Pathology

Doctor 2017 | Medicine | JU | MSS

Number >>	9
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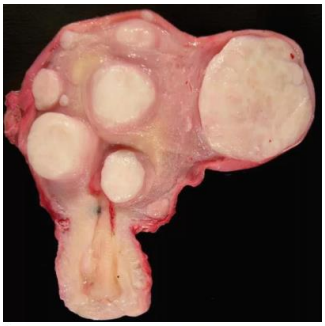
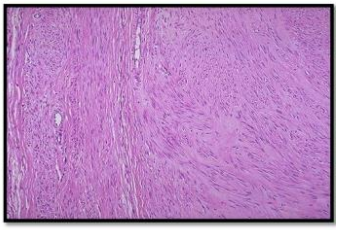
1st system - MSS

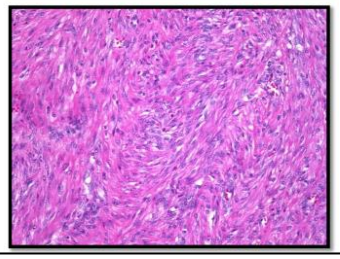
Smooth Muscle Tumors

Leiomyoma (benign) and leiomyosarcoma (malignant).

- **Leiomyomas** (very common) are benign tumours of smooth muscle, they **vary in size and location**, and can arise in any soft tissue site, but are **most common in the uterus**.
  - Uterine leiomyomas are common and may cause a variety of symptoms including **infertility** (masses impede proper implantation) and **menorrhagia** (heavy cycle bleeding, raises risk of anaemia).
  - **Features**: since they are benign, they are: well circumscribed, white (no aggressive angiogenesis), firm, and are most commonly found in uterine wall.
  - **Histological appearance**: normal bland (poor basophilia means no mitosis) smooth muscle with no necrosis, haemorrhage, or mitosis.
  - Few can have specific mutations: mutation in the fumarate hydratase (**FH**) **gene** located on chromosome **1q42.3**.
  - **DO NOT TURN MALIGNANT (DO NOT TURN INTO LEIYOMYOSARCOMAS).**

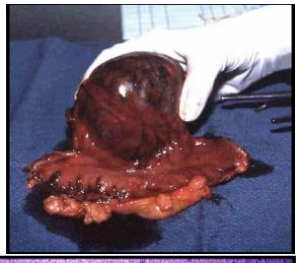


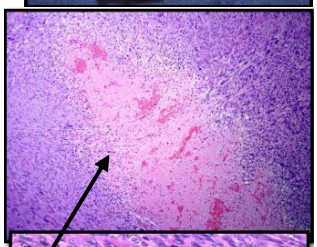


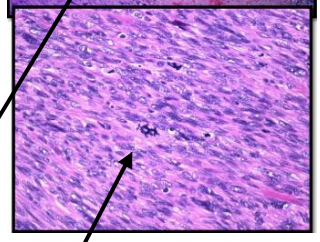


Notice the bland looking normal smooth muscle tissue.

- **Leiomyosarcomas** occur **in adults** and affect women more frequently than men. They account for 10% to 20% of soft tissue sarcomas.
  - Most develop in the **deep soft tissues of the extremities and retroperitoneum or arise from the great vessels**, also very **common in the uterus** → Flesh-like.
  - Leiomyosarcomas have complex genotypes.
  - **Hematogenous metastasis**.
  - **Histological appearance**: Hemorrhage, necrosis, increased mitosis and infiltration of surrounding tissue.
  - **Unlike leiomyomas, mitotic activity and necrosis are common in leiomyosarcoma**.
  - **Rx**: depends on location, size and grade







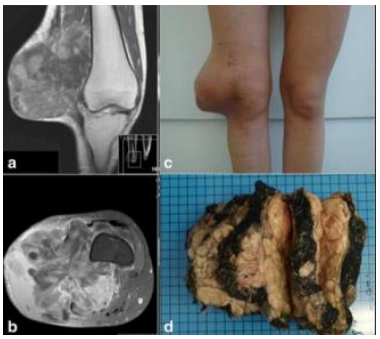
Necrosis

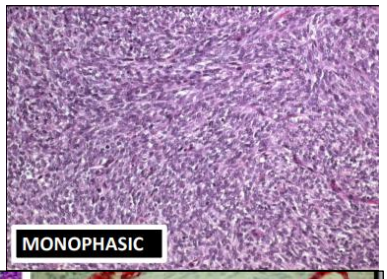
↑ Mitosis

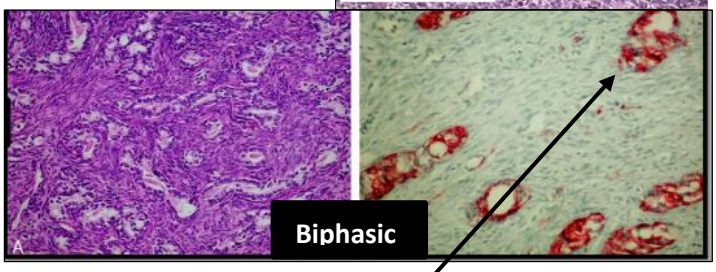
Tumors of Uncertain Origin

All have an **uncertain mesenchymal lineage**.

- **Synovial sarcomas**, their name is misnomer, because they do not actually have any connection to the synovium (they aren't arising from synoviocytes) except for they commonly occur near joints.
  - They are common tumors, and account for approximately 10% of all soft tissue sarcomas. Most occur in people in their 20s to 40s.
  - Patients usually present with a **deep-seated mass that has been present for several years**.
  - Most synovial sarcomas **show a characteristic chromosomal translocation t(x;18)(p11;q11)** producing fusion genes composed of portions of the *SS18* gene.
  - **Histological appearance**: can be **monophasic** (only spindle cells) or **biphasic** (spindle cells and glands), look like myosarcomas, notice the aggressive mitotic activity.
  - Synovial sarcomas are **treated aggressively with limb sparing surgery and frequently chemotherapy**. The 5-year **survival** varies from 25% to 62%, **related to stage**. Common sites of **metastases are the lung and lymph nodes** (usually only carcinomas met. to lymph nodes while sarcomas met. to the lungs, this is an exception).



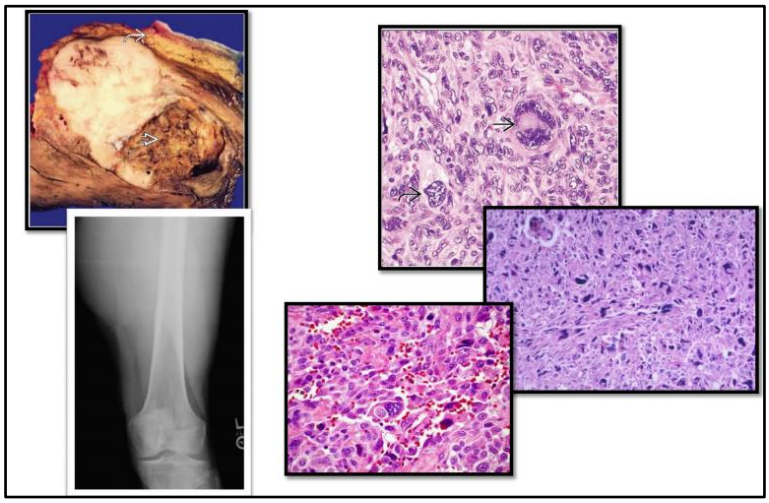




This is a keratin stain, which tests for keratin focal marks, usually (+) for carcinomas only but is also (+) for synovial sarcomas.

- **Undifferentiated Polymorphic Sarcoma** (UPS):
  - Are high grade mesenchymal sarcomas of **pleomorphic** (variable in shape) cells **that lack cell lineage**.
  - Mainly affect deep soft tissues and extremities.
  - Used to be called malignant fibrous histiocytoma (**MFH**)…not anymore.
  - Most tumors are aneuploid with complex genetic abnormalities.
  - **Rx: aggressive with surgery and adjuvant CT +/- RT; poor prognosis**.

- Morphology:
  - Large tumors
  - Anaplastic (no differentiation) and pleomorphic
  - Abnormal mitosis
  - Necrosis
  - ‘Ugly’ looking histological appearance.



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.

IMPORTANT NOTE: All soft tissue tumours are more common in benign form that in malignant, EXCEPT for skeletal muscle tumours (rhabdomyosarcoma is way more common that rhabdomyoma).