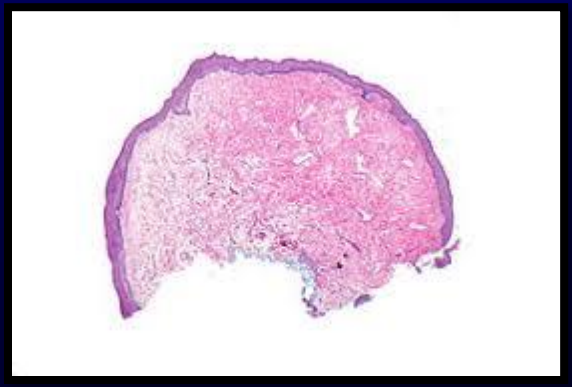
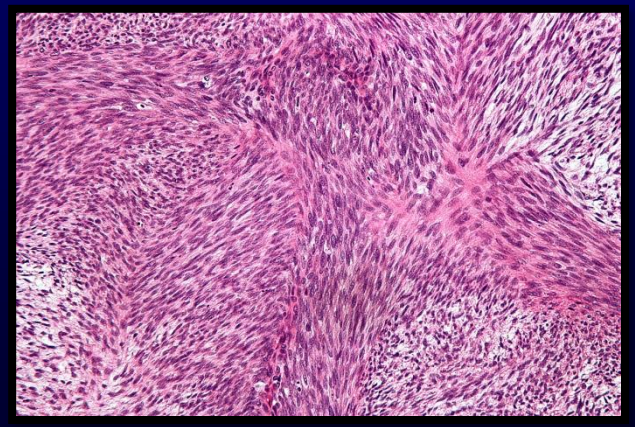
Lecture 10

FIBROMAS AND FIBROSARCOMAS:

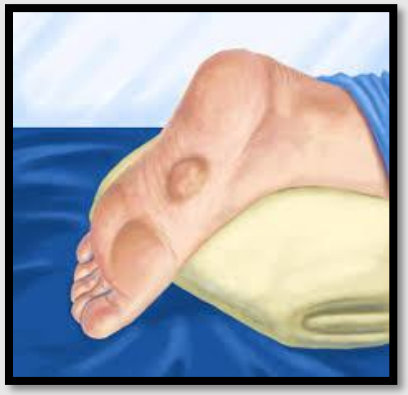
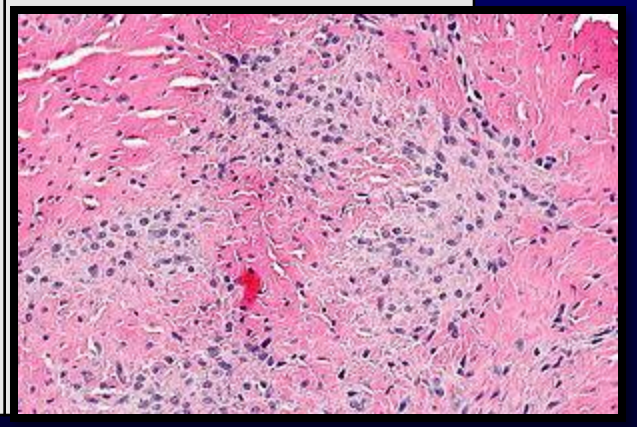
- **Fibromas: benign proliferation of fibroblasts, very common, skin and subcutaneous tissue**
- **Fibrosarcoma: malignant counterpart; usually superficial cutaneous tumors of fibroblasts, cellular, storiform pattern with increased mitosis**





SUPERFICIAL FIBROMATOSES:

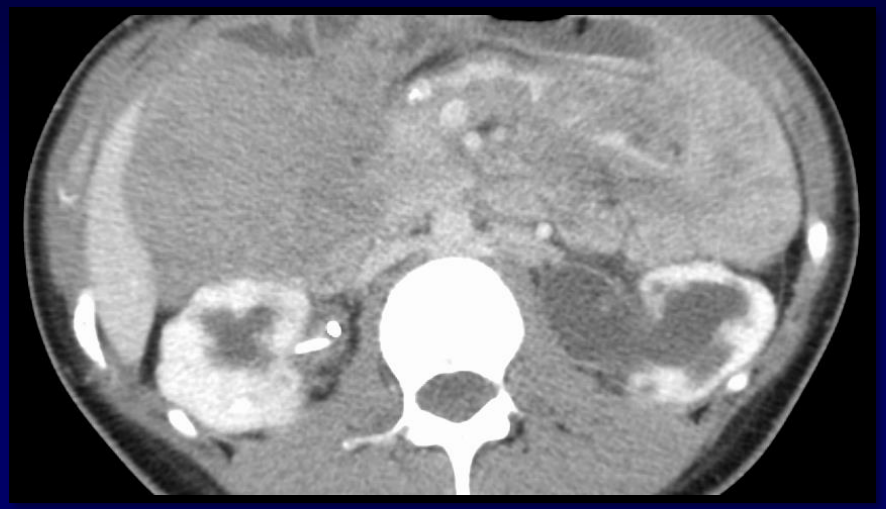
- **Infiltrative benign fibroblastic proliferation**
- **May run in families; may impact function**

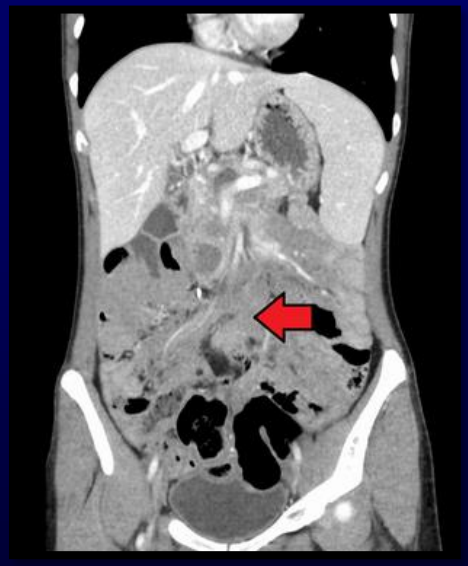
PALMAR (DUPUYTREN CONTRACTURE)	PLANTAR FIBROMATOSES	PENILE (PEYRONIE DISEASE)
Palmar fascia	Sole of foot	Dorsolateral aspect of the penis
		

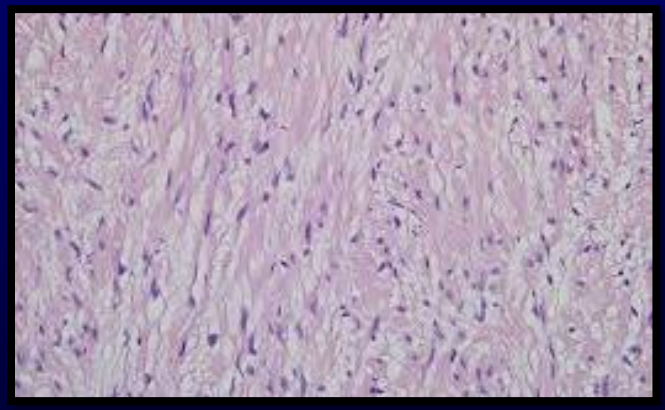
DEEP FIBROMATOSES (DESMOID TUMOR):

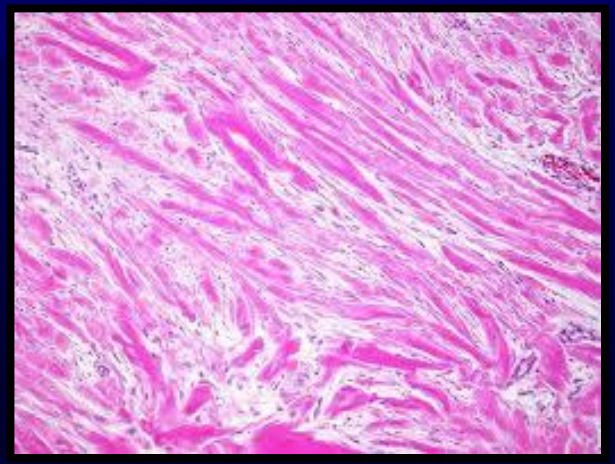
- **Deep infiltrative but bland fibroblastic proliferation; doesn't metastasize but recur**
- **20-30years, females more common**
- **Abdominal wall, mesentery and limbs**
- **Mutations in *CTNNB1* (β-catenin) or *APC* genes leading to increased Wnt signaling**
- **Mostly are sporadic; but patients with Gardner (FAP) syndrome are susceptible**
- **Complete excision is needed to prevent recurrence which is very common**
- **These tumors kill by local infiltration NOT metastasis**

DEEP FIBROMATOSES (DESMOID TUMOR):



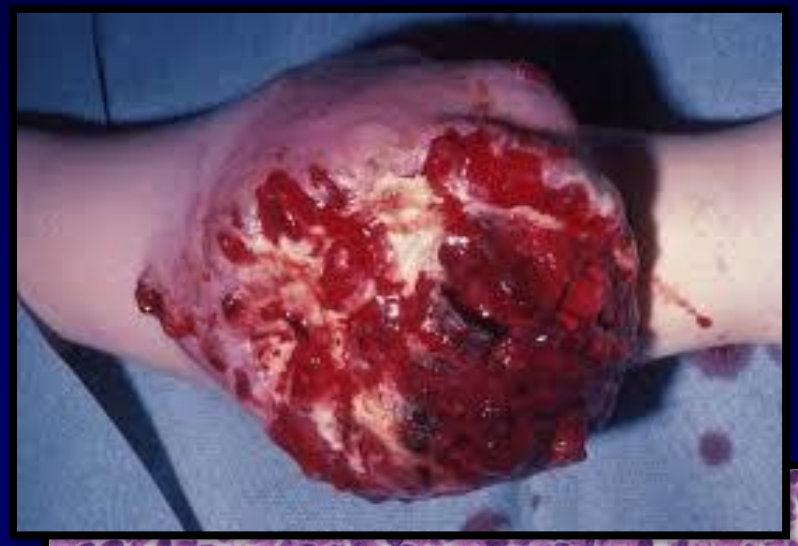


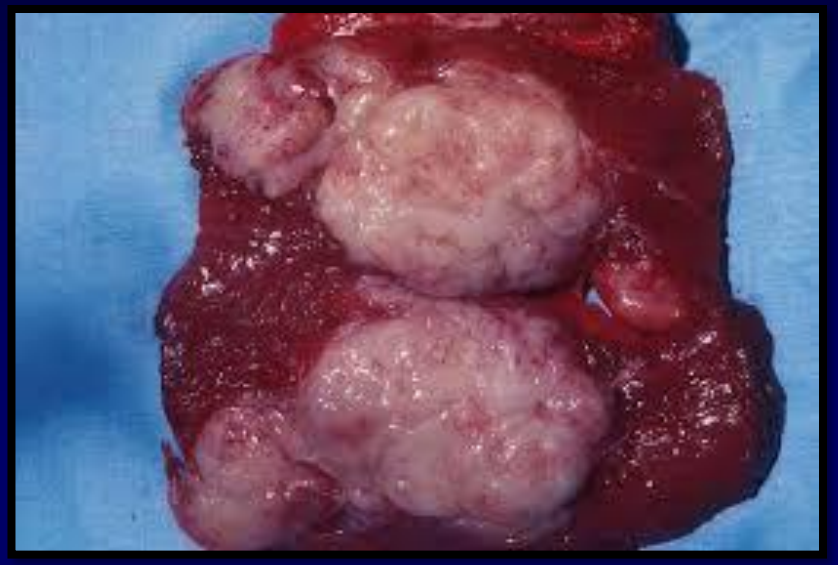


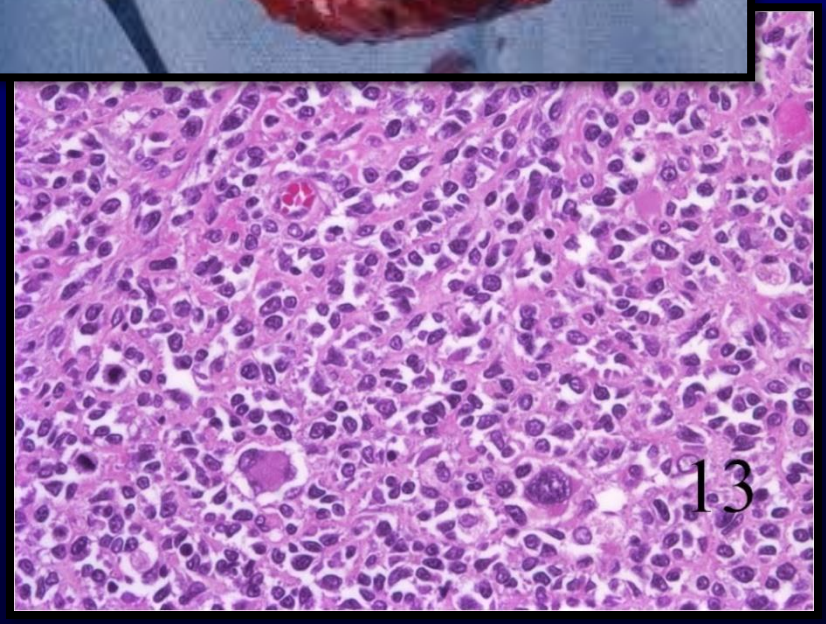


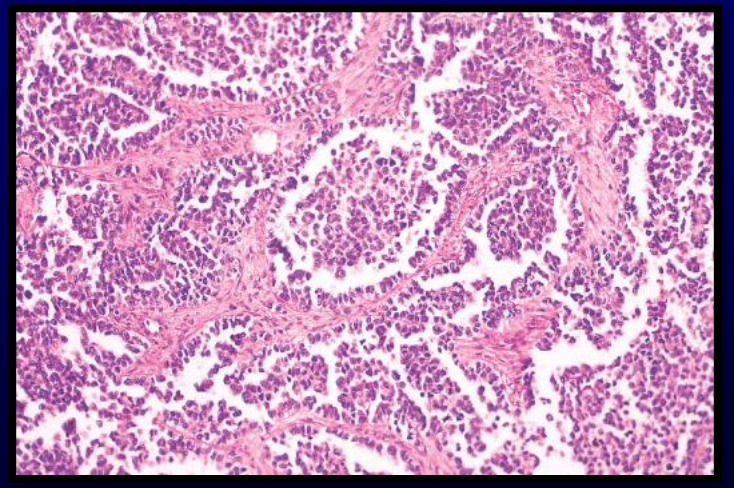
SKELETAL MUSCLE TUMORS:

- Almost all malignant; except rhabdomyoma which is benign, rare, occurs with tuberous sclerosis
- 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 +/- RT





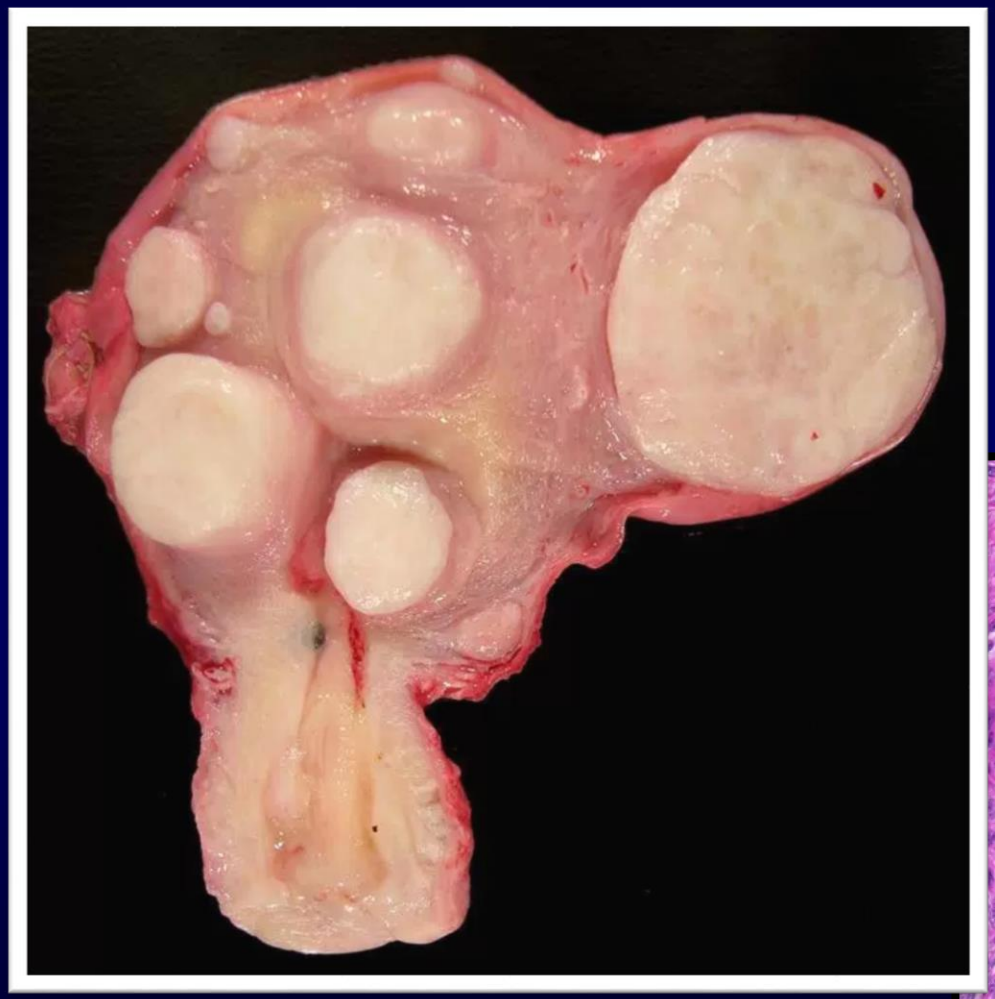


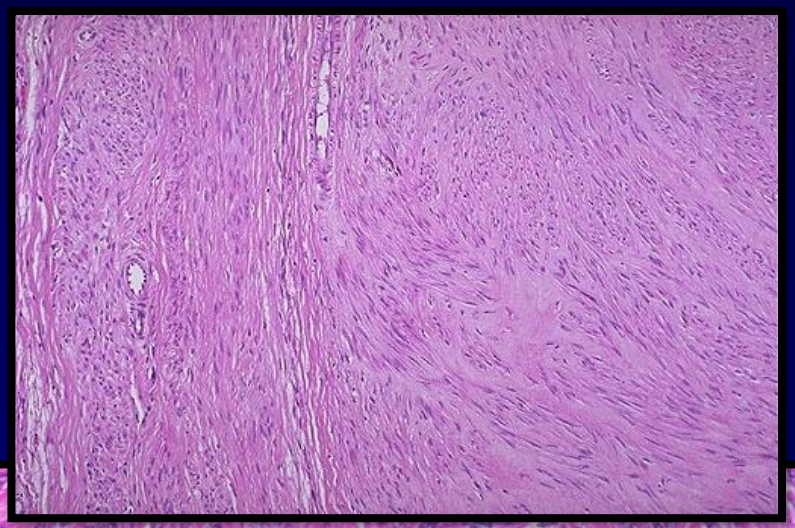


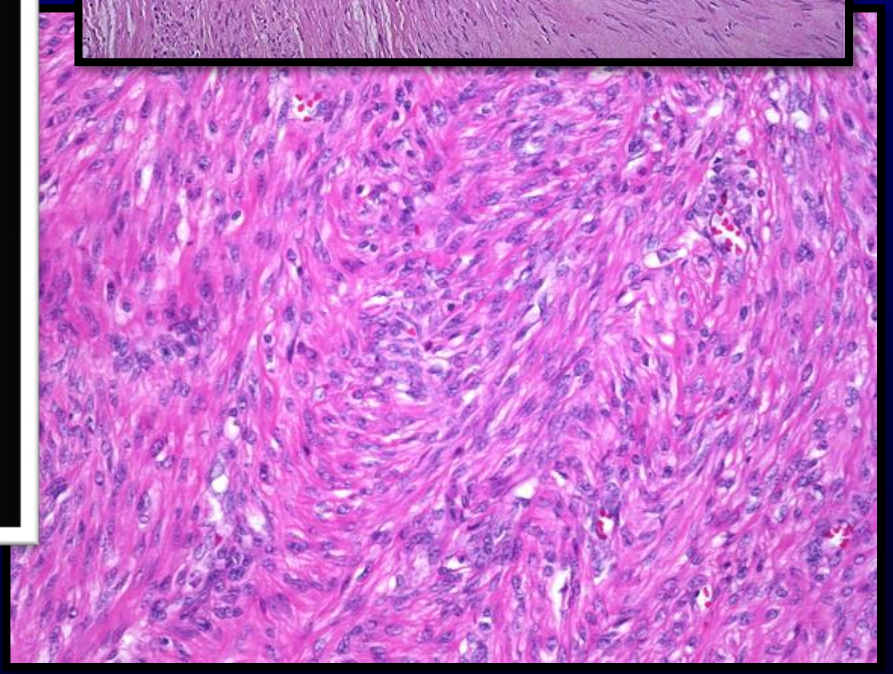
SMOOTH MUSCLE TUMORS:

- Leiomyoma (benign) and leiomyosarcoma (malignant)
- Leiomyoma (LYM): very common; any site but mostly uterus (fibroid)…menorrhagia and infertility
- LYM vary in size and location
- Few can have specific mutations (Fumarate hydratase on chromosome 1q42.3)

LEIOMYOMA FEATURES:



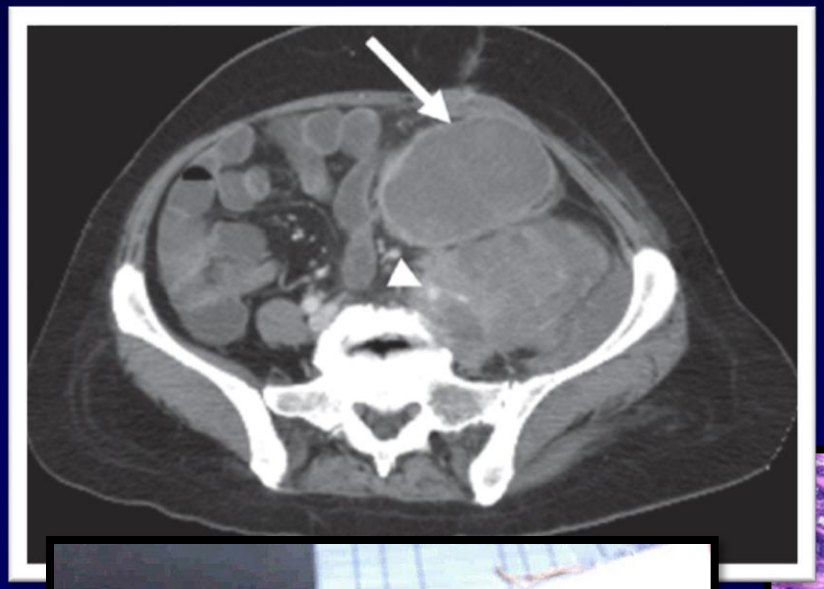


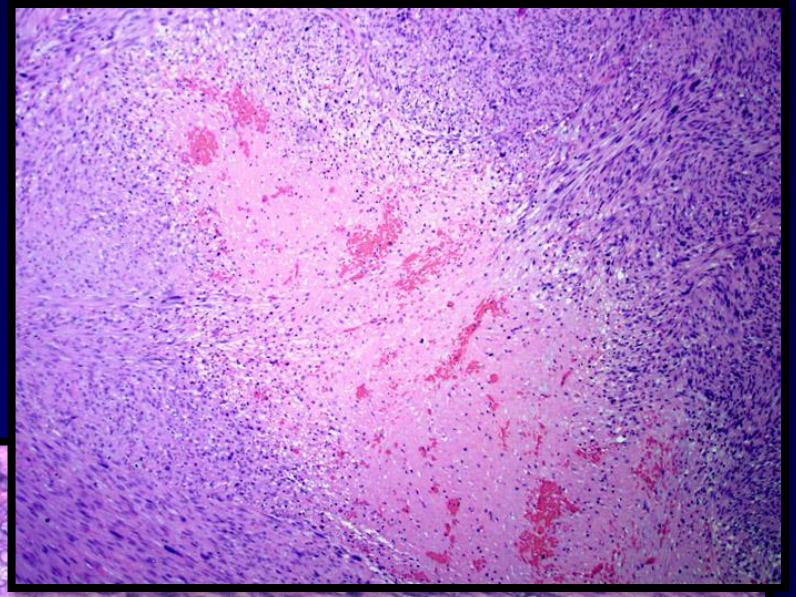


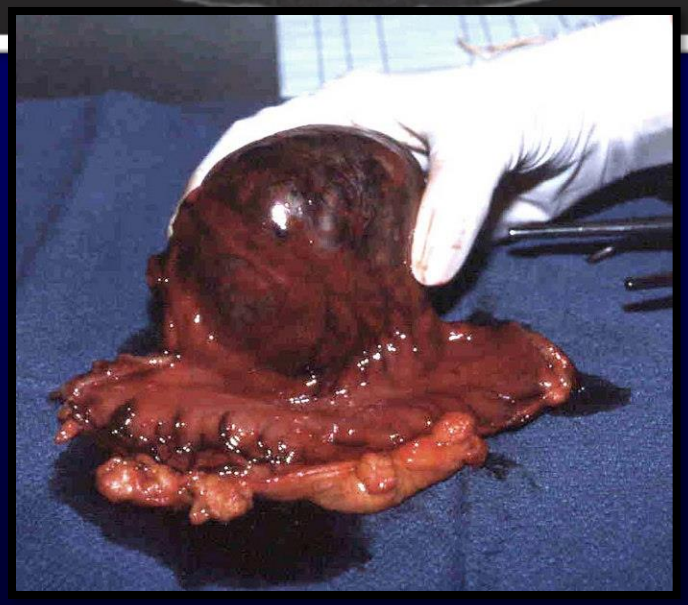
LEIOMYOSARCOMA:

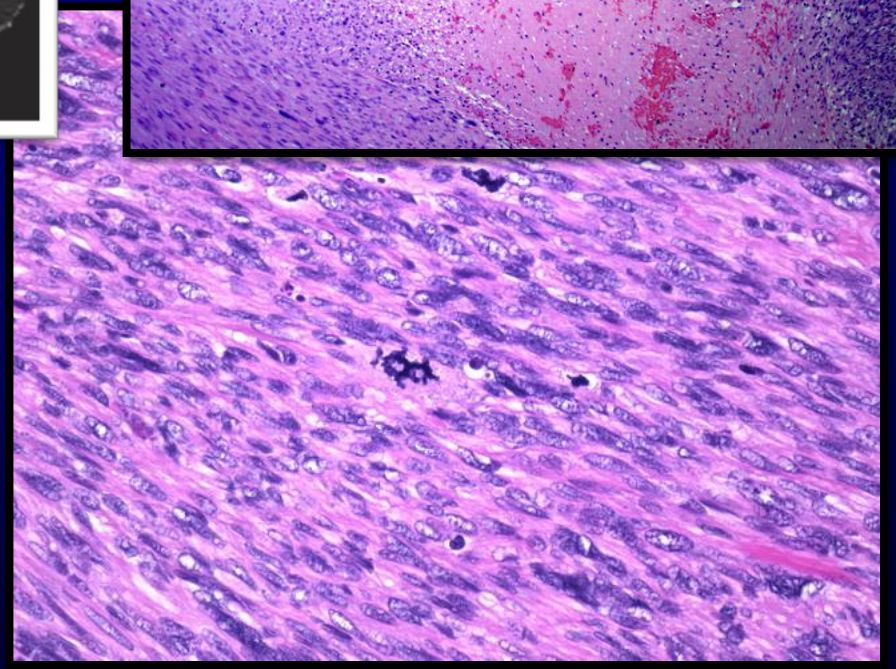
- 10-20% of soft tissue sarcomas
- Adults; more in females
- Deep soft tissue, extremities and retroperitoneum or from great vessels
- Complex genotypes
- Hemorrhage, necrosis, increased mitosis and infiltration of surrounding tissue
- Trx: depends on location, size and grade

LEIOMYOSARCOMA FEATYURES:









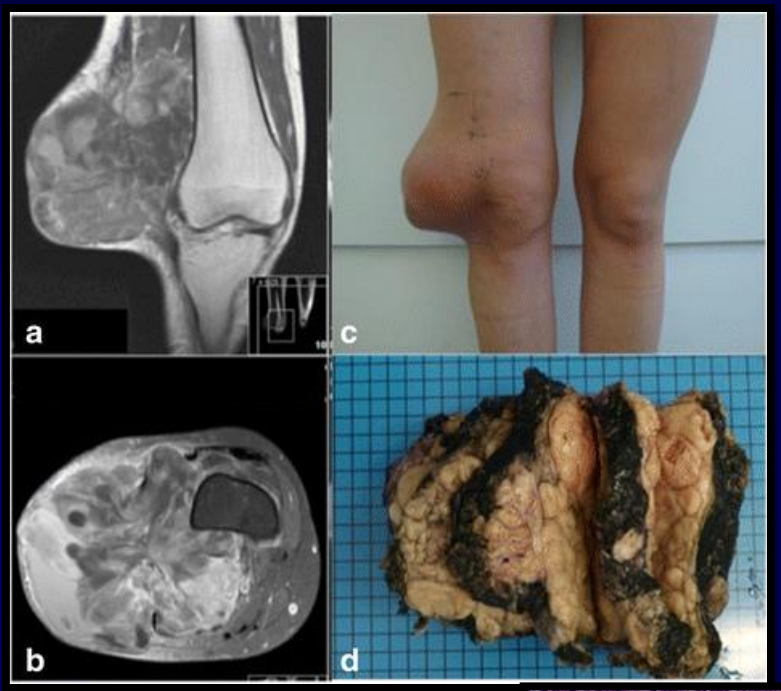
TUMORS OF UNCERTAIN ORIGIN:

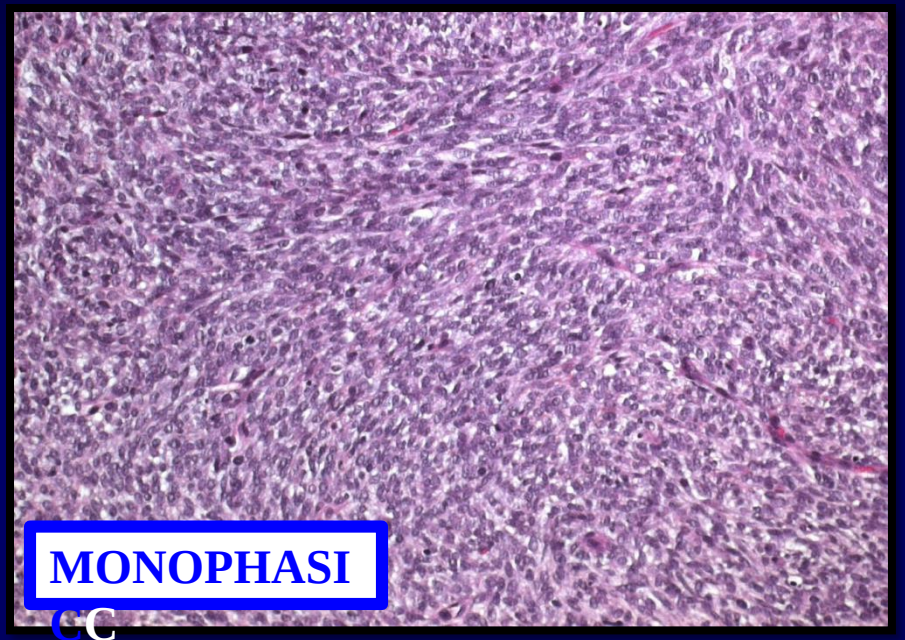
- Uncertain mesenchymal lineage
- Synovial sarcoma
- Undifferentiated pleomorphic sarcoma

SYNOVIAL SARCOMA:

- Name is misnomer
- 10% of all soft tissue sarcomas; 20-40s age
- Deep seated mass of long history
- T(X;18)(p11;q11) → fusion genes *SS18*…
- Monophasic (only spindle cells) or biphasic (spindle cells and glands)
- Trx: aggressive with limb sparing excision + CT
- 5 year survival 25-65% depending on stage
- Metastasis: lung and lymph nodes

SYN. SA. FEATURES:

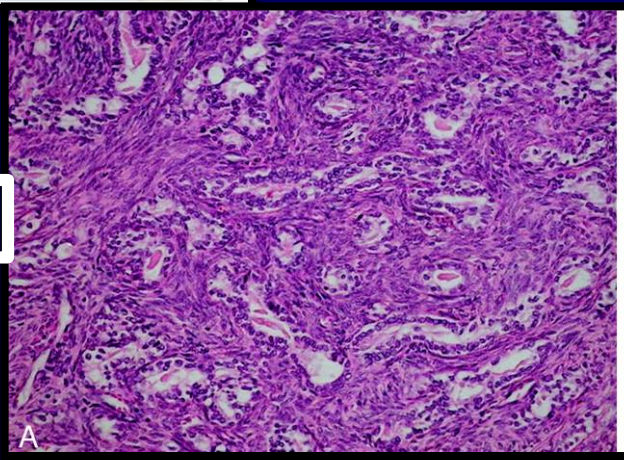


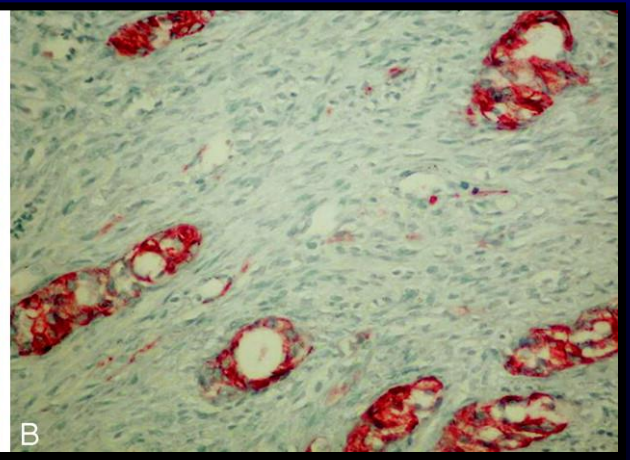


MONOPHASI

CC

BIPHASIC

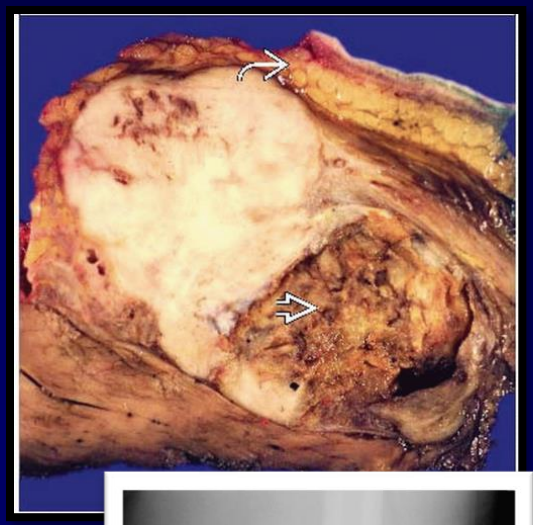


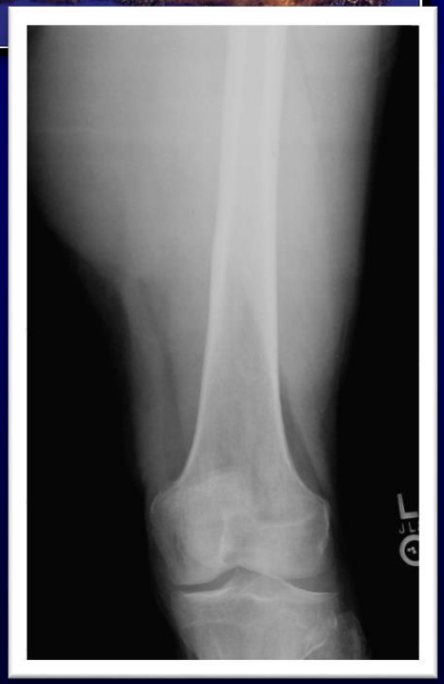


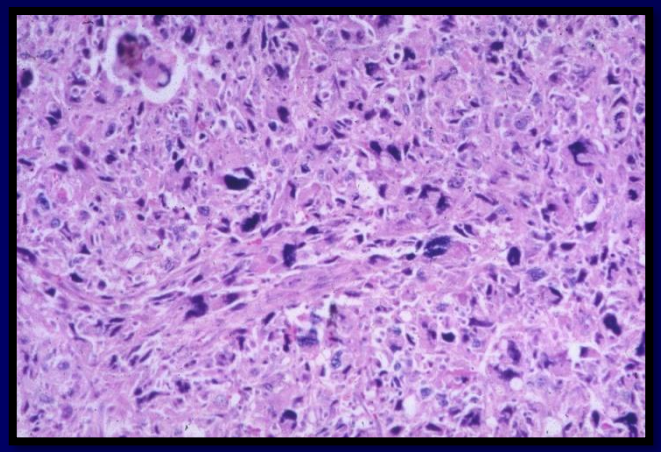
UNDIFFERENTIATED PLEOMORPHIC SARCOMA (UPS):

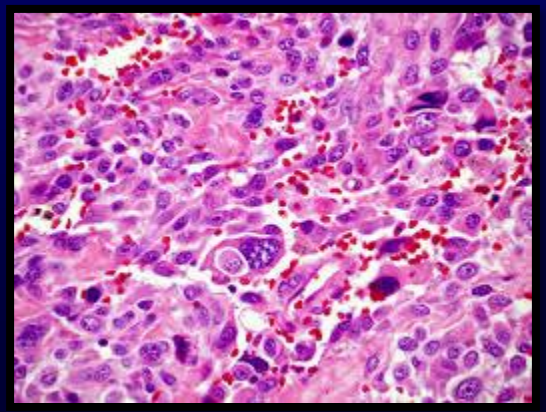
- High grade mesenchymal sarcomas of pleomorphic cells that lack cell lineage
- Deep soft tissue and extremities
- Old terminology: malignant fibrous histiocytoma (MFH)…not anymore
- Aneuploid and complex genetic abnormalities
- Large tumors; anaplastic and pleomorphic cells, abnormal mitoses, necrosis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

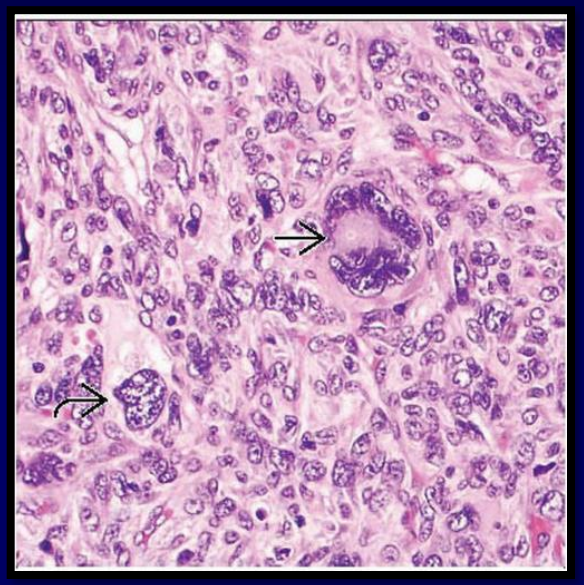
UPS FEATURES:











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.

GOOD

LUCK