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Pathology MSS

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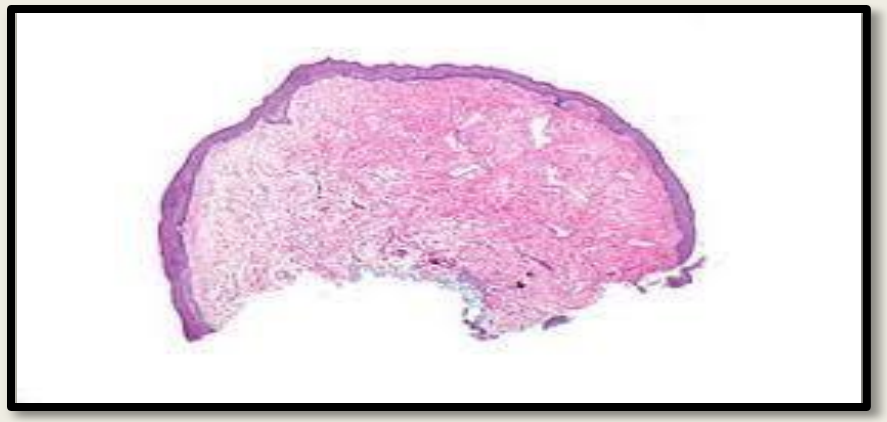
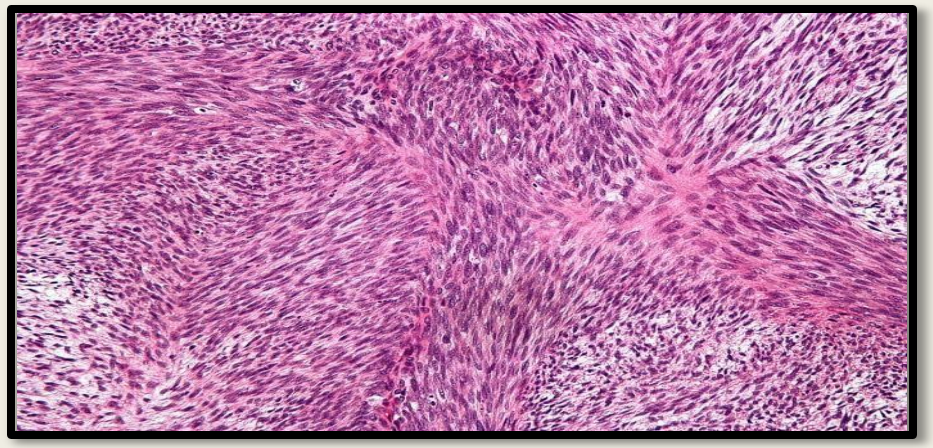
Lecture 10

FIBROMAS AND FIBROSARCOMAS:

(arise from fibroblasts)

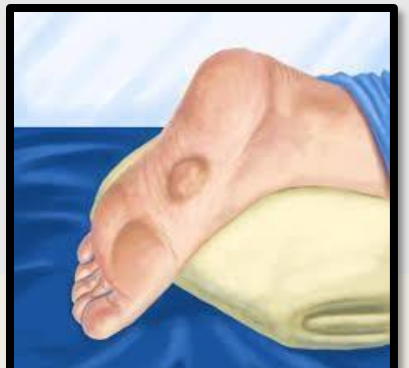
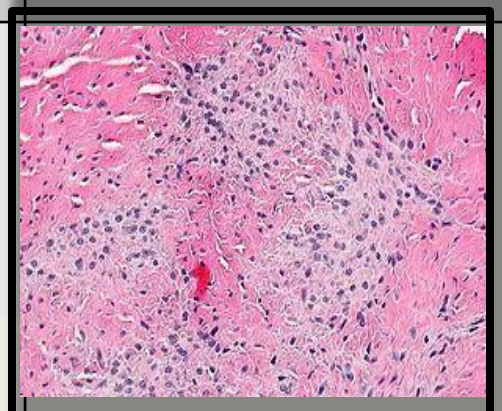
- **Fibromas (to the left): benign proliferation of fibroblasts, very common, skin and subcutaneous tissue (under the microscope they are planned benign appearing fibroblasts with immunostim chemical features)**
- **Fibrosarcoma (to the right) : malignant counterpart; usually superficial cutaneous tumors of fibroblasts, cellular, haphazard, storiform pattern with increased mitosis (positive fibroblast muscle marker is needed to confirm the case. Skeletal muscle marker and smooth muscle marker both are negative)**





SUPERFICIAL FIBROMATOSES (not deep in the soft tissue , can be examined by hand):

- **Infiltrative benign fibroblastic proliferation (no metastasis . But can embark local functions of that specific organ)**
- **May run in families; may impact function**

PALMAR (DUPUYTREN CONTRACTURE)	PLANTAR FIBROMATOSES	PENILE (PEYRONIE DISEASE)
Palmar fascia (in the palm , contracture-like, pain ,difficulty in flexing and extending the finger)	Sole of foot (pain, embarks walking)	Dorsolateral aspect of the penis
		

DEEP FIBROMATOSES (DESMOID TUMOR):

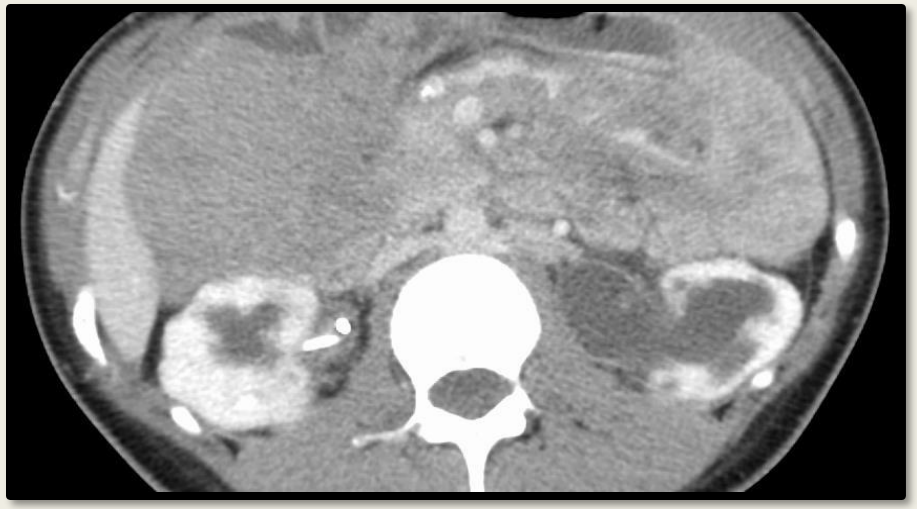
- **Deep infiltrative (not seen) but bland fibroblastic proliferation; doesn't metastasize but recur**
- **20-30years, females more common**
- **Abdominal wall, mesentery and limbs**
- **Mutations in *CTNNB1* (β-catenin); b-catenin stain positive or *APC* genes leading to increased Wnt signaling (patients with multiple polyps in the colon are at high risk of developing fibromatoses)**
- **Mostly are sporadic; but patients with Gardner (FAP) syndrome are susceptible**

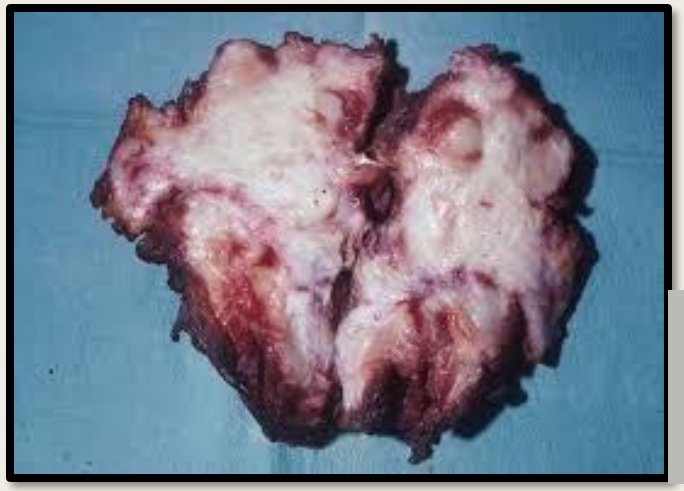
- **Complete excision is needed to prevent recurrence which is very common. (NOT easy ; the nature of the tumor makes it hard to determine the origin and end of the tumor in most cases. Hence the need of taking safe margins (4-5cm) during excision)**

- **These tumors kill by local infiltration (destroying kidneys, distal obstruction and different complications) NOT metastasis**

DEEP FIBROMATOSES (DESMOID TUMOR):

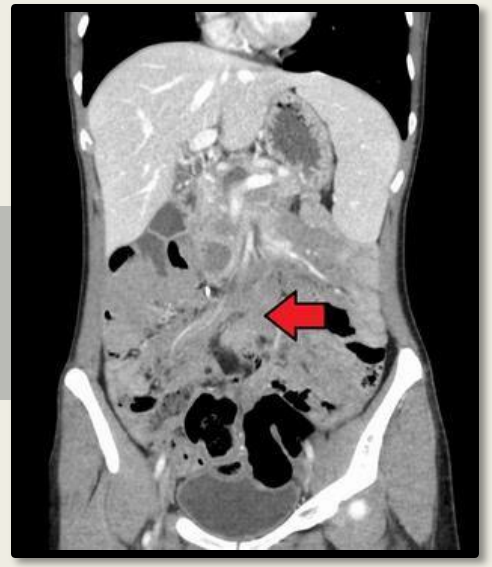
CT-Scan showing proliferations of gland appearing tissue

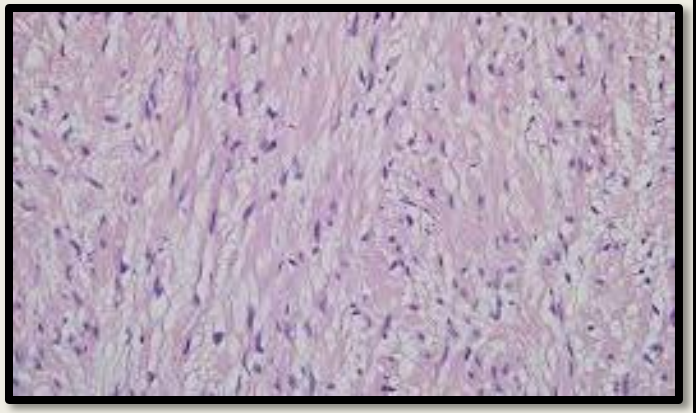


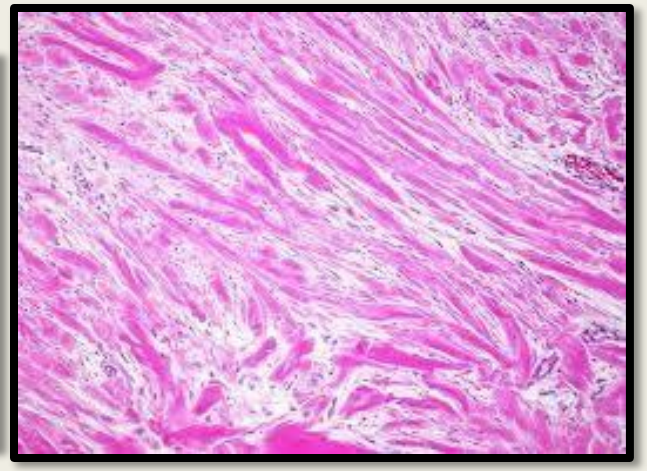


Whitish dense fibroblastic appearance.

Internal organs are destroyed



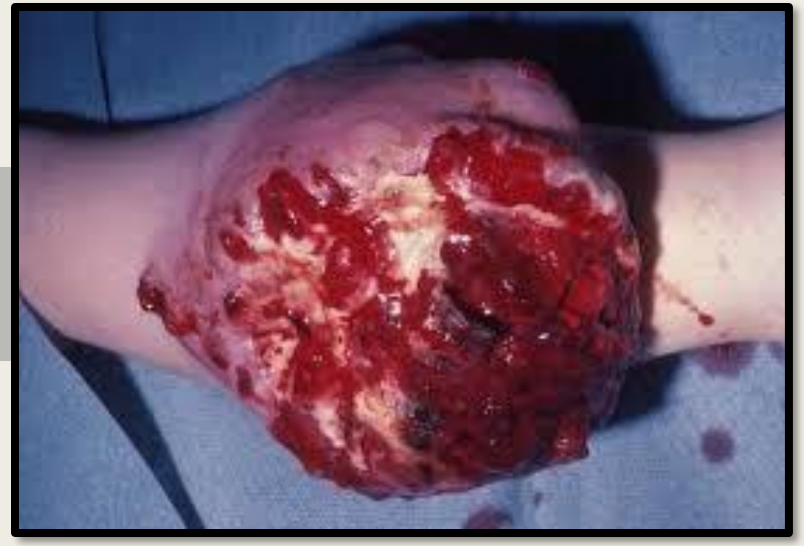


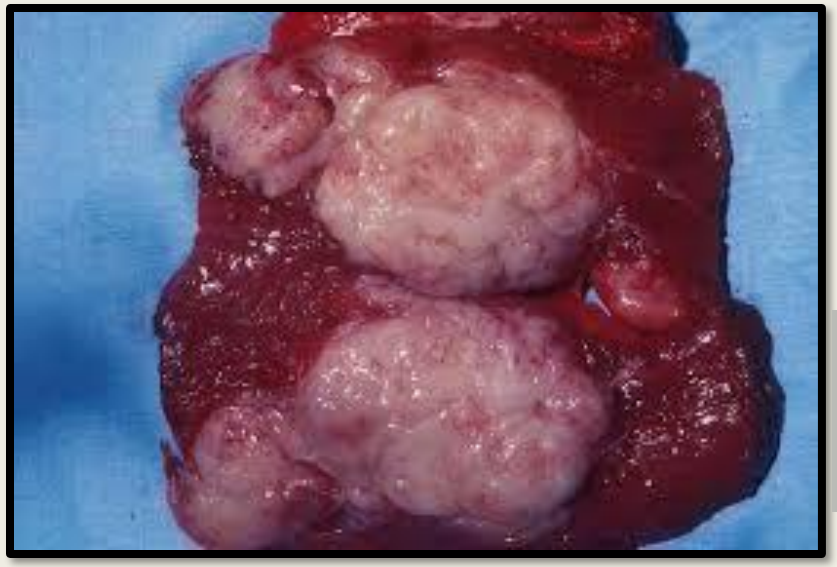


SKELETAL MUSCLE TUMORS:

- **Almost all malignant; except rhabdomyoma which is benign, rare, occurs with tuberous sclerosis (tung and heart are the most common organs)**
- **Rhabdomyosarcoma (RMS) is the malignant prototype; most common child sarcoma**
- **3 types (embryonal 60%; alveolar 20%; pleomorphic 20%)**
- **Specific mutations are common**
- **Aggressive tumors (high grade); treated by surgery, CT +/- RT**

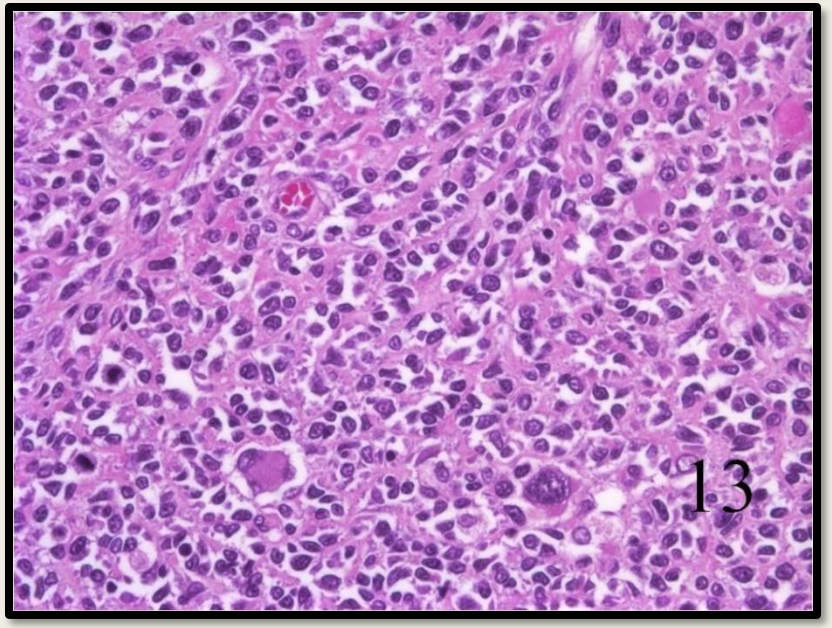
Large, fleshy, hemorrhagic tumor

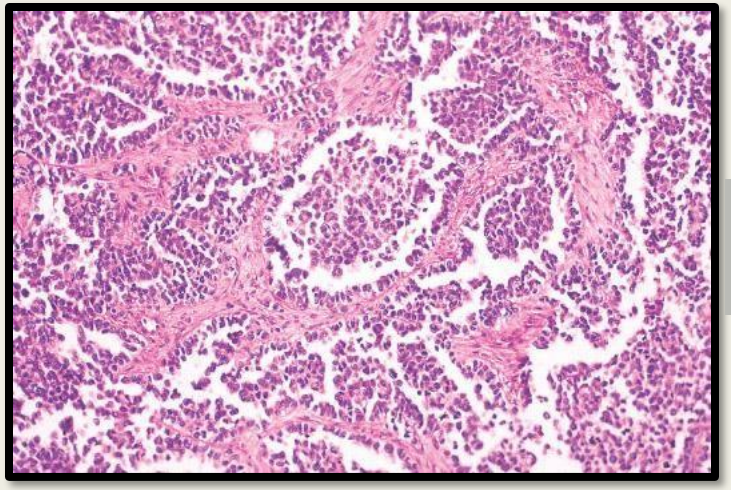




a lot of infiltrations

Cross-sections which tell that this is a skeletal muscle malignant tumor





Alveolar type

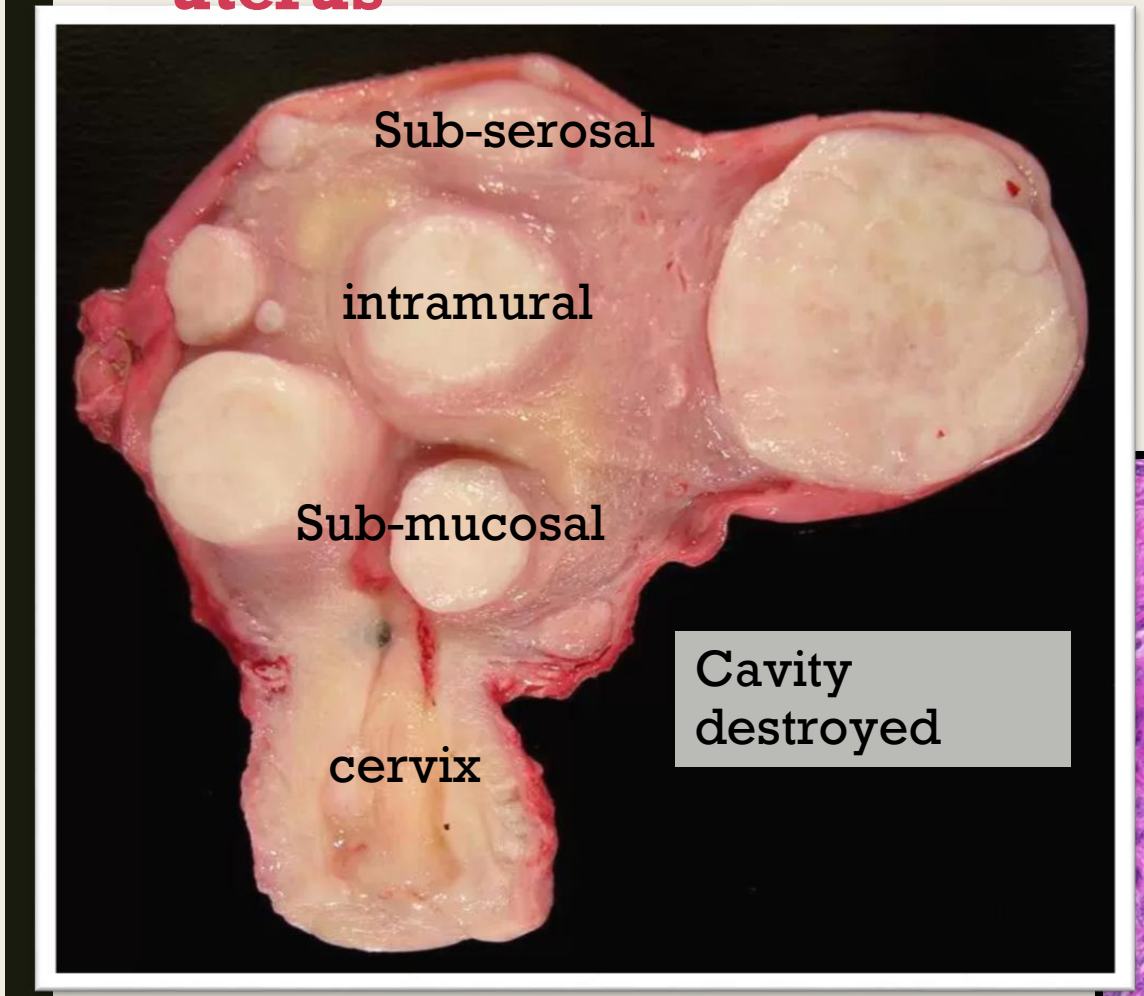
Polymorphic type composed of small blue cell tumors with rhabdomyosarcoma differentiations (pink cytoplasmic material)

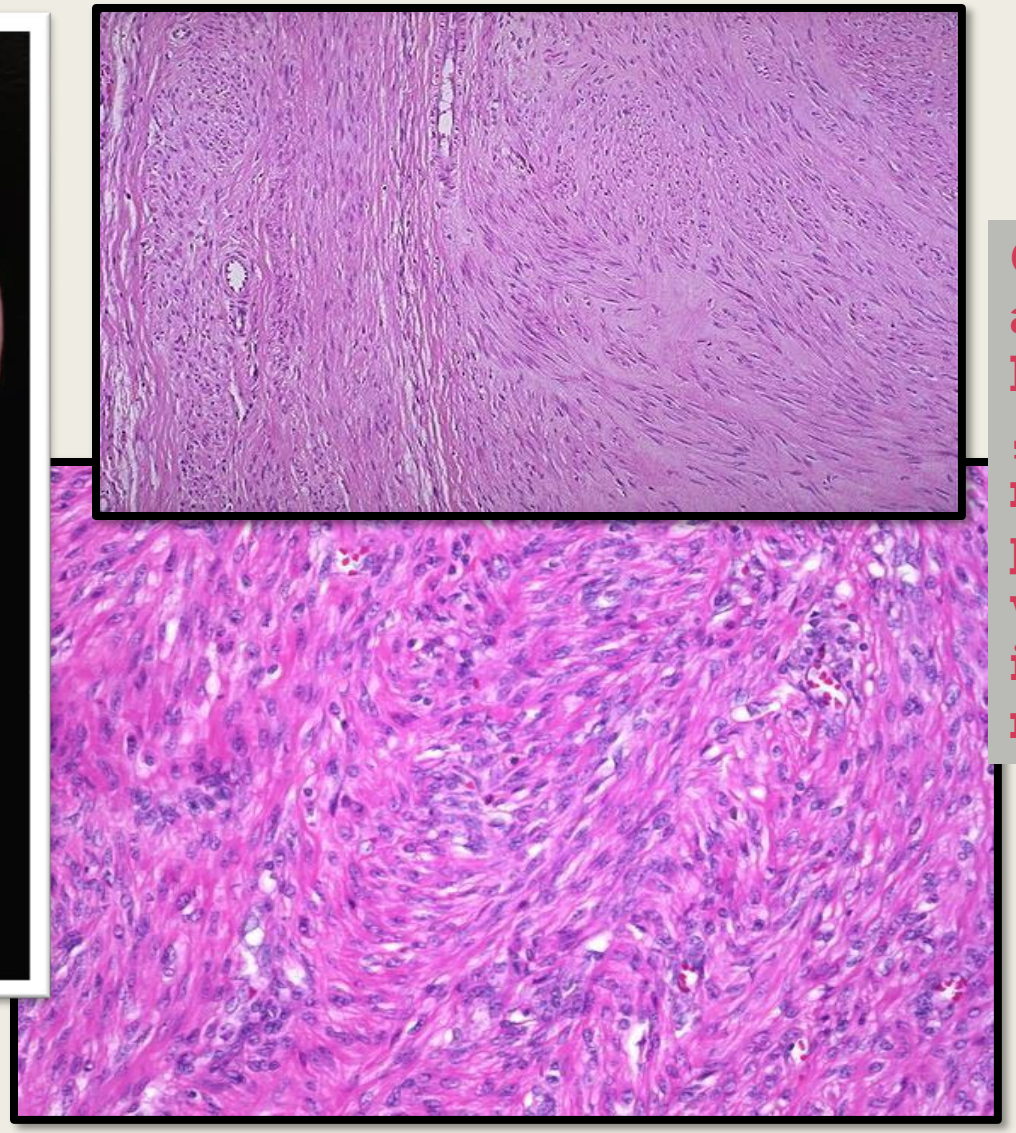
SMOOTH MUSCLE TUMORS:

- **Leiomyoma (benign) and leiomyosarcoma (malignant)**
- **Leiomyoma (LYM): very common; any site (appear in any organ) but mostly uterus (fibroid)…menorrhagia and infertility**
- **LYM vary in size and location (benign tumors are mostly well circumscribed, not infiltrative, no hemorrhagic necrosis, and firm white appearance).**
- **Few can have specific mutations (Fumarate hydratase on chromosome 1q42.3)**

LEIOMYOMA FEATURES:

uterus





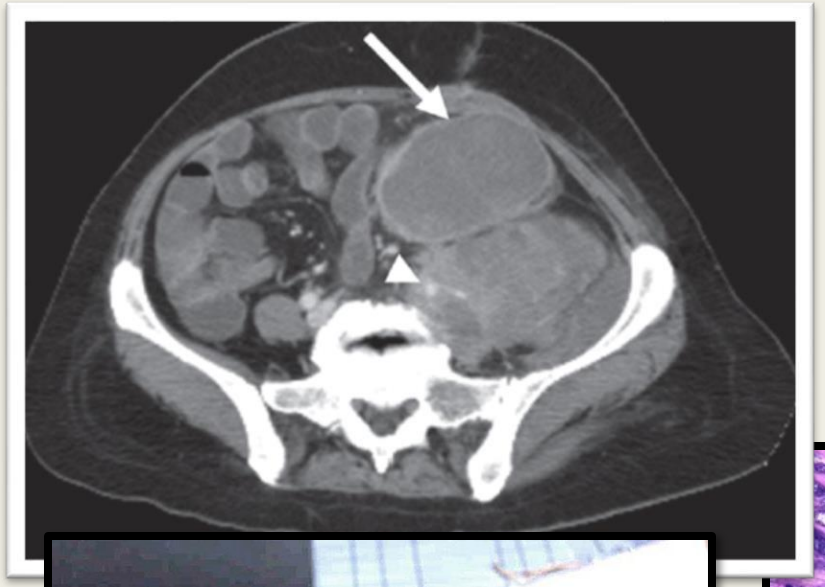
Classical appearance; benign ,smooth muscle proliferation with no increased mitosis.

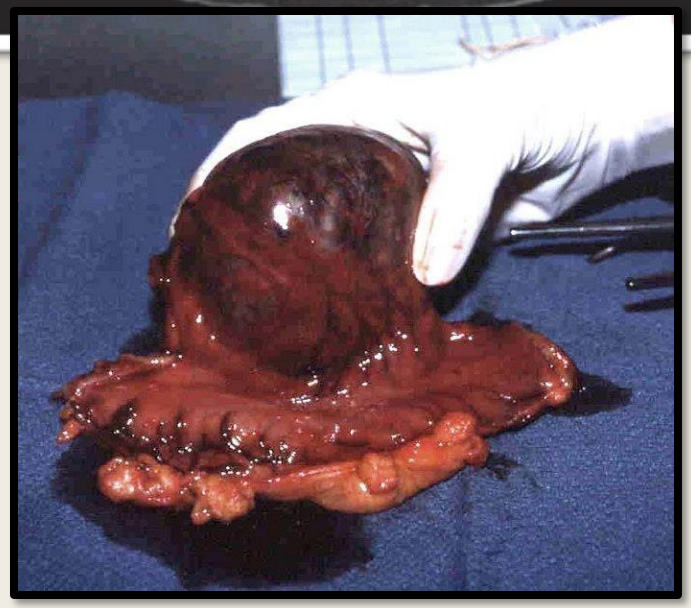
LEIOMYOSARCOMA (malignant counterpart):

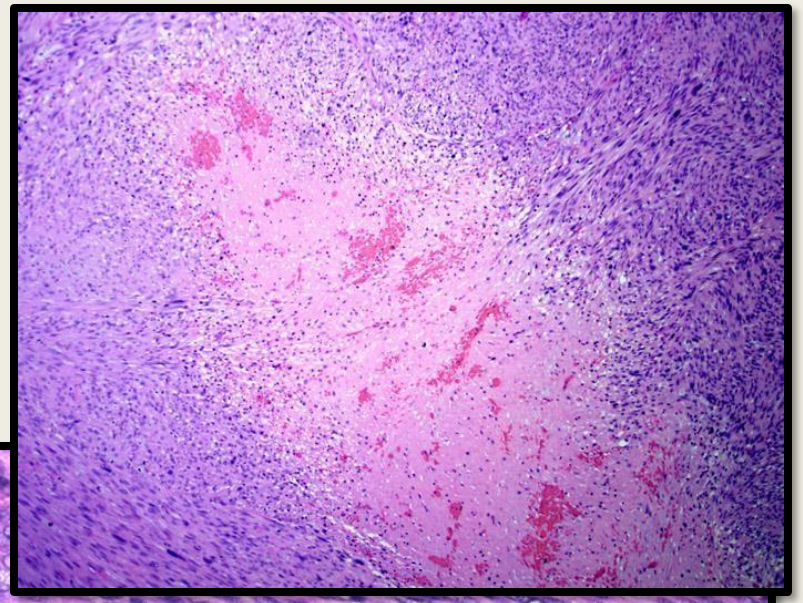
- **10-20% of soft tissue sarcomas**
- **Adults; more in females**
- **Deep soft tissue, extremities and retroperitoneum or from great vessels (major blood vessels),uterus**
- **Complex genotypes, no specific signature.**
- **Hemorrhage, necrosis, increased mitosis (abnormal/malignant) and infiltration of surrounding tissue**
- **Trx: depends on location, size and grade**

LEIOMYOSARCOMA FEATYURES:

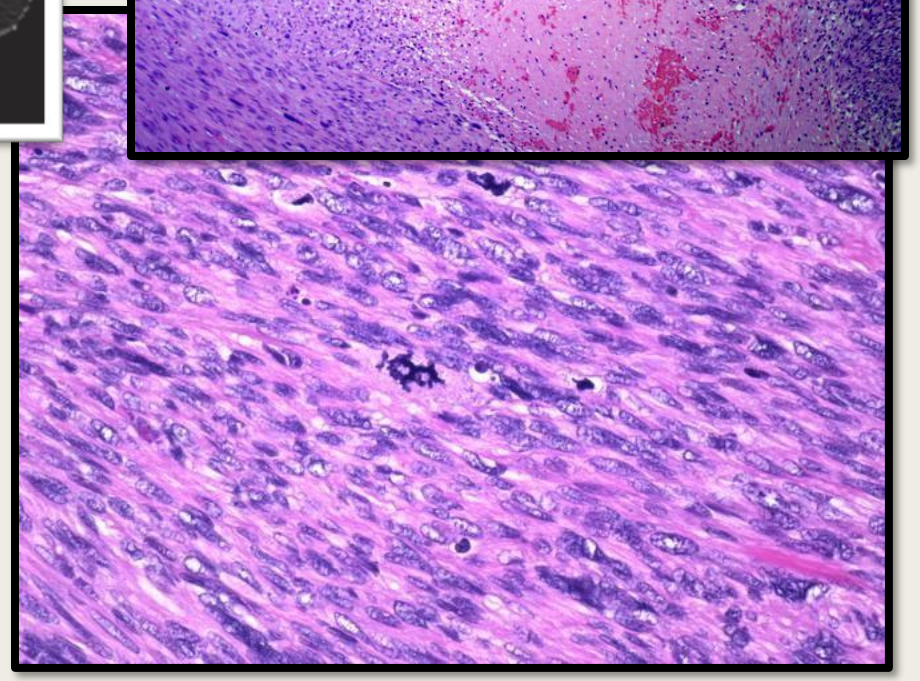
Big (10-15cm), spreading inside abdomen







Cellular, hemorrhage , necrosis



increased mitosi

TUMORS OF UNCERTAIN ORIGIN:

- **Uncertain mesenchymal lineage:**

1) Synovial sarcoma

2) Undifferentiated pleomorphic sarcoma

SYNOVIAL SARCOMA:

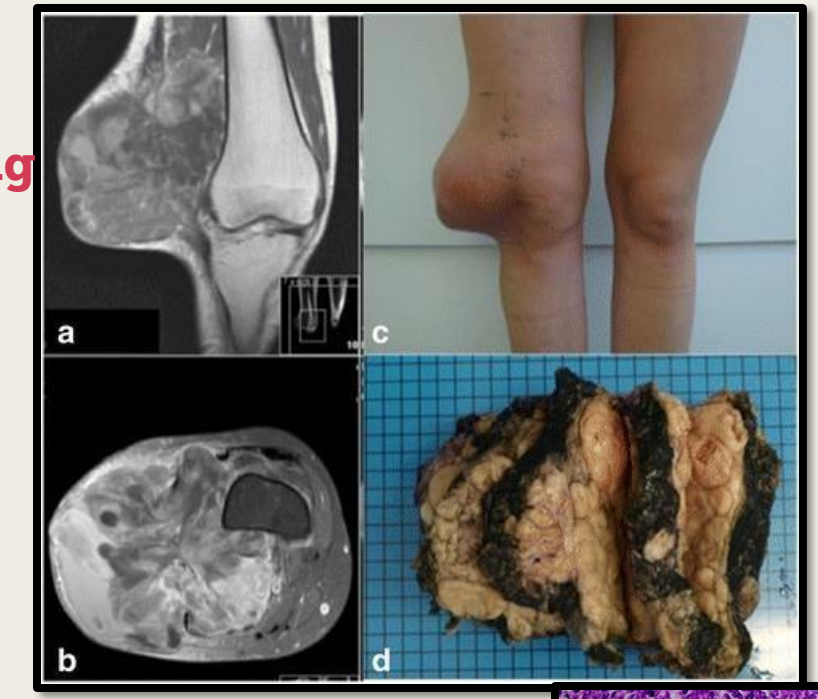
- Name is misnomer: it can occur anywhere
- 10% of all soft tissue sarcomas; 20-40s age (young adults)
- Deep seated mass of long history

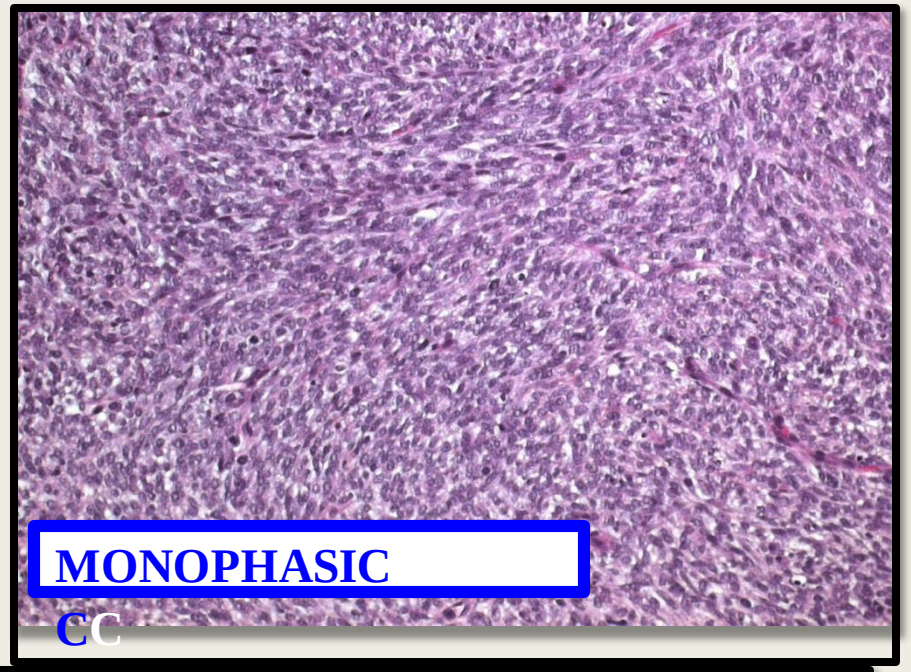
- **T(X;18)(p11;q11)** → **fusion genes *SS18*… (confirmed through FISH analysis)**

- **Monophasic (only spindle cells) or biphasic (spindle cells and glands)**
- **Trx: aggressive with limb sparing excision + CT (chemotherapy)**
- **5 year survival 25-65% depending on stage (and availability of multi-disciplinary team approach of these tumors)**
- **Metastasis: lung and lymph nodes**

SYN. SA. FEATURES:

Fleshy big mass



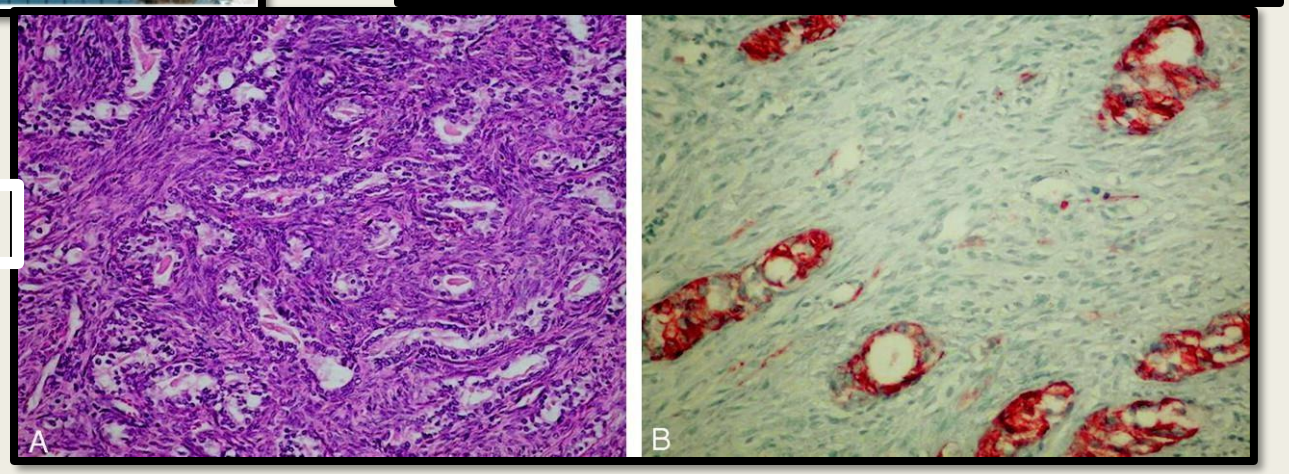


MONOPHASIC

CC

Spindle cells

BIPHASIC



Glands, keratin stain positive; epithelial markers

UNDIFFERENTIATED PLEOMORPHIC SARCOMA (UPS):

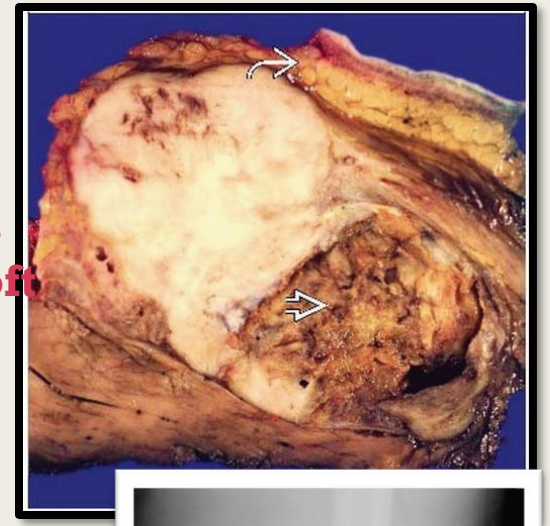
- **High grade mesenchymal sarcomas of pleomorphic cells that lack cell lineage**
- **Deep soft tissue and extremities**
- **Old terminology: malignant fibrous histiocytoma (MFH)…not anymore**

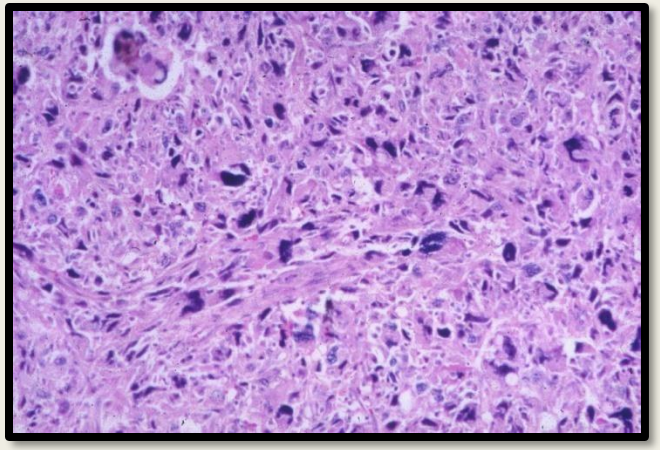
- **Aneuploid (abnormal DNA content) and complex genetic abnormalities (5-10/20 abnormalities)**

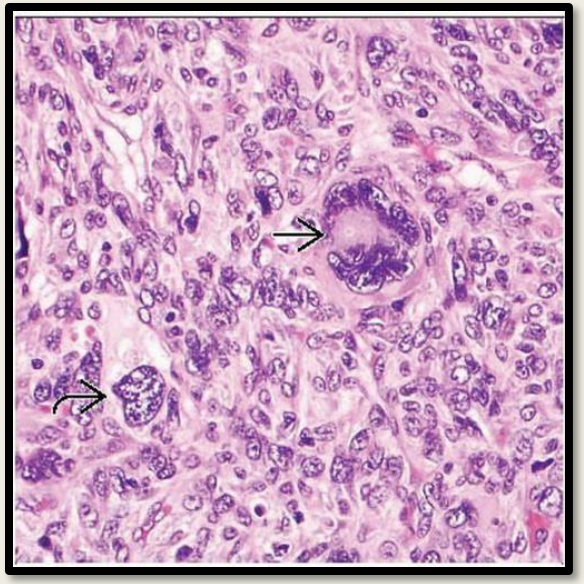
- **Large tumors; anaplastic and pleomorphic cells (ugly looking appearance), abnormal mitoses, necrosis**
- **Trx: aggressive with surgery and adjuvant CT +/- RT; poor prognosis**

UPS FEATURES:

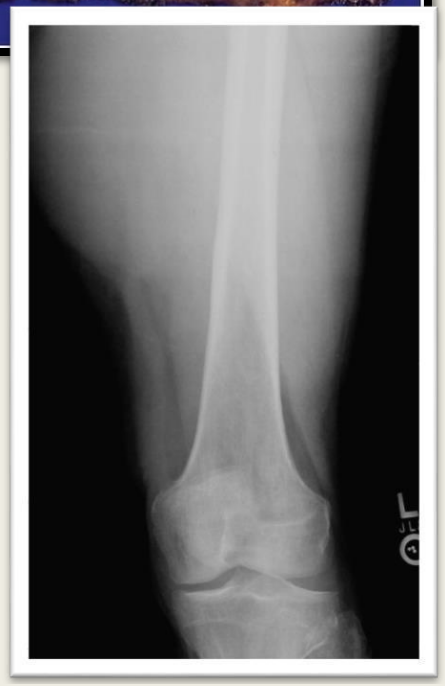
Large infiltrative ,deep in soft tissue

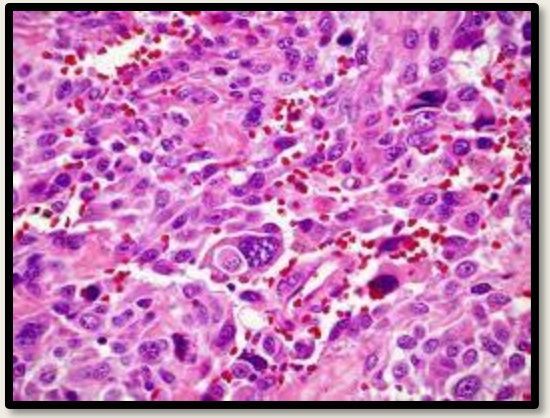






Thigh UPS





high grade UPS

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