## Joint tumor and tumor-like conditions:

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| -Joint tumors are rare<br>-Ganglion cyst and tenosynovial giant cell tumor are the most frequent | **Ganglion cyst**<br>-common condition<br>-close to a joint, dorsum of wrist<br>-not true cyst, no communication with synovial joint<br>-may cause pressure pain<br>-treated by surgical removal<br>**True synovial cyst**<br>(Baker cyst around the knee): herniation process | **tenosynovial giant cell tumor**<br>-Benign neoplasm of synovium<br>-Diffuse (pigmented villonodular synovitis, PVNS, large joints) or localized small hands tendons<br>-T (1;2); affecting type IV collagen α3 |

## 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.
-**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 pleomorphic Sarcoma
-Wide range (benign-highly malignant)
-Diagnosis, **grade** and stage are all important

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| **Adipose tissue tumors** | **Lipoma** | Most common soft T tumor<br>Well-encapsulated, subcutis<br>Mature fat cells<br>Treatment: excision |
| | **Liposarcoma** | Most common sarcomas in adults. >50 years<br>Extremities and retroperitoneum<br>3 types: WD, Myxoid, Pleomorphic (aggressive) |
| **Fibrous tumors** | **Nodular fasciitis** | thought to be reactive process<br>Now, clonal, t (17;22) producing MYH9- USP6 fusion gene<br>Trauma history, recent rapid size increase<br>Maybe self-limiting<br>***IMPORTANT: not to diagnose it malignant***<br>Culture-like histology |
| | **Fibromas** | benign proliferation of fibroblasts, ***very common***, skin and subcutaneous tissue |
| | **Fibrosarcoma** | malignant counterpart<br>usually superficial cutaneous tumors of fibroblasts, cellular, ***storiform pattern*** with increased mitosis |

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| | **Superficial fibromatoses** | -Infiltrative ***benign*** fibroblastic proliferation<br>-May run in families; **may impact function**<br>-Palmar (Dupuytren contracture) -> Palmar fascia<br>-Planter fibromatosis -> sole of foot<br>-Penile (Pyeronie disease) |
| | **Deep (Desmoid tumor) fibromatoses** | -Deep infiltrative but bland fibroblastic proliferation; ***doesn't metastasize*** but recur<br>-20-30years, females more common<br>-Abdominal wall, mesentery and limbs<br>-Mutations in CTNNB1 (**β-catenin**) or APC genes leading to increased Wnt signaling<br>-Mostly are sporadic; but patients with **Gardner (FAP) syndrome** are susceptible<br>-Complete excision is needed to prevent recurrence which is very common<br>-**These tumors kill by local infiltration NOT metastasis** |
| **Skeletal muscle tumors** | Almost all malignant; except **rhabdomyoma** which is **benign**, rare, occurs with tuberous sclerosis | **Rhabdomyosarcoma (RMS)** is the **malignant** prototype; most common child sarcoma<br>Specific mutations are common<br>Aggressive tumors; treated by surgery, CT +/- RT |
| **Smooth muscle tumors** | **Leiomyoma (LYM)** | -benign<br>-very common; any site but mostly uterus (fibroid)<br>-menorrhagia and infertility<br>-vary in size and location<br>-Few can have specific mutations |
| | **leiomyosarcoma** | -malignant<br>-10-20% of soft tissue sarcomas<br>-Adults; more in females<br>-Deep soft tissue, extremities and retroperitoneum or from great vessels<br>-Complex genotypes<br>- Hemorrhage, necrosis, **increased mitosis** and infiltration of surrounding tissue<br>- Treatment: depends on location, size and grade |
| **Tumors of uncertain origin<br>-> Uncertain *mesenchymal* lineage** | **Synovial sarcoma** | -Name is misnomer<br>-10% of all soft tissue sarcomas; 20-40s age<br>-Deep seated mass of long history<br>-**T(X;18)** (p11;q11) fusion genes SS18<br>-Monophasic (only spindle cells) or biphasic (spindle cells and glands)<br>-Treatment: aggressive with limb sparing excision + CT -5-year survival 25-65% depending on stage<br>-Metastasis: lung and ***lymph nodes*** |
| | **Undifferentiated pleomorphic sarcoma** | -High grade mesenchymal sarcomas of pleomorphic cells that lack cell lineage<br>-Deep soft tissue and extremities |

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| | | -Old terminology: **malignant fibrous histiocytoma (MFH)**…not anymore<br>-Aneuploid and complex genetic abnormalities<br>-Large tumors; anaplastic and pleomorphic cells, abnormal mitoses, necrosis<br>-Treatment: aggressive with surgery and adjuvant CT +/- RT; poor prognosis |