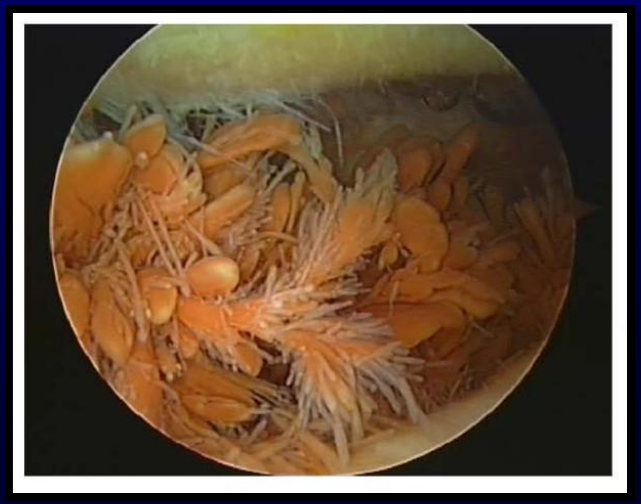
# Lecture 9

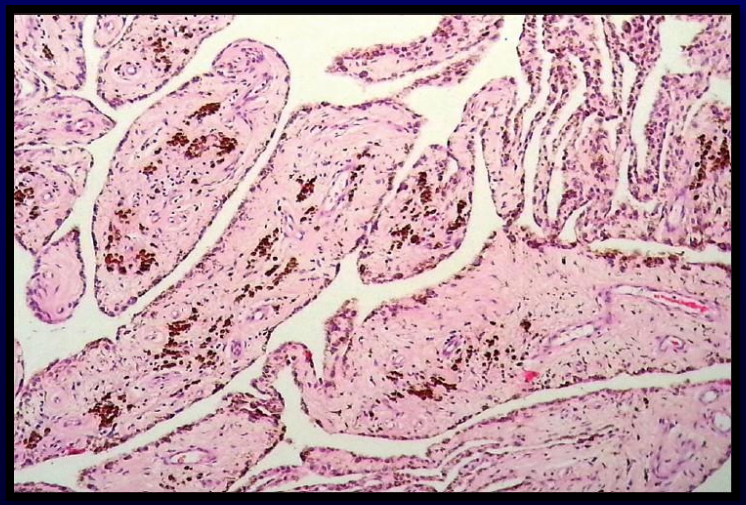
# JOINT TUMORS & TUMORLIKE CONDITIONS:

- Joint tumors are rare
- Ganglion cyst and tenosynovial giant cell tumor are the most frequent
- Ganglion cyst: common condition; close to a joint, dorsum of wrist; not true cyst, no communication with synovial joint; may cause pressure pain; treated by surgical removal
- True synovial cyst (Baker cyst around the knee): herniation process

# TENOSYNOVIAL GIANT CELL TUMOR:

- Benign neoplasm of synovium
- Diffuse (pigmented villonodular synovitis, PVNS, large joints) or localized small hands tendons
- T(1;2)(p13q;37); affecting type IV collagen α-3



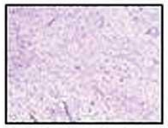
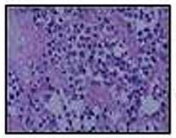
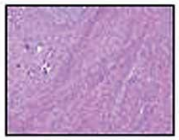
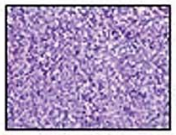


# SOFT TISSUE TUMORS:

- **Benign >>>>>>malignant**
- **Incidence: 1% and cause 2% cancer death**
- **Sarcomas are aggressive and metastasize mainly to lungs, hematogenous spread**
- **Most are in extremities (thigh)**
- **Most are sporadic; very few arise from tumor suppressor gene mutations (NF1, Gardner syndrome, Li-Fraumeni syndrome, Osler-Webber-Rendu Syndrome)**
- **Few occur after exposure to radiation, burns & toxins.**

# SOFT TISSUE TUMORS:

- No precursor lesions; theory that they arise from pluripotent mesenchymal stem cell which acquire somatic mutation
- 15-20% simple karyotype, single signature mutation (Ewing and synovial sarcoma)
- 80-85% complex karyotype (genomic instability), LMS and pleomor. Sarcoma
- Wide range (benign-highly malignant)
- Diagnosis, grade and stage are all important

| DIFFERENTATION | Subtypes | Chromosomal traslocations | Fusion trascripts |
|---|---|---|---|
|  ADIPOCYTIC TUMORS | *Lipoblastoma*: | t(7;8)(q31;q13); t(8;8)(q24;q13) | PLAG1-COL1A2;PLAG1-HAS2 |
| | *Myxoid liposarcoma* | t(12;16)(q13;p11); t(12;22)(q13;q12) | CHOP-TLS; CHOP-EWS |
|  FRIBLOBLASTIC/ MYOFIBROBL.TUMORS | *Inflammatory myofibroblastic tumor* | t(1;2)(q25;p23); t(2;19)(p23;q13); t(2;17)(p23;q23) | TPM3-ALK; ALK-TPM4; ALK-CLTC |
| | *Infantile fibrosarcoma* | t(12;15)(p13;q25) | ETV6-NTRK3 |
| | *Dermatofibrosarcoma protuberans/ Giant cell fibroblastoma* | t(17;22)(q22;q13) | COL1A1-PDGFB |
|  SKELETAL MUSCLE TUMORS | *Alveolar rhabdomyosarcoma* | t(2;13)(q35;q14); t(1;13)(p36;q14) | PAX3-FKHR; PAX7-FKHR |
|  TUMORS OF UNCERTAIN DIFFERENTIATION | *Angiomatoid fibrous histiocytoma* | t(12;22)(q13;q12); t(12;16)(q13;p11) | |
| | *Synovial sarcoma* | t(X;18)(p11.2;q11.2) | SYT-SSX1/2/4 |
| | *Alveolar soft part sarcoma* | t(X;17)(p11;q25) | TFE3/ASPL |
| | *Clear cell sarcoma* | t(12;22)(q13;q12) | EWS-ATF1 |
| | *Extraskeletal myxoid chrondrosarcoma* | t(9;22)(q22;q12); t(9;15)(q22;q21) | EWS-TEC; CHN-TFC12 |
| | *Desmoplastic small round cell tumor* | t(11;22)(p13;q12) | EWS-WT1 |
|  EWING SARCOMA | | t(11;22)(q24;q12);t(21;22)(q22;q12); t(17;22)(q12;q12); t(7;22)(p22;q12); | FLI1-EWS; ERG-EWS E1AF-EWS; ETV1-EWS |

# ADIPOSE TISSUE TUMORS:

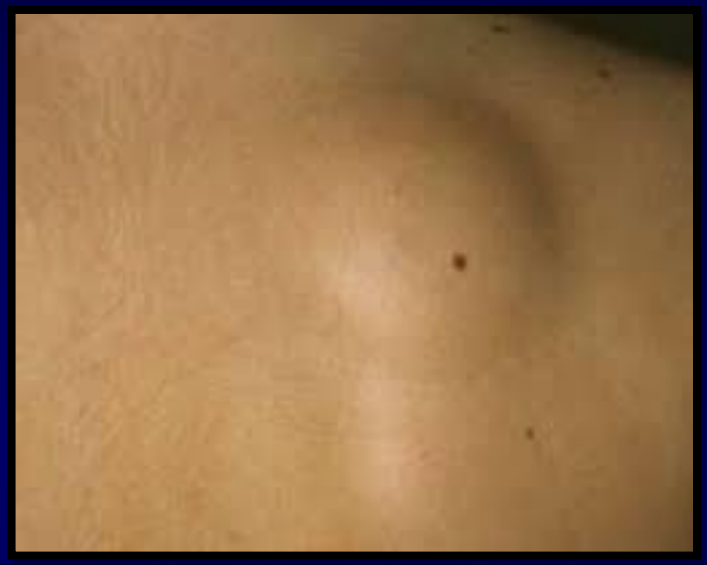
## LIPOMA

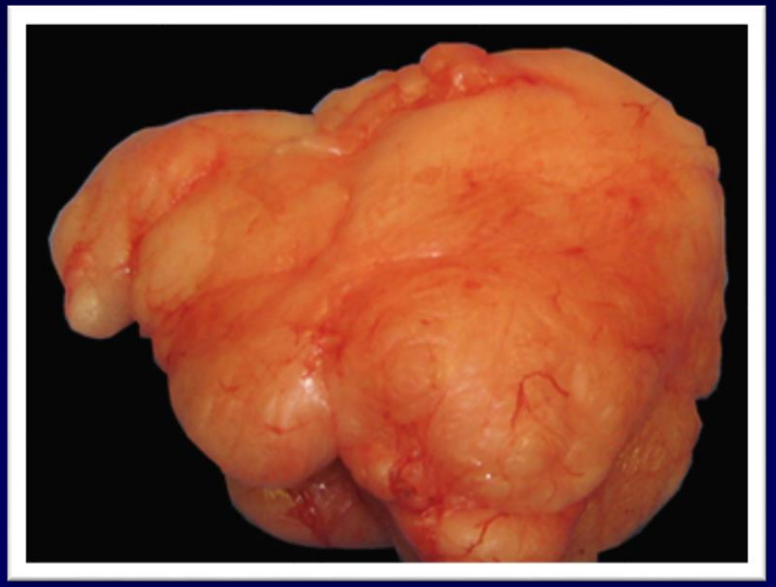
- Most common soft T tumor
- Well-encapsulated, subcutis
- Mature fat cells
- Trx: excision

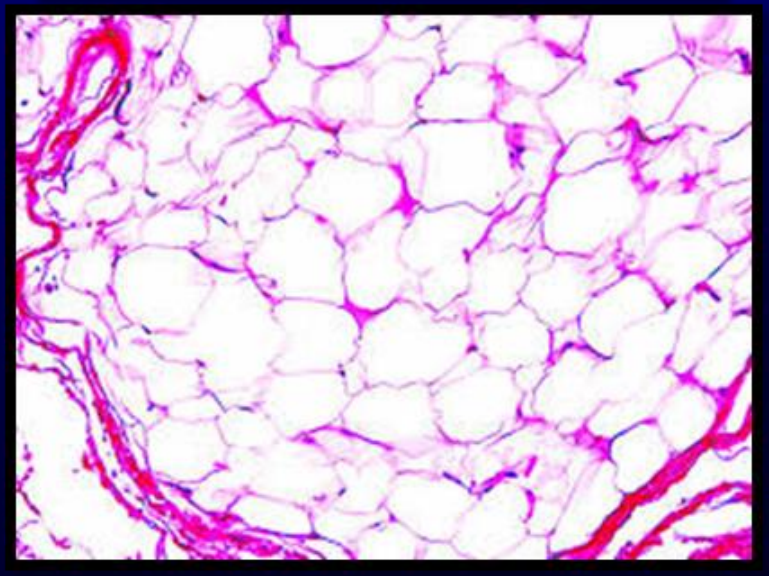
## LIPOSARCOMA

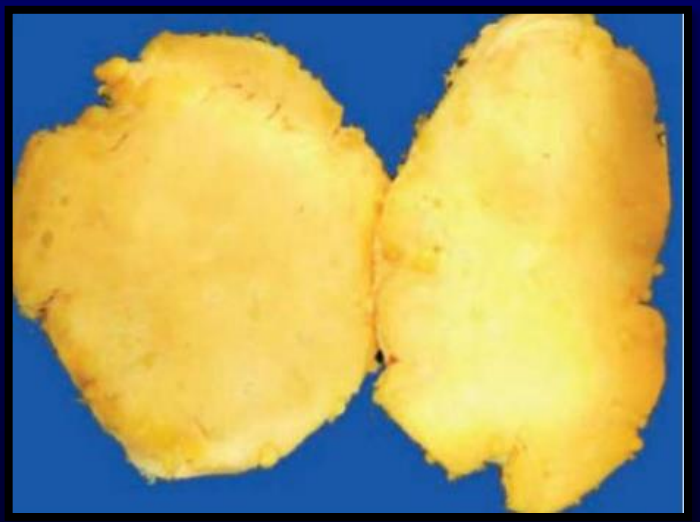
- Most common sarcomas in adults. >50 years
- Extremities and retroperitoneum
- 3 types:
  - WD (MDM2 gene chr 12)
  - Myxoid, t(12,16)
  - Pleomorphic (aggressive)

# LIPOMA PATHOLOGIC FEATURES:

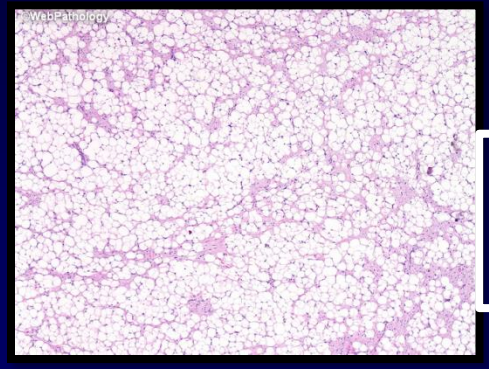




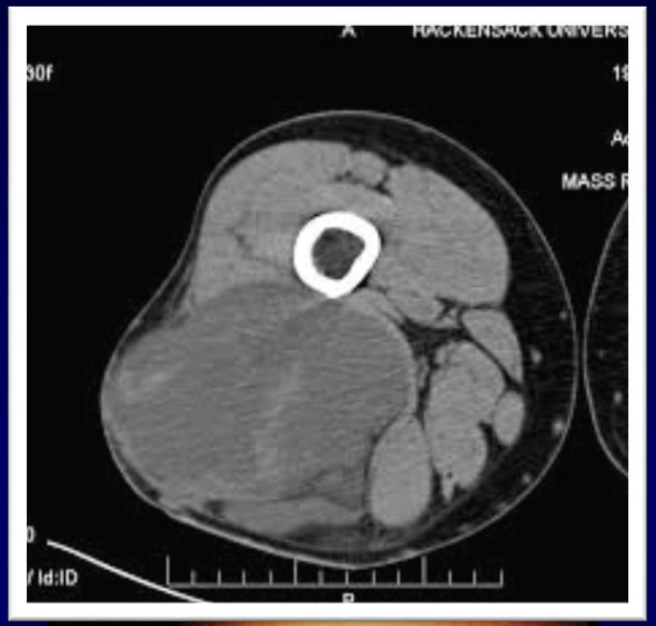


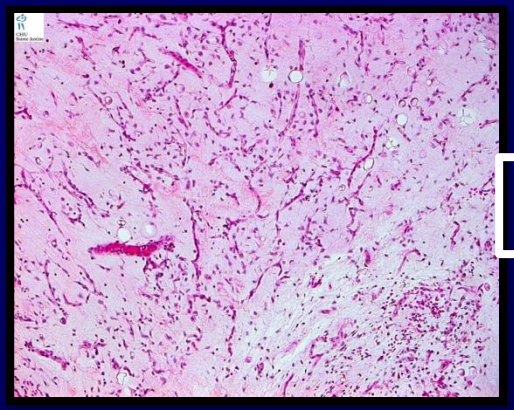


# LIPOSARCOMA FEATURES:

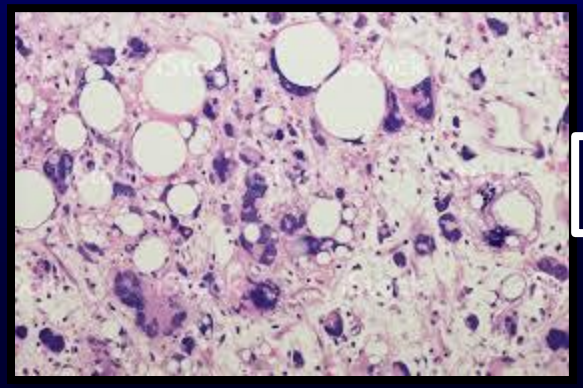



**Well-differentiated**

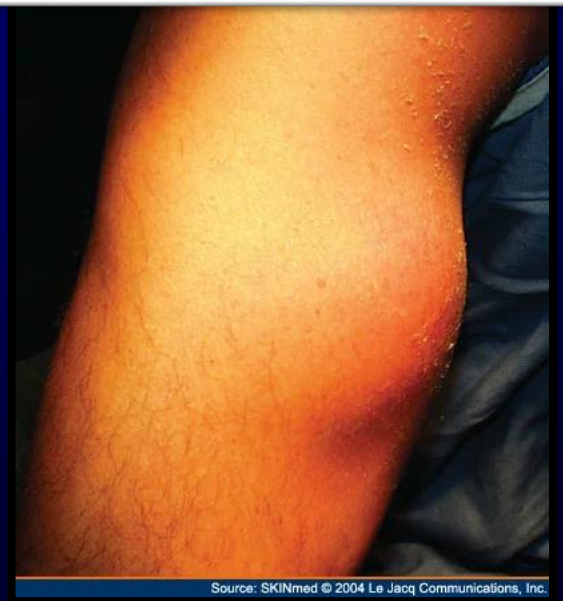




**Myxoid**



**Pleomorphic**



# FIBROUS TUMORS:

- Nodular fasciitis
- Fibromas and Fibrosarcoma
- Fibromatoses:
  - Superficial
  - Deep (Desmoid tumor)

# NODULAR FASCIITIS:

- **Nodular fasciitis: thought to be reactive process**
- **Now, clonal, t(17;22) producing *MYH9-USP6* fusion gene**
- **Trauma history, recent rapid size increase**
- **Maybe self-limiting**
- **IMPORTANT: not to diagnose it malignant**
- **Culture-like histology**

# NODULAR FASCIITIS: