Soft Tissue Sarcomas

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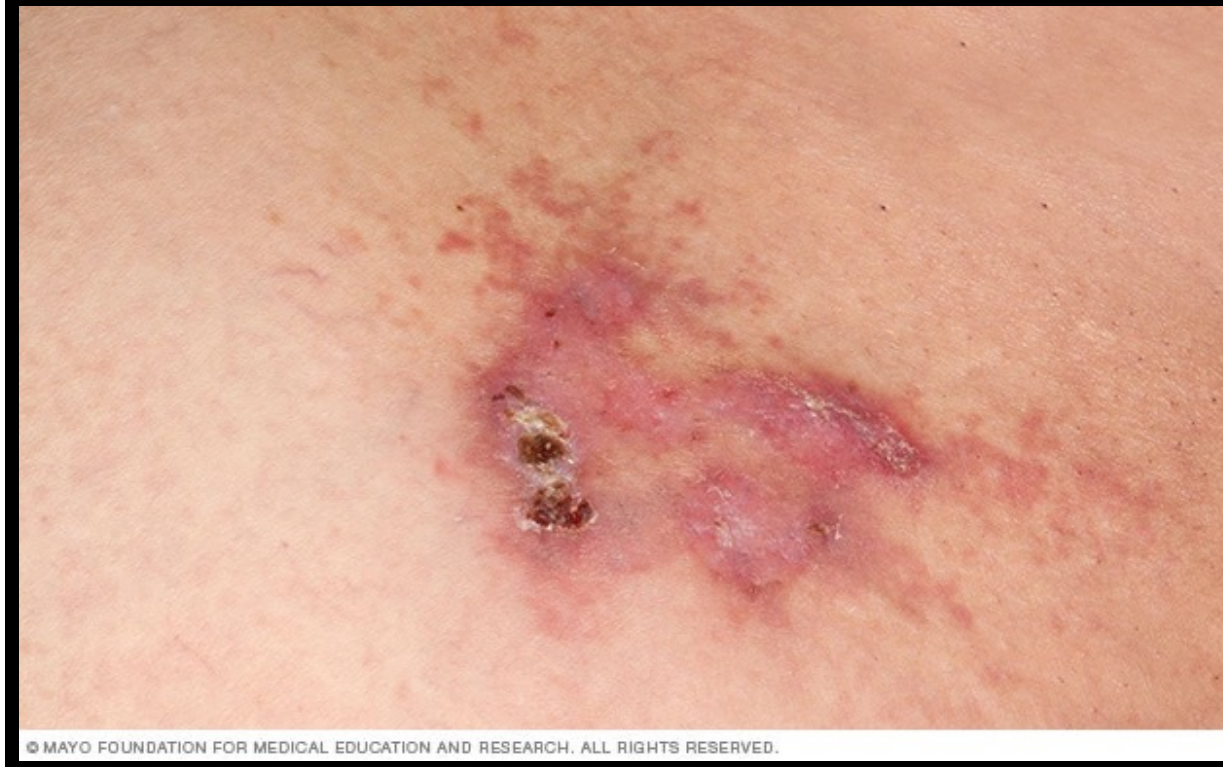
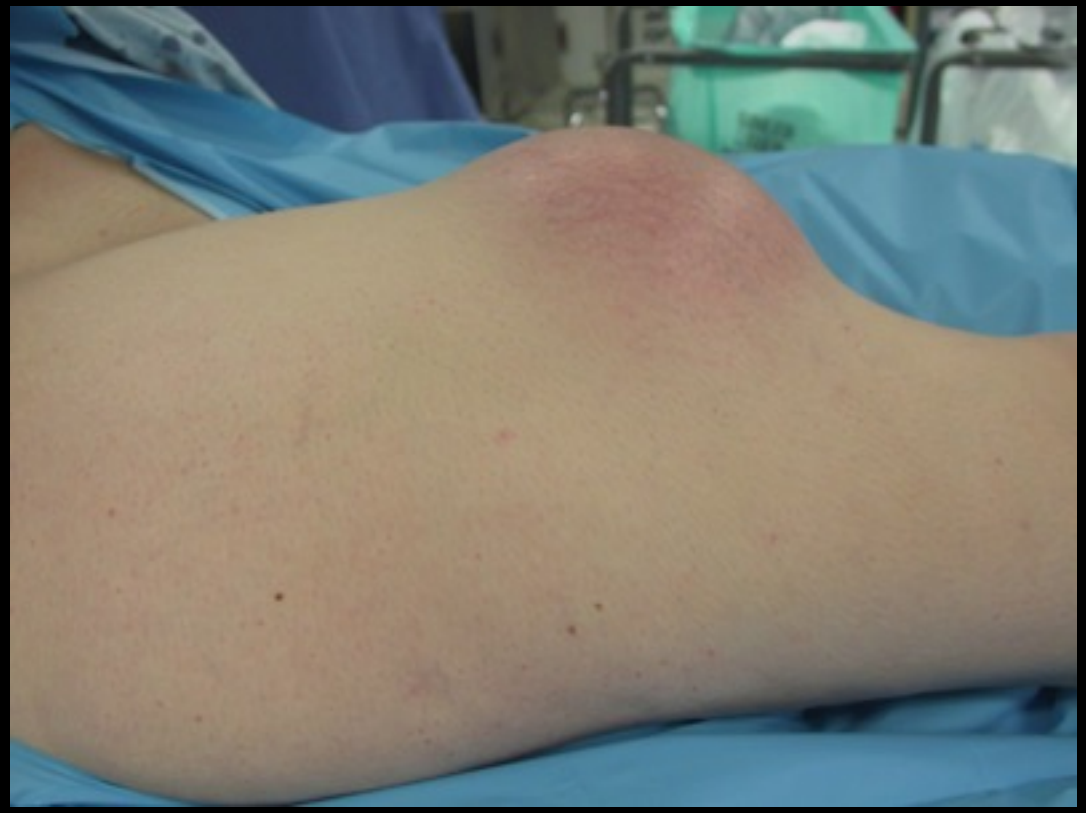


Presentation

- Enlarging painful/painless mass
- Most are large >5cm, deep to fascia, and firm (prognostic factors)

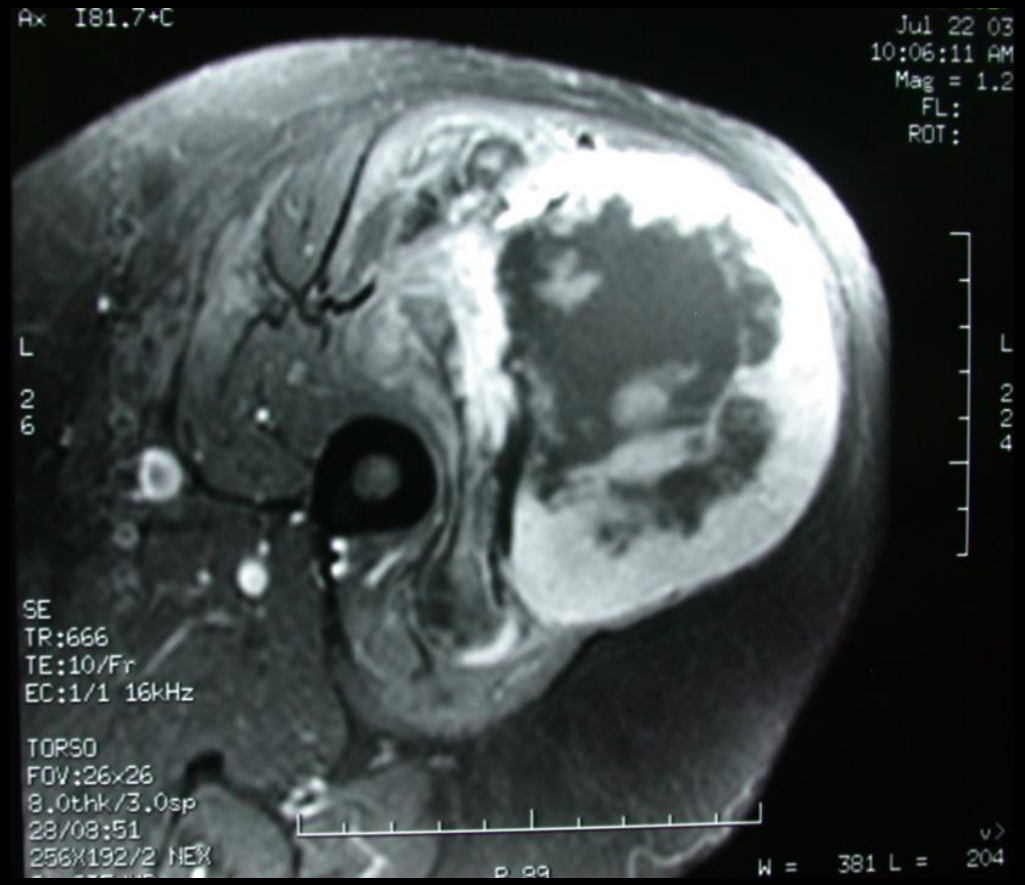


Predisposing factors

- NF-1: MPNST
- Gardners familial polyposis- low grade fibrosarcoma
- Digoxin (agent orange)- high grade sarcoma

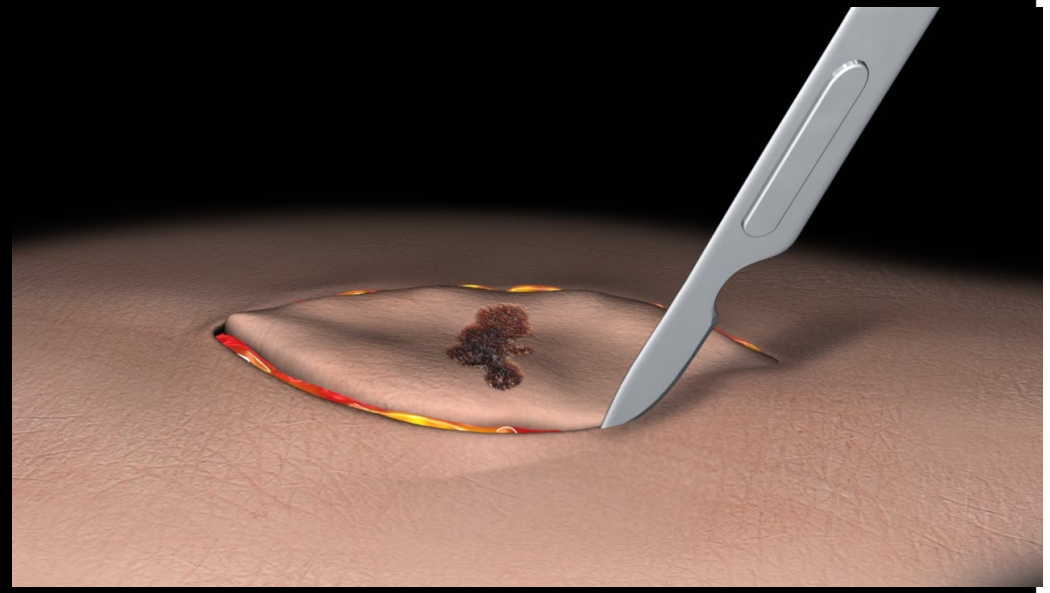
Imaging

- MRI is the best imaging modality to help define anatomy and to help characterize lesion
- T1 Dark, T2- Bright (except low grade liposarcoma)
- CT chest to eval for mets
- CT abdomen pelvis- w/ liposarcoma because synchronous retroperitoneal liposarcoma



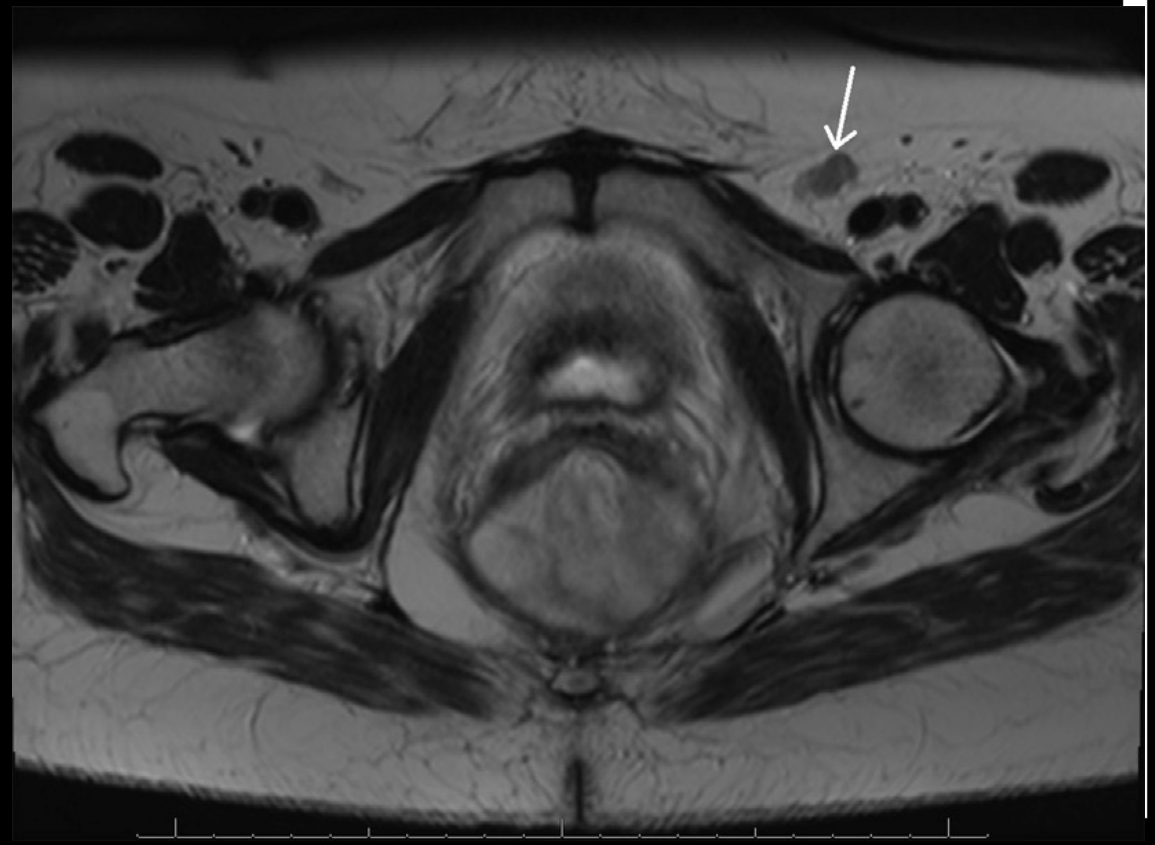
Diagnosis

- Must be systematic to avoid errors
- Excisional biopsy
- Unplanned removal of soft tissue sarcoma is the most common error
  - Repeat excision in this case to get residual tumor



Mets

- Most soft tissue sarcomas metastasis to the lung
- SCARE tumors- lymph node mets
  - Synovial sarcoma, clear cell sarcoma, angiosarcoma, rhabdomyosarcoma, epithelioid sarcoma



Treatment

- Typically wide resection
- + Radiation therapy (>5cm)
- Adjuvant radiation
  - Pre op 50Gy (wound healing complications)
  - Post-op 60 Gy (fibrosis/ fracture)

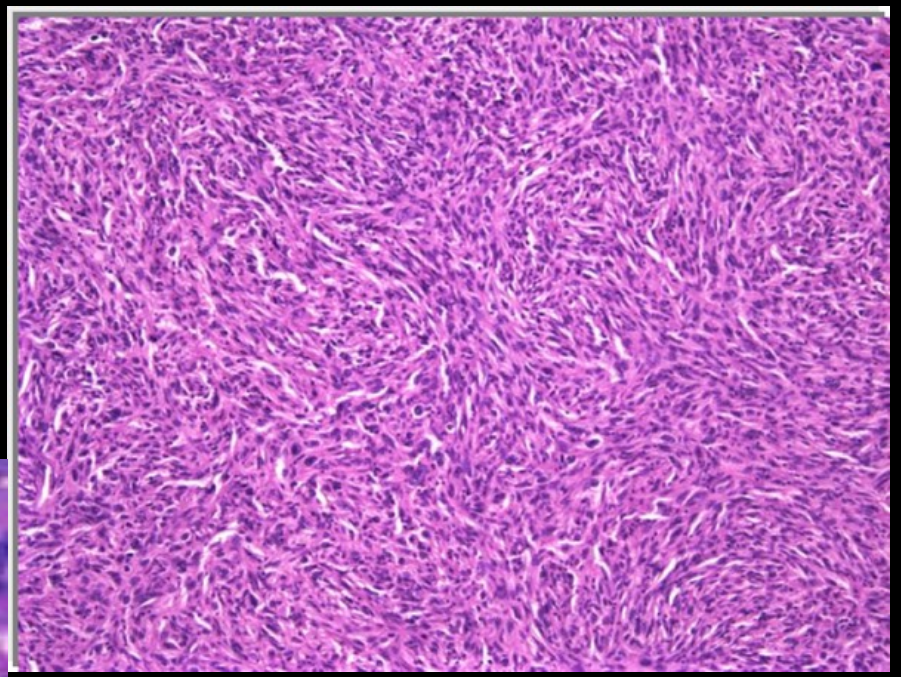
Fibrous Tumors

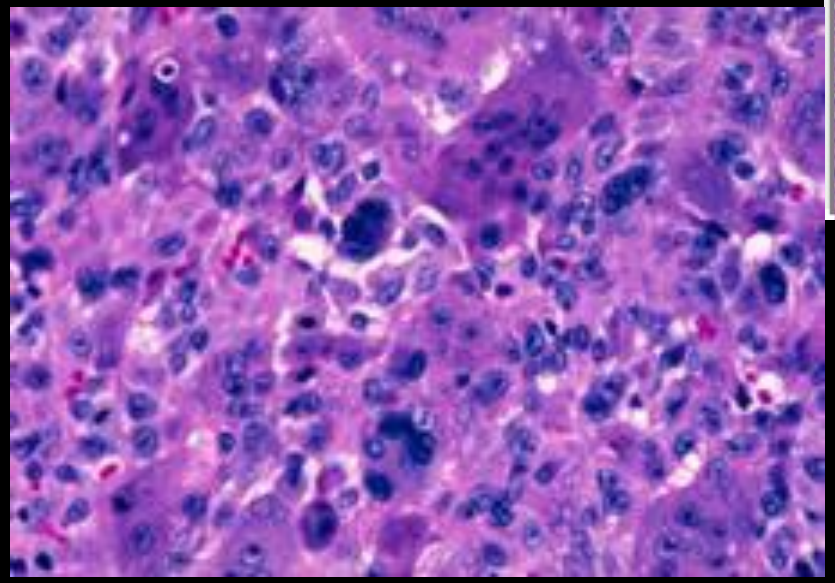
- Benign- (calcifying aponeurotic fibroma, fibromatosis, extrabdumonal desmoid tumor, nodular fasciitis)
- Malignant- Fibrosarcoma + NFH

- Typically age 30-80, manifests as enlarging, generally painless mass
- Imaging: deep seated inhomogeneous mass

Undifferentiated pleomorphic sarcoma (MFH)

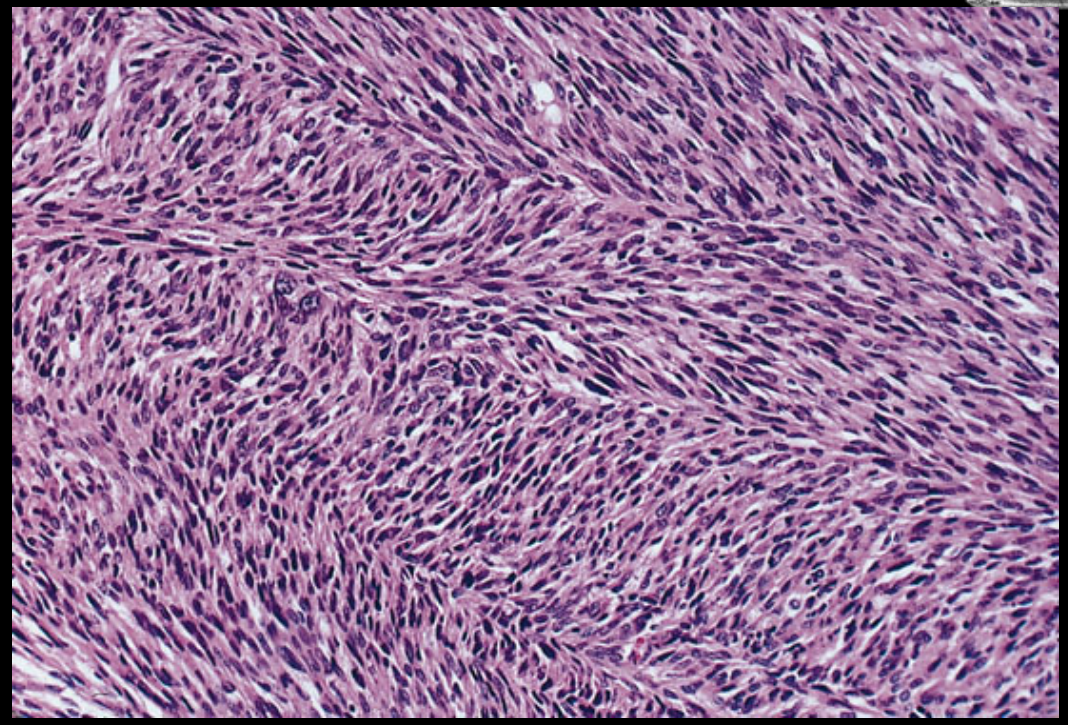
- Spindle + histiocytic cells in a storiform (cartwheel) pattern.
- Chronic inflammatory cells may be present





Fibrosarcoma

- Fasciculated growth pattern w/ spindle-shaped cells, scant cytoplasm.
- Herringbone pattern w/ intersecting fascicles in which nuclei in one fascicle is viewed transversely but adjacent fascicle viewed longitudinal

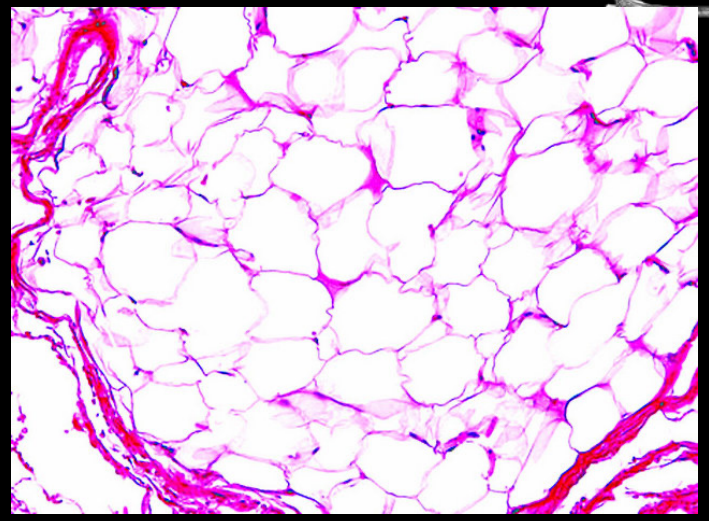


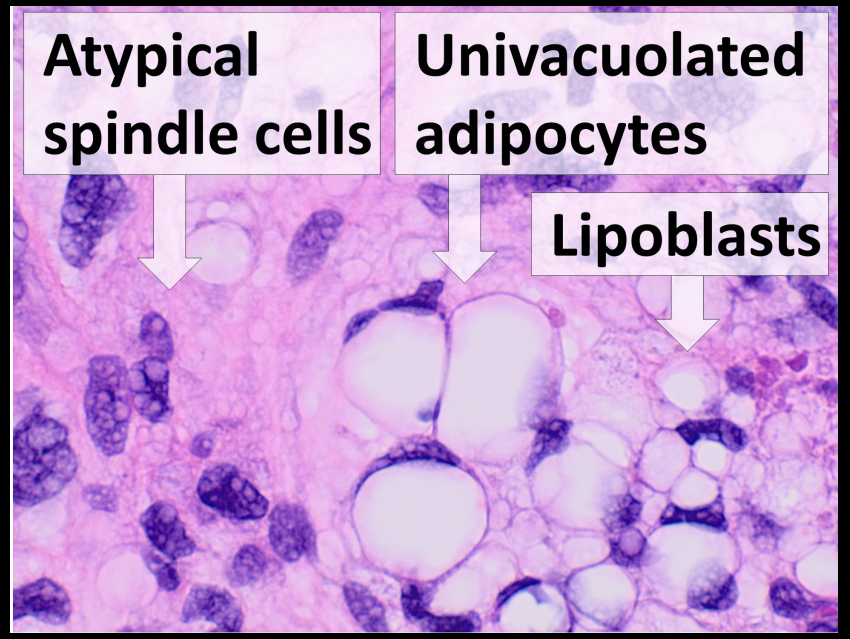
UPS + Fibrosarcoma tx

- Tx for both: wide margin local excision. If tumor >5cm, +/- radiation

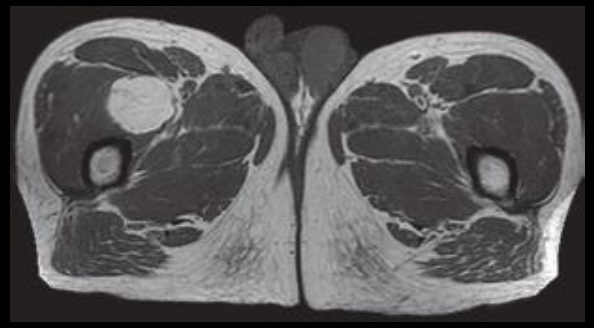
Fatty Tissue Tumors

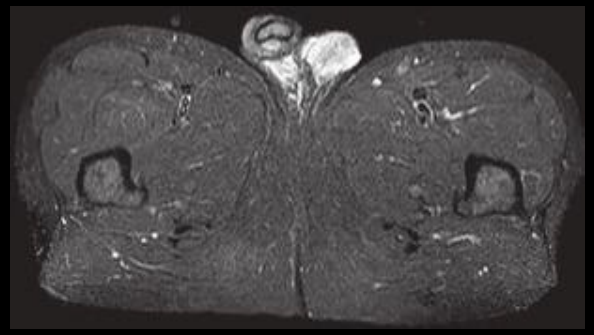
- Benign: (lipomas- spindle cell lipoma, pleomorphic lipoma, angiolipoma)
- Malignant- Liposarcomas
  - heterogenous group of tumors , have in common the presence of lipoblasts in the tissue
  - Virtually never in subcutaneous tissues
  - Metastaze according to grade of lesion
  - MDM2+

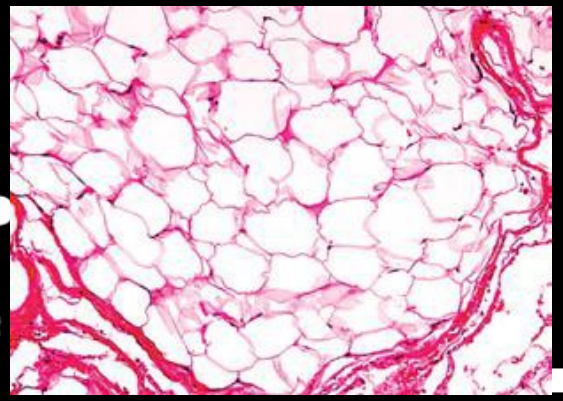


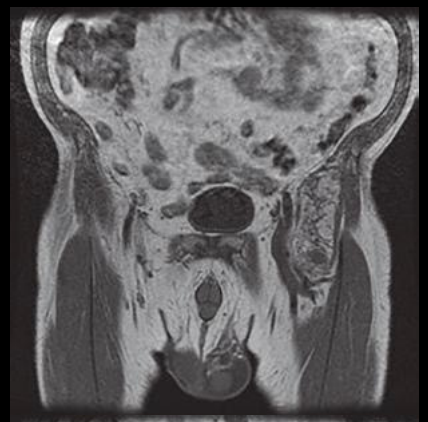


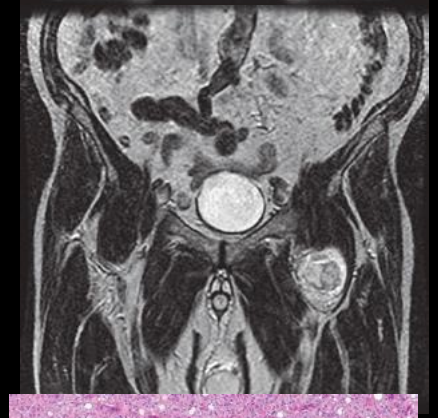
Lipoma v Liposarcoma v Dedifferentiated Liposarcoma

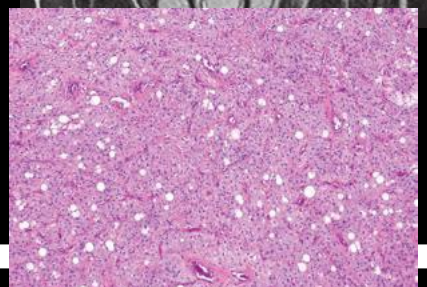


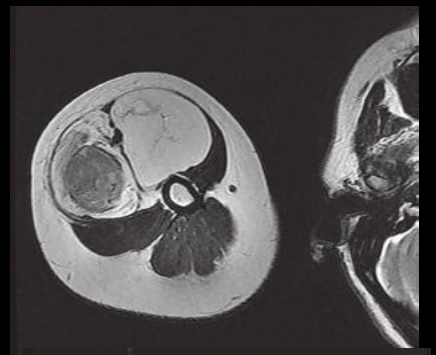


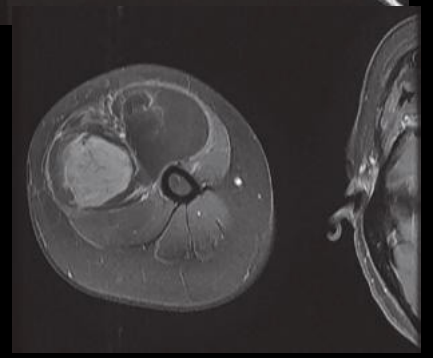


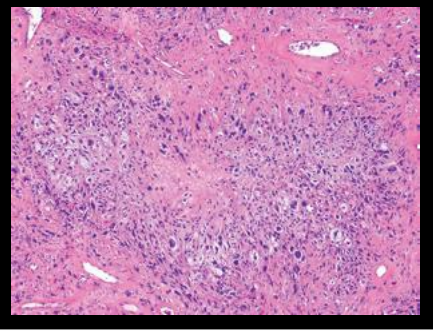






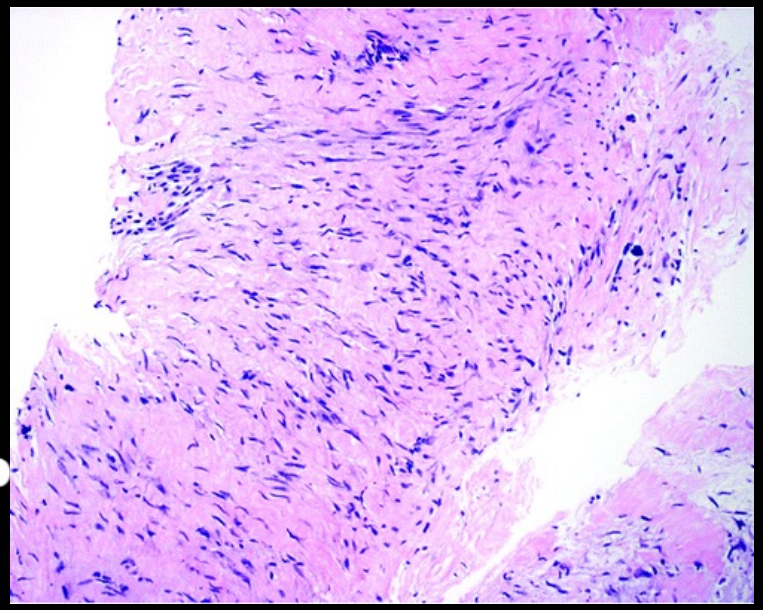


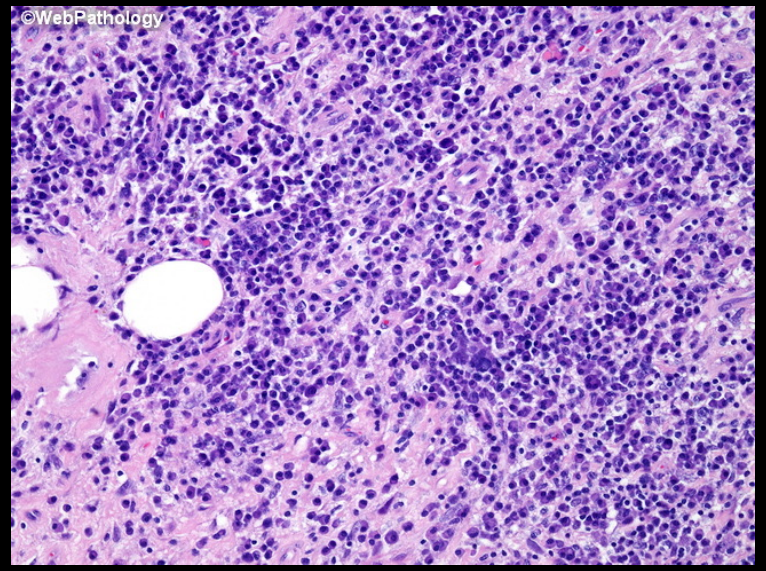




Liposarcoma—low grade

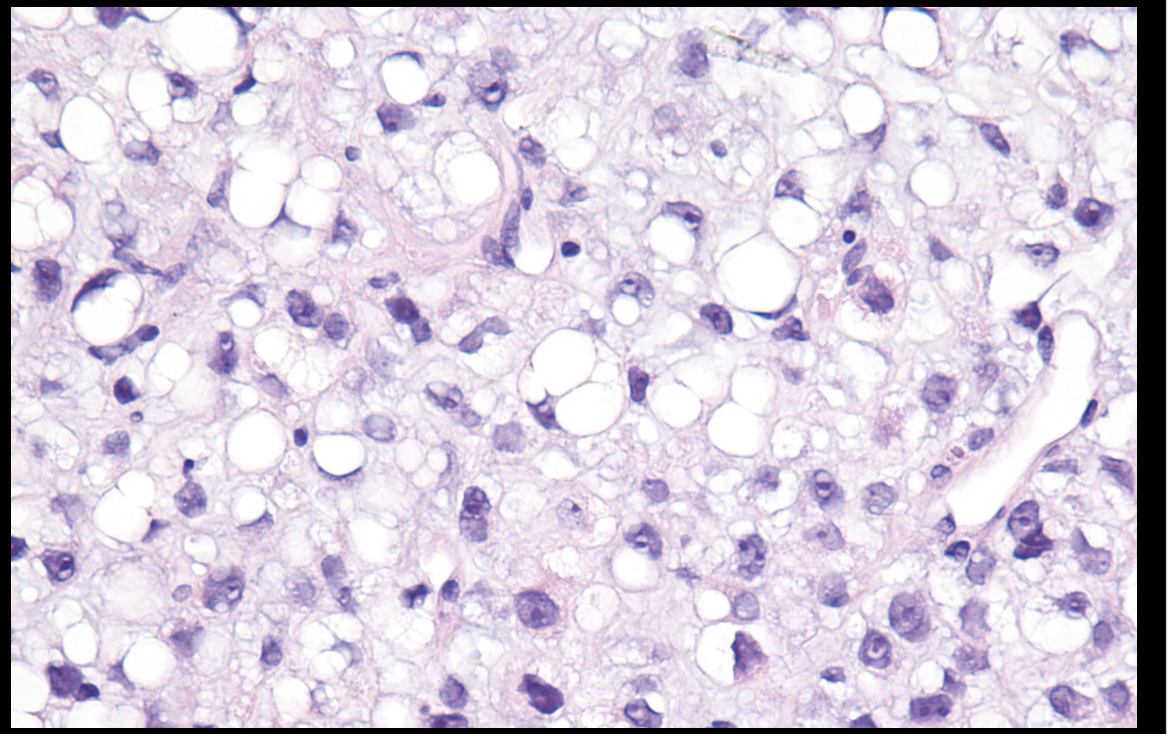
- Low grade (mets <10%)
  - Well differentiated liposarcoma
    - Lipoma like
    - Sclerosing
    - Inflammatory





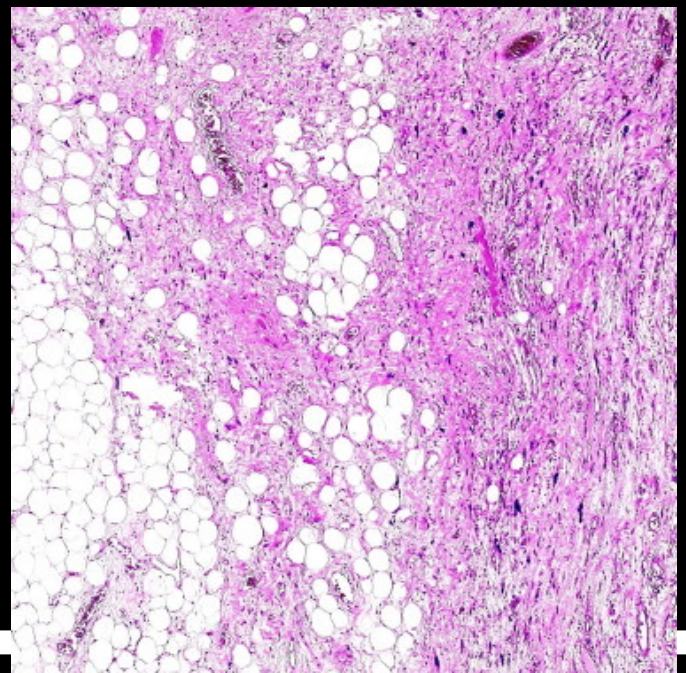
Liposarcoma- Intermediate

- Intermediate grade (mets 10-30%)
  - Myxoid liposarcoma(12:16)

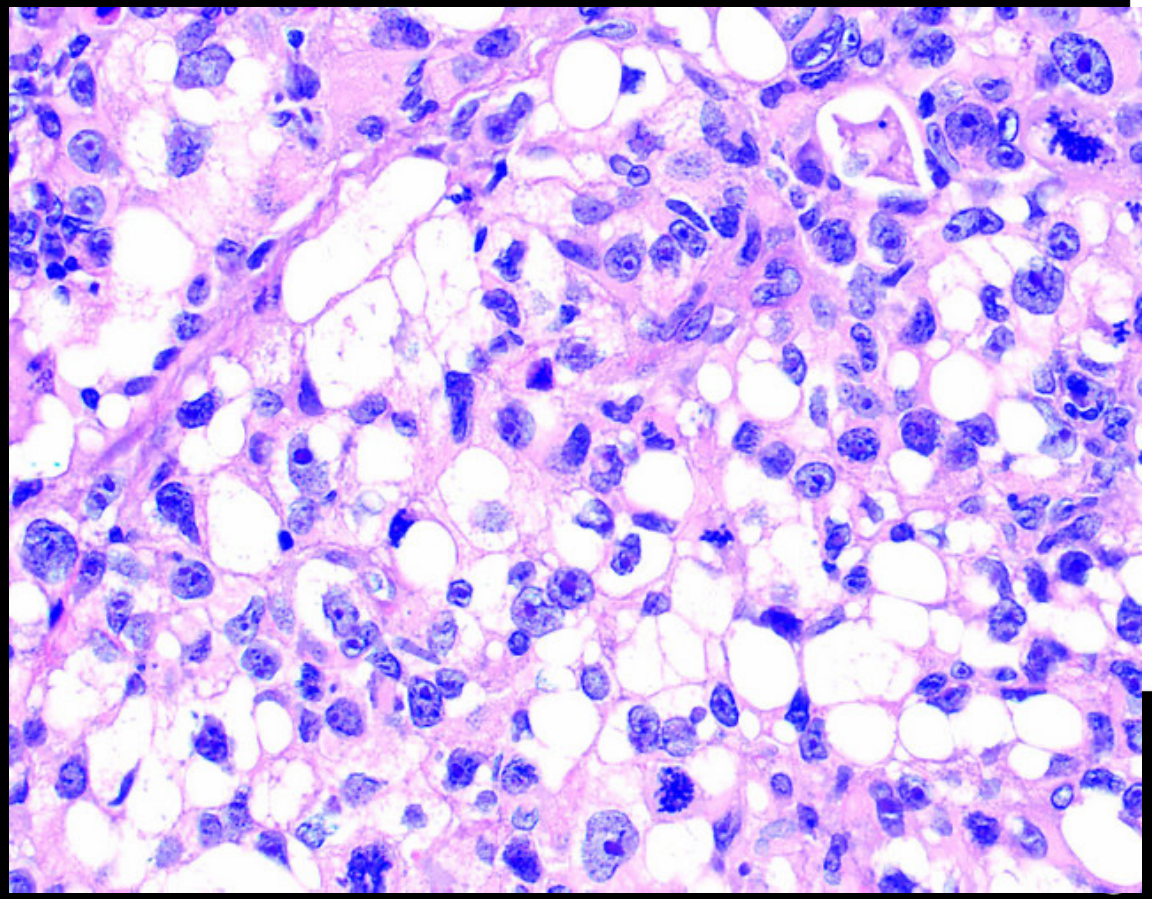


Liposarcoma- High grade

- High grade (mets >50%)
  - Round cell liposarcoma
  - Dedifferentiated
    - On histo: areas w/ fat and other w/ soft tissue sarcoma

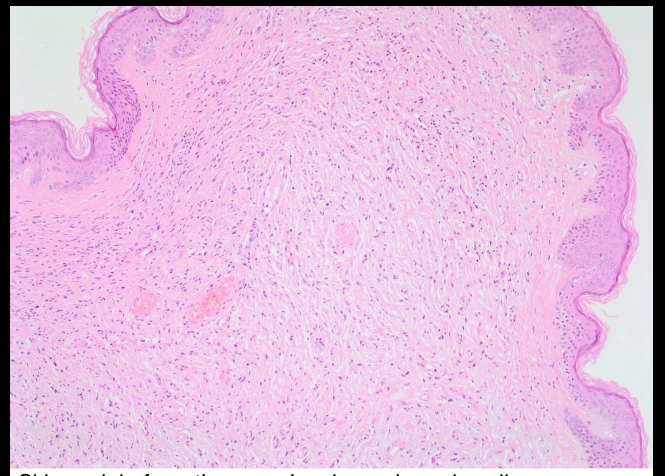


- Pleomorphic liposarcoma

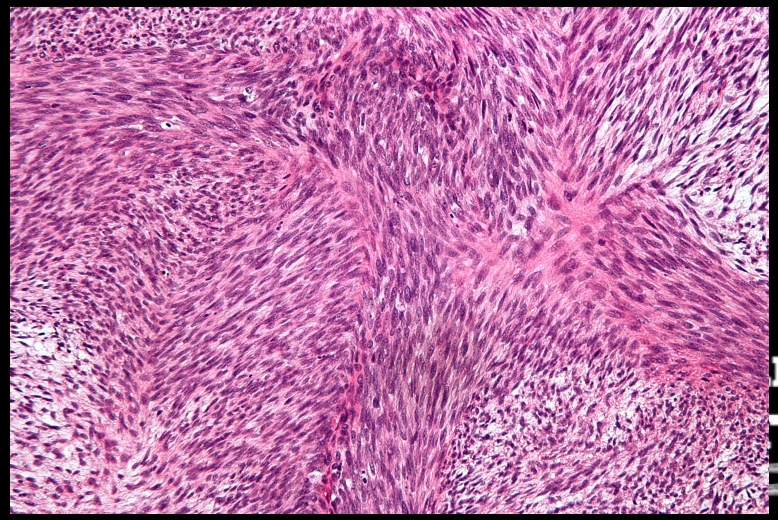


Neural Tissue Tumors

- Benign (neurilemoma + neurofibroma)
- Malignant- neurofibrosarcoma/ malignant peripheral nerve sheath tumor
  - May arise de novo or in setting of neurofibromatosis
  - Spindle cells in whirling pattern
  - S100 positive
- Presentation?

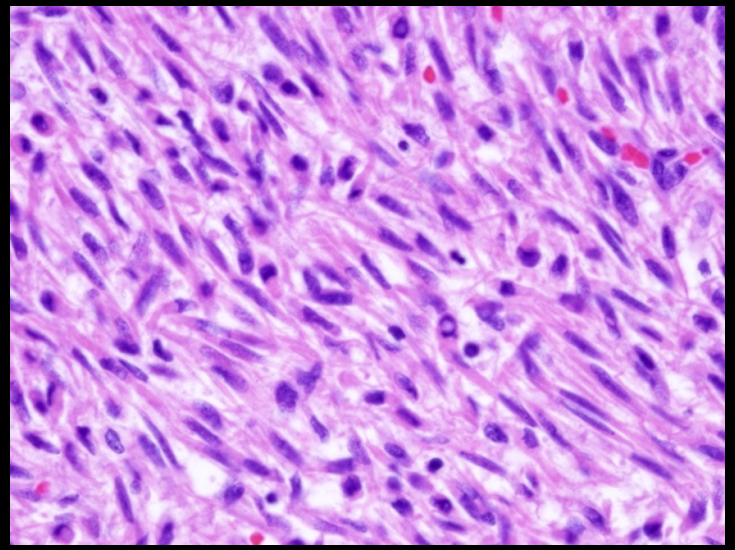


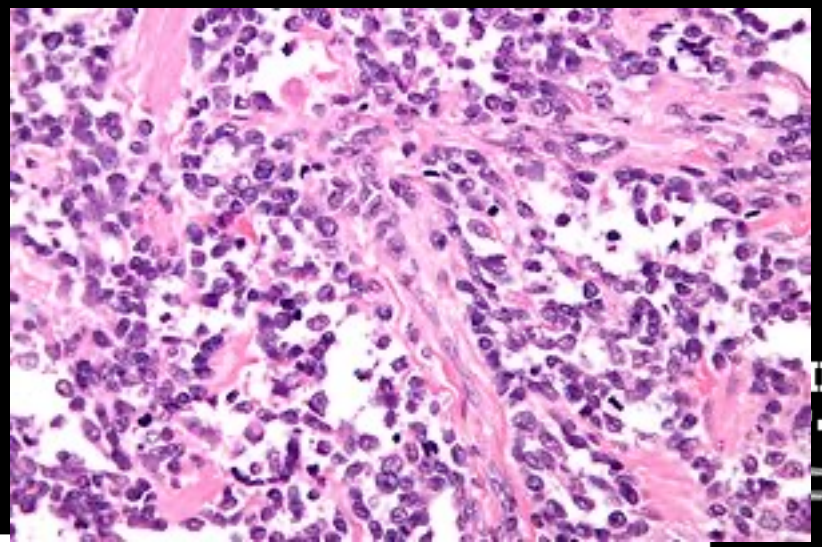
Skin nodule from the arm showing a dermal, well circumscribed non encapsulated proliferation of hypocellular spindle cells.



Muscle Tissue Tumors

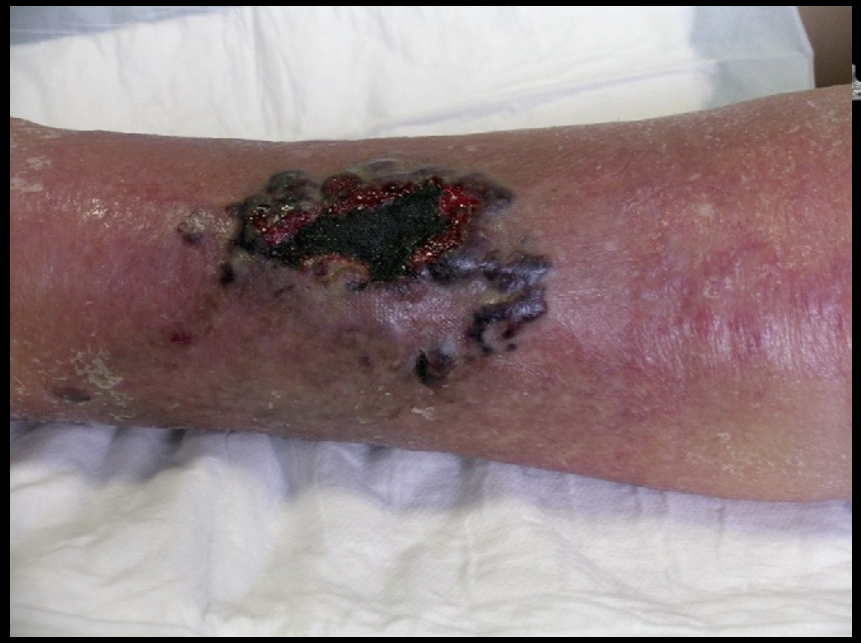
- Leiomyosarcoma
  - Spindle cells intersect at 90 degrees
- Rhabdomyosarcoma
  - MC sarcoma in young patients
  - Spindle cells in parallel bundles, multinucleated giant cells, racquet shaped cells
  - Sensitive to chemo + wide margin resection. + external beam irradiation
  - t(2;13), PAX-FKHR gen

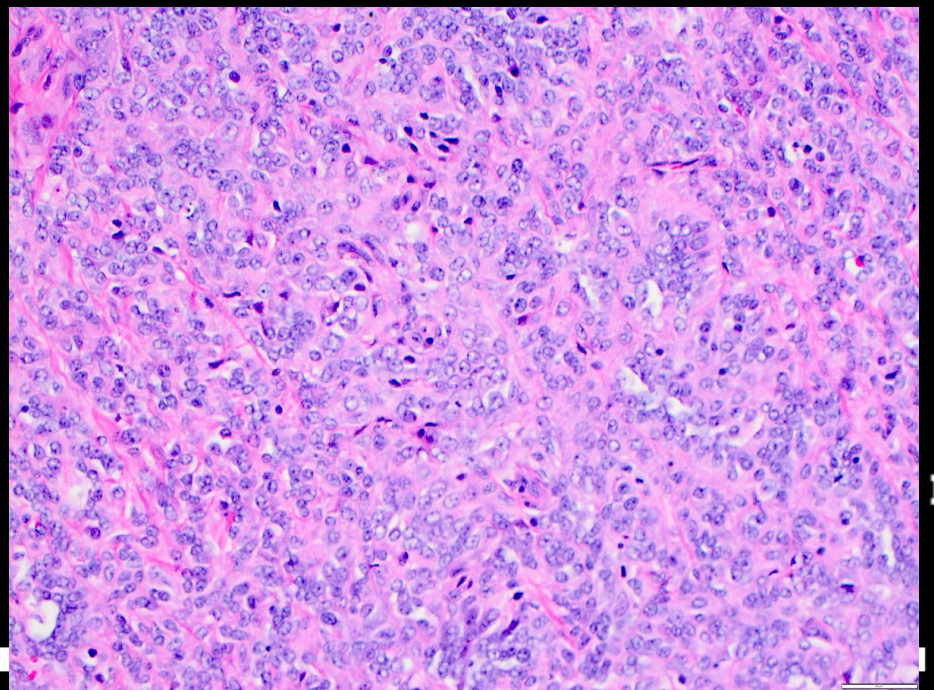




Vascular Tumors

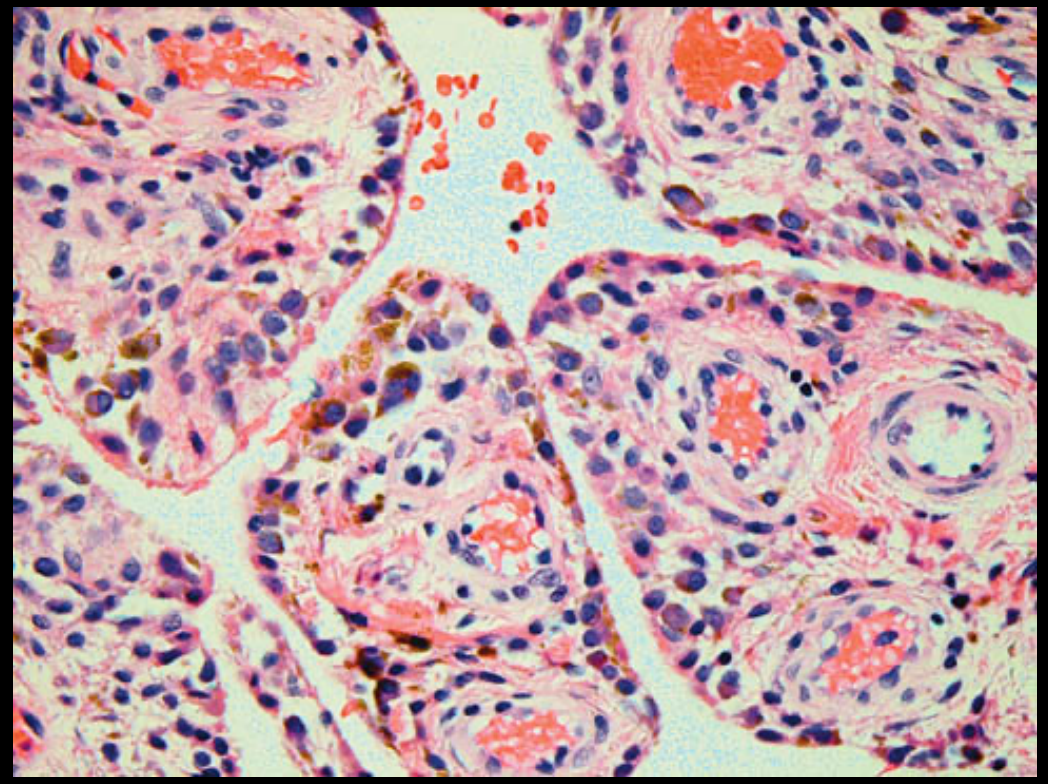
- Benign (hemangioma)
- Angiosarcoma
  - In the setting of chronic vascular stasis, trauma, radiation, lymphedema. Breast ca, radiation
  - Cells resemble blood vessel endothelium
  - Lymph node + pulmonary mets common
  - Cutaneous + Highly malignant
  - TX: wide excision and radiation





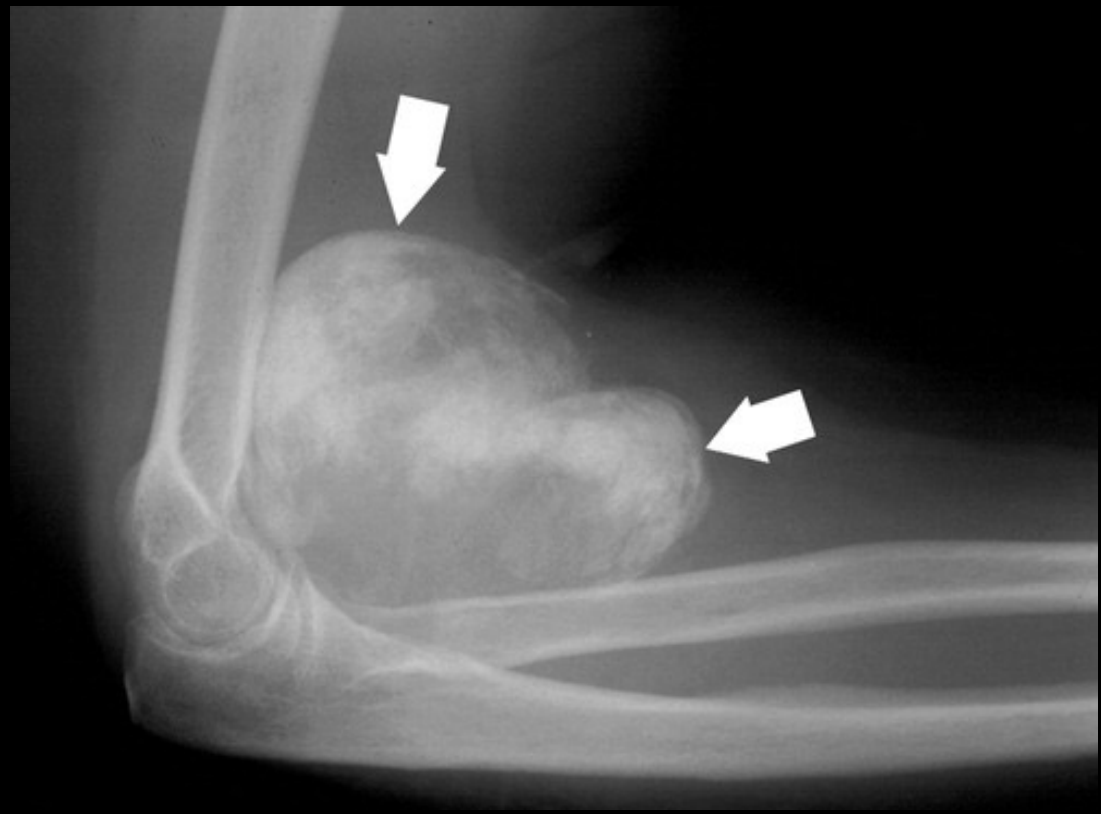
Synovial Disorders

- Benign (ganglia, PVNS, Giant cell tumor of tendon sheath, synovial chondromatosis)
- Synovial sarcoma



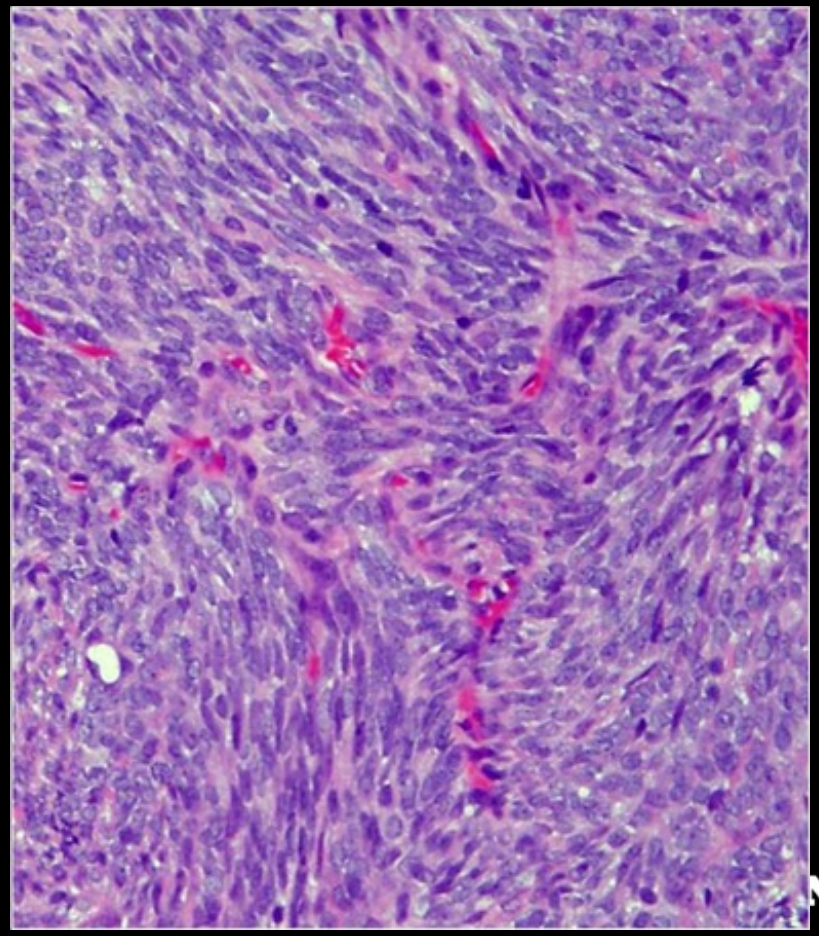
Synovial Sarcoma

- Ages 15-40
- Malignant high grade tumor, lymph mets common
- Most common sarcoma in the foot
- Xrays/CT- may show mineralization within lesion in up to 25% of cases
- Translocation X:18> results in gene fusion products, SYT-SSX1 and SYT-SSX2



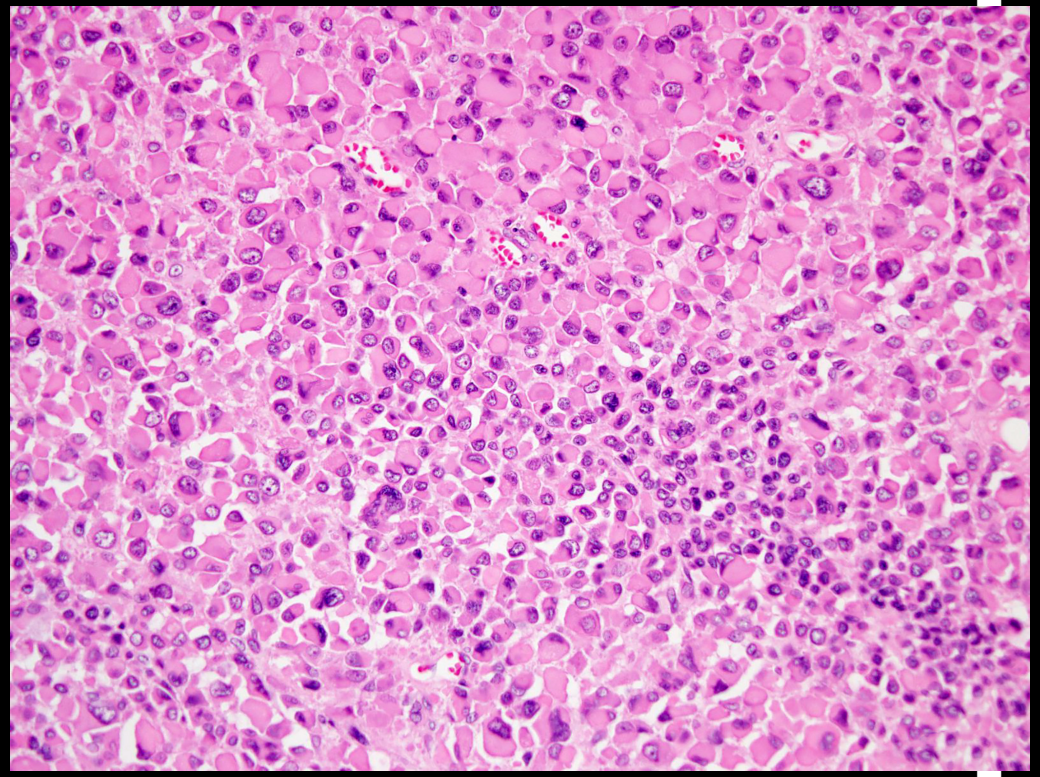
Synovial Sarcoma

- Histo: often biphasic w/ both spindle cells and epithelial cells.
  - Epithelioid cells are ovoid and can form lumina or nests containing mucin
  - Spindle cell components run in fascicles and have small nuclei



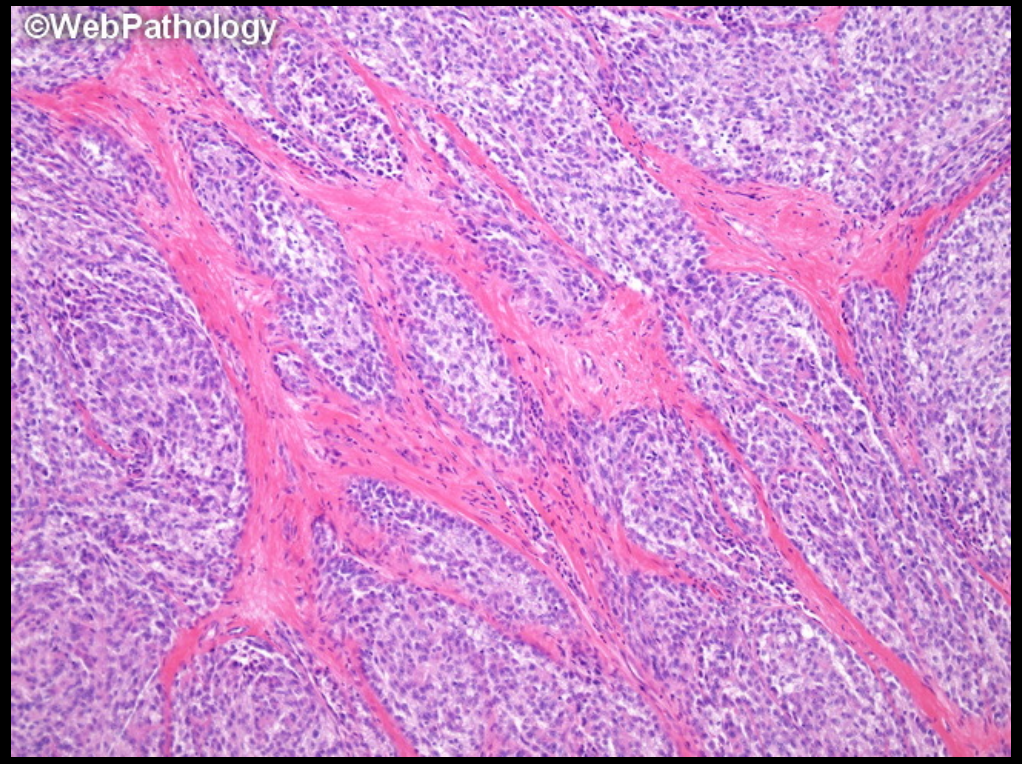
Epithelioid sarcoma

- Rare nodular tumor common in upper extremities of young adults
- Most common sarcoma of the hand
- Cell shape range from ovoid to polygonal w/ deeply eosinophilic cytoplasm
- Tx: wide margin surgical resection



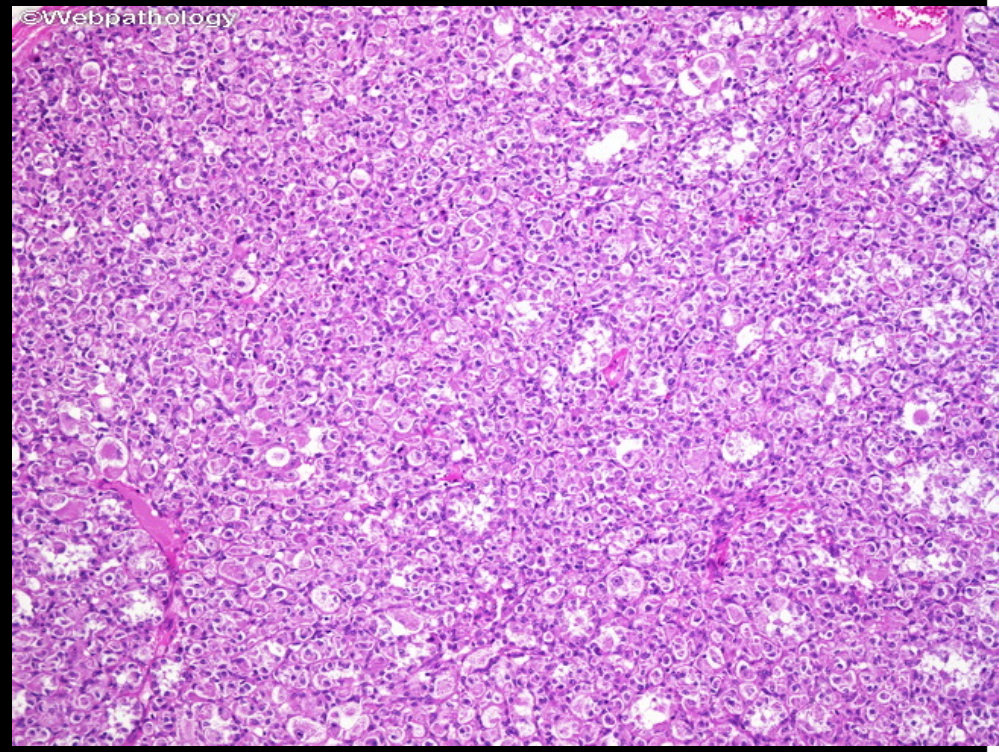
Clear cell sarcoma

- Slow growing mass in association w/ tendons or aponeuroses
- Compact nests or fascicles of rounded or fusiform cells w/ clear cytoplasm. Giant cells
- Tx: wide margin resection w/ adjuvant irradiation



Alveolar cell sarcoma

- Slow growing painless mass in young adult
- Common in anterior thigh
- Histo: dense fibrous trabeculae dividing tumor into organoid or nestlike arrangement
- Tx: wide margin resection a/ adjuvant irradiation in select cases



Thank you Dr. Balach!

- Sources: Millers review of Orthopaedics