SUPERFICIAL FIBROMATOSES:

proliferation of fibroblasts (Fibromatoses syndromes), has 2 types:

1. superficial fibromatoses:

- in cutaneous and subcutaneous area
- infiltrative but benign (they do not metastasize)
- Hereditary

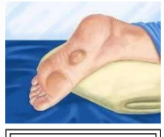
Has 3 major forms:

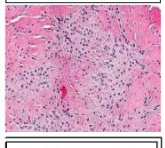
A. **Palmar fibromatoses (DUPUYTREN CONTRACTION)**, in the palmar fascia in the hand.

B. **Planter fibromatoses:** in the sole of the foot.

C. **Penile fibromatosis (PEYRONITE DISEASE)**: in the dorsolateral aspect of the penis.

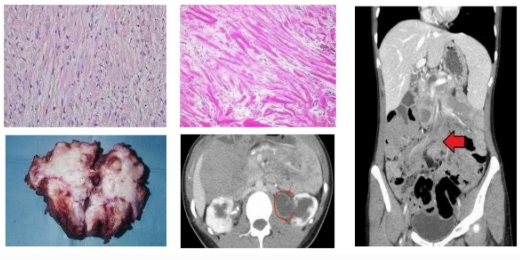

Palmar fibromatoses


Planter fibromatoses


Penile fibromatosis

2. deep fibromatoses (DESMOID TUMOR):

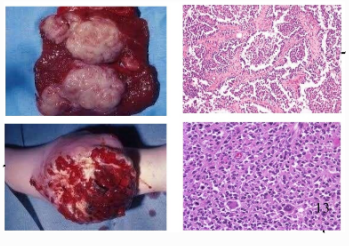
- deep inside the tissue, invisible.
- Lethal, kill by local infiltration NOT metastasis.
- 20-30 years, females (more).
- in abdominal wall, mesentery and limbs.
- mutations in **CTNNB1** that produce a fusion protein called **β-catenin**.
- or mutations in **APC** (Adenomatous polyposis coli) genes leading to increased **Wnt** signaling.



- sporadic; (mostly), but patients with Gardner (FAP -familial adenomatous polyposis syndrome) are susceptible.
- **Trx**: complete excision.

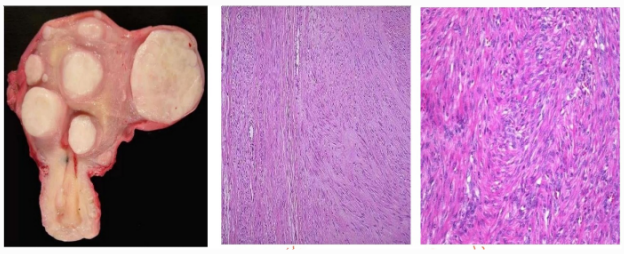
SKELETAL MUSCLE TUMORS:

- all are malignant; except rhabdomyoma (benign).
- **rhabdomyoma:** benign, occurs with **tuberous sclerosis** in the brain and on other vital organs, common locations of rhabdomyomas is the tongue and the heart.
- **Rhabdomyosarcoma (RMS):** malignant; in **children**, has 3 types (embryonal 60%; alveolar 20%; pleomorphic 20%), small bleu cell tumor, in EM you can see the cross striations of the skeletal muscles. Grossly it is large, fleshy and hemorrhagic tumor.



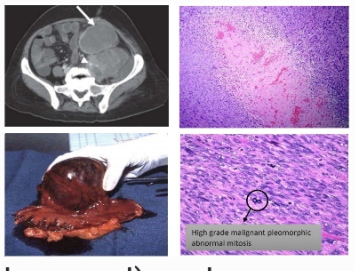
SMOOTH MUSCLE TUMORS:

1. Leiomyoma (LYM):



- benign
- **Uterus** (mostly).
- causes menorrhagia and infertility.
- 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**.

2. LEIOMYOSARCOMA:

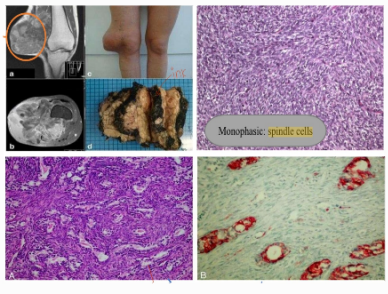


- malignant
-In adults; **females** (more).
- in deep soft tissue, extremities and retroperitoneum or from great vessels and uterus.
• Complex genotypes
- Causes **hemorrhage, necrosis, increased mitosis** (many of them are abnormal) and infiltration of surrounding tissue (**infiltrative**).
- **Trx**: depends on location, size and grade
- Histology: very **cellular with clear hemorrhage and necrosis.**

TUMORS OF UNCERTAIN ORIGIN:

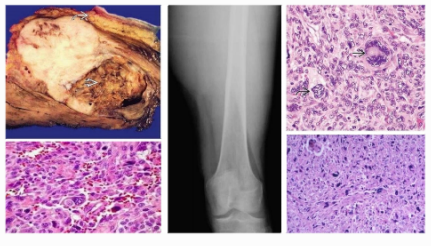
1. Uncertainmesenchymal lineage.

2. Synovial sarcoma (misnomer): around joints (mostly), but can occur anywhere



- 20-40s age.
- Translocation **T(X;18) (p11;q11)**, Makes fusion genes **SS18**.
Histologically: **Monophasic** (only spindle cells) or **biphasic** (spindle cells and glands (epithelial cells))
• **Trx**: aggressive with limb sparing excision + CT
- 5 years survival 25-65% depending on stage.
- **Metastasis**: lung and lymph nodes.

3. Undifferentiated pleomorphic sarcoma (UPS):



- Old terminology: malignant fibrous histiocytoma (MFH).
- **High grade** mesenchymal sarcomas of pleomorphic cells that lack cell lineage
- In deep soft tissue and extremities.
- Aneuploid and complex genetic abnormalities
- **Large tumors; anaplastic and pleomorphic cells, abnormal mitoses, necrosis.**
- Very ugly bizarre abnormal cells, **bad prognosis.**