

Benign Soft Tissue Tumors

Lipoma, Hemangioma, Fibromatosis
Myxoma, Schwannoma, GCT of Tendon Sheath
Pigmented Villonodular Synovitis (PVNS)

James C. Wittig, MD
Orthopedic Oncologist
Sarcoma Surgeon
www.TumorSurgery.org

Lipoma

- **Definition:** Benign tumor composed of mature adipocytes with uniform nuclei identical to the cells of normal adult fat.
- Benign/Mature adipocytes have cytoplasmic lipid in a single large vacuole that pushes the nucleus to the periphery of the cell and compresses it into a thin crescent.
- **Lipoblasts:** Malignant or Immature adipocytes.
 - Larger peripheral or central nucleus indented by one or more fat vacuoles so that spikes of chromatin project between fatty vacuoles. Usually smaller and have smaller vacuoles than mature lipocytes

Lipoma

- Types of Lipomas:
 - Lipoma
 - Fibrolipoma
 - Angiolipoma
 - Spindle Cell/Pleomorphic Lipoma
 - Myxolipoma
 - Lipoblastoma
 - Myolipoma
 - Hibernoma
 - Chondroid Lipoma

Lipoma

- Most common soft tissue tumor
- Adults
- Location:
 - Subcutaneous: Most Common
 - Back, shoulder, neck, abdomen, proximal extremities
 - Uncommon in hands, feet, distal extremities
 - Rarely recur after excision
 - No sex predilection: Males=Females
 - Deep
 - Intramuscular: develops within skeletal muscle
 - Usually affects adults but occasionally found in children
 - Trunk, large muscles of arm, thigh, shoulder
 - Males affected greater than females
 - Up to 20% recurrence after excision especially if infiltrative



Lipoma

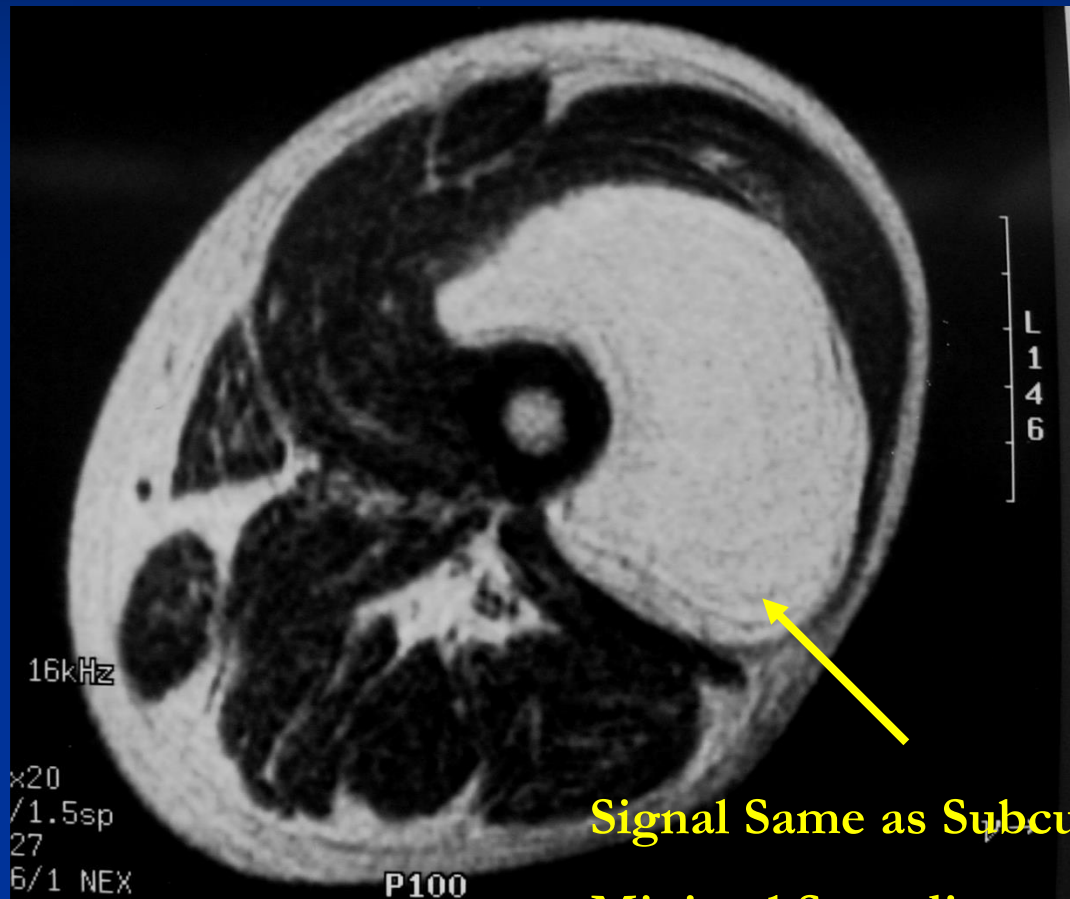
- Clinical Presentation:
 - Slowly growing, asymptomatic, painless mass
 - Some patients may have multiple lipomas

Radiology: Lipoma

- Radiology: Follows that of subcutaneous fat
- Xray: Radiolucent soft tissue mass; Calcification may be present particularly in areas of fatty necrosis
- CT Scan: Well defined, homogeneous
 - **No enhancement following contrast administration**
 - Tissue Attenuation Coefficient is Low (-65 to -120 HU)
- MRI: Follows the signal characteristics of subcutaneous fat on all sequences
 - T1: High Signal
 - T2: Intermediate Signal; Low Signal with Fat Suppression
 - Minimal or No enhancement with gadolinium

MRI T1: Intramuscular Lipoma of Thigh

High Signal on T1



Signal Same as Subcutaneous Fat

Minimal Stranding within Tumor



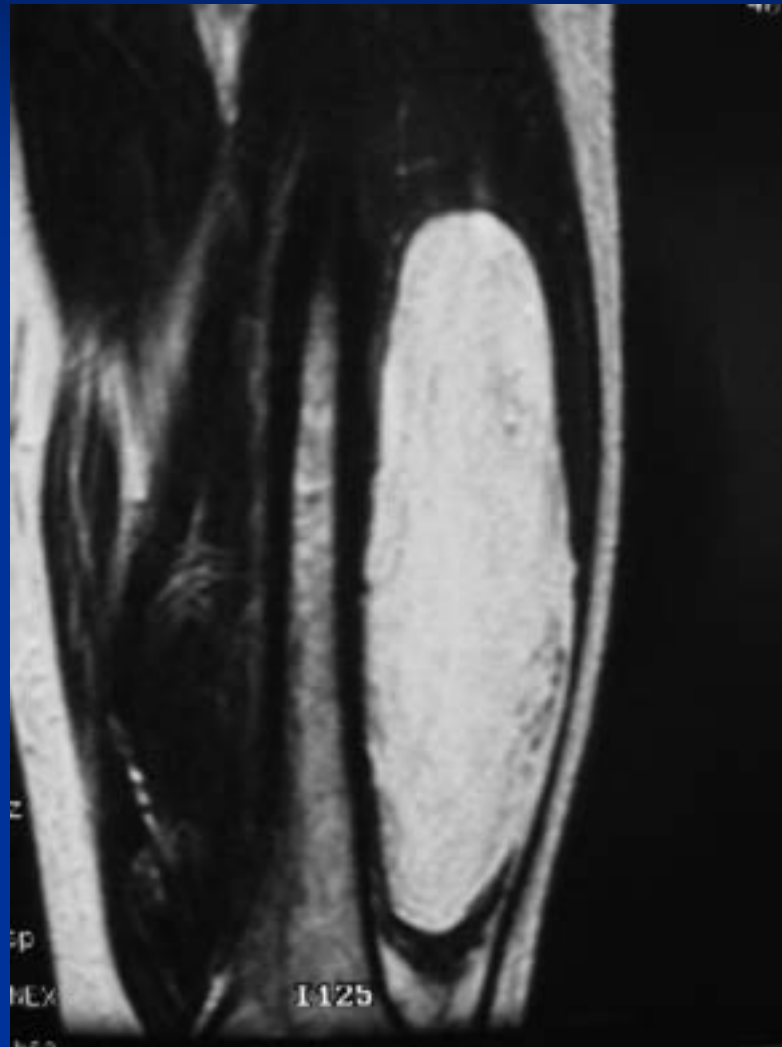
MRI T2 Fat Suppressed Image



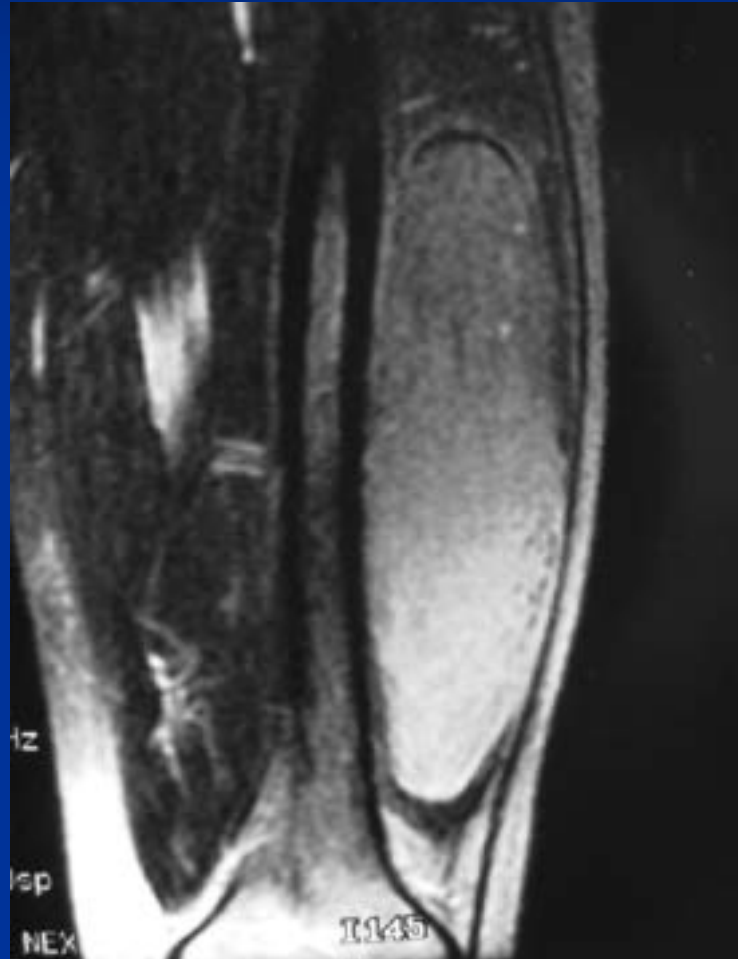
Fat in Lesion Suppressed Same as Subcutaneous Fat



MRI T1: Lipoma of Posterior Thigh



MRI T2 (not Fat Suppressed)
Fat is Intermediate Signal on T2
Low Signal on Fat Suppressed



MRI T1: Lipoma Right Shoulder

Fibrolipoma: Notice Stranding



Stranding within Tumor
indicates
Collagenous/Fibrous
Septae

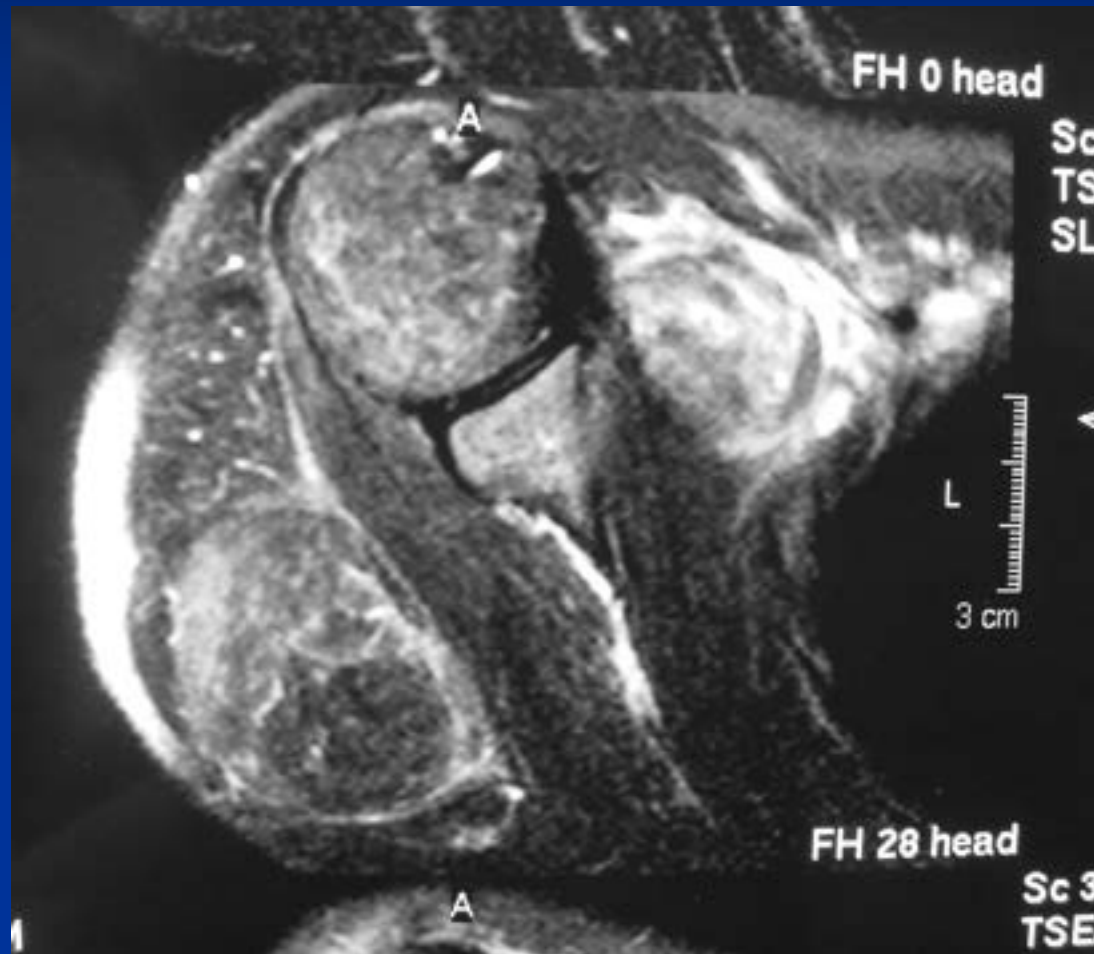
Can Also Occur with Low
Grade
Liposarcomas/Atypical
Lipomas



MRI T1: Lipoma of Shoulder

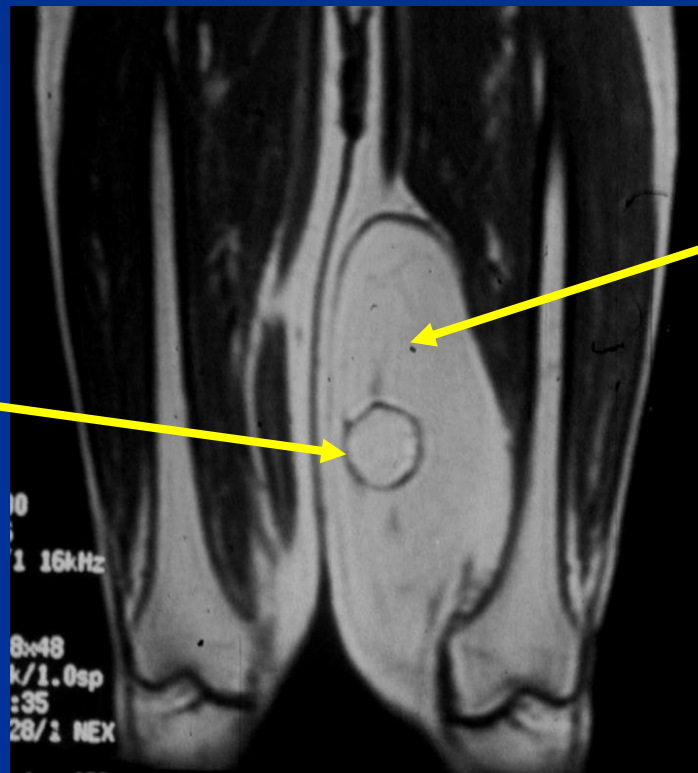


MRI T2 Fat Suppressed



MRI T1: Large Lipoma Left Thigh with Small Area of Fat Necrosis

Area of Fat
Necrosis



Large Lipoma



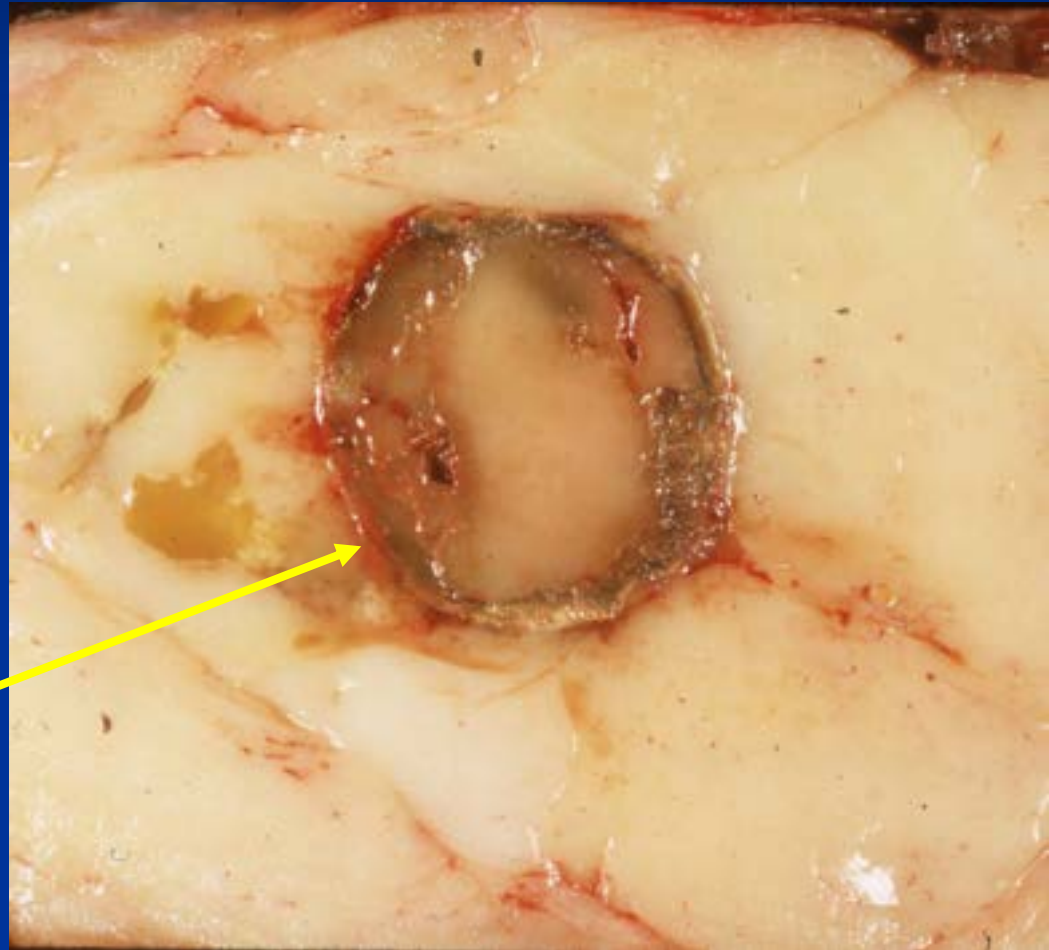
Pathology: Lipoma

- Gross Pathology:
 - Soft yellow fatty mass indistinguishable from normal fat
 - Lobular growth in some lipomas
 - May have fibrous component reflected as white tissue
 - Muscle fibers may be present with intramuscular lipomas



Gross Pathology Lipoma

- Yellow fat
- Area of necrosis (arrow)

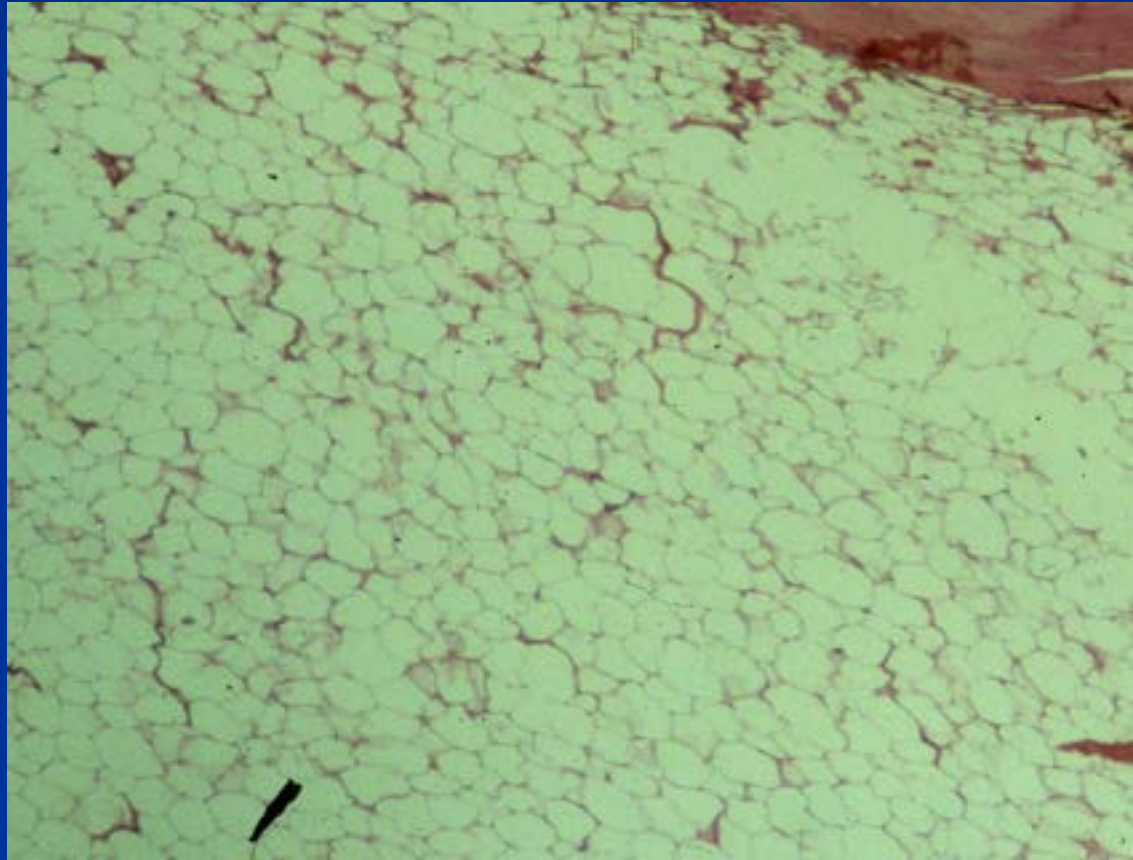


Pathology: Lipoma

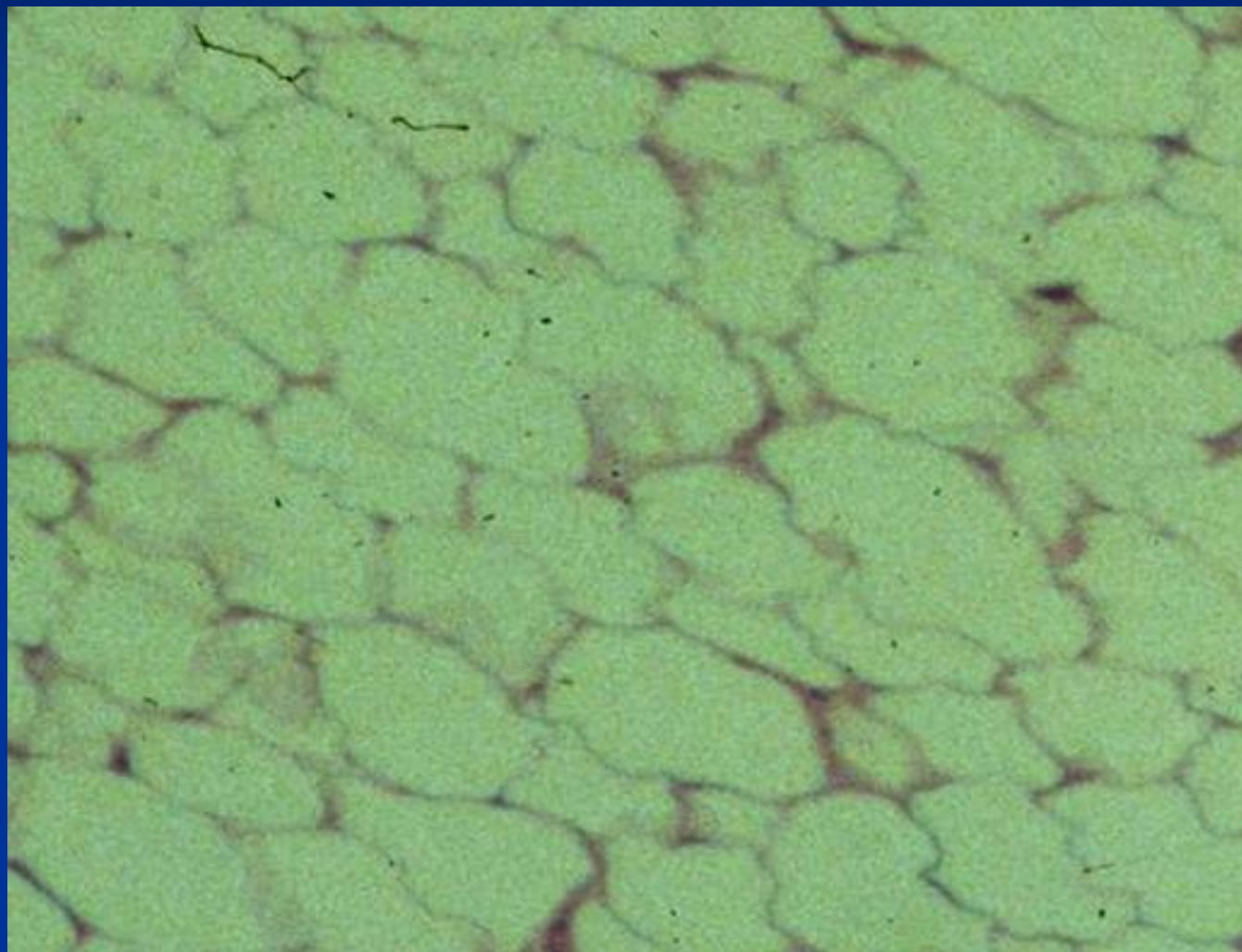
- Microscopic Pathology:
 - Mature fat cells with small, uniform, eccentric nuclei
 - Nucleus compressed against the periphery of the cell membrane by a fat vacuole
 - No mitotic figures
 - Can have areas that undergo fat necrosis
 - Muscle fibers interspersed amongst mature adipocytes (intramuscular lipomas)
 - Other components
 - Fibrous tissue
 - Myxoid tissue
 - Blood vessels

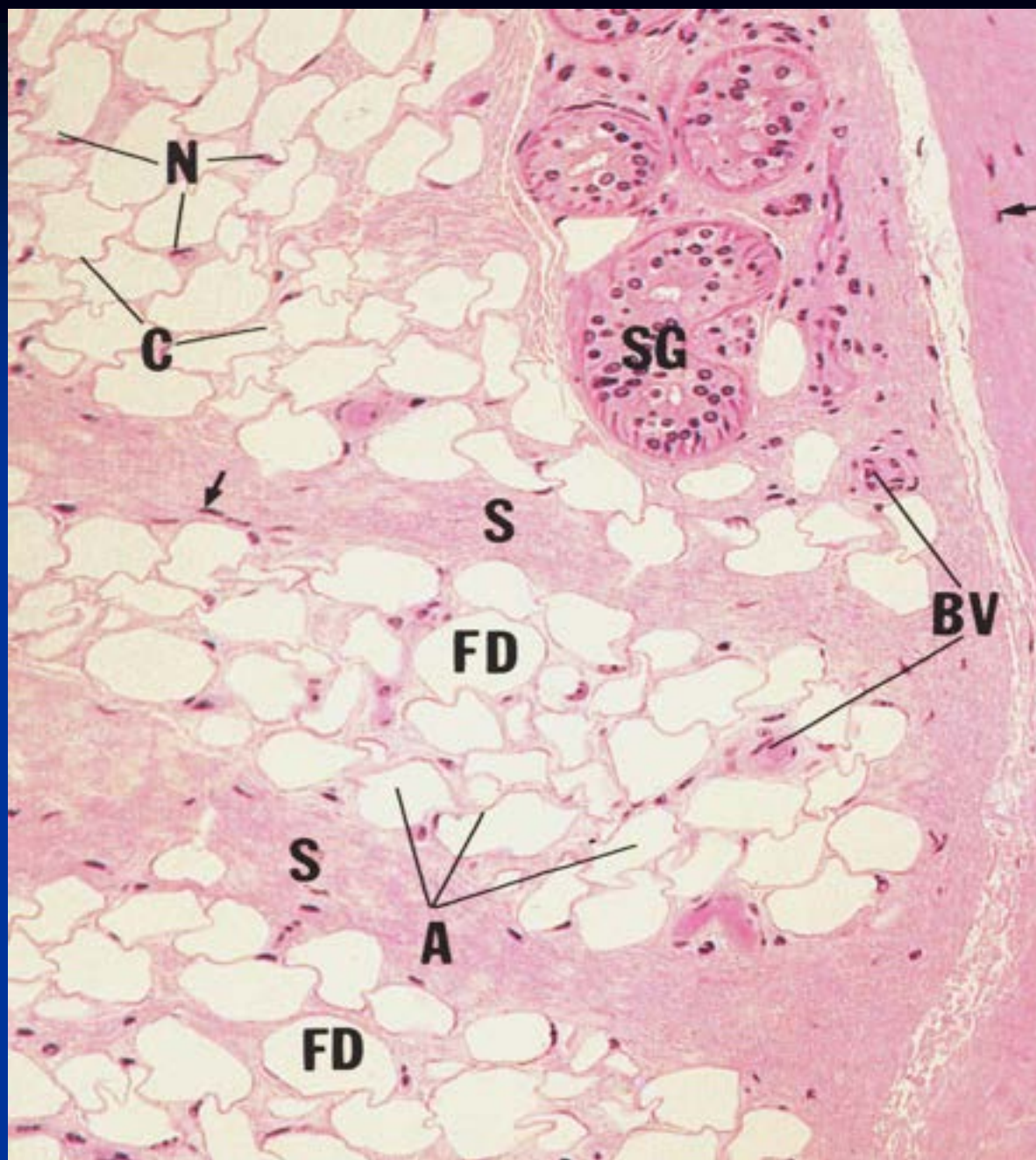
Pathology Microscopic Lipoma

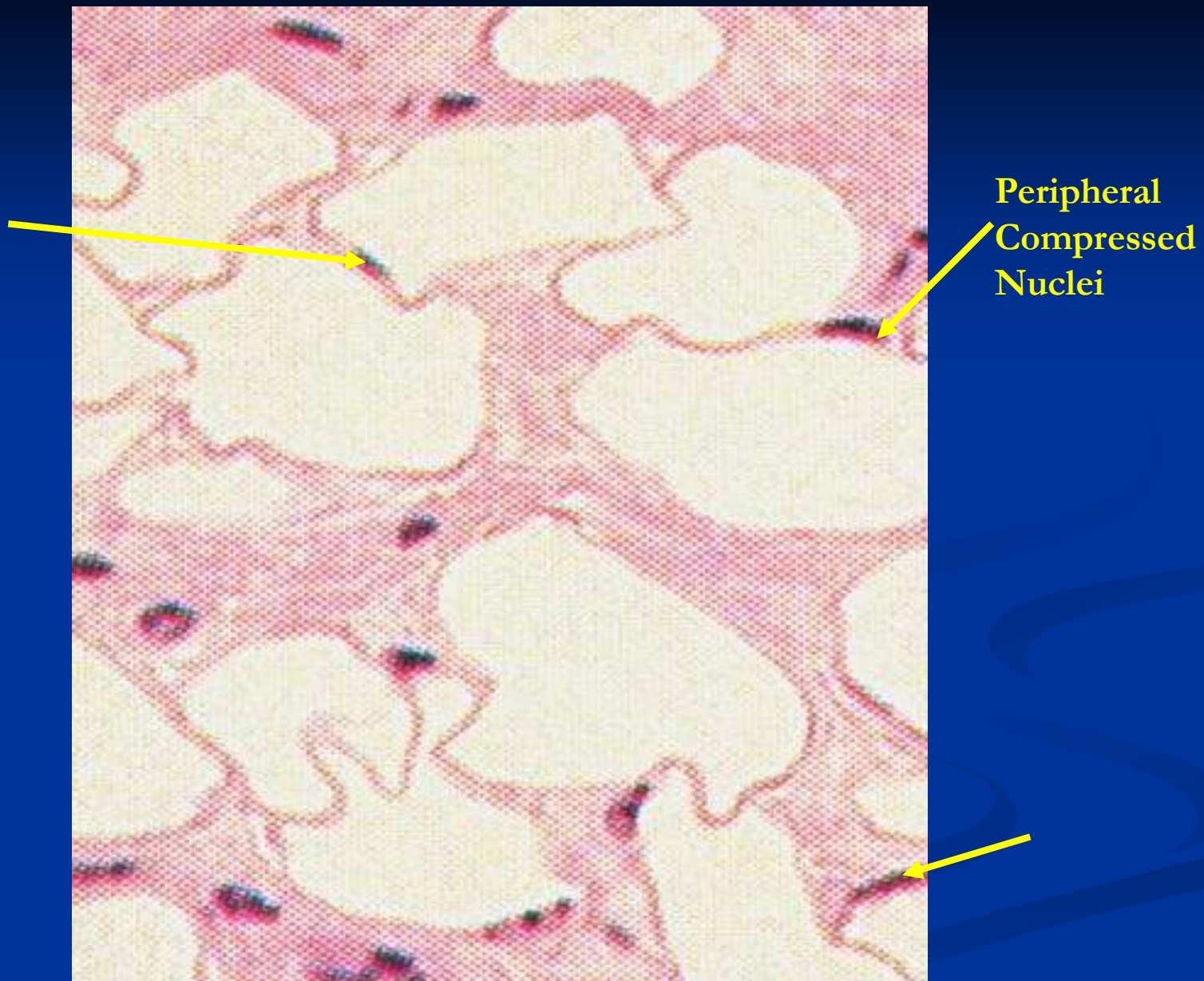
- Uniform Cells
- Peripheral compressed nuclei barely discernible
- Mature adipocytes
- No mitoses

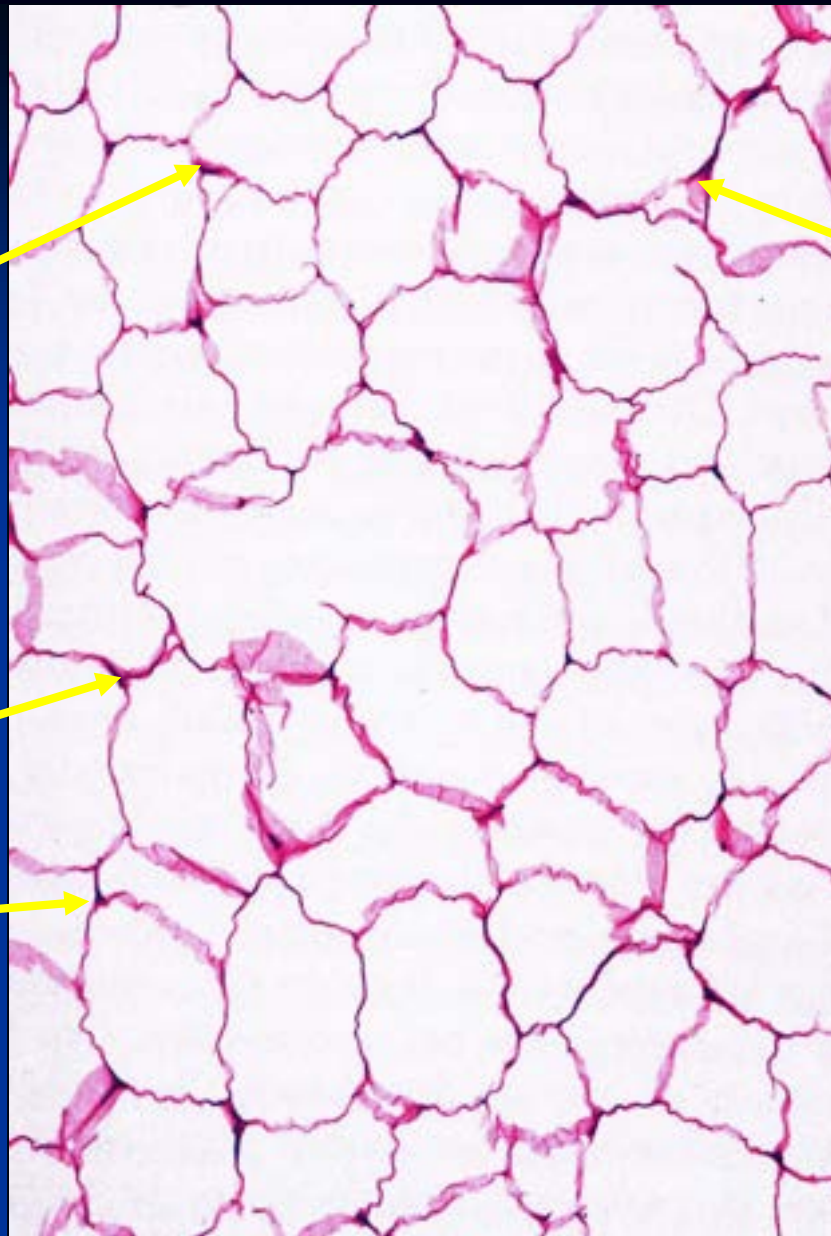


Pathology Microscopic Lipoma









**Compressed
nuclei are
barely
discernible**



Differential Diagnosis

- Lipoma vs atypical lipomatous tumor (well differentiated liposarcoma)
- Both may look similar on MRI
- Well differentiated liposarcoma:
 - Larger nuclei
 - Lipoblasts
 - Considerable variation in fat cell size

Differential Diagnosis

- Lipomas do not occur in the retroperitoneum.
- Retroperitoneal fatty tumors are capable of recurring and dedifferentiating even if they look histologically bland similar to a lipoma
- Retroperitoneal fatty tumors should be considered malignant

Treatment and Prognosis

- Lipomas: Benign and do not metastasize
 - Not treated with any forms of chemotherapy nor radiation.
- Observation for small asymptomatic lipomas
- Surgery: **Marginal** excision for symptomatic, large or deep lesions
- Local recurrence:
 - Risk depends on size and location of lipoma
 - Rarely occurs for subcutaneous tumors
 - Intramuscular tumors--up to 20% local recurrence rate.

Hemangioma

- **Defintion:** Benign proliferation of mature vessels.
 - Composed of capillaries, veins or a combination of both
 - Vessels vary in size and shape.
 - Broad variety of hemangiomas with varying clinical presentations and biological activity.
- Sites:
 - Skin
 - Subcutaneous
 - Intramuscular
 - Intraarticular/Synovial
 - Bone

Intramuscular Hemangioma

- Often larger than cutaneous hemangiomas
- More often symptomatic
- Recur more often than cutaneous hemangiomas

Intramuscular Hemangioma

- Composed of morphologically benign vascular channels occurring within skeletal muscle
- Almost always associated with varying amounts of **fatty tissue**
- Blood vessels/vascular channels are often mixed type of venous and capillary.

Intramuscular Hemangioma

- Adolescent and Adult most frequently affected
- Male=Female
- Sites:
 - Lower Limb most common
 - Head and Neck
 - Upper Limb
 - Trunk
 - Rare: Retroperitoneum and Mediastinum

Intramuscular Hemangioma

- Clinical:
 - Slowly enlarging often longstanding mass
 - **Painful**
 - Pain worse after exercise
 - Changes size according to position of limb

Radiology:

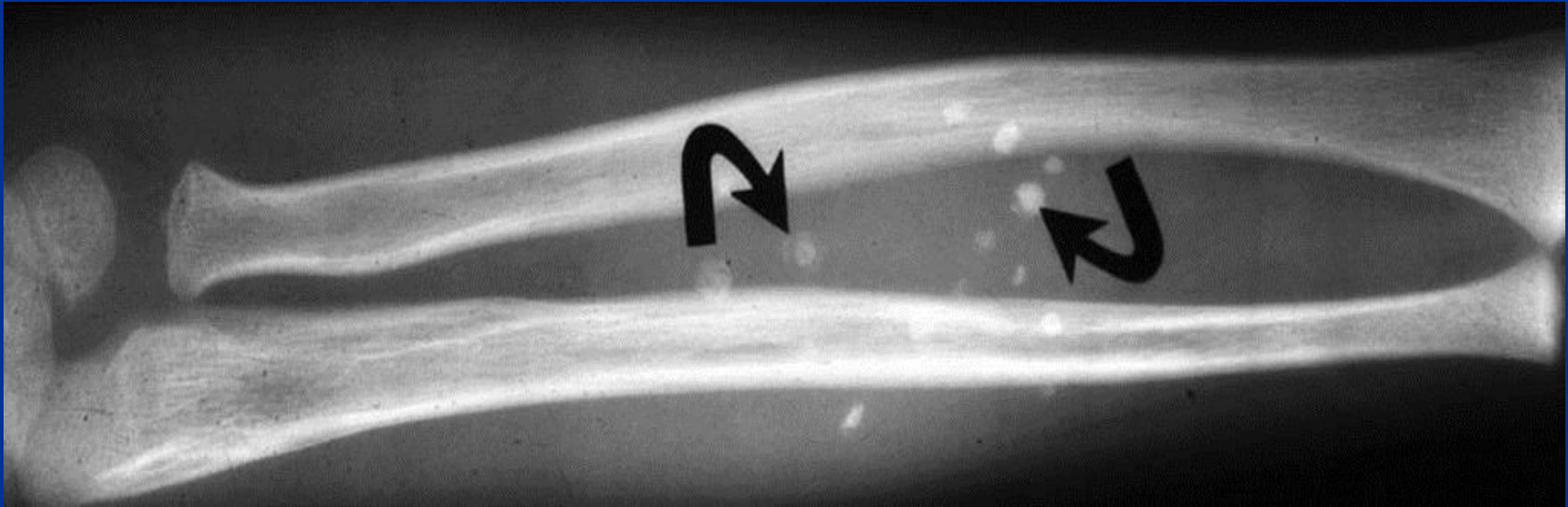
Intramuscular Hemangioma

- Xray: Usually Normal
 - **Phleboliths** or stromal bone formation (30-50%)
 - Pressure erosion of adjacent bone or extension into bone (channel like radiolucencies)
- CT: Poorly defined lesion attenuation similar to muscle
 - Marked contrast enhancement of serpentine vascular channels
 - Subtle phleboliths
 - Lesions adjacent to bone rarely stimulate a subtle periosteal reaction

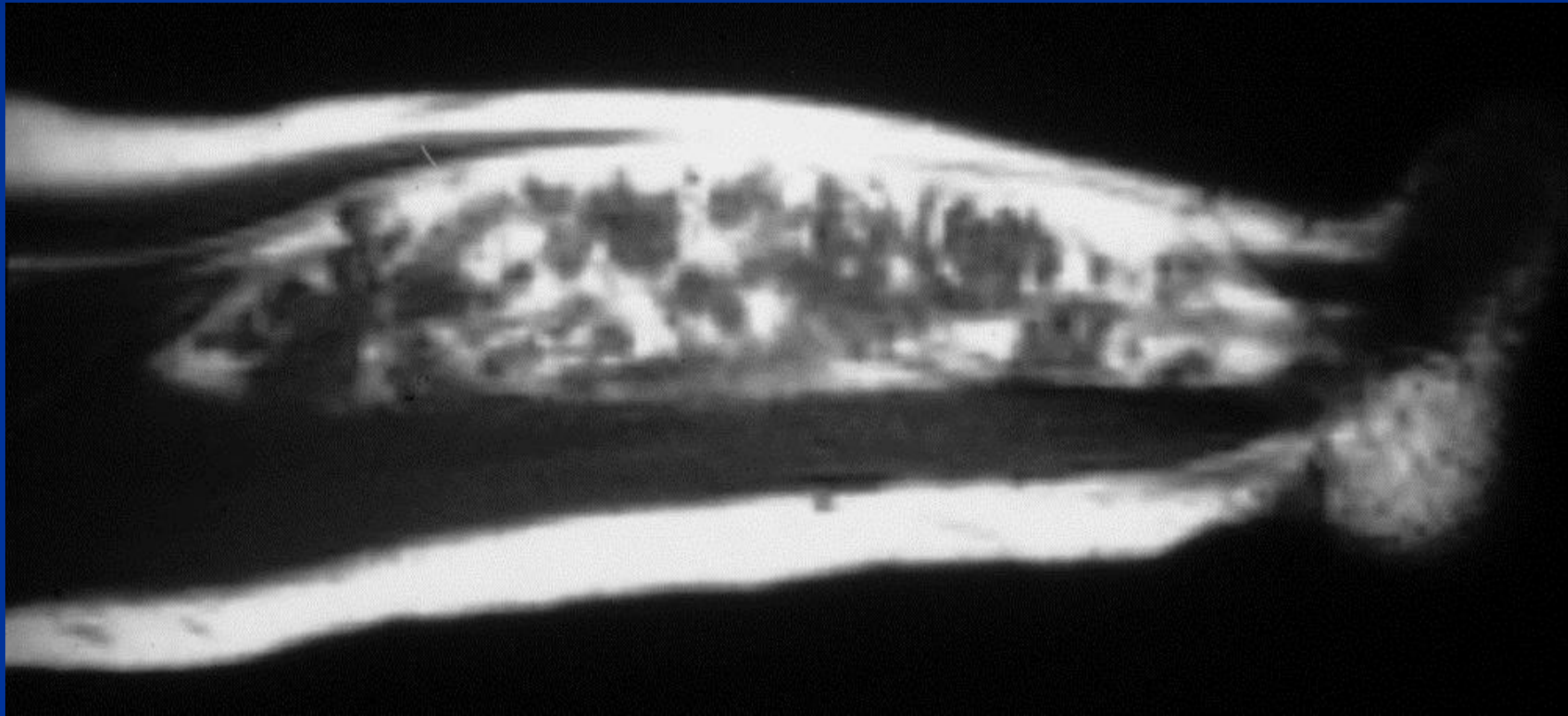
MRI: Hemangioma

- Hetero. poorly marginated mass on T1
- High SI areas on T1 (fat overgrowth)
- Heterogeneous well-defined mass on T2
- Vascular channels high T2 SI (slow flow)
- Hemorrhage/prominent enhancement
- Infiltrate (don't displace or destroy) surrounding structures

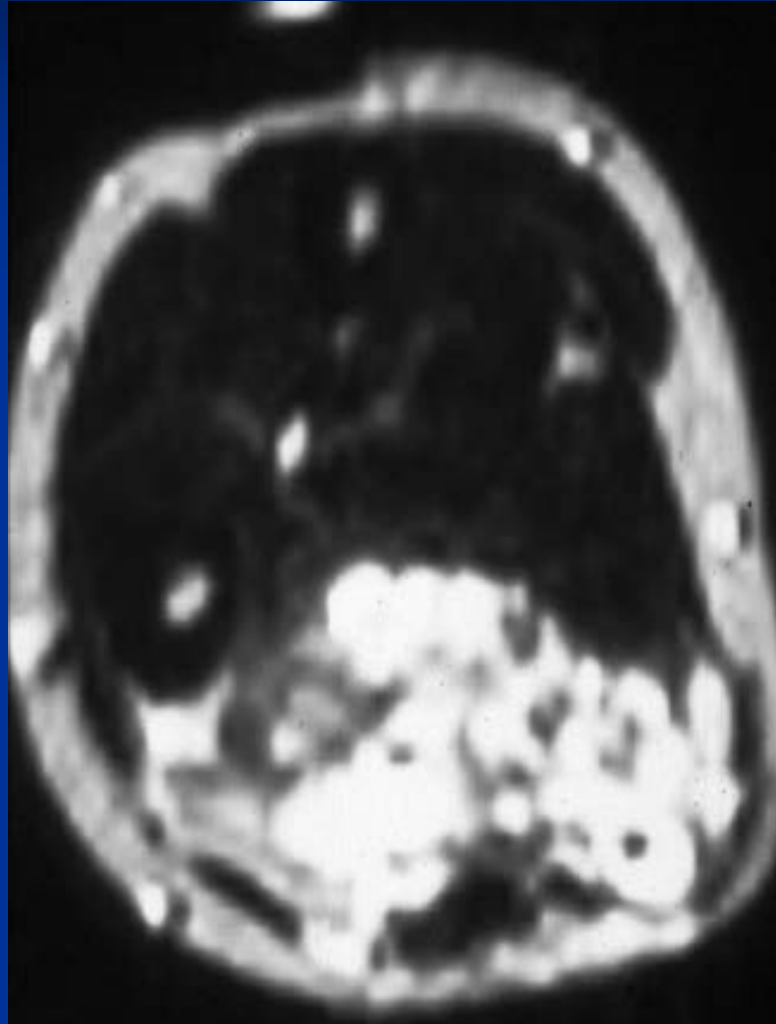
Hemangioma: Phleboliths (arrows)



Hemangioma



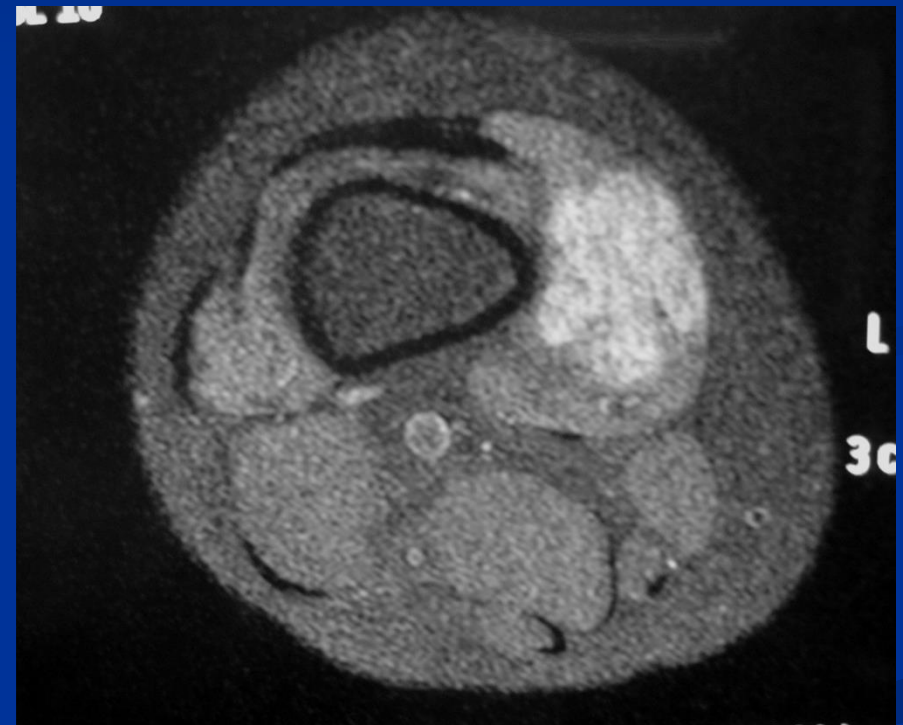
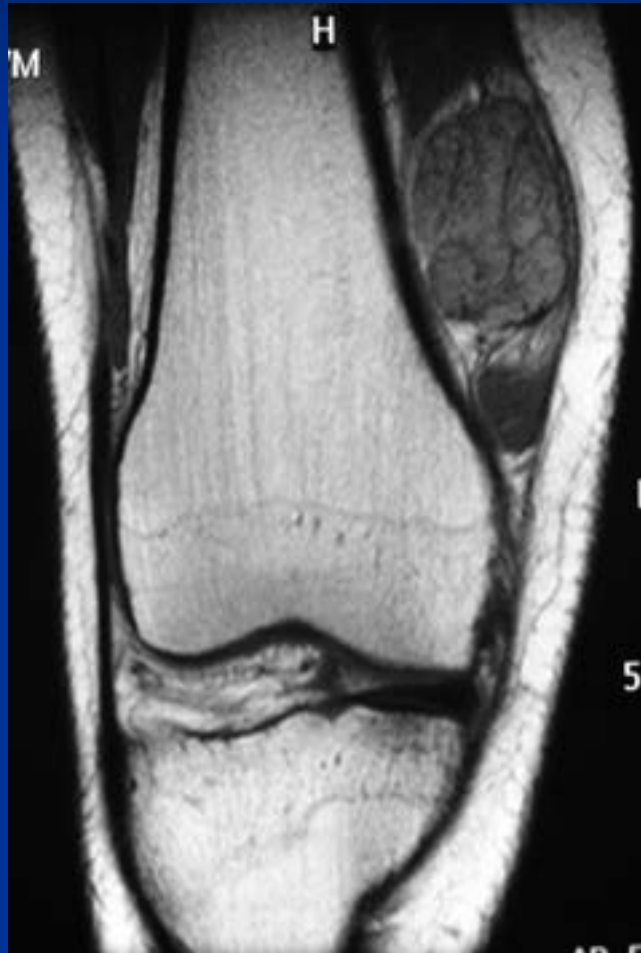
Hemangioma



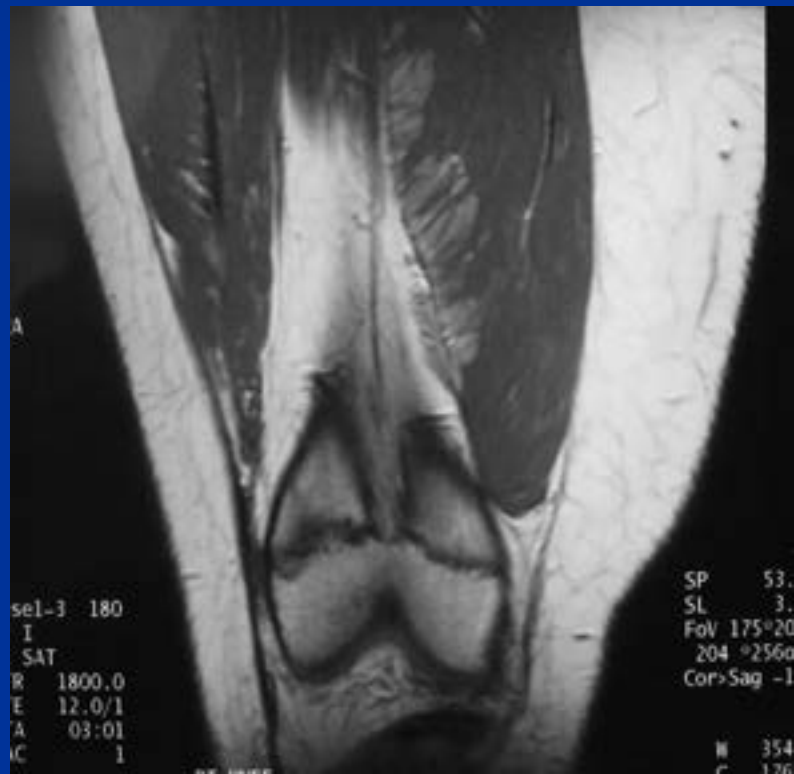
Hemangioma



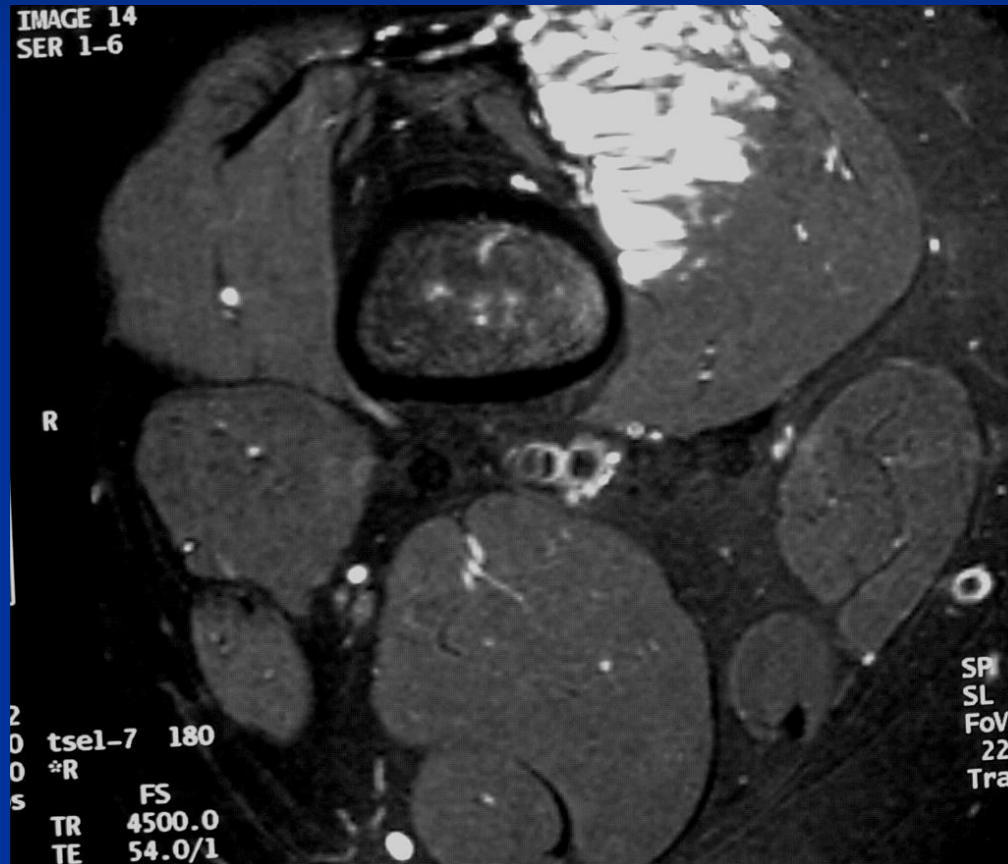
Hemangioma



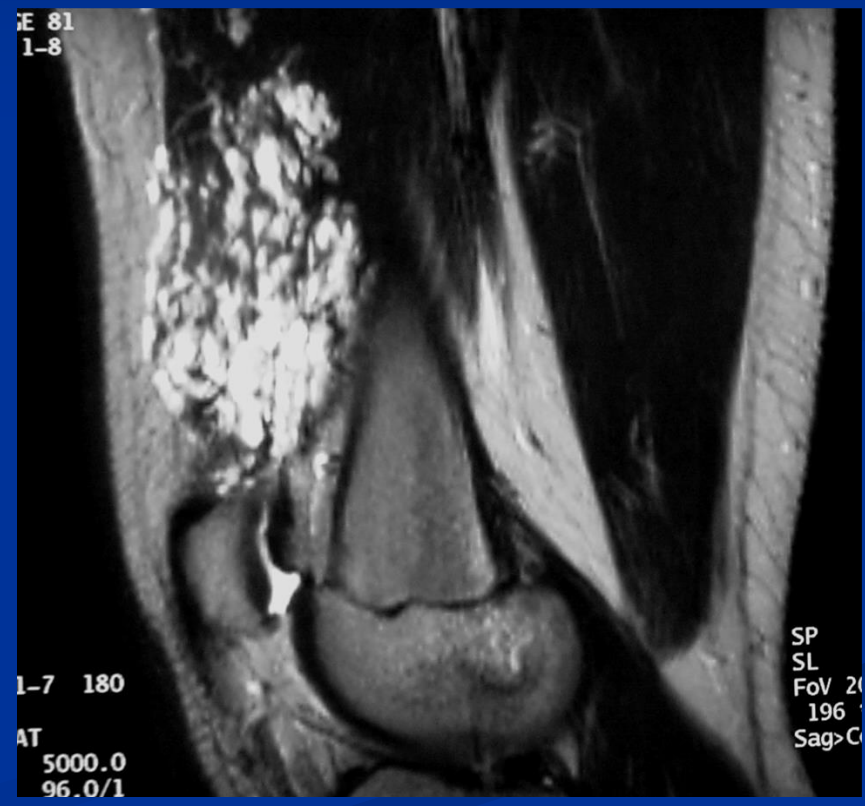
Hemangioma



MRI T2 Hemangioma



Hemangioma



Pathology

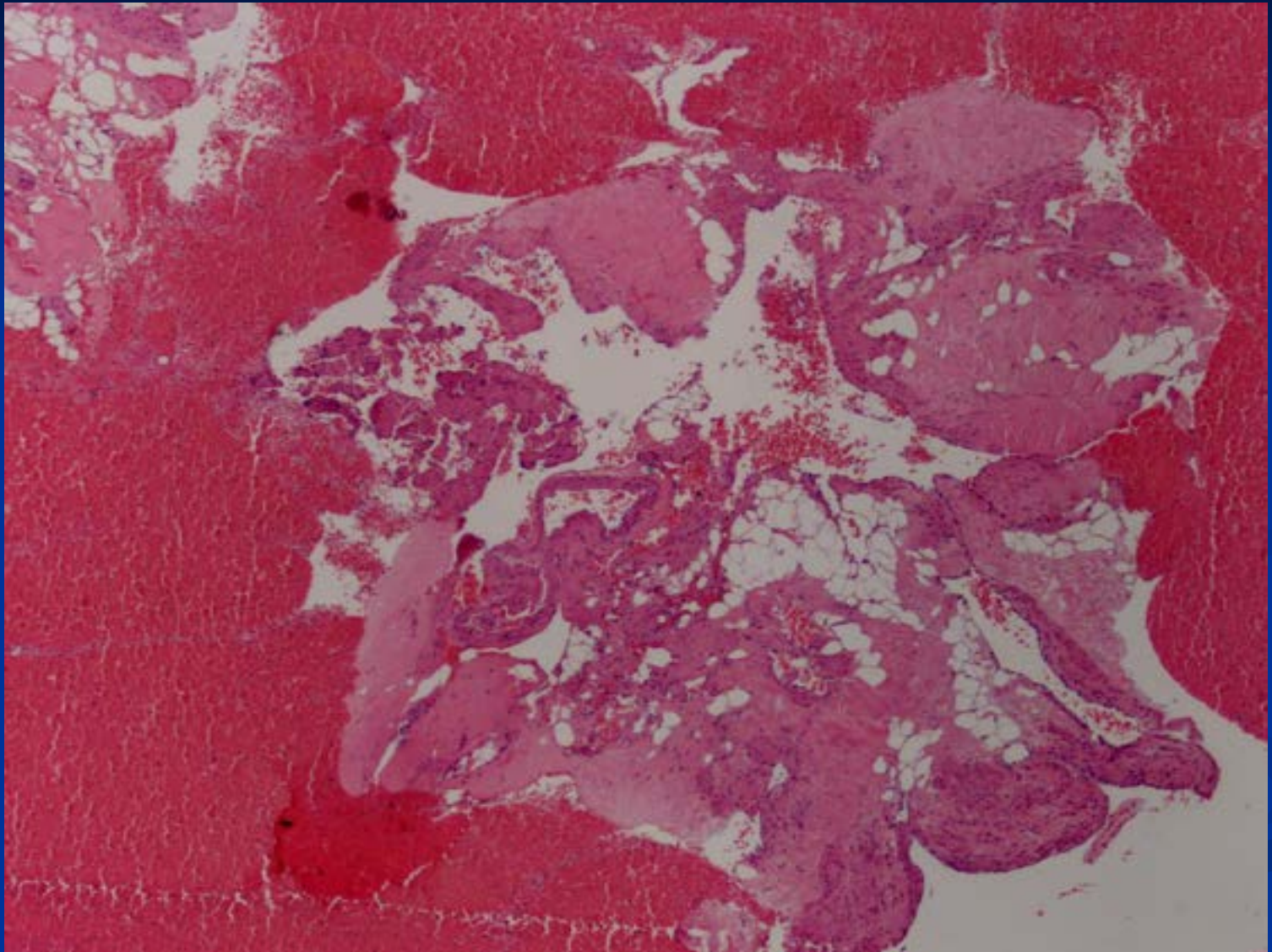
Intramuscular Hemangioma

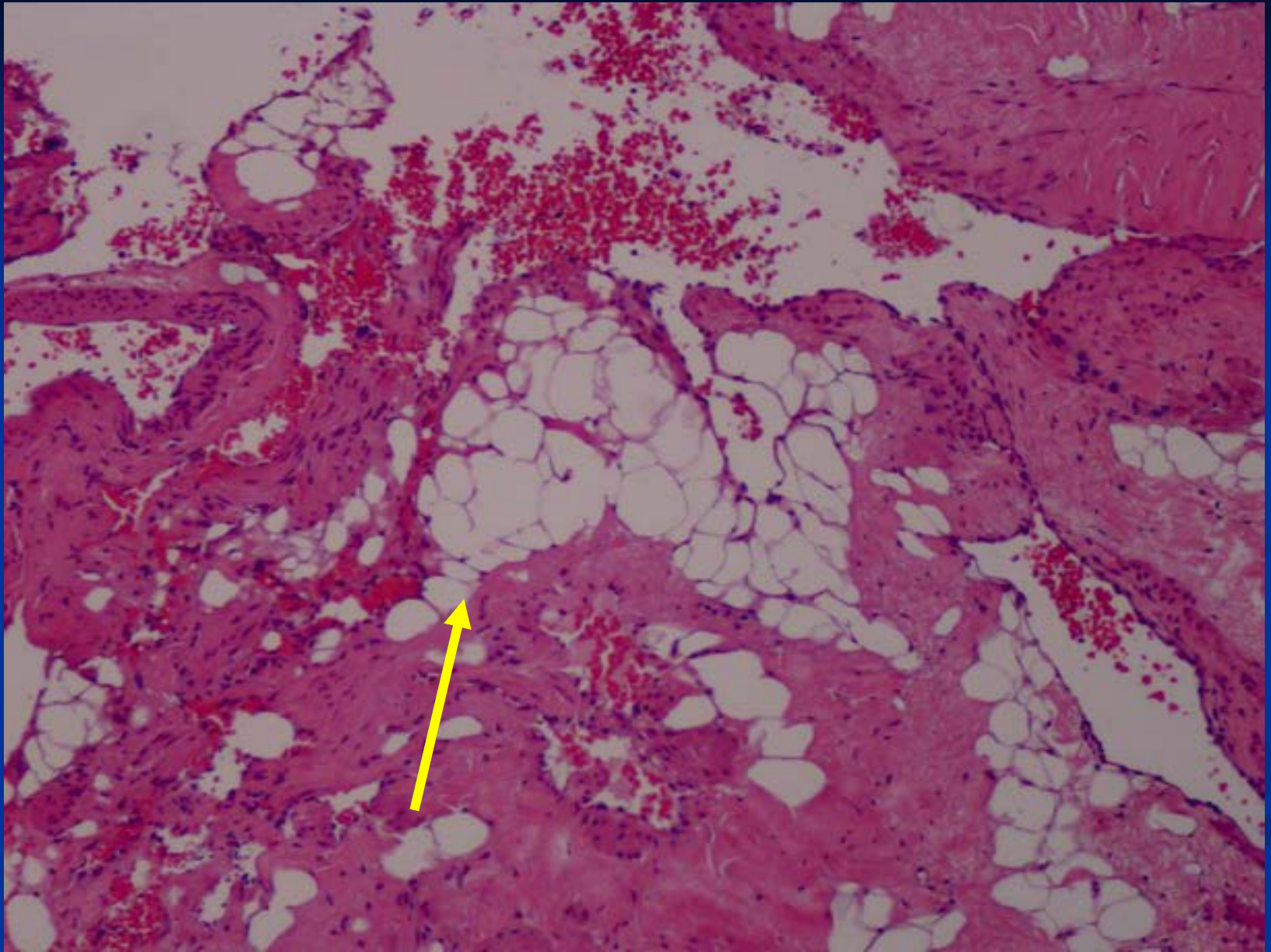
- **Gross Pathology:**

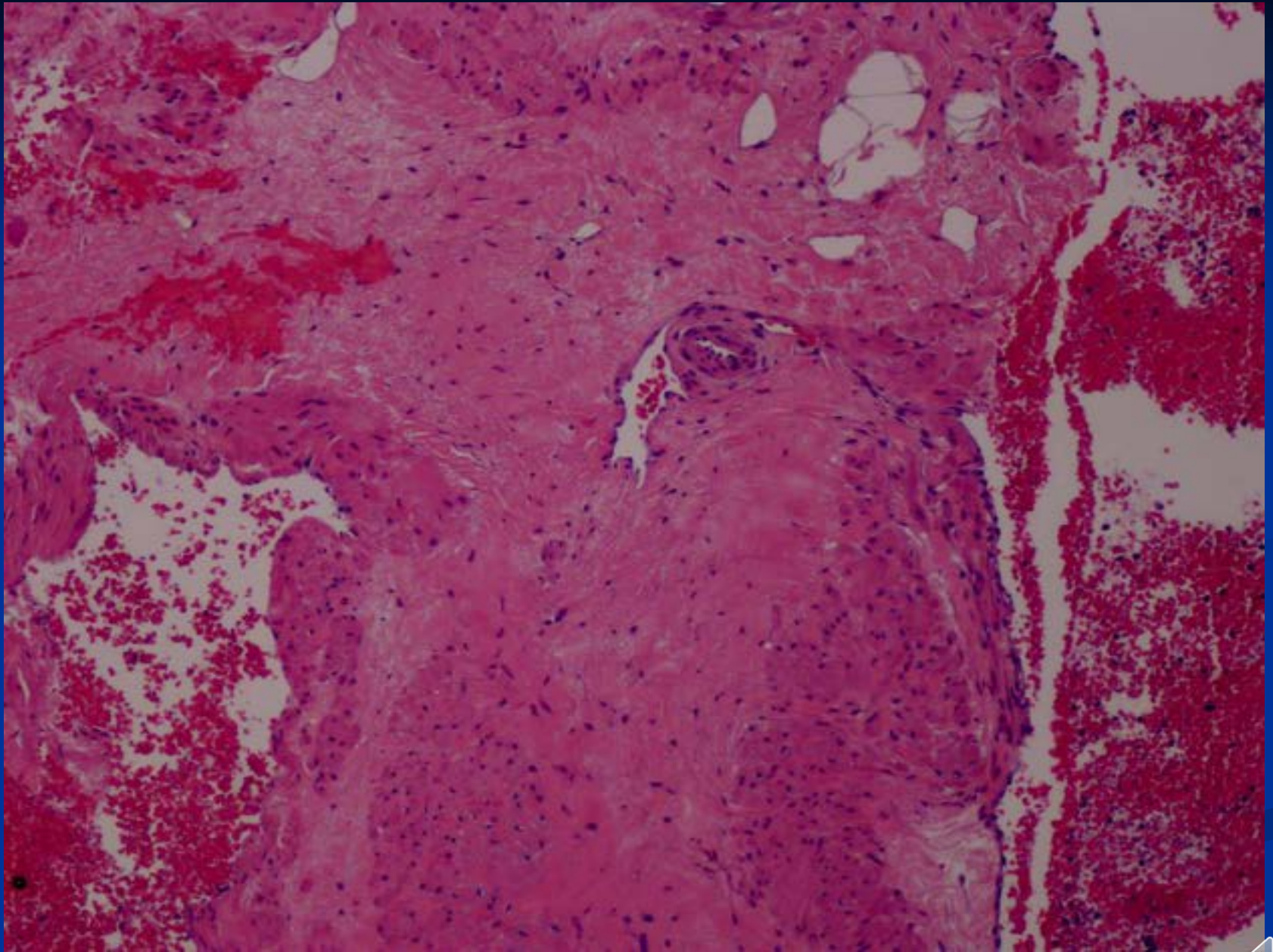
- Large, poorly demarcated, yellowish (fatty nature)
- Vascular and hemorrhagic areas
- Focal calcification or ossification

- **Microscopic Pathology:**

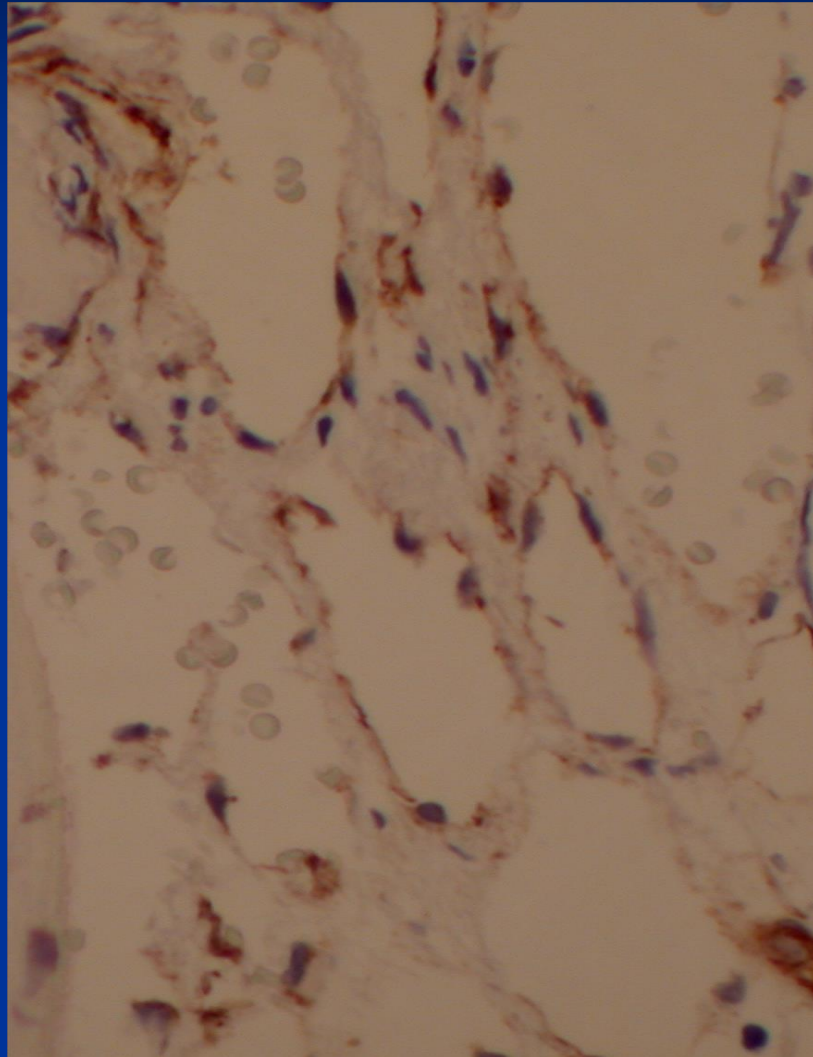
- Usually mixed vessel type
- Lined by normal appearing single layer of epithelial cells
- Diffusely infiltrate muscle and entrap muscle cells leading to degenerative bizarre muscle cells
- Prominent adipose component

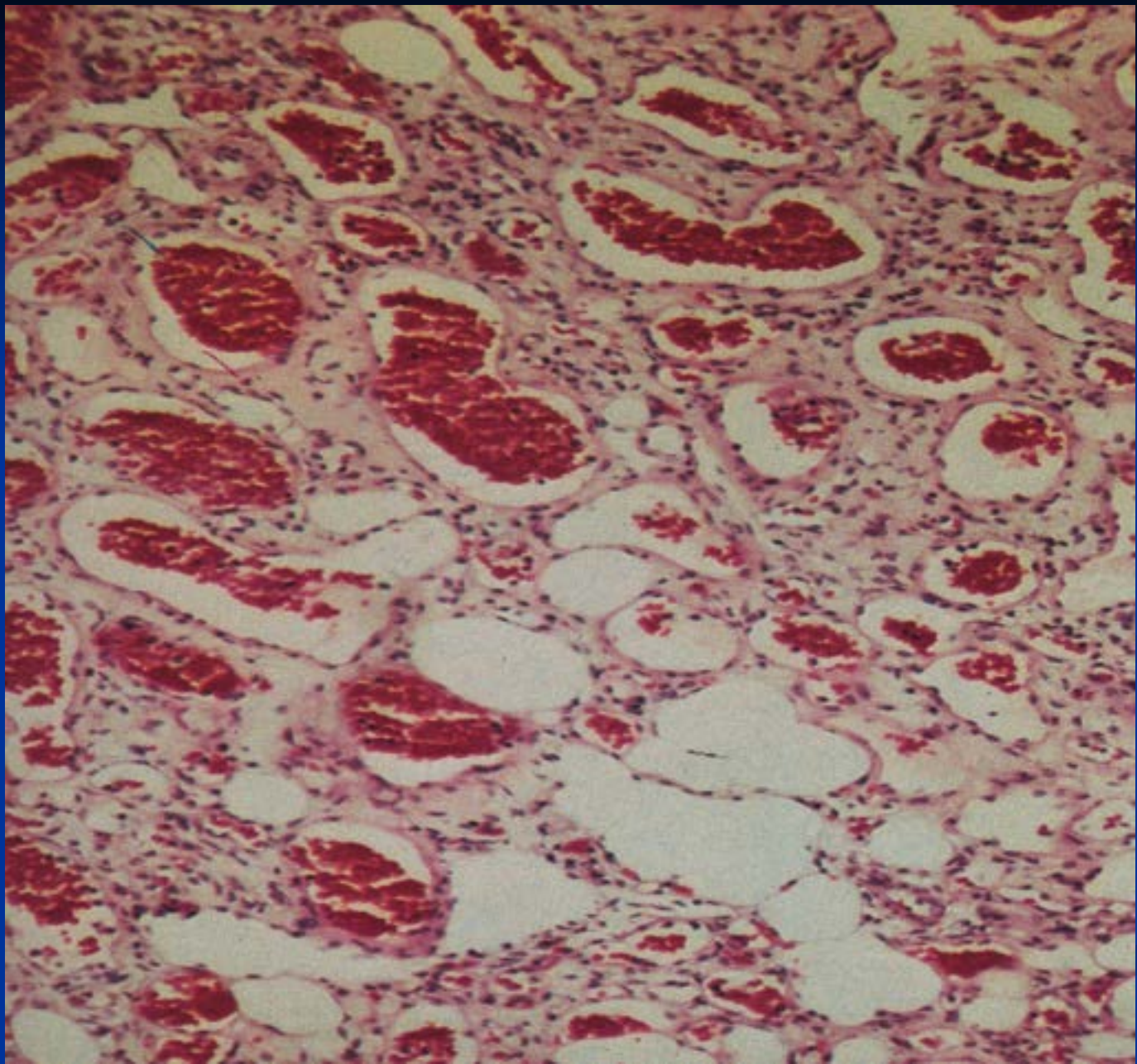






Hemangioma





Fibromatosis/Desmoplastic Fibroma

Extra-abdominal Desmoid tumor

- **Definition:**

- Benign, nonmetastasizing, infiltrating fibroproliferative neoplasm
- Composed of fibrocytes, fibroblasts, and myofibroblasts within a collagenous to myxoid stroma.
- The cells have uniform, bland nuclear features.
- Infiltration of skeletal muscle occurs routinely.
- High propensity for local recurrence.

Fibromatosis

- 15 to 40 most common; described in all ages
 - Age < 5: Infantile Fibromatosis
 - Females > Males, slightly
- Sites:
 - Shoulder Girdle/Upper Arm (most common)
 - Buttock
 - Trunk
 - Head and Neck 10%
 - Hands and Feet: rare
 - 10% are Multicentric and usually involve an anatomic area although may develop in unrelated areas
- Clinical Presentation:
 - Mass or swelling that may be mildly painful or painless



Radiology: Fibromatosis

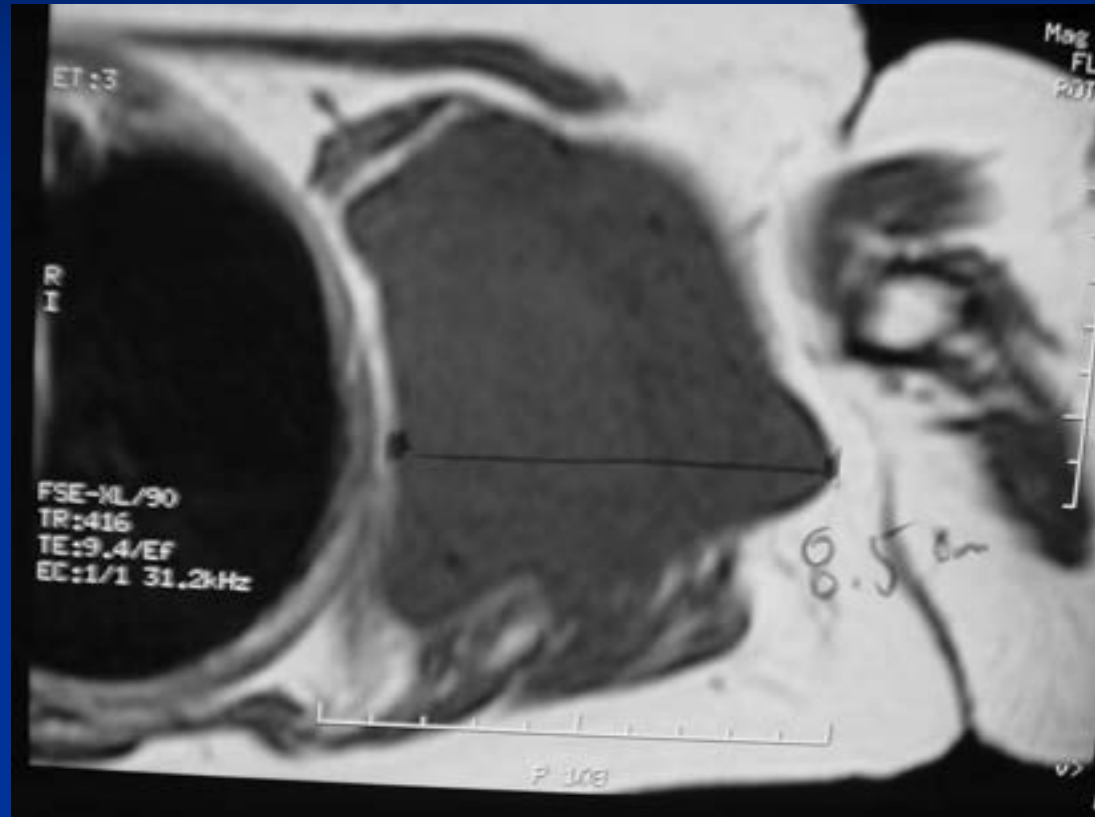
- Plain Radiographs:
 - Usually a nonspecific mass
 - Bone involvement usually pressure erosion/scalloping if present
 - No mineralization (rare)
 - Bone involvement is more common with recurrences
- CT:
 - **Poorly defined margin of mass**
 - Enhance with IV contrast (does not correlate with lesion vascularity)

Radiology: Fibromatosis

- MRI:
 - Infiltrative growth margin
 - Tentacles; Invades adjacent tissues; Not ball-like as with other sarcomas
 - T1: Intermediate Signal Similar to Muscle
 - T2: Variable: usually heterogeneous
 - Low Signal if hypocellular and significant collagen
 - High signal with hypercellularity, myxoid change / mucopolysaccharides
 - Identification of low signal areas on all pulse sequences of dense collagenous areas
 - Usually enhance markedly with Gado (10% no enhancement)

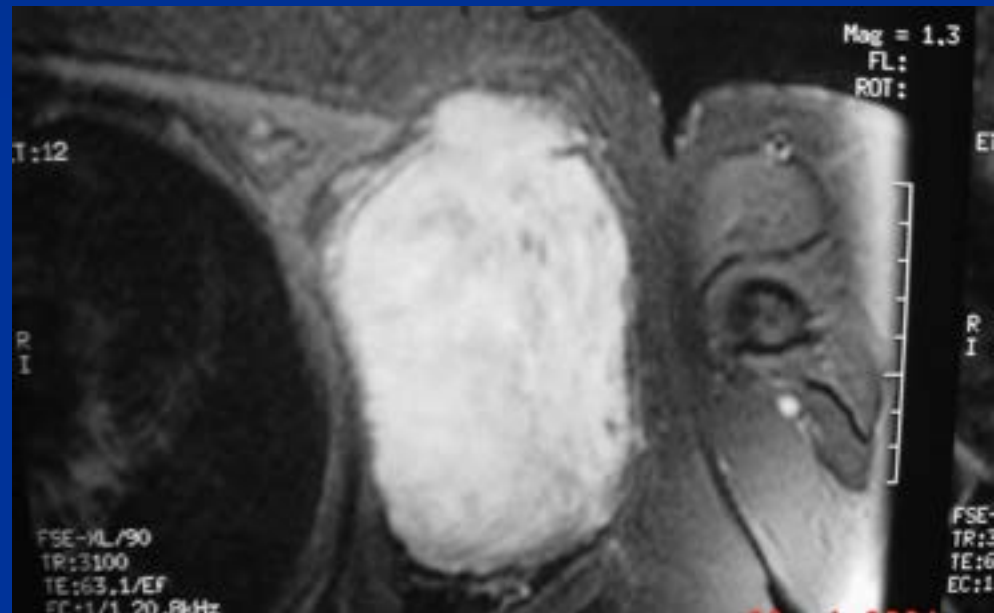
MRI T1: Fibromatosis

- Intermediate Signal
- Not Ball Like
- Shoulder Girdle Area
- Serratus Anterior

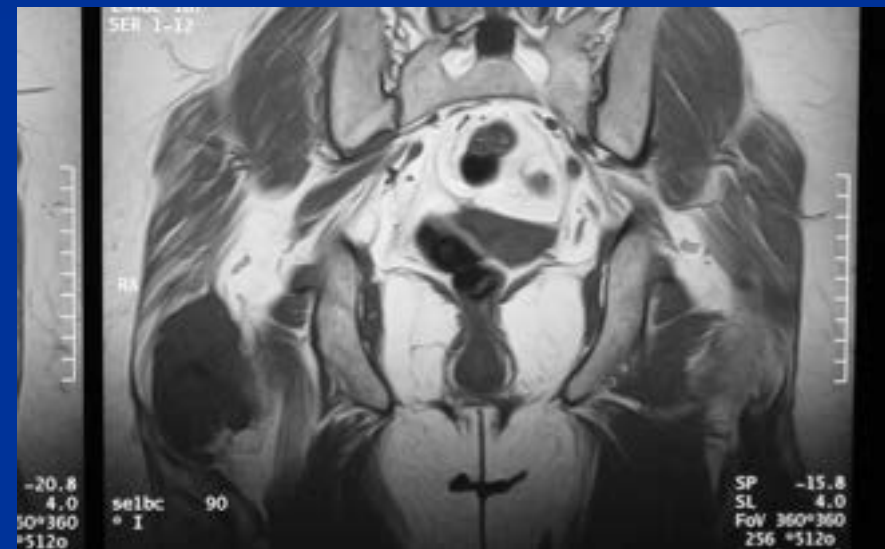
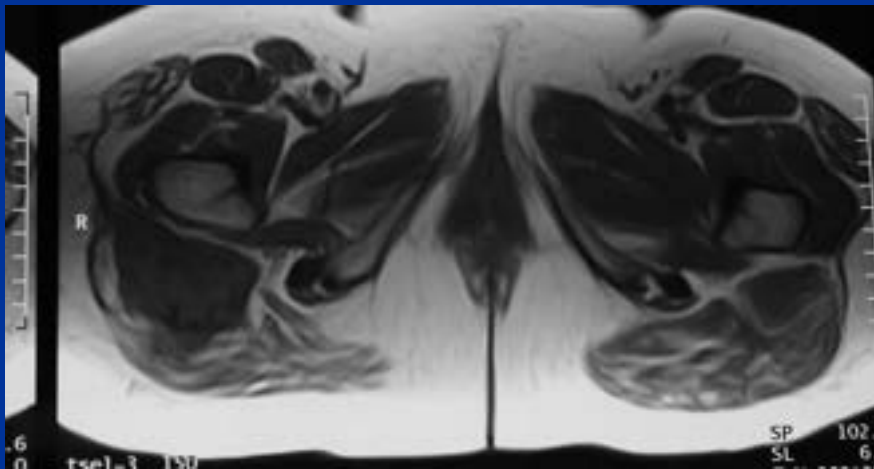


MRI T2: Fibromatosis

- Hyperintense (myxoid or more cellular)
- Subtle low signal areas in tumor (fibrous tissue)

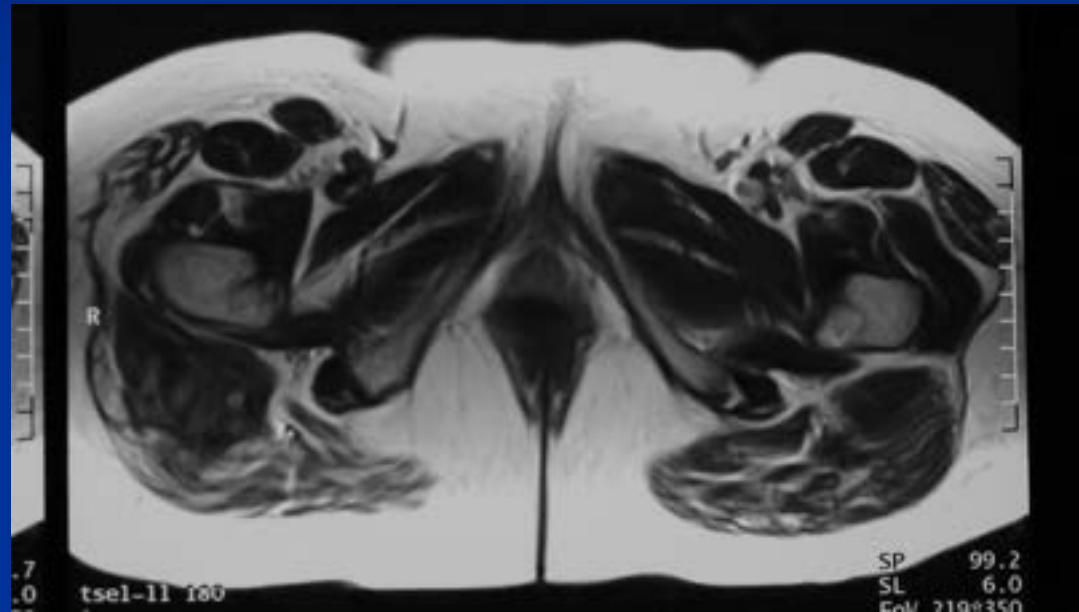


MRI T1: Right Buttock Fibromatosis



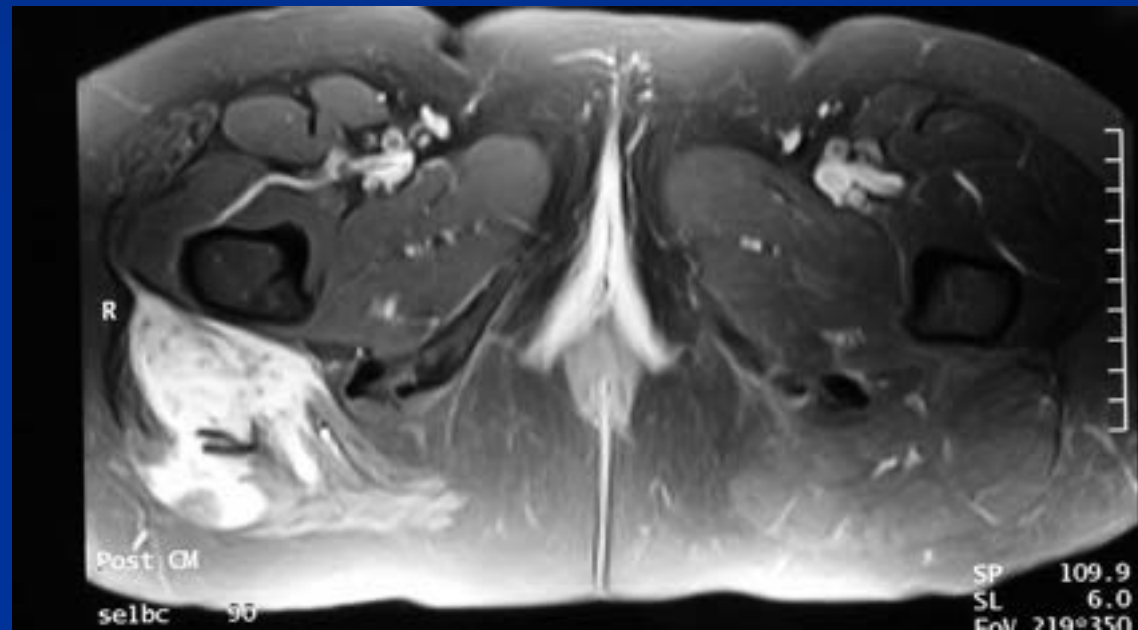
MRI T2: Fibromatosis Right Buttock

- Low to Intermediate Signal on T2
- Significant Fibrous Component

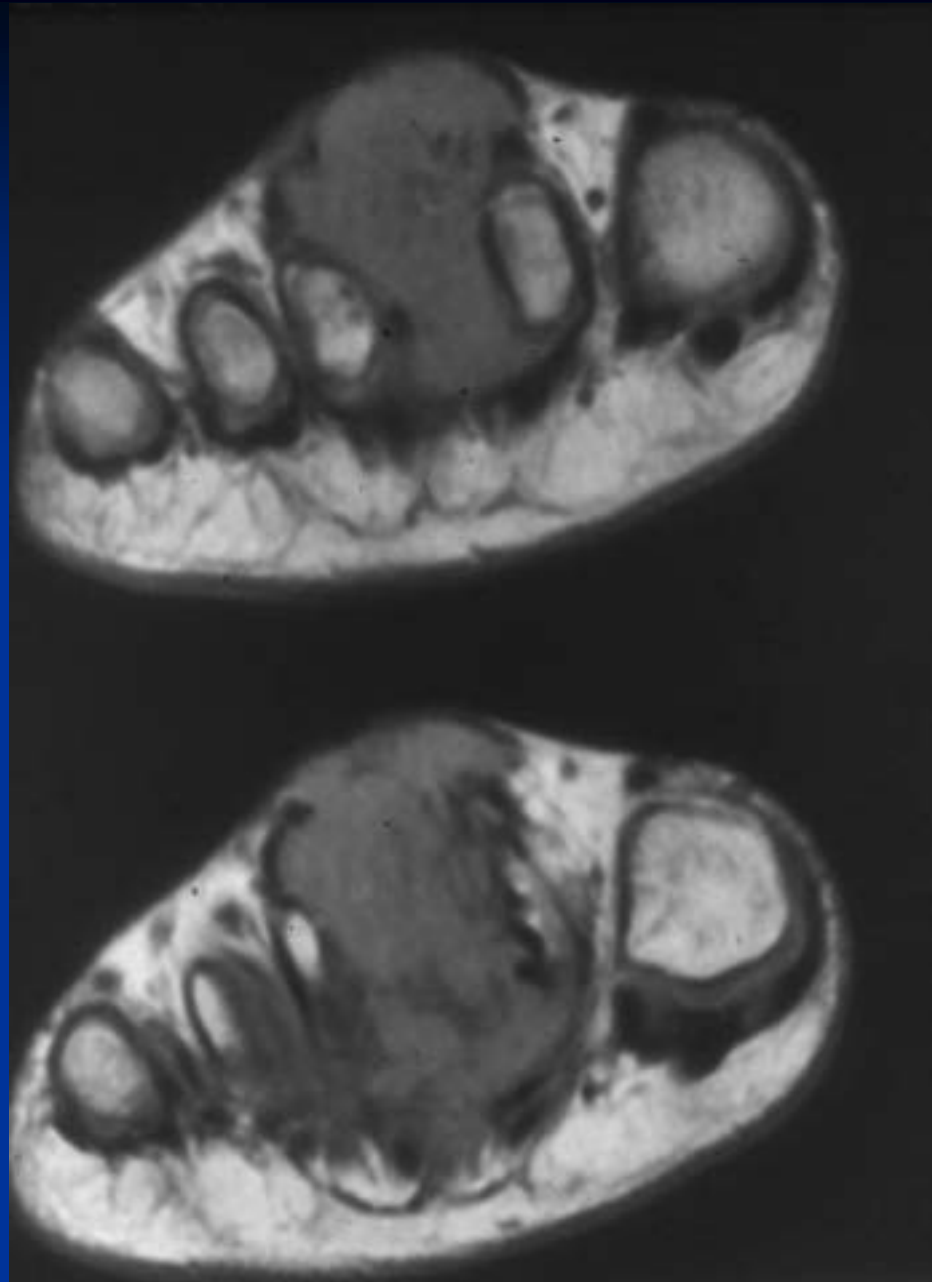


MRI with Gadolinium: Fibromatosis

- Enhances Diffusely
- Very Infiltrative
- Poor margins





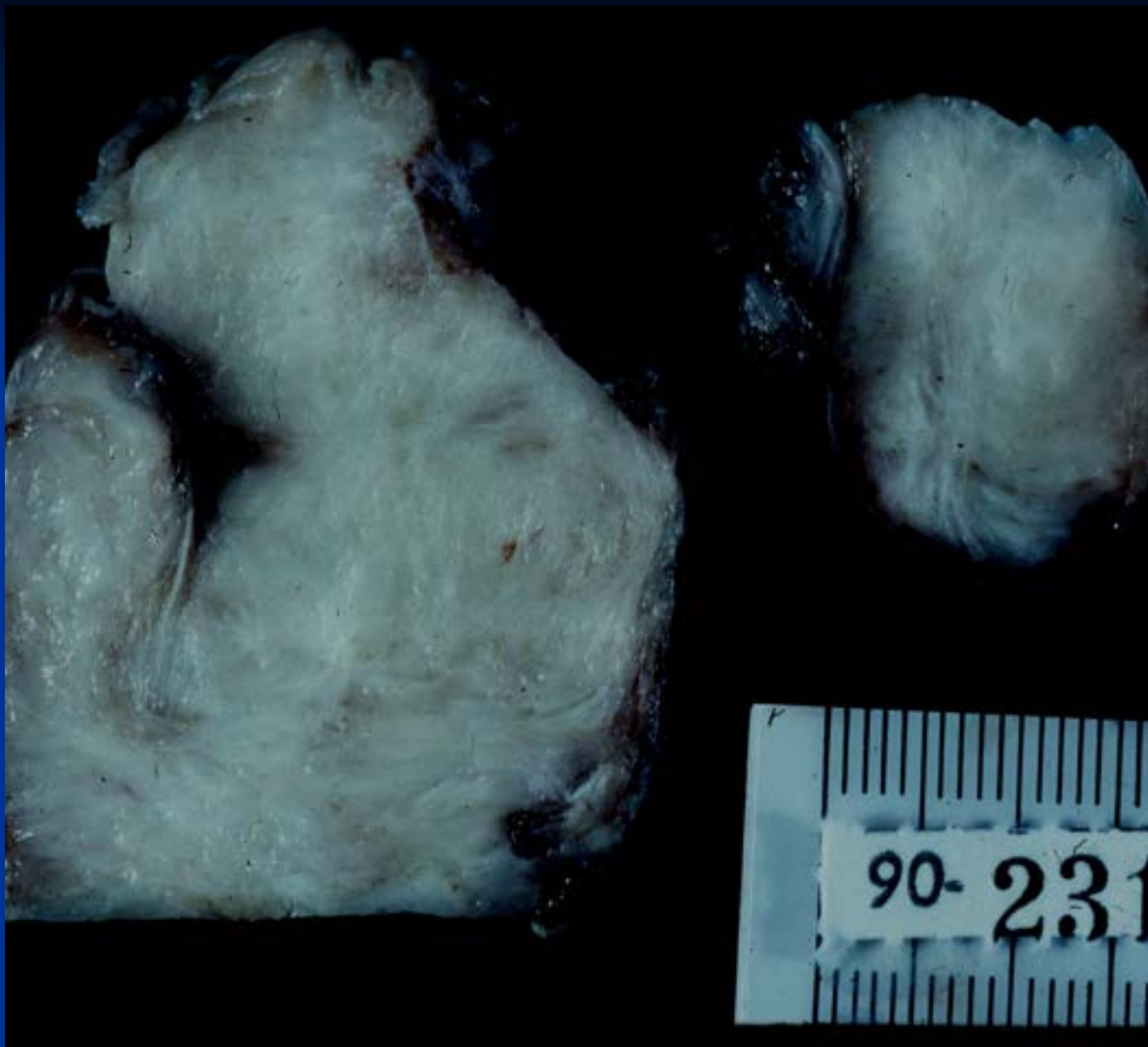


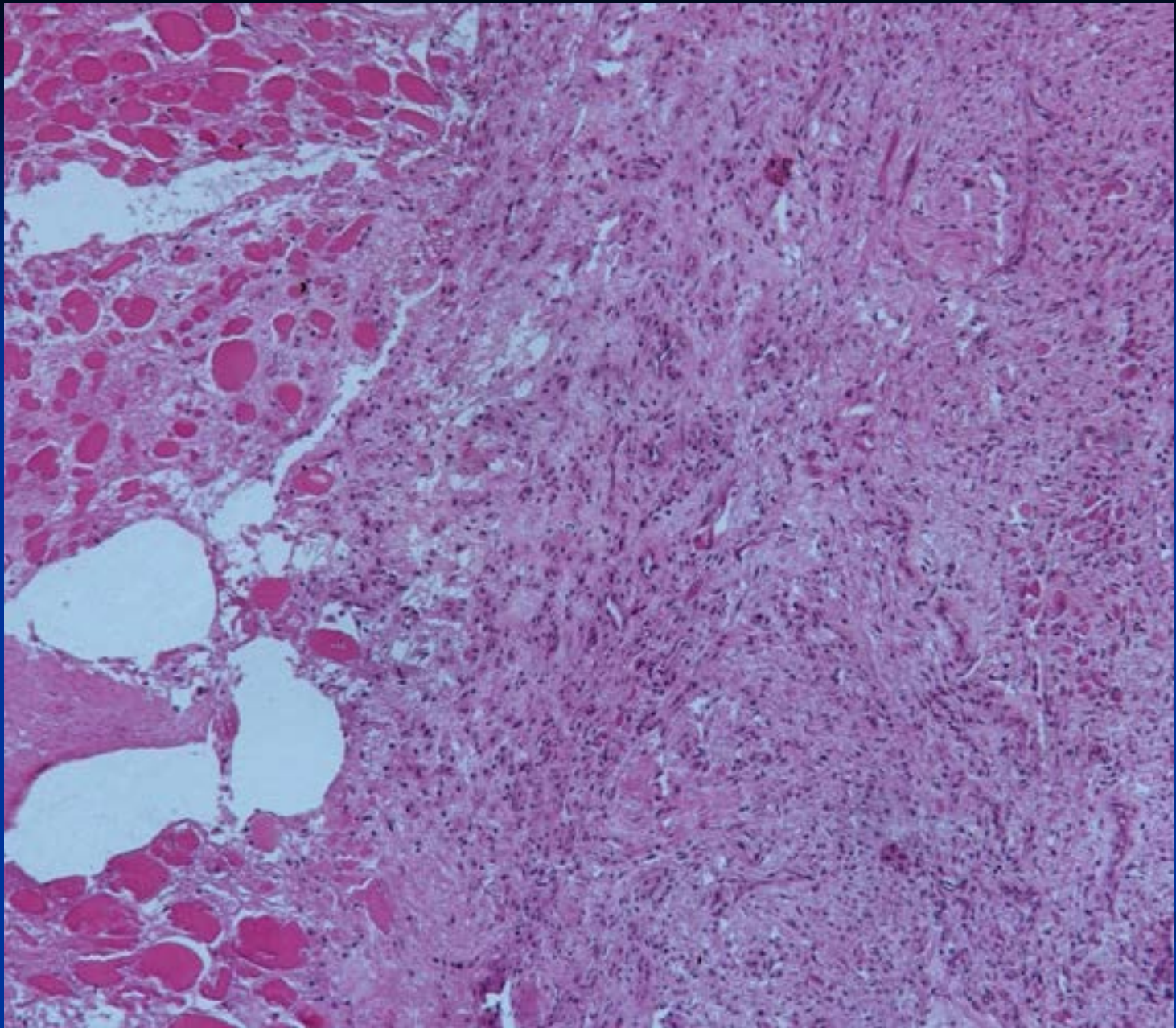
Pathology: Fibromatosis

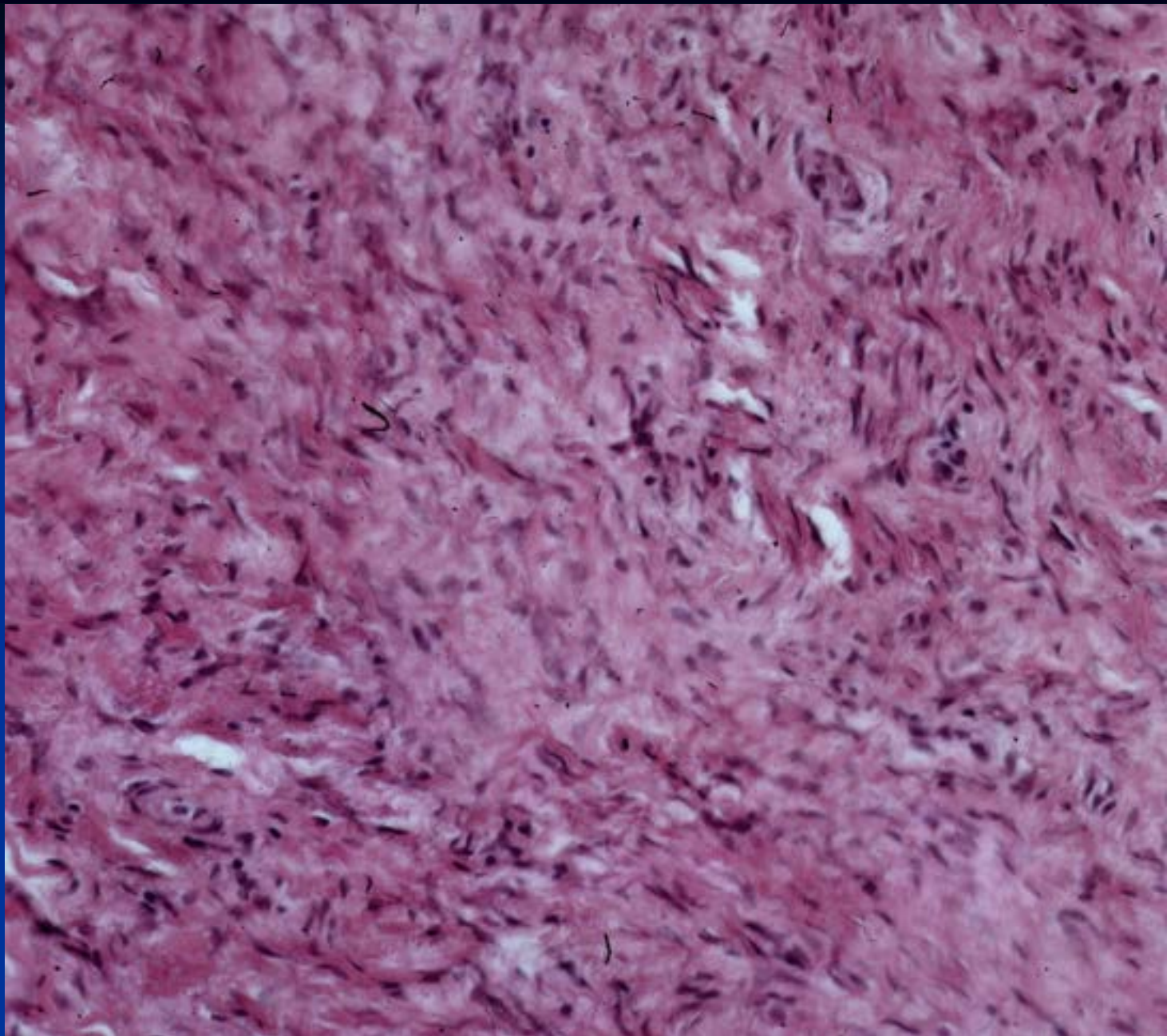
- Gross Pathology:
 - Most tumors are large (5 to 10 cm and up to 20 cm)
 - Glitening white fibrous tissue
 - Myxoid areas may be noted
 - Infiltrates skeletal muscle at the periphery

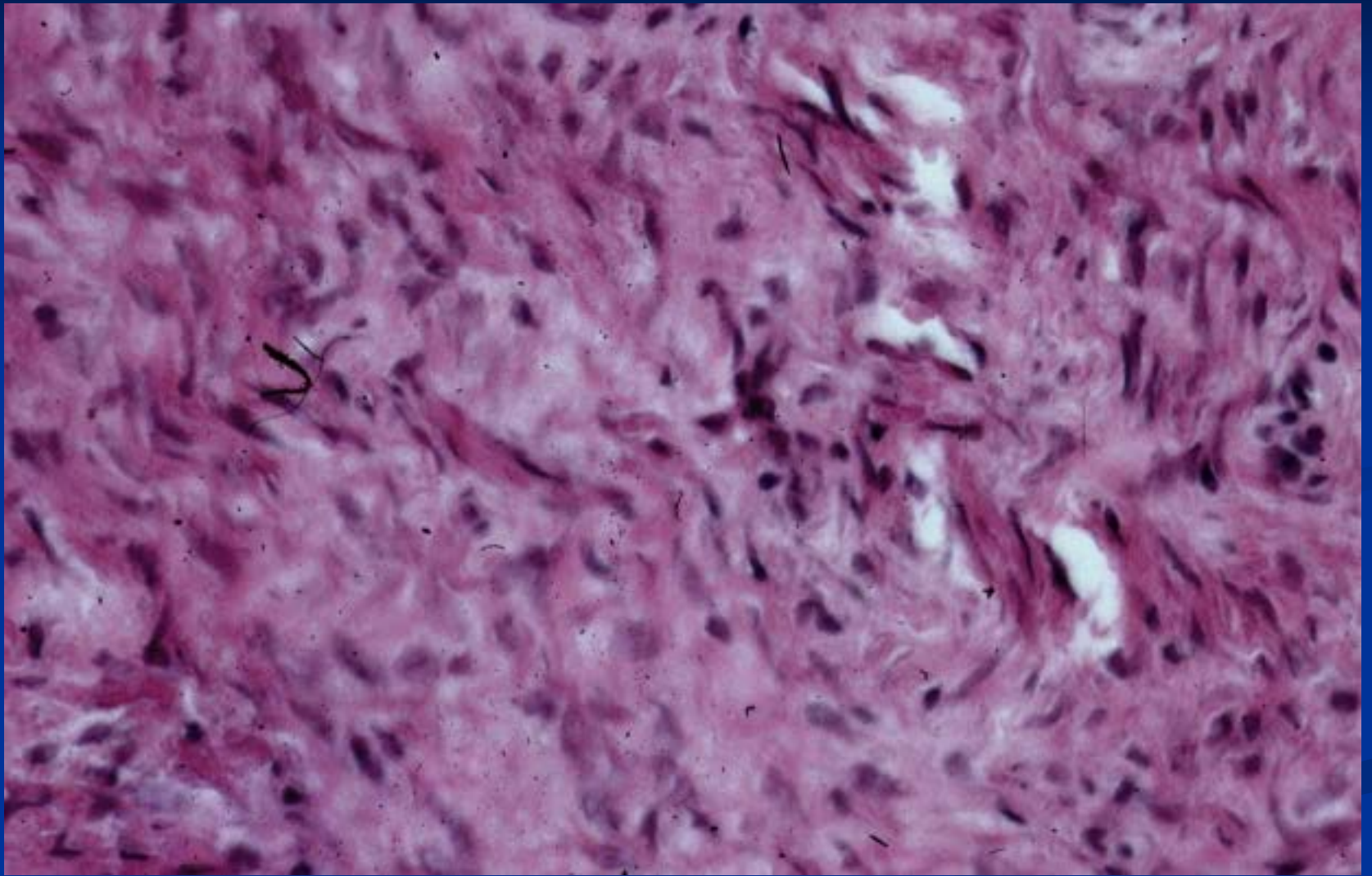
Pathology: Fibromatosis

- Microscopic Pathology:
 - Hypocellular to moderately cellular lesion; No Atypia
 - Elongate, uniform, bland fibroblasts and myofibroblasts loosely arranged in bundles
 - Collagenous to myxoid matrix (resembles scar, fascia, fibrous tissue, tendon)
 - Often wavy collagen bundles; sometimes large thickened collagen bundles
 - Infiltrates skeletal muscle at periphery
 - Numerous thin walled compressed vessels appear slit-like

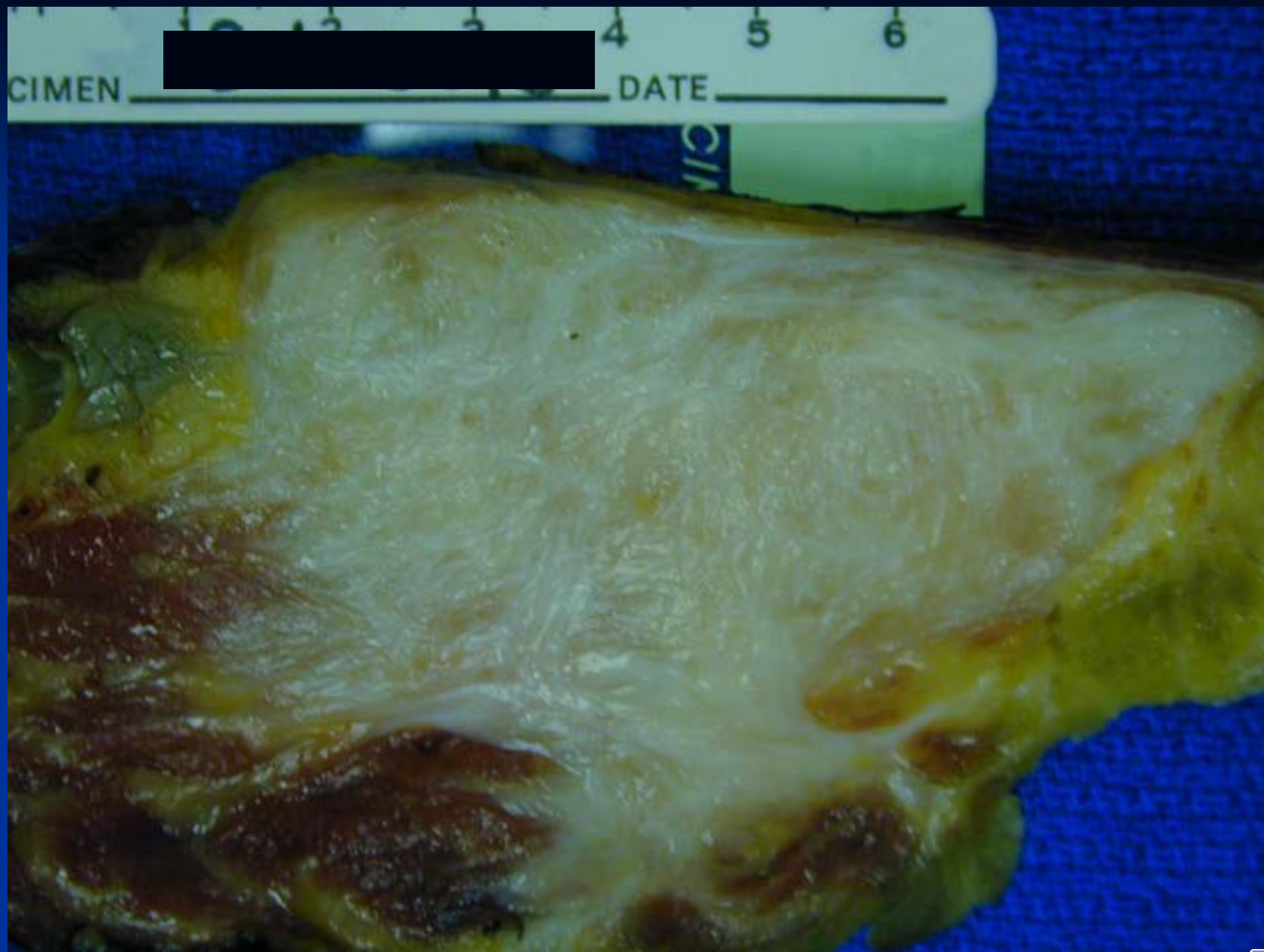


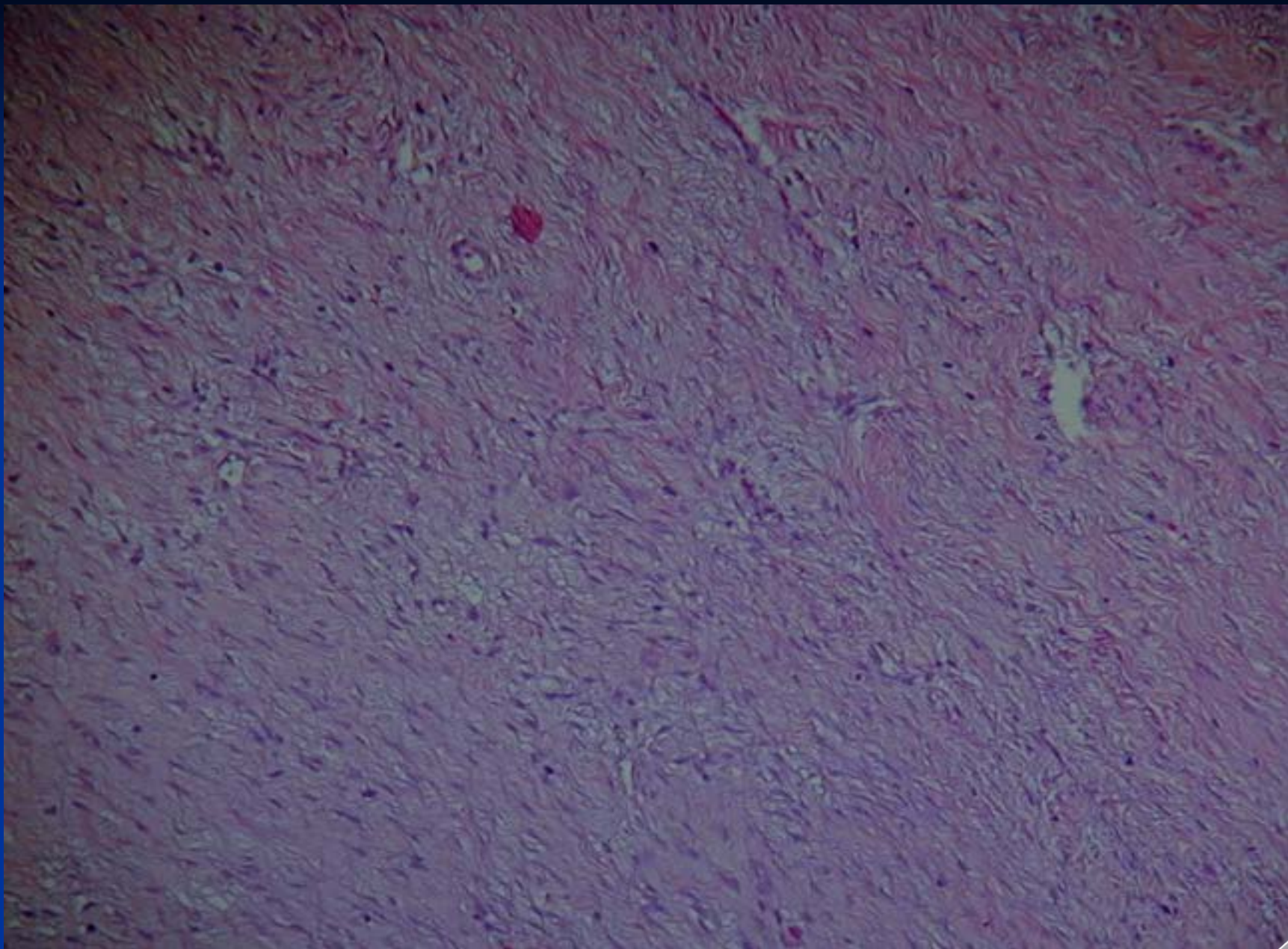


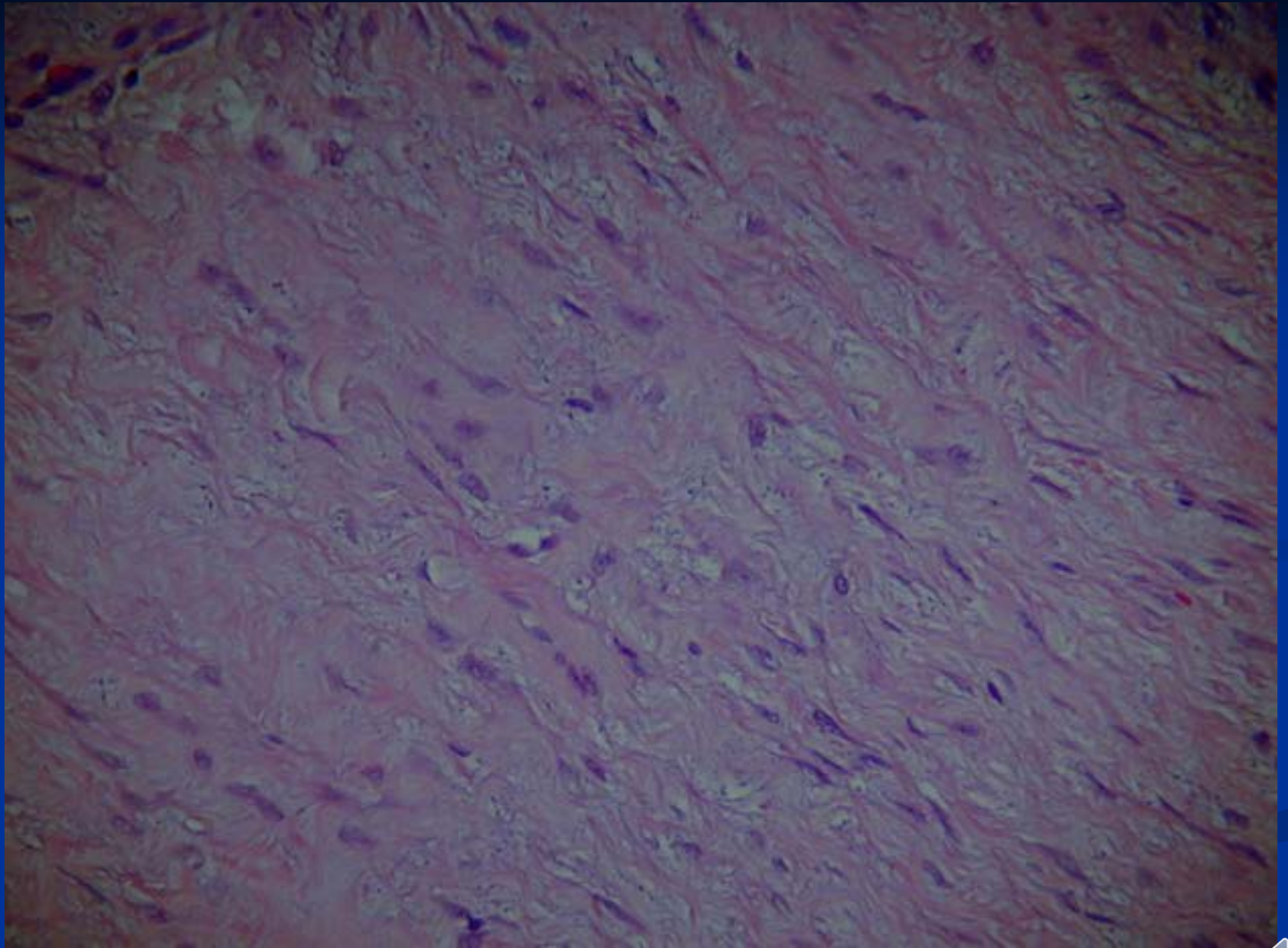


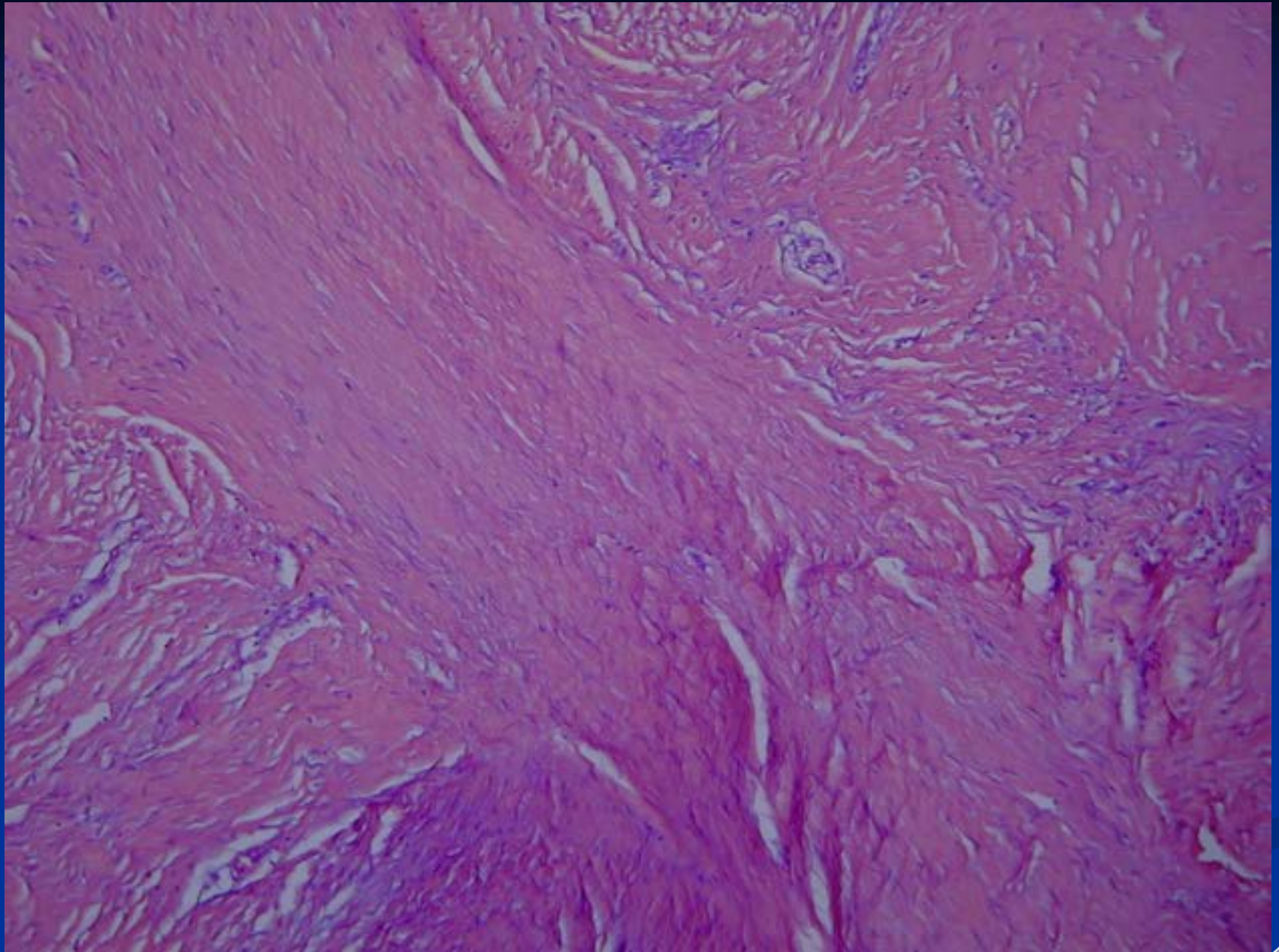


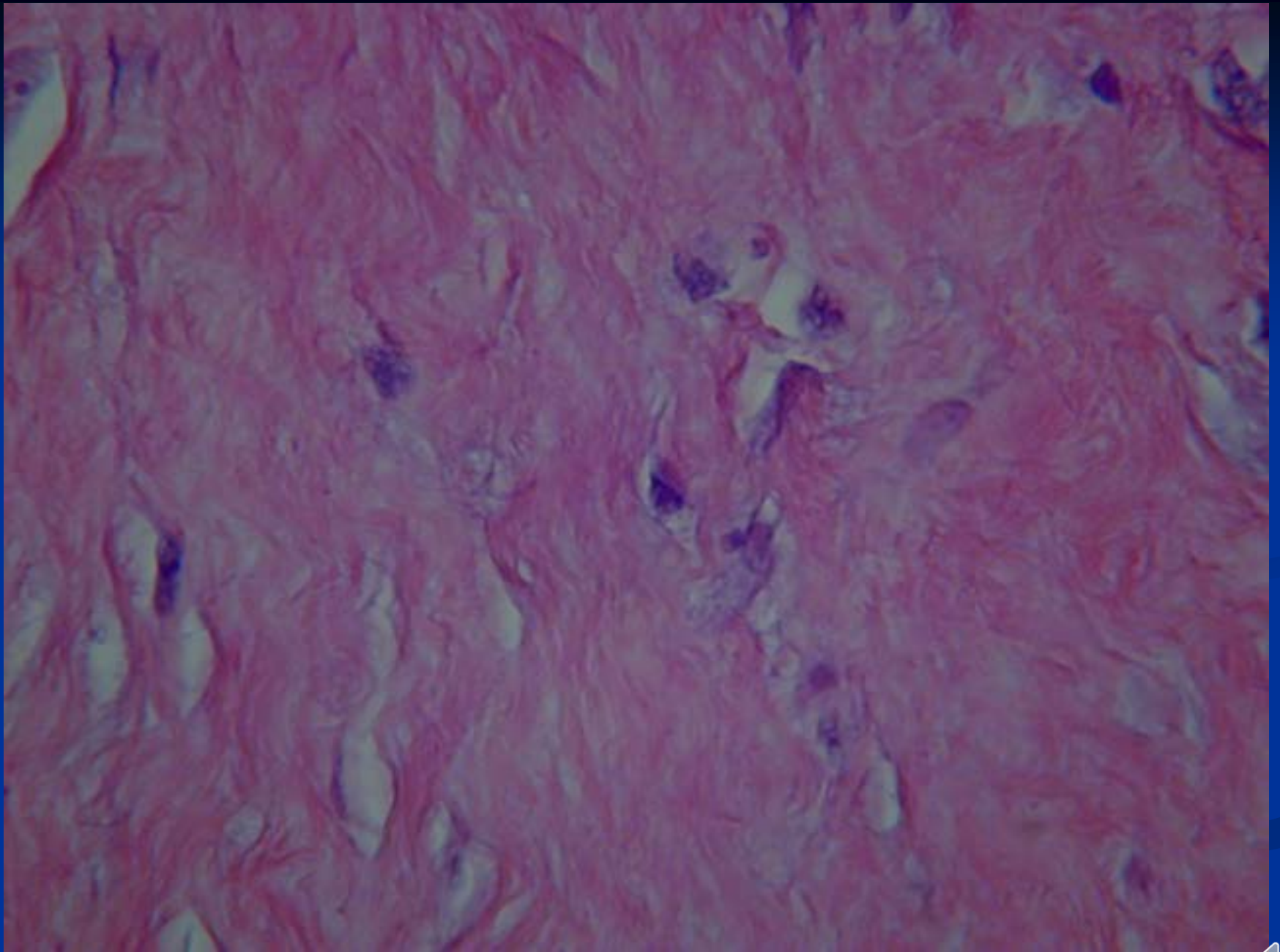












Differential Diagnosis

- Differentiate from Fibrosarcoma
 - Difficult to differentiate from Grade 1 Fibrosarcoma
- Fibrosarcomas:
 - More cellular; abnormal chromatin, Mitoses (>5 per high power field)
 - Grow more ball-like and less infiltrative although radiographic appearance can be similar to fibromatosis

Treatment and Prognosis

- Fibromatosis is a benign, nonmetastasizing locally aggressive neoplasm with a locally destructive growth pattern and propensity for local recurrence following resection.
- Local destruction of vital structures can lead to death (ie. growth into thoracic outlet and mediastinum)

Treatment and Prognosis

- Treatment:
 - Surgery: Wide Excision
 - Local Recurrence: 20%; Depends on size, location and surgical margin
 - Radiation: Controversial
 - May be considered postoperatively for close or microscopically positive margins depending on site and multiple patient factors and if the tumor is recurrent
 - Chemotherapy: Controversial; Some regimens noted to shrink some fibromatoses; utilized in selected situations

Myxoma

- **Definition:** Benign hypocellular tumor with sparse vasculature.
- Bland spindle to stellate fibroblasts within an abundant **myxoid/mucinous** stroma.
- Rare tumor: 1 case per 1 million people
- Adults 40 to 60 years old
- Females more affected than males
- Rare childhood cases
- **Sites:**
 - Thigh, Gluteus, Upper Arm

Myxoma

- **Clinical:**
 - Slowly growing mass
 - Painless
 - Size usually 5 to 10 cm
 - Multiple tumors (rare; 5% of patients)
 - Associated with fibrous dysplasia of underlying bone in about 5% of cases and in cases of multiple myxomas (Mazabraud Syndrome)

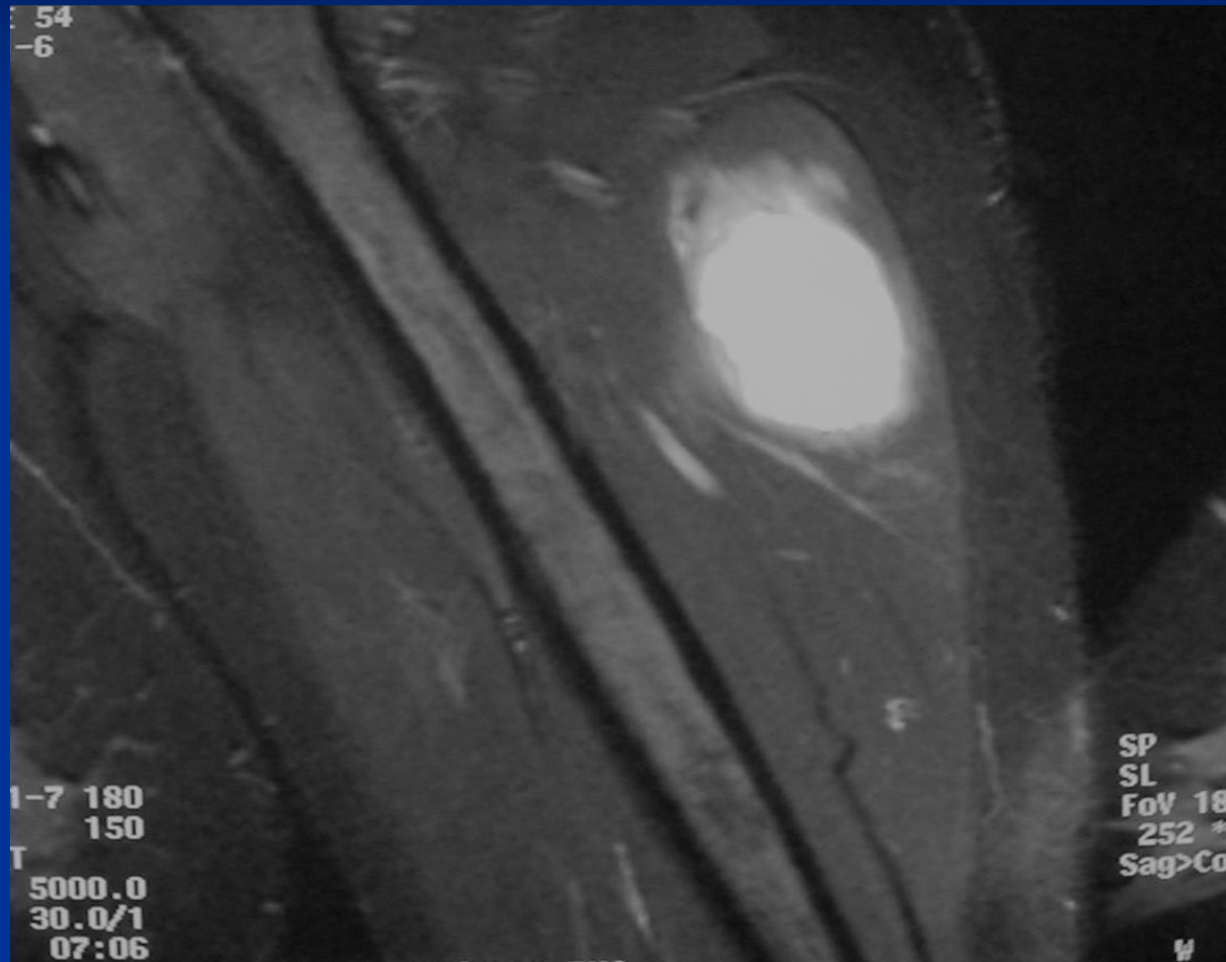
Radiology: Myxoma

- Radiographs:
 - Normal or a nonspecific soft tissue mass
 - Mineralization is extremely rare
- CT:
 - Well defined, homogeneous soft tissue mass
 - No enhancement with CT contrast
- MRI: Appear similar to fluid or ganglion cysts
 - T1: Homogeneous mass; Signal lower than muscle
 - T2: Markedly high signal
 - Usually peripheral and septal enhancement with gadolinium although may see heterogeneous enhancement

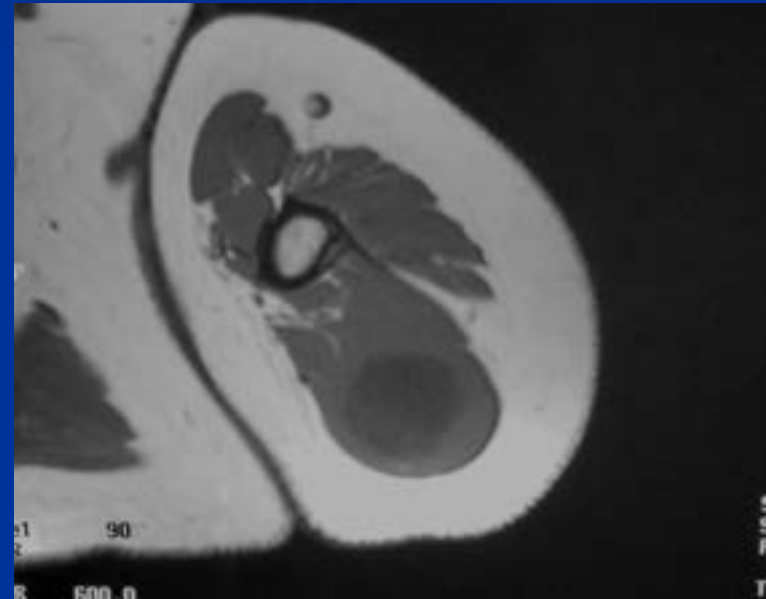
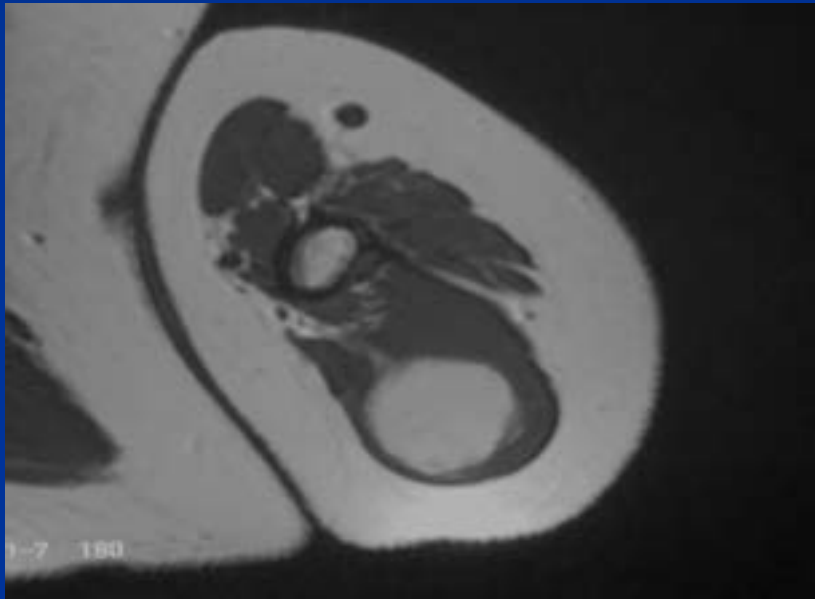
MRI T1: Myxoma of Triceps



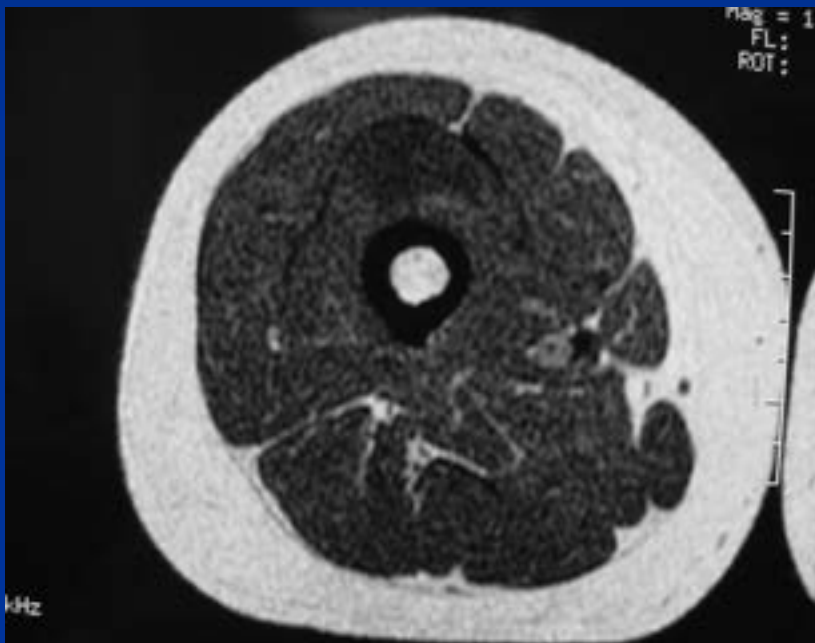
MRI T2: Myxoma of Triceps



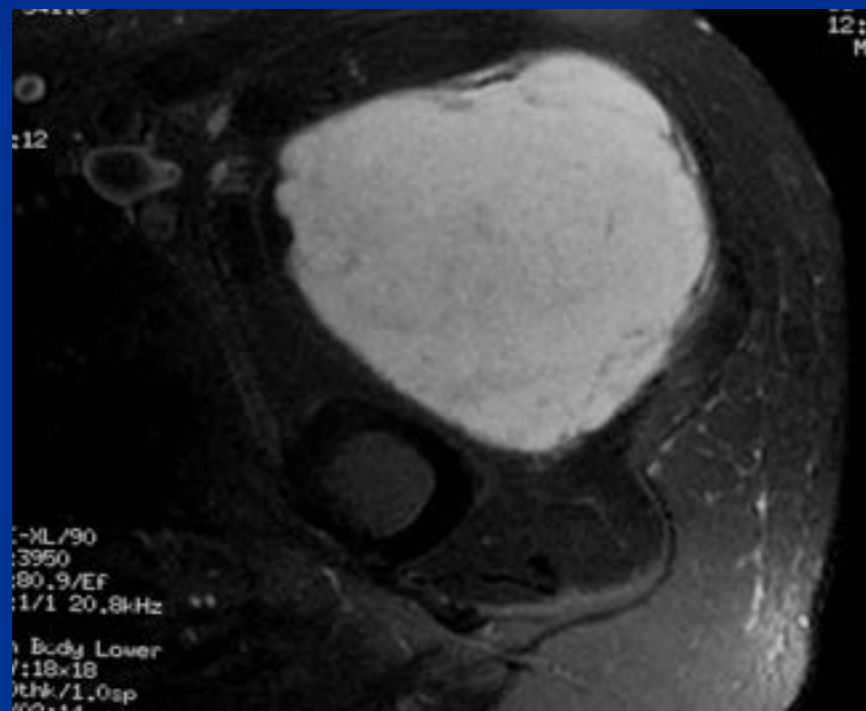
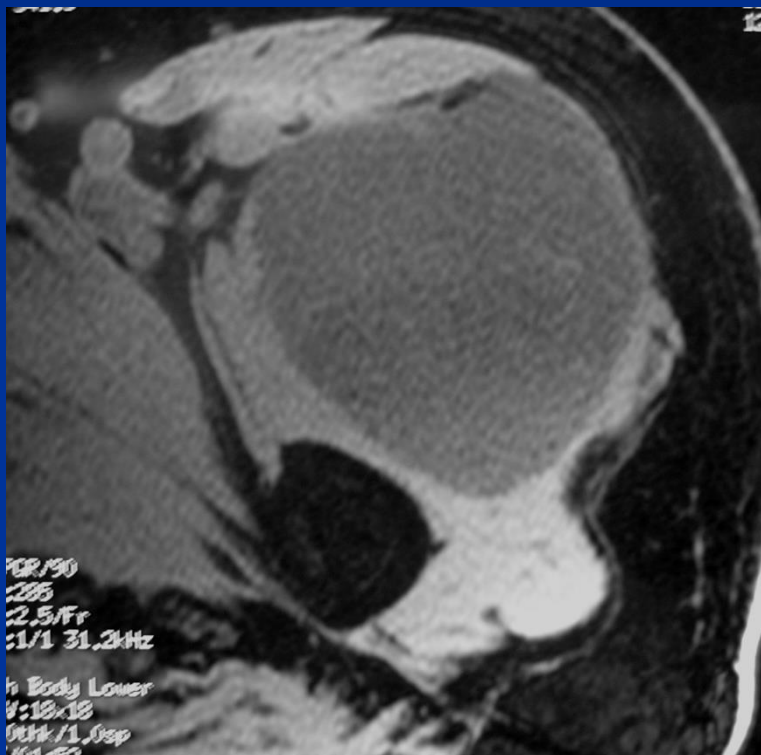
MRI Myxoma of Triceps



MRI: Myxoma of Thigh



Myxoma of Thigh



Pathology: Myxoma

- Gross Pathology:
 - Round to ovoid, well circumscribed
 - Pale mucinous to gelatinous surface
 - No Necrosis and hemorrhage

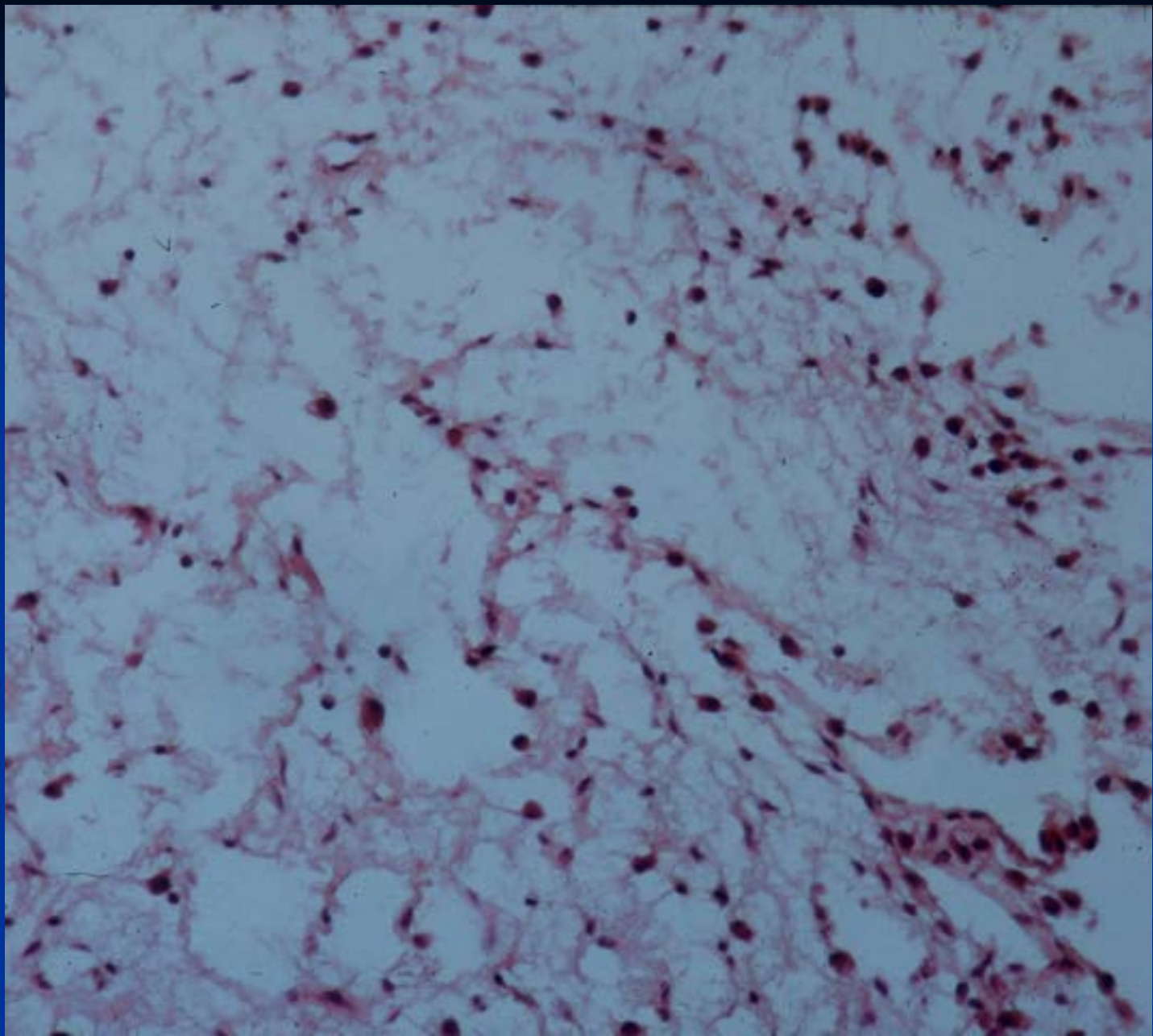


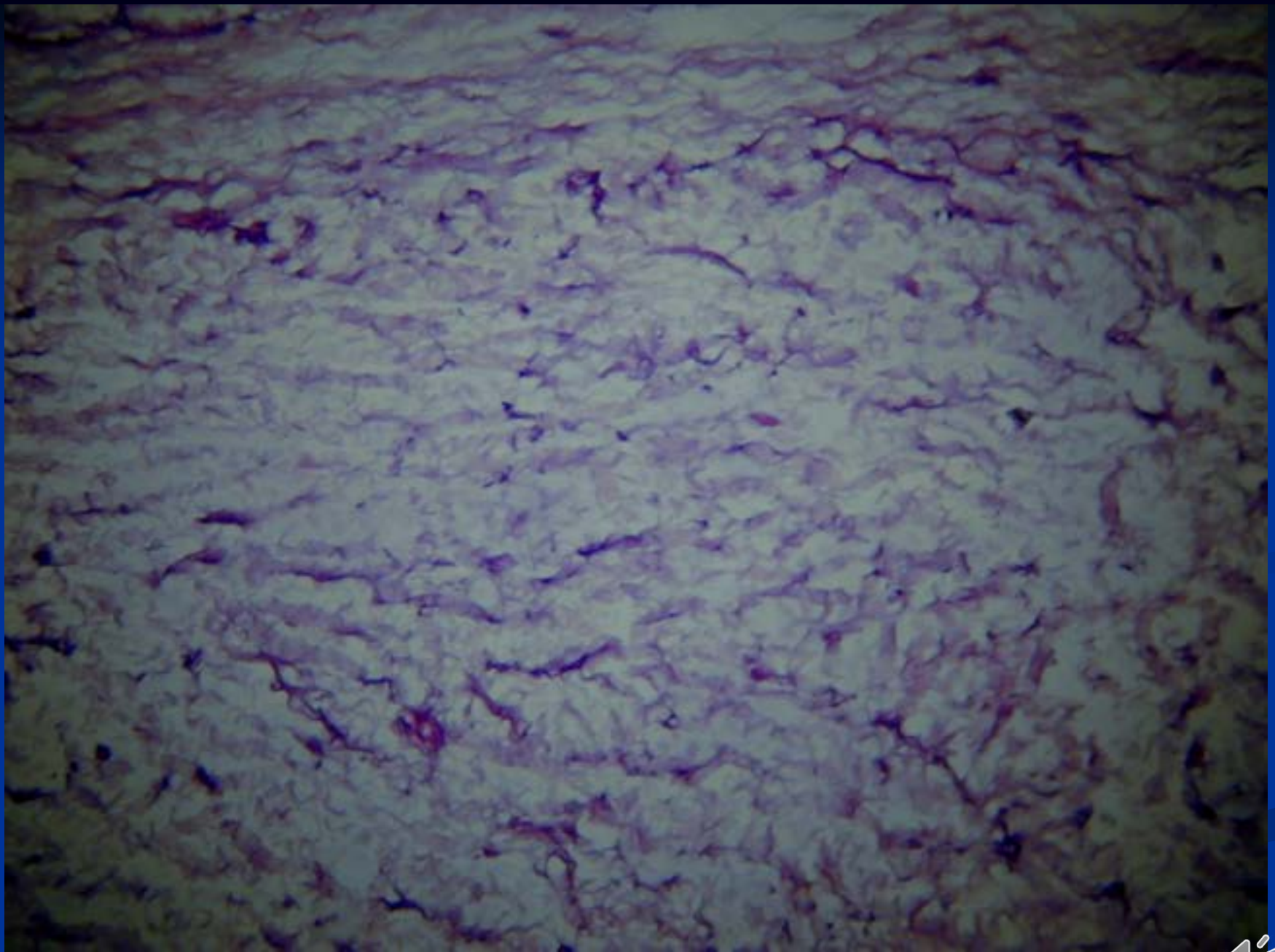
Myxoma



Myxoma

- Microscopic Pathology:
 - Poorly circumscribed, infiltrative lesion
 - Merges with surrounding skeletal muscle and fascial tissue
 - Hypocellular spindle and stellate cells with pale cytoplasm and dark, hyperchromatic small nuclei
 - Myxoid/Mucinous Stroma/Matrix that stains pale blue (Hyaluronic acid)
 - No nuclear atypia or pleomorphism
 - Very rare mitoses; No abnormal mitoses
 - Vimentin positive





Differential Diagnosis

- Other Myxoid Neoplasms can appear identical on MRI
 - Myxoid MFH
 - Myxoid Liposarcoma
 - Myxoid Schwannoma
 - Myxoid Leiomyosarcoma
 - Myxoid MPNST
 - Ganglion Cyst

Treatment and Prognosis

- Benign, does not metastasize; grows slowly but in a nondestructive manner
- Treatment: **Marginal** Excision
 - Local recurrence < 5%
 - Very low risk of recurrence even when residual microscopic disease is left behind

Schwannoma/Neurilemmoma

- **Definition:** Benign tumor arising from a peripheral nerve sheath (epineurium) derived from a schwann cell.
- 5% of benign soft tissue tumors
- Adults; 20 years to 50 years old
- Major peripheral nerves in upper and lower extremities, head and neck region
- **Clinical:** Usually painful, slowly growing mass
 - Usually <5cm
 - +Tinel's sign
 - Mobile in transverse plane but no in longitudinal plane

Schwannoma/Neurilemmoma

- Arises from the **periphery** of the nerve, epineurium
- The nerve is displaced eccentrically
- Small cutaneous nerves may appear to be obliterated by the tumor

Radiology: Schwannoma

- Plain Radiographs:
 - Usually normal
- CT:
 - Attenuation on noncontrast CT is lower than muscle

Radiology: Schwannoma

- MRI:
 - Intermuscular mass closely related to a neurovascular bundle with a distinct margin surrounded by rim of fat (**split fat sign**)
 - Fusiform, elongated cigar shaped mass
 - Nerve: Tubular structure visualized entering and exiting the mass (may be difficult to see with small lesions or lesions of the trunk or retroperitoneum)
 - Spinal lesions: usually dumbbell shaped exiting from and enlarged neural foramina

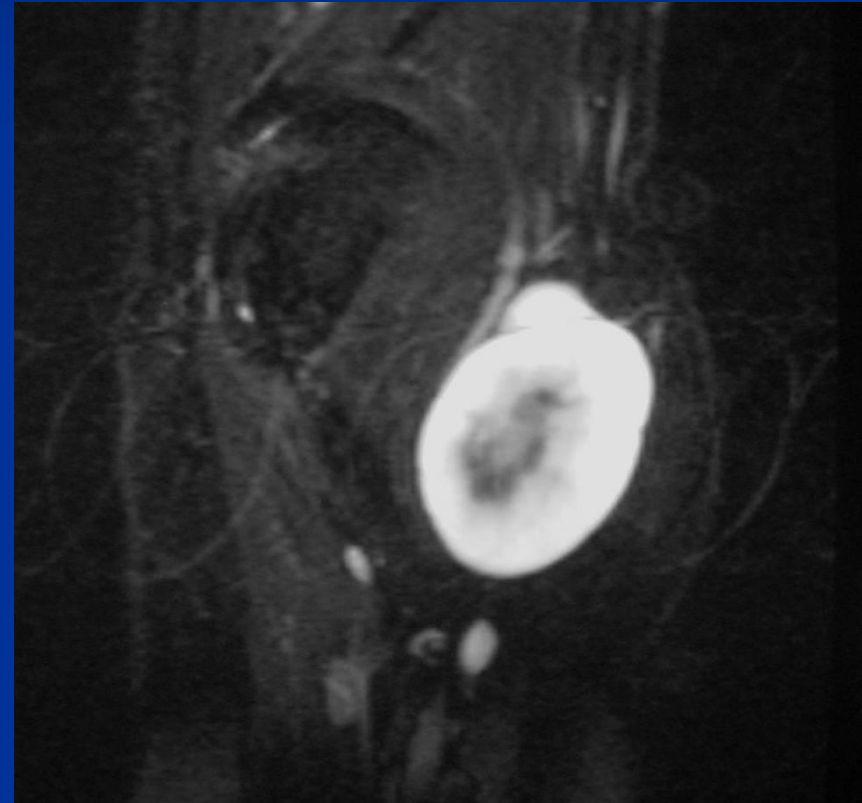
Radiology: Schwannoma

- MRI:
 - T1: Isointense to muscle; Variable enhancement with gado
 - T2: High signal (myxoid areas) with some heterogeneity
 - **Target Sign:** Low signal central area (cellular/collagenous area) and high signal peripheral area (myxoid area)

Pathology: Schwannoma

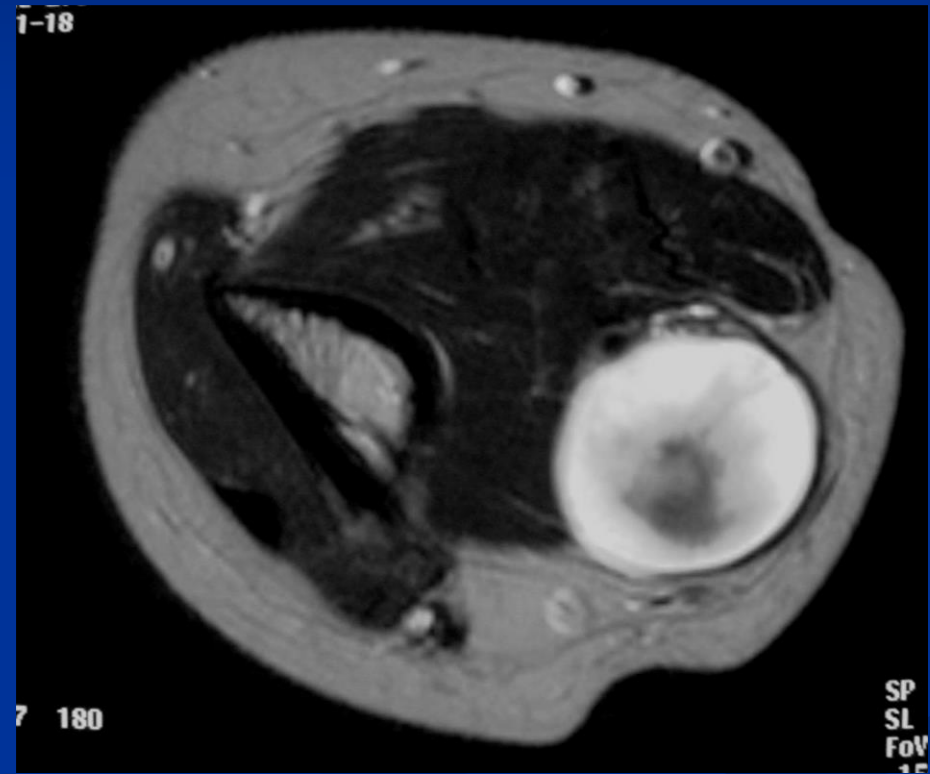
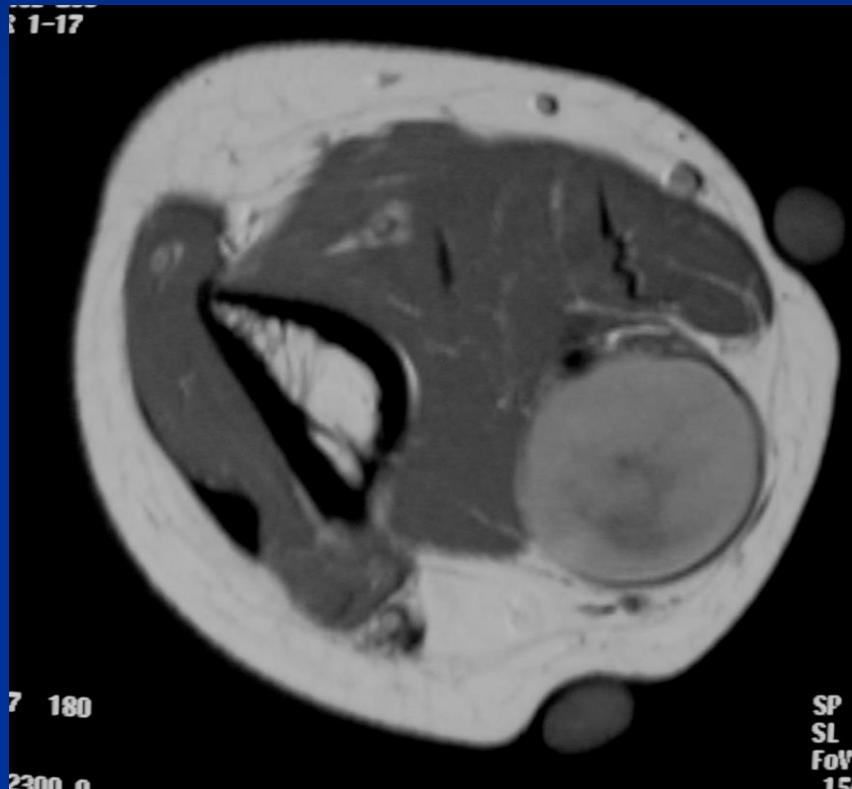
- Microscopic Pathology:
 - Antoni A Area: Cellular area arranged in short bundles or interlacing fascicles
 - Antoni B Area: Less cellular and more myxoid
 - Ancient schwannomas: cyst formation, calcification, hemorrhage, fibrosis
 - S-100 positive staining

MRI T1 and T2: Schwannoma Target Sign



MRI: Schwannoma

Intermuscular, Neurovascular Region, Target Sign

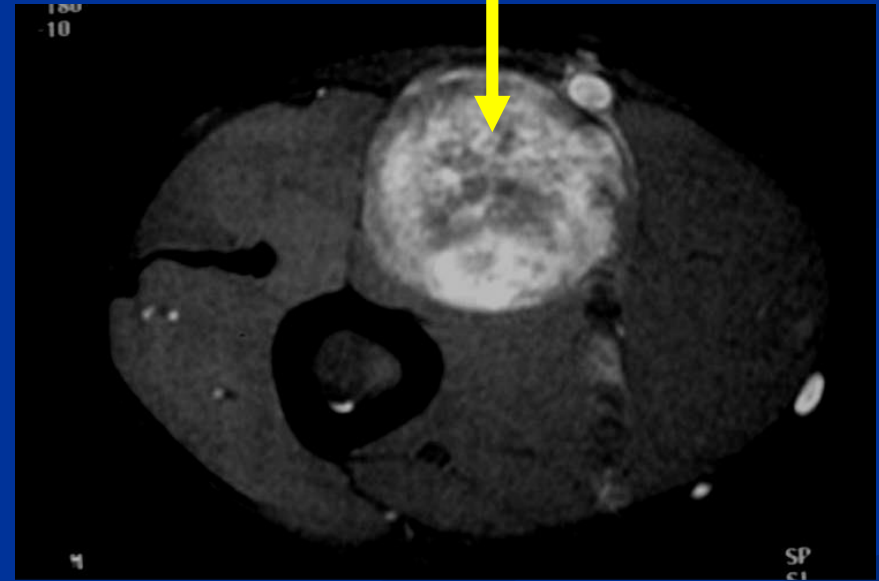
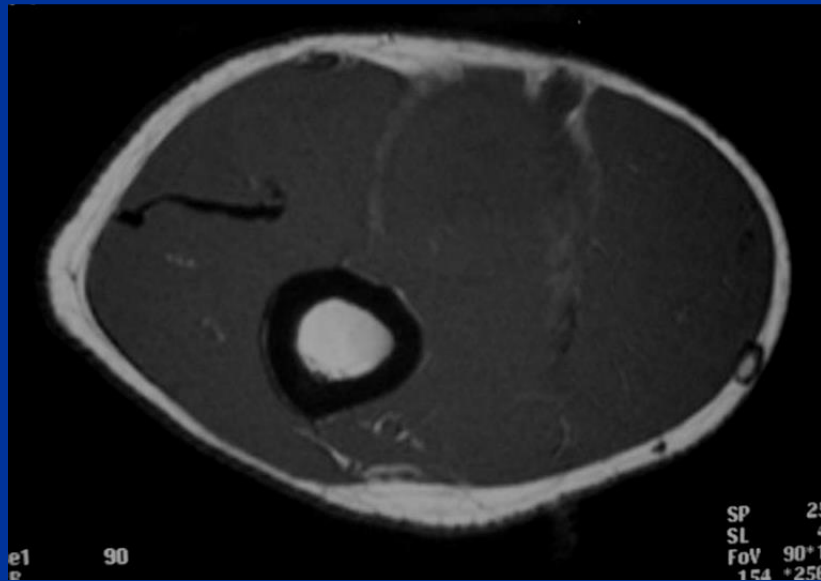


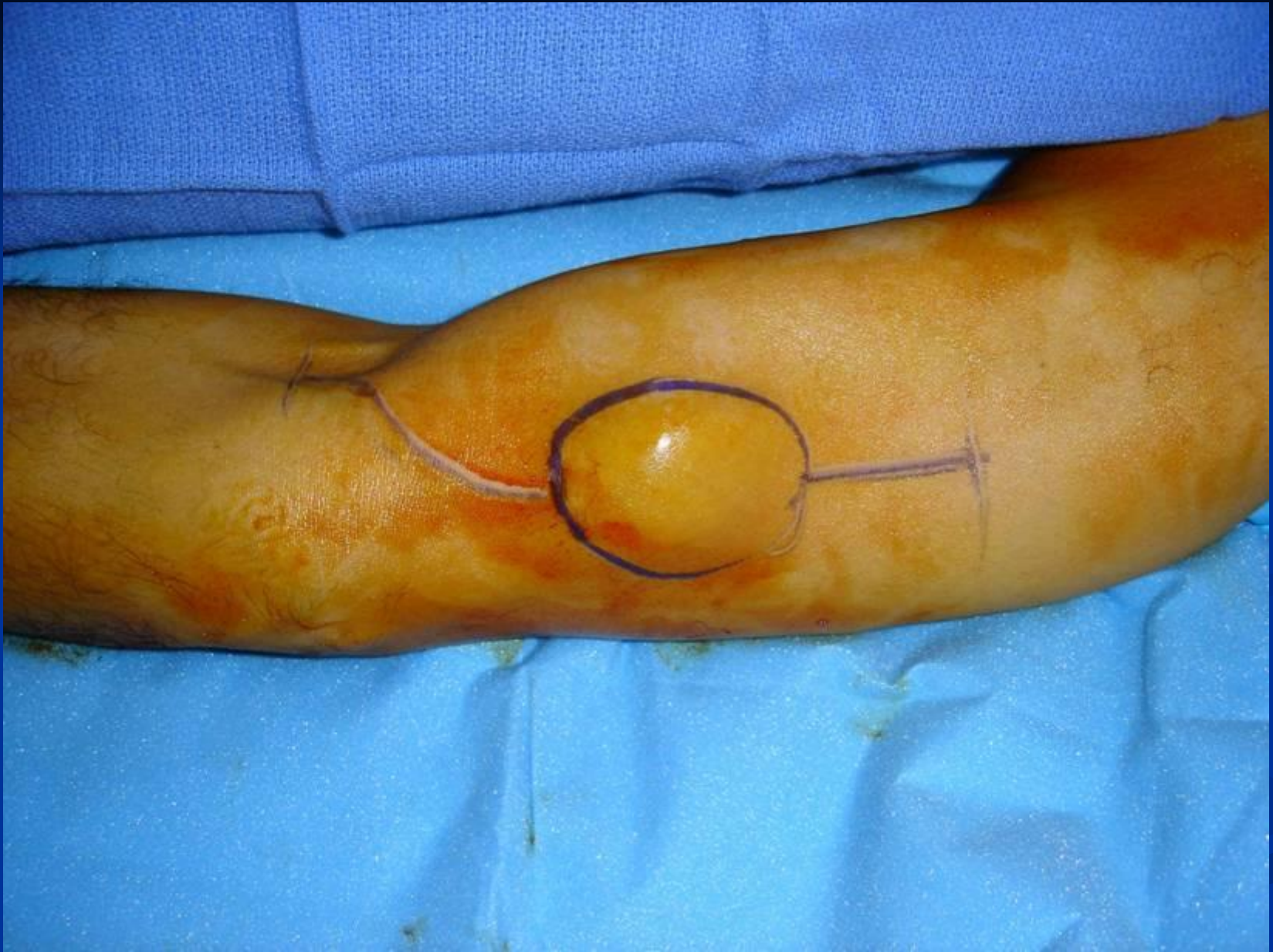
MRI Schwannoma: Nerve/Tubular Structure Entering Oblong Mass

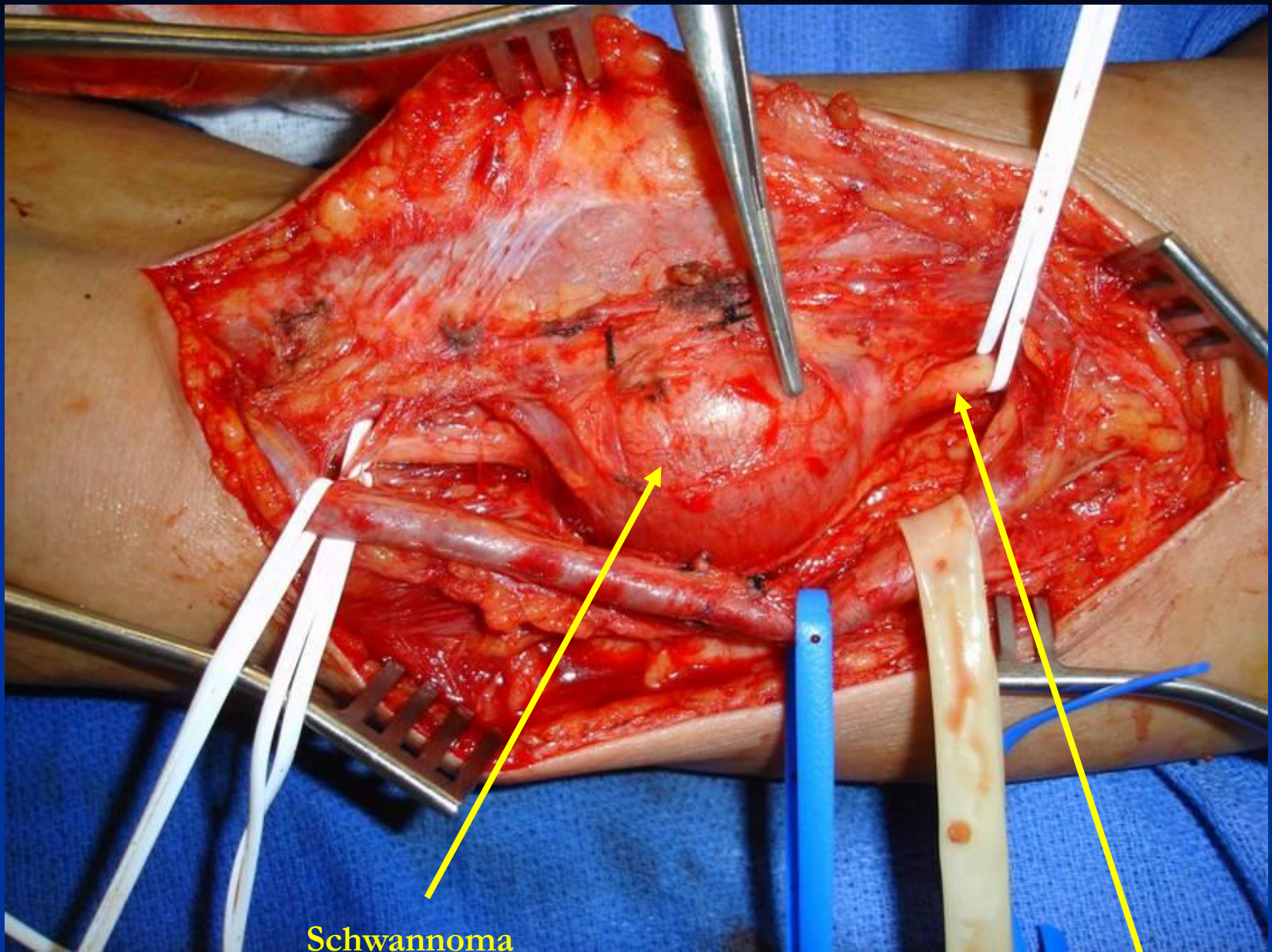


MRI: Schwannoma Median Nerve

Target Sign on T2 (arrow)
Hyperintense Area: Myxoid Antoni B
Split Fat Sign on T1

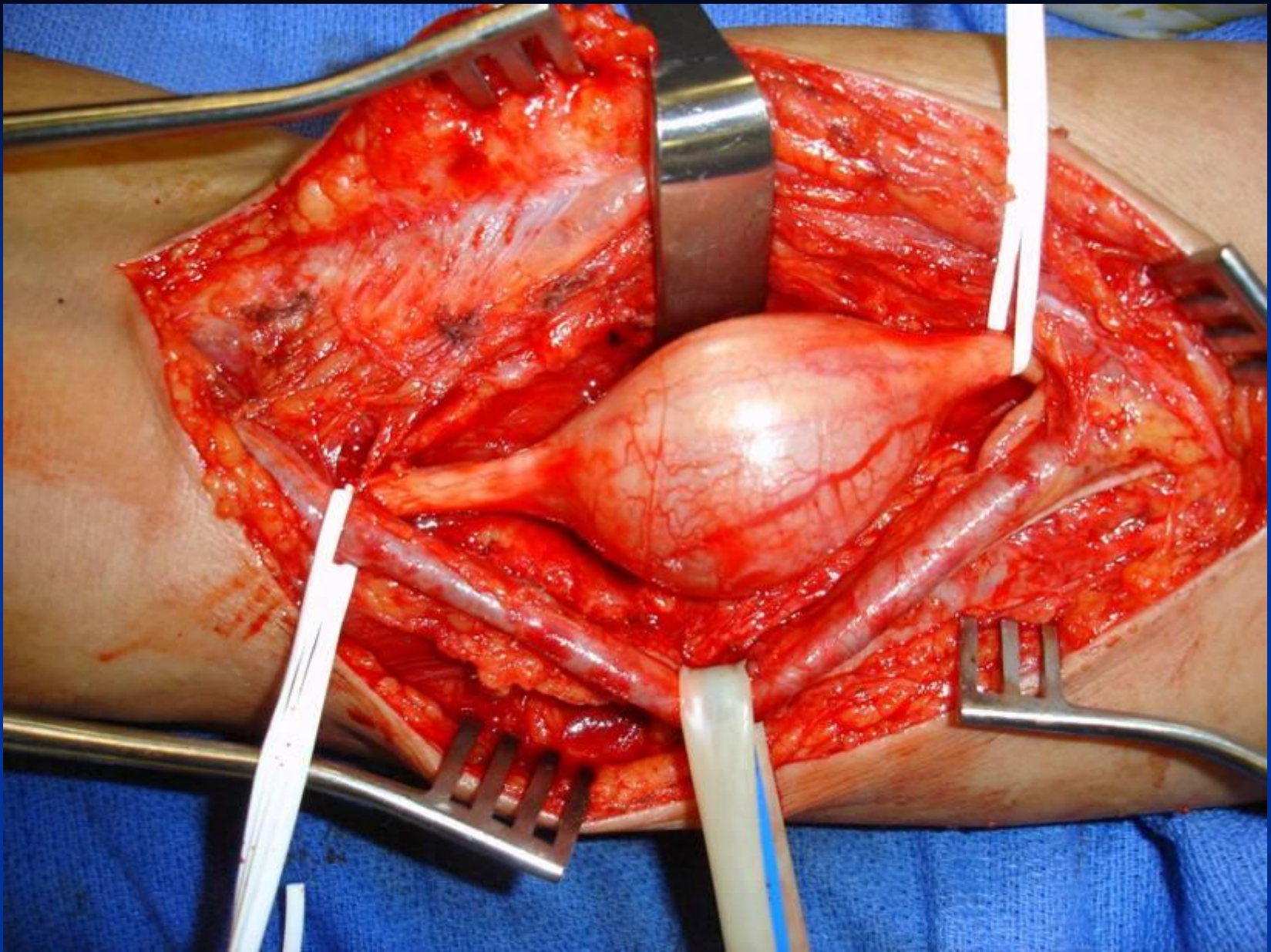


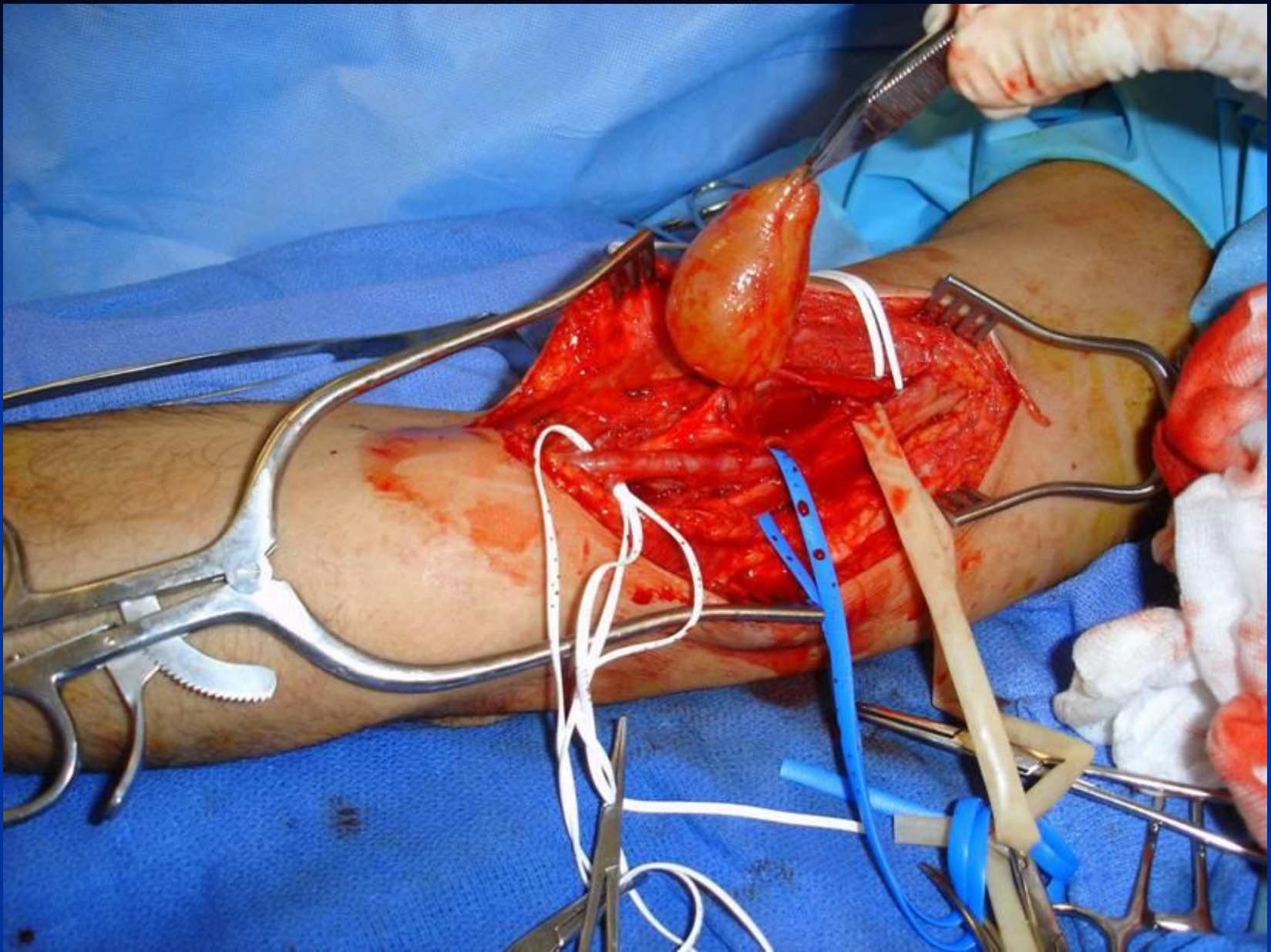




Schwannoma

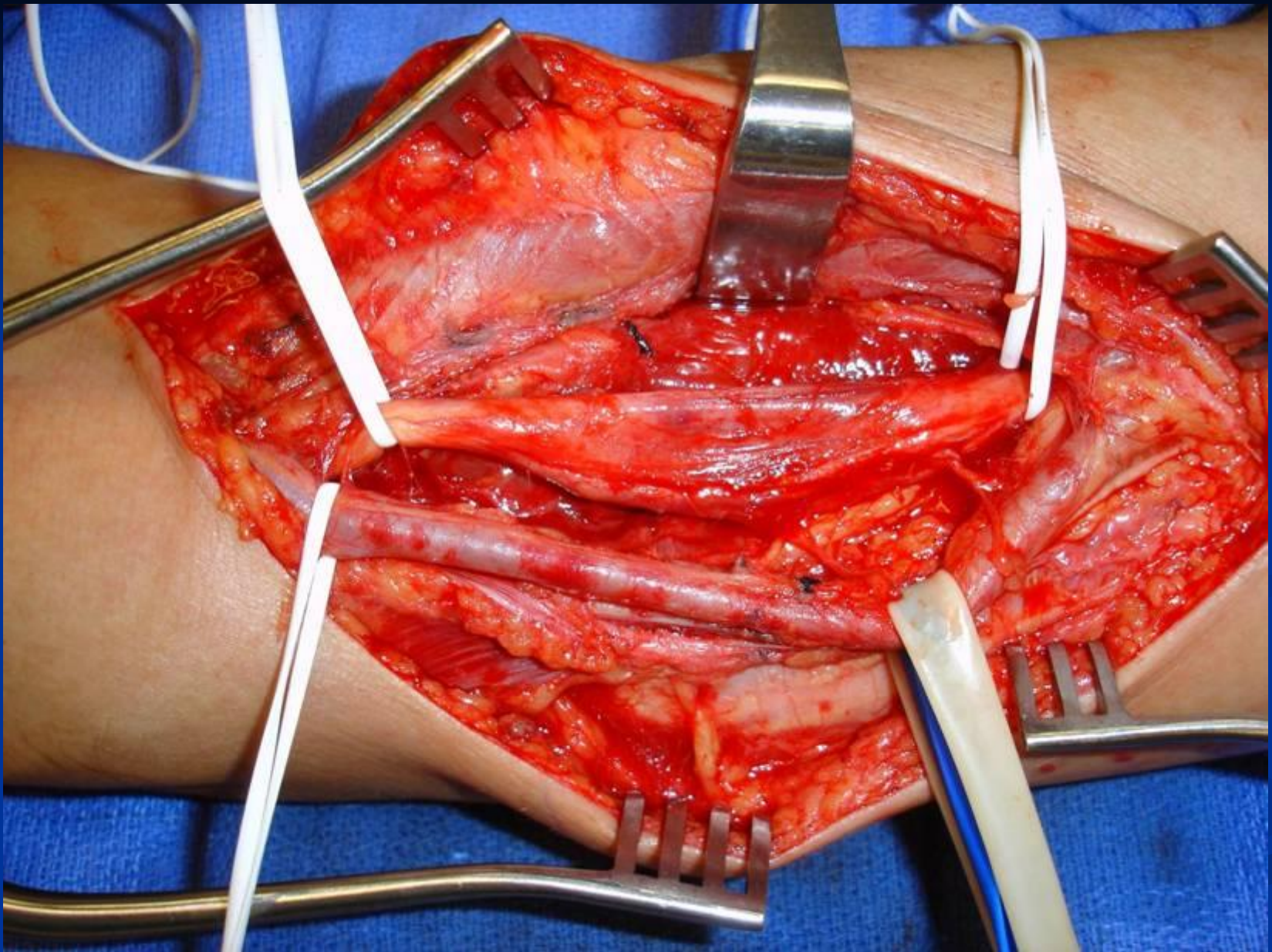
Median Nerve





Nerve Sheath Opened and Schwannoma Removed; Median Nerve Left Intact

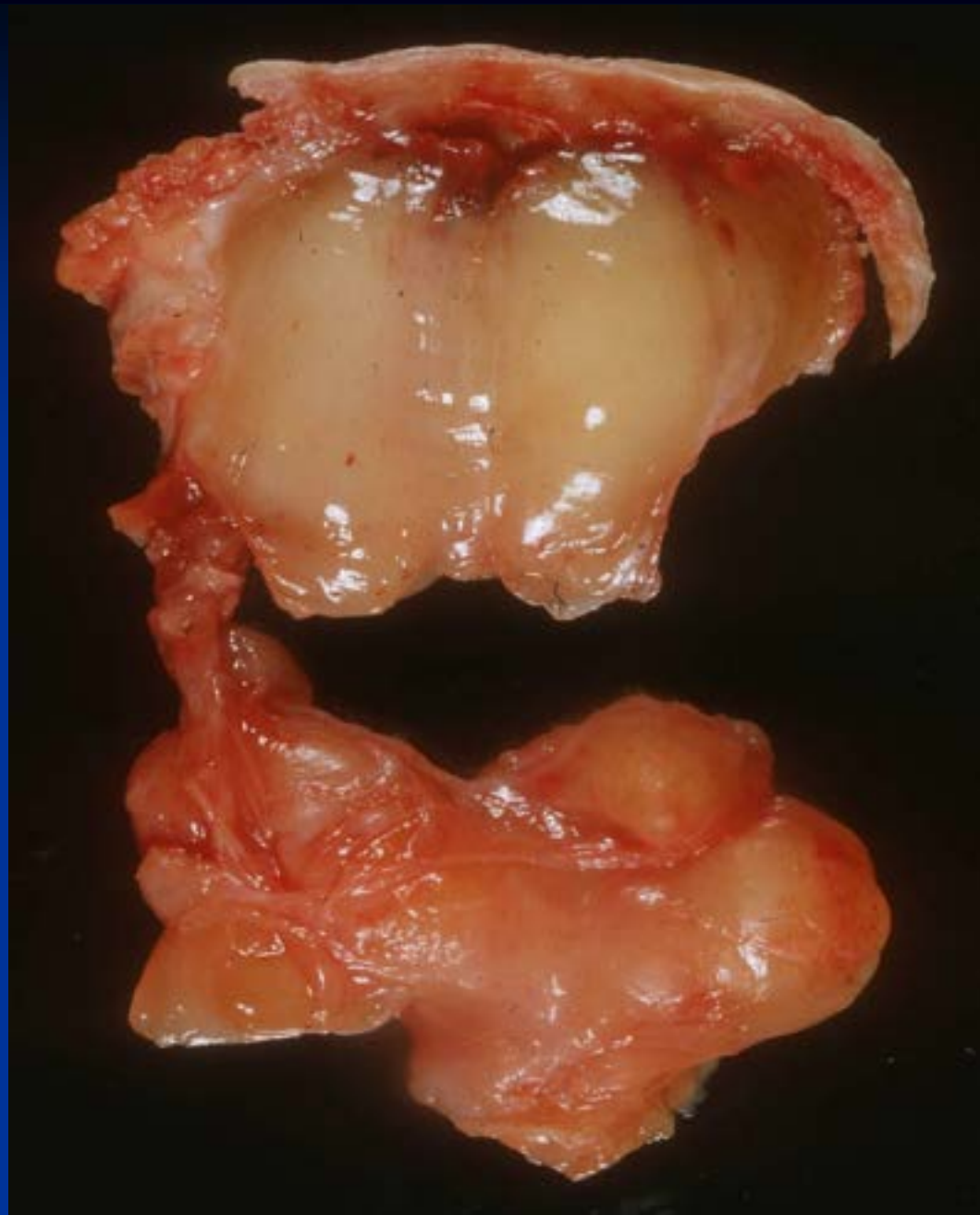




Median Nerve Intact



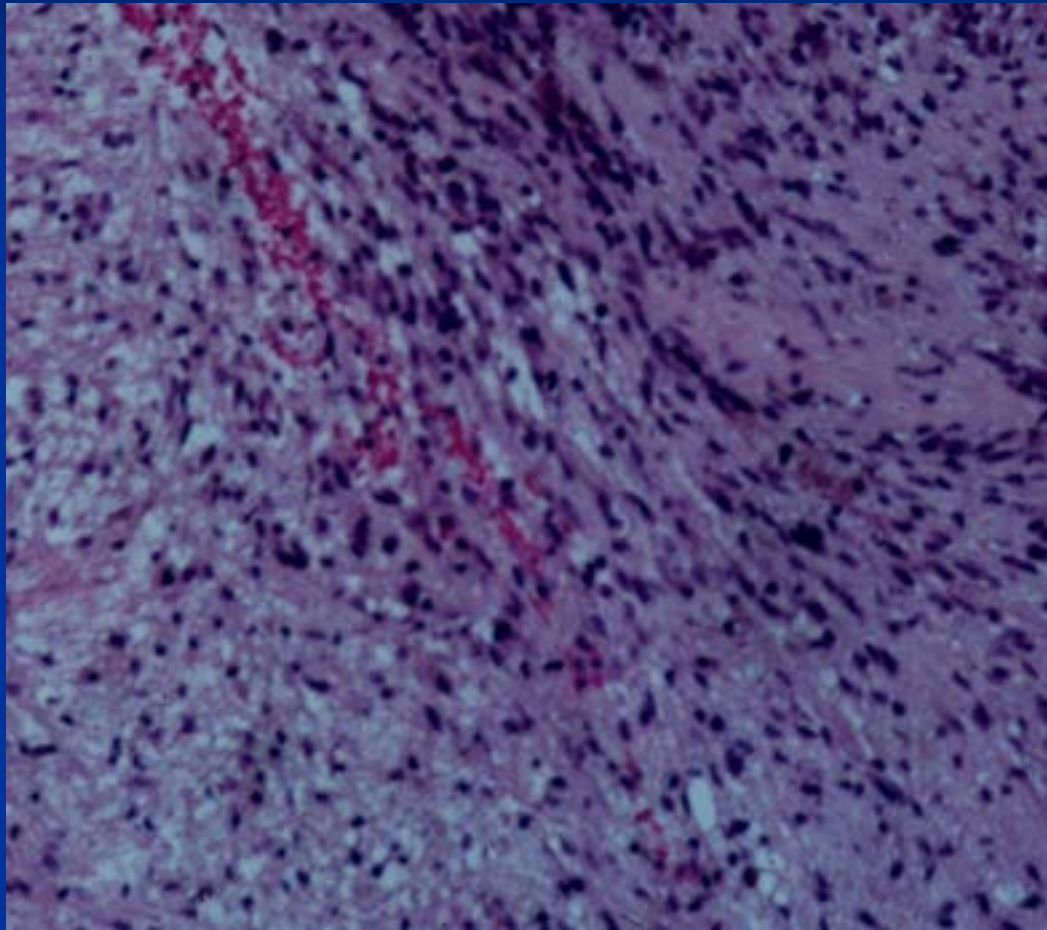






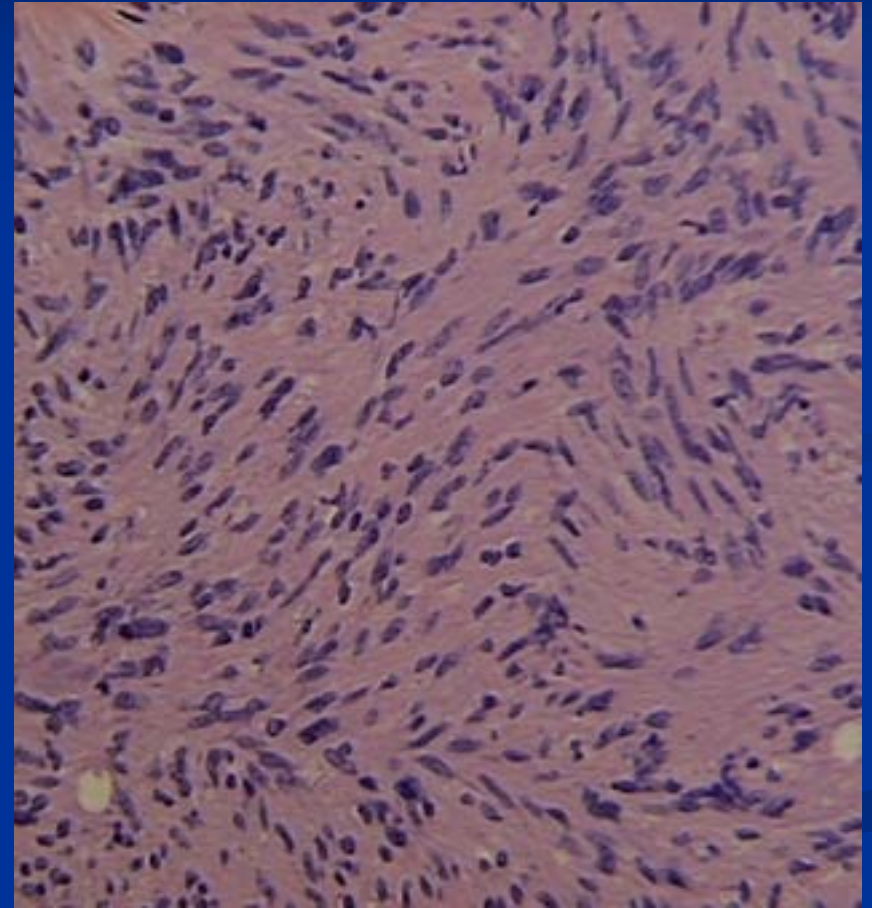
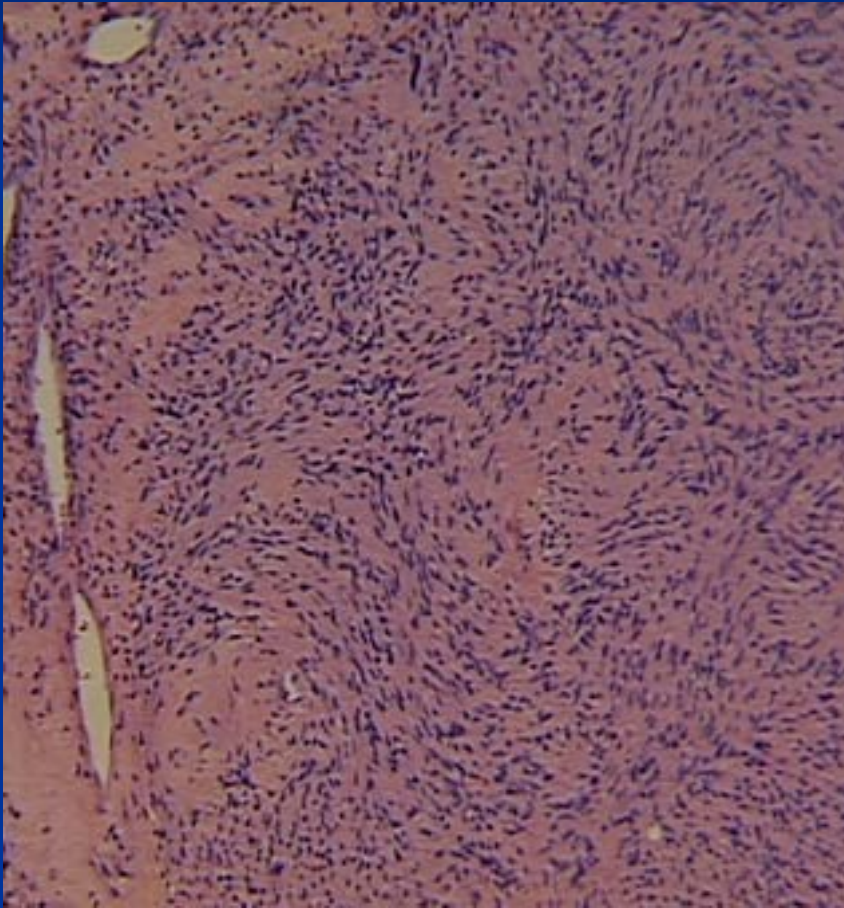
Schwannoma/Neurilemmoma

Antoni A: Cellular Area



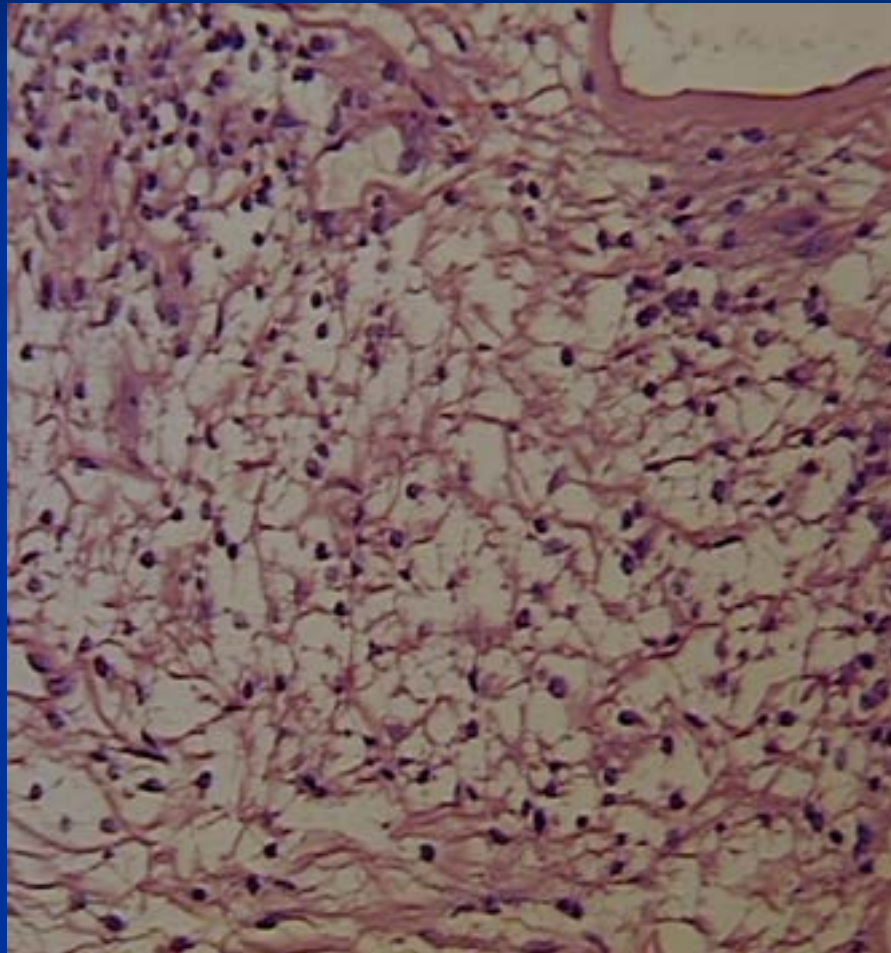
Pathology: Schwannoma

Antoni A: Cellular Area



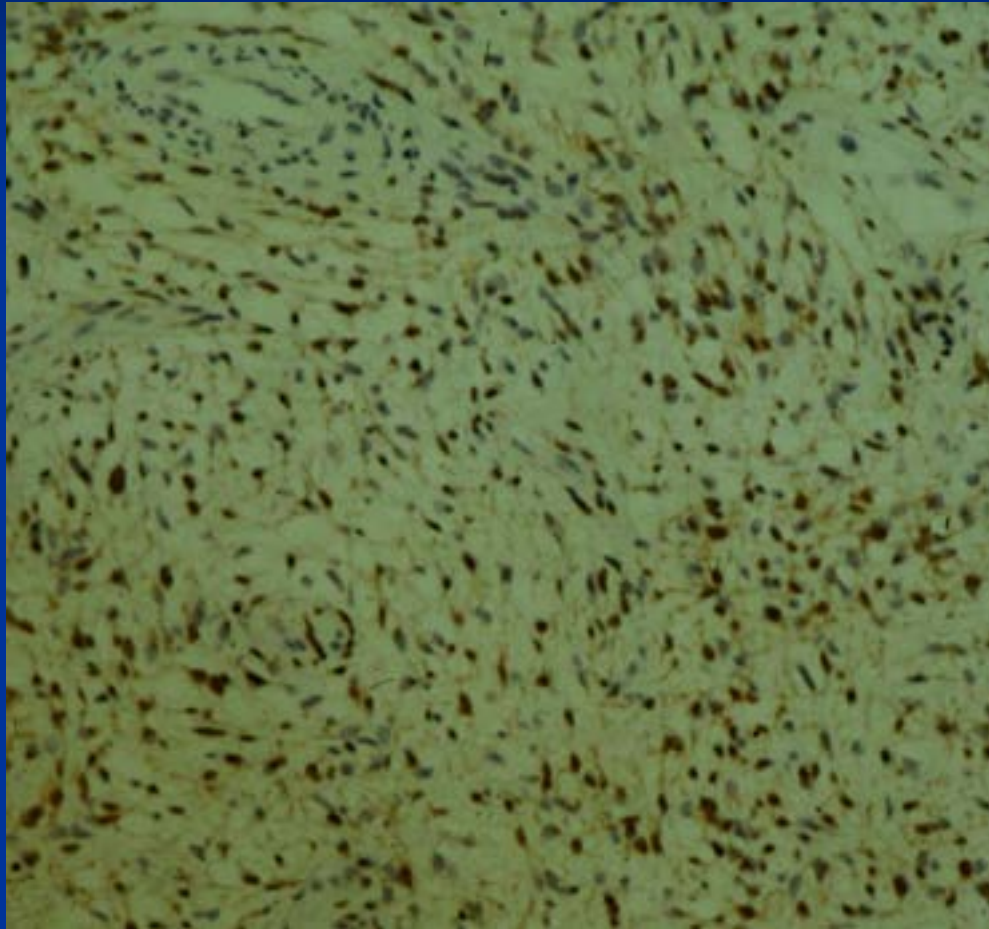
Pathology: Schwannoma

Antoni B: Myxoid Area



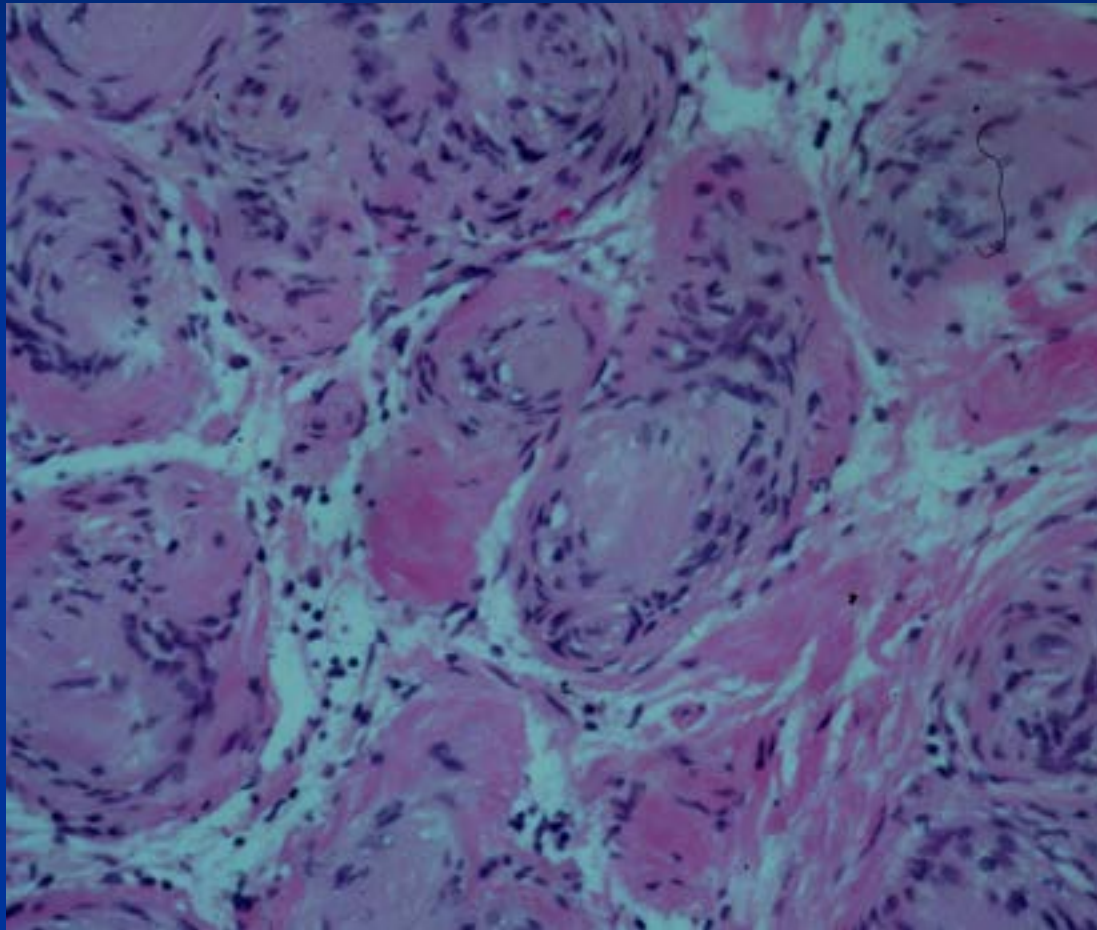
Pathology: Schwannoma

Antoni B: Myxoid Area



Pathology: Schwannoma

Verrucae Bodies



Treatment and Prognosis

- Treatment:
 - Surgical Excision: **Marginal** Excision; Spare nerve from excision
- Prognosis:
 - Benign tumor, does not metastasize
 - Local recurrence rare <1%
 - Malignant degeneration is very rare

Neurofibromas

- 5% of all benign soft tissue tumors
- 90% solitary lesions unrelated to neurofibromatosis
- **Ages:** Young adults 20-30 years old
- **Sites:** usually superficial nerves in the dermis or subcutaneous tissue
 - Rarely affect larger nerves
- **Clinical:**
 - Painless mass less than 5 cm; slowly growing
- Infiltrate beyond epineurium into nerve; Not encapsulated; cannot be separated from the nerve

Neurofibroma

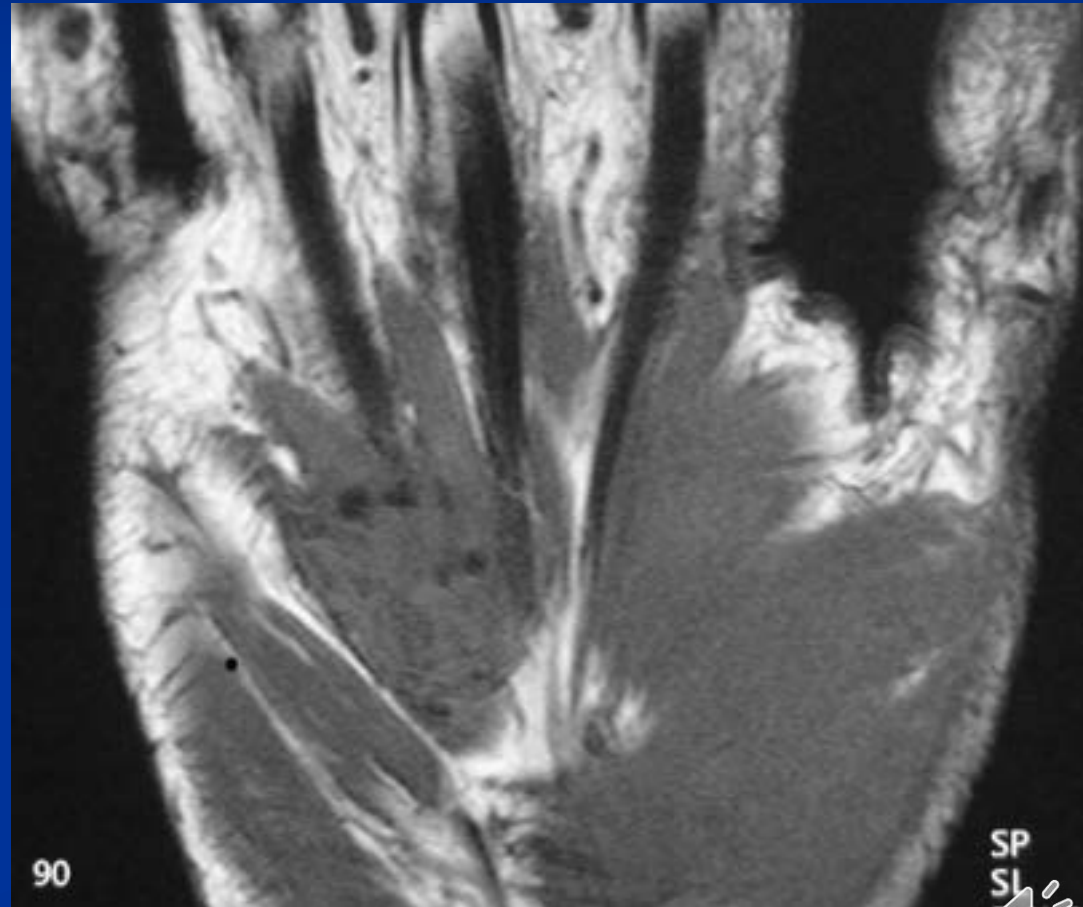
- **Pathology:** interlacing bundles of elongated cells with wavy, darkly staining nuclei and significant amounts of collagenization
- No Antoni A or Antoni B areas
- Solid and fleshy tumors
- Rarely removed surgically since they are not painful; Can not remove without removing nerve
- Solitary neurofibromas rarely if ever undergo malignant change.
- Malignant transformation usually occurs in the setting of neurofibromatosis; Persistently painful lesions usually indicate malignant transformation to a MPNST which are usually >5cm

Giant Cell Tumor of Tendon Sheath

- Localized nodular tenosynovitis
- Localized or diffuse proliferation of synovial-like cells, giant cells, inflammatory cells and xanthoma cells along tendon sheaths
- 3rd-5th decades
- Most common benign soft tissue tumor of the hand
- Can erode into bone and destroy it

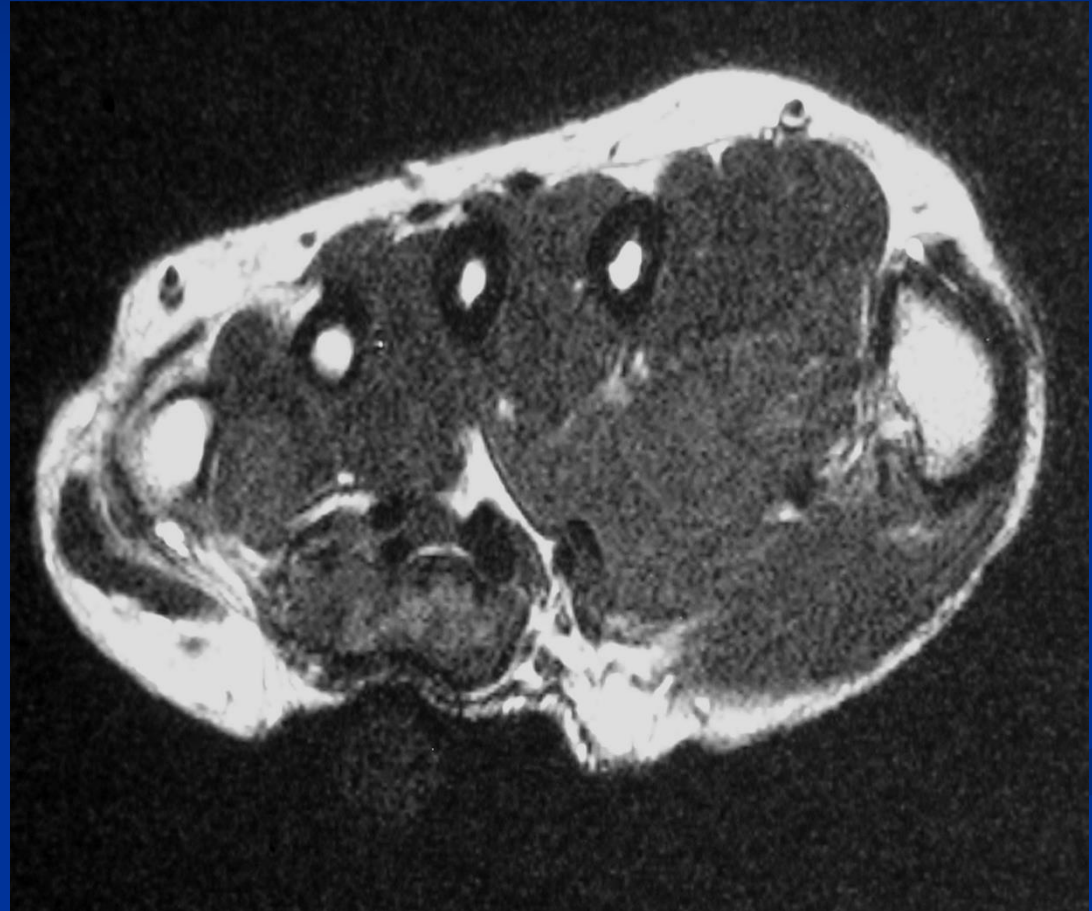
Giant Cell Tumor of Tendon Sheath

- Hand and wrist m/c locations (65-89%)
- Foot and ankle (5-15%)
- Pressure erosions in 15% (esp. ankles/feet)
- Ca^{+2} uncommon

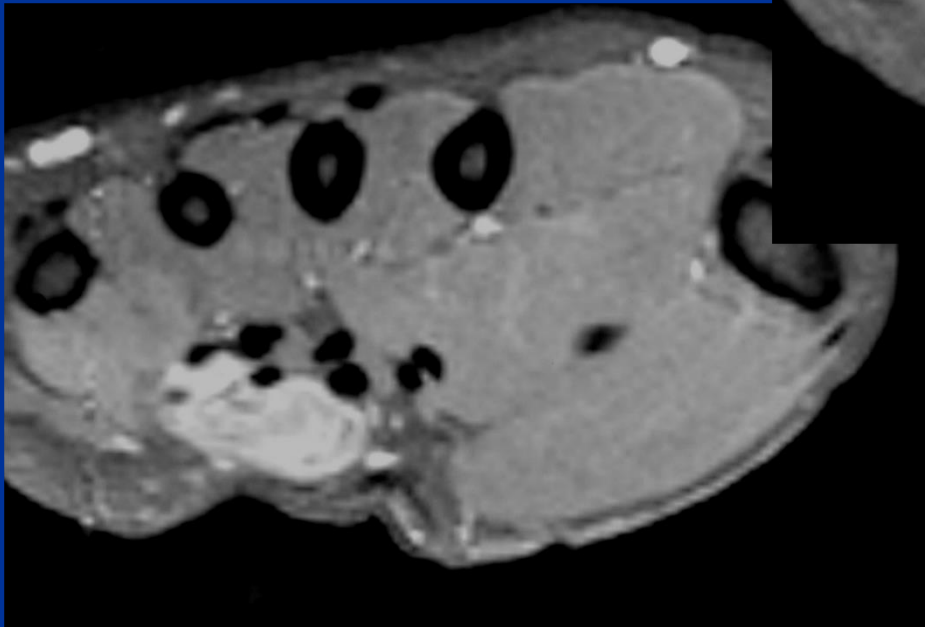
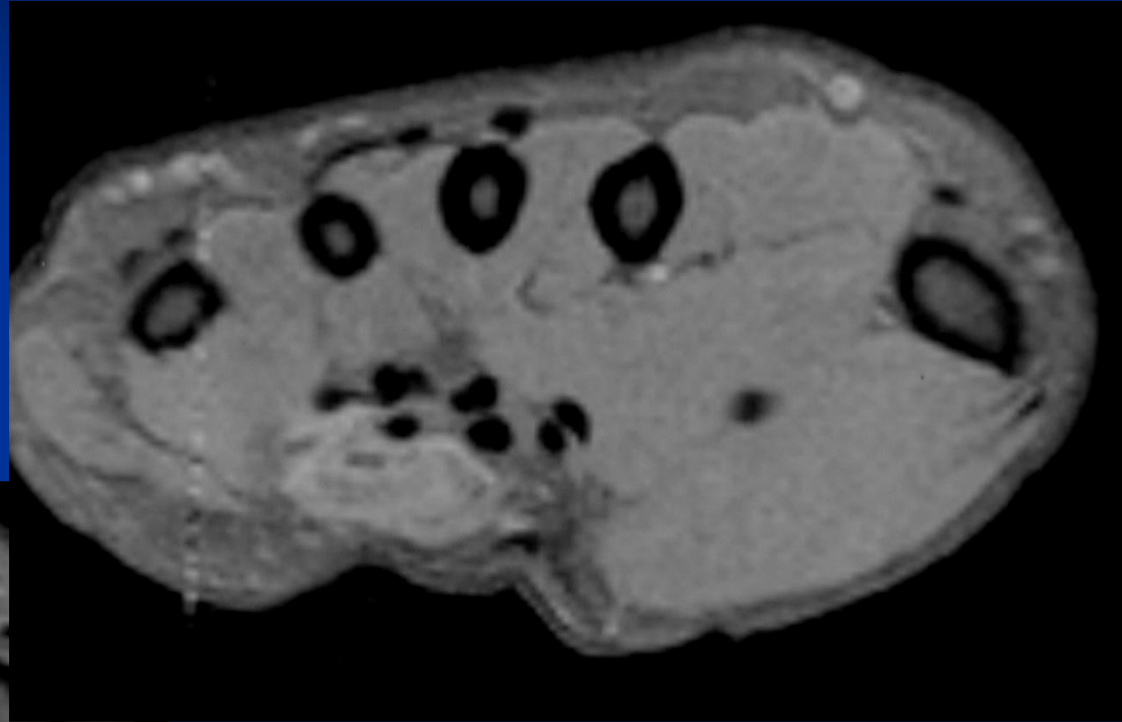


GCTTS

- Isointense to muscle T1
- Heterogeneous on T2 (Low and High Signal)
- Dark areas on T2: hemosiderin
- May bloom on gradient echo (hemosiderin)
- May demonstrate intense enhancement



Giant Cell Tumor of Tendon Sheath



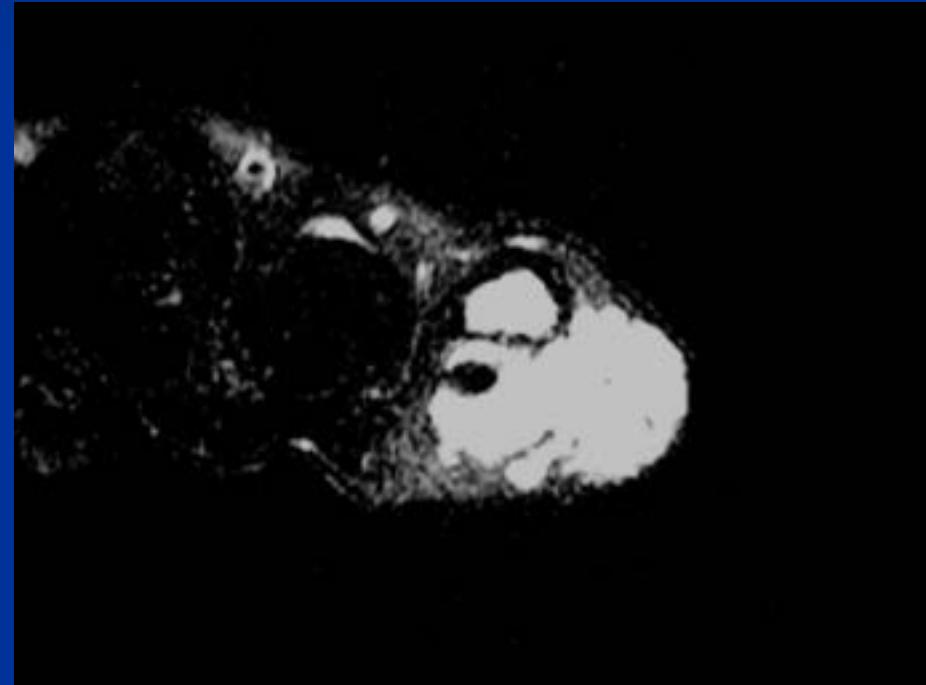
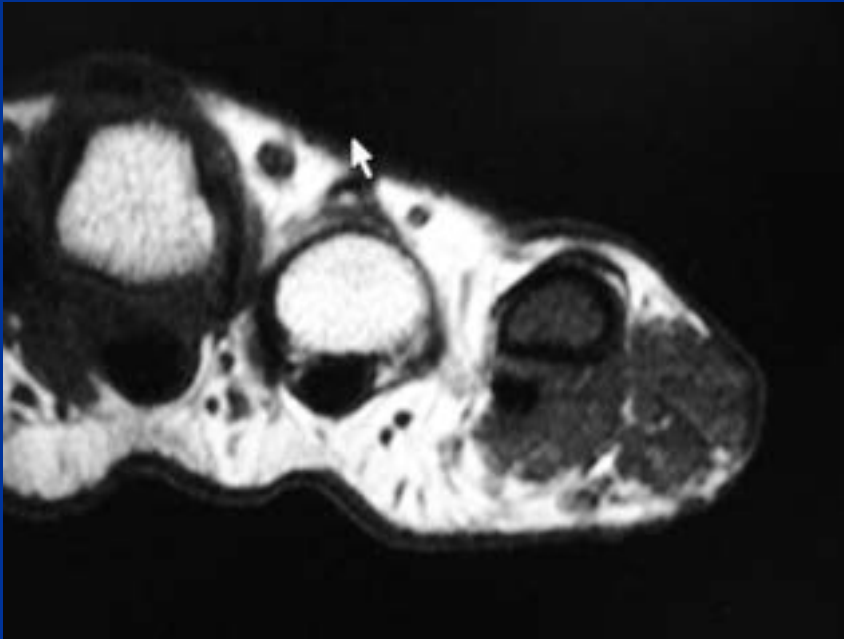
GCTTS Hand



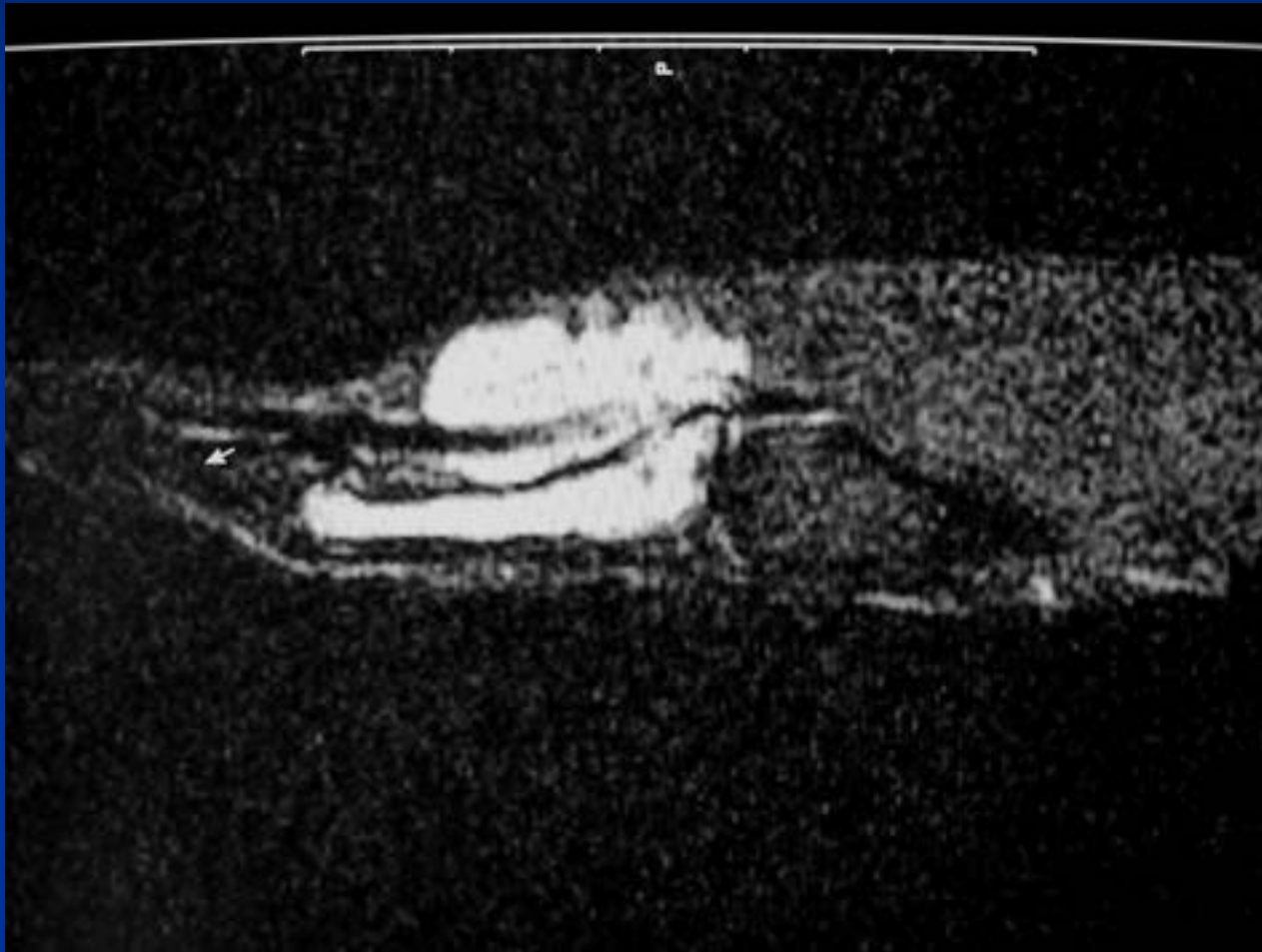
GCTTS of Hand



MRI T1: Giant Cell Tumor of Tendon Sheath



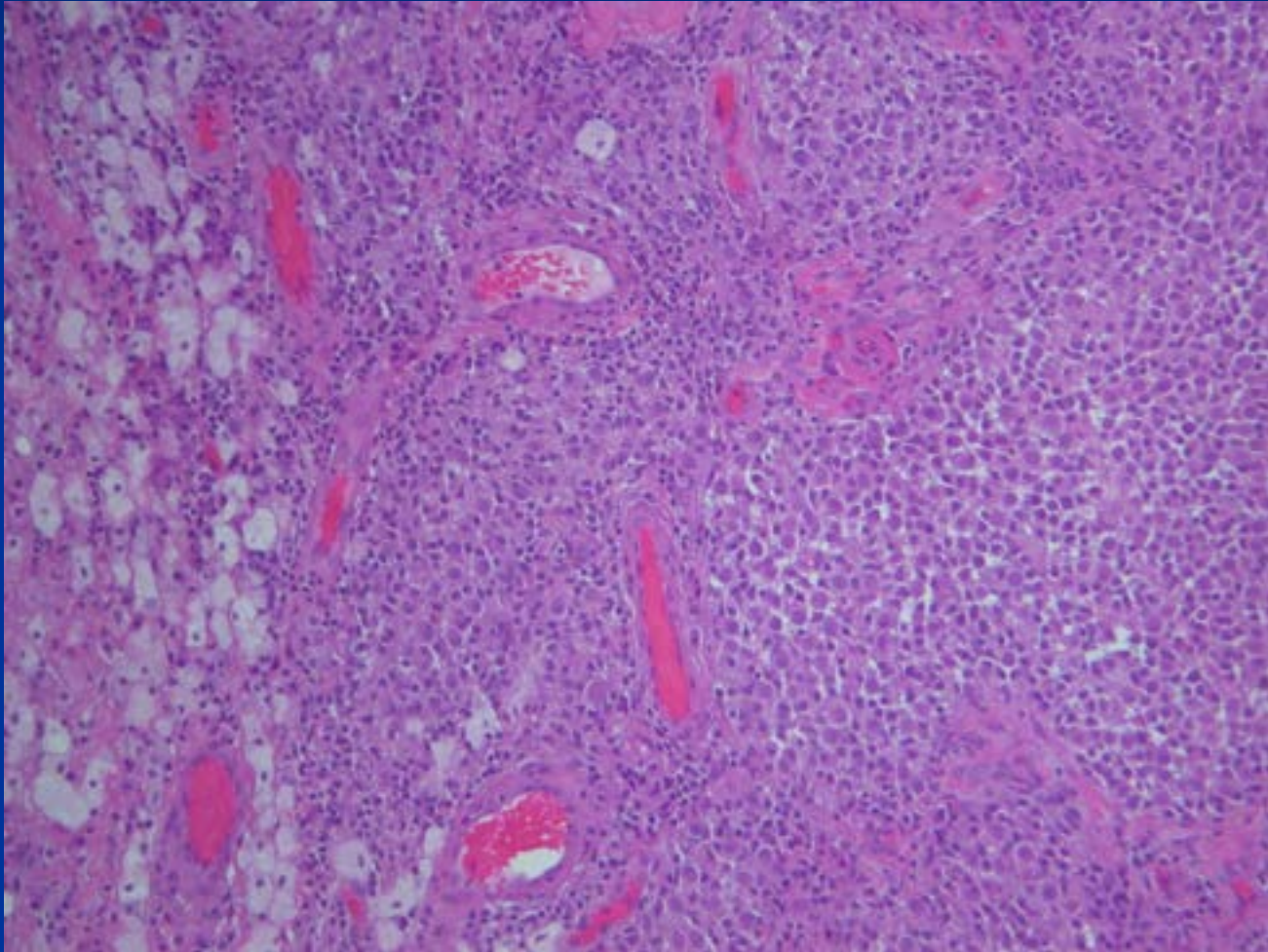
MRI T2: Giant Cell Tumor of Tendon Sheath of Hand



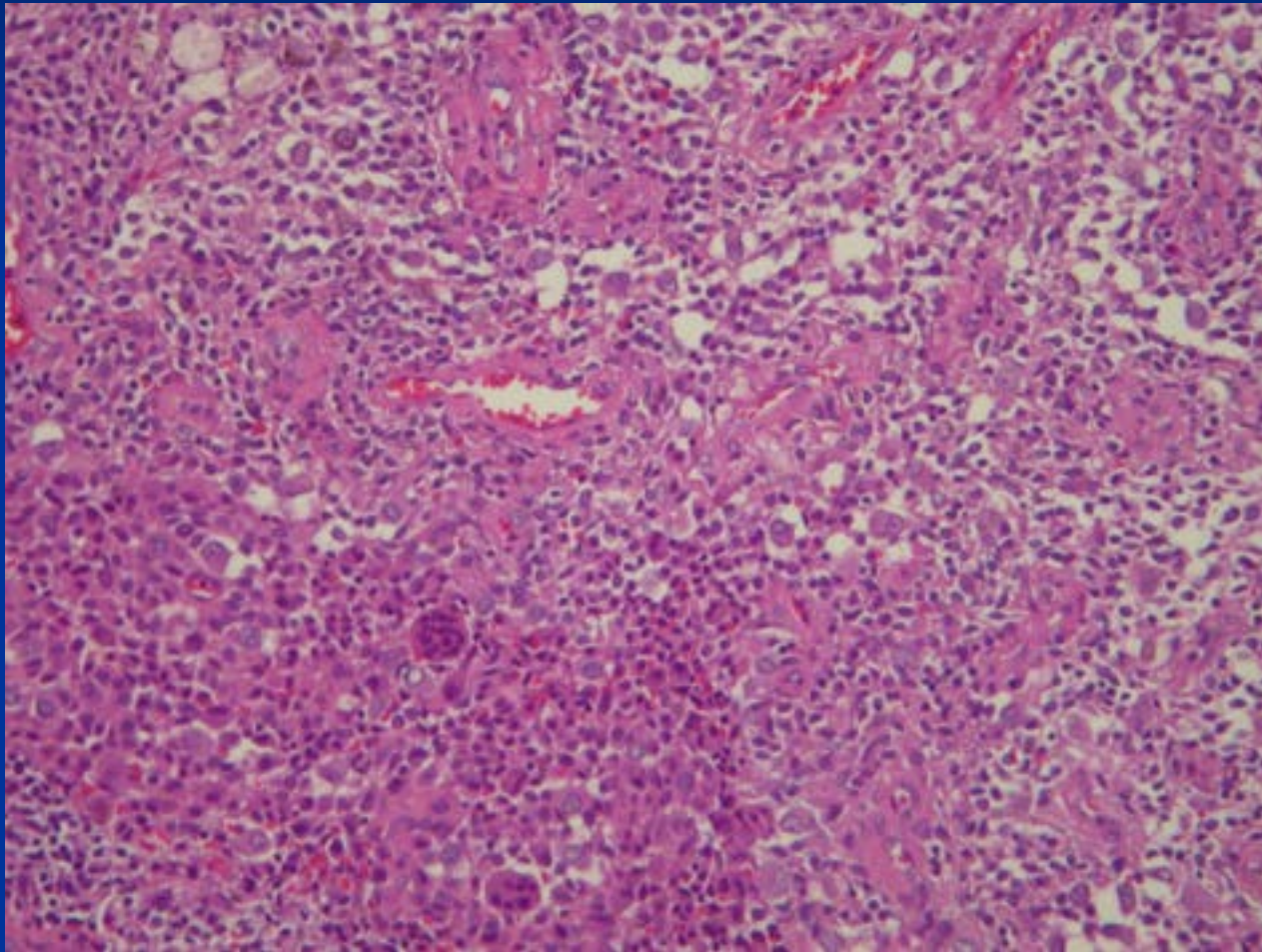
Pathology: Giant Cell Tumor of Tendon Sheath

- Localized or diffuse proliferation of synovial-like cells, giant cells, inflammatory cells and xanthoma cells along tendon sheaths
- Same histology as PVNS (Pigmented Villonodular Synovitis)

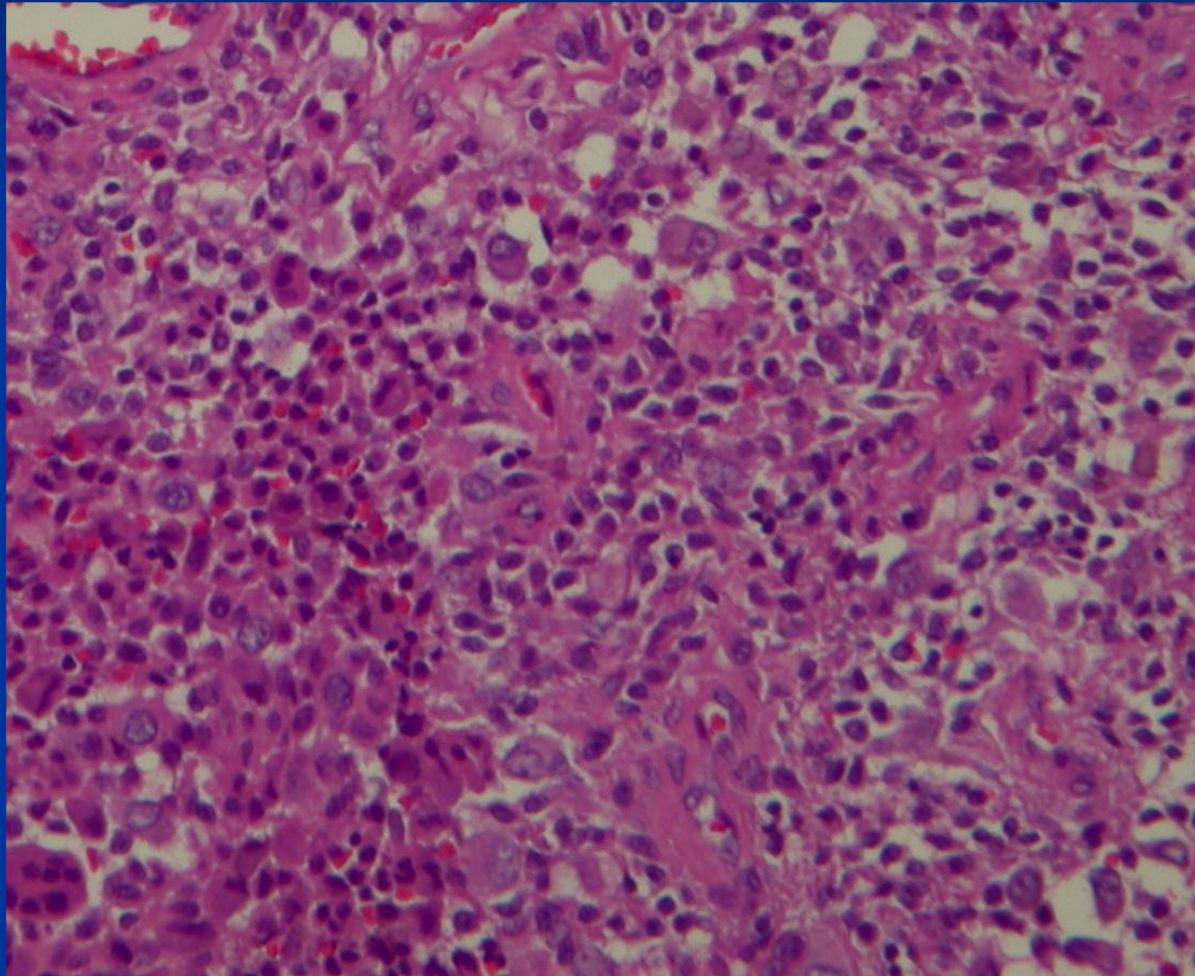
Pathology: Giant Cell Tumor of Tendon Sheath



Pathology: Giant Cell Tumor of Tendon Sheath

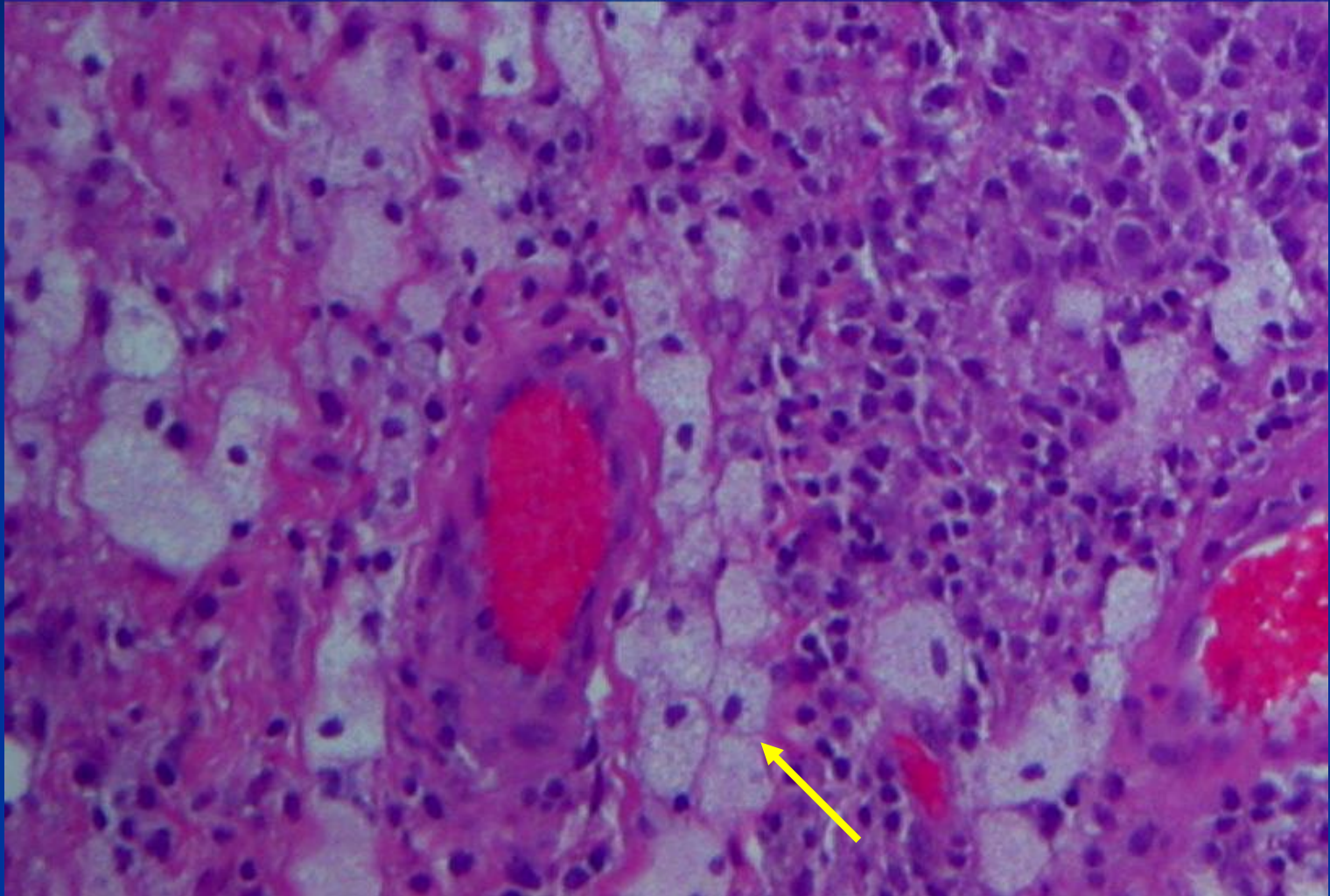


Pathology: Giant Cell Tumor of Tendon Sheath



Pathology: Giant Cell Tumor of Tendon Sheath

Foamy Histiocytes (arrow)



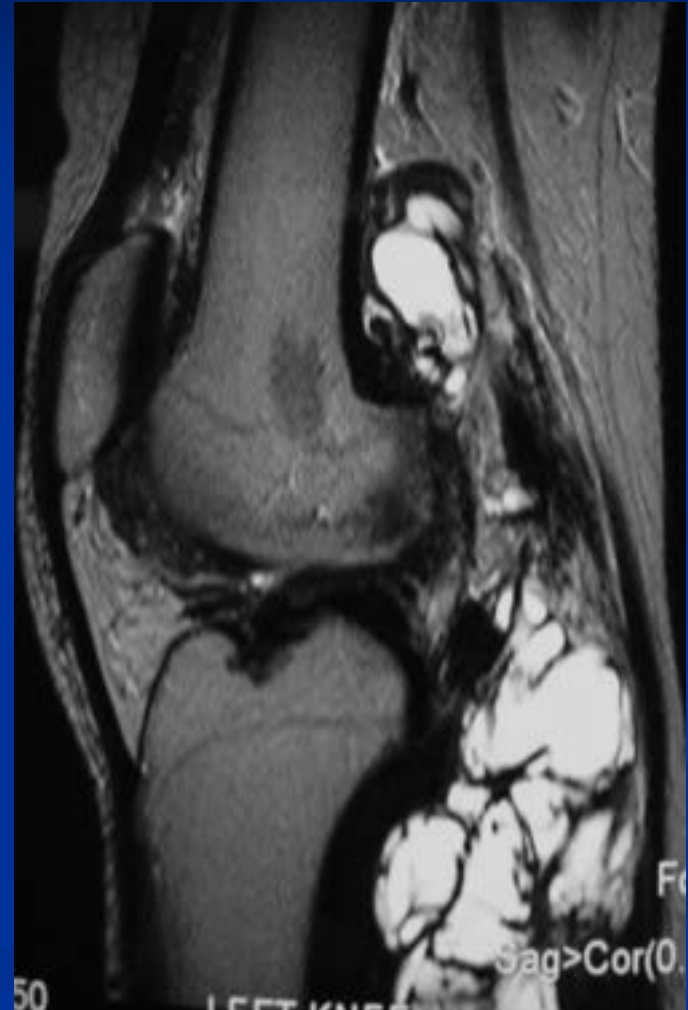
Pigmented Villonodular Synovitis

- Large joints (80% in knee)
- Joint effusion/pain
- Diffuse synovial process with pathology similar to GCTTS
- Synovial hyperplasia with multinucleated GC
- Intra- and extracellular hemosiderin



PVNS Imaging

- Effusion or mass may be seen on radiographs
- Erosions (50%) m/c in smaller tighter joints, especially in hip (93%) and shoulder (75%)
- Erosions are geographic/lytic + sclerotic rim
- Joint space usually preserved
- Ca^{+2} very rare
- Possible increased attenuation on CT (hemosiderin)



PVNS Imaging

- Heterogeneous synovial mass extending away from joint on MR
- SI $\leq$ muscle on T1 and T2
- Scattered areas of high SI on T2 possible
- Cystic lesions uncommon (10%)
- Blooming on gradient images (hemosiderin)



Thank You!

