Tumors of Soft Tissue

Anatomy, Work-Up, and MR Features

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Outline

I. Soft Tissue Anatomy
   - Compartmental

I. Imaging Work-Up
   - Post-Treatment Imaging

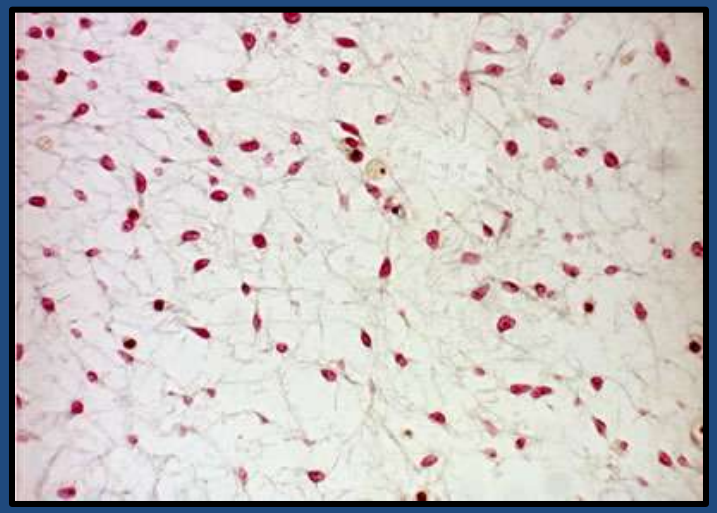
II. Soft Tissue Tumors—MR Features
   - WHO Classification

Soft Tissue Anatomy

Soft Tissue

- Derived from mesenchyme:

1. Skeletal muscle
2. Fat
3. Fibrous tissue
4. Vascular structures
5. Associated peripheral nervous system



Compartmental Anatomy

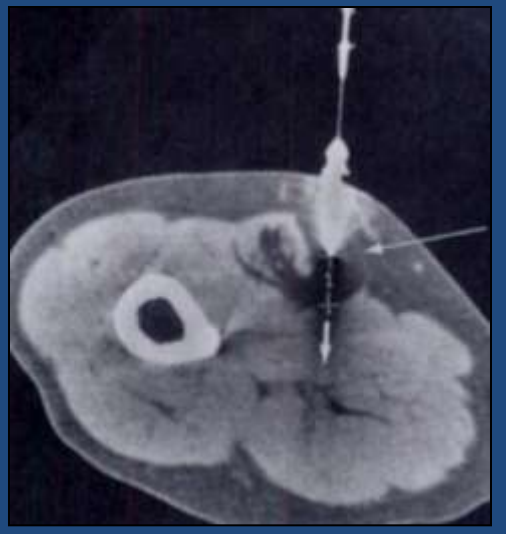
1. Local staging
   - Depends on which anatomic spaces (compartments) are involved
   - Intracompartmental lower stage

2. Biopsy
   - Risk of seeding malignant cells along needle track
   - Determines subsequent surgical approach; track usually resected

Compartmental Anatomy

- Natural Barriers define compartments:
  - Joint capsule
  - Cortex/periosteum
  - Tendon origins/insertions
  - Major fascial septae

- Extracompartmental spread by:
  - Direct tumor invasion
  - Fracture
  - Hemorrhage
  - Poorly planned biopsy



Anderson MW, et al. AJR 1999; 173: 1663-1671

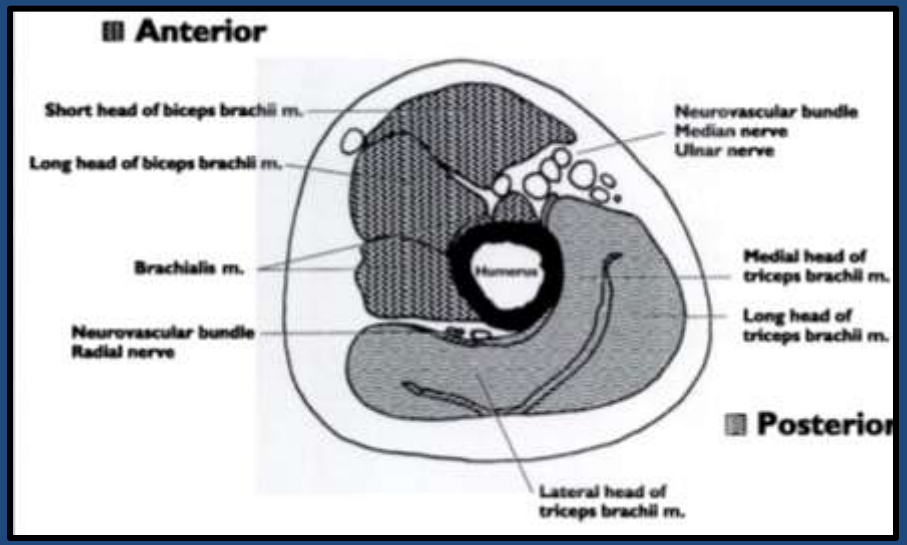
Compartmental Anatomy
General

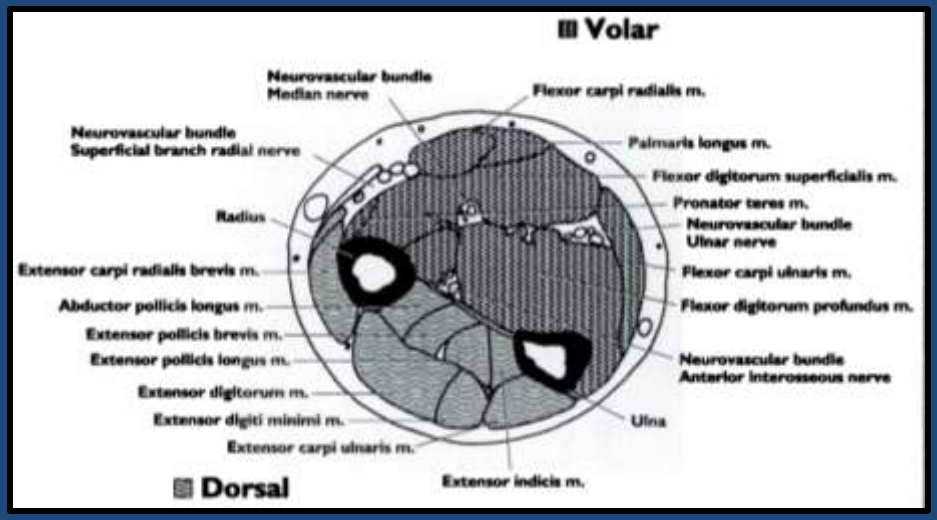
- Skin/Subcutaneous fat
- Bone
- Paraosseous
  - Space between bone and overlying tissues
- Intraarticular
- Muscle
- Neurovascular
  - Not a compartment, but can provide route of extracompartmental spread

Compartmental Anatomy
Upper Extremity

- Upper Arm
  - Anterior
  - Posterior
- Forearm
  - Dorsal
  - Volar

- Purely Extracompartmental
  - Periclavicular
  - Axilla
  - Antecubital fossa
  - Wrist
  - Dorsum of hand

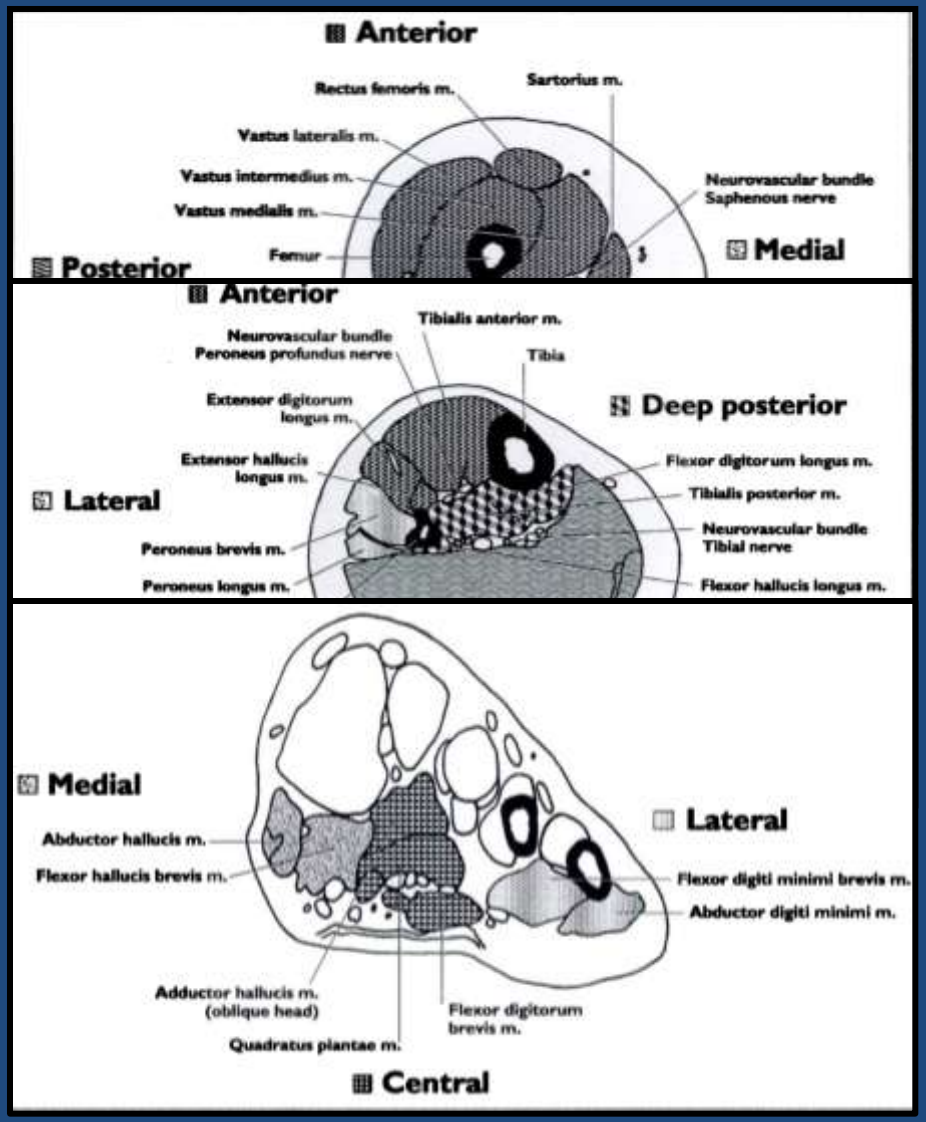




Compartmental Anatomy

Lower Extremity

- Thigh
  - Anterior
  - Posterior
  - Medial
- Lower Leg
  - Anterior
  - Deep posterior
  - Superficial posterior
  - Lateral
- Foot
  - Medial, central, lateral plantar
- Purely Extracompartmental
  - Inguinal
  - Popliteal fossa
  - Ankle
  - Dorsum of foot



Work-Up

Soft Tissue Tumors
Preliminary Evaluation

- Clinical History

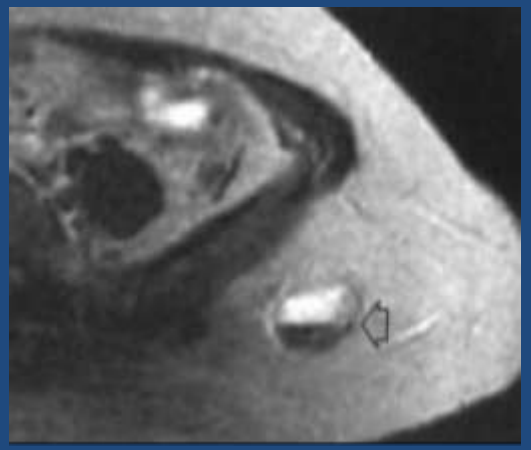
  - ✓ Previous lesion/underlying malignancy?
  - ✓ Prior surgery/radiation?
  - ✓ Painful vs painless
  - ✓ Trauma
  - ✓ Anticoagulation
  - ✓ Stability over time/Variation in size

Soft Tissue Tumors

Initial Evaluation

- > 1 lesion limits DDx

- Multiple soft tissue tumors:
  - Lipomas
  - Fibromatoses
  - Neurofibromas
  - Angiomatous lesions
  - Myxomas
  - Mets (rare)

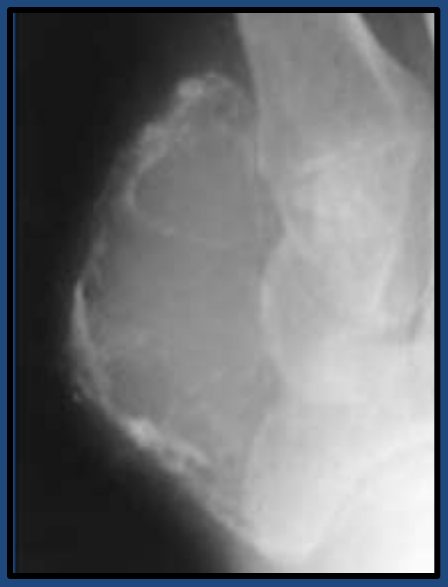


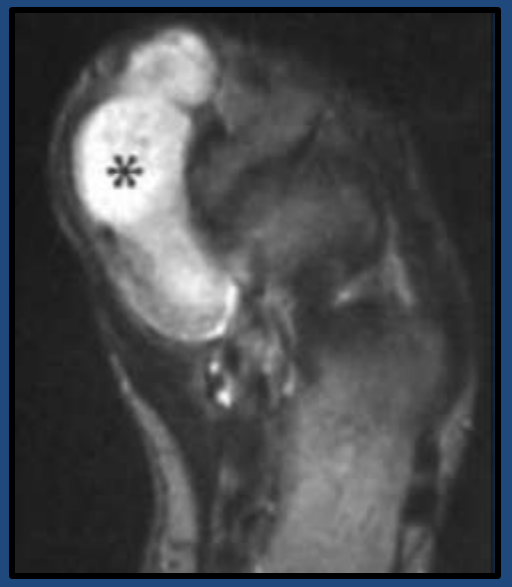
Kransdorf MJ, Murphey MD. AJR 2000; 175: 575-587

Soft Tissue Tumors

Imaging

- Radiographs (Always)
  - Specific calcifications (exostosis, phleboliths, synovial chondromatosis, myositis ossificans)
  - Non-specific calcifications (dystrophic in slow growing mass suggests synovial sarcoma)
  - Osseous Involvement





Soft Tissue Tumors
Imaging

- Sonography
  - Fast
  - Inexpensive
  - Ideal for solid vs. cystic when anatomically accessible

- CT
  - Further evaluate pattern of mineralization
  - Relationship to nearby complex osseous structures (Pelvis, shoulder, paraspinal)

- MRI
  - Modality of choice
  - Superior soft tissue contrast

Soft Tissue Tumors

Imaging

- MR cannot reliably distinguish benign from malignant soft tissue masses

- Non Specific:
  - Contrast enhancement (solid v. cystic, hematomas, necrosis for biopsy or trtmt response)

- Suggestive of malignancy:
  - Larger (5% benign tumors > 5 cm)
  - Heterogenous signal (infarction, necrosis)
  - Well-defined borders
  - Deep (1% benign tumors are deep) > superficial

Kransdorf MJ, Murphey MD. AJR 2000; 175: 575-587

Post-Treatment Imaging

Soft Tissue Tumors
Post-Treatment Imaging

- 50% patients with soft tissue sarcomas have local recurrence

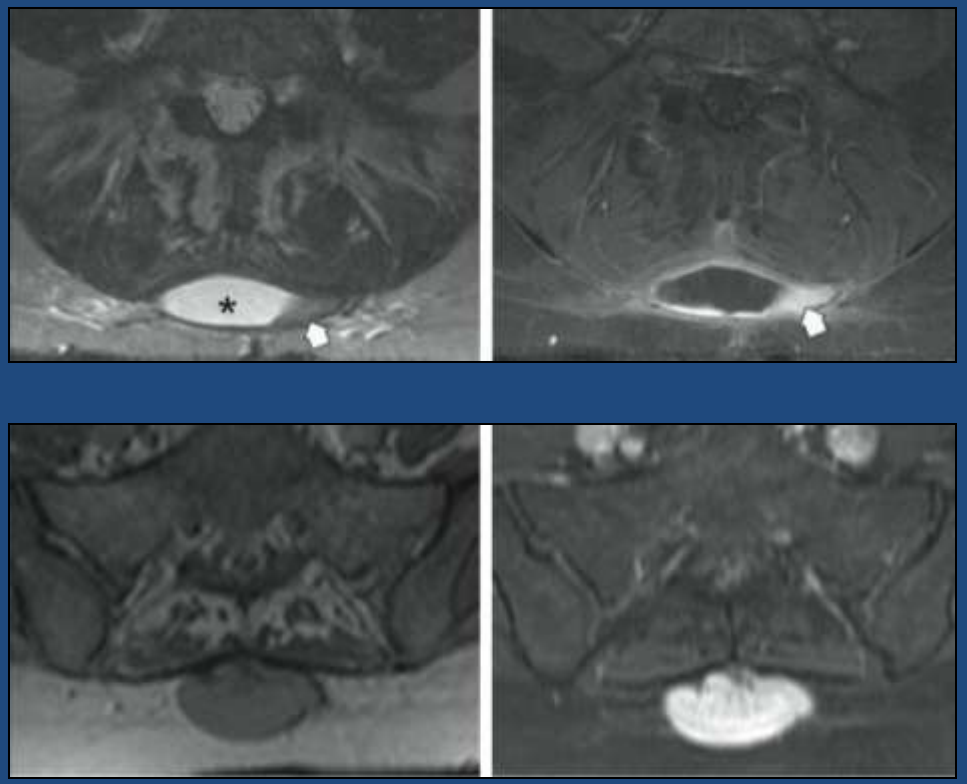
- Increase Risk for Local Recurrence:
  - Tumor diagnosis
  - High Grade
  - Deep location
  - Unable to obtain wide margins
  - Radical resection vs marginal excision
  - Positive Surgical margins

- Radiation or chemotherapy (time course)
- Reconstructive surgery (time course)

Soft Tissue Tumors

Post-Treatment Imaging

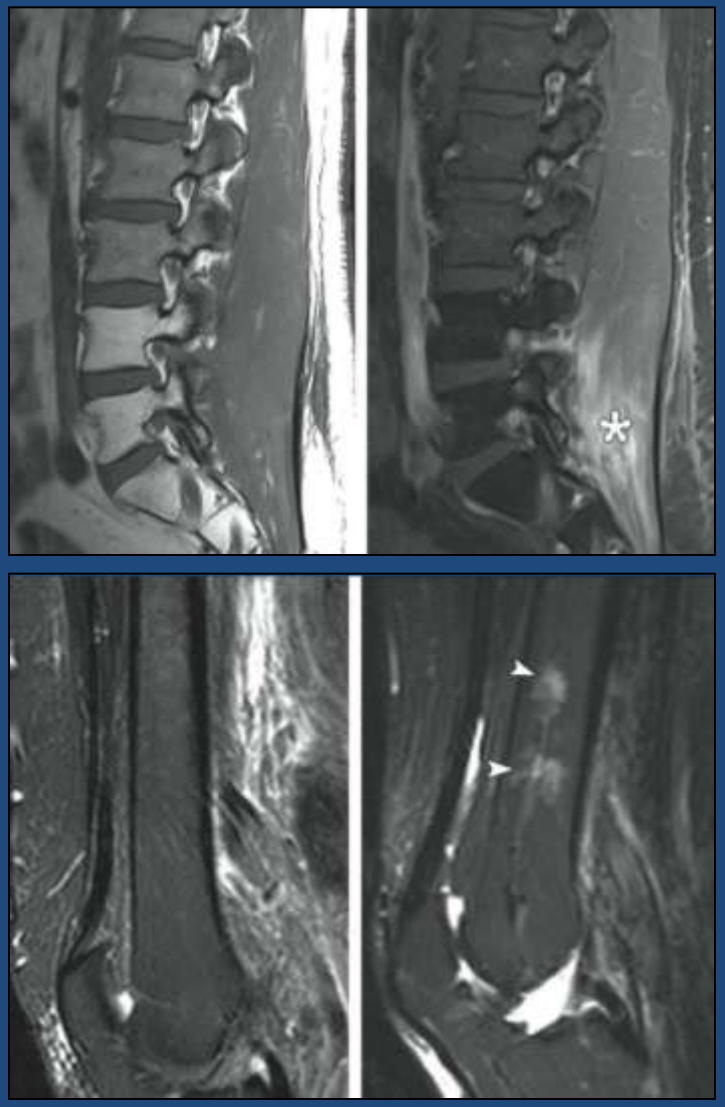
- MR
  - Discrete nodule (Post surgical changes more variable)
  - Recurrent tumor looks like the primary tumor (review pre-op)
  - Markers noting scar margins
  - Contrast (necrosis/response, hematoma)



Garner HW, Kransdorf MJ, Bancroft LW, Peterson JJ, Berquist TH, Murphey MD. Radiographics 2009; 29:119-134.

Soft Tissue Tumors
Radiation

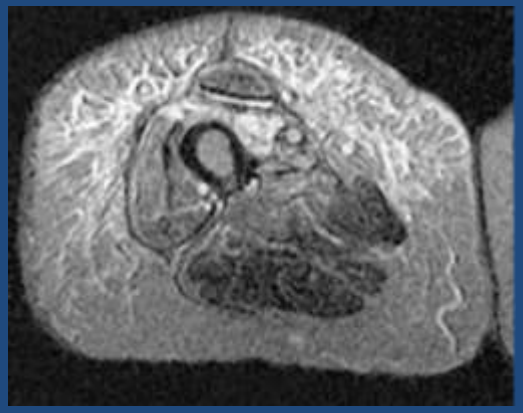
- Marrow changes
  - As early as 8 days
  - Increasing fatty signal (1-6 wks)
  - Complete fatty replacement in 6-8 wks
  - Can see focal non specific signal (radiation osteitis), mean 9 months



Garner HW, Kransdorf MJ, Bancroft LW, Peterson JJ, Berquist TH, Murphey MD. Radiographics 2009; 29:119-134.

Soft Tissue Tumors
Radiation

- Soft Tissue Changes
  (Peak 12-18 mo; half return to nl in 2-3 yrs)
  - Trabecular/lattice-like subcutaneous signal
  - Diffuse muscle enhancement, preservation of shape and architecture
  - Signal persists in intermuscular septae longer
  - Pseudotumor
  - Sarcoma



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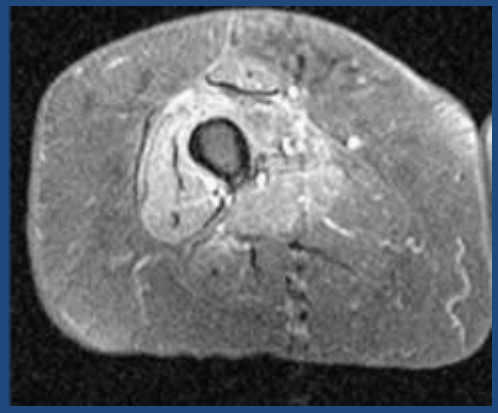
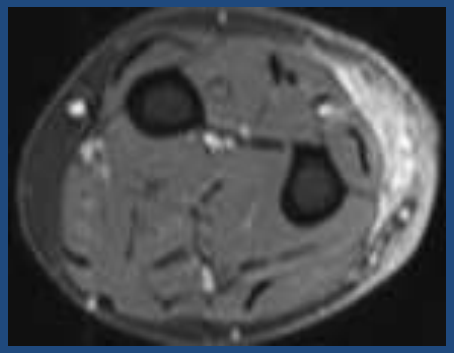
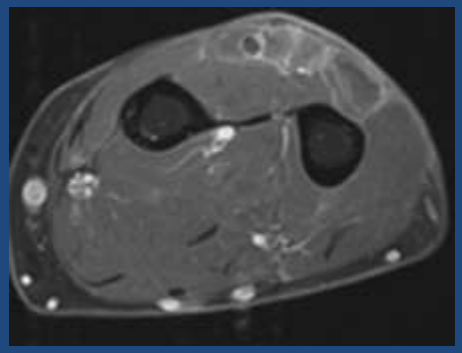


Post





3 yrs

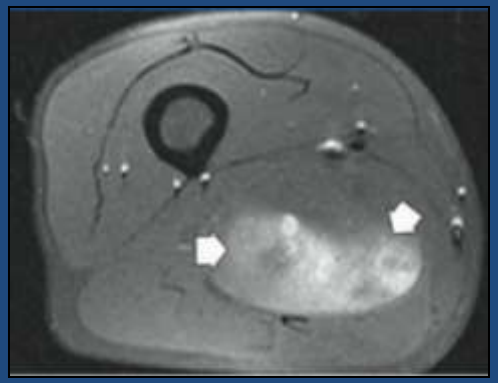
Kransdorf MJ, Murphey MD. RCNA 2006; 44:463-472

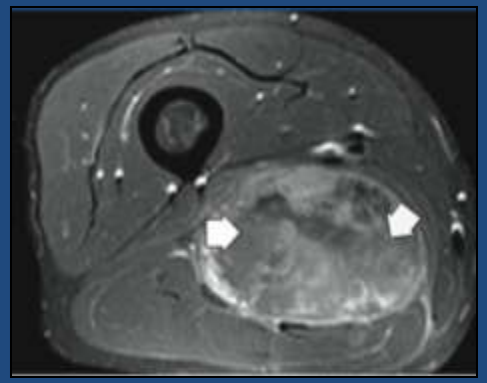
Soft Tissue Tumors

Post-Treatment Changes

Chemotherapy

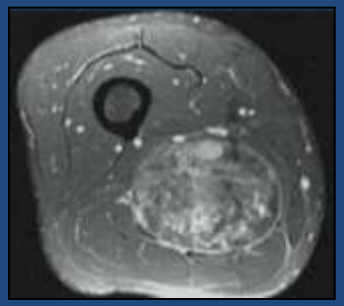
- May increase tumor size at first due to hemorrhage
- Necrosis predicts response

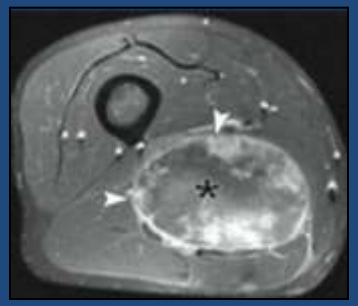


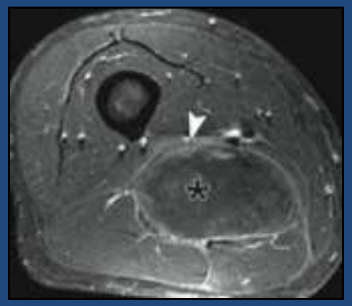


Postoperative Fluid and Hemorrhage:

- Similar appearance seen with non-oncologic procedures
- Most seromas resolve in 3-18 months







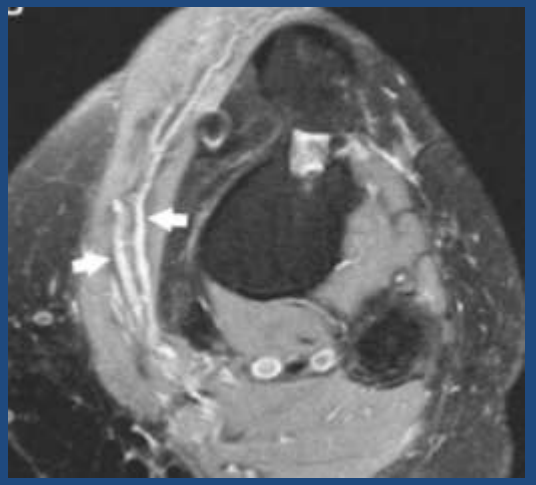
Soft Tissue Tumors
Post-Treatment Changes

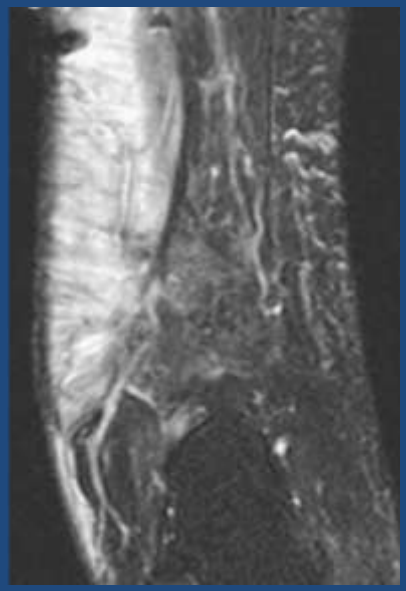
- Reconstructive Surgery
  - Myocutanous flaps used in > 2/3
  - Rotational Flaps
    - Rotated into position preserving native neurovascular pedicle
  - Free Flaps
    - Completely detached with vascular pedicle reanastomosed

Soft Tissue Tumors

Reconstructive Surgery

- Atrophy with time (less with those providing function)
- Increased T2 signal initially
- Signal returns to baseline within 2 yrs (1/3 cases)
- Enhancement in 3/4; returns to baseline in 18 months in 1/3





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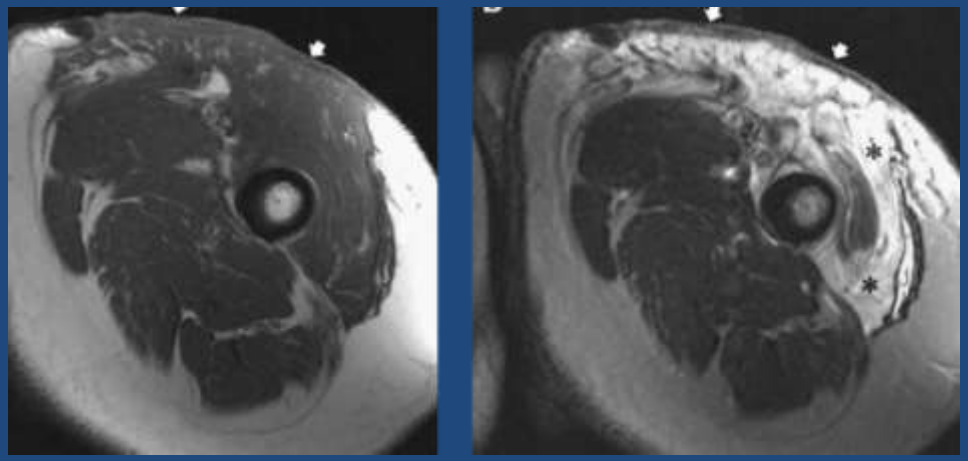
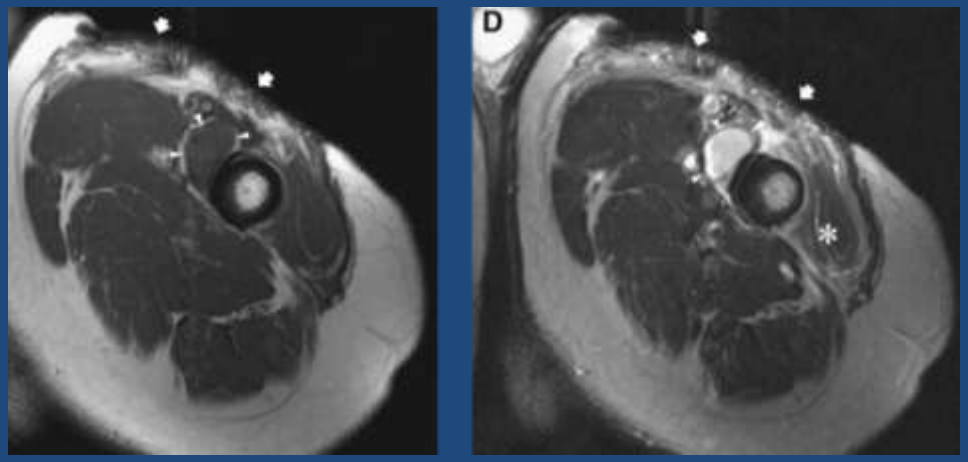
Soft Tissue Tumors

Post-Treatment Changes

4 months



31 months

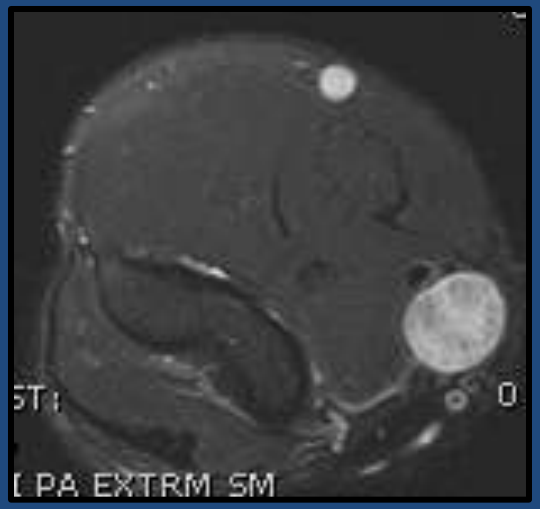


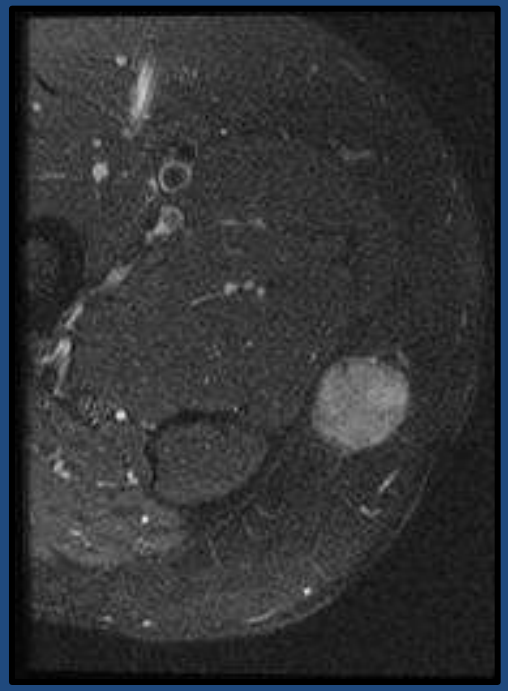
Kransdorf MJ, Murphey MD. RCNA 2006; 44:463-472.

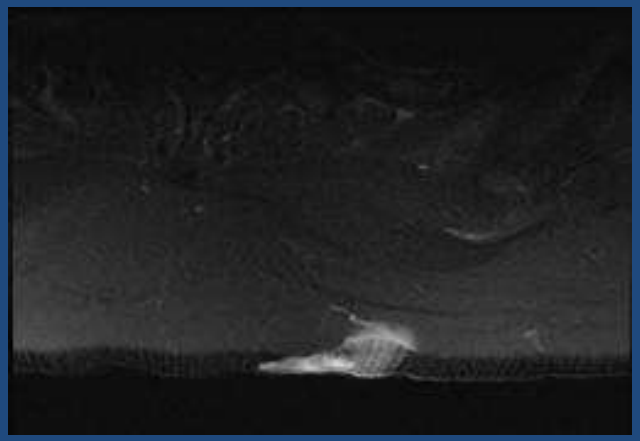
Soft Tissue Tumors

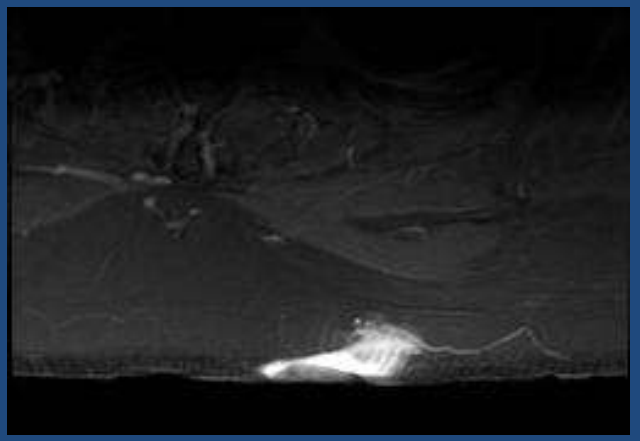
Soft Tissue Tumors

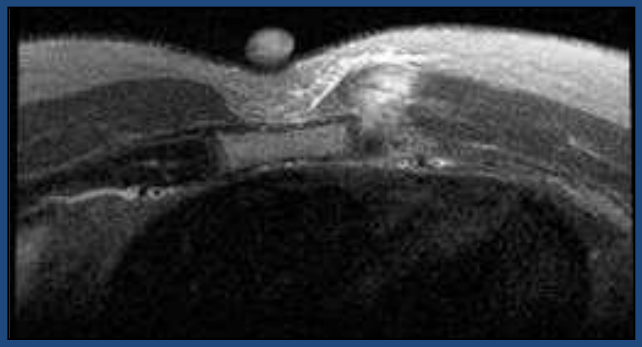
- Benign 100X more common than malignant
- Soft tissue sarcomas 2-3X more common than primary malignant bone tumors
- Tumors classified histologically based on adult tissue they resemble
- Many demonstrate specific MR features, but majority are nonspecific

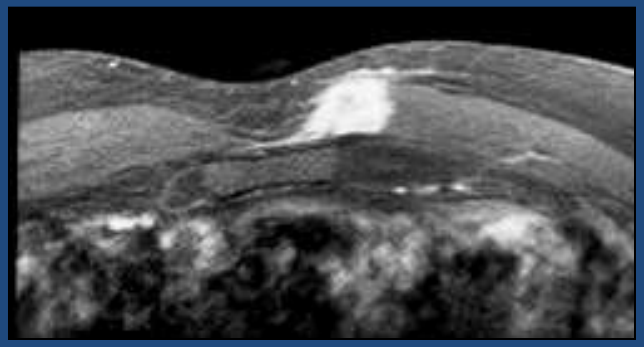












Courtesy Tudor Hughes, M.D.

Soft Tissue Tumors

WHO Classification

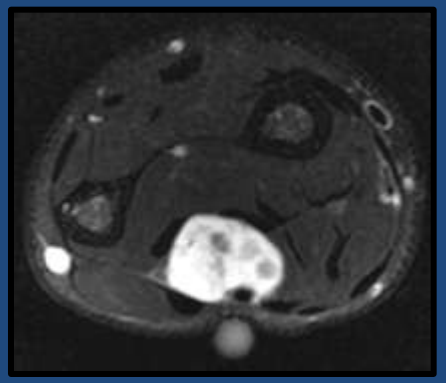
- Neurogenic
- Vascular
- Fibroblastic
- Adipocytic
- Fibrohystiocytic
- Smooth Muscle
- Perivascular
- Skeletal Muscle
- Chondro-osseous
- Tumors of uncertain differentiation

Neurogenic Tumors

Peripheral Nerve Sheath Tumors

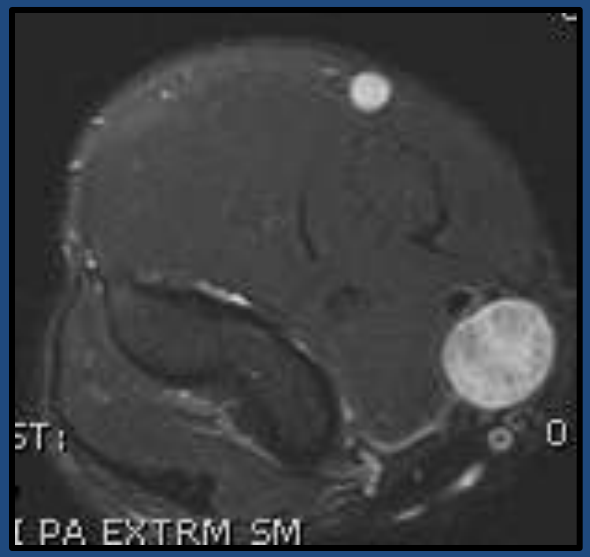
Benign

- Schwannomas/Neurofibromas
  - Fascicular Sign
  - Split Fat Sign



- Neurofibroma
  - Target pattern (T2)
  - Infiltrative: resection sacrifices nerve

- Schwannoma
  - Eccentric
  - Displaces nerve: resection spares nerve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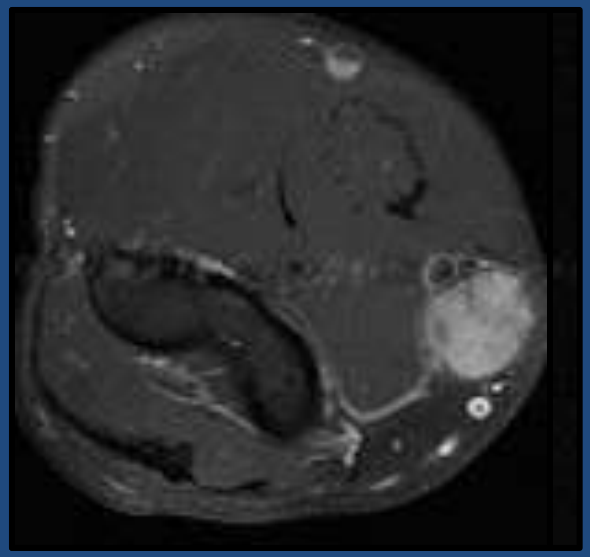
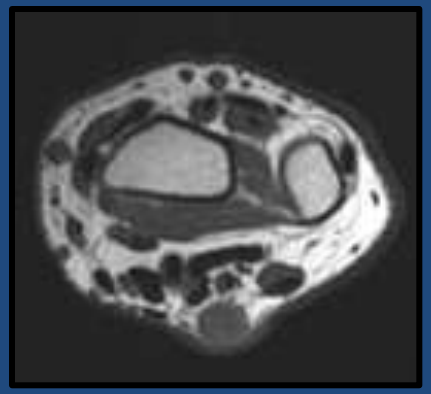
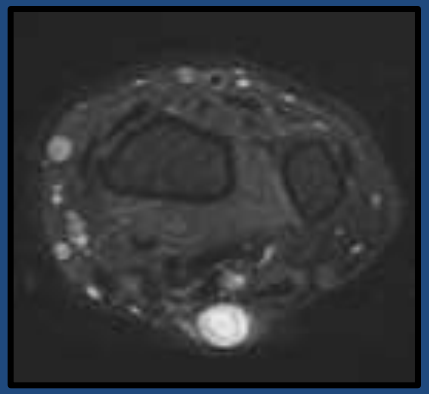
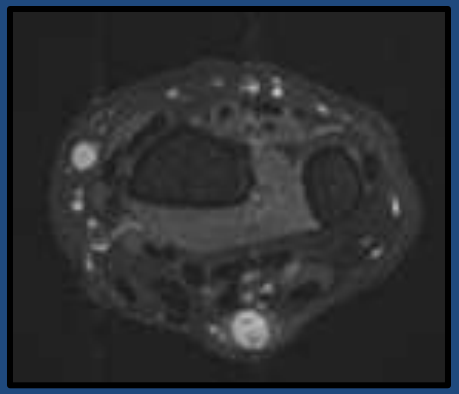
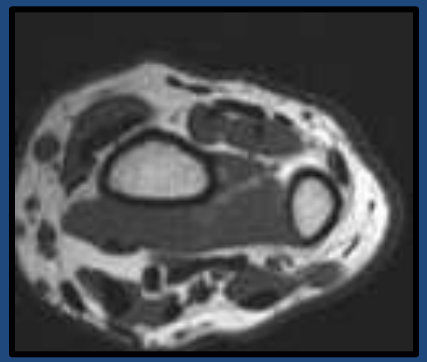
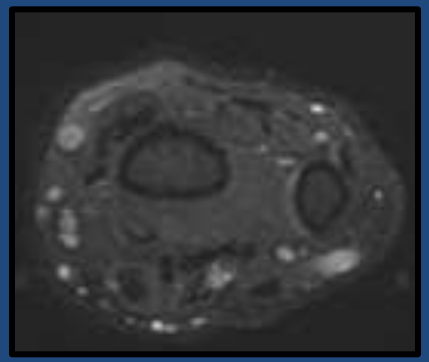


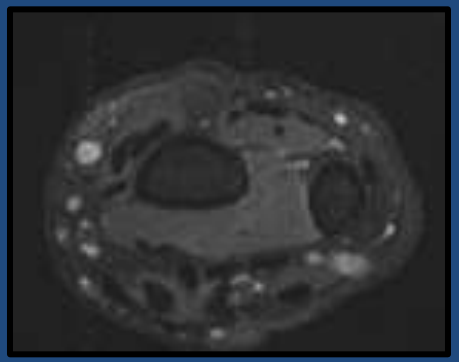


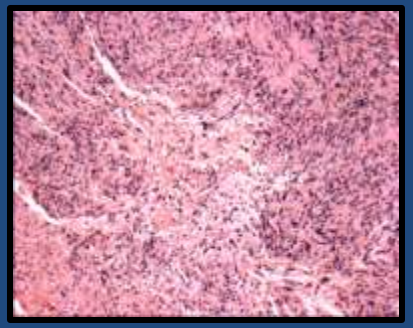








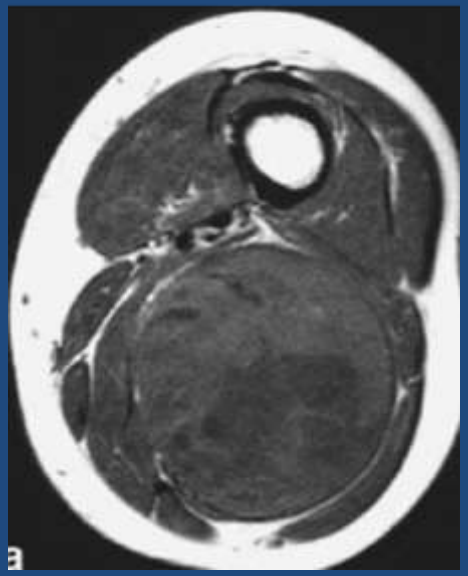


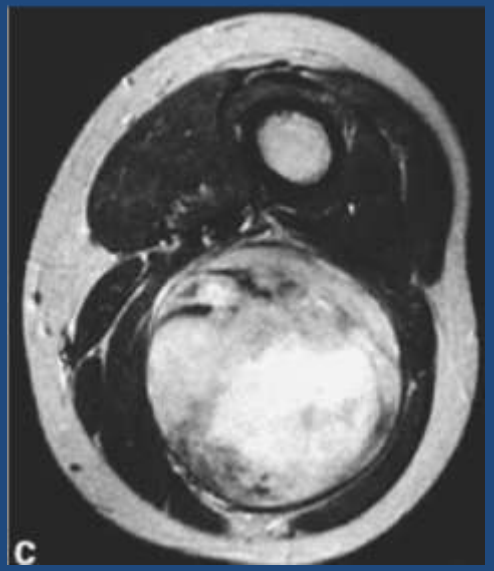


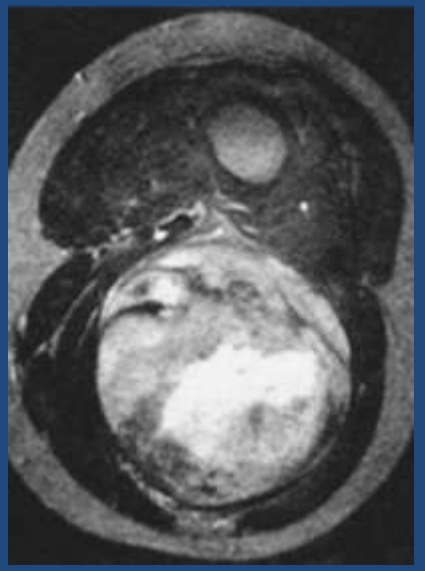
Peripheral Nerve Sheath Tumors

Malignant

- Pain, rapid growth
- No target, split fat, or fascicular sign
- Intralesional hemorrhage and necrosis (peripheral enhancement)
- Inhomogenous (T1, T2, Post)
- Nodular
- Along course of large nerve

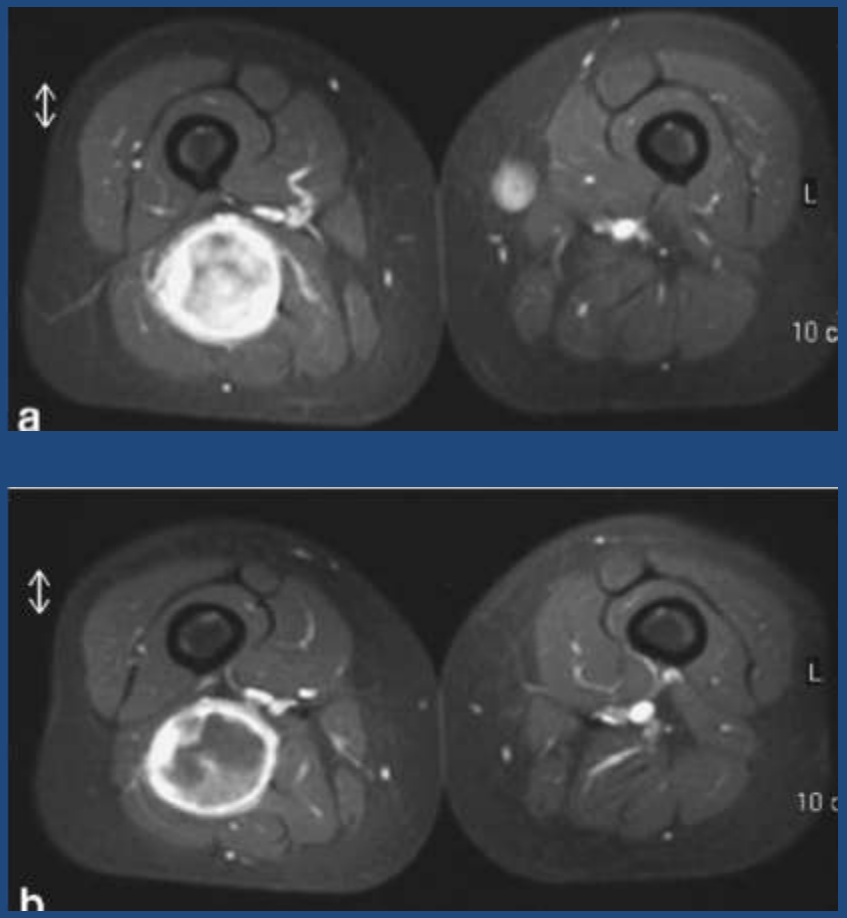






Peripheral Nerve Sheath Tumors

Malignant

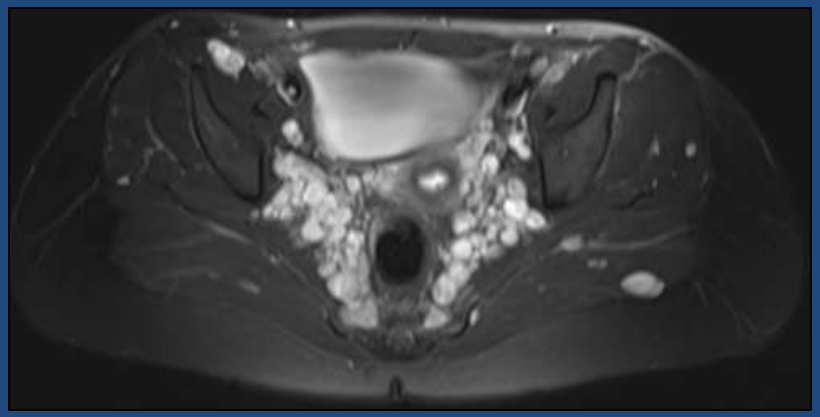


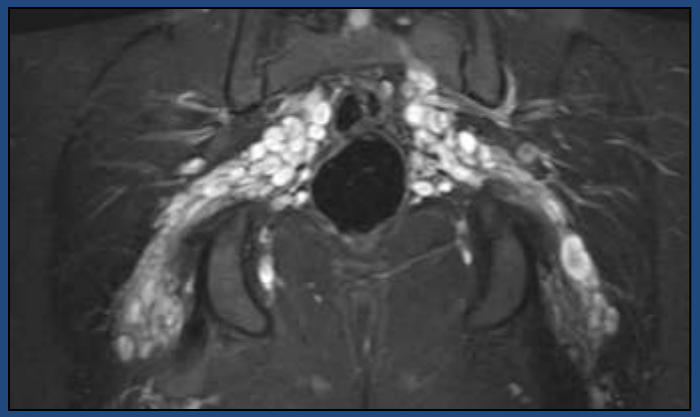
T2

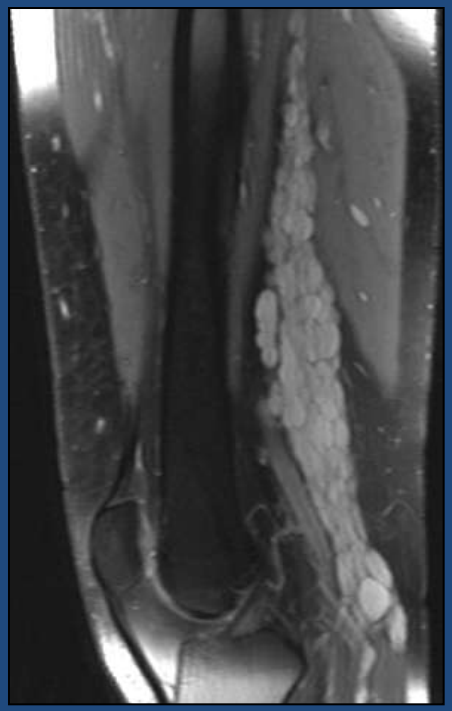
T1 Post-Gad

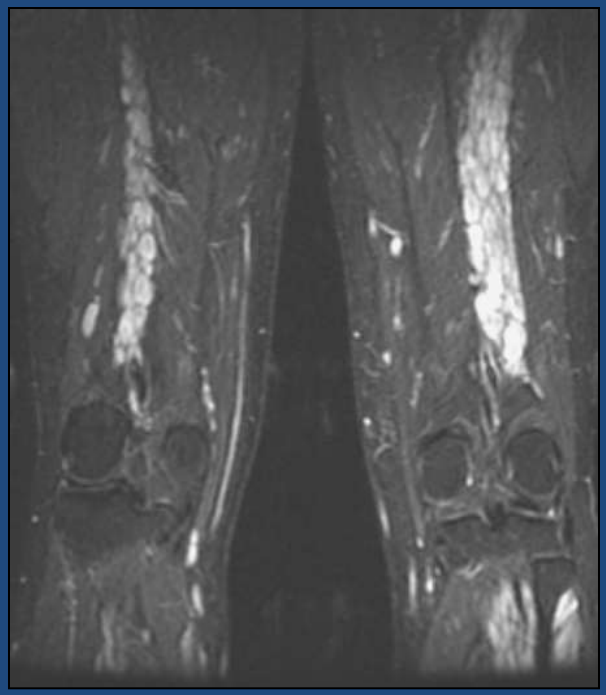
Van Herendael BH, et al. Skeletal Radiology 2006; 35: 745-753.

Malignant Peripheral Nerve Sheath Tumor?



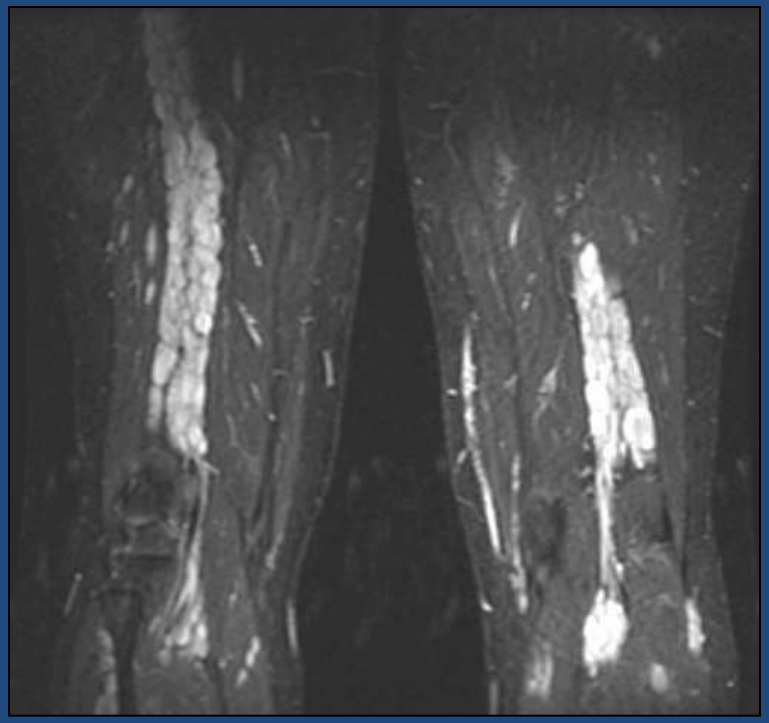






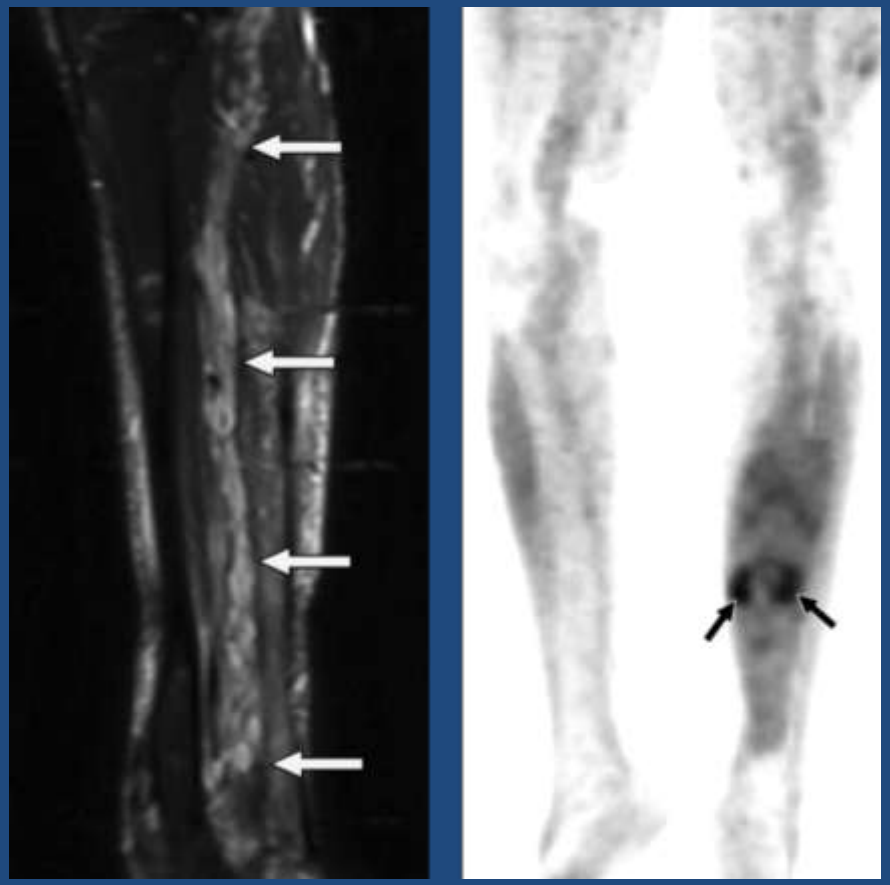
Malignant Peripheral Nerve Sheath Tumors

- MR and CT not reliable in characterizing benign vs. malignant
- Surgical resection of entire lesion often not feasible
- Biopsy may yield false negative due to sampling error



MPNST and FDG-PET

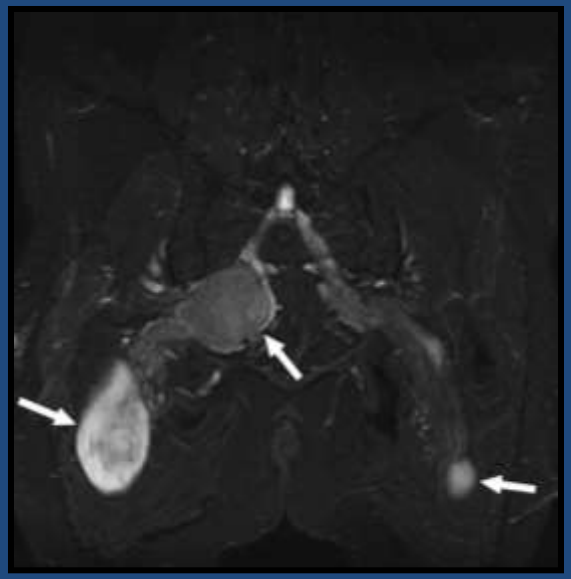
- FDG PET sensitive (95%) in detecting MPNST in patients with NF1
- Can also detect mets or second primaries (GIST which is associated with NF1)

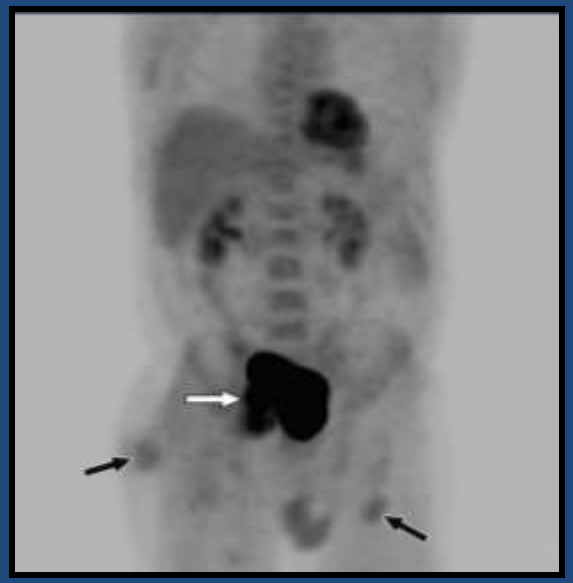


Bredella MA, et al. AJR 2007; 189:928–935

MPNST and PET

- PET specificity lower (72%)
- Can use 11-C Methionine PET to increase specificity (91%)





Bredella MA, et al. AJR 2007; 189:928–935

Vascular Tumors

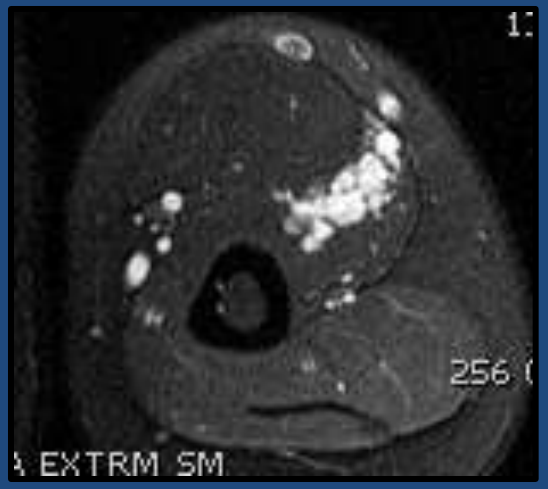
Vascular Anomalies

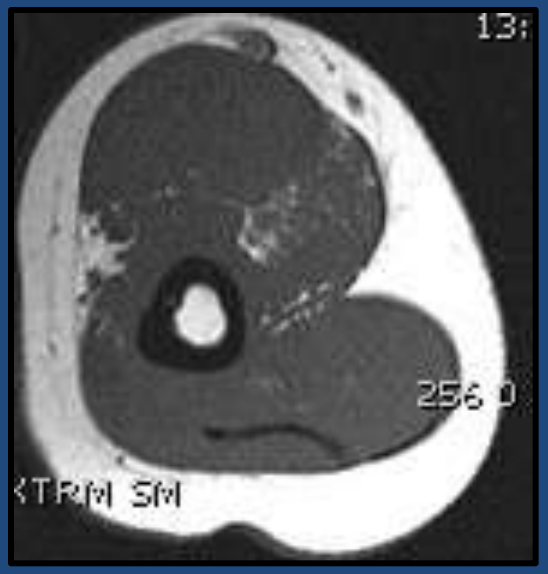
Tumors of Childhood	Vascular Malformations
• Hemangioma ➢ *Childhood neoplasm with a proliferative and involutive phase (not applicable to any adult lesion)*	• Capillary • Venous • Lymphatic • Ateriovenous • Mixed

Fayad L, et al. Vascular Malformations in the Extremities: Emphasis on MR Imaging Features that Guide Treatment Options. Skeletal Radiology 2006; 35:127-137.

Vascular Malformations

- Prevalence 1.5%
- Pelvis, extremities, intracranial most common
- Not neoplastic (do not proliferate or involute)





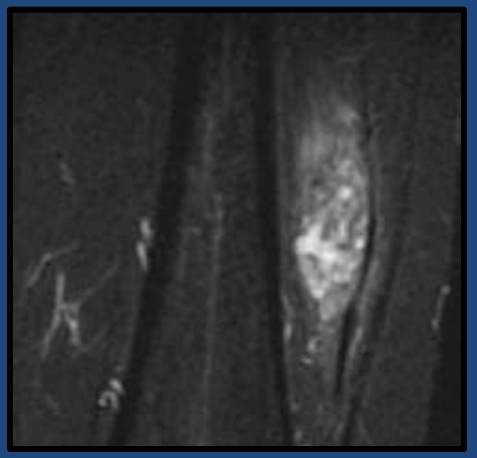
Vascular Malformations

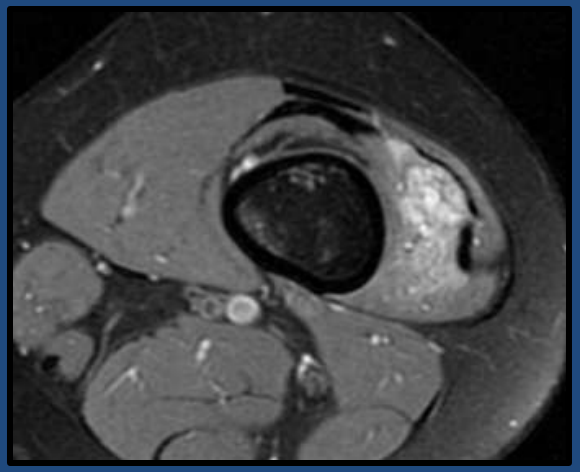
Low Flow	High Flow
• Venous • Capillary • Lymphatic	• Arteriovenous malformation • Arteriovenous fistula

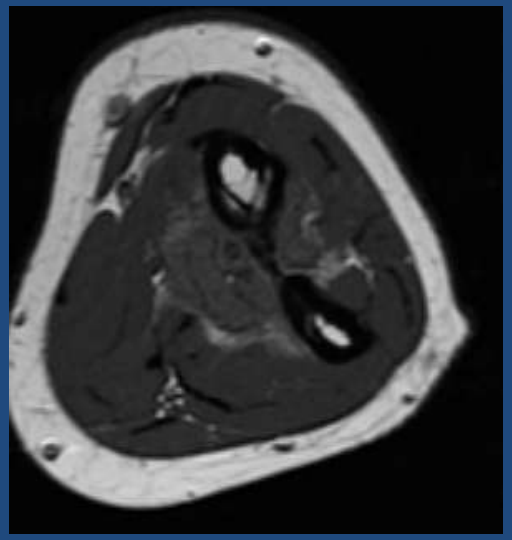
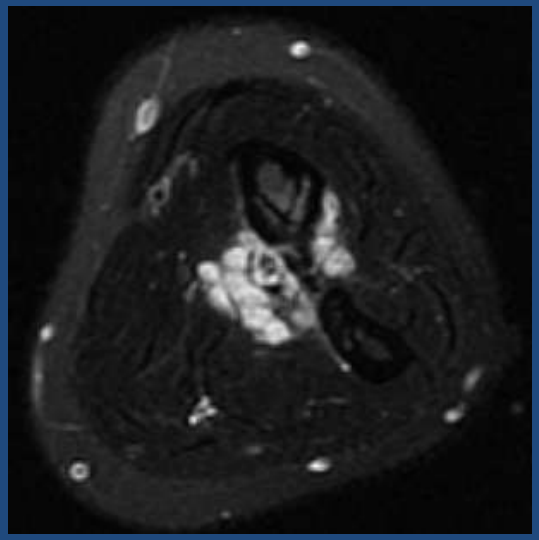
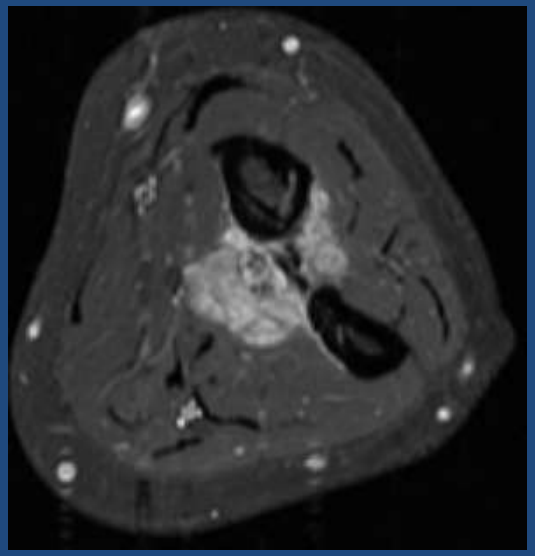
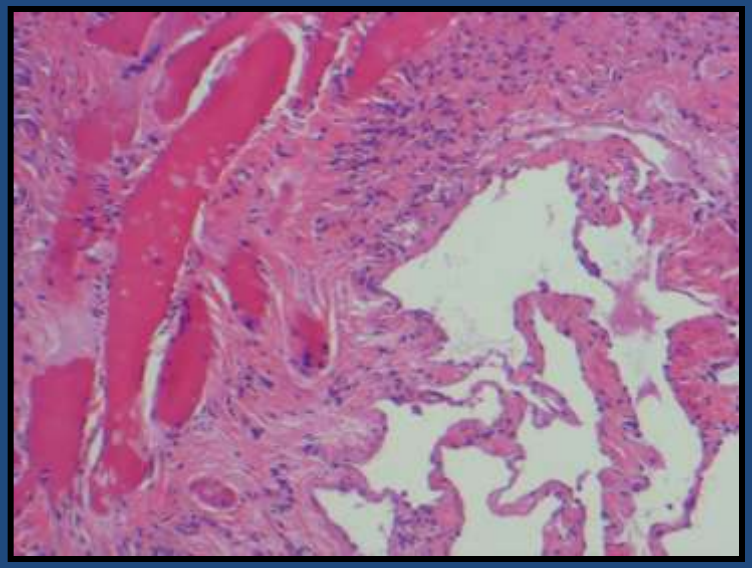
Fayad L, et al. Vascular Malformations in the Extremities: Emphasis on MR Imaging Features that Guide Treatment Options. Skeletal Radiology 2006; 35:127-137

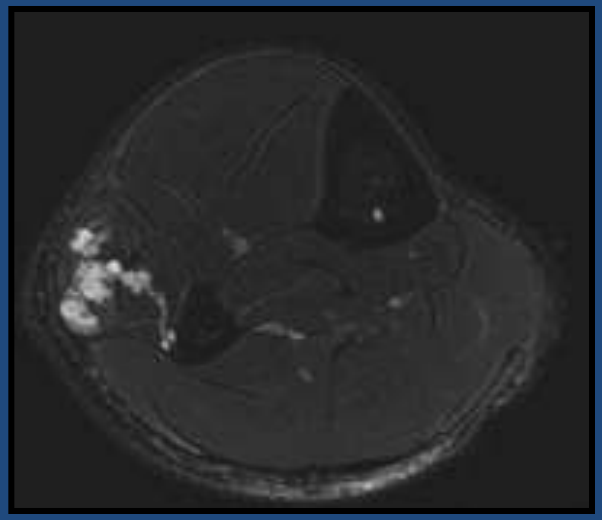
Low Flow Malformations

- Venous most common of the extremities
- Present at birth, grow proportionately with patient, do not regress
- Forearm flexors and quadriceps muscle most common (venous)





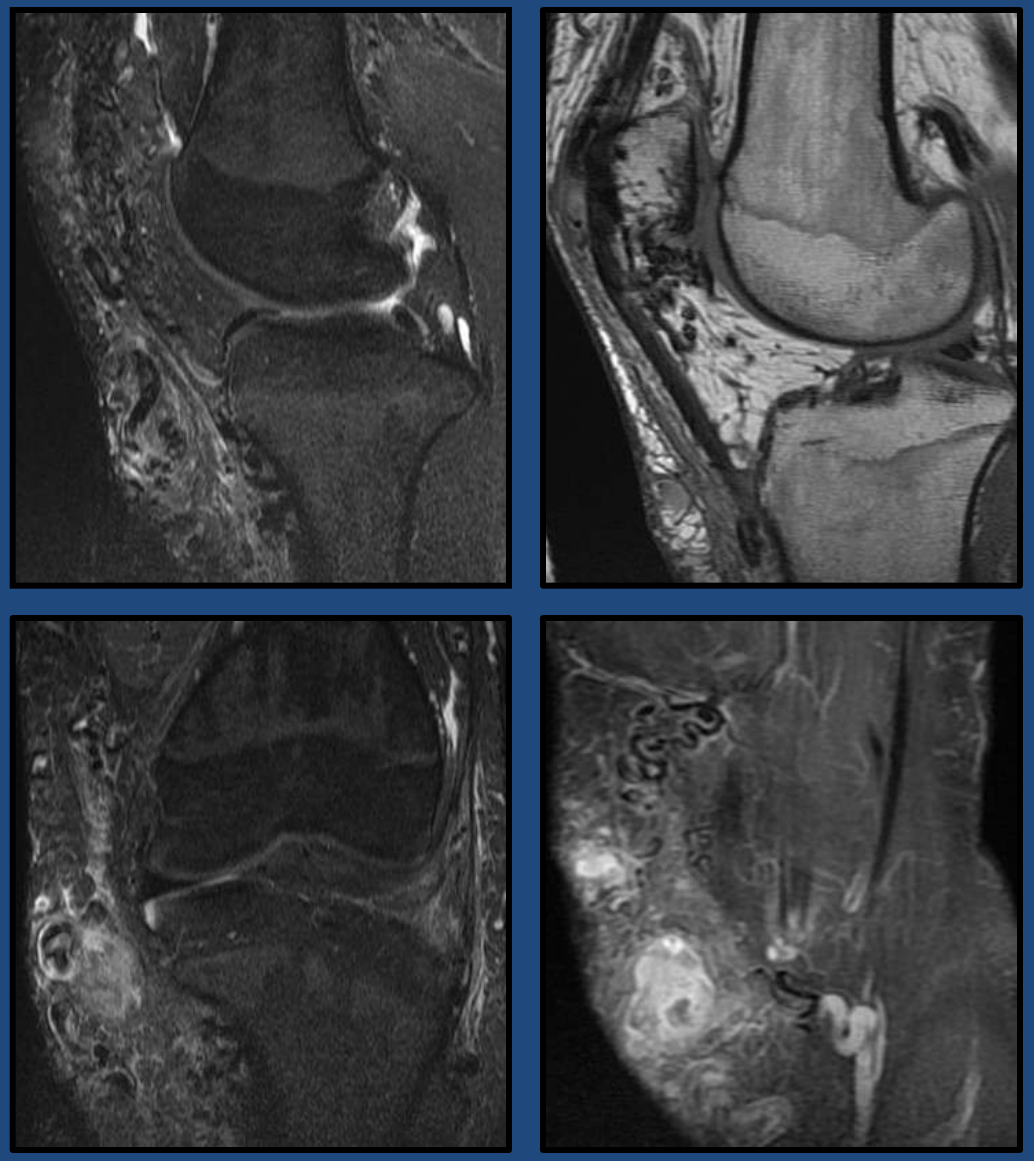









TR/free PA
ET:1
C:1

High Flow Malformations

- AVM
  - Feeding arteries and draining veins connected by multiple dysplastic vessels
- AVF
  - Direct connection between arteries and veins, bypassing capillary bed



Vascular Malformations

MR Assessment

1. **Distinguish from Hemangioma**
   - Age + no mass effect (caution atypical low flow lesions which can appear mass-like and share features of hemangiomas, angiosarcomas, myxoid, fibrosarcoma)
2. **Low vs. High Flow**
   - Flow voids
   - Feeding arteries, draining veins, dysplastic vessels
3. **Focal, multifocal, or diffuse**
4. **Adjacent tissue involvement**
   - Skin, subcutaneous, muscle, tendon, bone
   - Can contain fat, hemosiderin, Ca++, thrombus
5. **Connection to normal vessels**
   - Arterial vs. Deep Venous (DVT risk)

Fibrous Tumors

Fibrous Tumors

I. Benign Fibrous Proliferations

- Nodular Fasciitis
- Proliferative Fasciitis
- Proliferative Myositis
- Fibroma of the Tendon Sheath
- Keloid/Hypertrophic Scar
- Elastofibroma

II. Fibromatoses

- Superficial (Palmar, Plantar, Penile)
- Deep (Intraabdominal, extraabdominal)

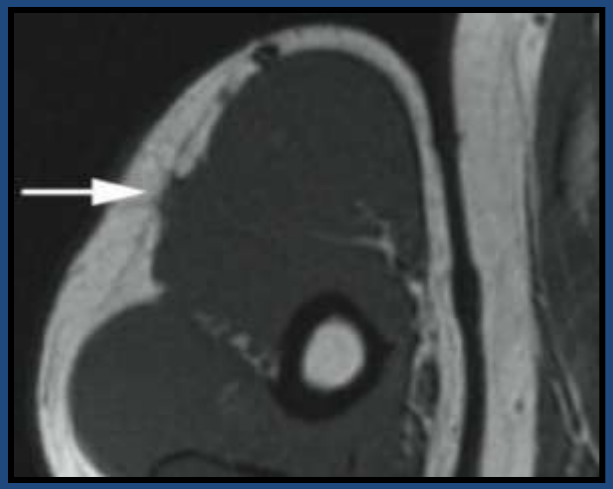
III. Fibrosarcomas

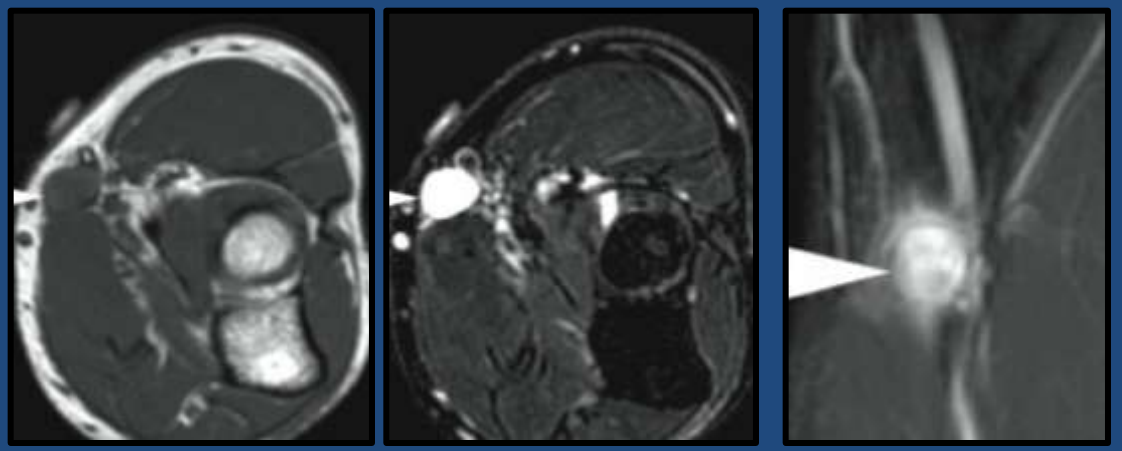
I. Fibrous Proliferations of Infancy/Childhood

Benign Fibroblastic Proliferations

Nodular Fasciitis

- Most common benign mesenchymal lesion histopathologically misdiagnosed as sarcoma
- 20-40 years
- < 4 cm, rapidly growing
- Upper extremity (volar forearm)



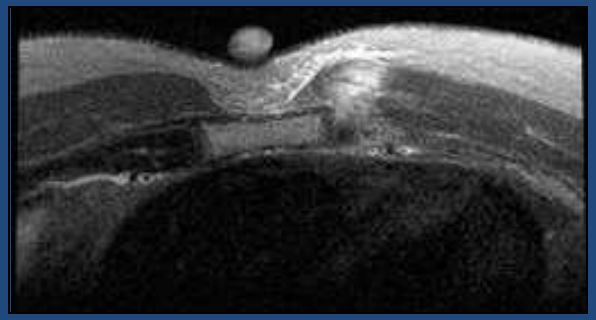


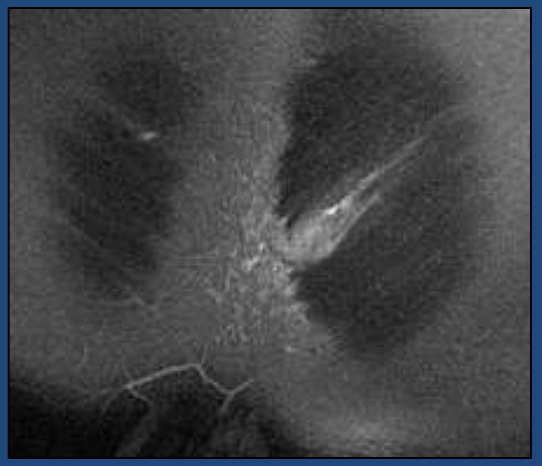
Dinauer PA, et al. Pathologic and MR Imaging Features of Benign Fibrous Soft Tissue Tumors in Adults. Radiographics 2007; 27: 173-187

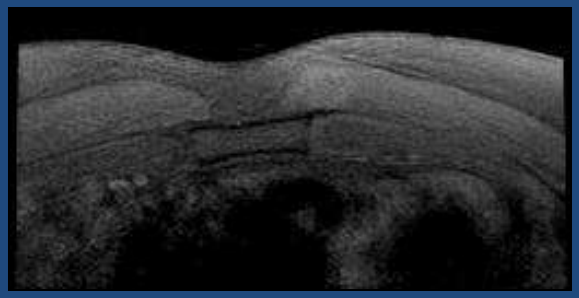
Benign Fibroblastic Proliferations

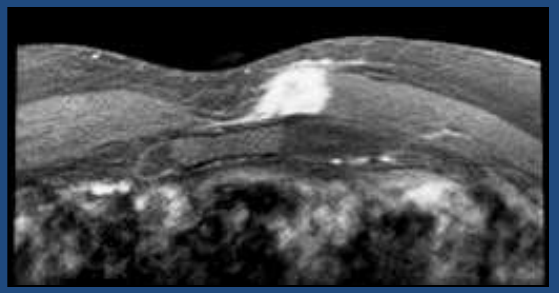
Nodular Fasciitis

- Typically subcutaneous, and attached to superficial fascia
- Low to intermediate signal on T1 and Intermediate to high signal on T2
- Enhance
- Fascial tail sign







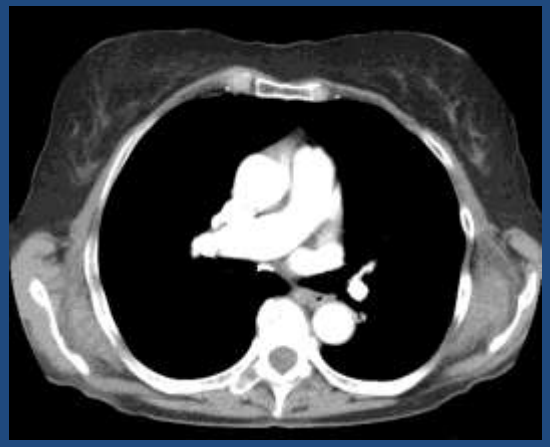


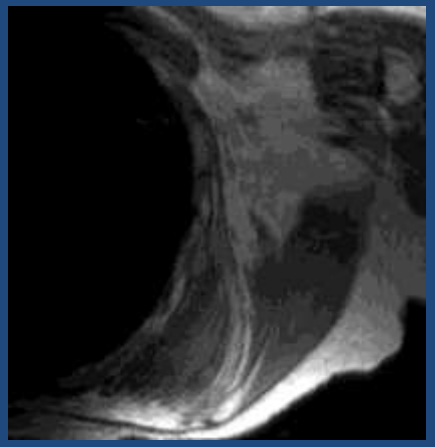
Courtesy Tudor Hughes, M.D.

Benign Fibroblastic Proliferations

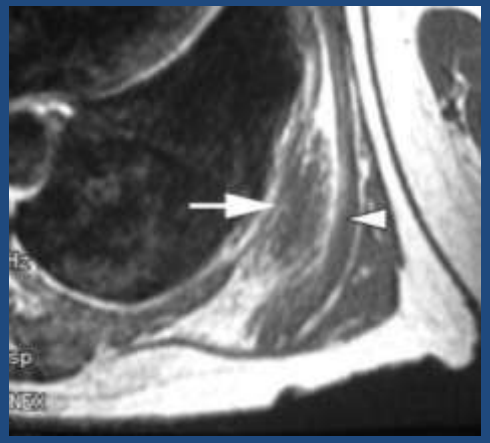
Elastofibroma

- > 55 years
- Between posterior chest wall and inferomedial scapula border (also about greater trochanter and olecrenon)
- Bilateral (25%)
- Signal similar to skeletal muscle intermixed with streaky fat signal
- Heterogenous enhancement



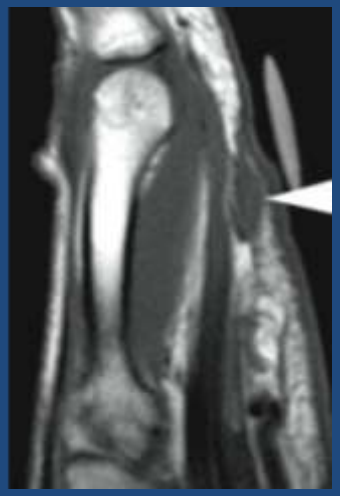


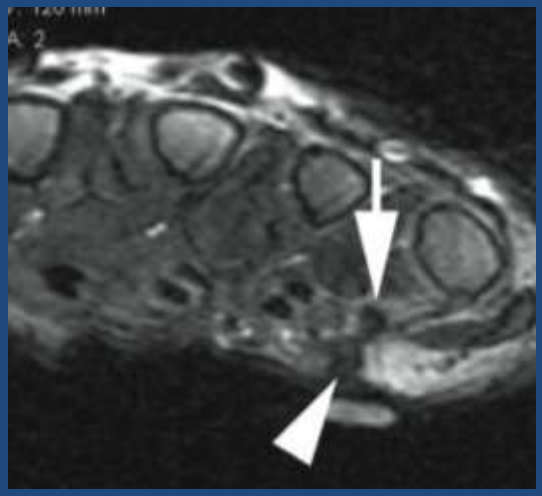
Courtesy Tudor Hughes, M.D.



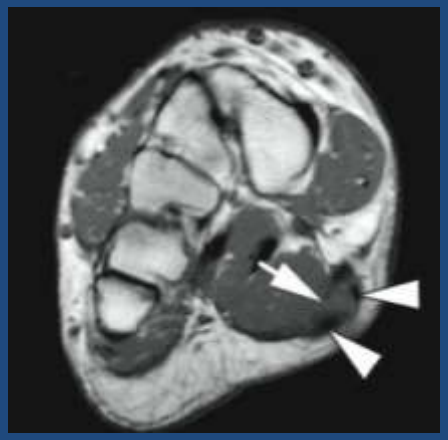
Fibromatoses
Superficial

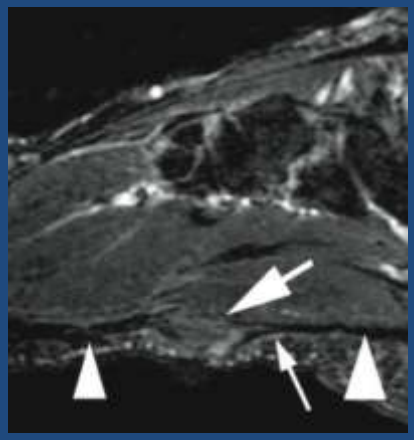
- Palmar Fibromatosis (Dupuytren Disease)
  - Volar aponeurosis of hand
  - > 30 years
  - Variable T2 depends on collagen maturity and may suggest propensity to recur





- Plantar Fibromatosis (Ledderhose Disease)
  - Bilateral 20-50%
  - M > F (2X)
  - Associated palmar fibromatosis (10-65%)





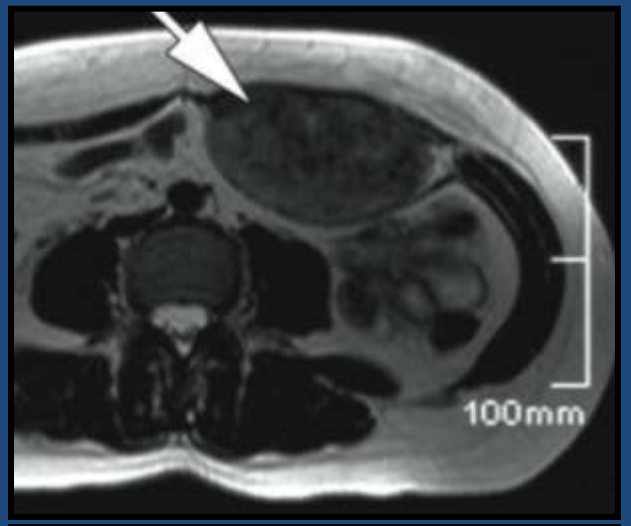
Dinauer PA, et al. Radiographics 2007; 27: 173-187

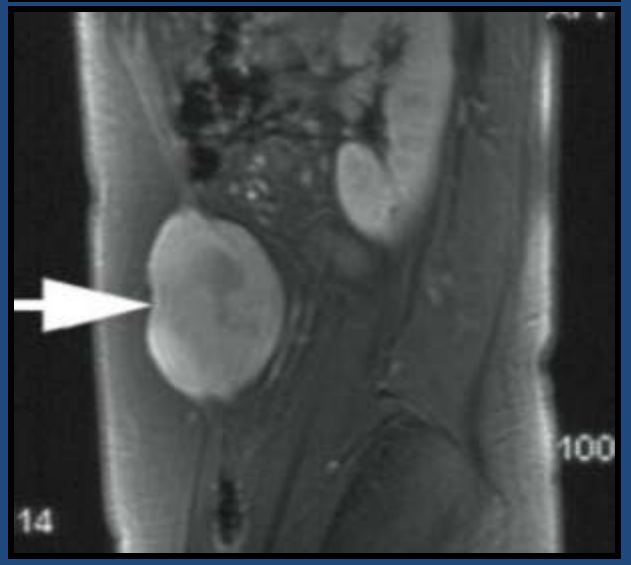
Fibromatoses
Deep (Desmoid Tumors)

I. **Intraabdominal**
   - FAP (Gardner Syndrome)

II. **Abdominal**
   - Pregnant women, or OCP
   - Rectus abdominis and Internal Oblique

III. **Extraabdominal**
   - > 5 cm
   - Typically solitary
   - Can be aggressive, local recurrence high (87% in < 20 yo)

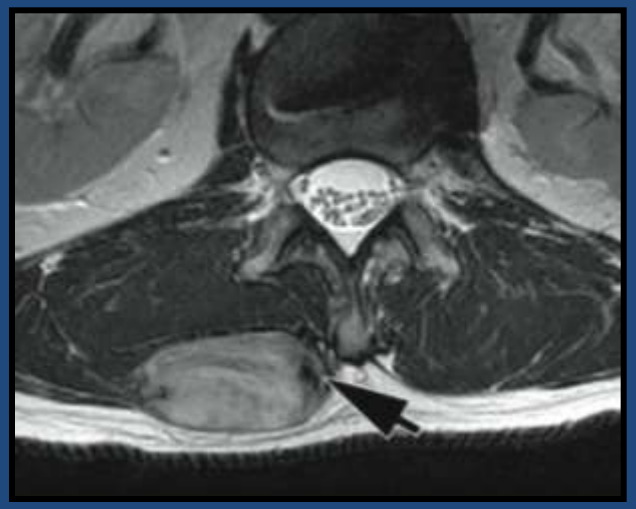


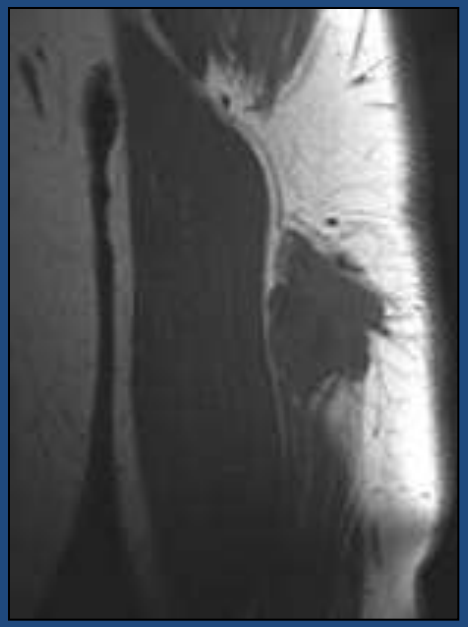


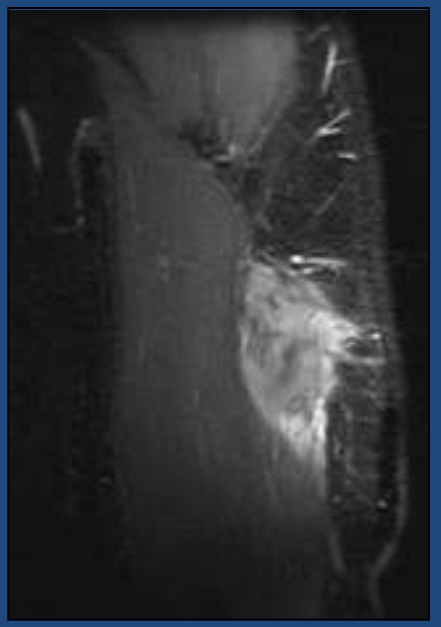
Deep Fibromatoses

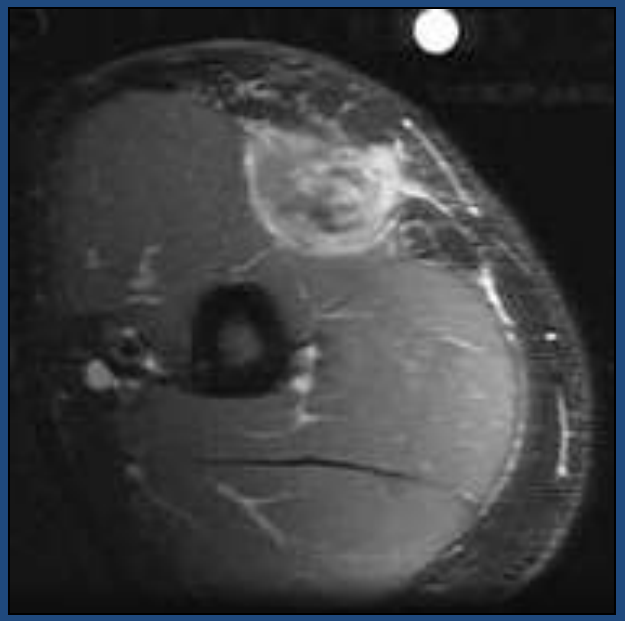
MR Features

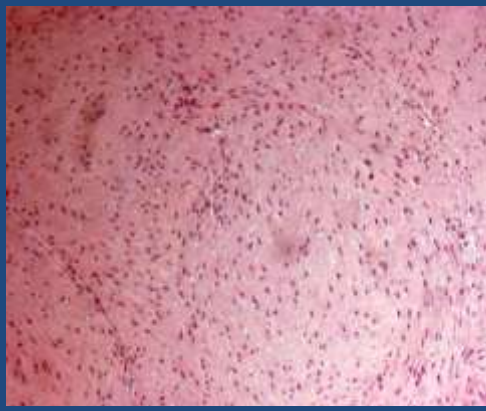
- Non-enhancing, T2 hypointense bands corresponding to collagen bundles (86%)
- Infiltrative border or fascial tail (80%)
- Evaluation of response to treatment:
  - Decreased cellularity and increased collagen show low T2 signal (positive response)











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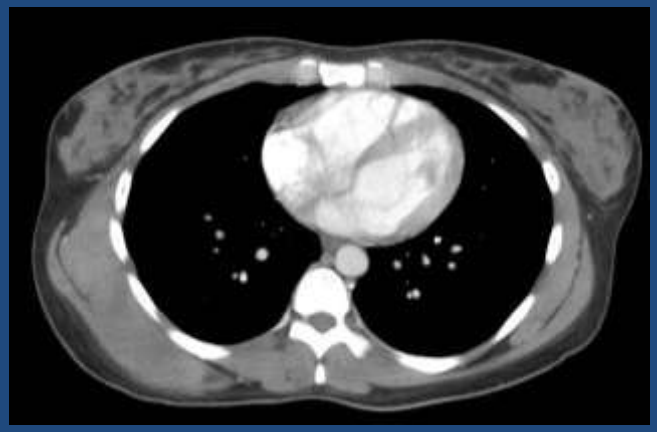
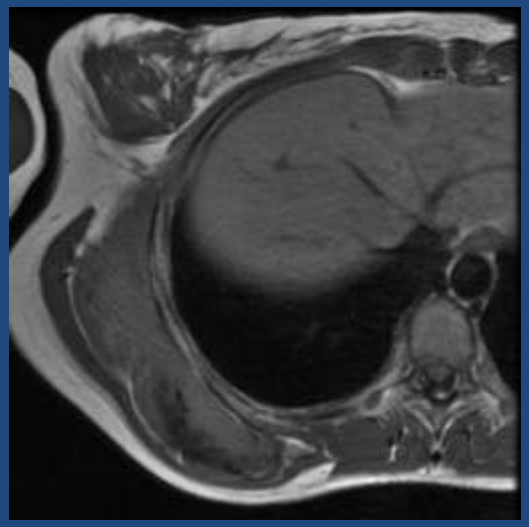
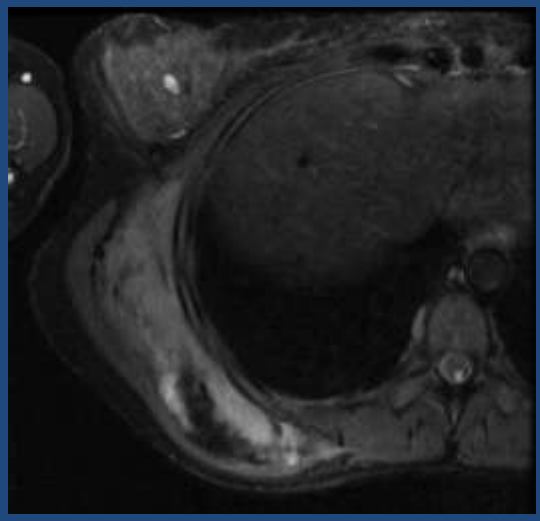
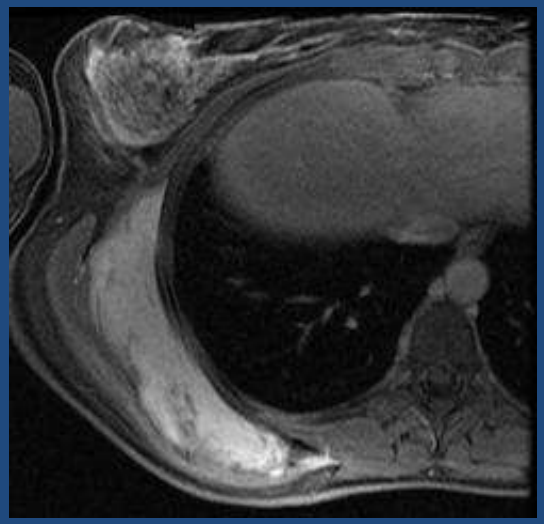
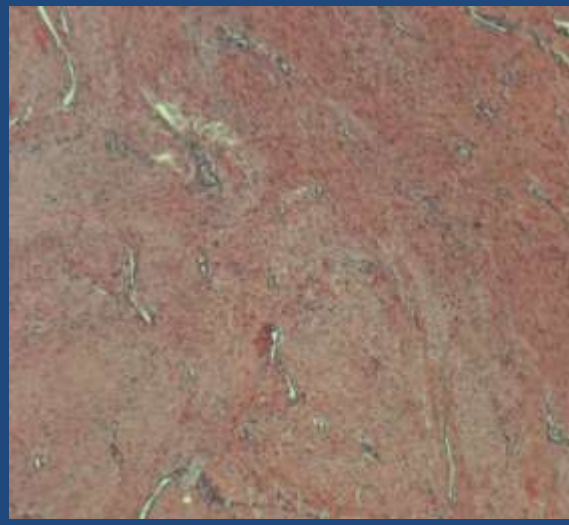










Lipomatous Tumors

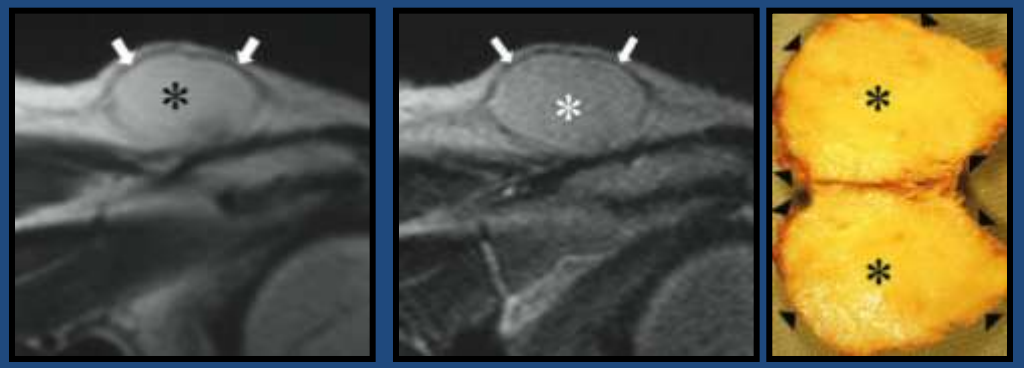
Lipomatous Tumors

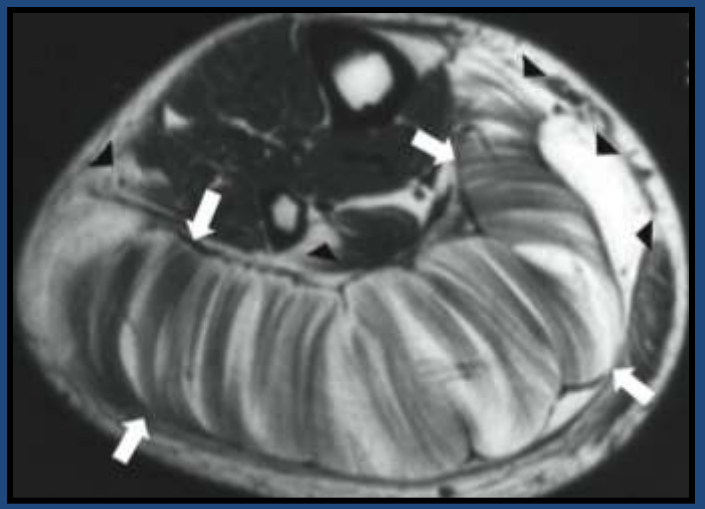
Benign	Malignant

• Lipoma
• Lipomatosis
• Lipomatosis of nerve
• Lipoblastoma
• Angiolipoma
• Spindle cell/Pleomorphic lipoma
• Myolipoma
• Chondroid lipoma
• Hibernoma
	• Liposarcoma
	  • Well-differentiated
	  • Dedifferentiated
	  • Myxoid
	  • Pleomorphic
	  • Mixed-type

Lipoma

- Most common soft tissue tumor (50%)
- Benign neoplasm vs. local hyperplasia of fat cells
- Superficial
  - Upper back, neck, proximal extremities, abdomen
  - < 5 cm
- Deep
  - Intra vs. Intermuscular (arbitrary) (if both = *infiltratng*)

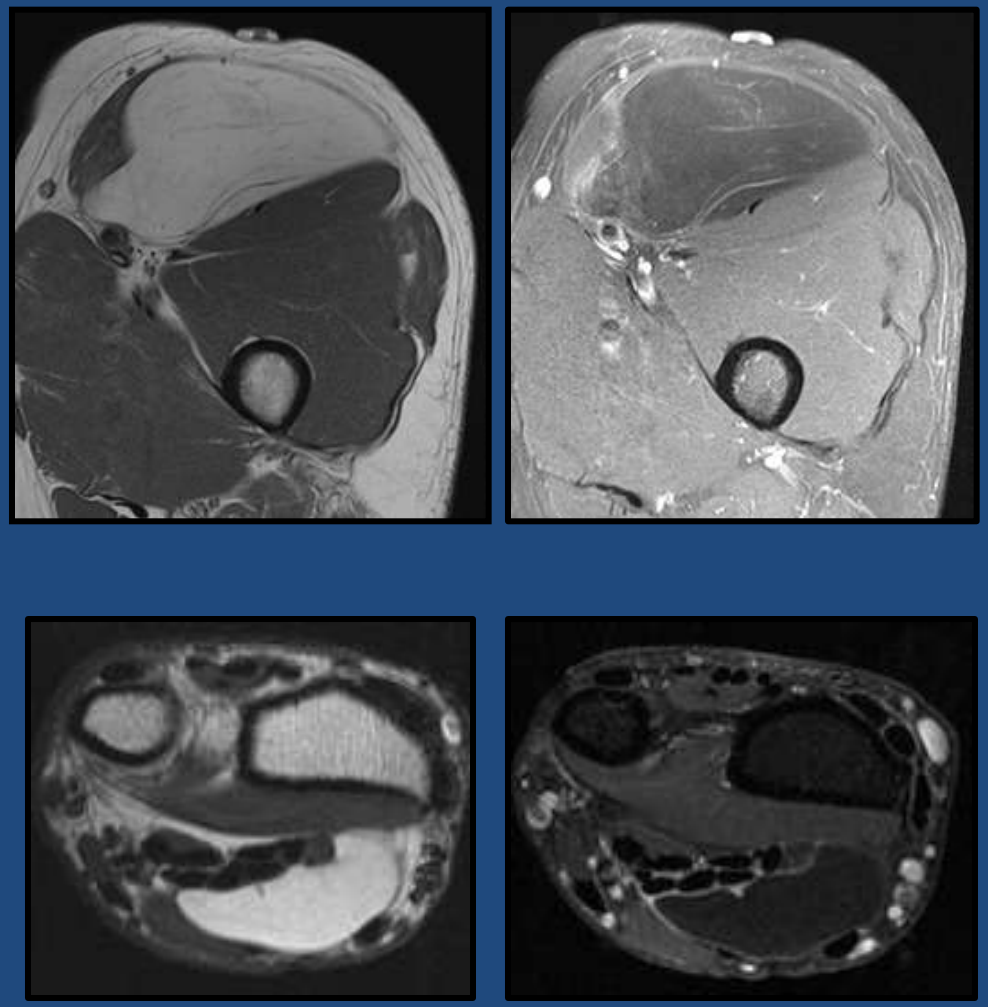




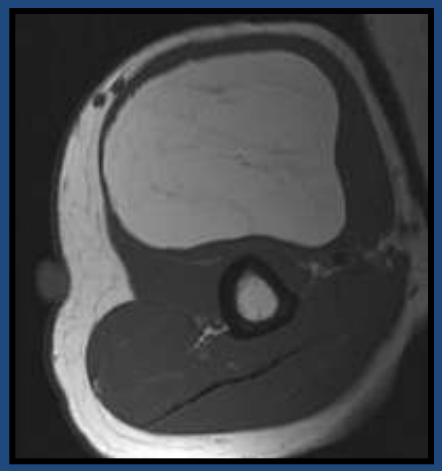
Murphey MD, et al. *Benign Musculoskeletal Lipomatous Lesions*. Radiographics 2004; 24: 1433-1466

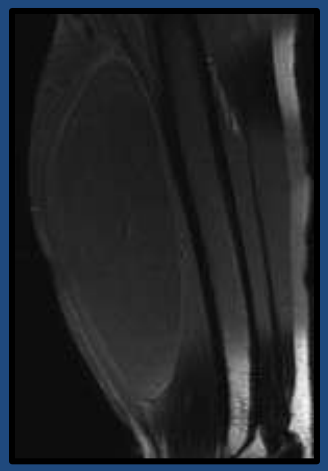
Lipoma

- Multiple (5-15%)
- Thin, non enhancing septa (< 2 mm)
- No capsule with intramuscular and some subcutaneous lipomas
- Intramuscular lipomas have irregular margins, striated
- No malignant transformation



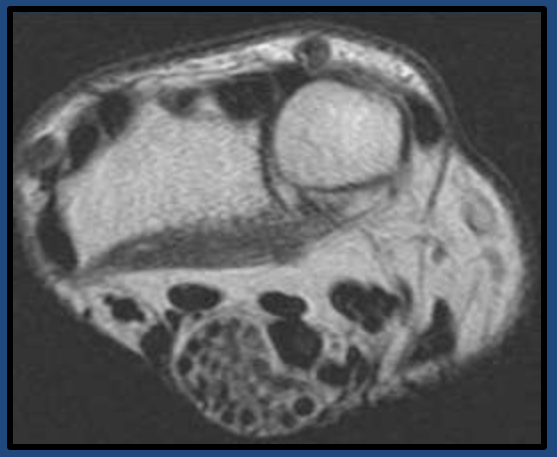
Lipoma

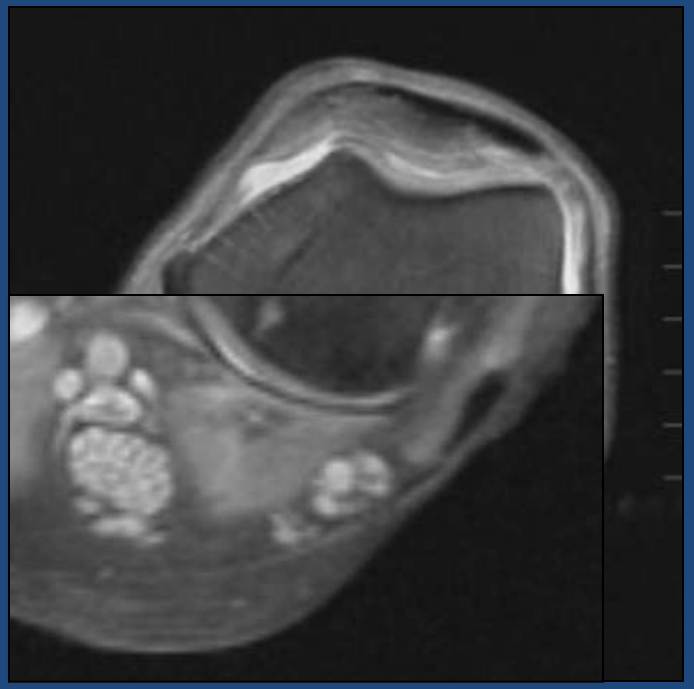




Lipomatosis of Nerve
(Fibrolipomatous Hamartoma)

- < 30 years old
- Median nerve (85%)
- Macrodactyly (27-67%) (Macrodystrophia lipomatosa)
- *Lipomatosis of the nerve with or without macrodactyly*

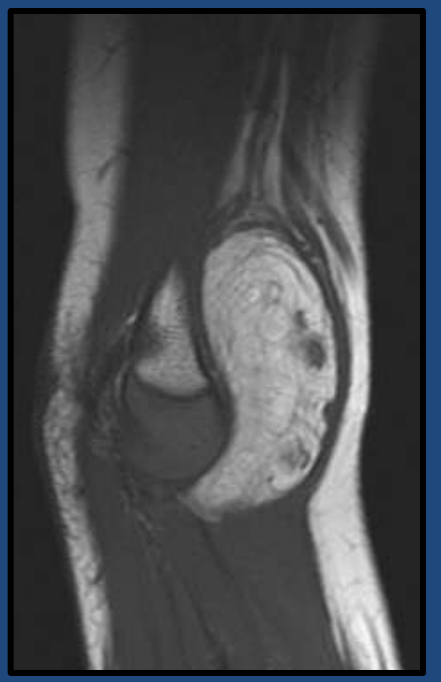


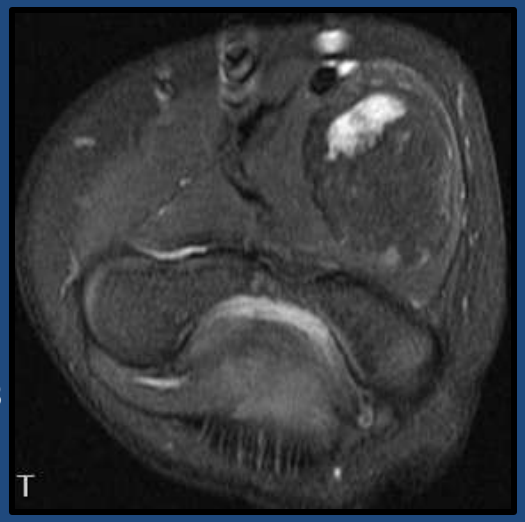


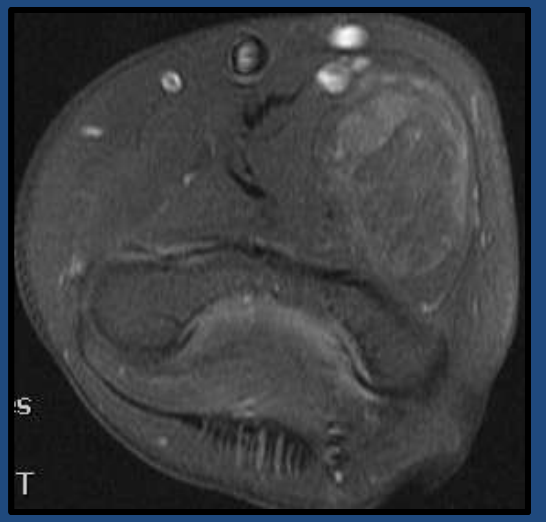
Courtesy Tudor Hughes, M.D.

Lipoblastoma

- < 3 years old
- Superficial, extremities
- Progress to mature lipomas
- Imaging appearance can be similary to myxoid liposarcoma (rare < 10 yrs old)







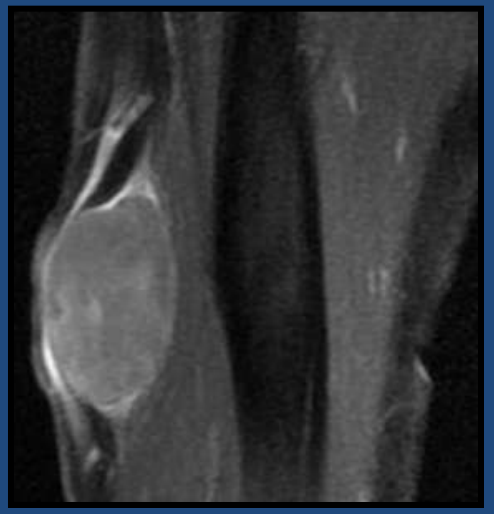
Bancroft LW, et al. Skeletal Radiology 2006; 36: 719-733

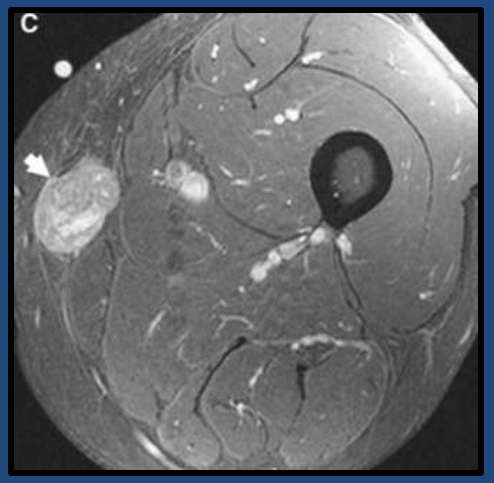
Soft Tissue Sarcomas

Soft Tissue Sarcomas

- 75% arise in extremities
- Usually develop de novo (not from dedifferentiation of benign tumor)
- Hematogenous metastasis (lungs)

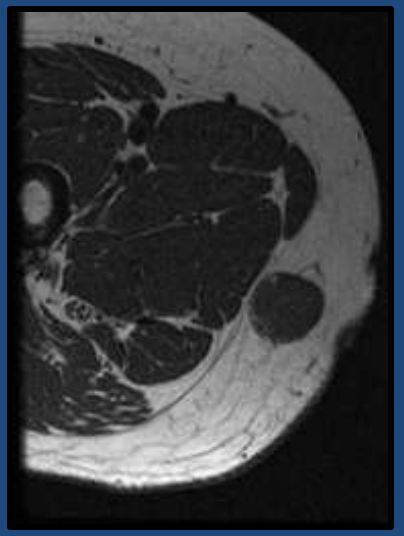
- > 50 subtypes (75% are the following):
  - Undifferentiated Pleomorphic Sarcoma (MFH)
  - Liposarcoma
  - Leiomyosarcoma
  - Synovial Sarcoma
  - Malignant Peripheral Nerve Sheath Tumor

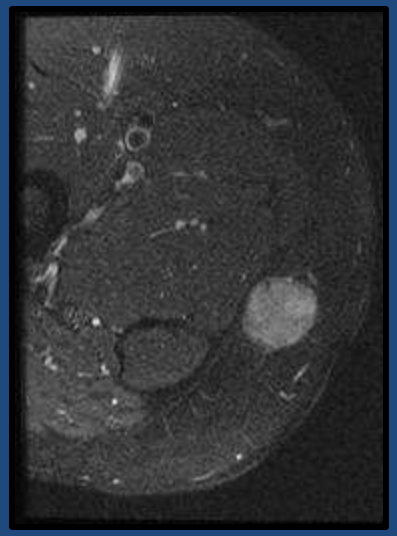


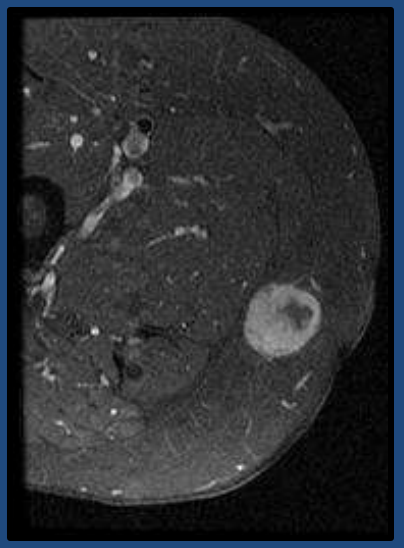


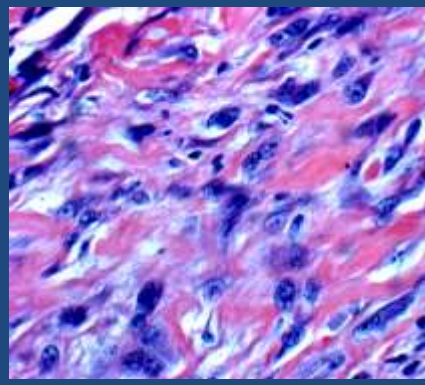
Undifferentiated Pleomorphic Sarcoma (MFH)

- Histologic diagnosis of exclusion
- Non specific MR features
- Peripheral enhancement common (necrosis, hemorrhage, or myxoid content)

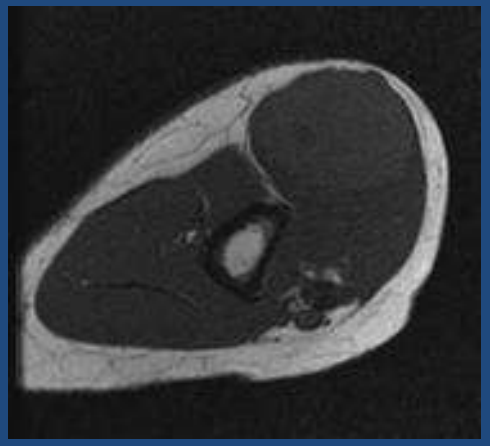


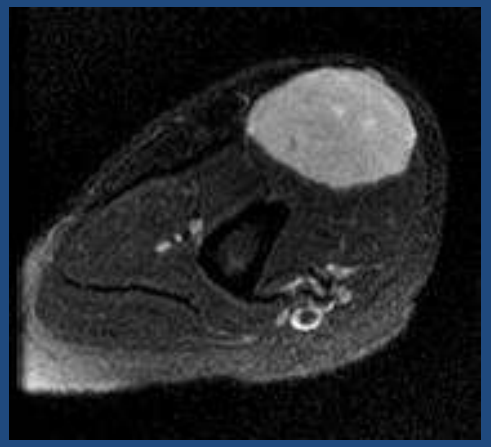


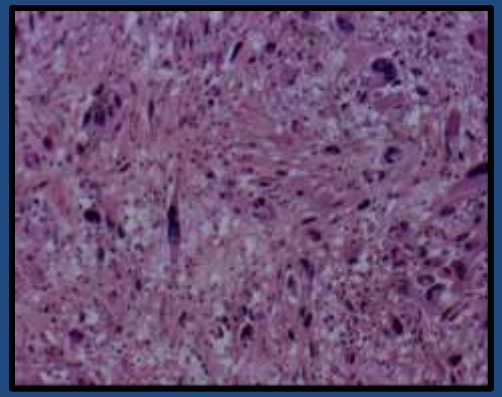




Undifferentiated Pleomorphic Sarcoma (MFH)





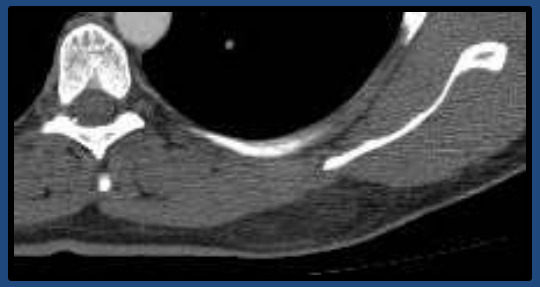


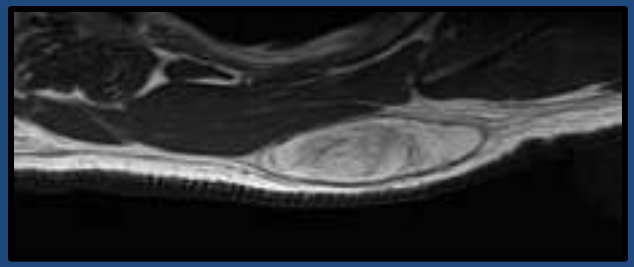
Liposarcoma

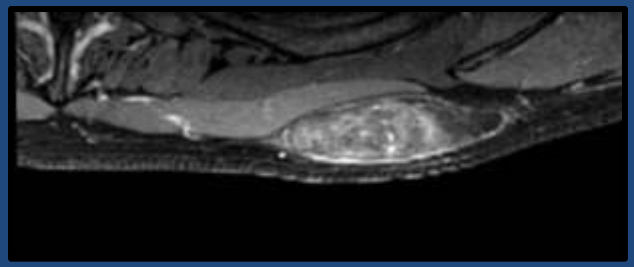
- Second most common type of soft tissue sarcoma
- Five histologic subtypes:
  - Well-differentiated
  - Dedifferentiated
  - Myxoid
  - Pleomorphic
  - Mixed-type

Well-Differentiated Liposarcoma

- Most common subtype (50%)
- Deep soft tissues of extremities (65-75%), retroperitoneum (20-33%)
- No metastatic potential
- *Atypical lipomatous lesion* reserve for subcutaneous lesions



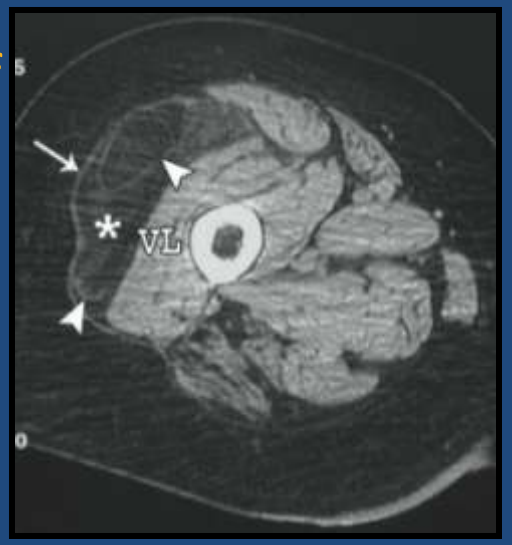


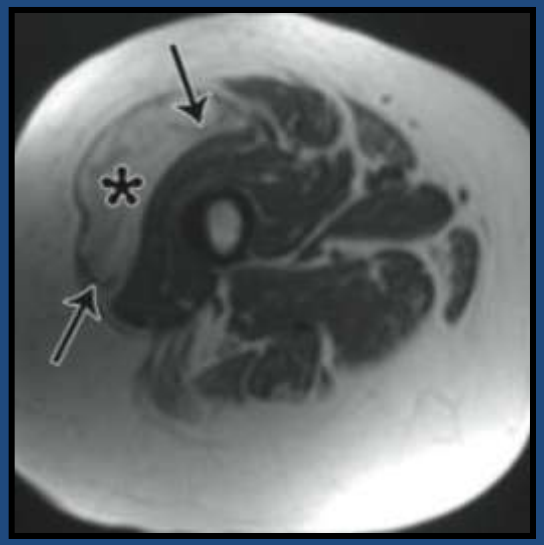


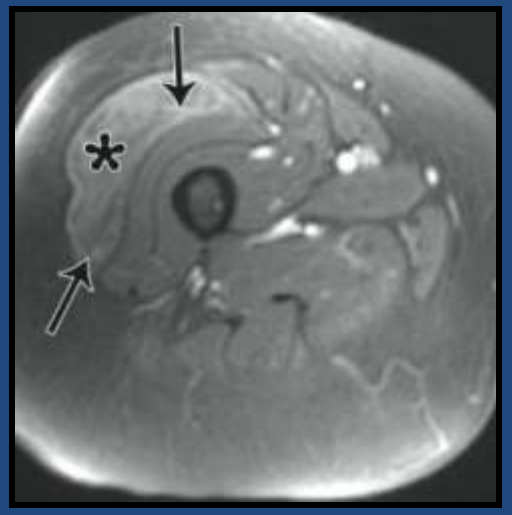
Murphey MD, et al. *Imaging of Musculoskeletal Liposarcoma with Radiologic-Pathologic Correlation.* Radiographics 2005; 25: 1371-1395

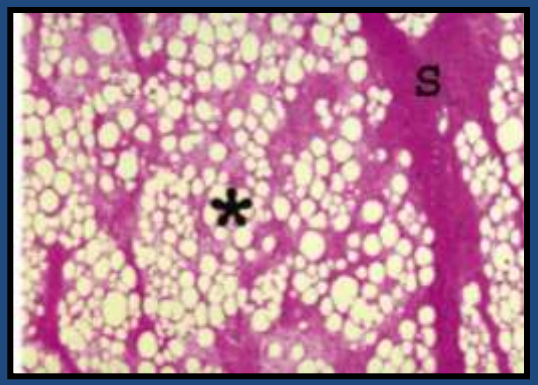
Well-Differentiated Liposarcoma

- > 75% of the lesion composed of fat
- Thick, enhancing septa (> 2mm)
- Liposarcoma > lipoma:
  - Male
  - > 66 years old
  - < 75% fatty
  - Calcifications
  - Size > 10 cm
  - Septa > 2 mm
  - Nonlipomatous nodular or globular foci





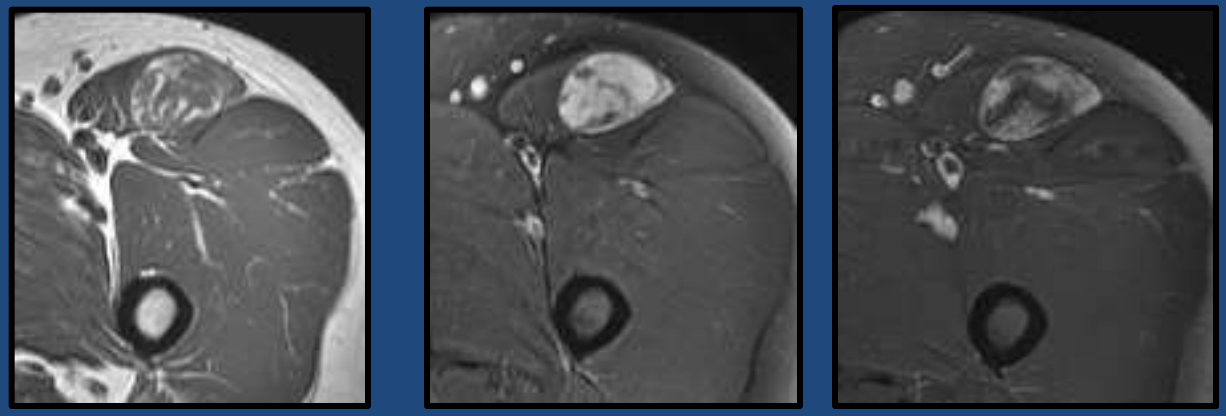




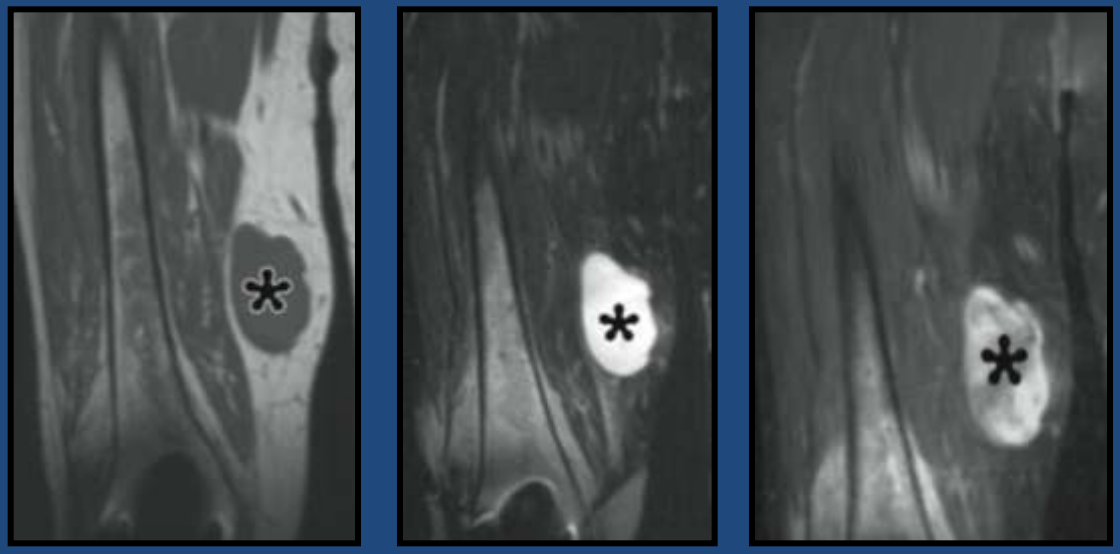
Kransdorf MJ, et al. Radiology 2002; 224:99-104

Myxoid Liposarcoma

- Second most common subtype
- Younger pt (4th-5th decade)
- *Inter*muscular, lower extremity

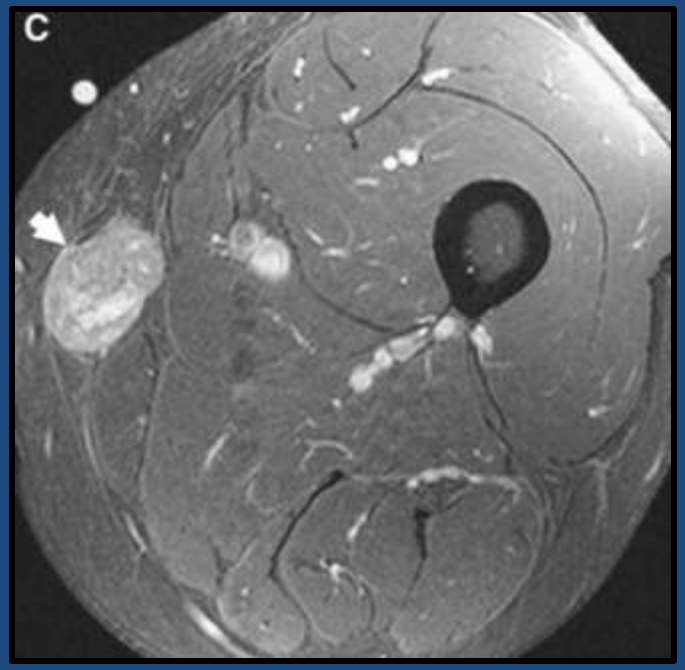


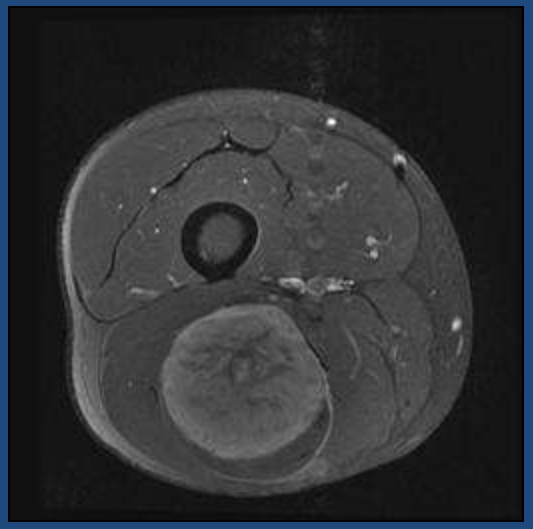
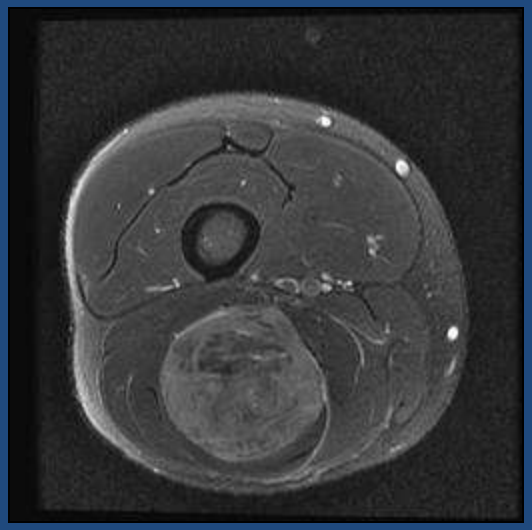
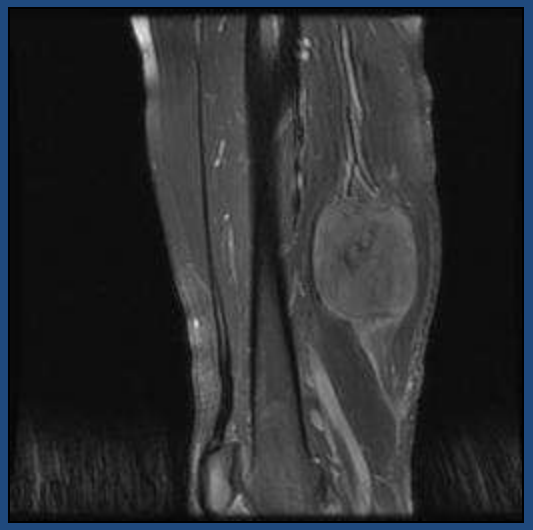
- Pathognomonic MR:
  - Fatty septa or nodules in a myxoid mass
  - May simulate a cyst (unusual location) or myxoma (*intra*muscular)



Leiomyosarcoma

- Smooth muscle
- Intermuscular and subcutaneous, rarely in association with a vessel (vein)
- Irregular rim enhancement
- Ca++ uncommon

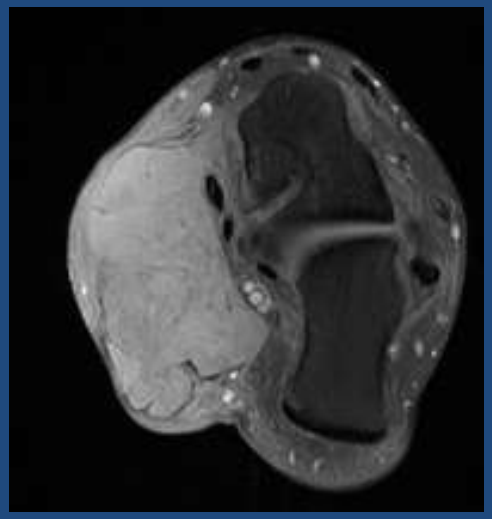


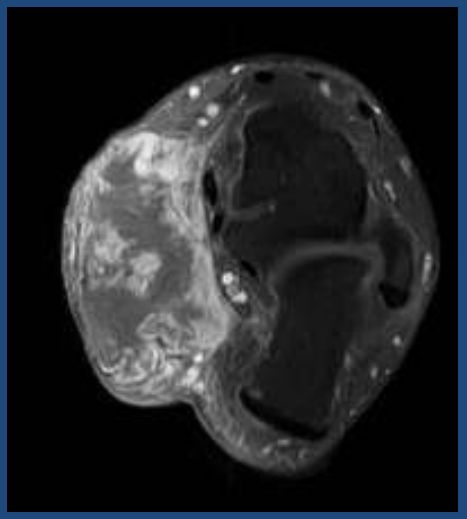





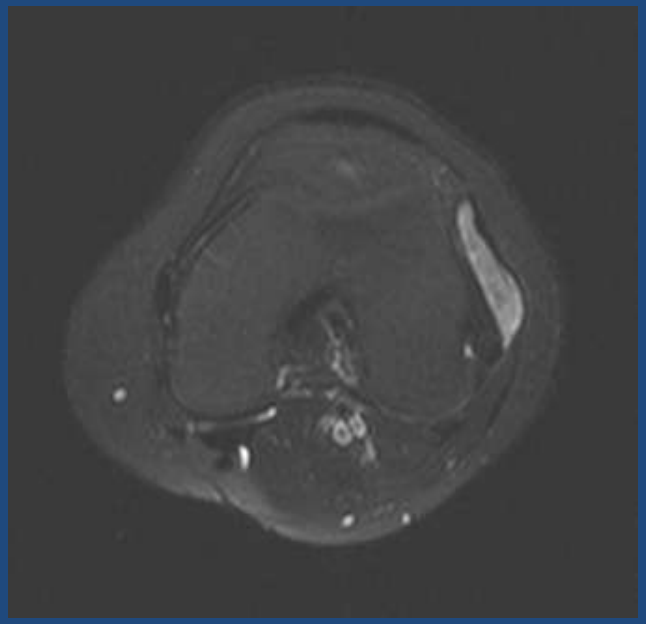

Courtesy Tudor Hughes, M.D.

Synovial Sarcoma

- 2nd-4th decade
- Deep soft tissues of extremities and adjacent to joints or tendon sheaths (popliteal fossa)
- Triple T2 signal (relative to fat)
- Heterogeneous signal and variable contrast enhancement
- Ca++ (33%)
- Fluid-fluid levels
- Bone erosion (20%)

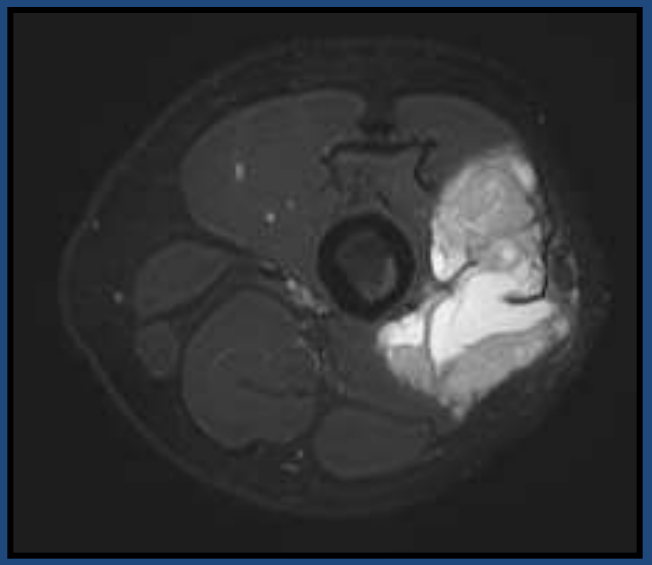






Courtesy of Tudor Hughes, M.D.

Synovial Sarcoma



Summary

I. Soft Tissue Anatomy
   - Compartmental

I. Imaging Work-Up
   - Post-Treatment Imaging

II. Soft Tissue Tumors—MR Features
   - WHO Classification

Thank You!

References

1. Anderson MW, Temple HT, Dussault RG, Kaplan PA. *Compartmental Anatomy: Relevance to Staging and Biopsy of Musculoskeletal Tumors*. AJR 1999; 173: 1663-1671.
2. Kransdorf MJ, Murphey MD. *Radiologic Evaluation of Soft Tissue Masses: A Current Perspective*. AJR 2000; 175: 575-587.
3. Kransdorf MJ, Murphey MD. Soft Tissue Tumors: Post-Treatment Imaging. Radiologic Clinics of North America 2006; 44:463-472.
4. Garner HW, Kransdorf MJ, Bancroft LW, Peterson JJ, Berquist TH, Murphey MD. Benign and Malignant Soft Tissue Tumors: Post Treatment MR Imaging. Radiographics 2009; 29:119-134.
5. Van Herendael BH, et al. *The Value of Magnetic Resonance Imaging in the Differentiation Between Malignant Peripheral Nerve Sheath Tumors and Non-Neurogenic Malignant Soft Tissue Tumors.* Skeletal Radiology 2006; 35: 745-753.
6. Bredella MA, et al. *Value of PET in the Assessment of Patients with Neurofibromatosis Type 1.* AJR 2007; 189: 928-935.
7. Vilanova JC, et al. *Soft tissue Tumors Update: MR Imaging Features According to the WHO Classification*. European Radiology 2007; 17: 125-138.
8. Blacksin MF, et al. *Superificial Soft Tissue Masses of the Extremeties.*Radiographics 2006; 26:1289-1304.
9. Van Vliet M, et al. *Soft Tissue Sarcomas at a Glance: Clinicial, Histological, and MR Imaging Features of Malignant Extremity Soft Tissue Tumors.* European Radiology 2009; DOI 10.1007.
10. Fayad L, et al. *Vascular Malformations in the Extremeties: Emphasis on MR Imaging Features that Guide Treatment Options*. Skeletal Radiology 2006; 35:127-137.

References

11. Olson KI, et al. *Soft Tissue Cavernous Hemangioma*. Radiographics 2004; 24: 849-854.
12. Dinauer PA, et al. *Pathologic and MR Imgaing Features of Benign Fibrous Soft Tissue Tumors in Adul*ts. Radiographics 2007; 27: 173-187.
13. Bancroft LW, Kransdorf MJ, Peterson JJ, O'Connor MI. *Benign Fatty Tumors: Classification, Clinical Course, Imaging Appearance, and Treatment*. Skeletal Radiology 2006; 36: 719-733.
14. Murphey MD, et al. *Benign Musculoskeletal Lipomatous Lesions*. Radiographics 2004; 24: 1433-1466.
15. Murphey MD, et al. *Imaging of Musculoskeletal Liposarcoma with Radiologic-Pathologic Correlation.* Radiographics 2005; 25: 1371-1395.
16. Bancroft LW, Peterson JJ, Kransdorf MJ. *MR Imaging of Tumors and Tumor-Like Lesions of the Hip*. Magnetic Resonance Imaging Clinics of North America 2005; 13:757-774.
17. Stacy GS, et al. *Pitfalls in MR Image Interpretation Prompting Referrals to an Orthopedic Oncology Clinic*. Radiographics 2007; 27:805-828
18. Kotilingam D, et al. *Staging Soft Tissue Sarcoma: Evolution and Change*. CA Cancer Journal for Clinicians 2006; 56: 282-291.
19. Clark MA, et al. *Soft Tissue Sarcomas in Adults*. The New England Journal of Medicine 2005; 353:701-711.
20. Bancroft LW, et al. *Soft Tissue Tumors of the Lower Extremities*. Radiologic Clinics of North America 2002; 40: 991-1011.
21. Beaman FD, et al. *Superficial Soft Tissue Masses: Analysis, Diagnosis, and Differential Considerations*. Radiographics 2007; 27: 509-523.