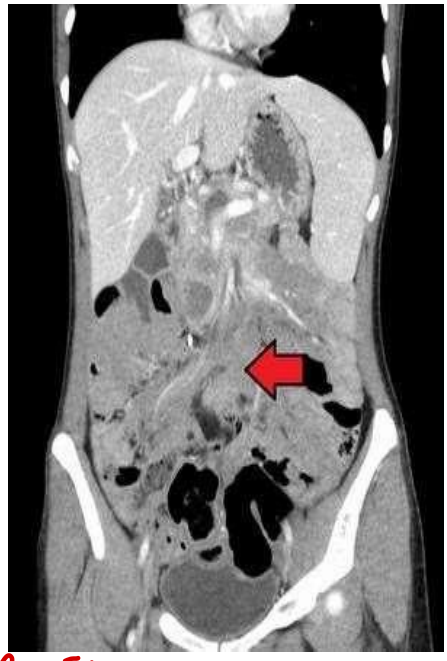
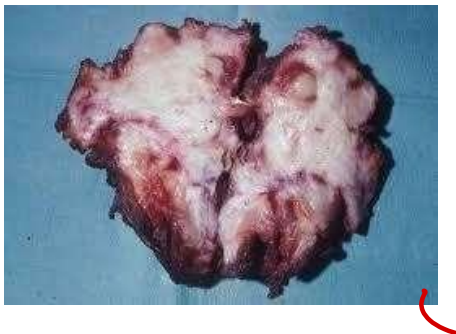
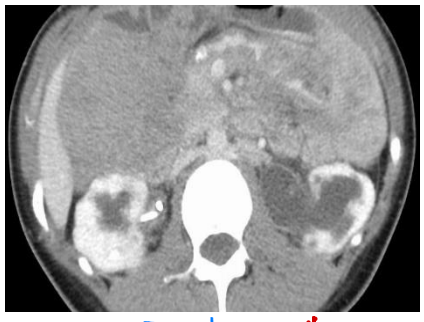
Positive (if we found it)then we are having deep fibromatosis

- ✓ Mostly are sporadic; but patients with Gardner (FAP -familial adenomatous polyposis syndrome) are susceptible There is association between them
- ✓ Difficult to treat Mutation on chromosome 5
- ✓ Complete excision is needed to prevent recurrence which is very common, but complete excision is very difficult because of the nature of this tumor, the surgeon doesn't know where the tumor start and end. The surgeon takes wide margin -will take 4-5 cm additional safe margins to make sure that he has complete excision

They look normal and same as Superficial fibromatoses

Destruction for other tissues

- ✓ These tumors (Lethal) kill by local infiltration NOT metastasis

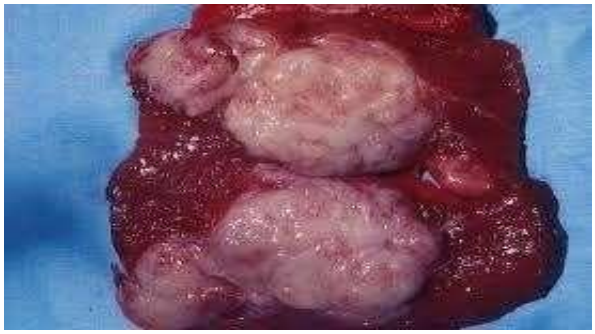
Borders بنقدرش نحدد ال

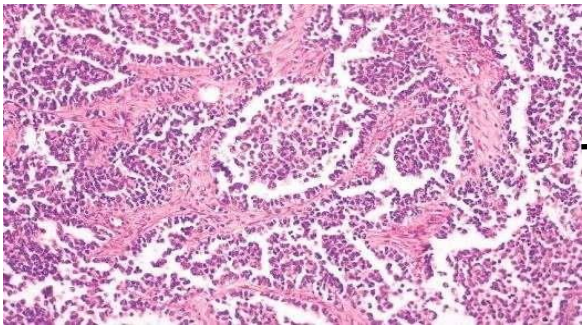
SKELETAL MUSCLE TUMORS:

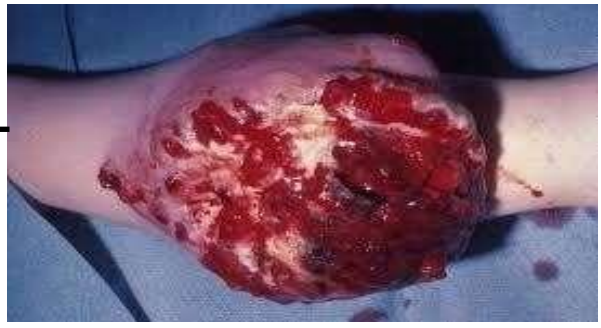
Arise from rhabdomyocyte cell

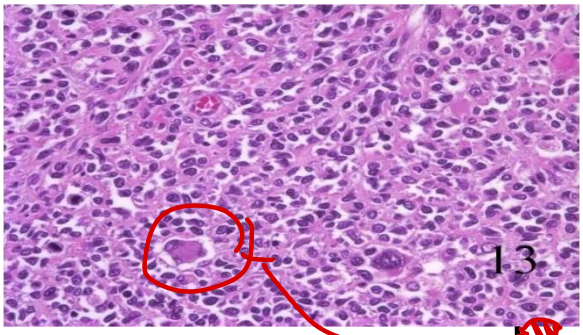
- ✓ Almost all malignant; except **rhabdomyoma** which is benign, rare, occurs with tuberous sclerosis (**TSC**-**Tuberous sclerosis**- is a rare multisystem autosomal dominant genetic disease that causes non-cancerous tumors to grow in the brain and on other vital organs such as the kidneys, heart, liver, eyes, lungs and skin), common locations of rhabdomyomas is the tongue and the heart

- ✓ Rhabdomyosarcoma (RMS) is the malignant prototype; most common child sarcoma
- ✓ 3 types (embryonal 60%; alveolar 20%; pleomorphic 20%)
- ✓ Specific mutations are common
- ✓ Aggressive tumors; treated by surgery, CT (chemotherapy) +/- RT (radiotherapy) multimodality management









Pleomorphic rhabdomyosarcoma in which the tumor is composed of small bleu cell tumor, at electron microscopy you can see the cross striations that will tell us that this is a skeletal muscle malignant tumor

Reflect striated muscle

Alveolar type of rhabdomyosarcoma because it looks like the alveoli of the lung

→ Rhabdomyosarcomas are usually large, fleshy and hemorrhagic tumors

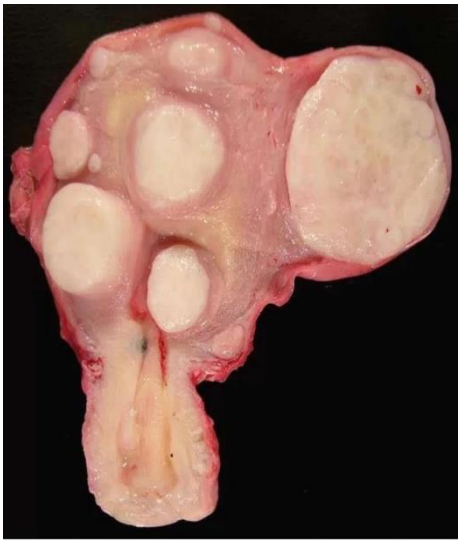
SMOOTH MUSCLE TUMORS

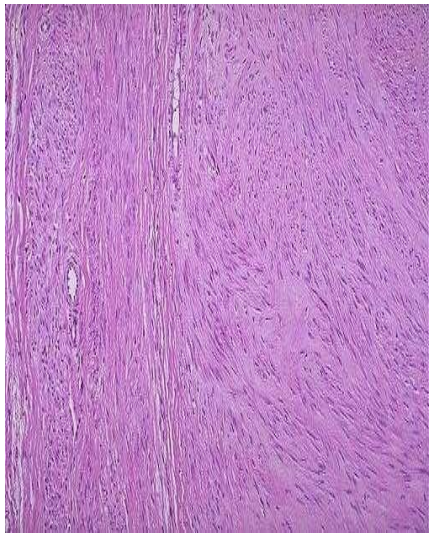
Leiomyocyte cell cell

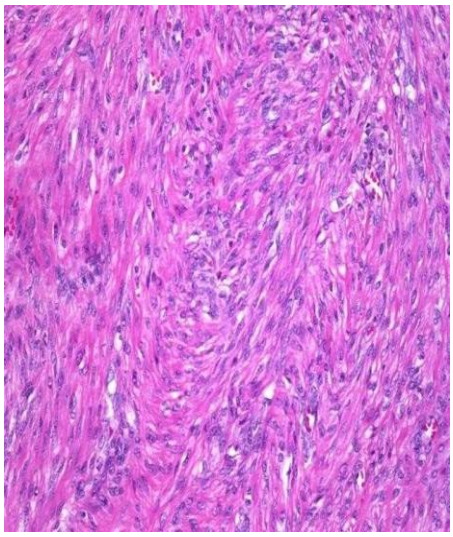
- ✓ Smooth muscles are present anywhere in the body, so smooth muscles tumors can arise in any organ
- ✓ Leiomyoma (benign) and leiomyosarcoma (malignant)

We differentiate between them from mitosis rate

- **Leiomyoma (LYM):** very common; any site but mostly uterus (fibroid-leiomyoma of the uterus- الألياف)…patient with one or multiple fibroid complains of menorrhagia and infertility
  - LYM vary in size and location, well circumscribed, not infiltrative
  - Few can have specific mutations (Fumarate hydratase on chromosome 1q42.3)
  - Dx: morphology and histology alone
  - Histology: Smooth muscle Cell proliferation, No necrosis, Little mitosis, no hemorrhage



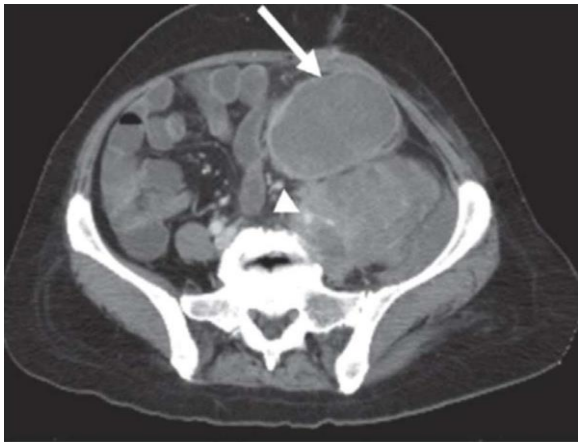


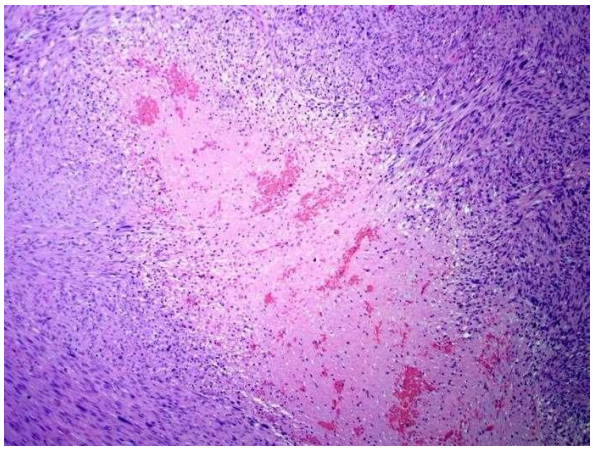


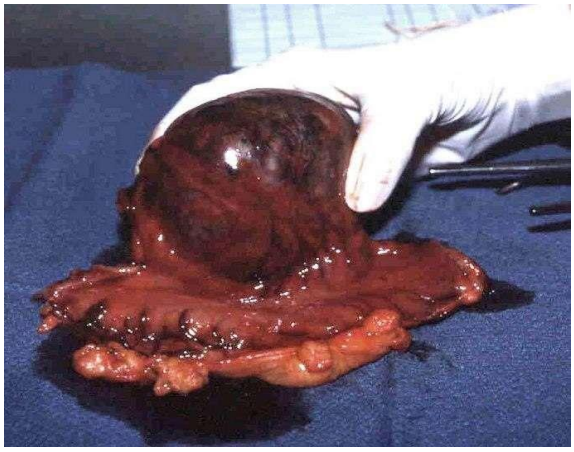
This is the cervix of the uterus and we can notice the well circumscribe, non-infiltrative, firm white leiomyoma

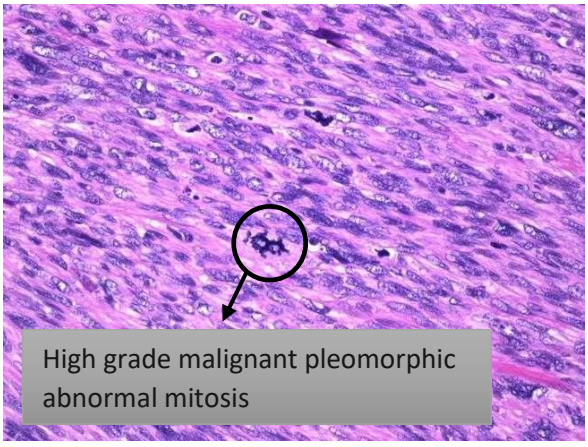
- **LEIOMYOSARCOMA:**
  - 10-20% of soft tissue sarcomas
  - Adults; more in females
  - Deep soft tissue, extremities and retroperitoneum or from great vessels and uterus (almost 98-99% of smooth muscle tumors of the uterus are leiomyomas, 1-2% are leiomyosarcoma)
  - Complex genotypes Smart important
  - Hemorrhage, necrosis, increased mitosis (many of them are abnormal) and infiltration of surrounding tissue

- Trx: depends on location, size and grade (high grade sarcomas in thigh or in the uterus are difficult to treat and sometimes we have to add probably additional modalities other than surgery)



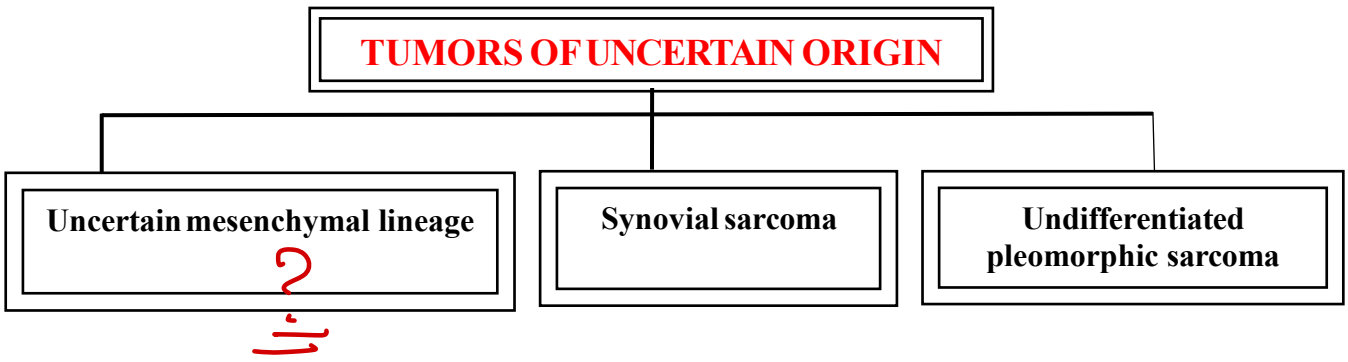






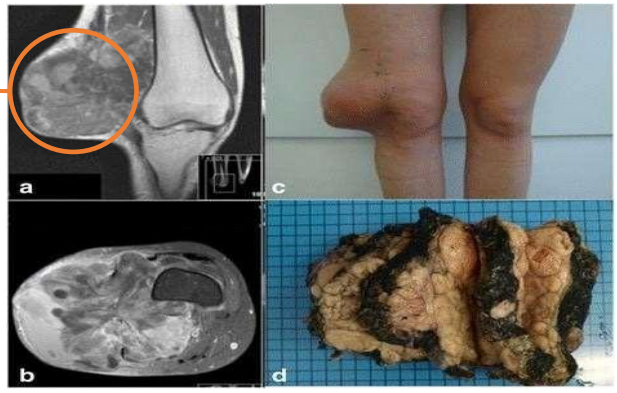
Abnormal mitosis then malignant

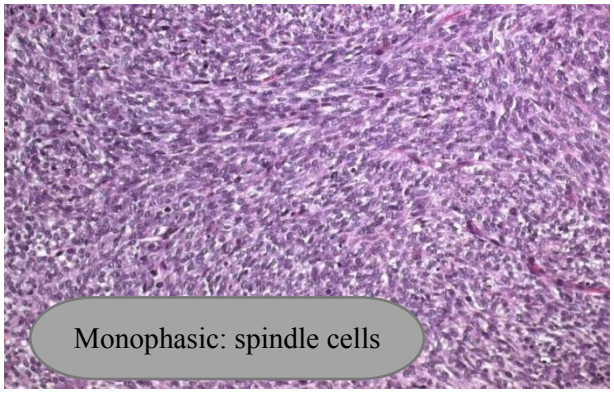
Radiologic, gross and histologic features of intraabdominal Leiomyosarcoma. Big tumor (10-15) cm with central hemorrhage and necrosis. Histologically this tumor is very cellular with clear hemorrhage and necrosis



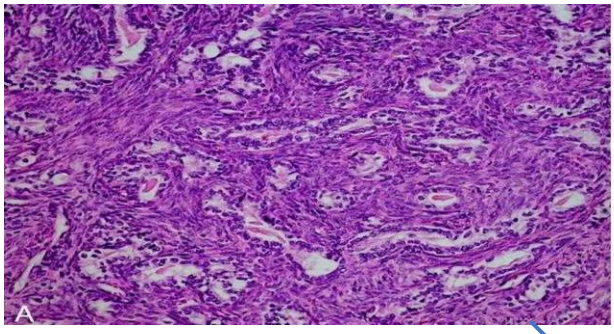
Synovial sarcoma

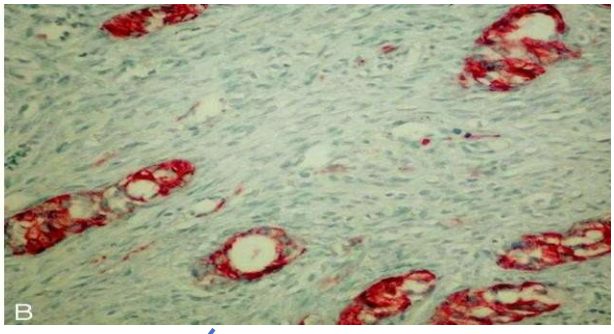
- Name is misnomer (Although they occur most commonly around joints, however they can occur anywhere) Doesn't arise from synovial cells
- 10% of all soft tissue sarcomas; 20-40s age
- Deep seated mass of long history, whether it's deep in the joint or deep in the chest or the abdomen
- Translocation T(X;18) (p11;q11) à Makes fusion genes SS18 (signature characteristic)
- Histologically: Monophasic (only spindle cells) or biphasic (spindle cells and glands (epithelial cells) There is a variability
- Trx: aggressive with limb sparing excision + CT to decrease the chance of hematogenous metastasis
- 5 years survival 25-65% depending on stage and multi-disciplinary team approach
- Metastasis: lung and lymph nodes





Monophasic: spindle cells





Radiologic appearance: Big mass with hemorrhage and heterogenous texture. It's close to the joint but there is no evidence that the origin is from the synovial cells

Biphasic

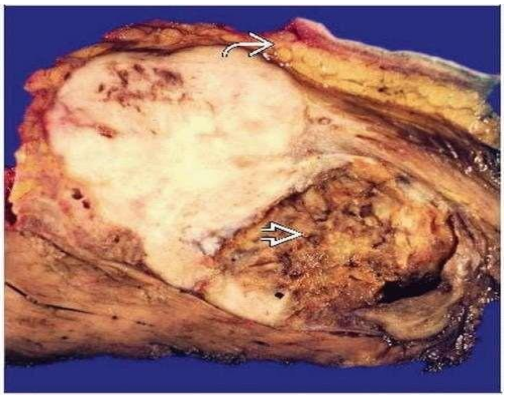
Cytokeratin + ve For glands

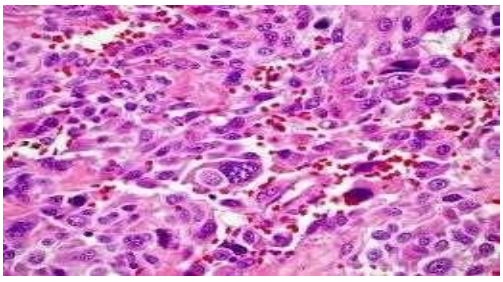
UNDIFFERENTIATED PLEOMORPHIC SARCOMA (UPS)

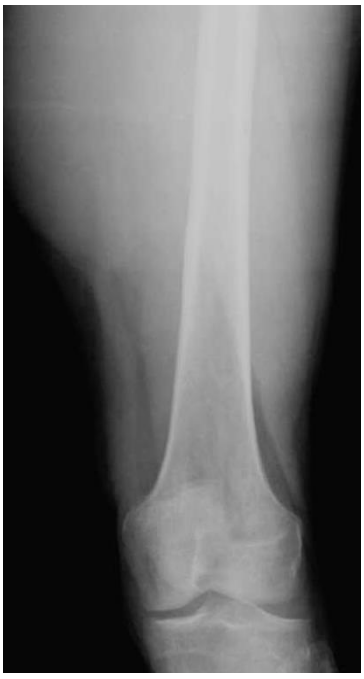
- High grade mesenchymal sarcomas of pleomorphic cells that lack cell lineage
- Deep soft tissue and extremities Retroperitoneum
- Old terminology: malignant fibrous histiocytoma (MFH)…not anymore
- Aneuploid and complex genetic abnormalities
- Large tumors; anaplastic and pleomorphic cells,abnormal mitoses, necrosis
- Trx: aggressive with surgery and adjuvant CT +/- RT; poor prognosis

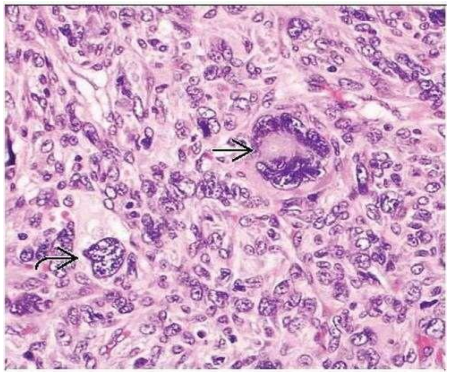
Can't know cell origin

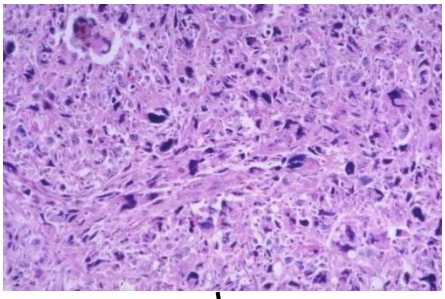
Stains are -ve











Very ugly bizarre abnormal cells

Summary

Soft Tissue Tumors

- The category of soft tissue neoplasia describes tumors that arise from non-epithelial tissues, excluding the skeleton, joints, central nervous system, and hematopoietic and lymphoid tissues. A sarcoma is a malignant mesenchymal tumor.
- Although all soft tissue tumors probably arise from pluripotent mesenchymal stem cells, rather than mature cells, they can be classified as
  - Tumors that recapitulate a mature mesenchymal tissue (e.g., fat). These can be further subdivided into benign and malignant forms.
  - Tumors composed of cells for which there is no normal counterpart (e.g., synovial sarcoma, UPS).
- Sarcomas with simple karyotypes demonstrate reproducible, chromosomal, and molecular abnormalities that contribute to pathogenesis and are sufficiently specific to have diagnostic use.
- Most adult sarcomas have complex karyotypes, tend to be pleomorphic, and are genetically heterogeneous with a poor prognosis.

"لا يطوي النسر جناحيه طالما أن هناك قمة لم يصلها بعد"

دمتم بتوفيق من الله وحفظ