Benign Soft Tissue Tumors

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

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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

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Lipoma

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

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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

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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

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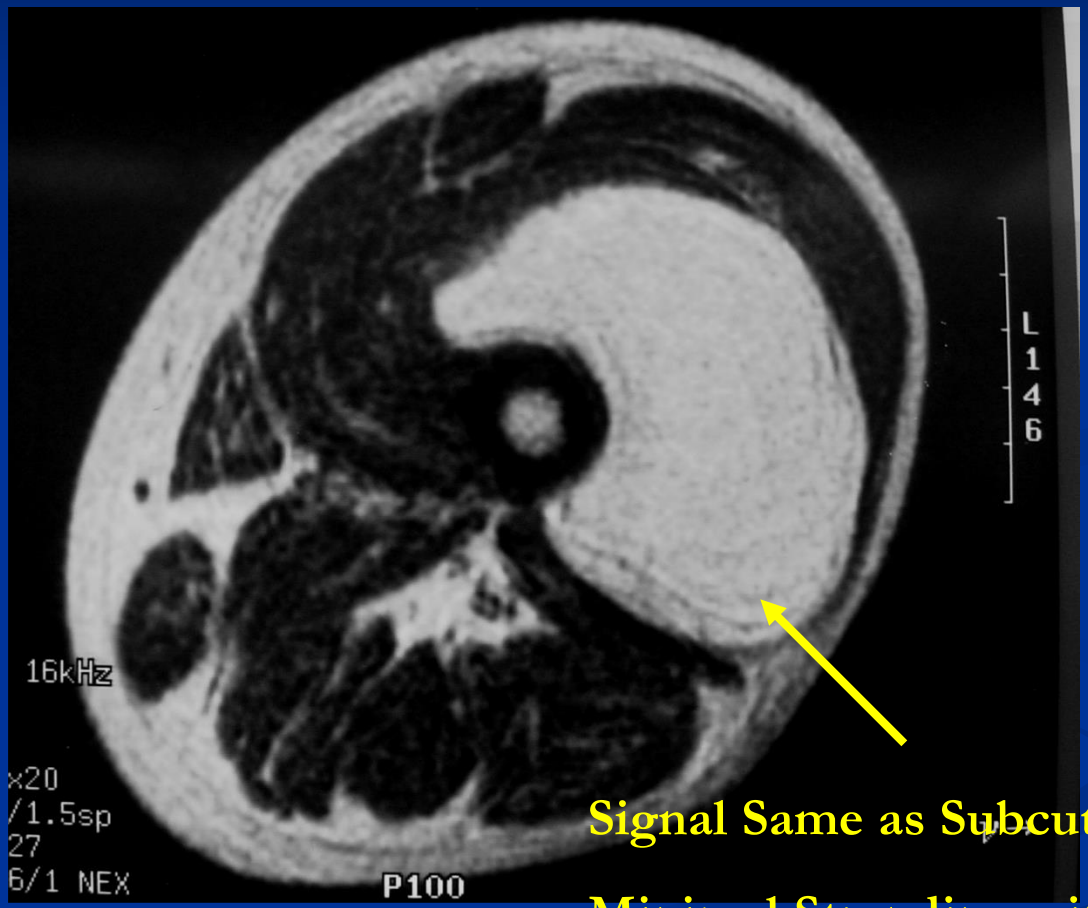
MRI T1: Intramuscular Lipoma of Thigh
High Signal on T1



Signal Same as Subcutaneous Fat

Minimal Stranding within Tumor

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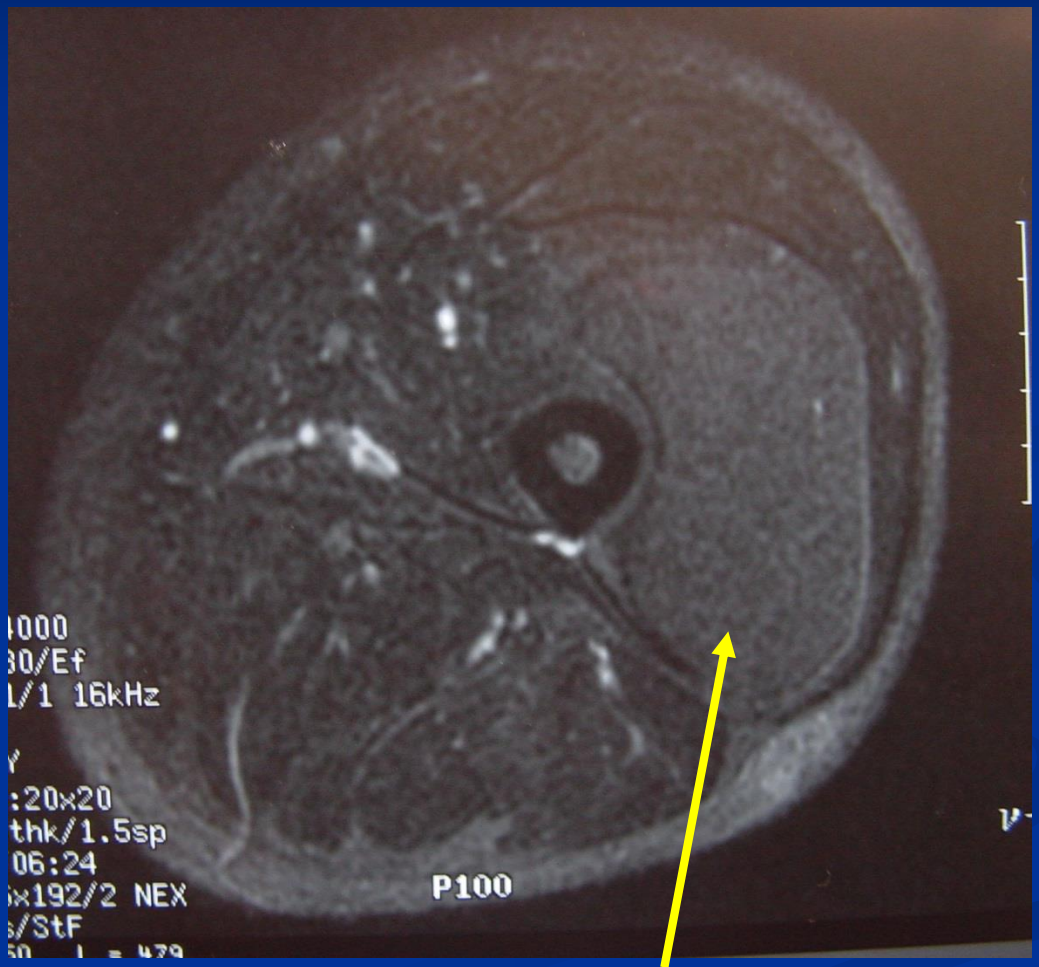
MRI T2 Fat Suppressed Image



Fat in Lesion Suppressed Same as Subcutaneous Fat

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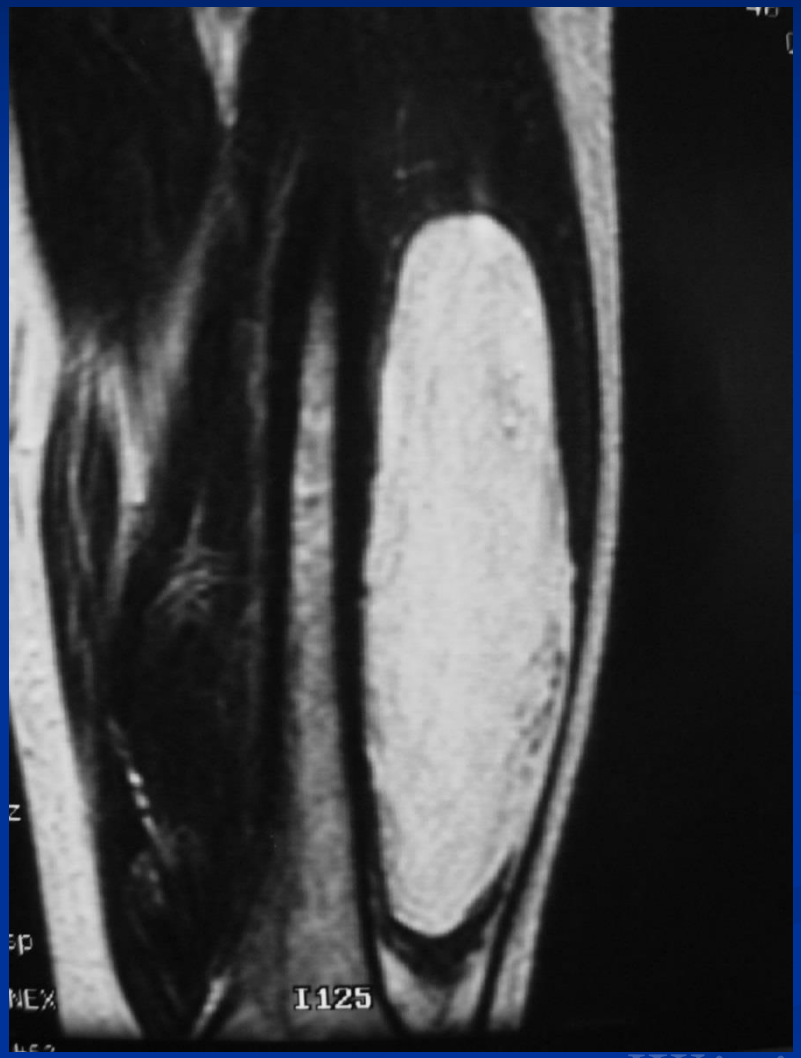
MRI T1: Lipoma of Posterior Thigh



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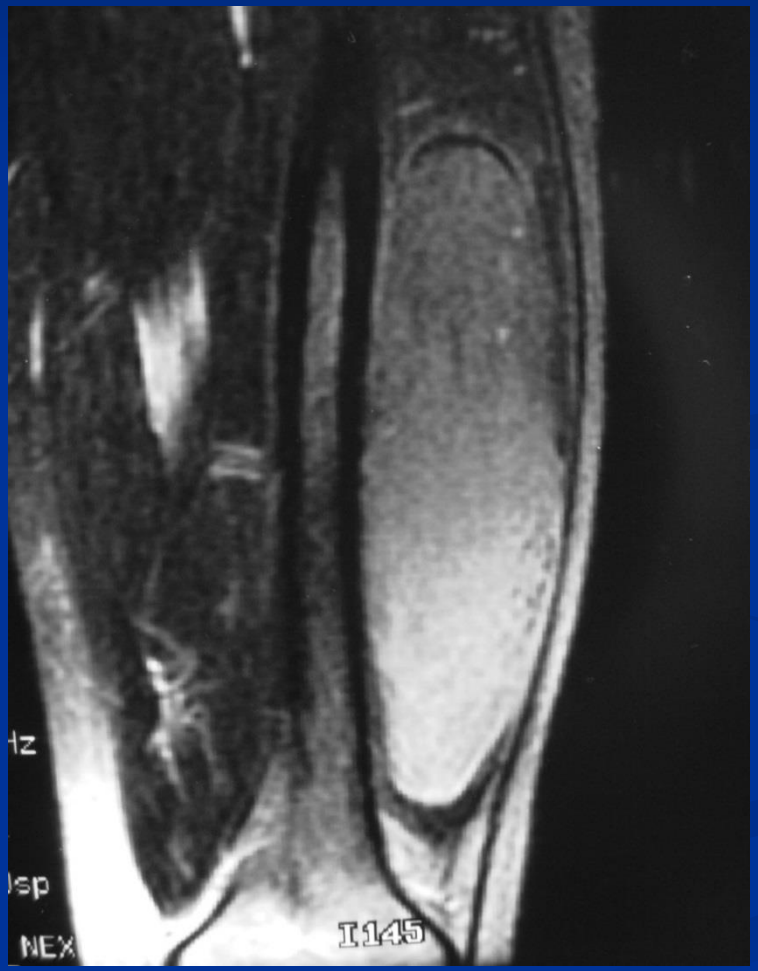
MRI T2 (not Fat Suppressed)
Fat is Intermediate Signal on T2
Low Signal on Fat Suppressed



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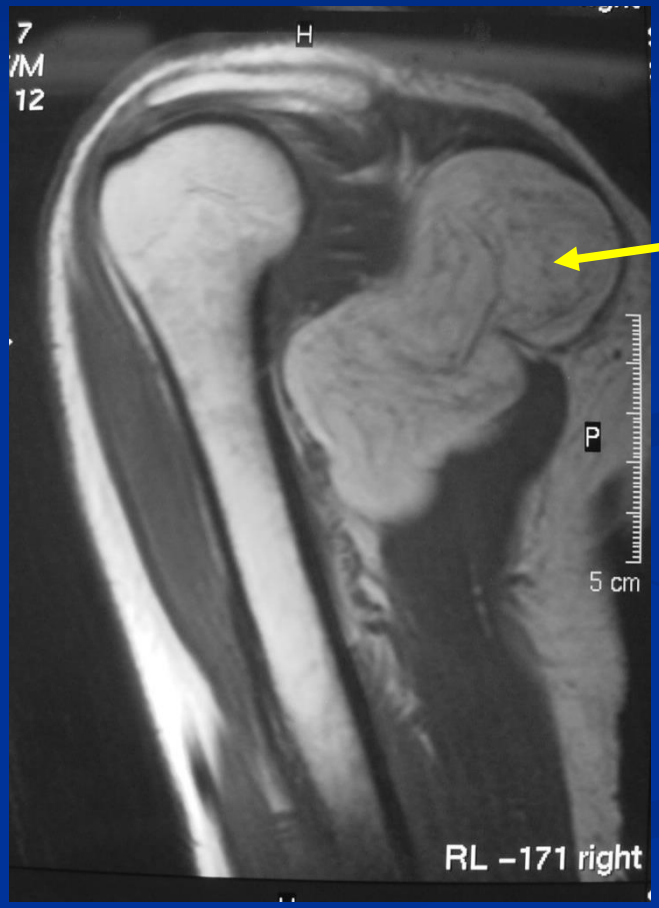
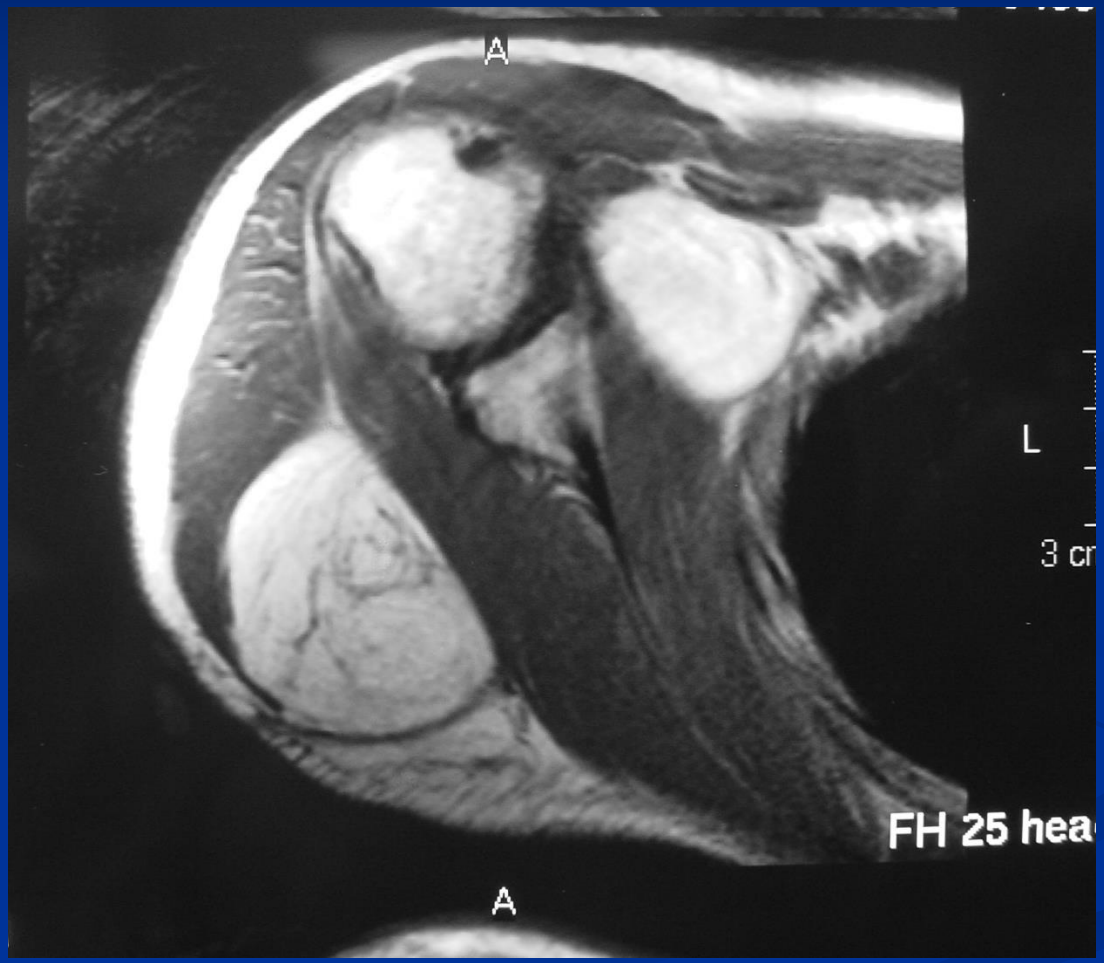
MRI T1: Lipoma Right Shoulder Fibrolipoma: Notice Stranding



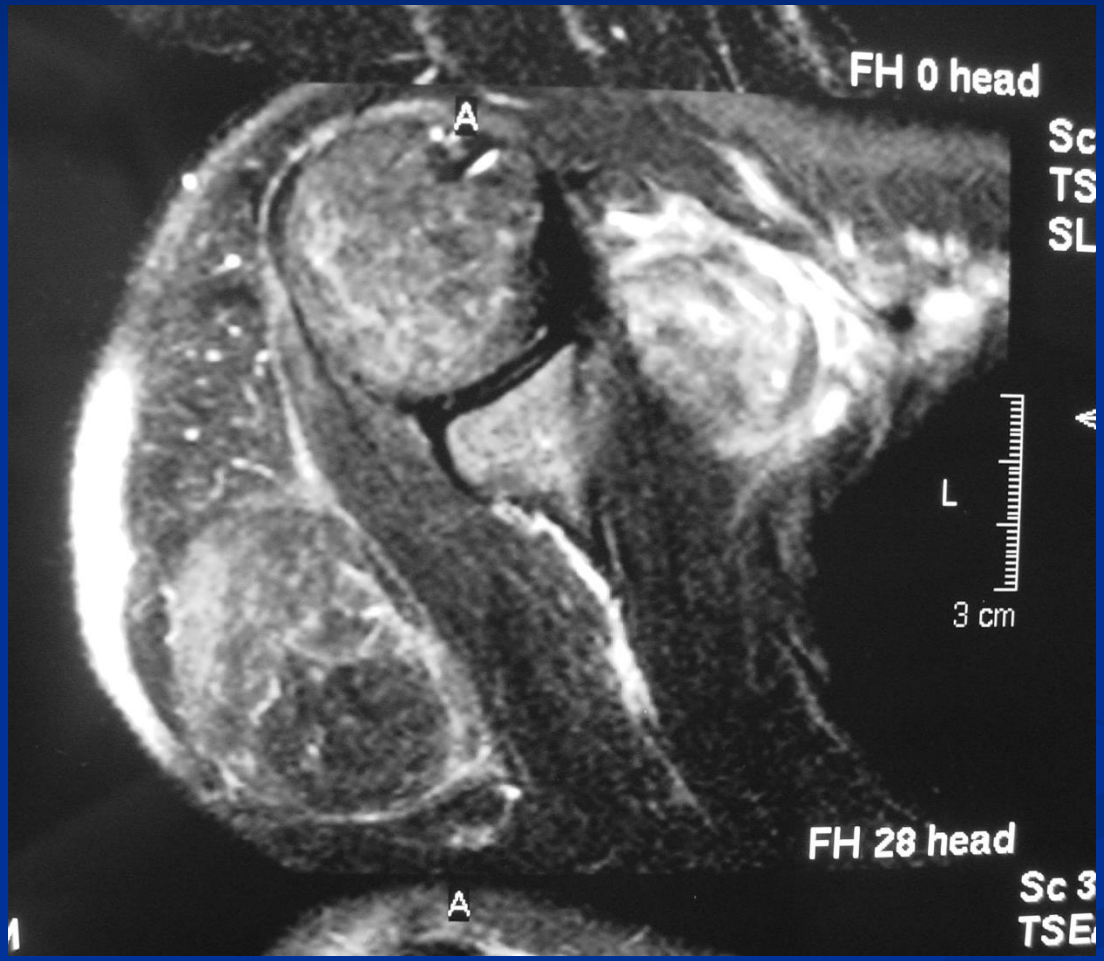
Stranding within Tumor indicates Collagenous/Fibrous Septae

Can Also Occur with Low Grade Liposarcomas/Atypical Lipomas

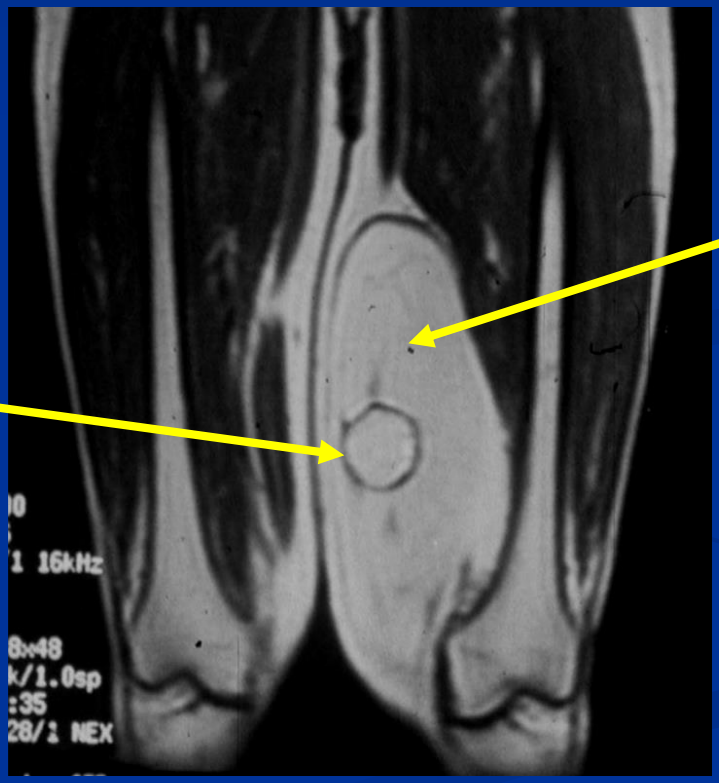
MRI T1: Lipoma of Shoulder



MRI T2 Fat Suppressed

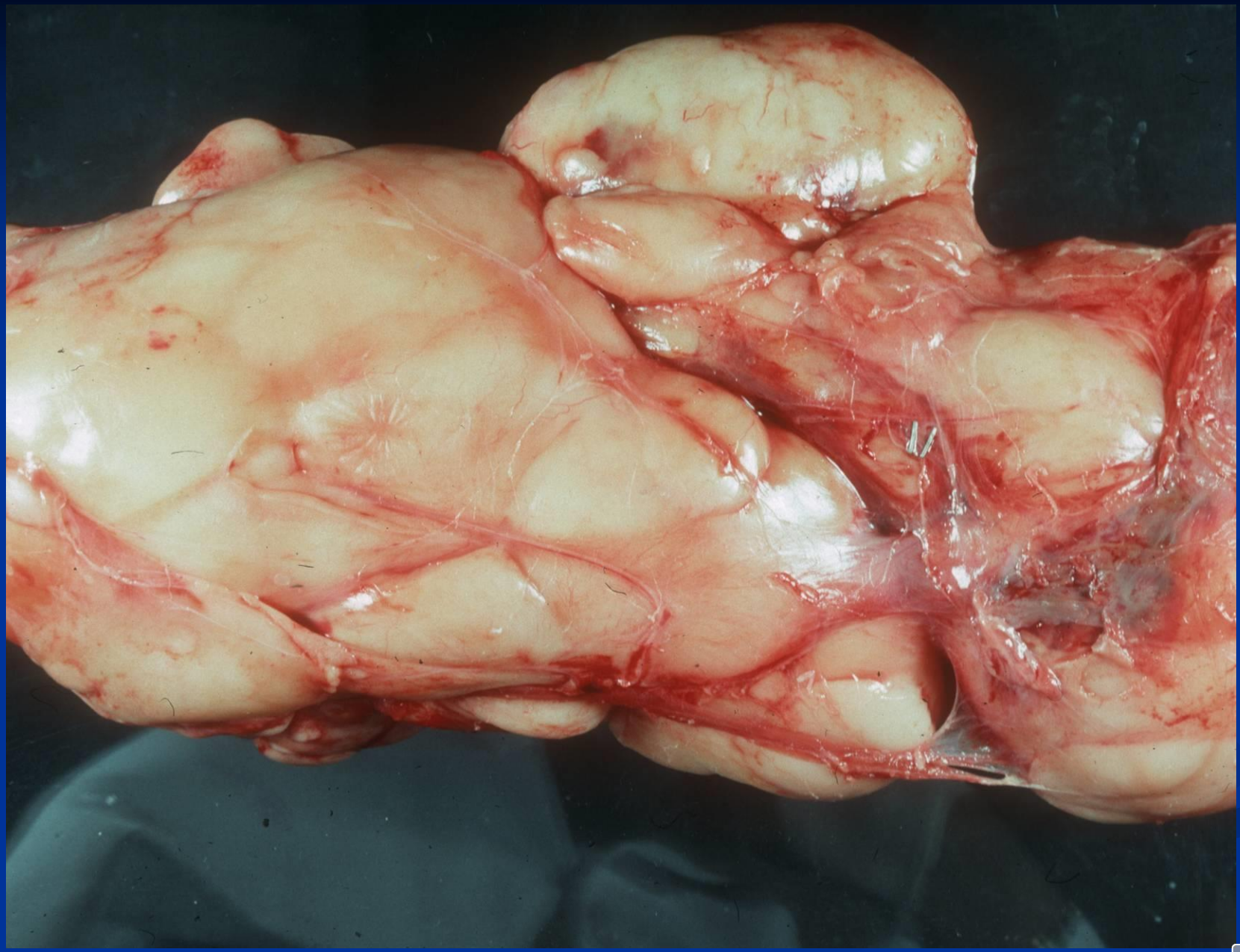


MRI T1: Large Lipoma Left Thigh with Small Area of Fat Necrosis



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



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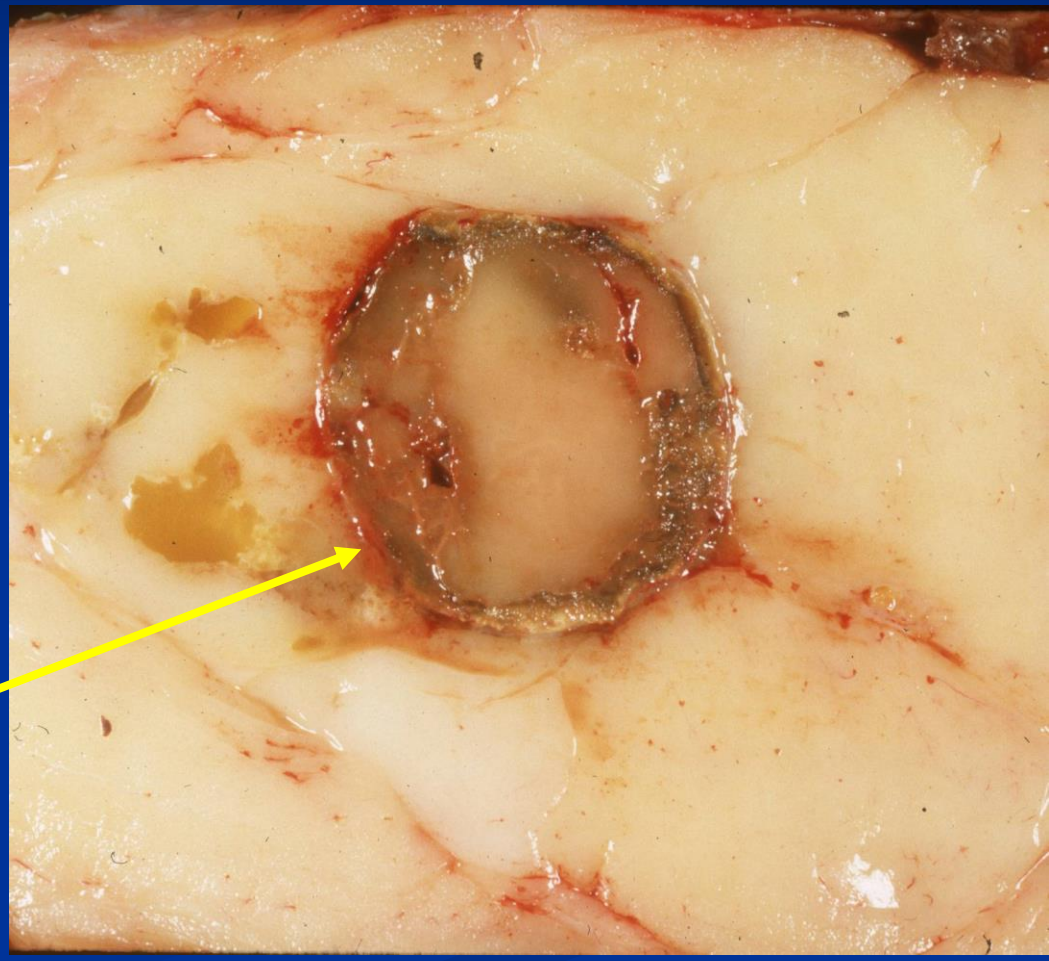
Gross Pathology Lipoma

- Yellow fat
- Area of necrosis (arrow)



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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

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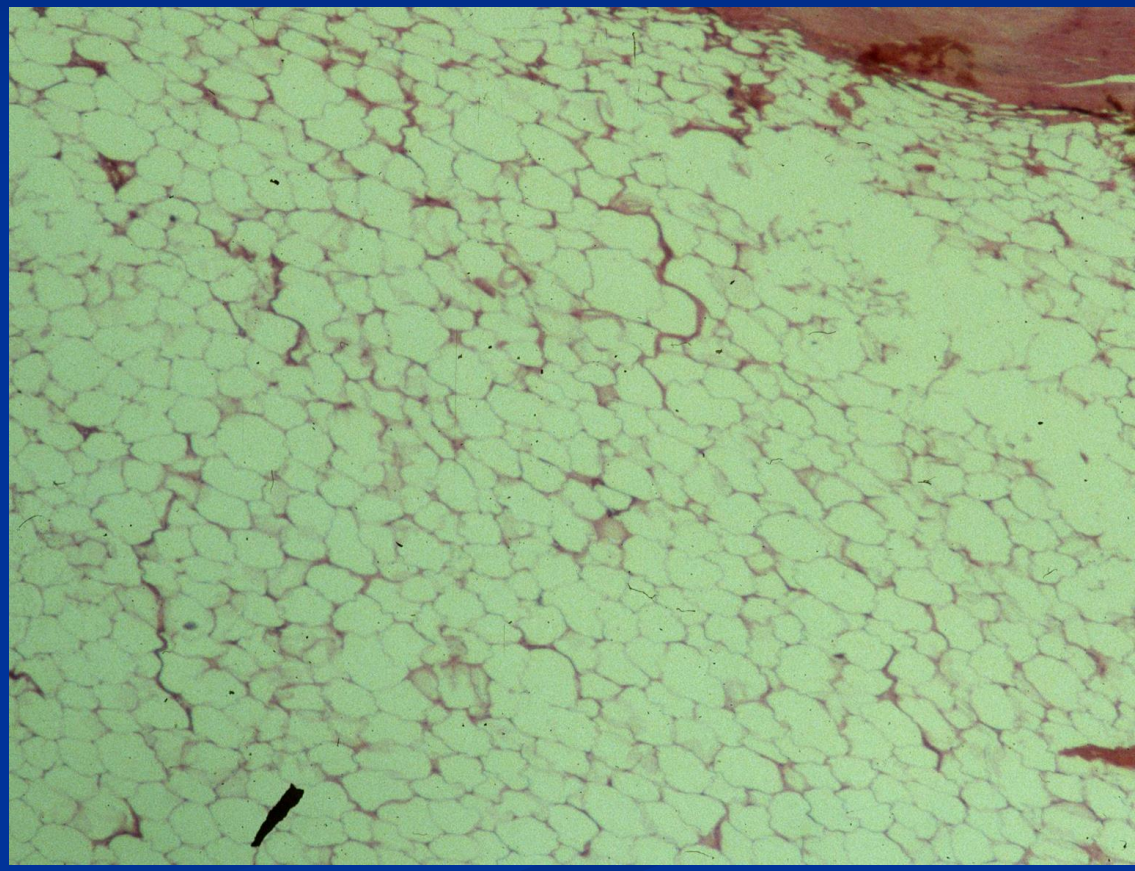
Pathology Microscopic Lipoma

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



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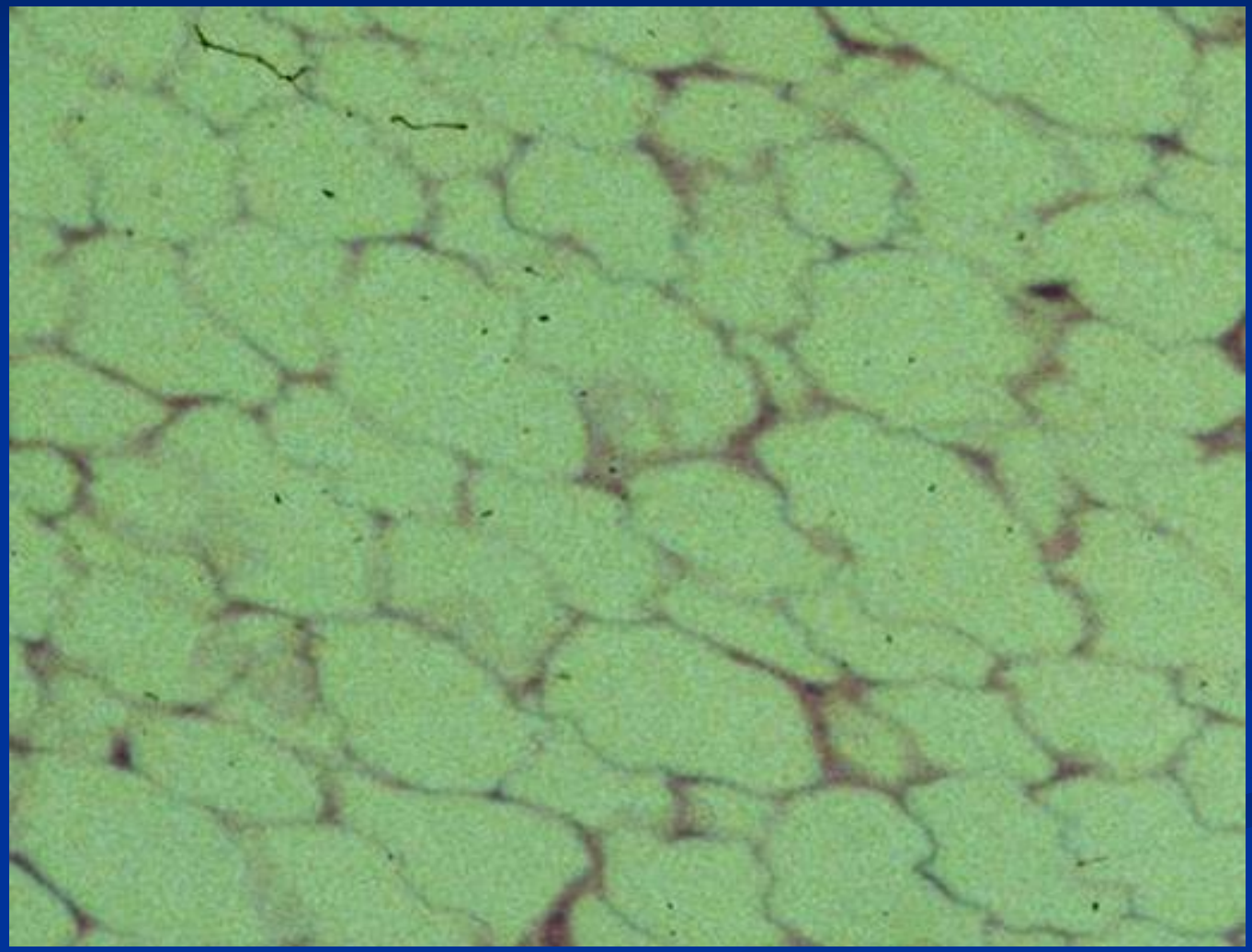
Pathology Microscopic Lipoma



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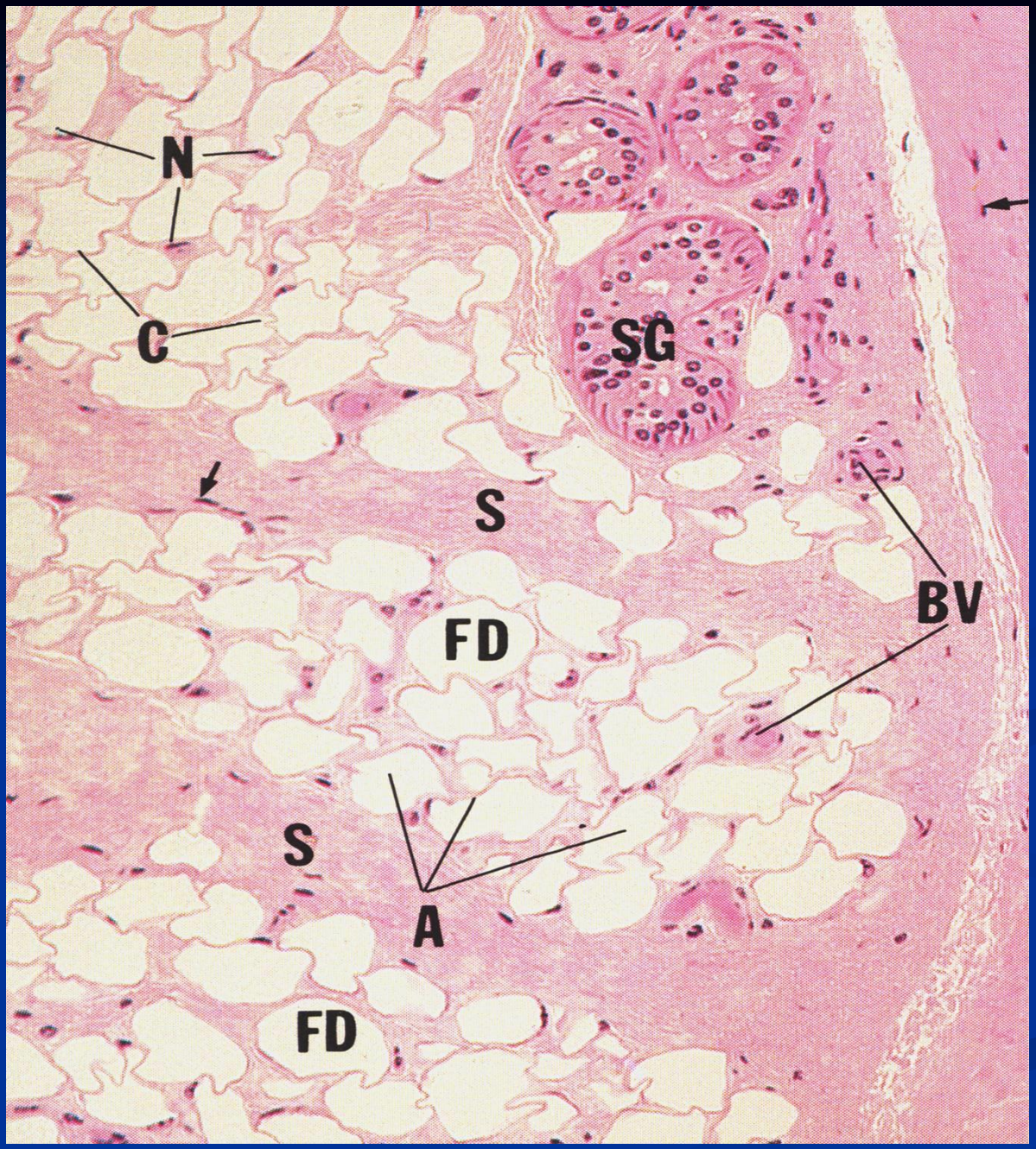

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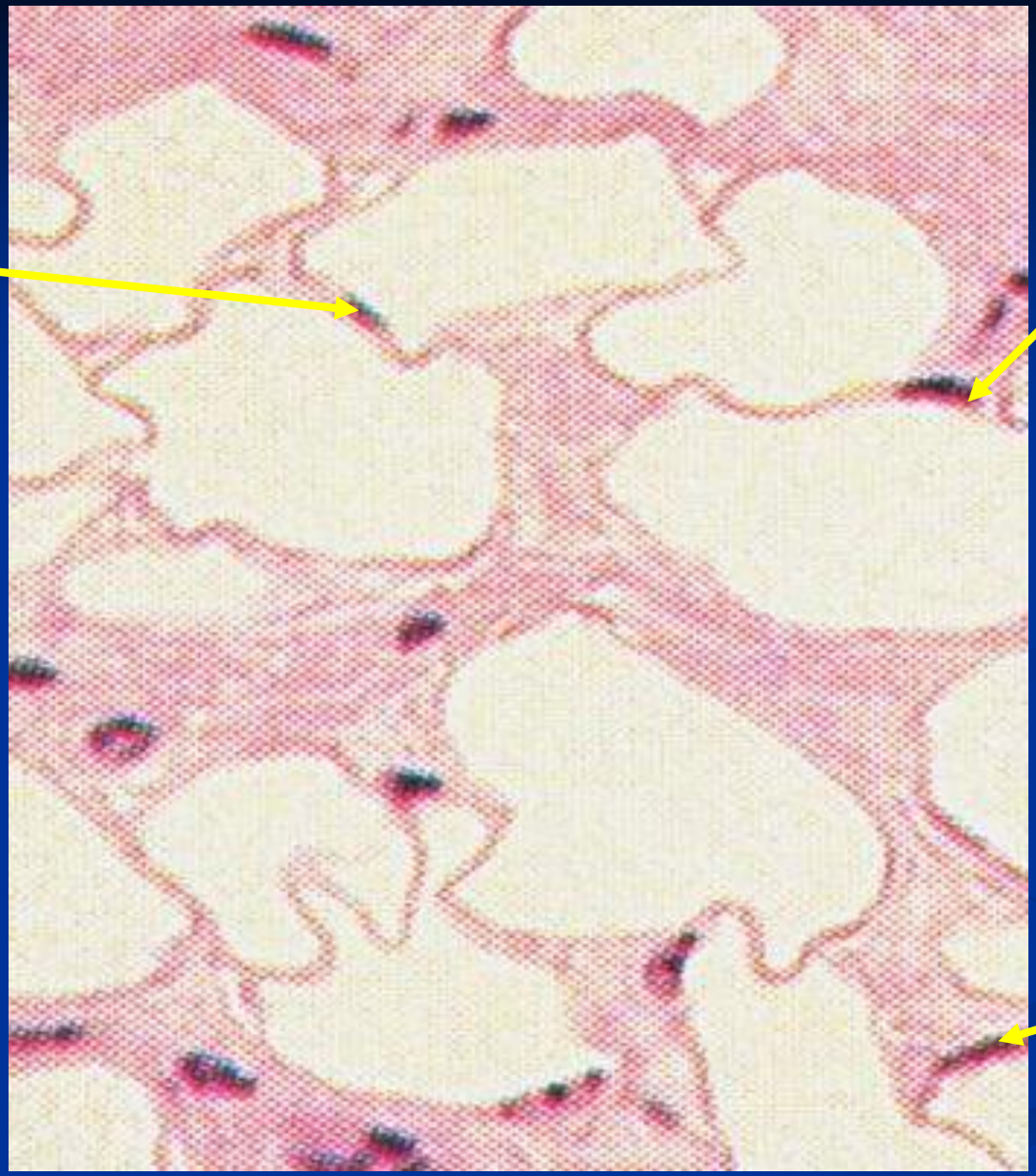

Peripheral
Compressed
Nuclei

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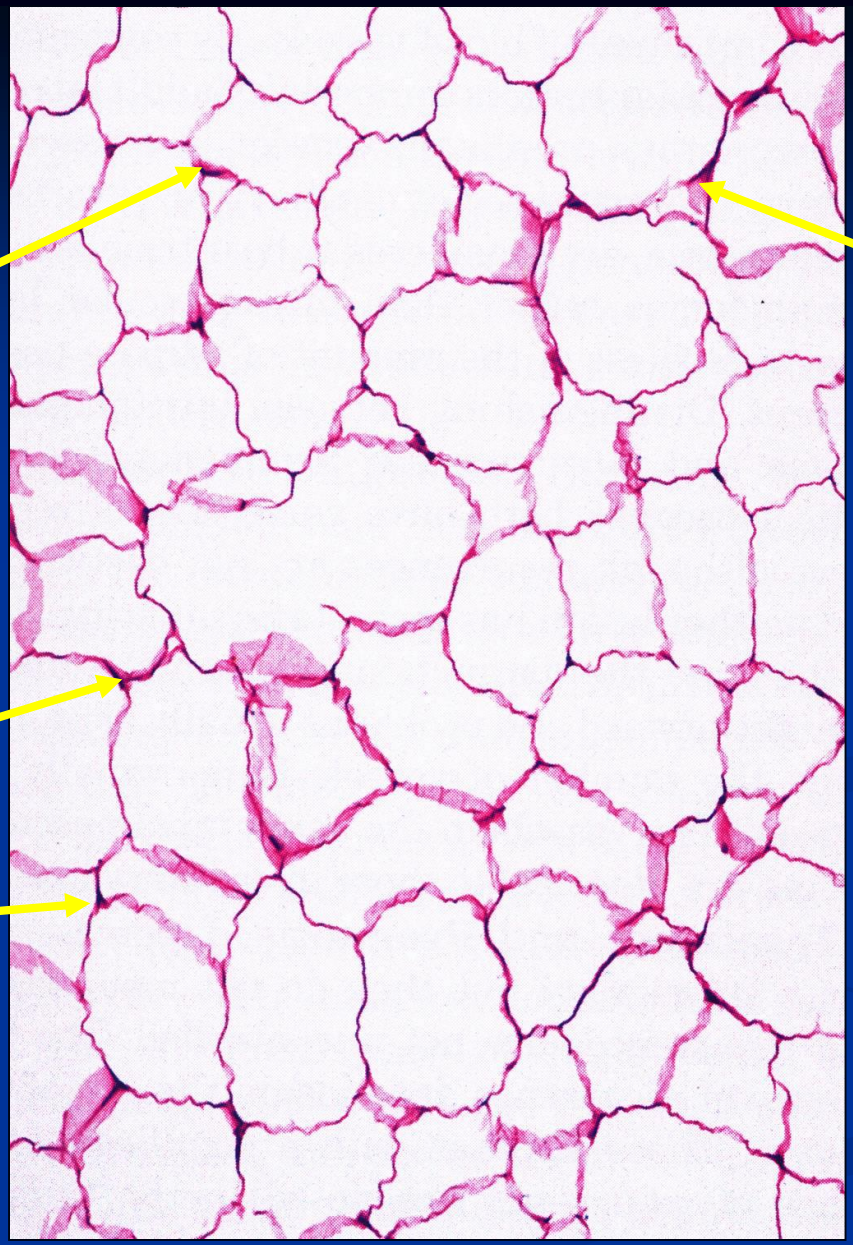

Compressed nuclei are barely discernible

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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

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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.

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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

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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.

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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

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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 enhacement of serpentine vascular channels
  - Subtle phleboliths
  - Lesions adjacent to bone rarely stimulate a subtle periosteal reaction

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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

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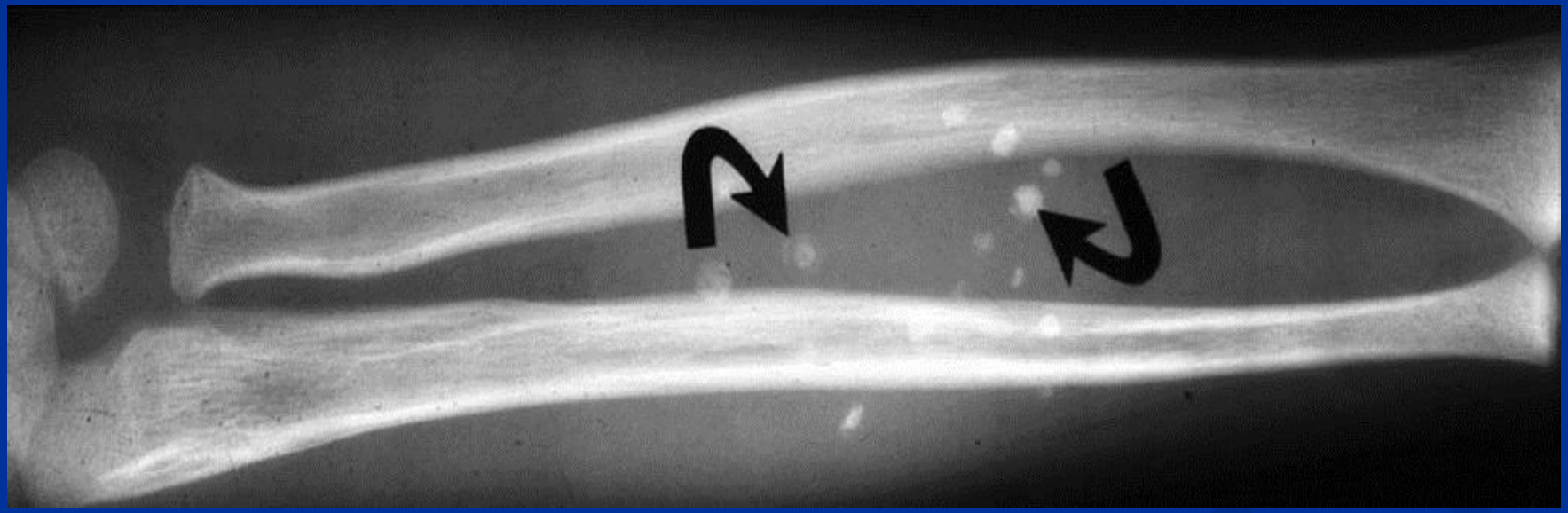
Hemangioma: Phleboliths (arrows)



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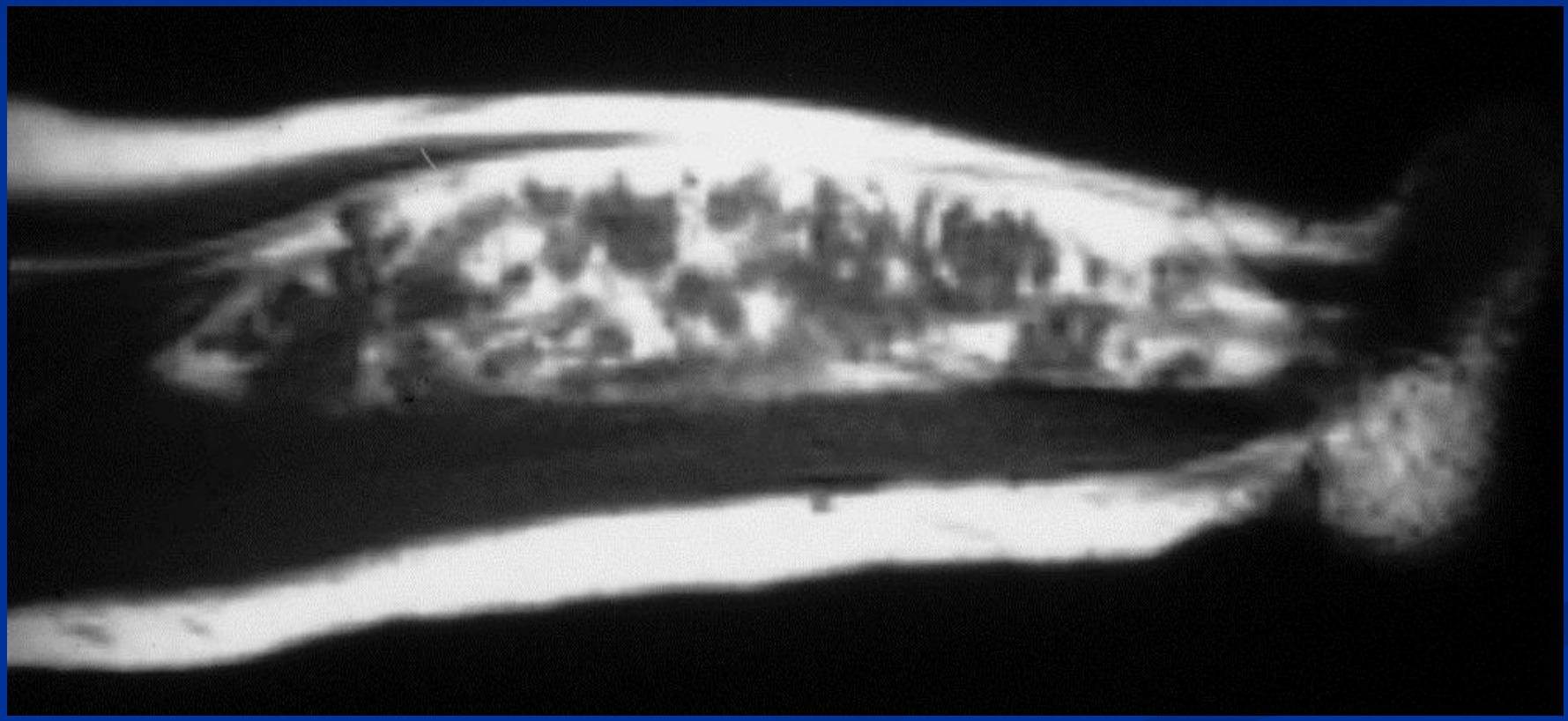
Hemangioma



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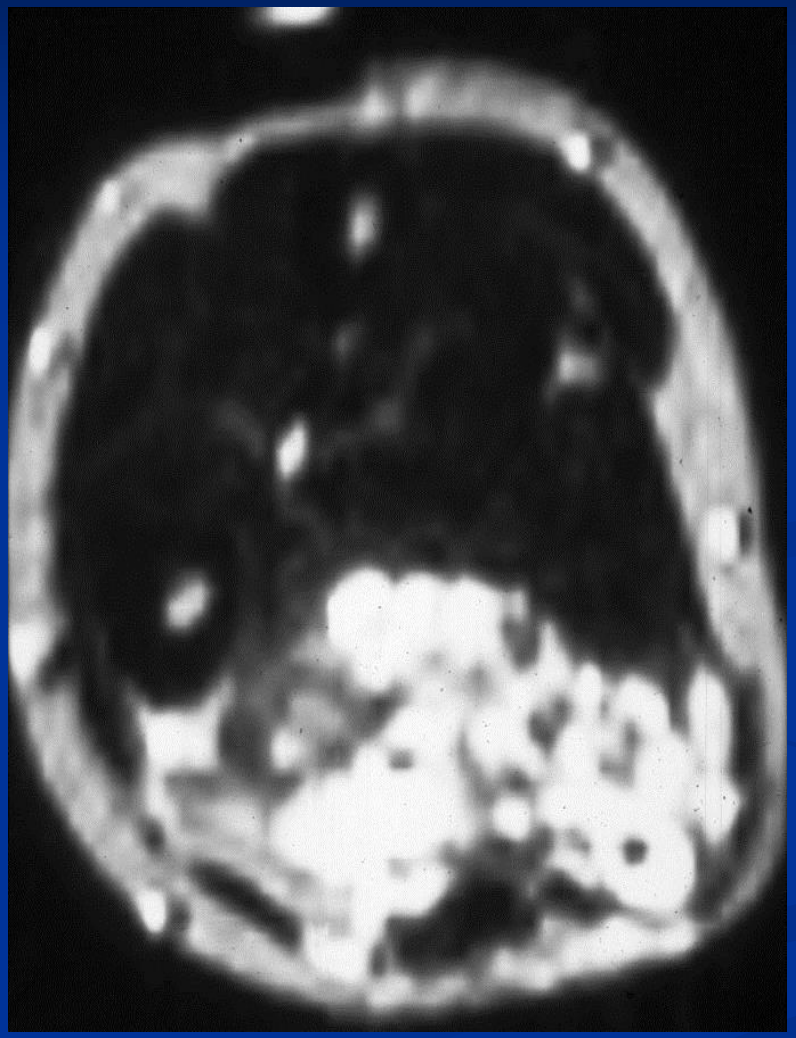
Hemangioma



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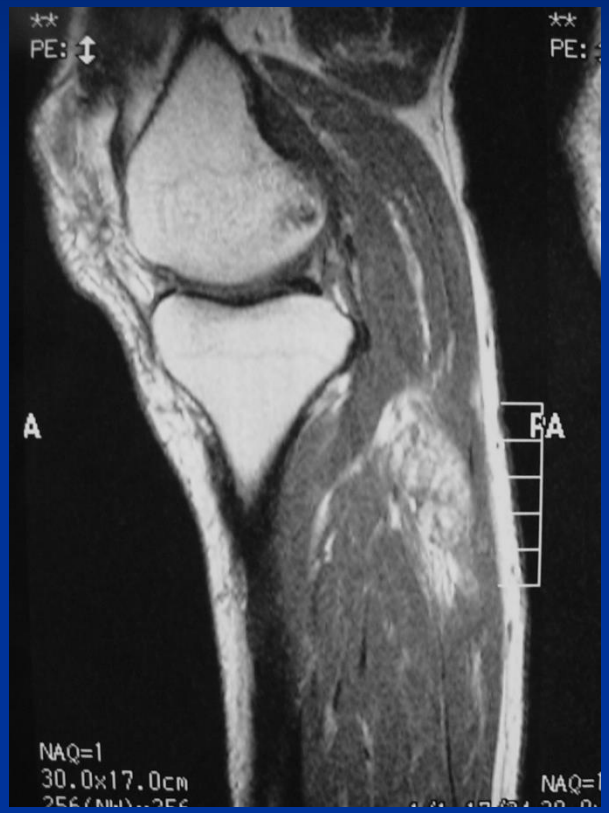
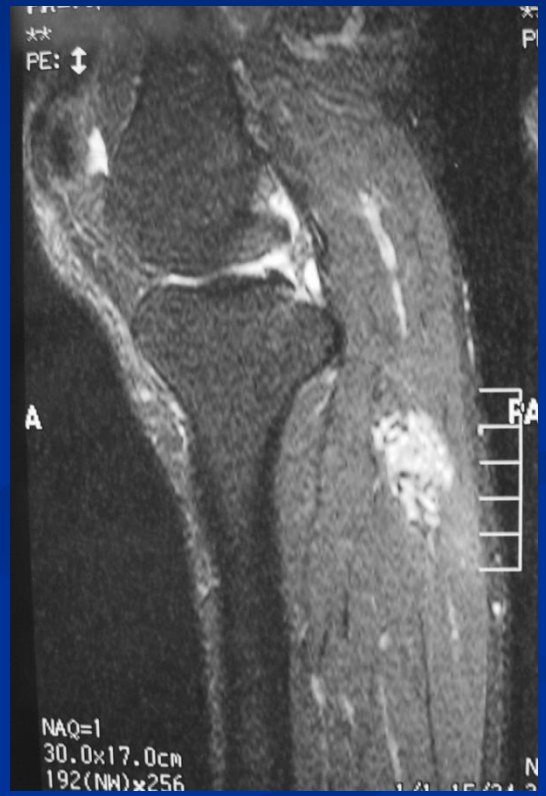
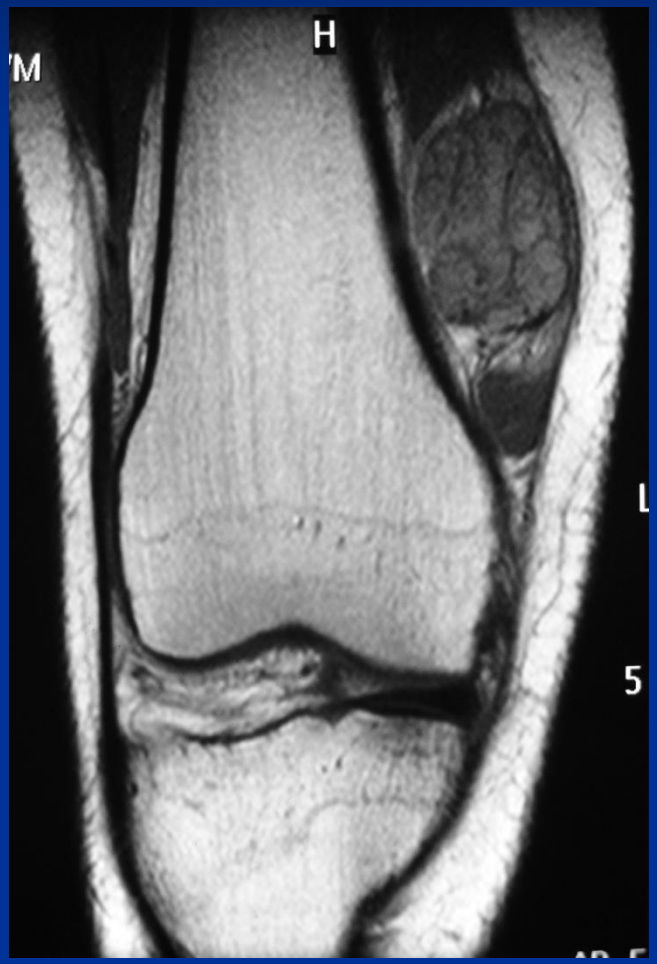
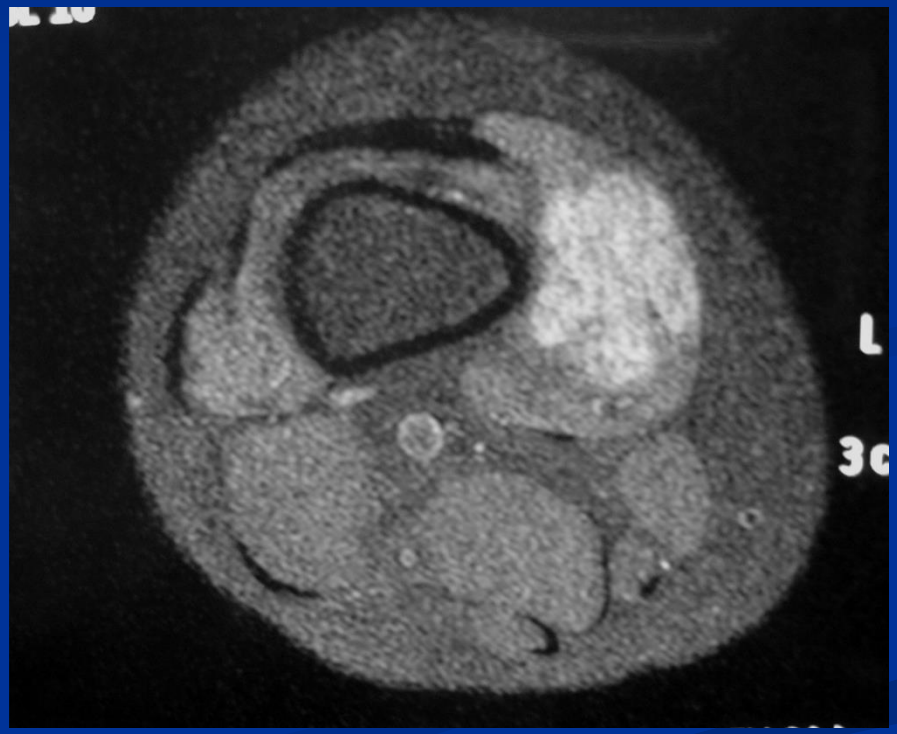
Hemangioma





Hemangioma





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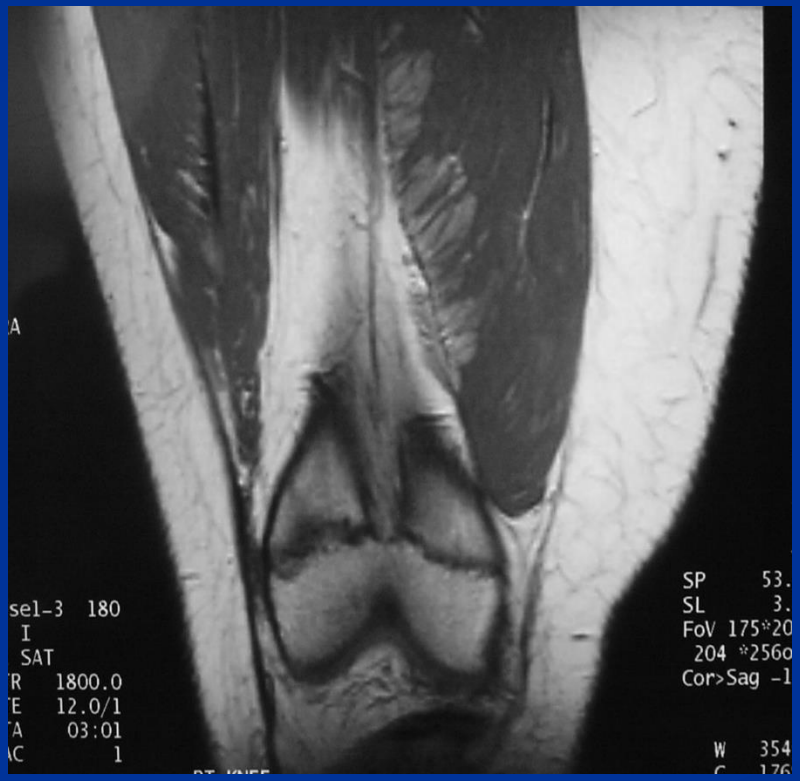
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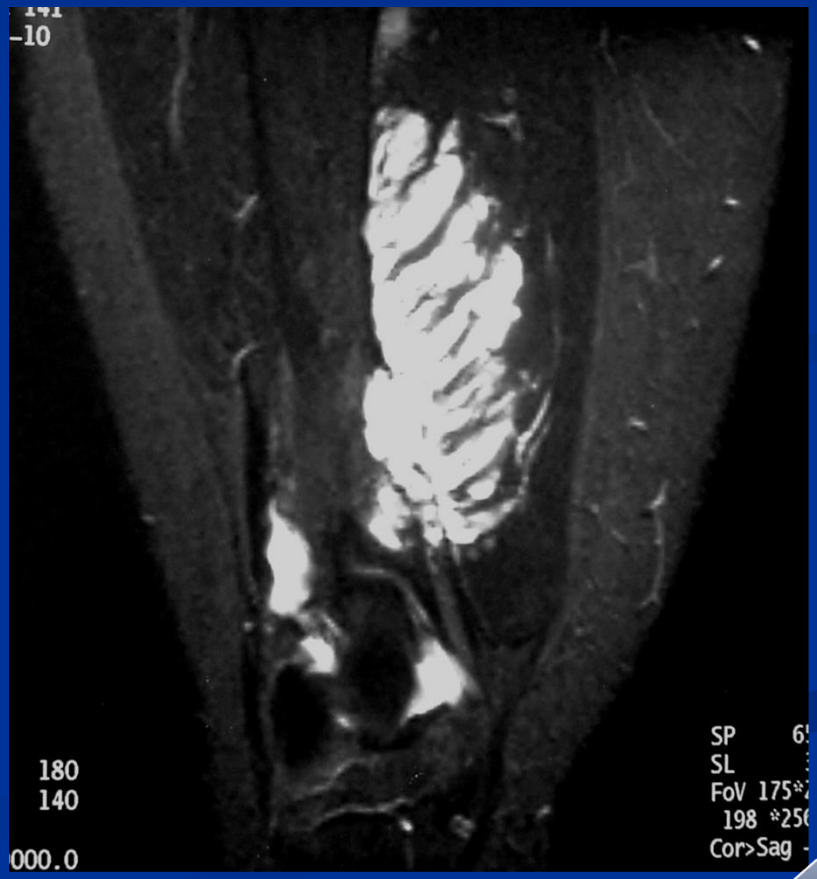
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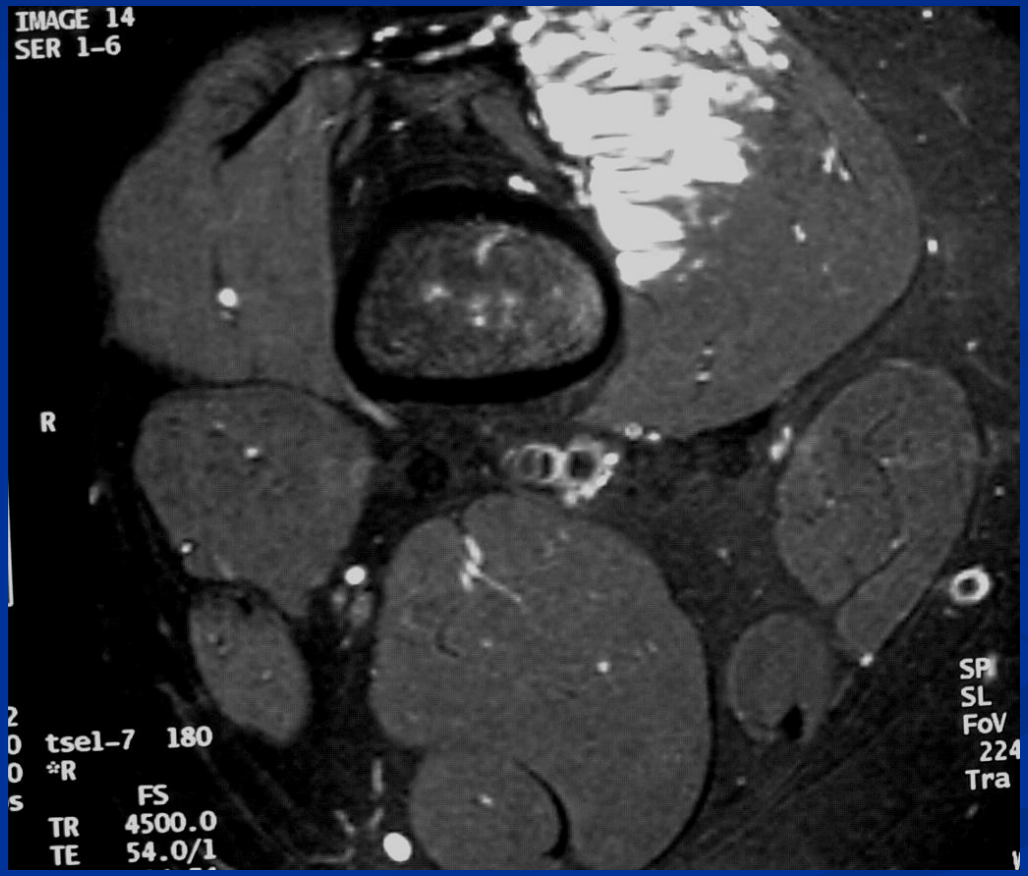
MRI T2 Hemangioma



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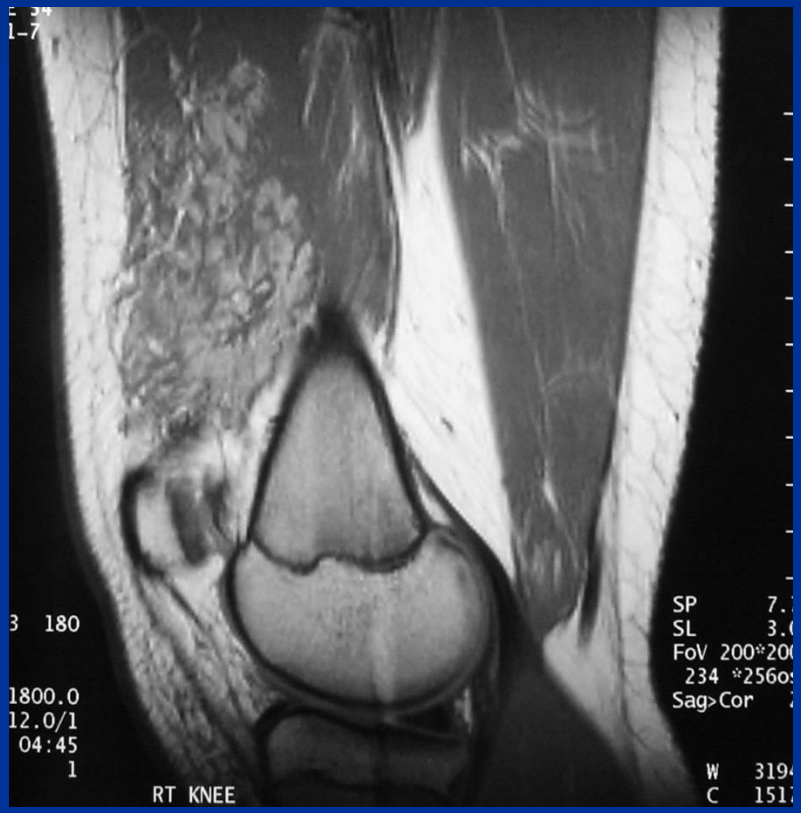
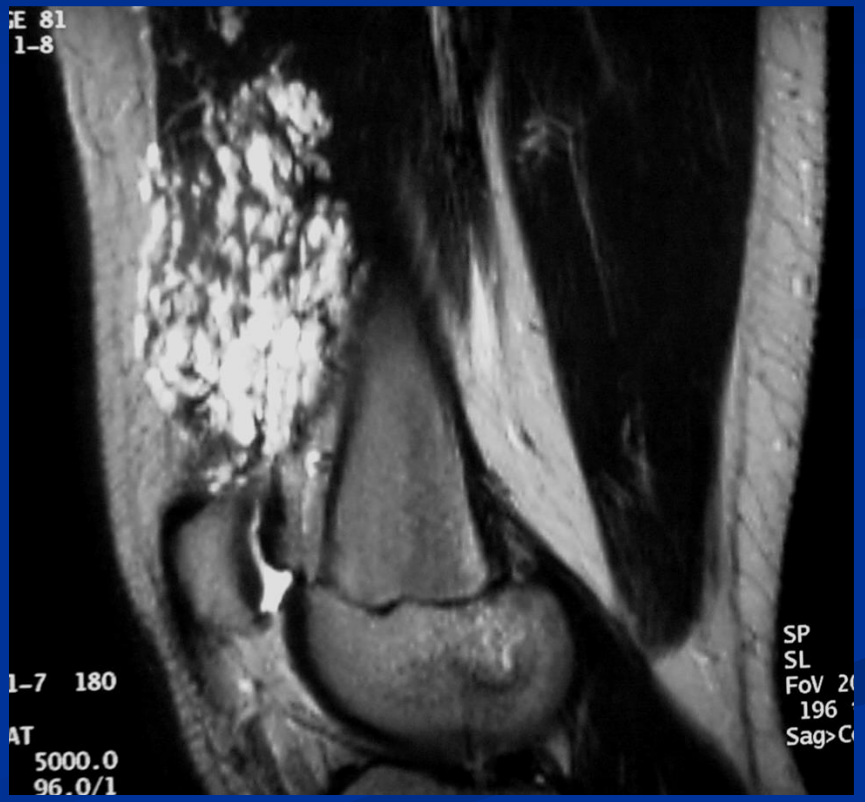
Hemangioma





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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

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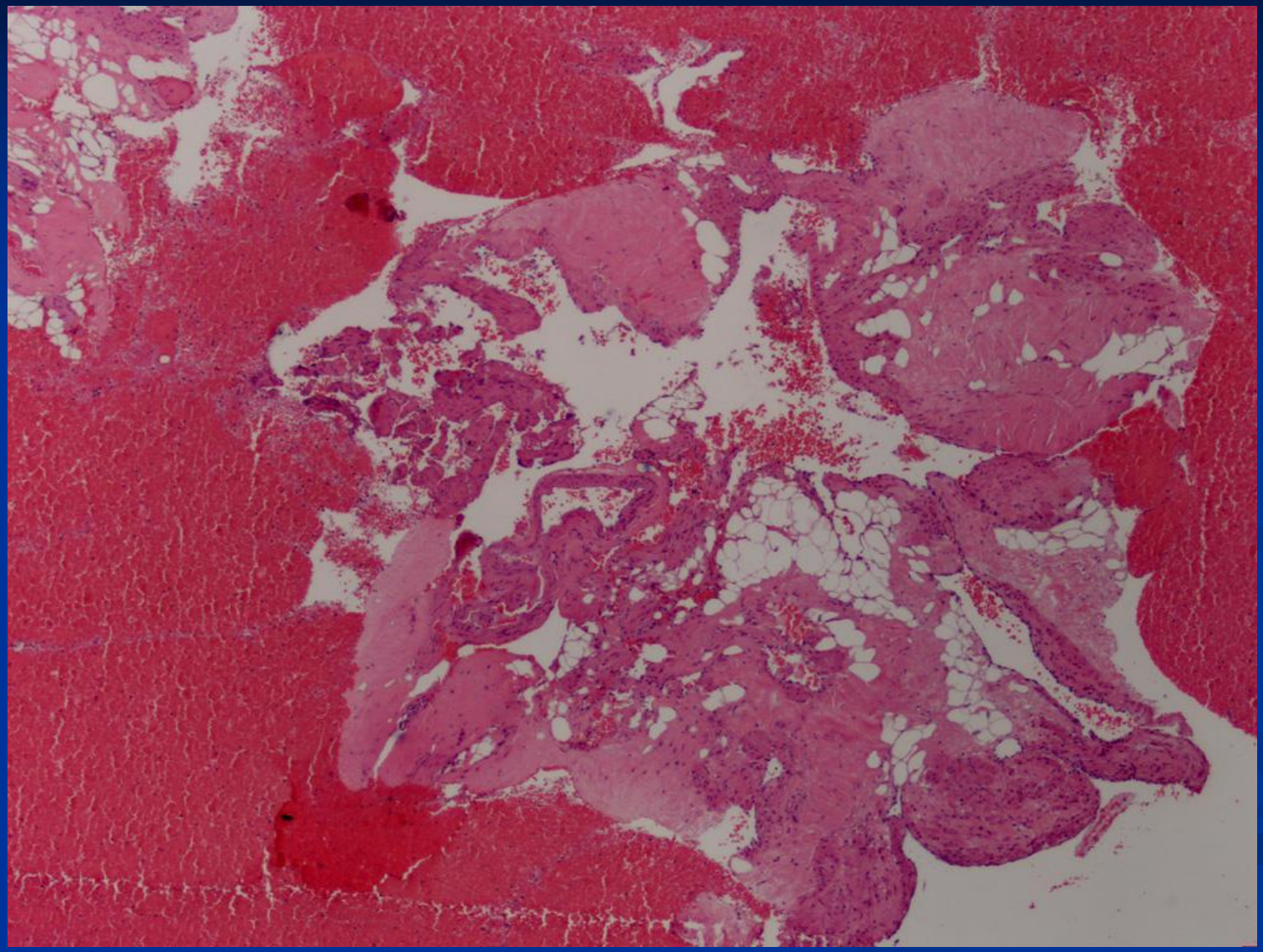

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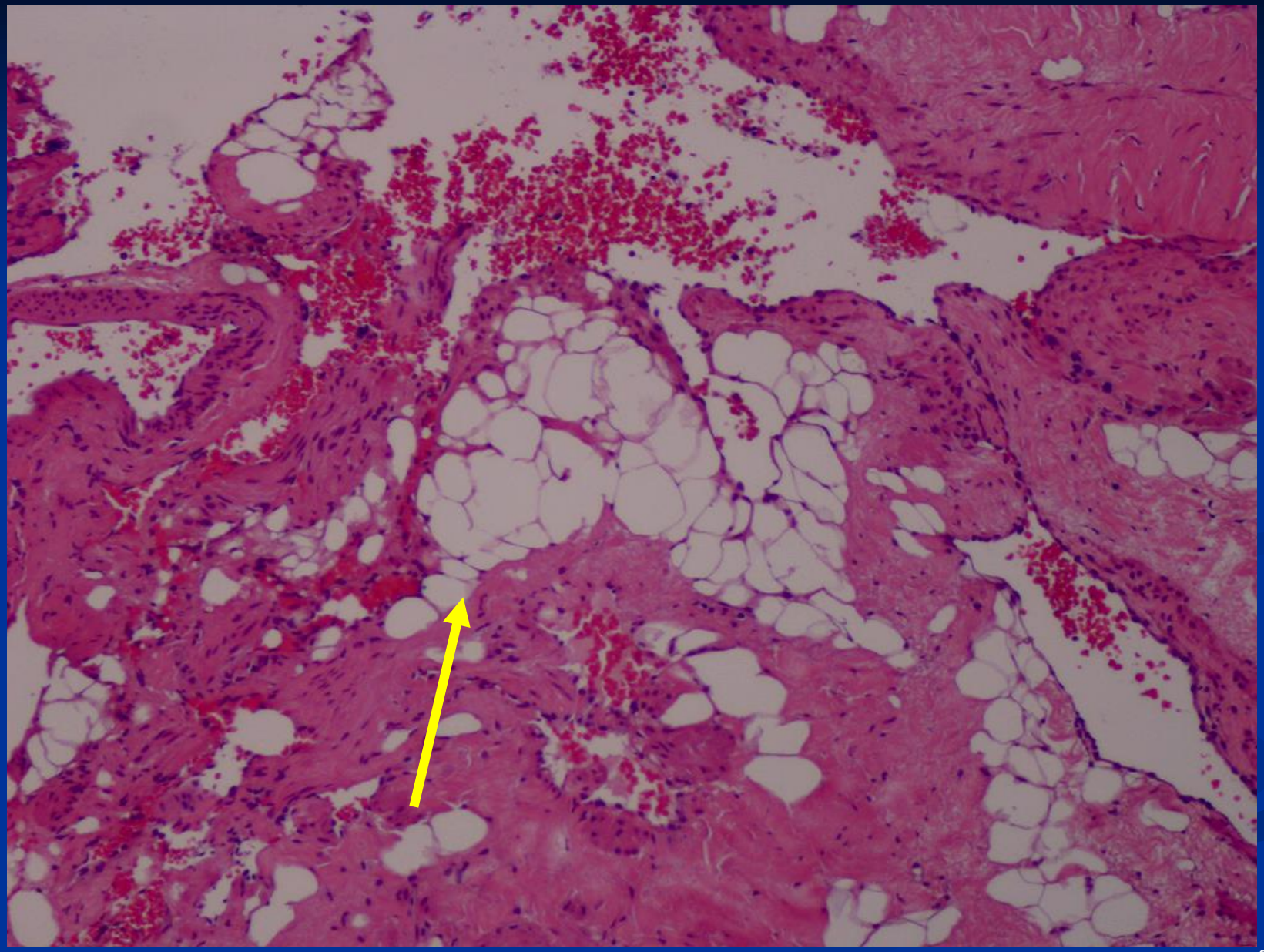

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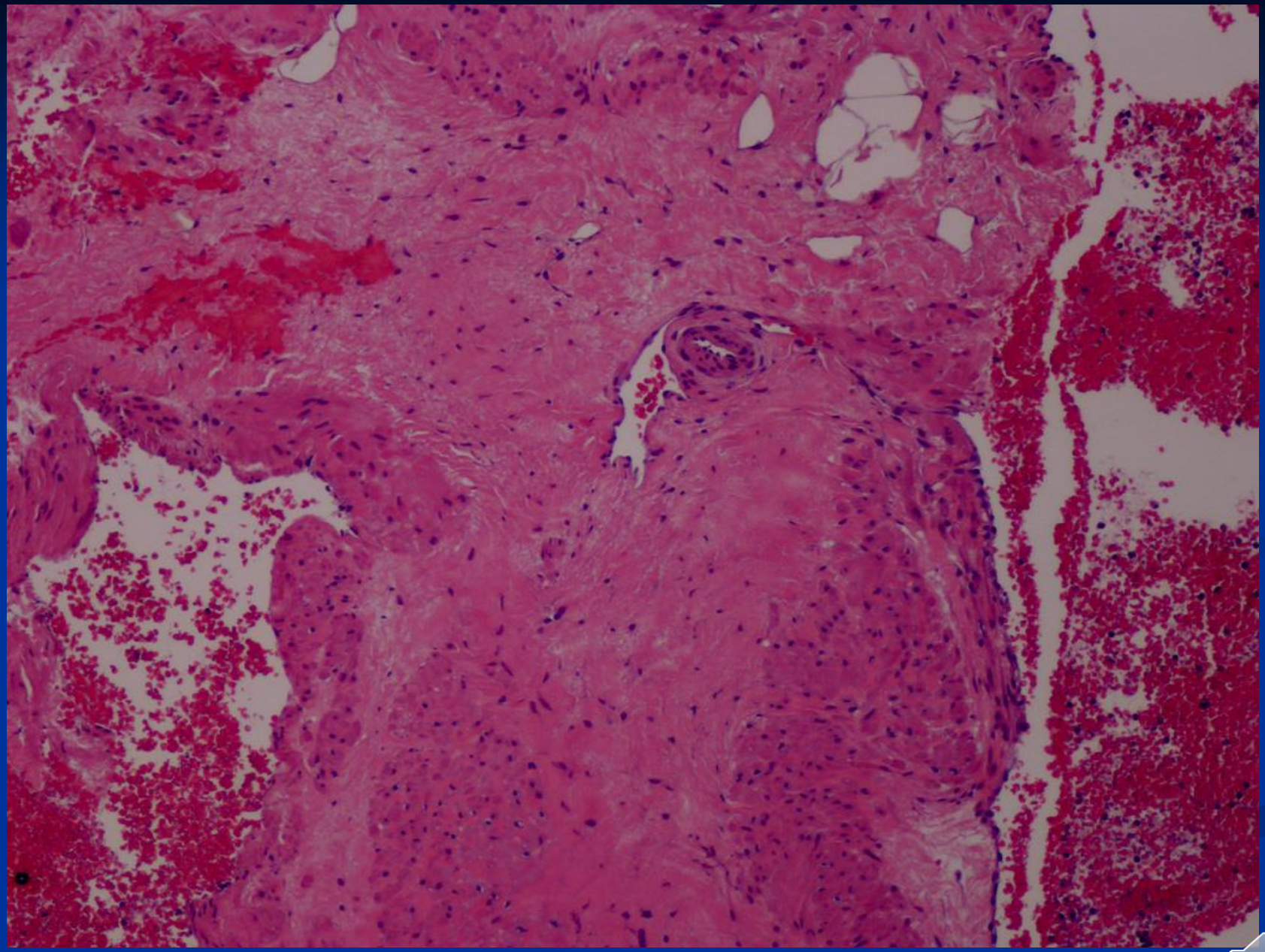

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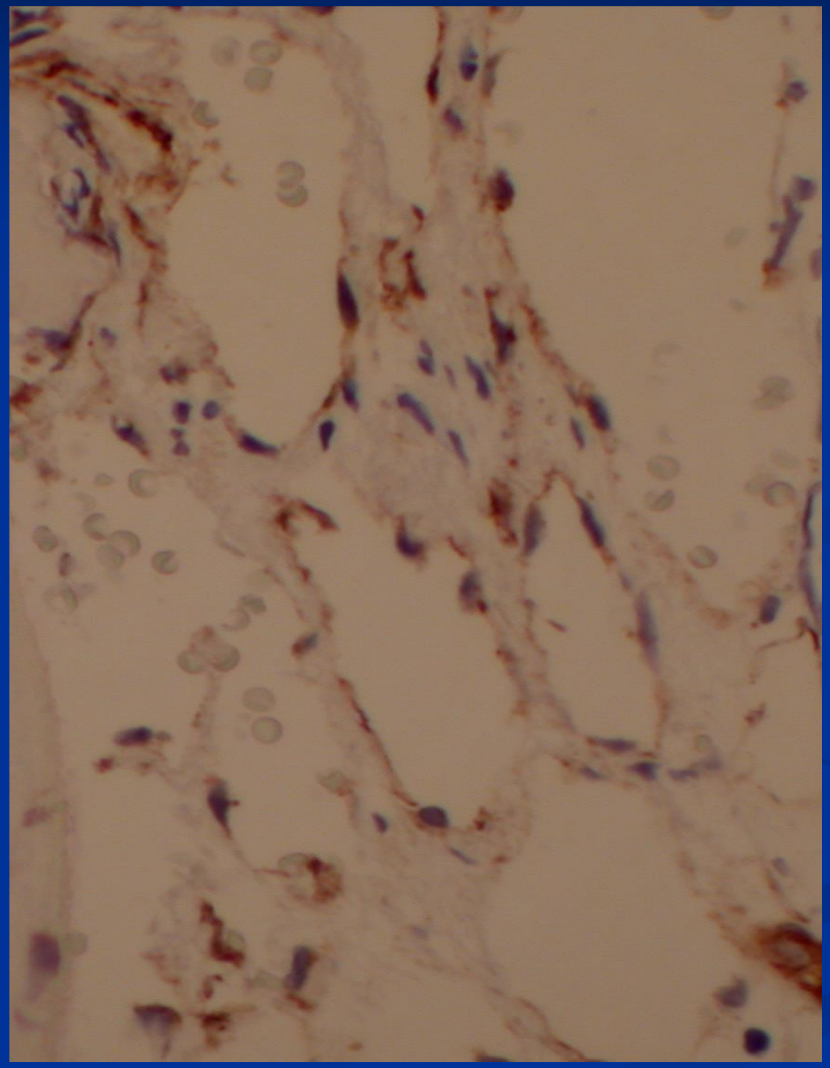
Hemangioma



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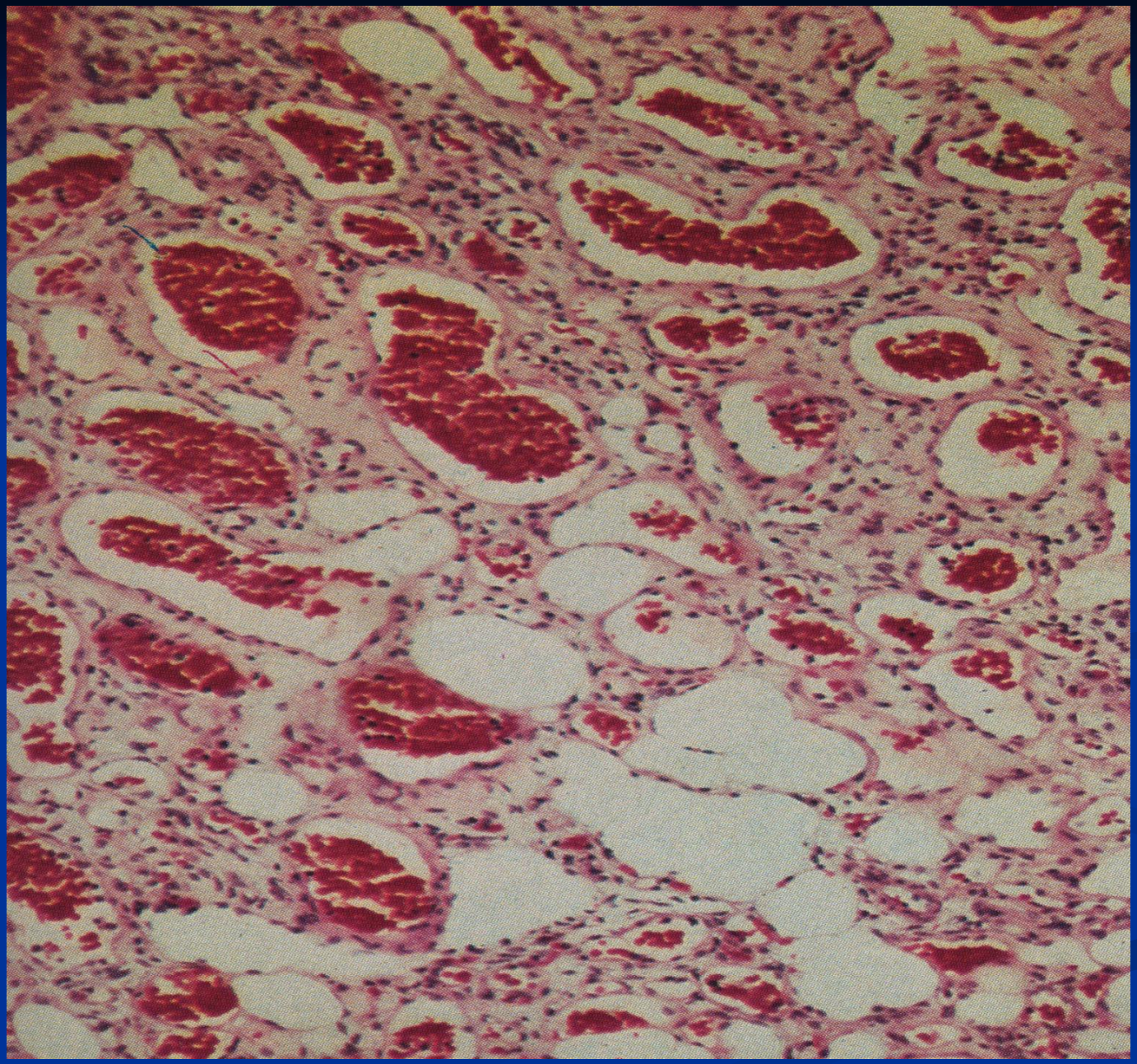

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Fibromatosis/Desmoplastic Fibroma

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.

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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

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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)

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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)

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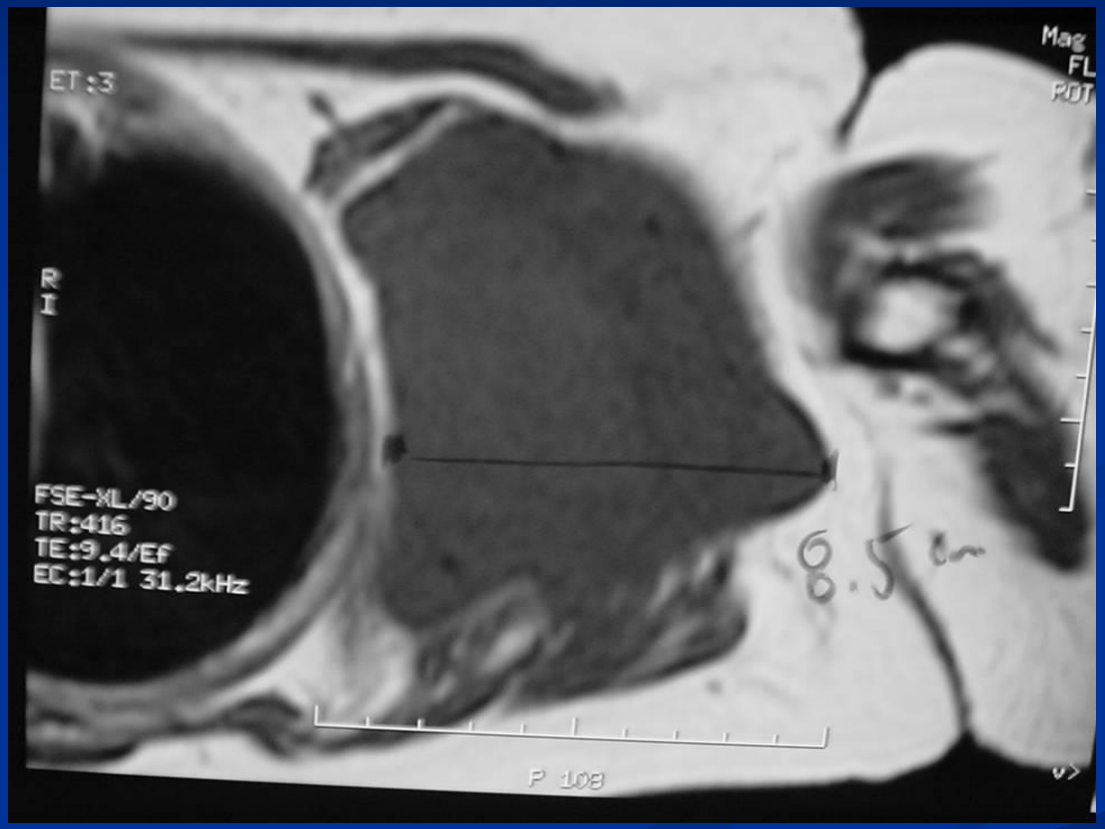
MRI T1: Fibromatosis

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



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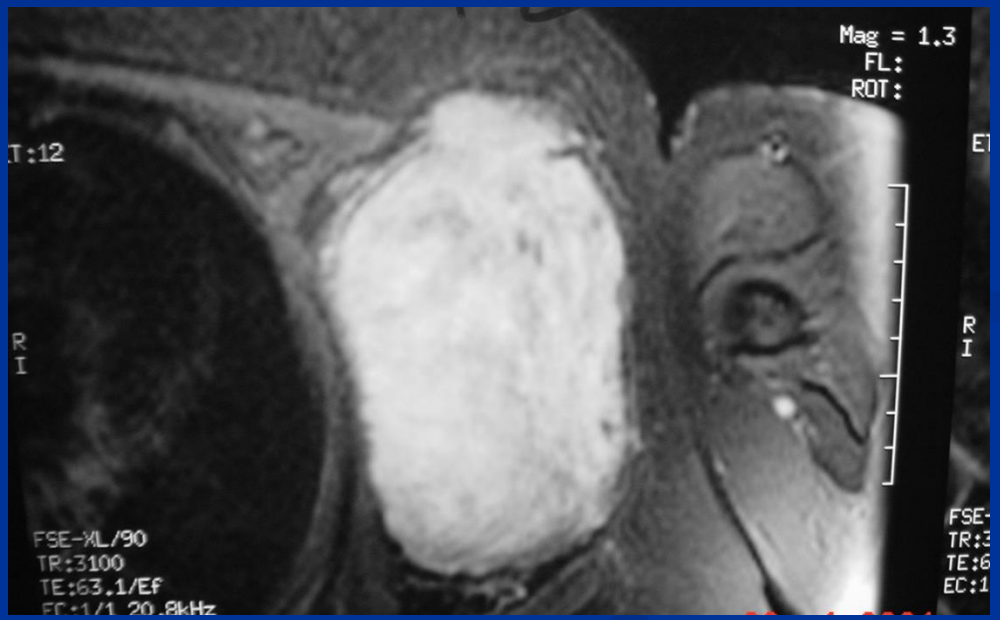
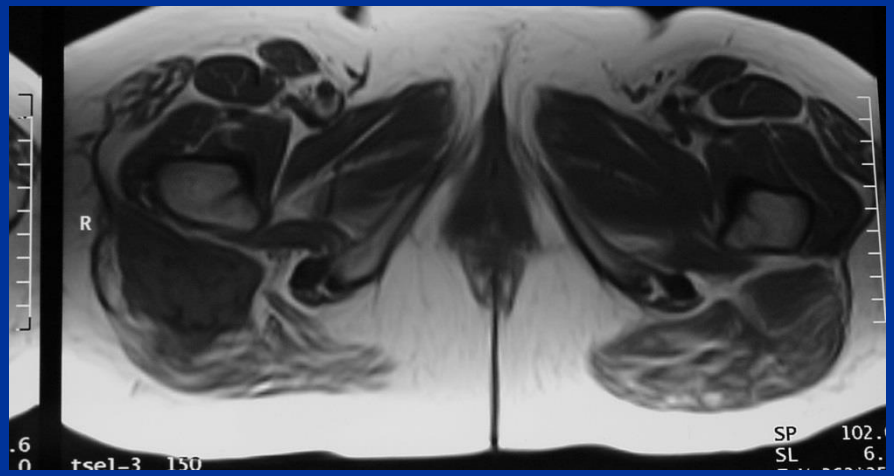
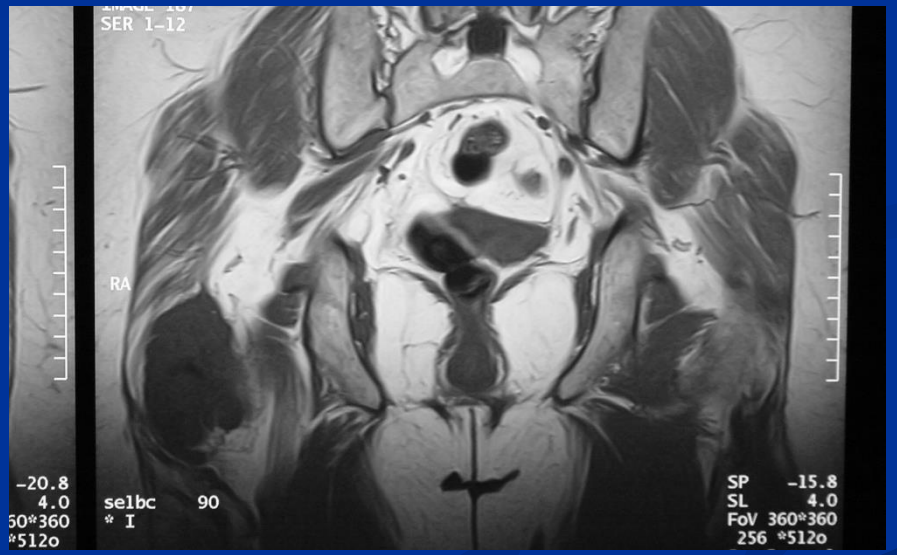
MRI T2: Fibromatosis

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



MRI T1: Right Buttock Fibromatosis





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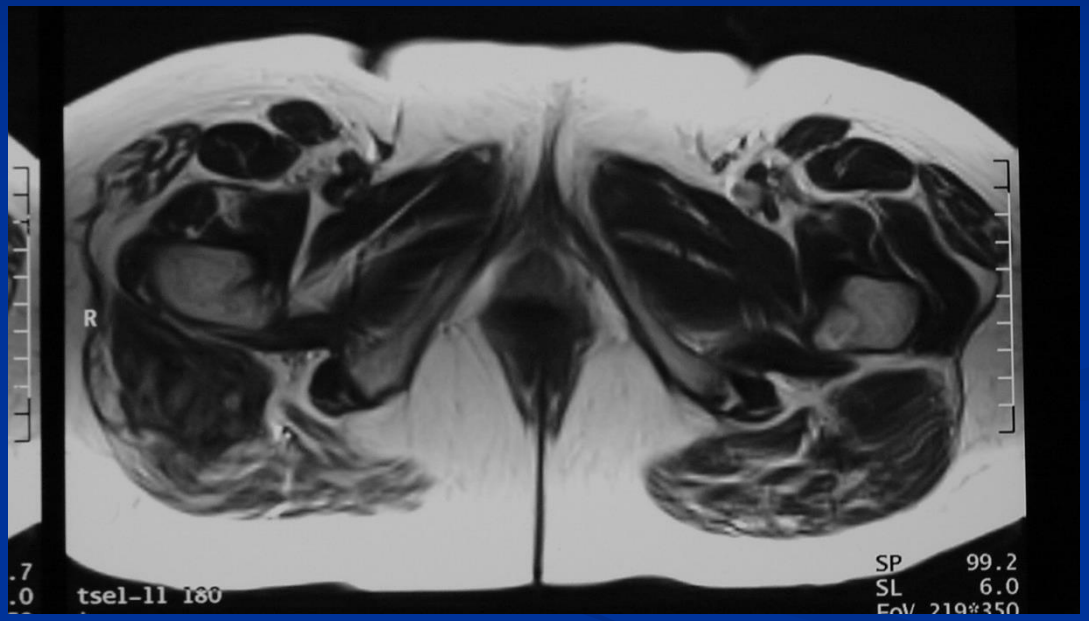
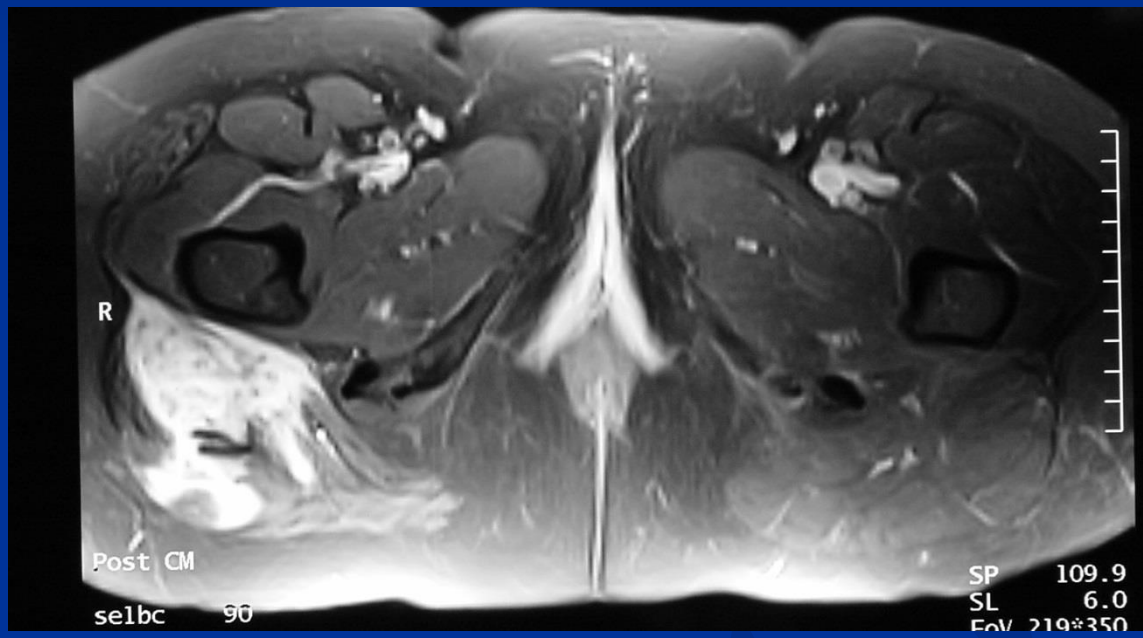
MRI T2: Fibromatosis Right Buttock

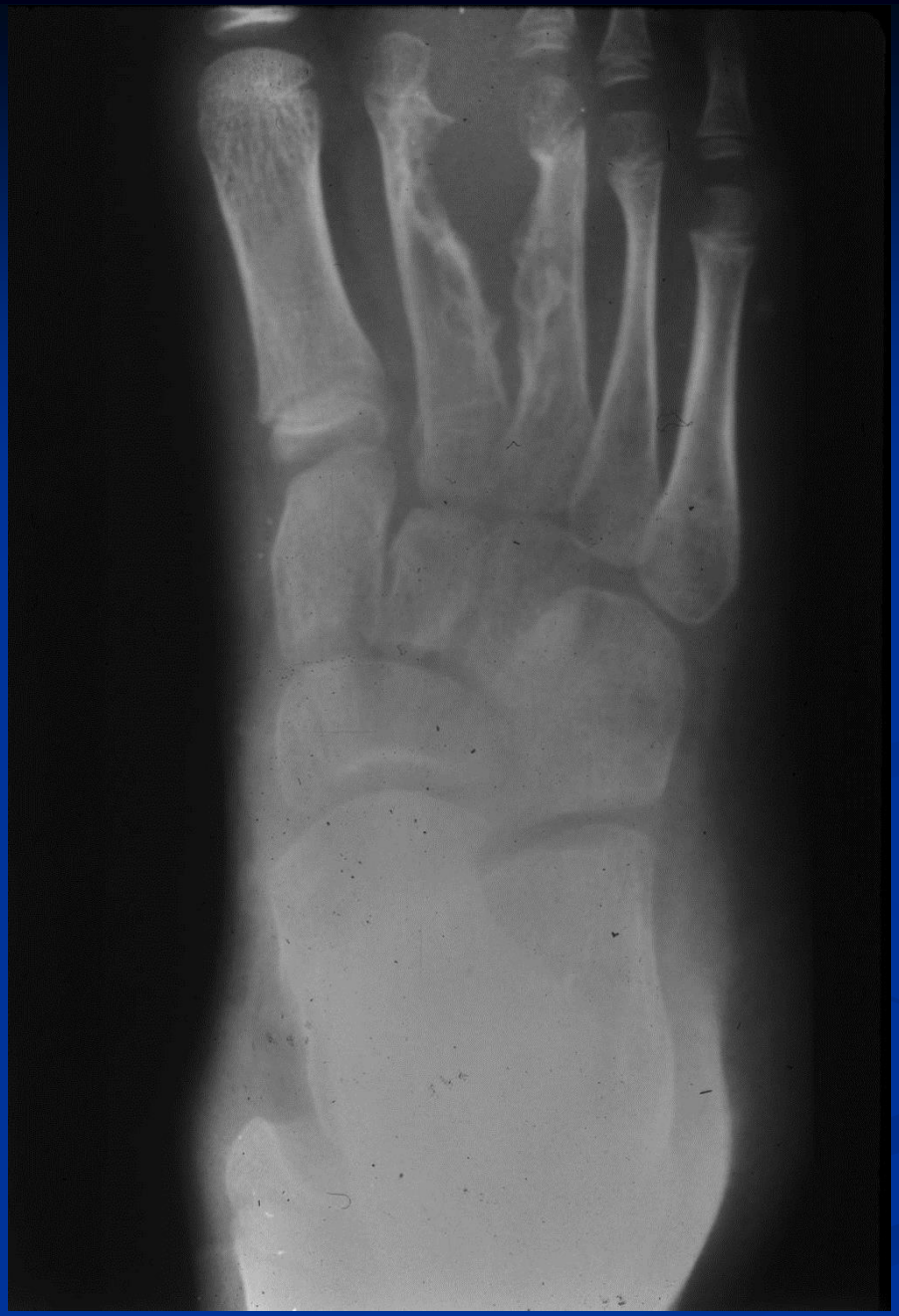
- Low to Intermediate Signal on T2
- Significant Fibrous Component



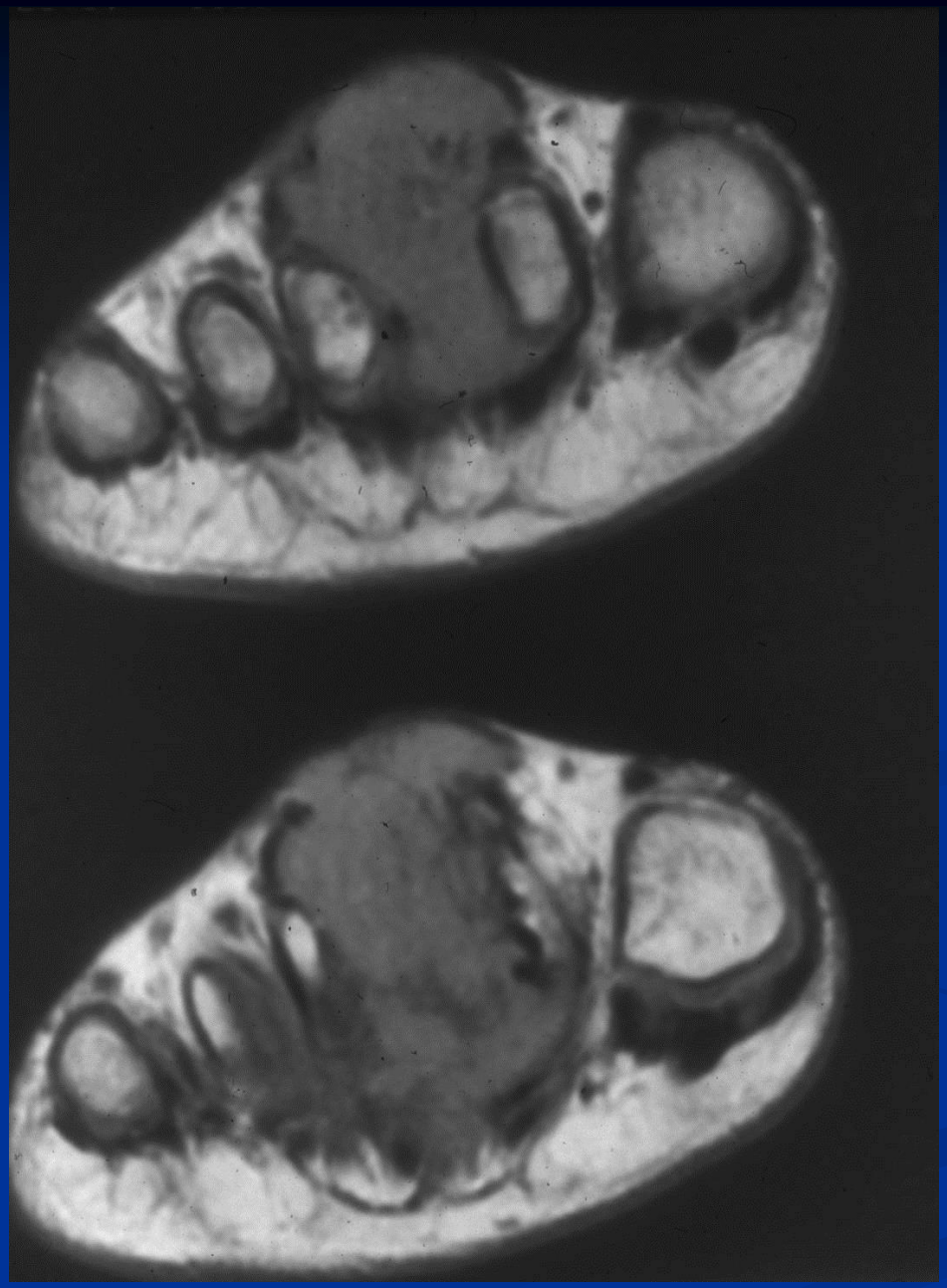
MRI with Gadolinium: Fibromatosis

- Enhances Diffusely
- Very Infiltrative
- Poor margins





Wittig Orthopedic Oncology



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

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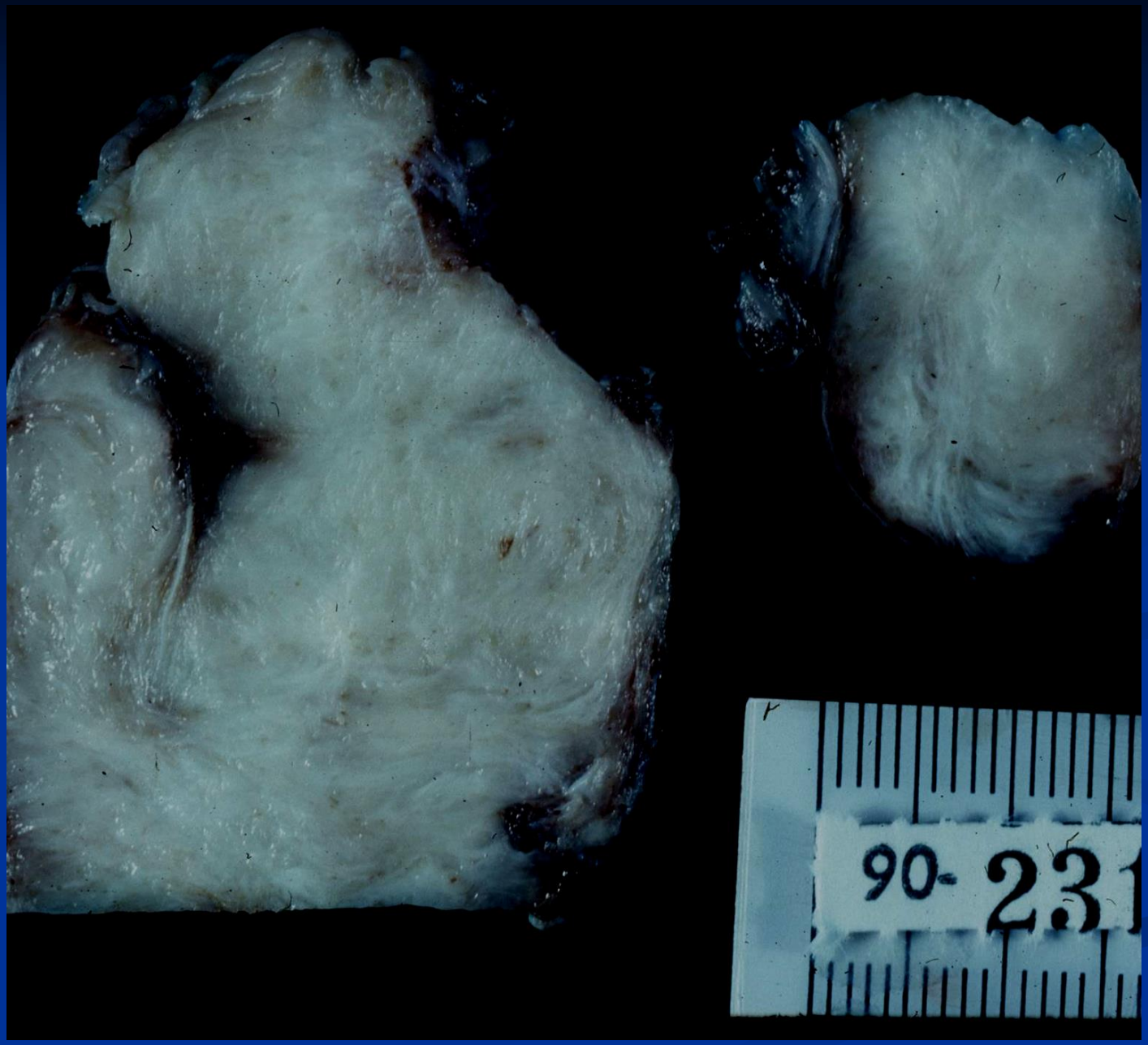

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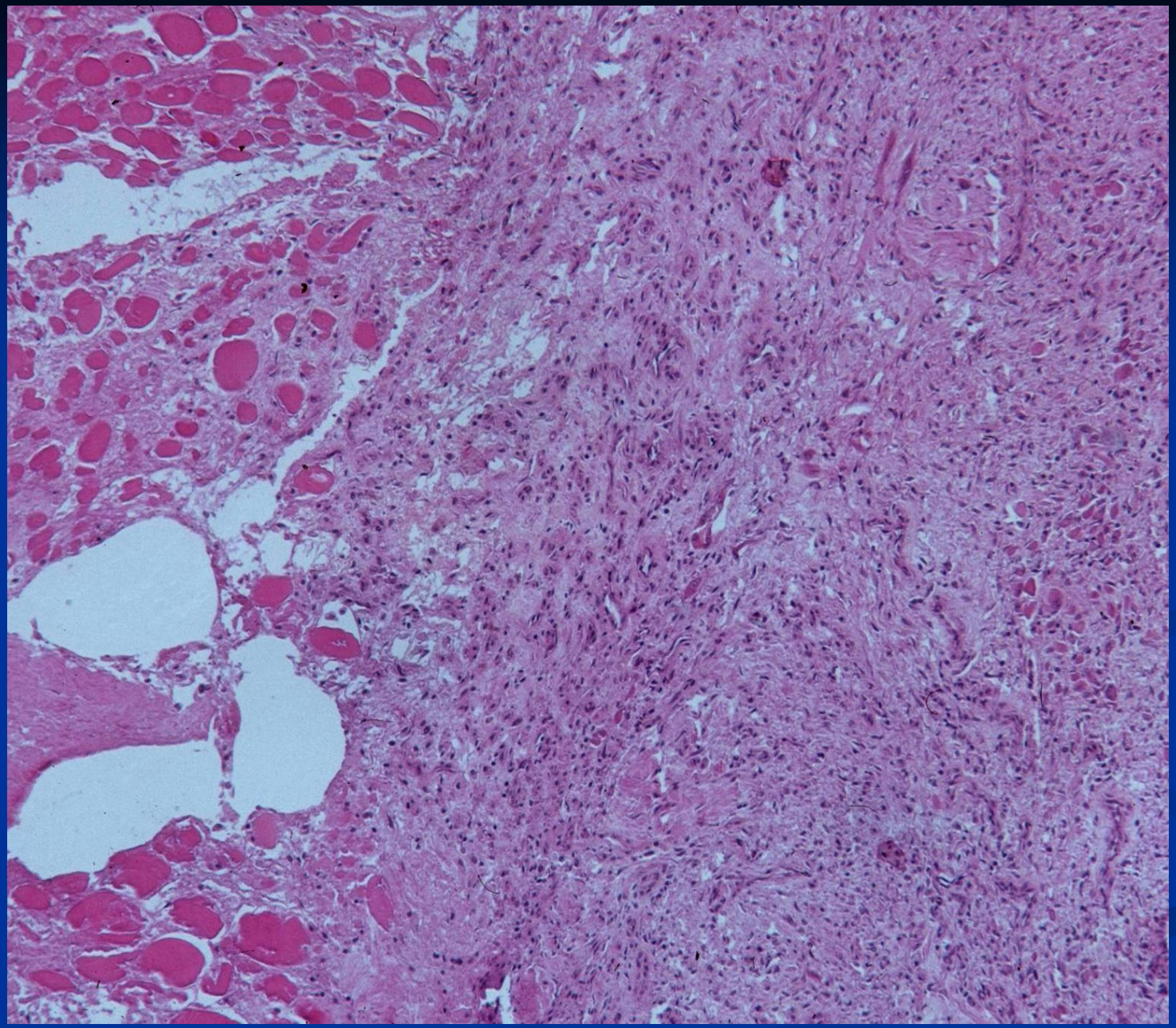
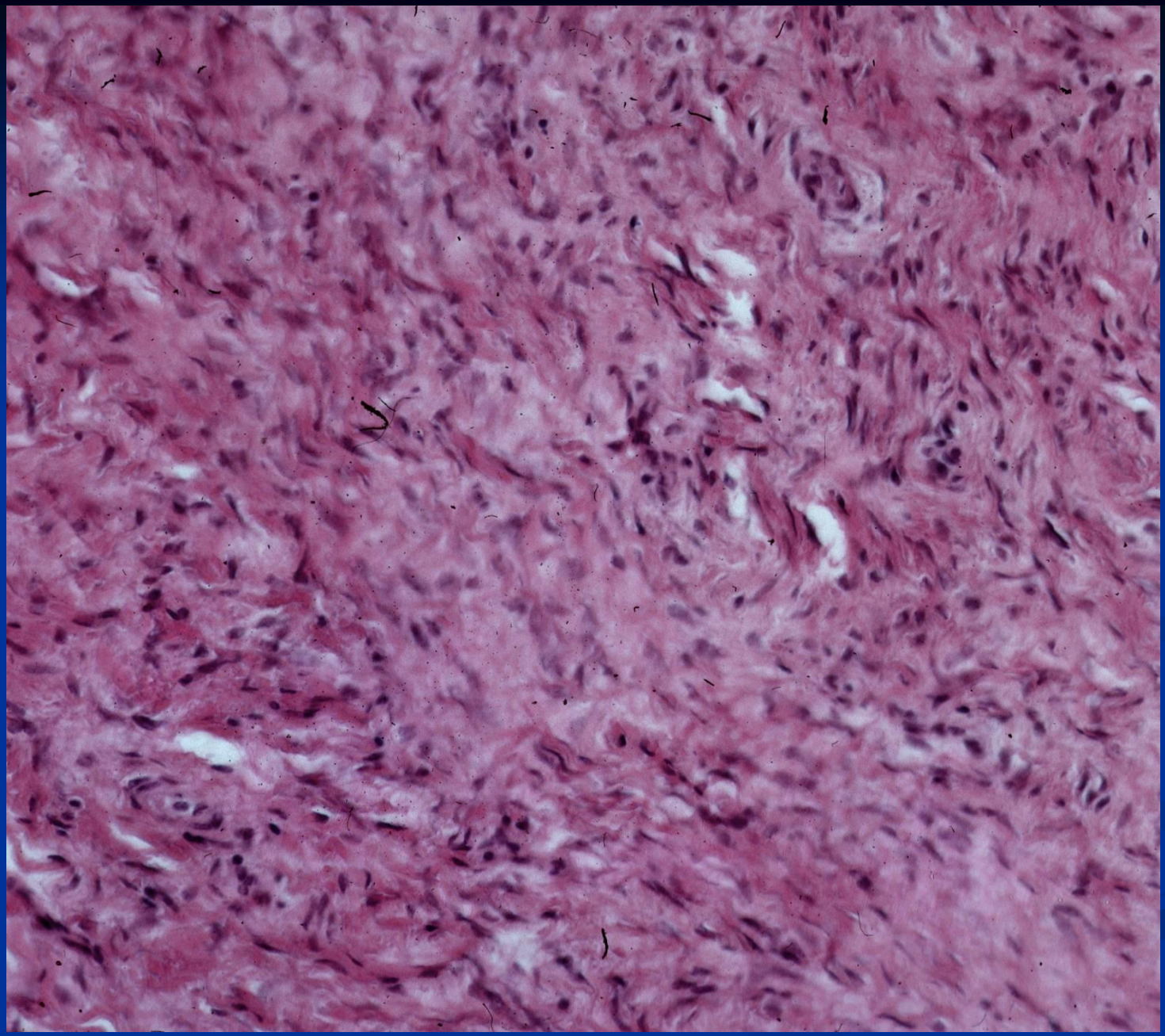


Wittig Orthopedic Oncology



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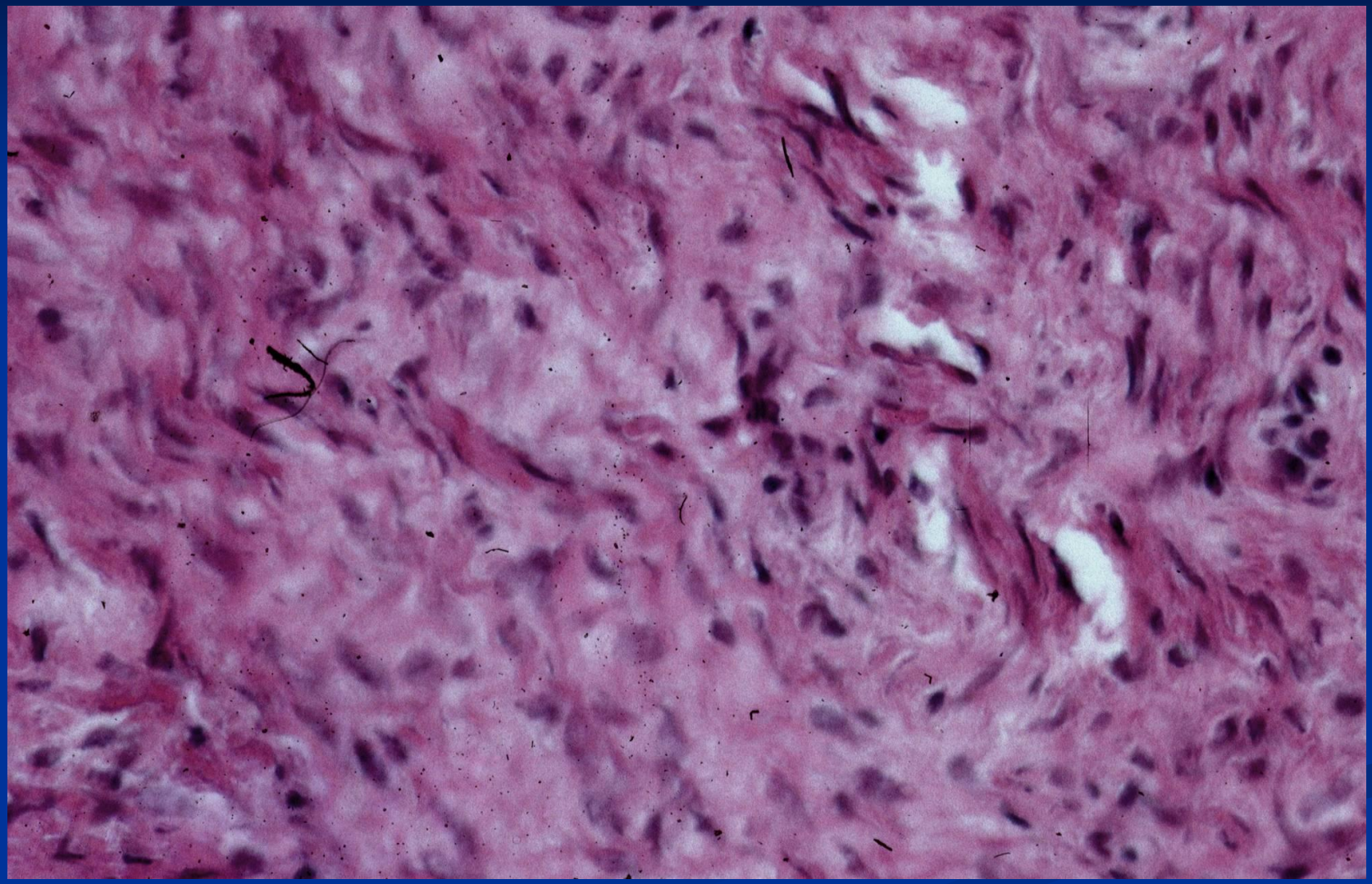

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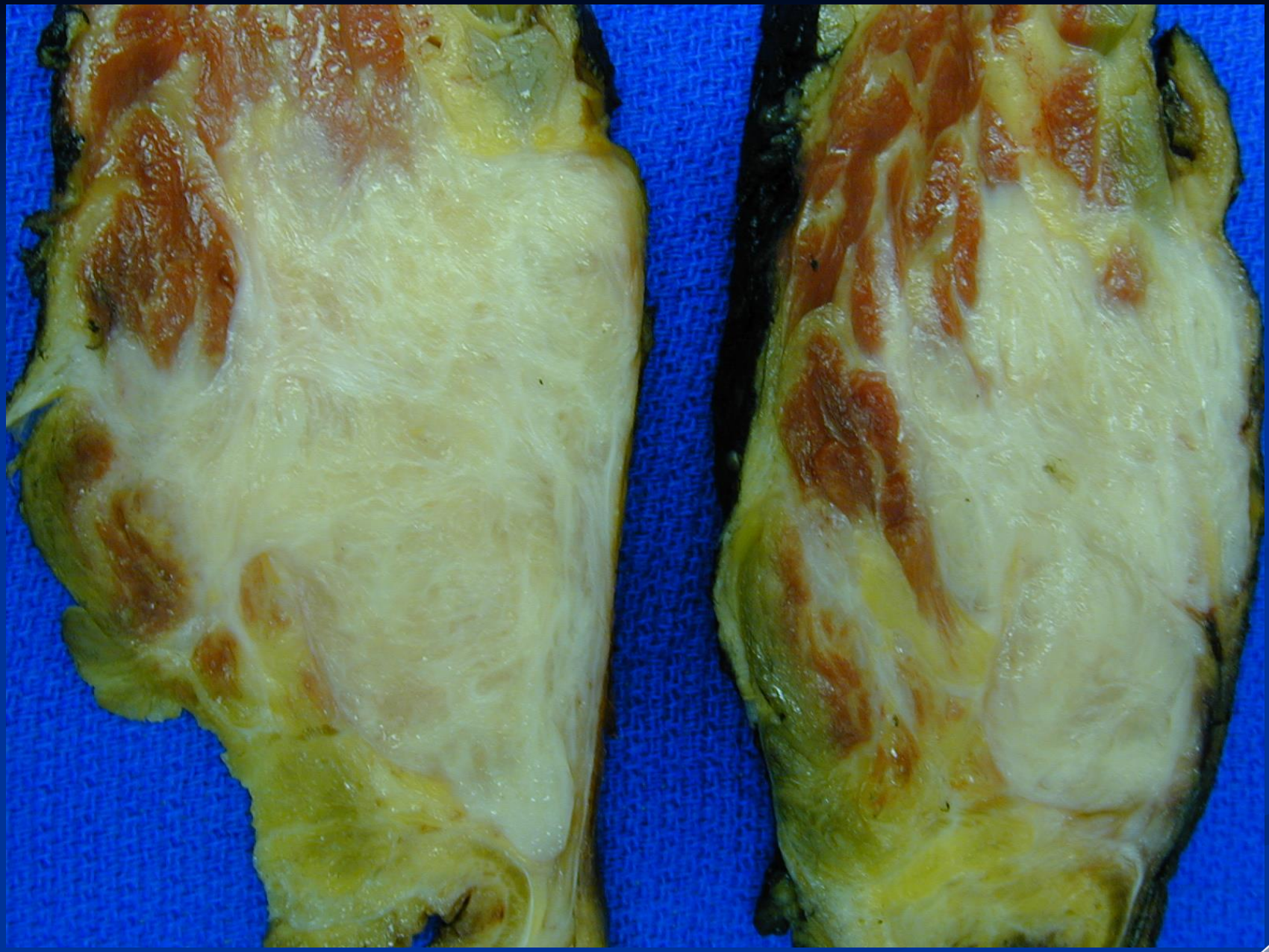

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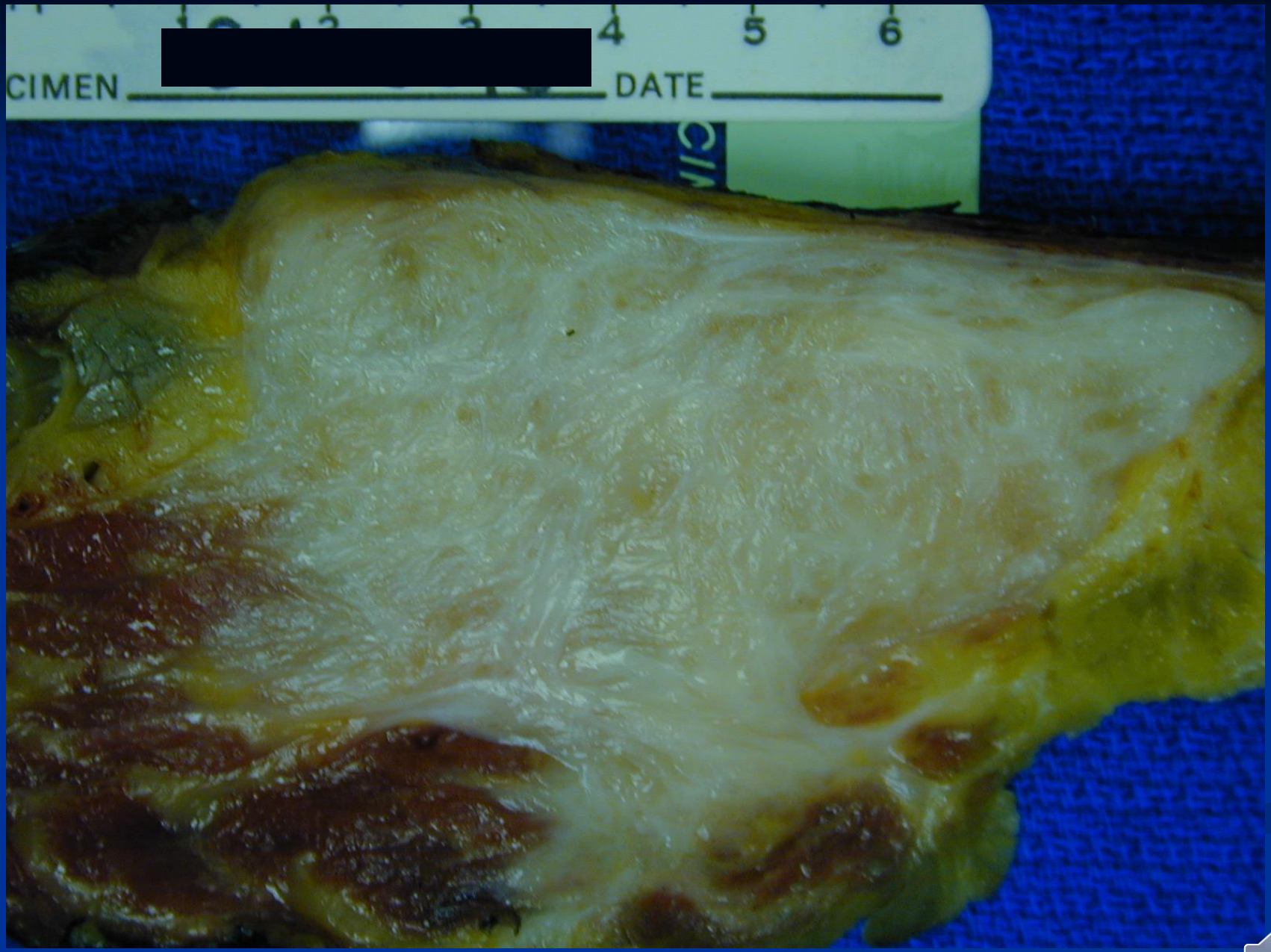

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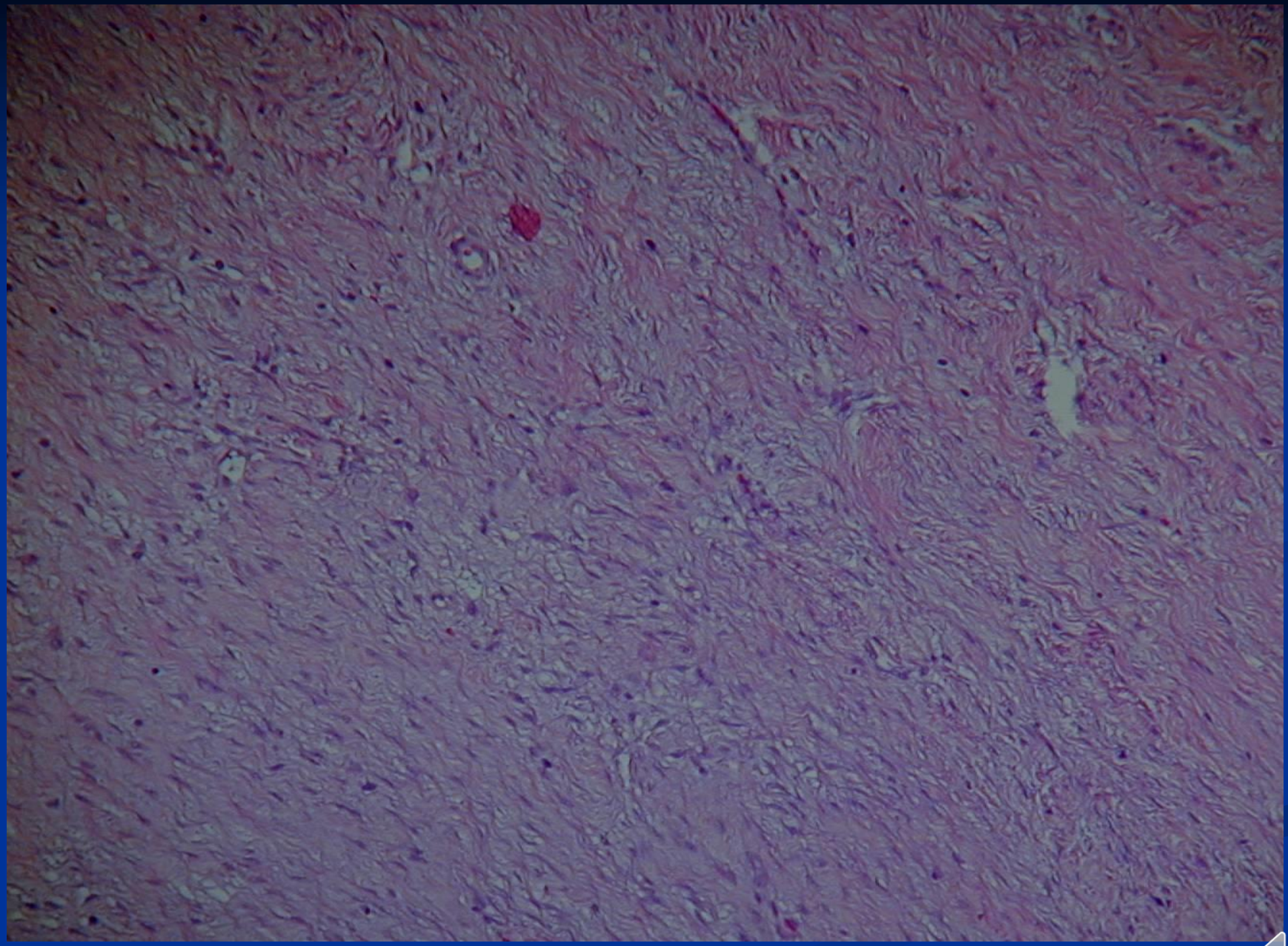

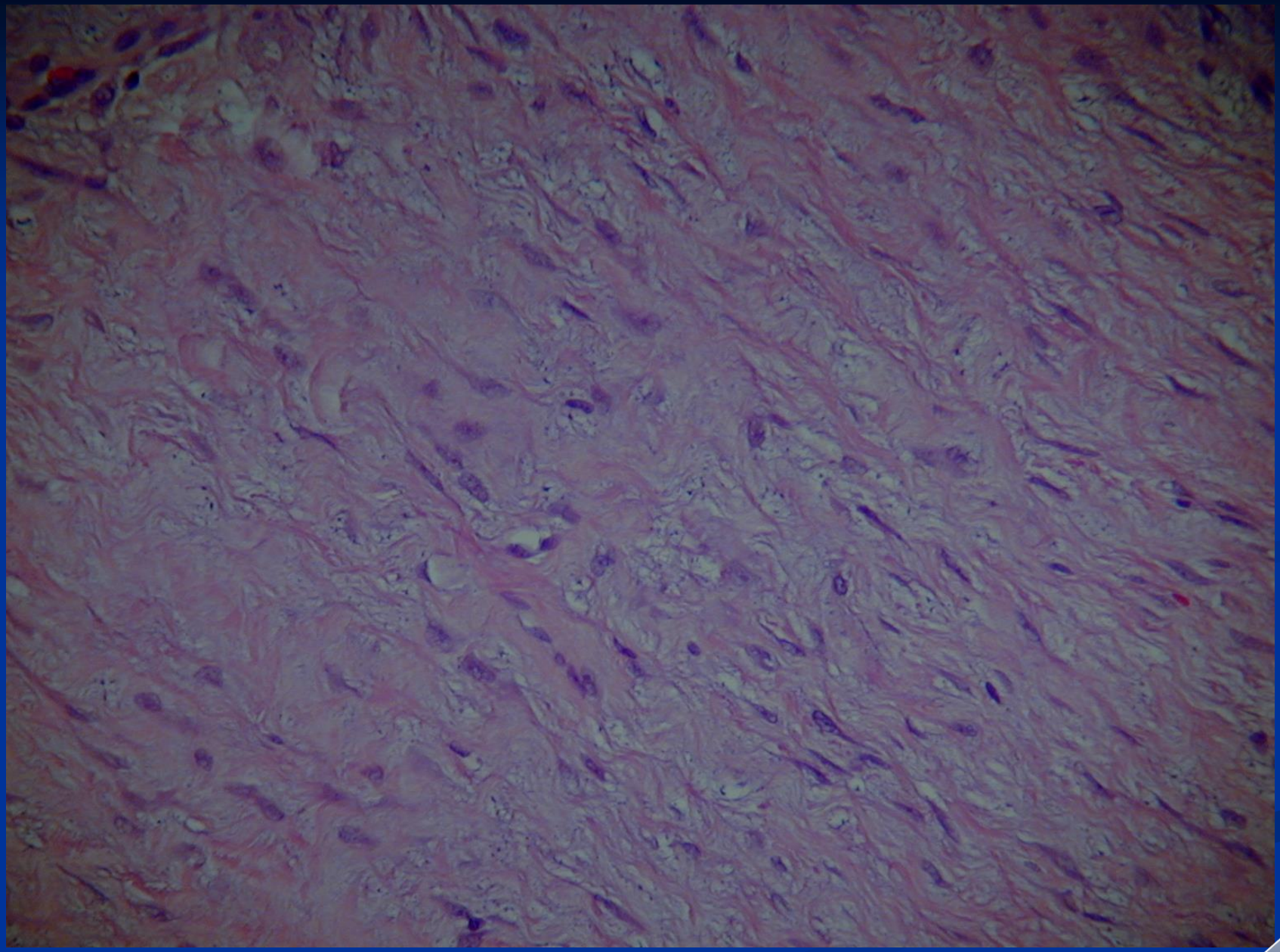

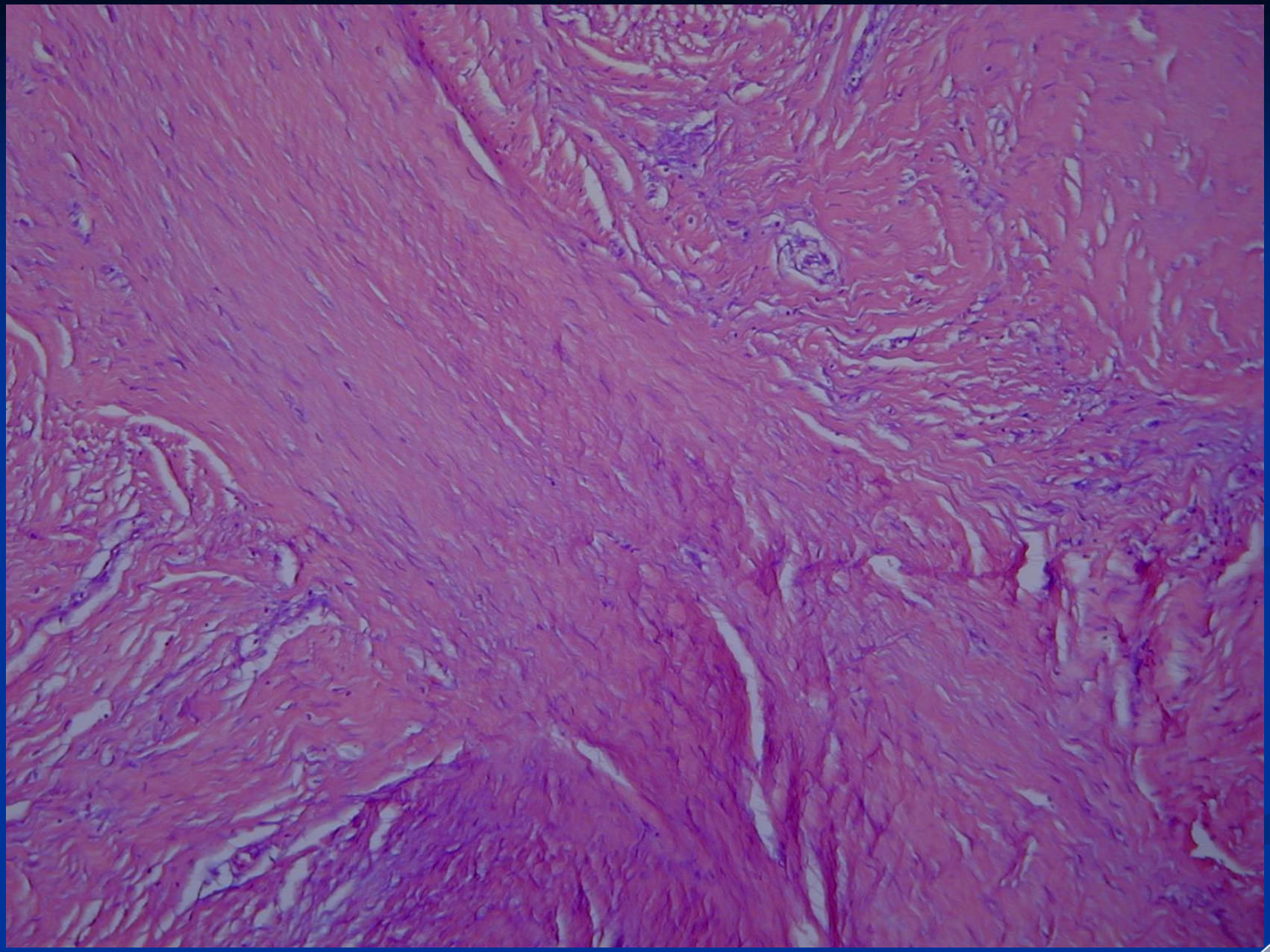

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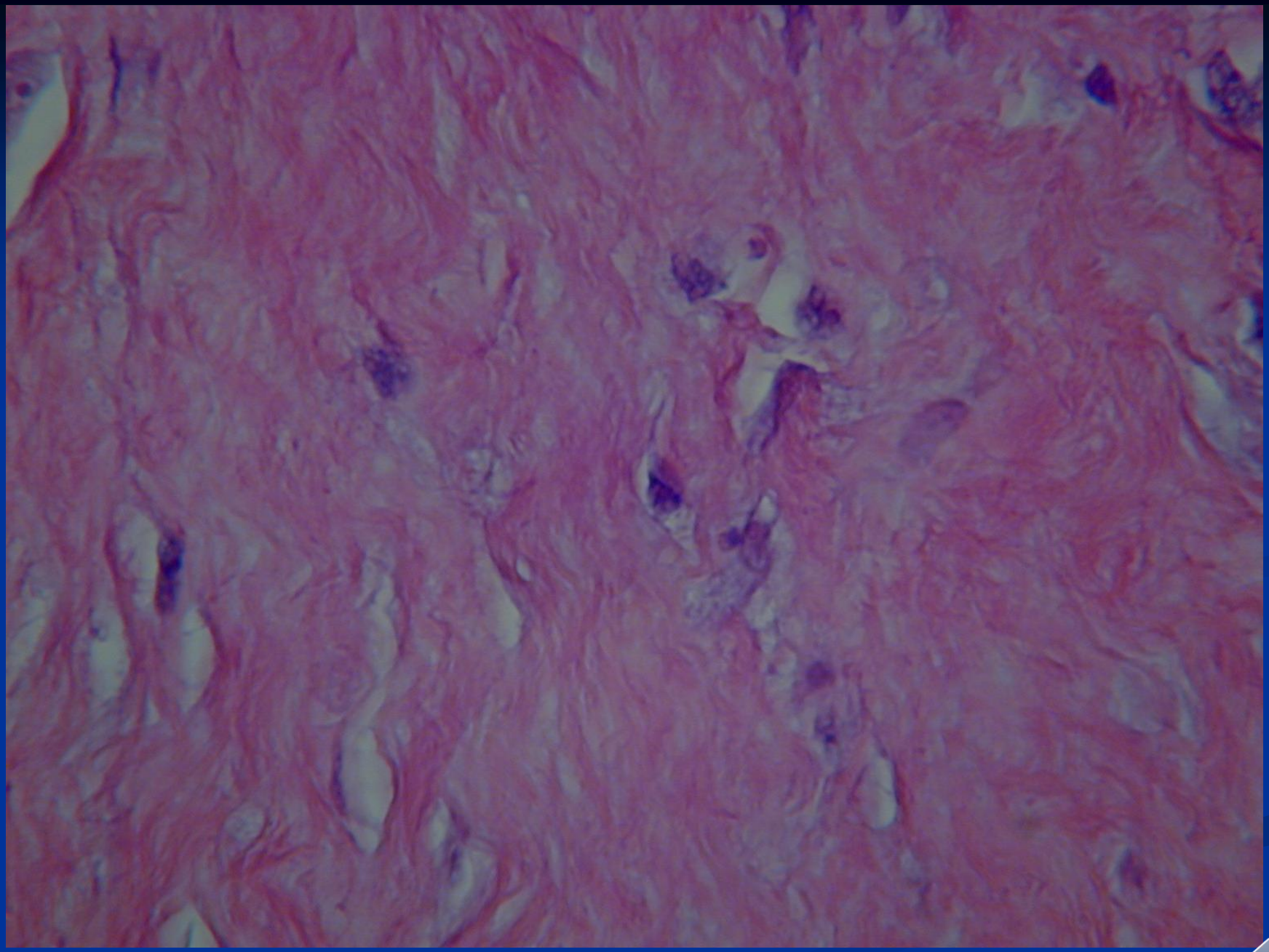

Wittig Orthopedic Oncology

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

Wittig Orthopedic Oncology

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)

Wittig Orthopedic Oncology

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

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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)

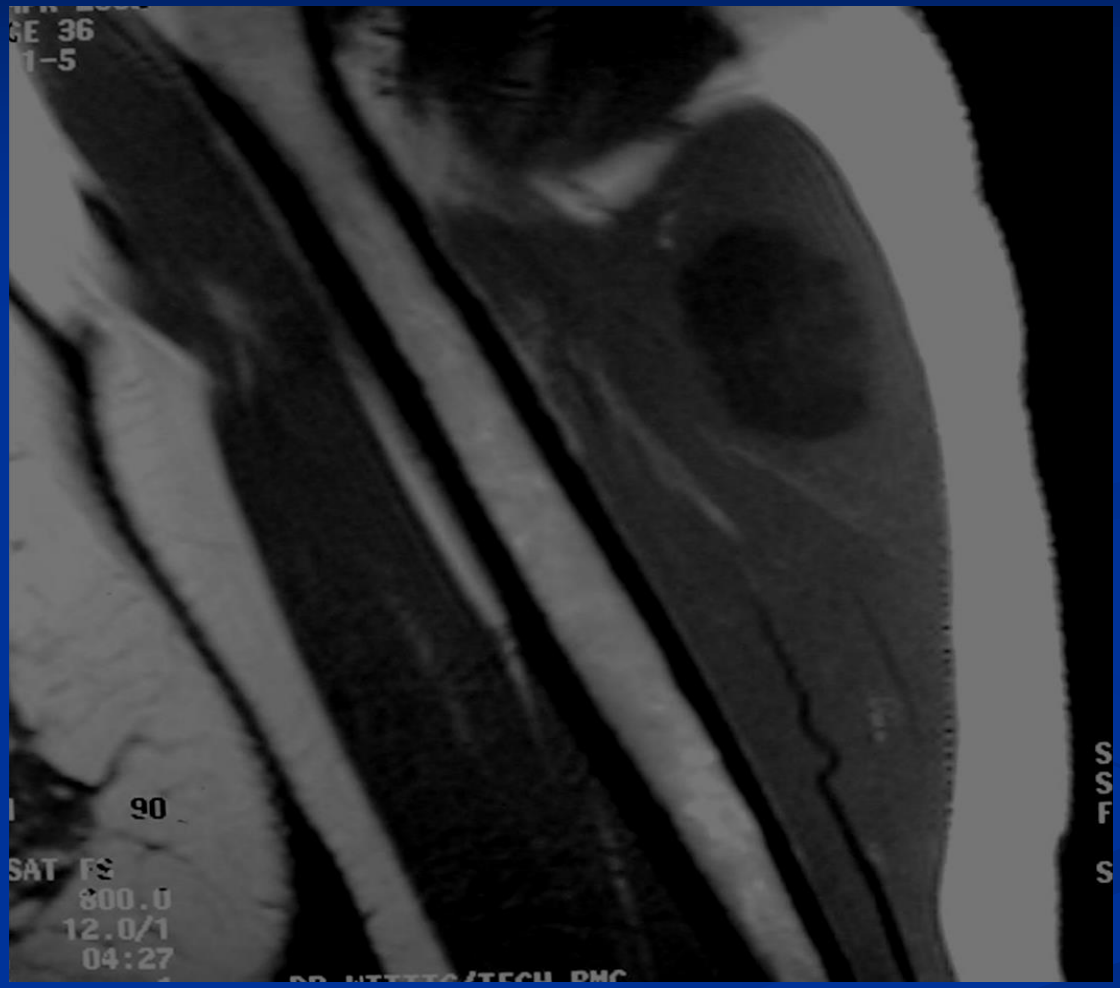
Wittig Orthopedic Oncology

Radiology: Myxoma

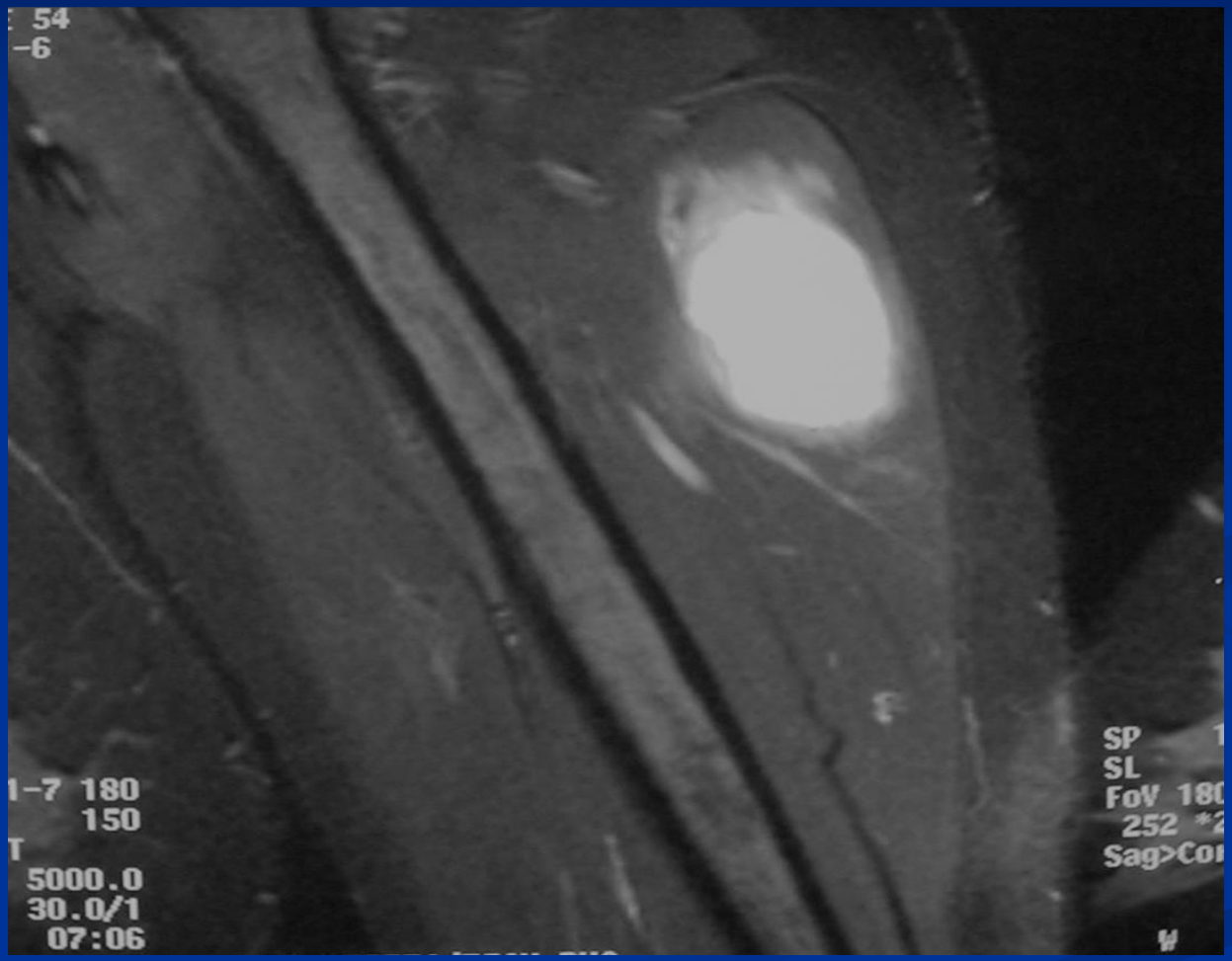
- Radiographs:
  - Normal or a nonspecific soft tissue mass
  - Mineralization is extremely rare
- CT:
  - Well defined, homogeneous soft tissue mass
  - No enhancement with CT contrast
- MRI: Appear similar to fluid or ganglion cysts
  - T1: Homogeneous mass; Signal lower than muscle
  - T2: Markedly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

Wittig Orthopedic Oncology

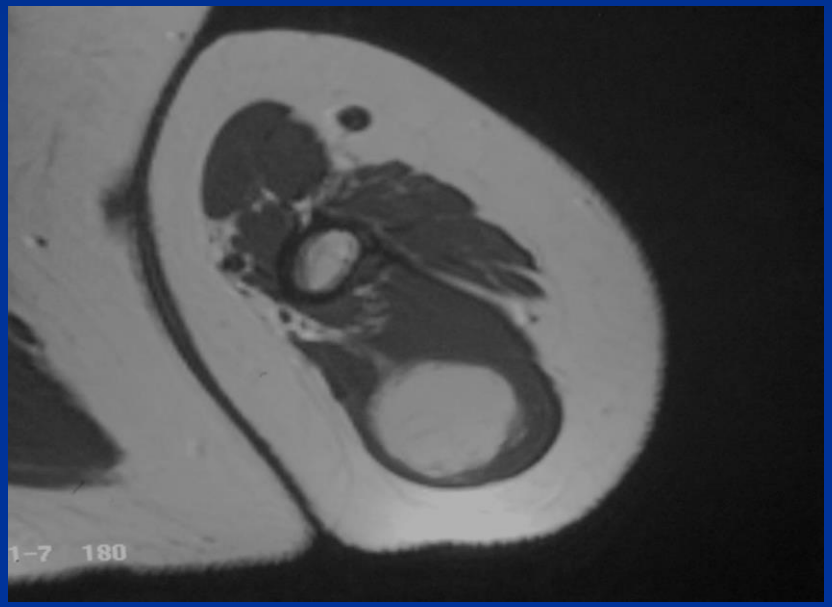
MRI T1: Myxoma of Triceps

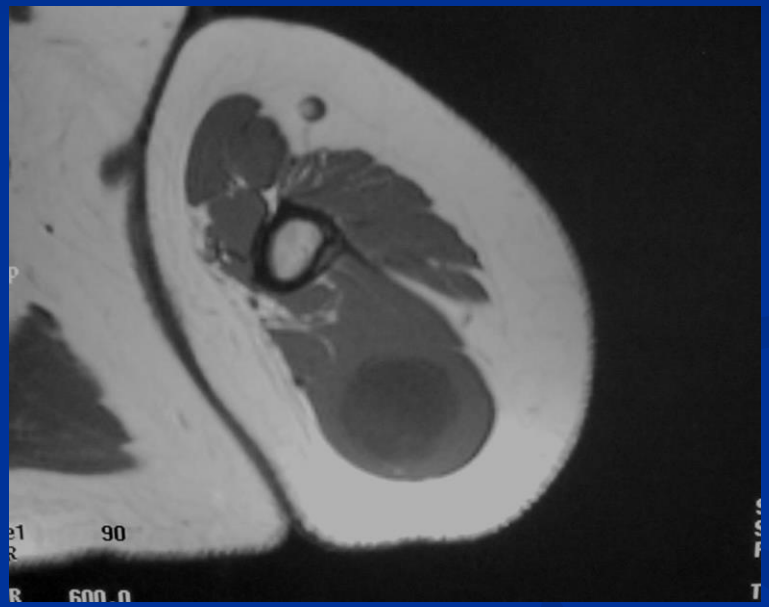


MRI T2: Myxoma of Triceps



MRI Myxoma of Triceps





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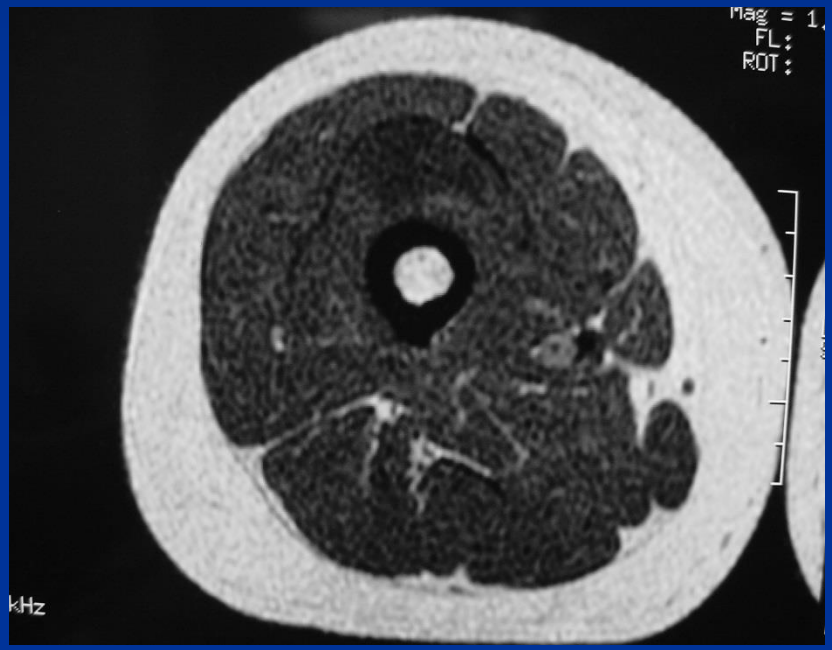
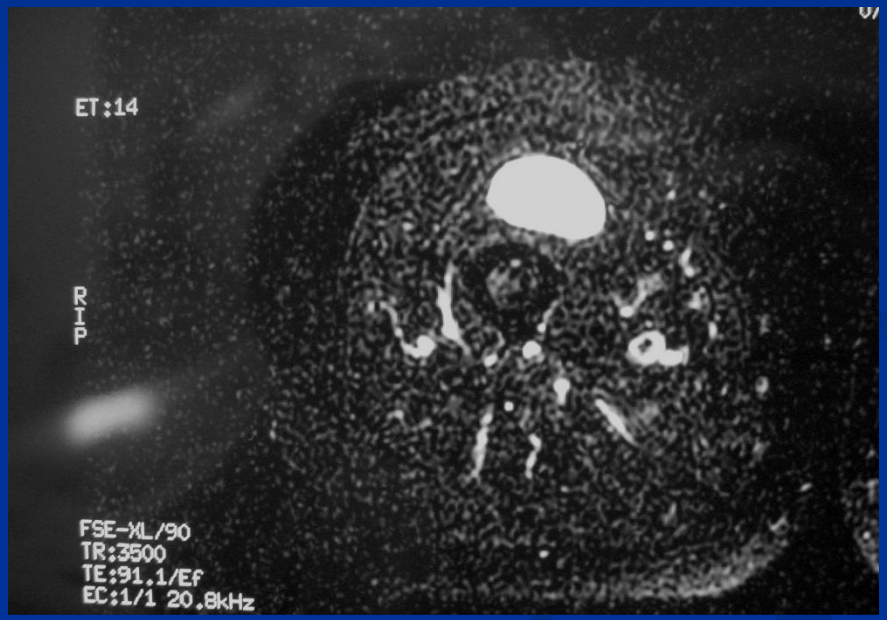
MRI: Myxoma of Thigh





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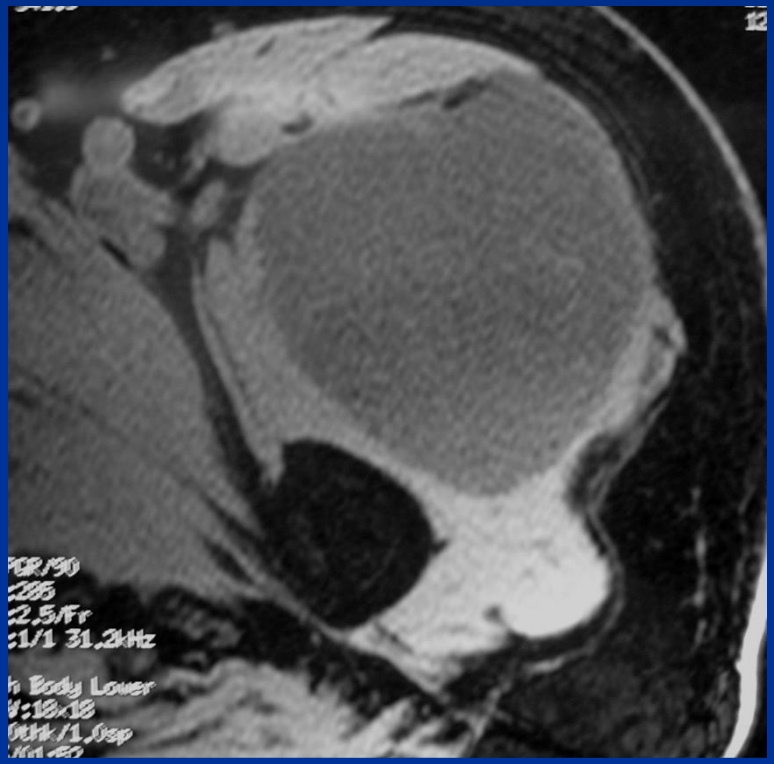
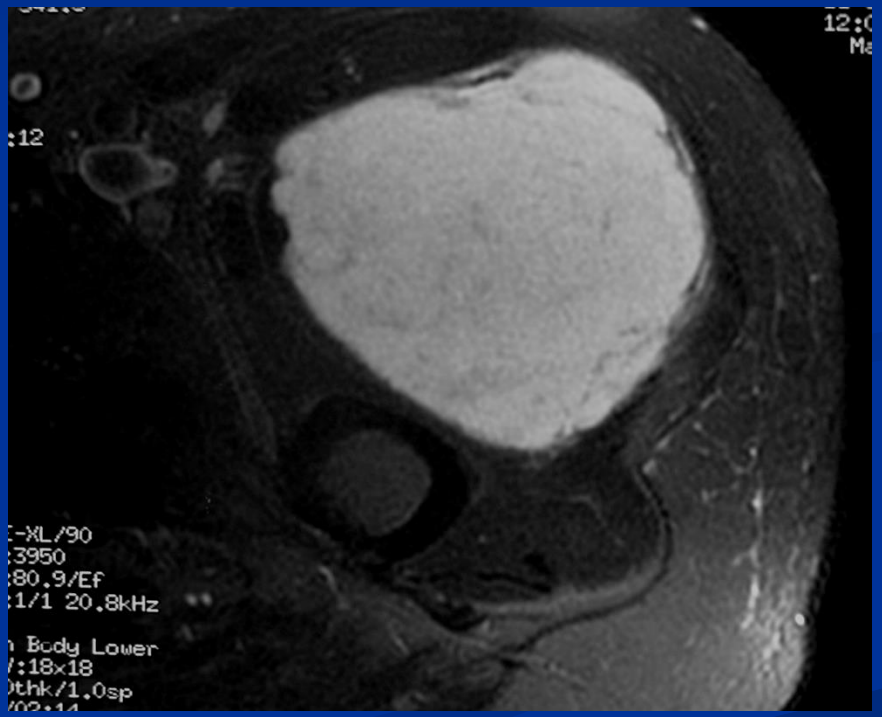
Myxoma of Thigh





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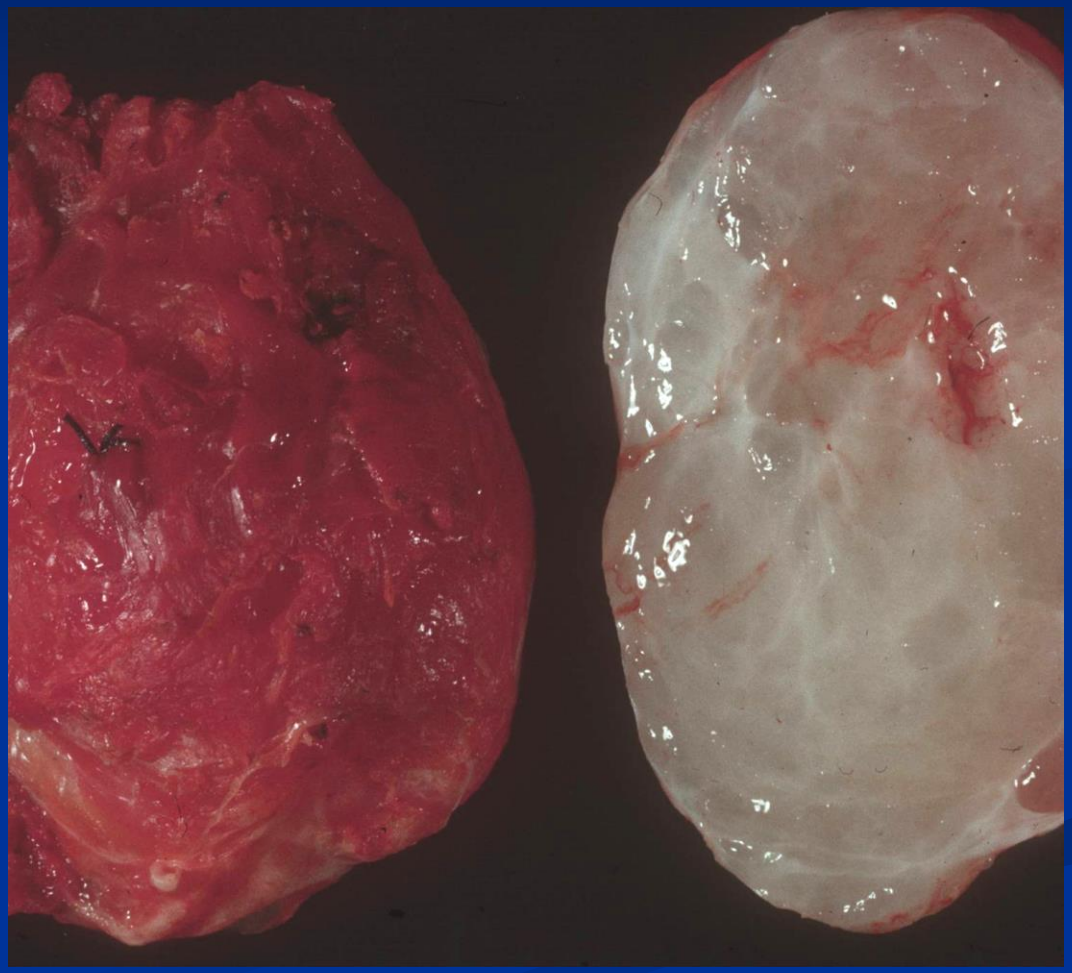
Pathology: Myxoma

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



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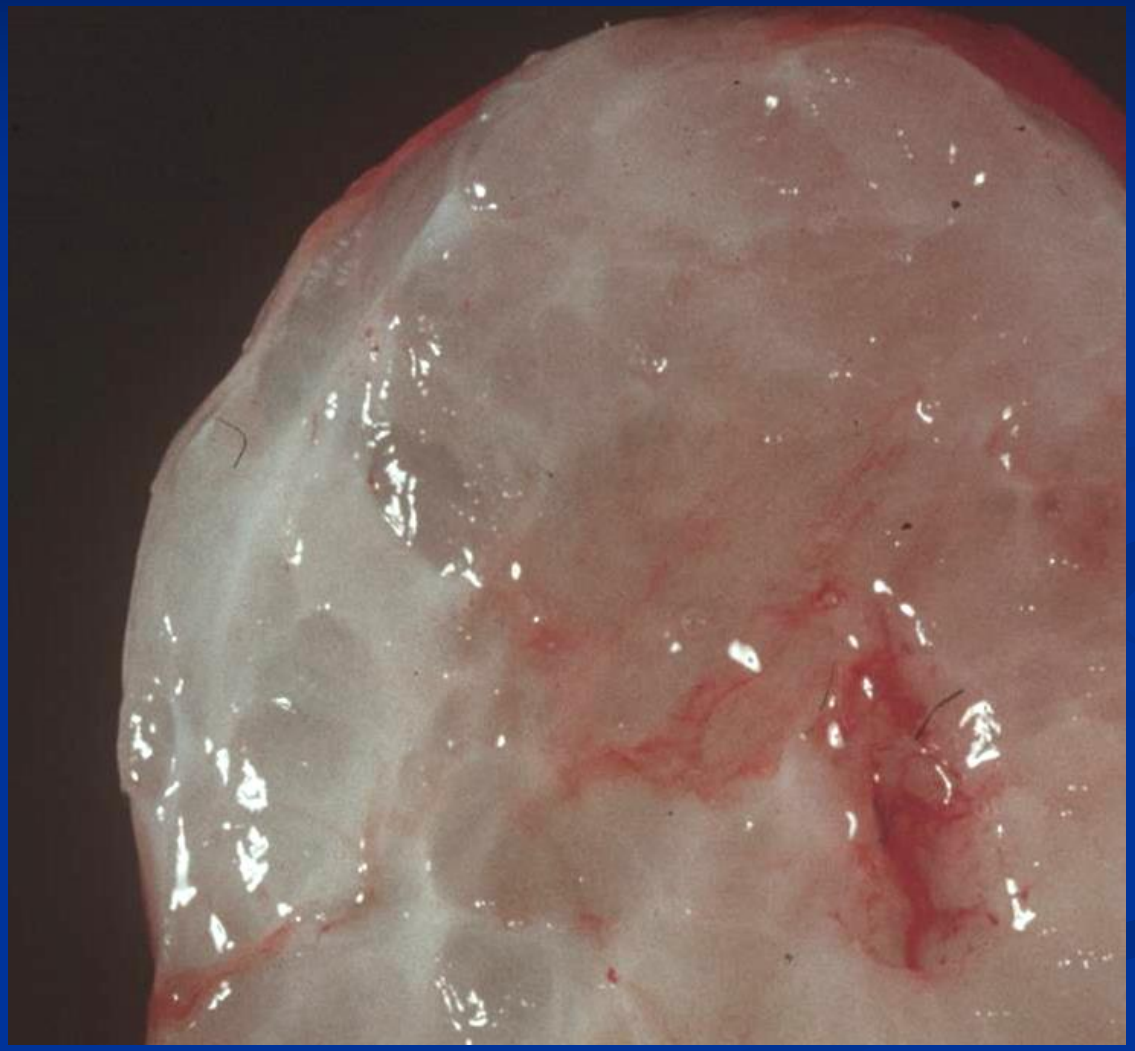
Myxoma



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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

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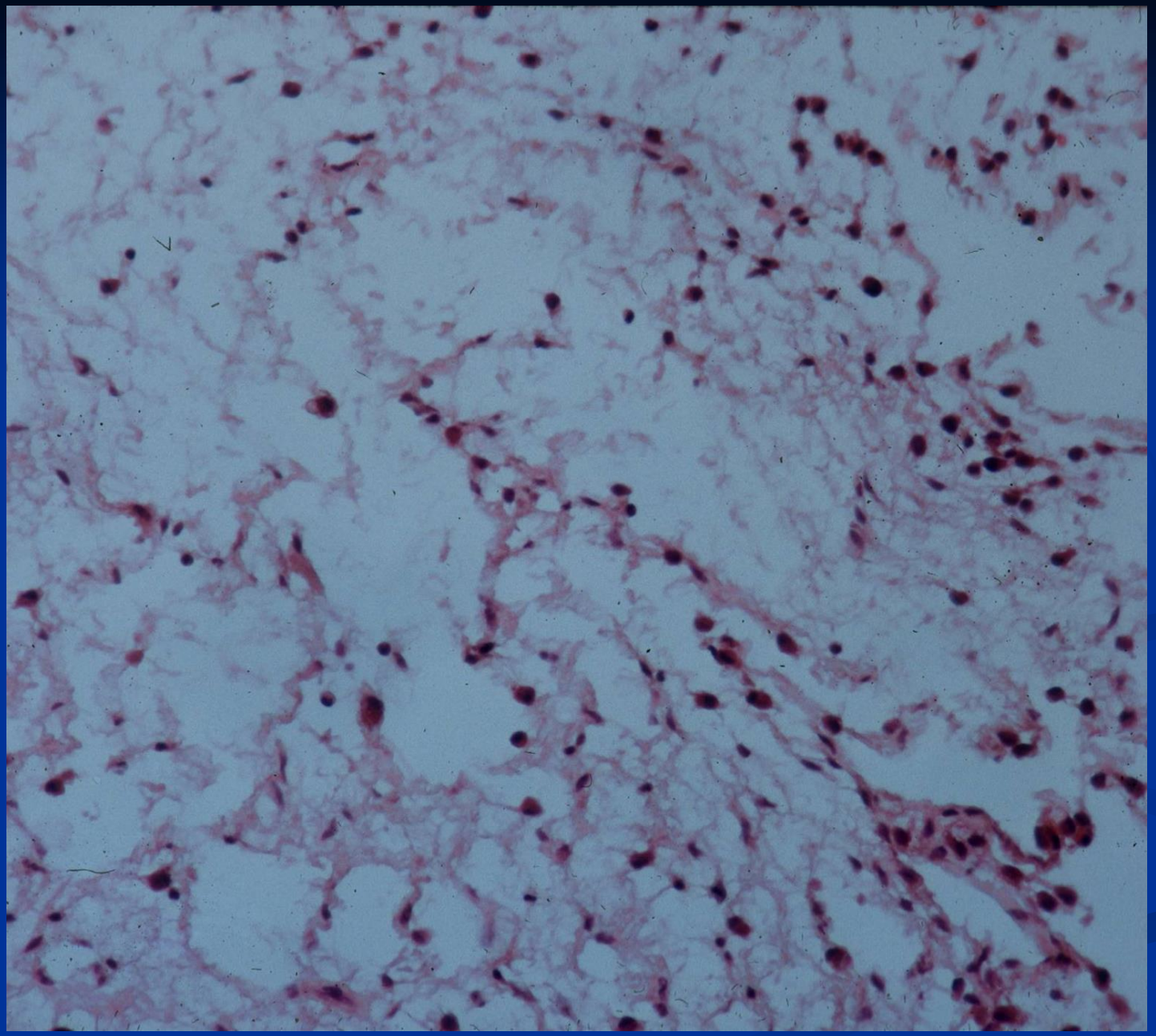
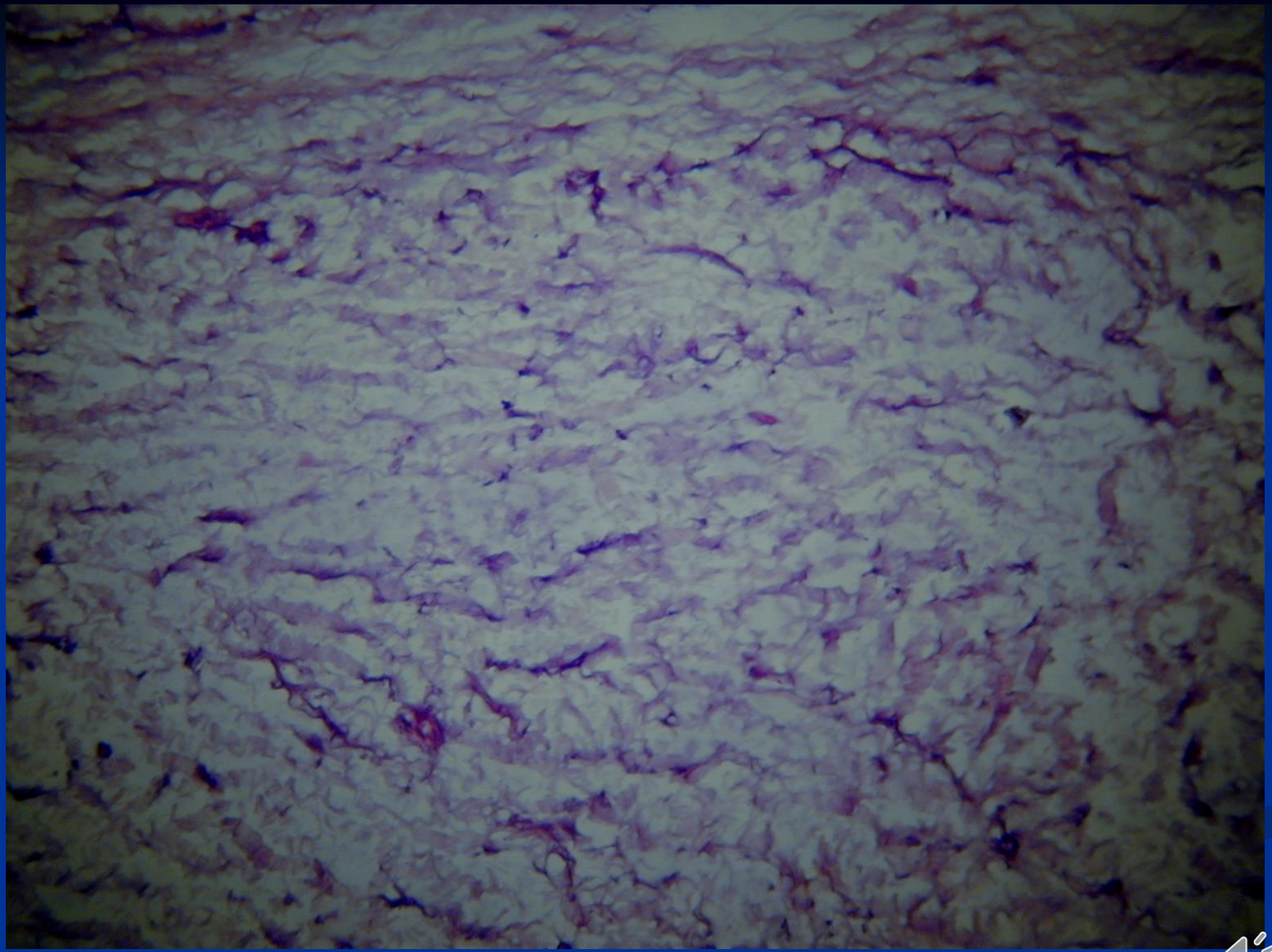


Wittig Orthopedic Oncology



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

Wittig Orthopedic Oncology

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

Wittig Orthopedic Oncology

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

Wittig Orthopedic Oncology

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 dumbell 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

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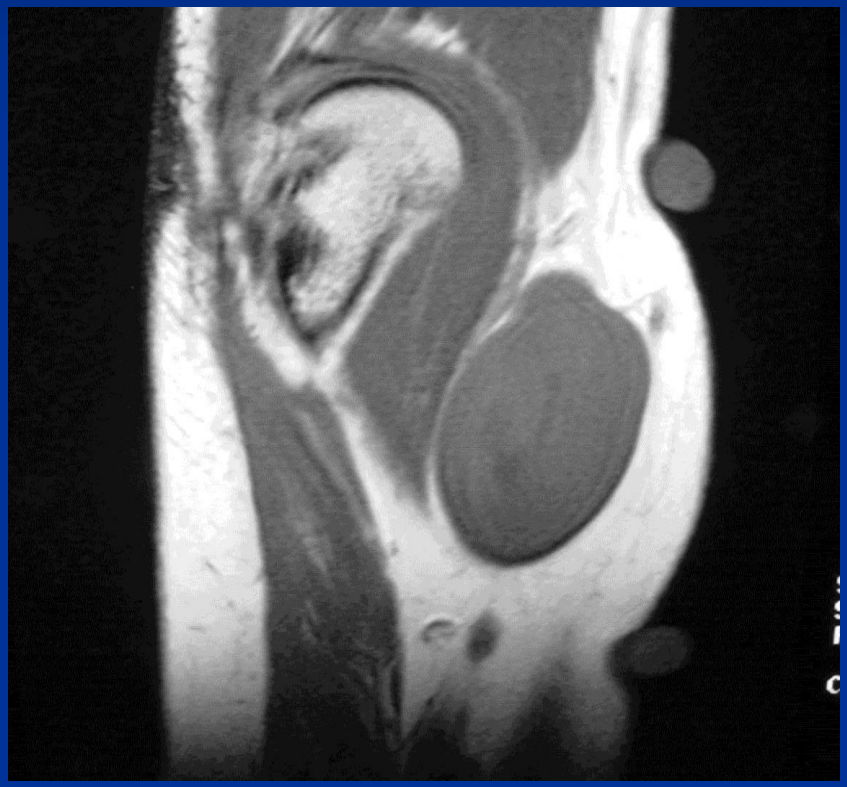
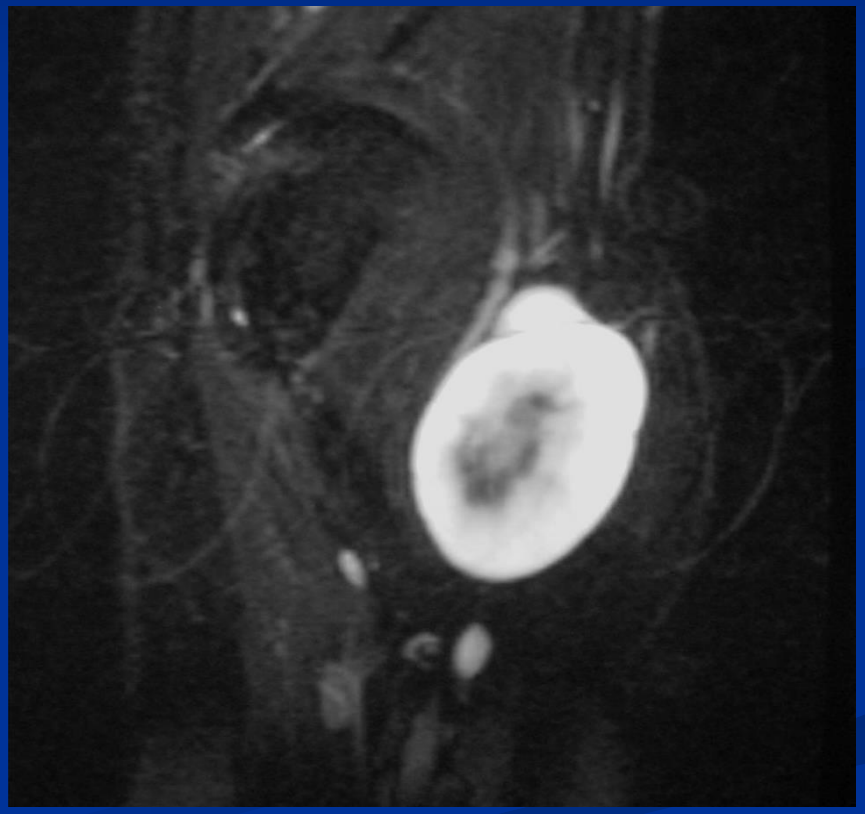
MRI T1 and T2: Schwannoma Target Sign





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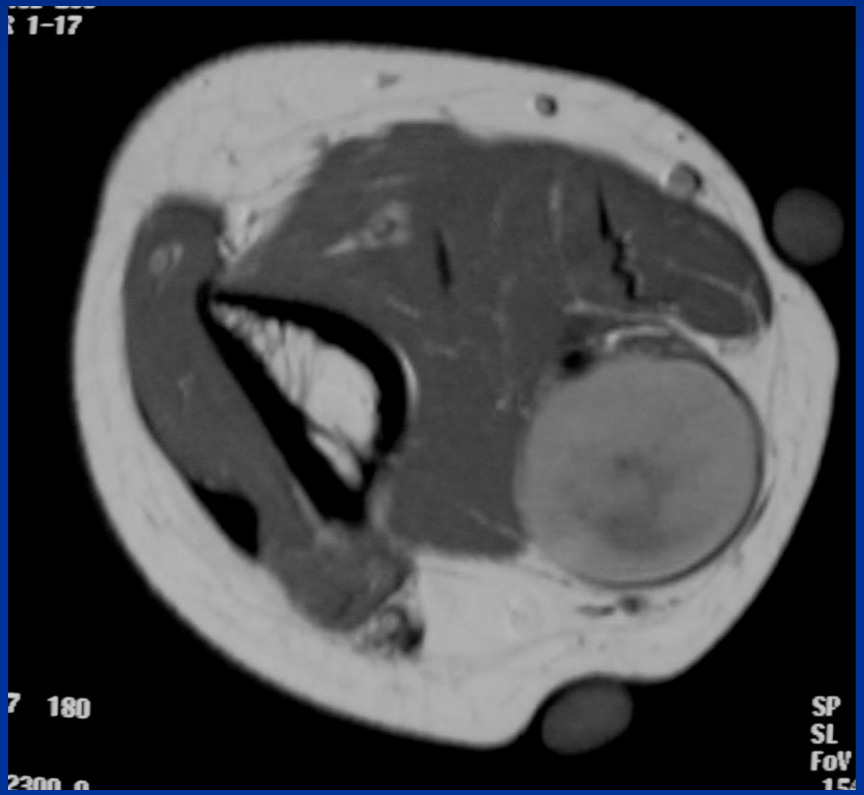
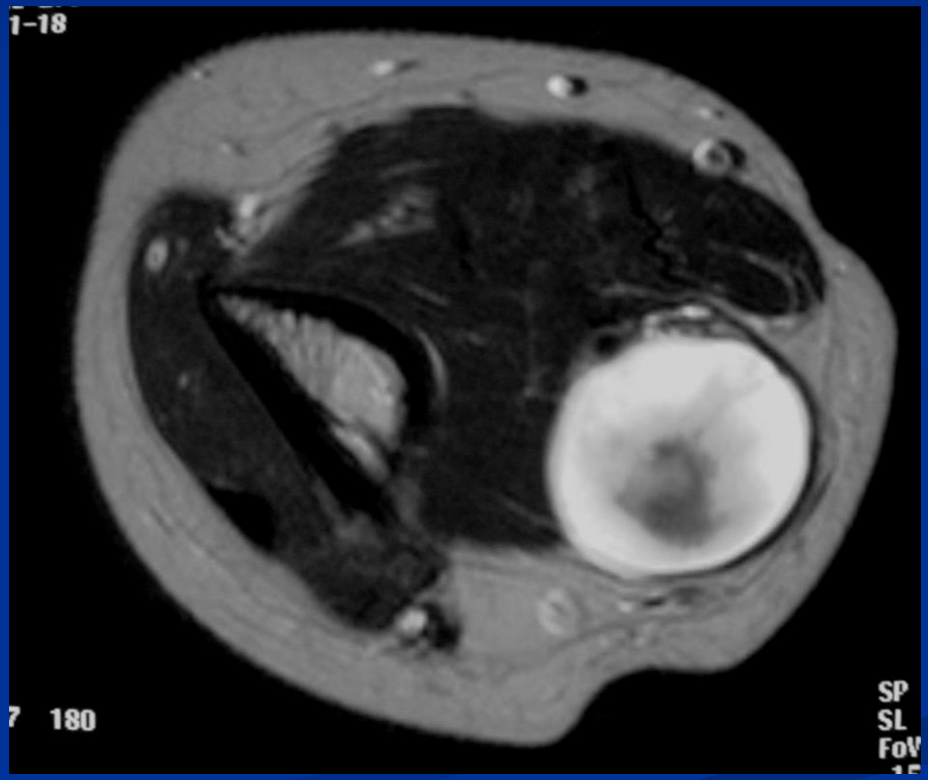
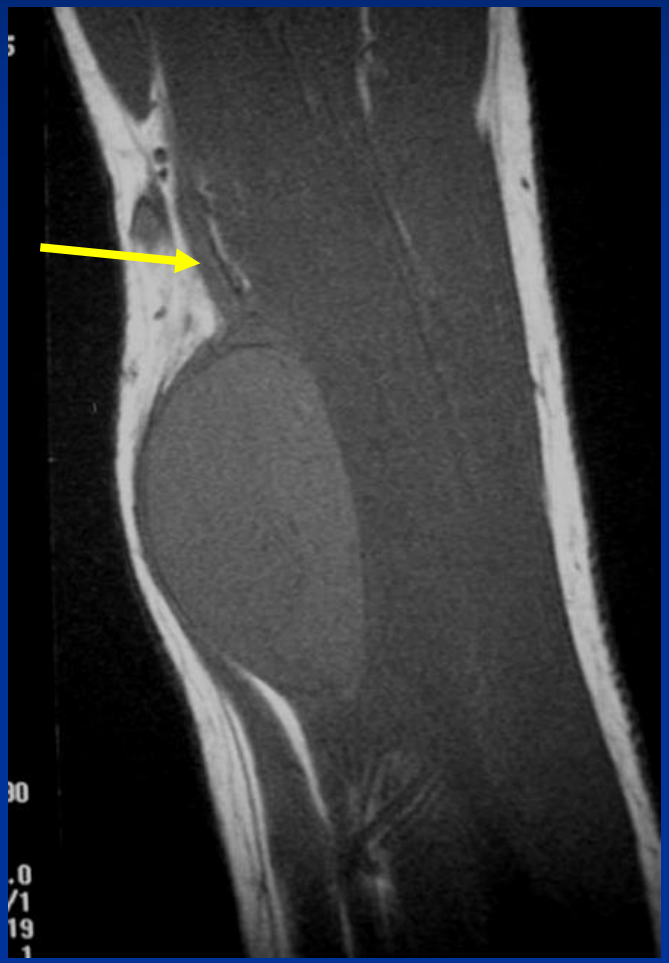
MRI: Schwannoma

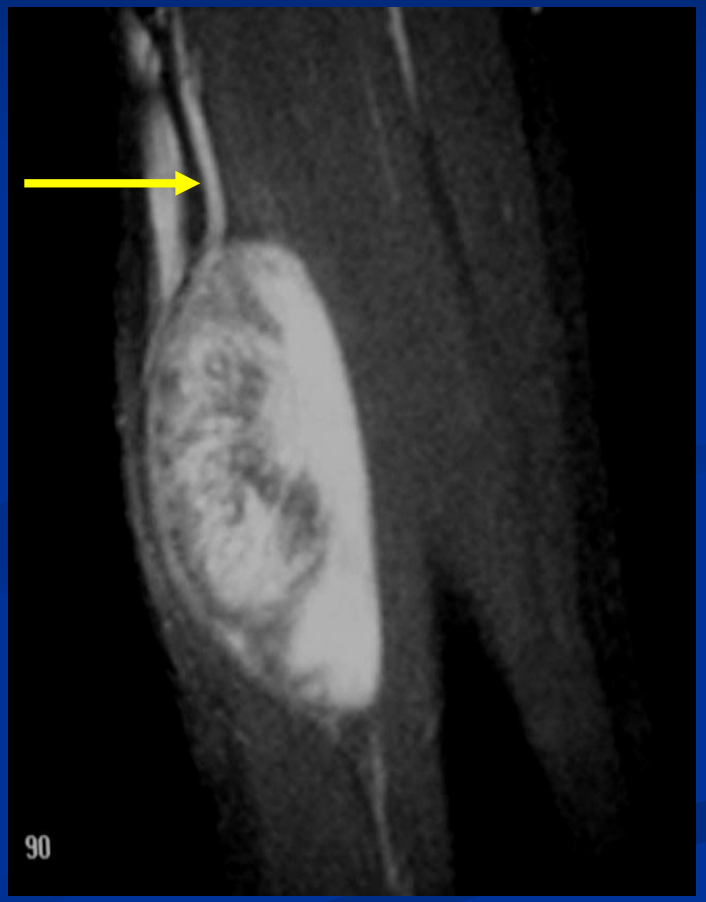
Intermuscular, Neurovascular Region, Target Sign





MRI Schwannoma: Nerve/Tubular Structure Entering Oblong Mass



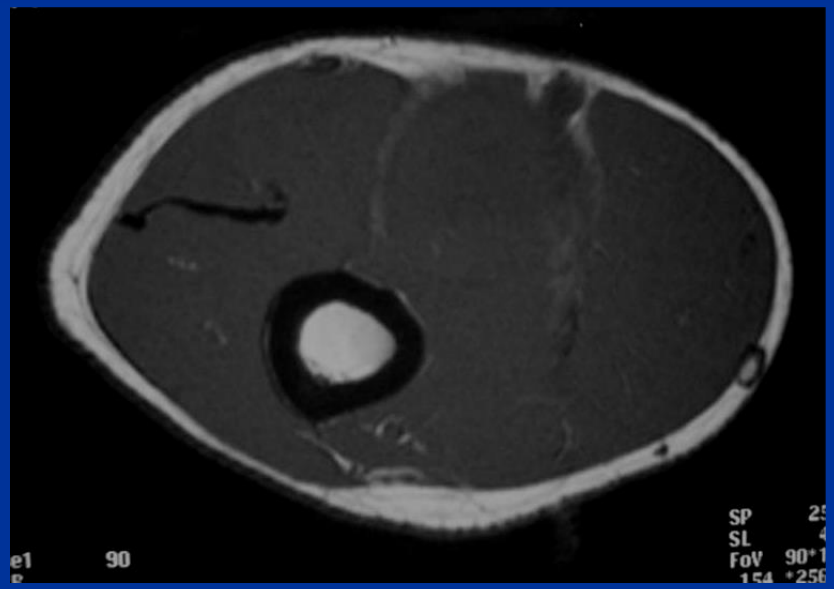


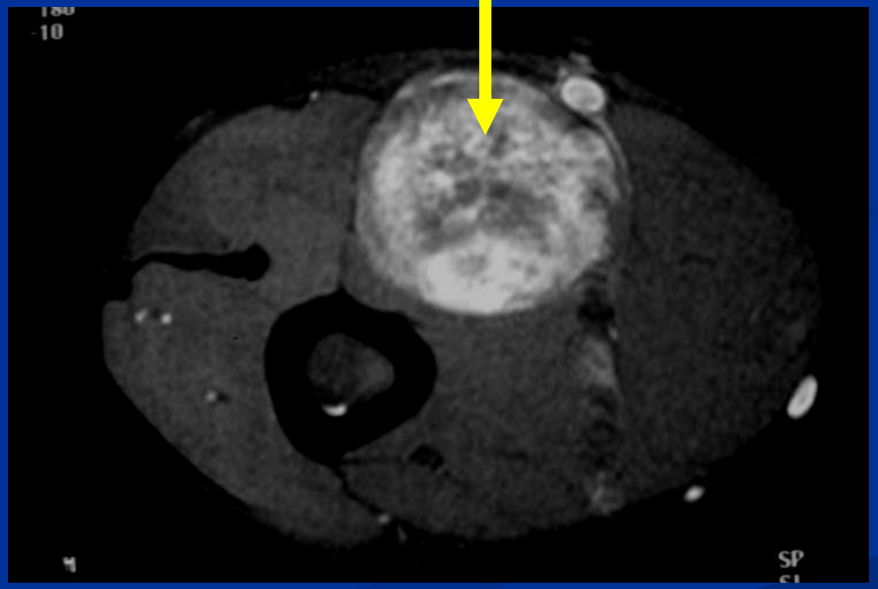
MRI: Schwannoma Median Nerve

Target Sign on T2 (arrow)

Hyperintense Area: Myxoid Antoni B

Split Fat Sign on T1





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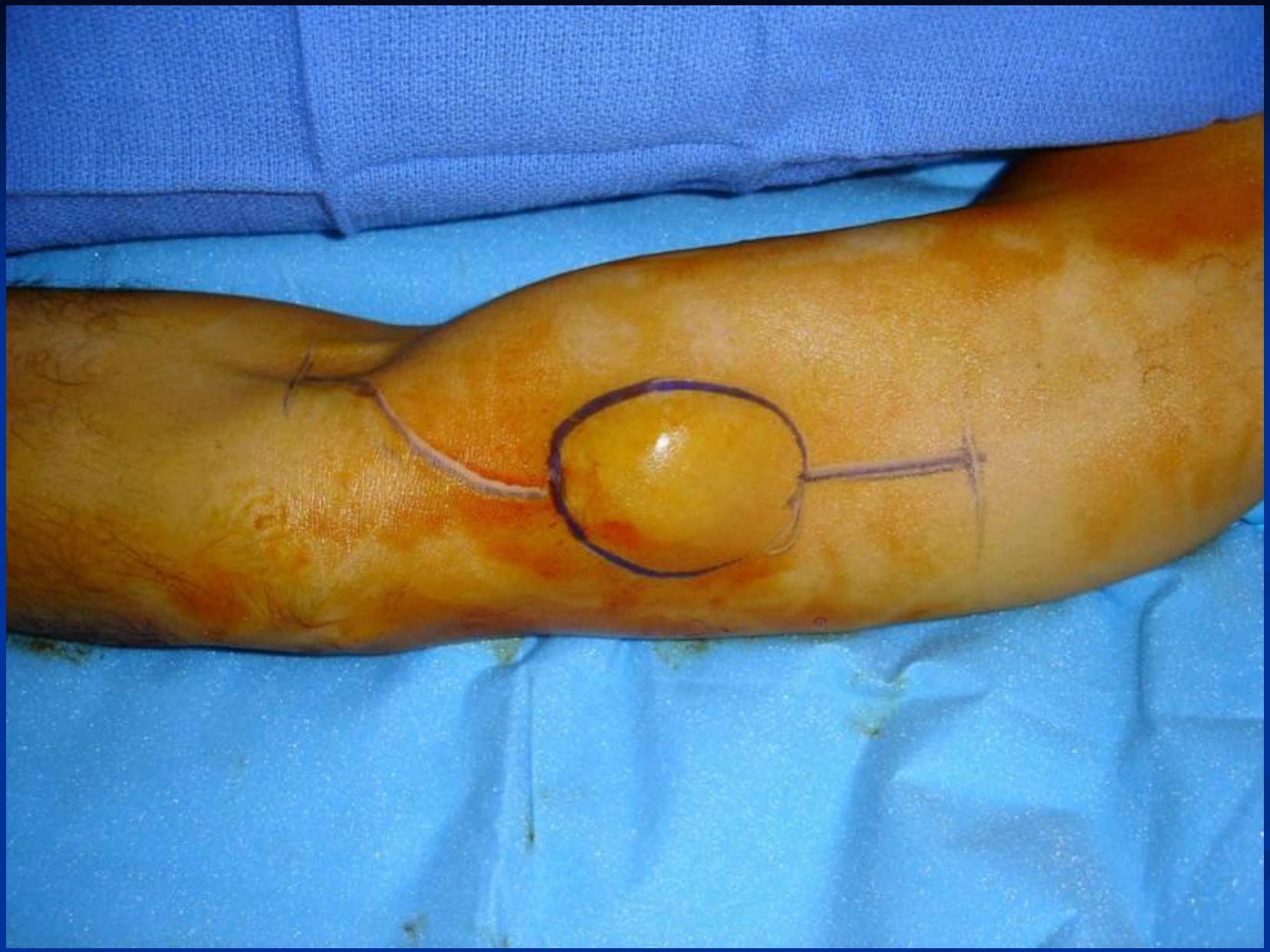

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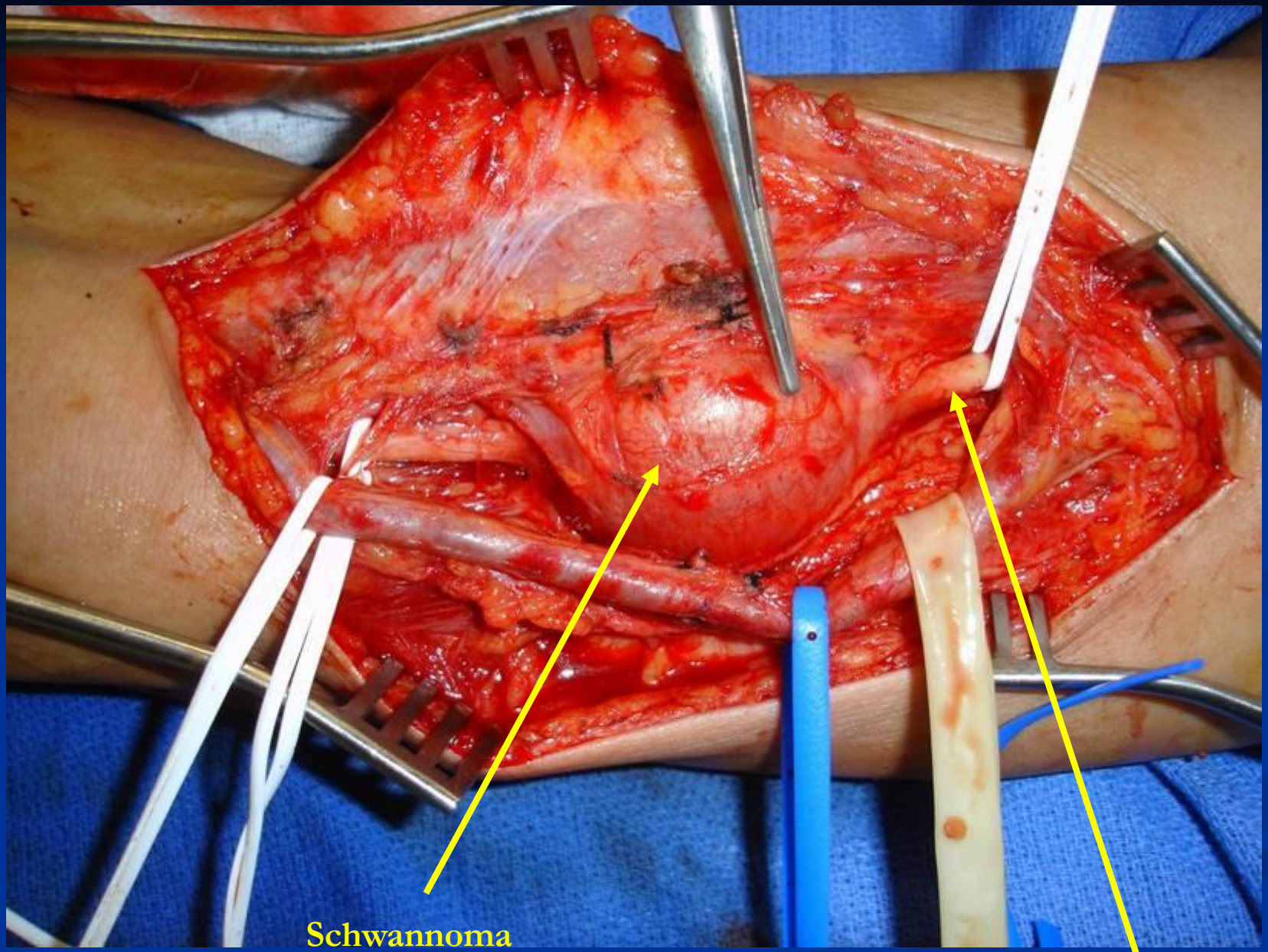

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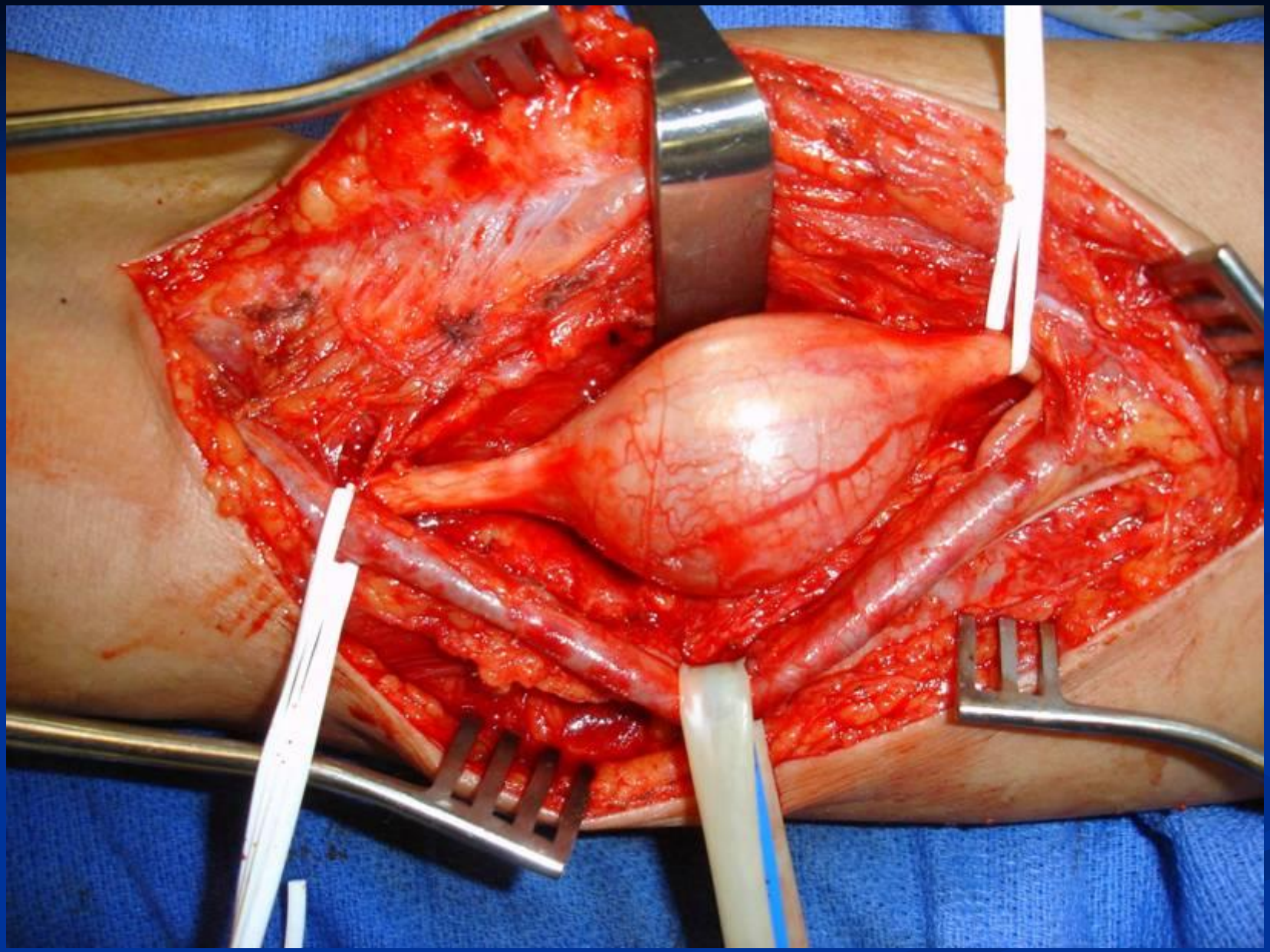

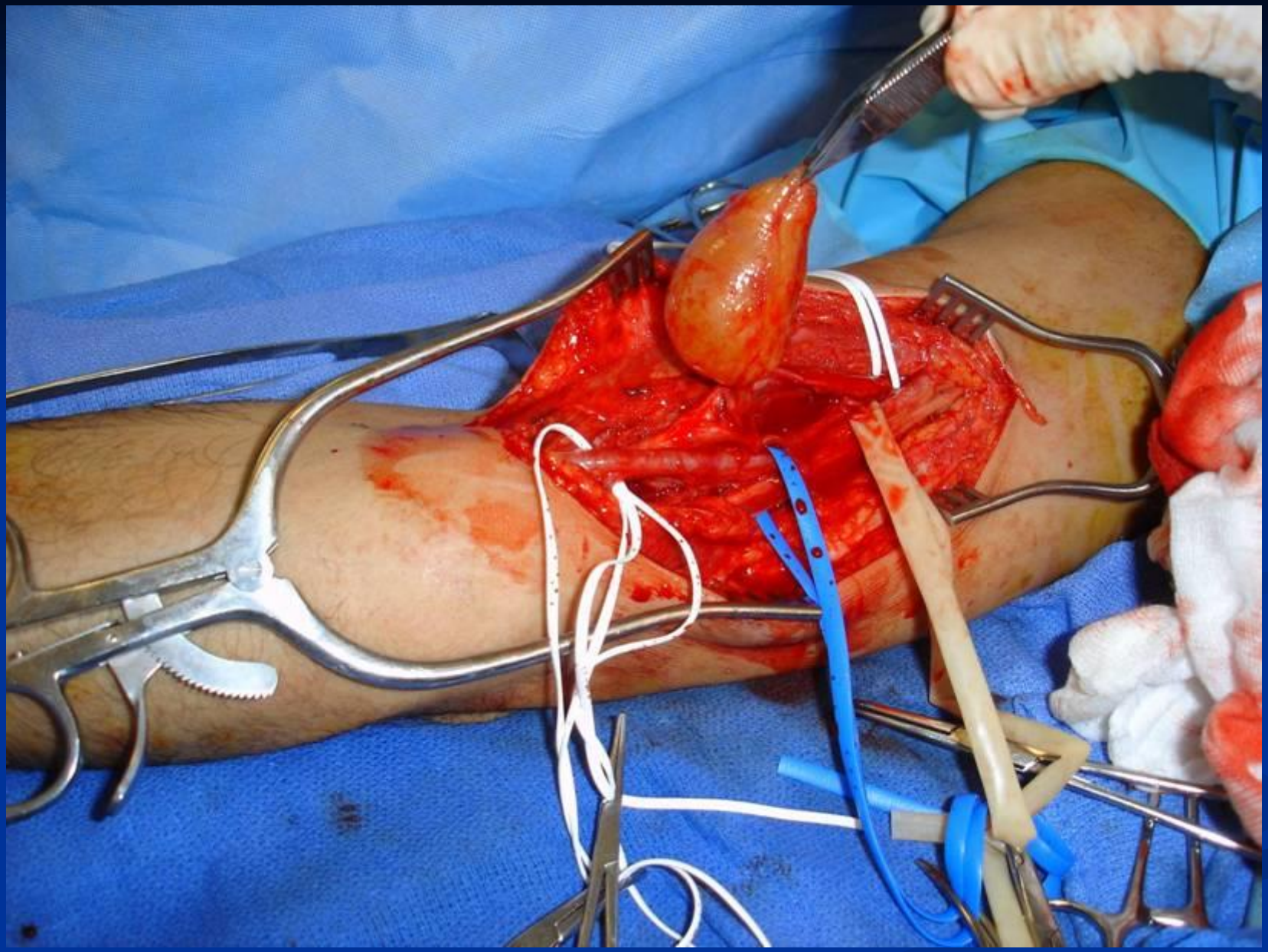

Nerve Sheath Opened and Schwannoma Removed; Median Nerve Left Intact

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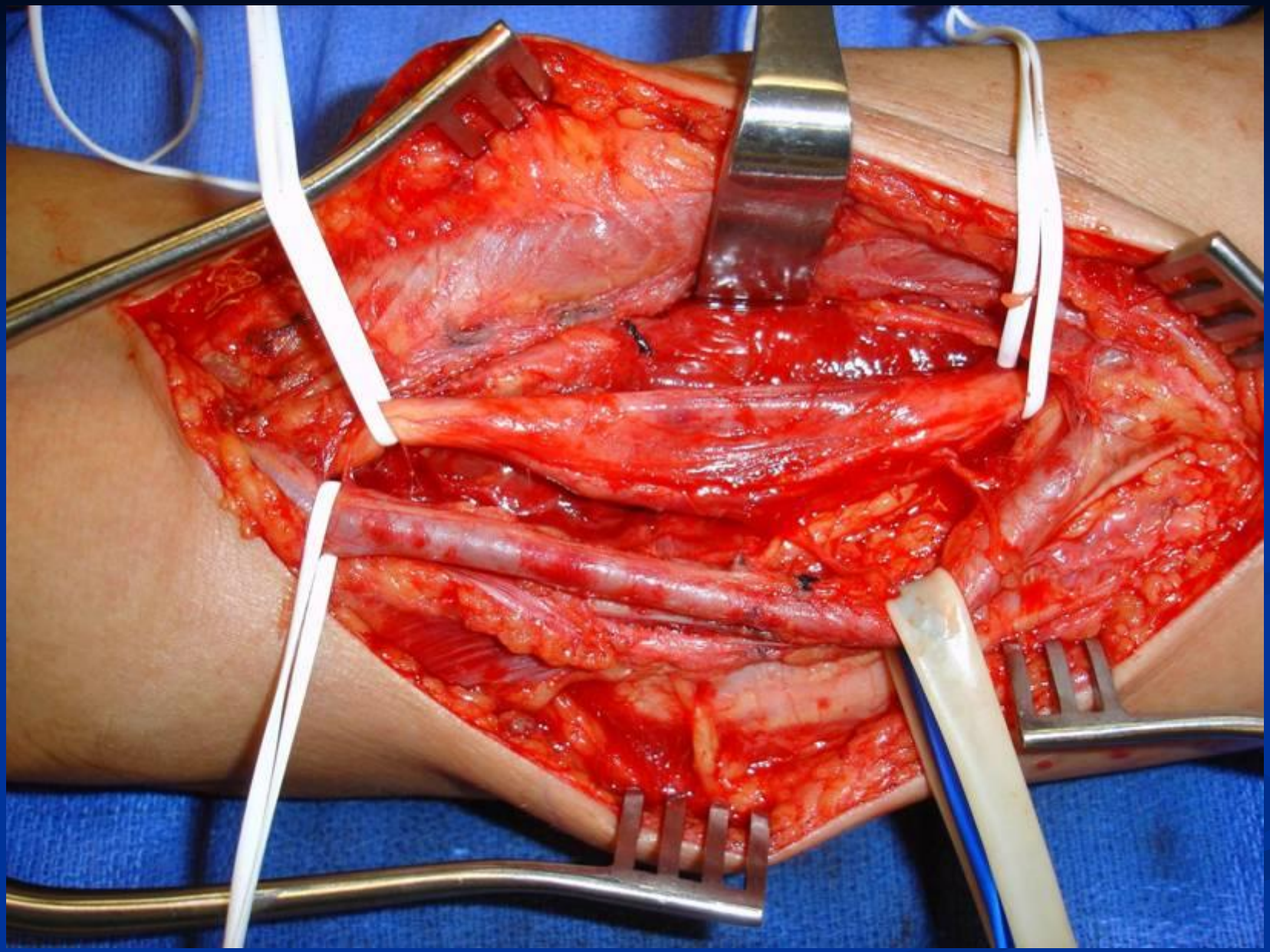

Median Nerve Intact

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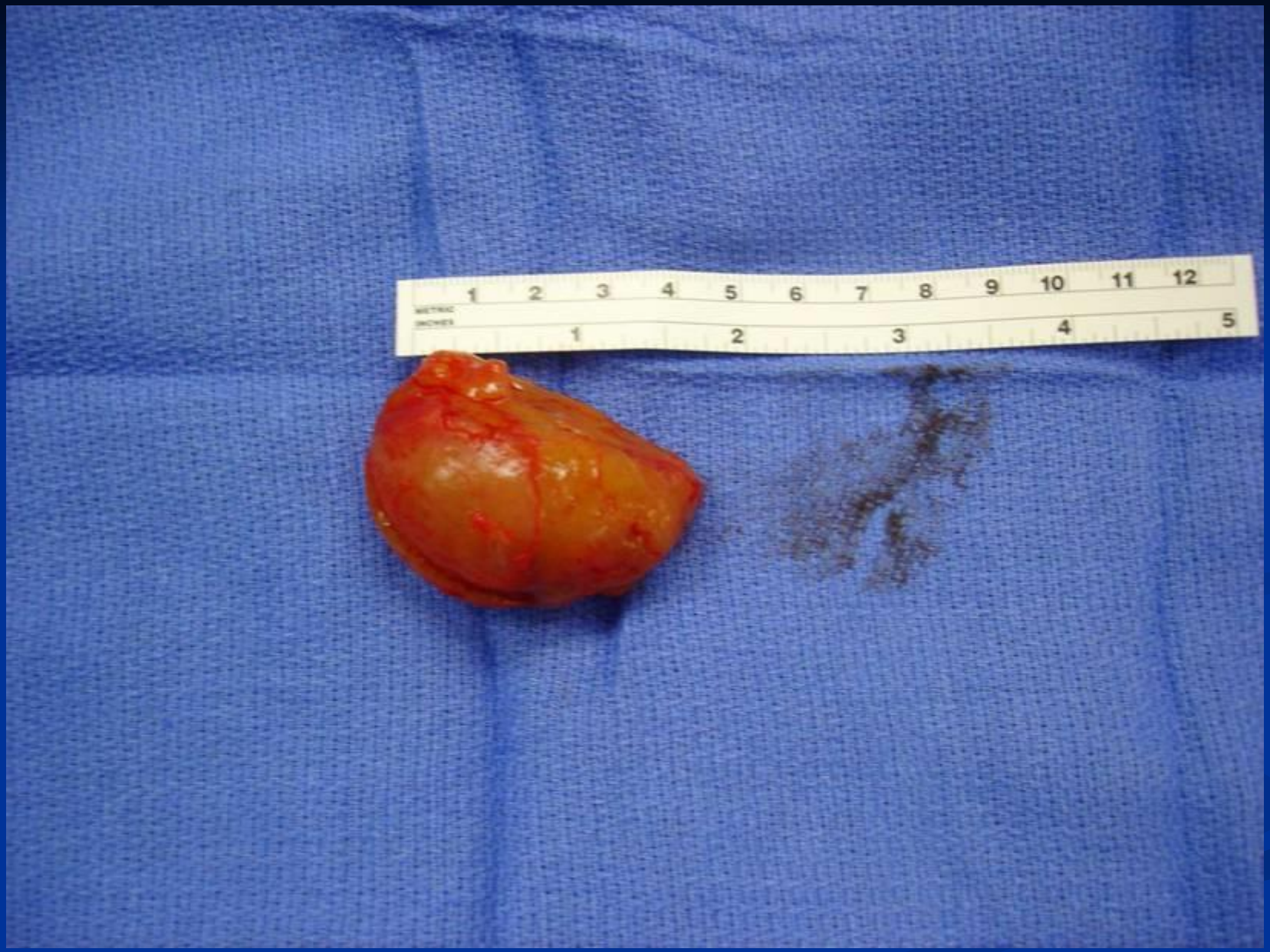
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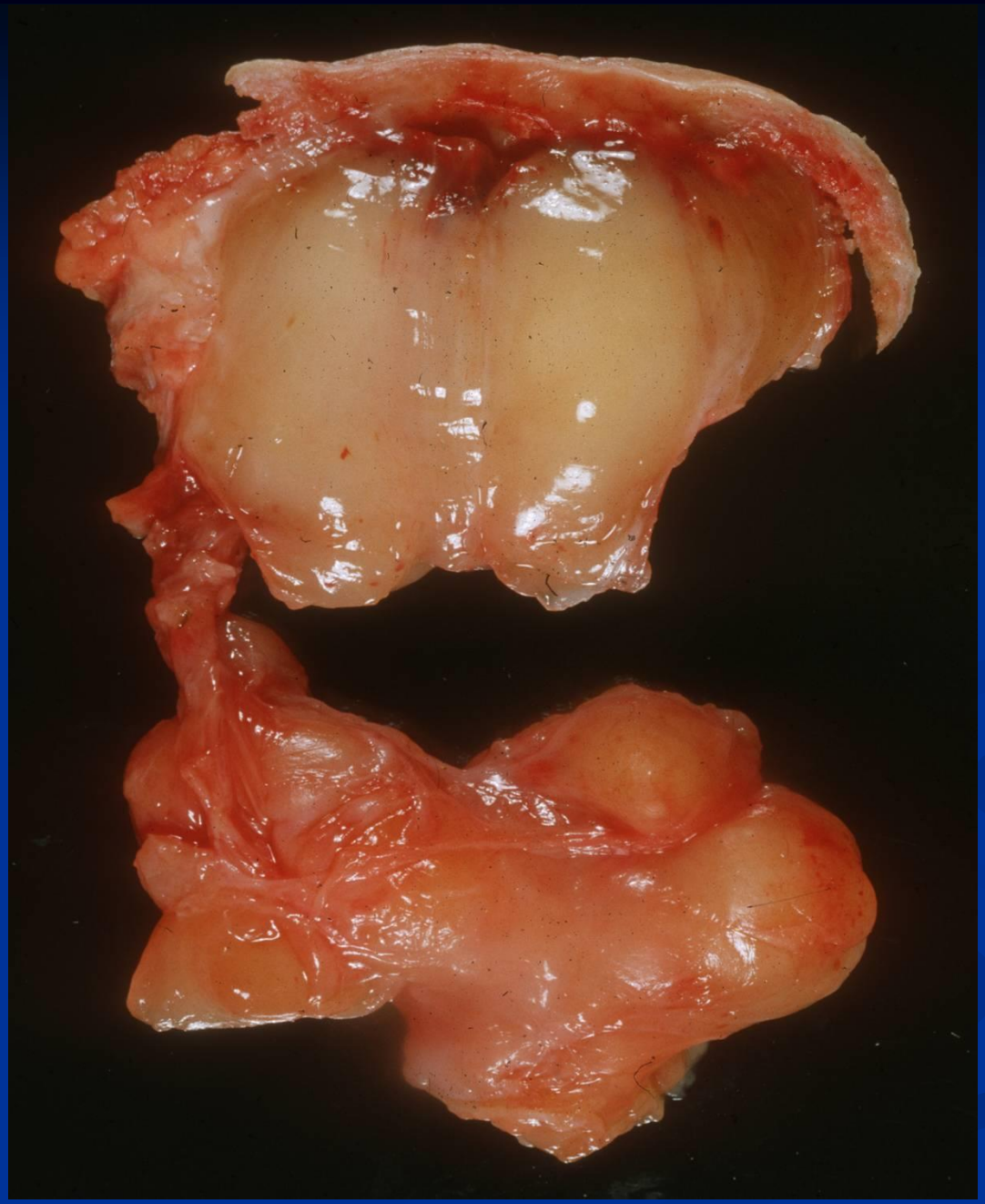
1 2 3 4 5



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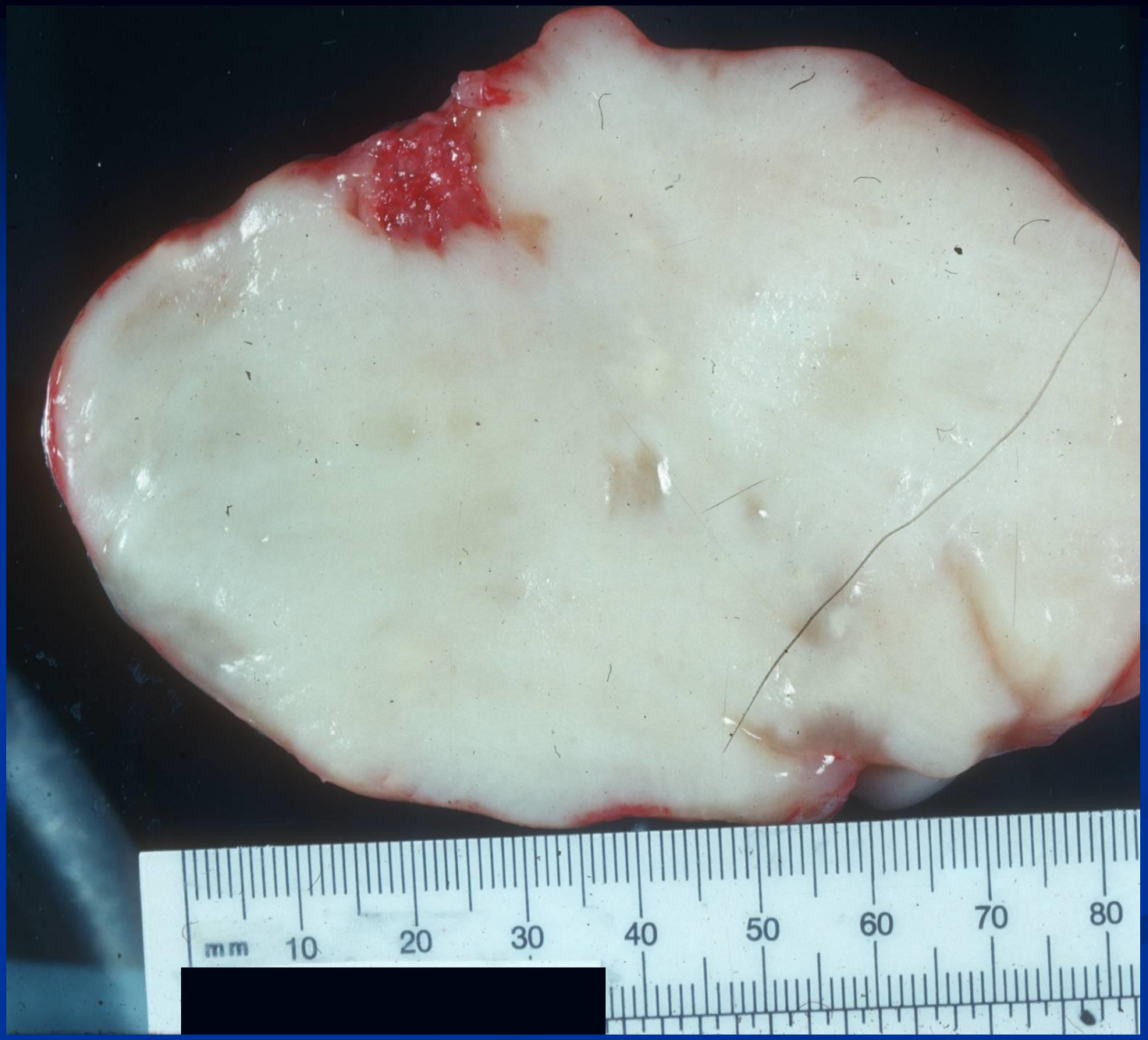
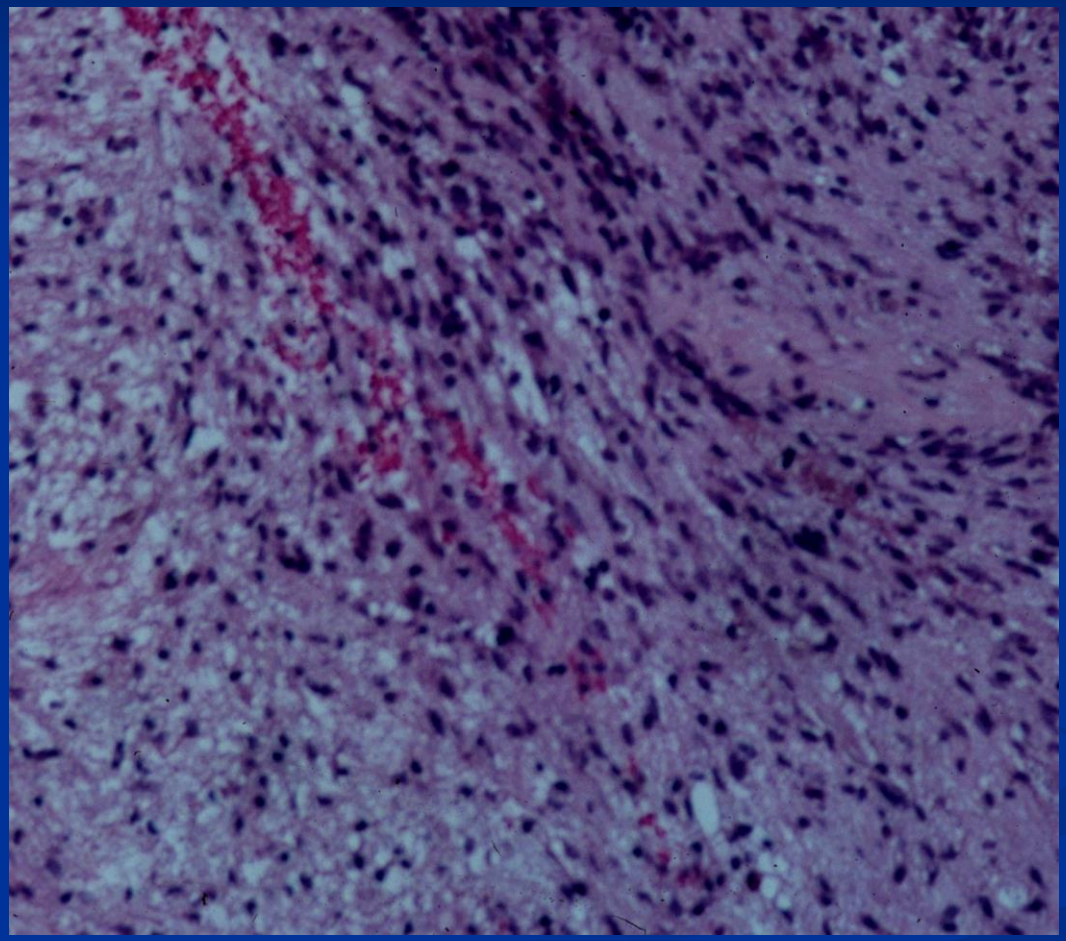
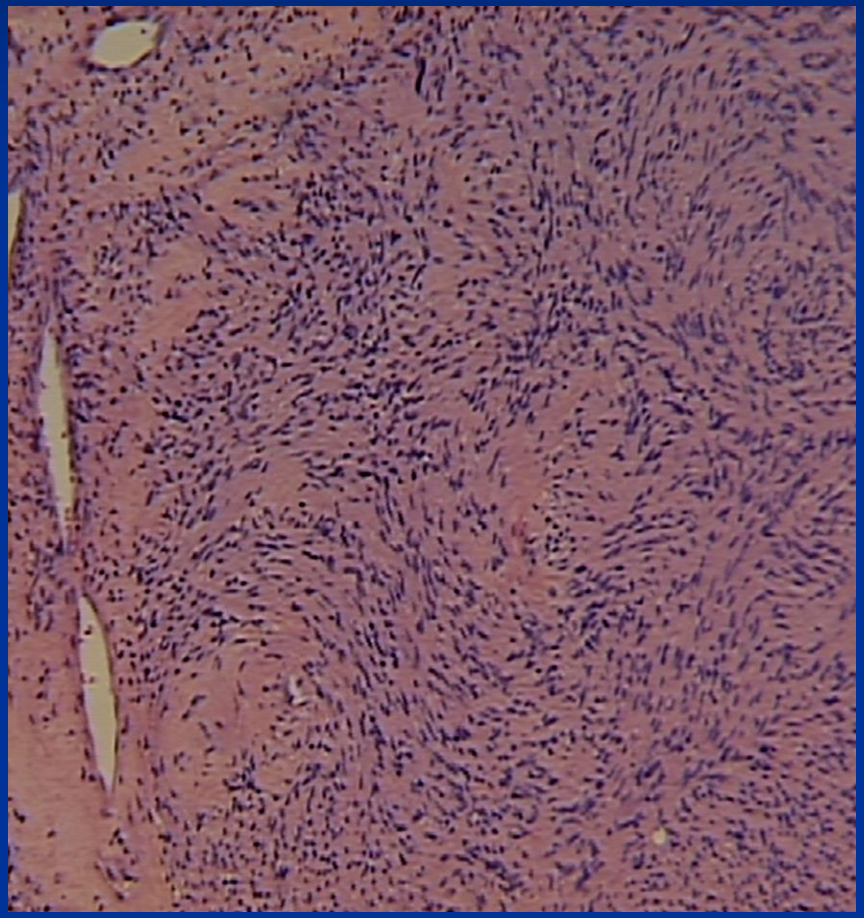


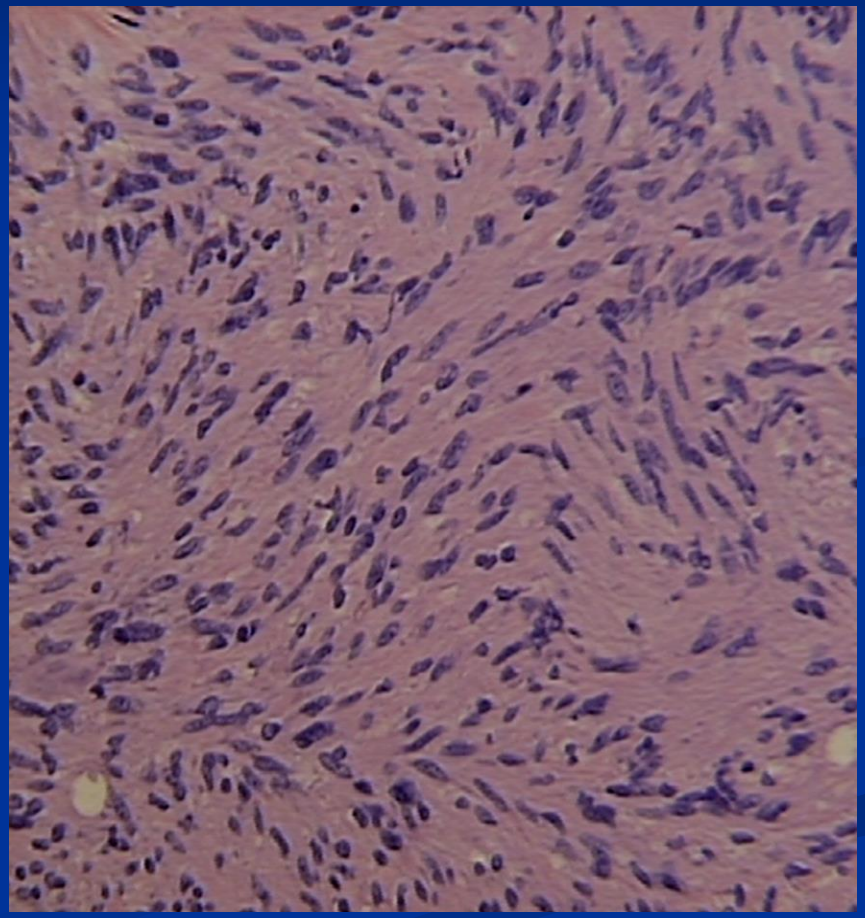
Wittig Orthopedic Oncology

Schwannoma/Neurilemmoma
Antoni A: Cellular Area

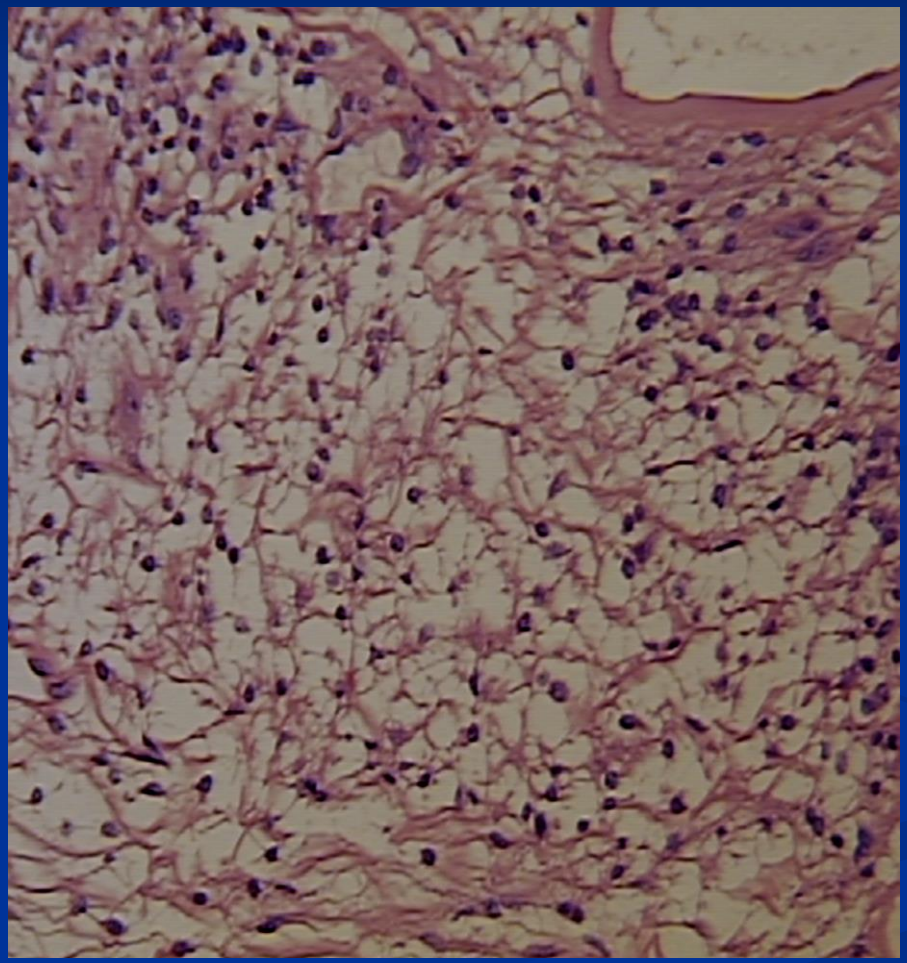


Pathology: Schwannoma
Antoni A: Cellular Area

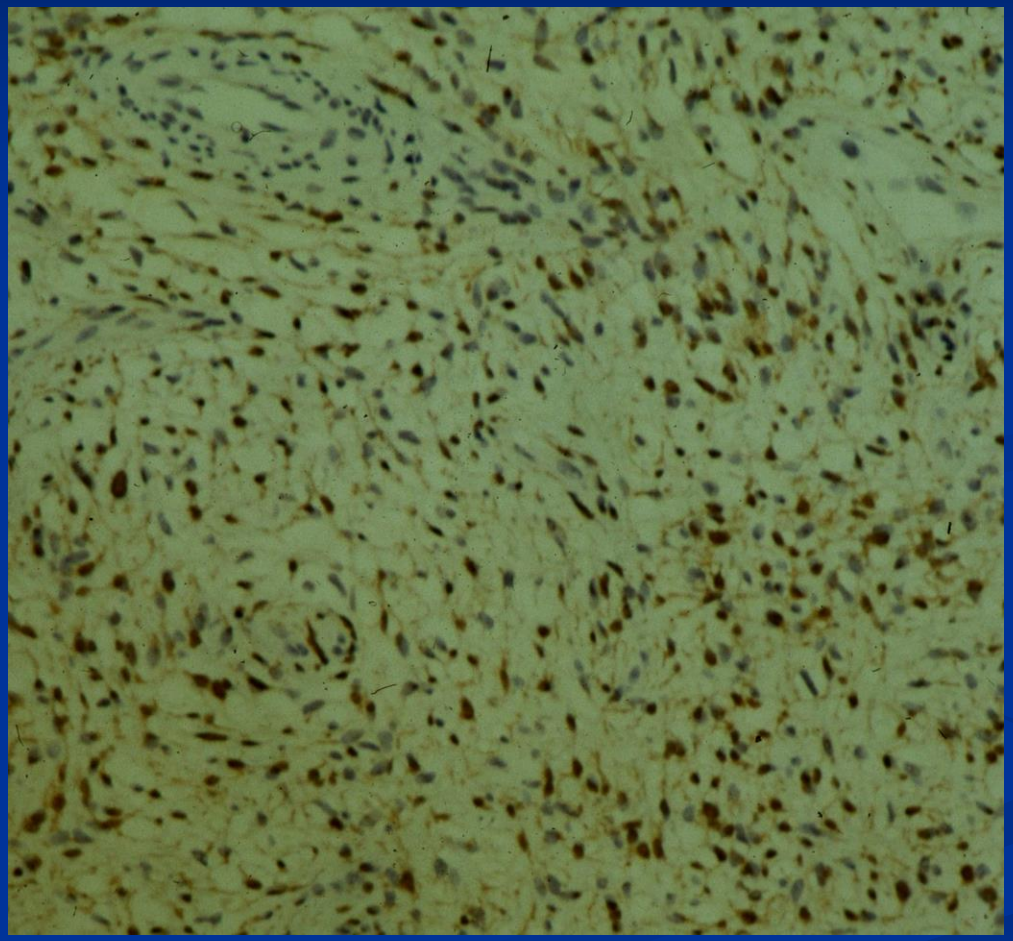




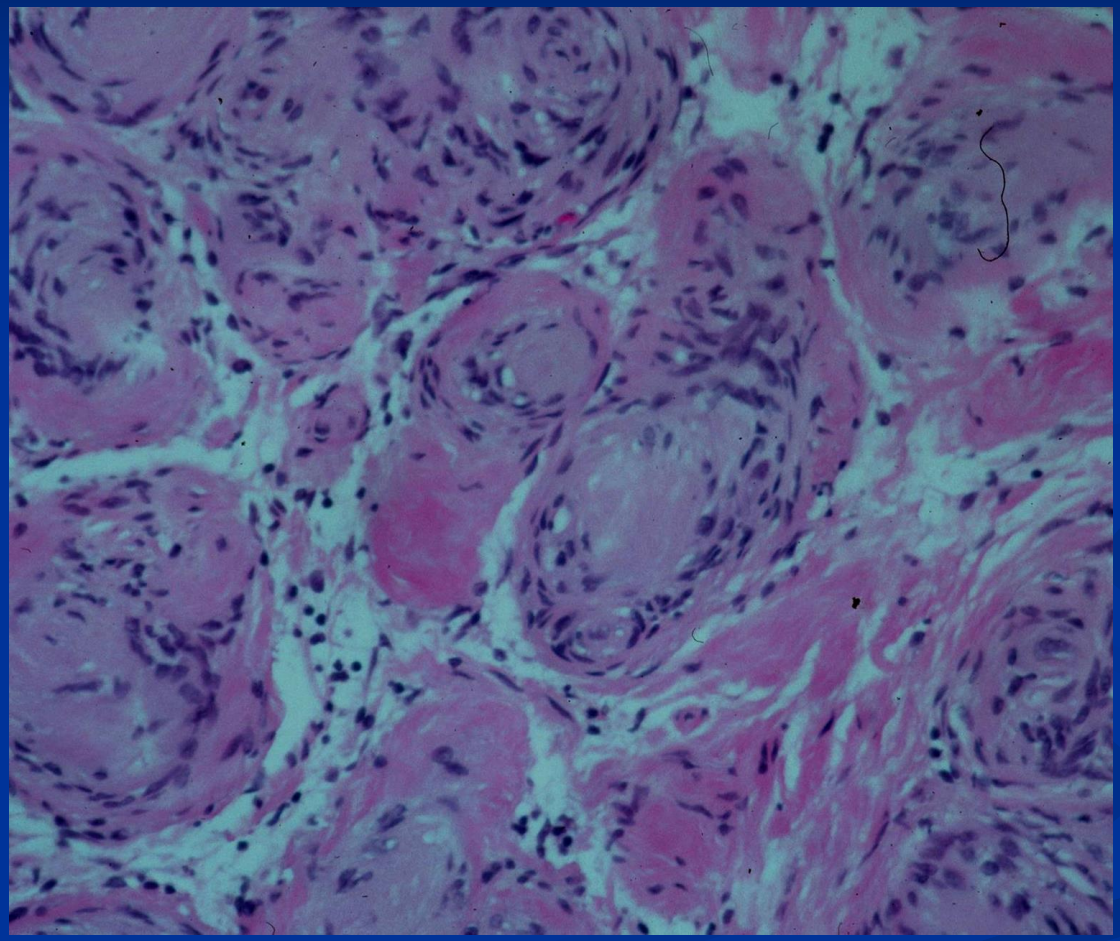
Pathology: Schwannoma
Antoni B: Myxoid Area



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

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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

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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

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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

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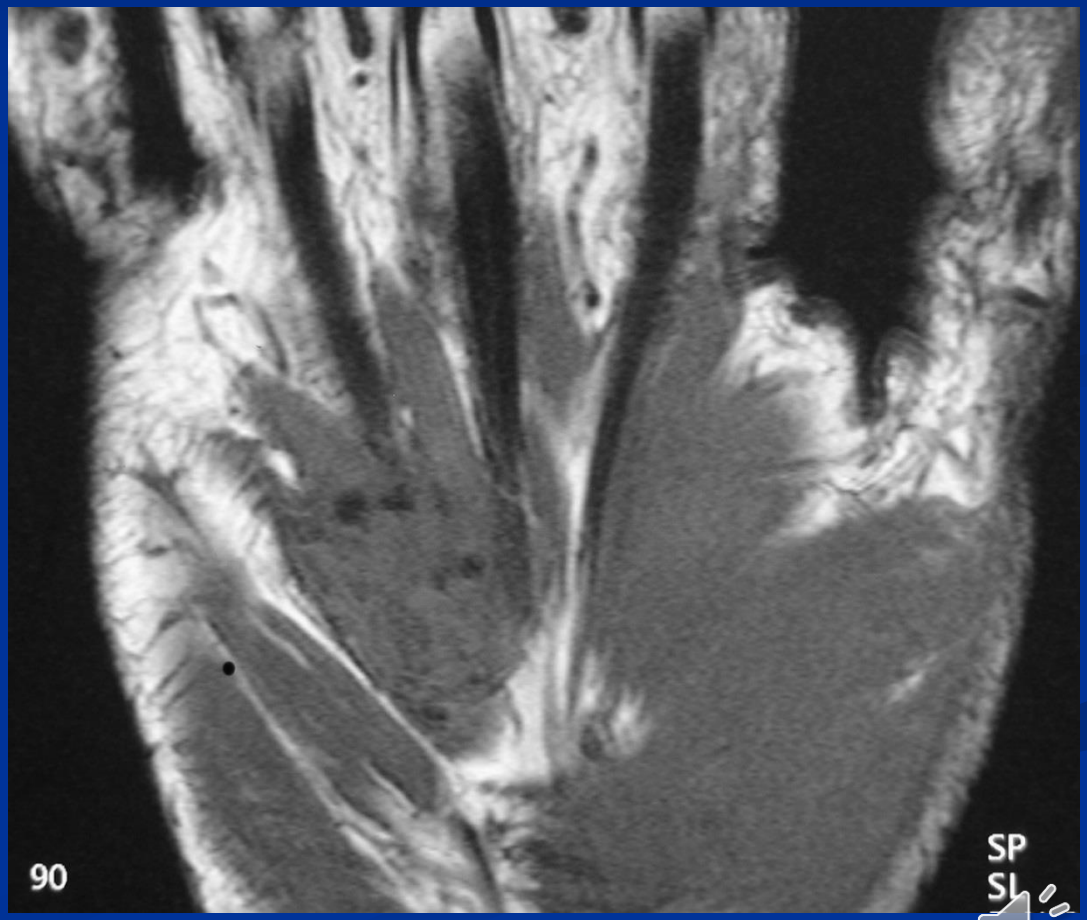
Giant Cell Tumor of Tendon Sheath

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



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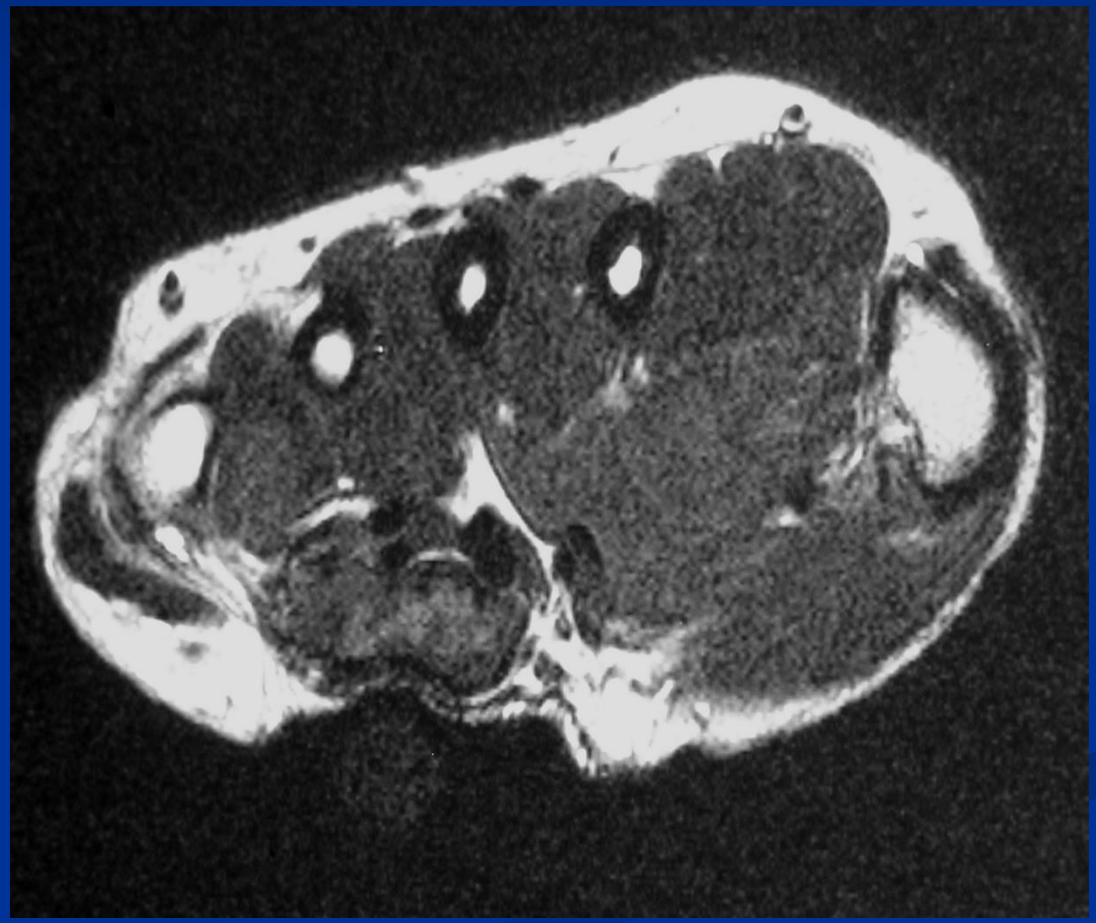
GCTTS

- 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



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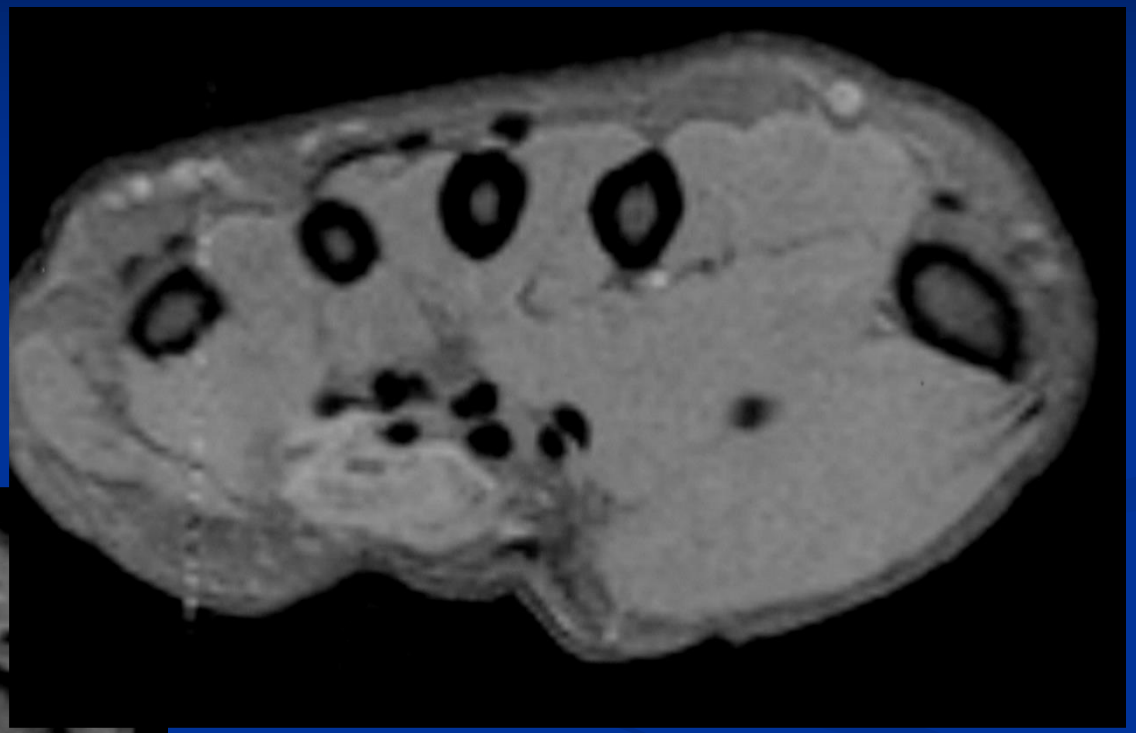
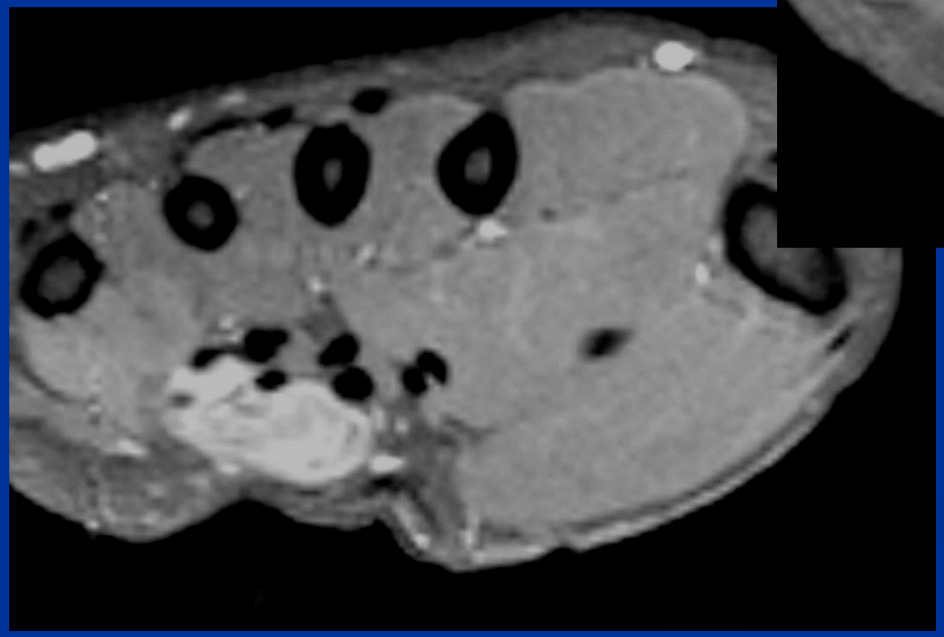
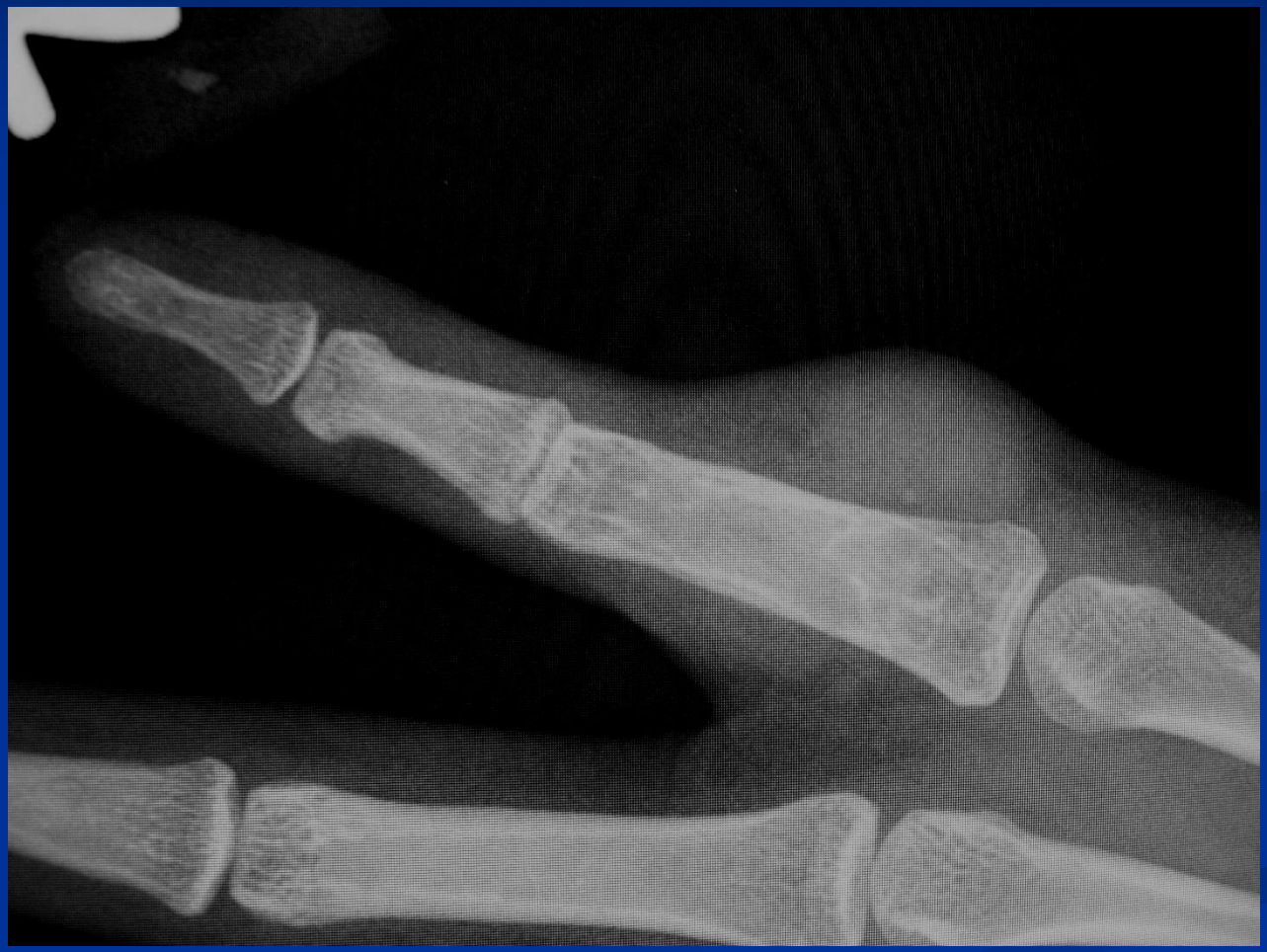
Giant Cell Tumor of Tendon Sheath





GCTTS Hand



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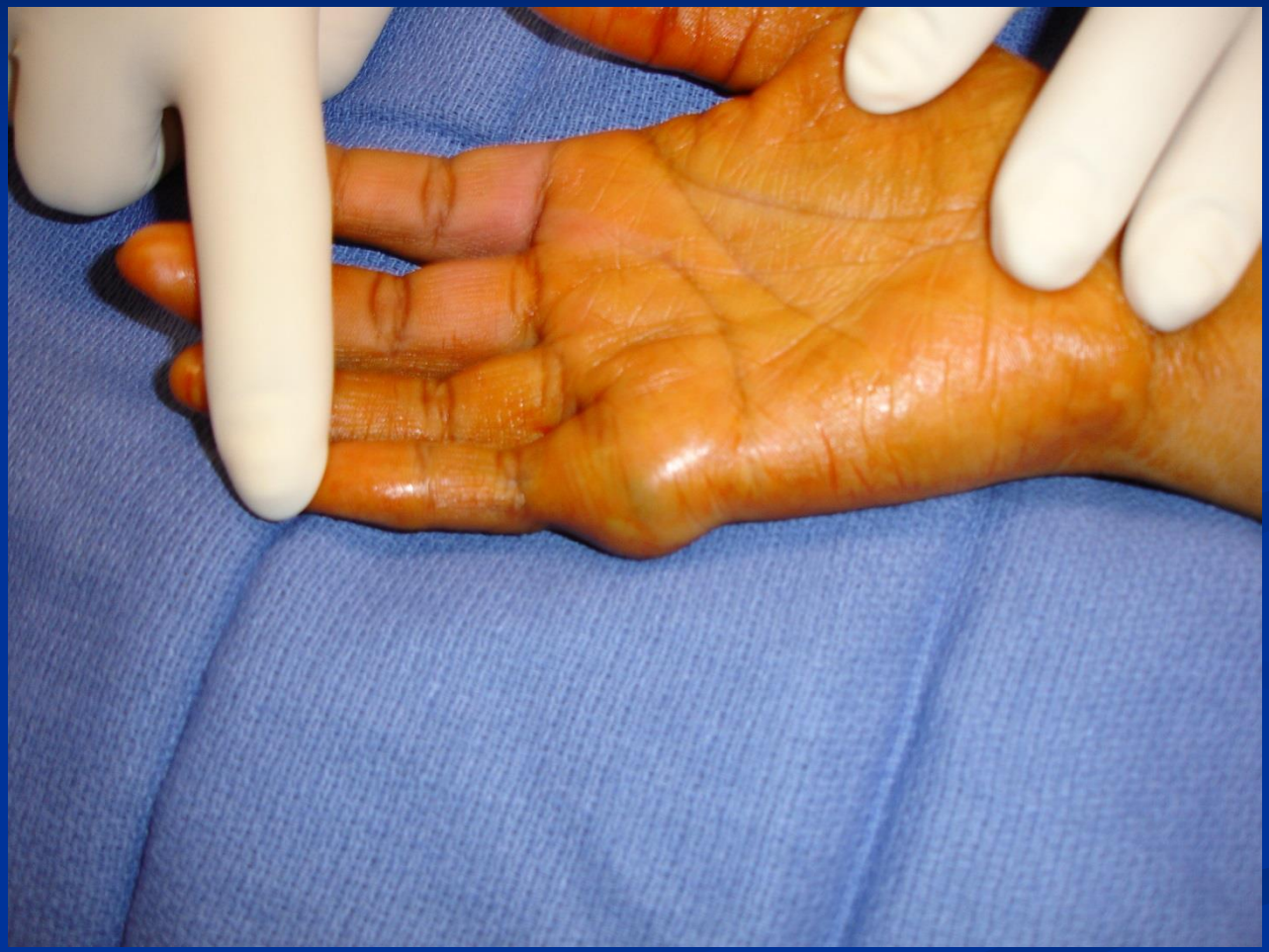
GCTTS of Hand



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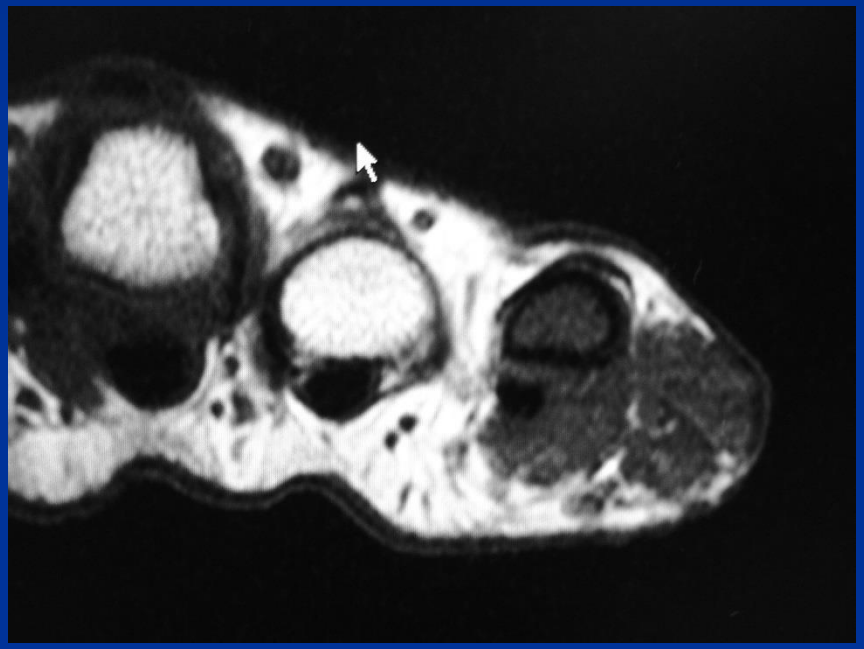
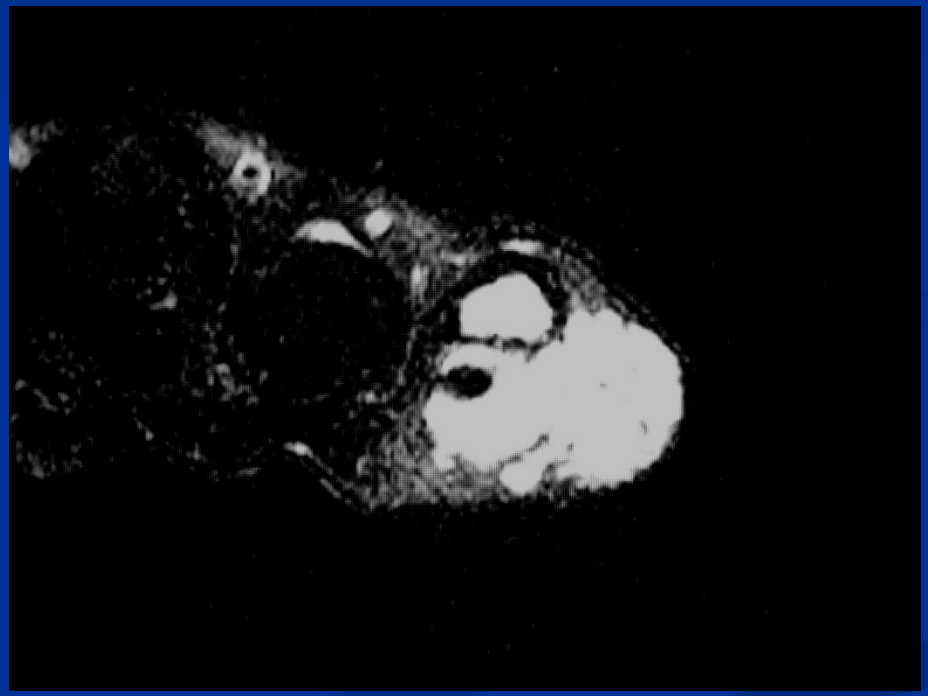
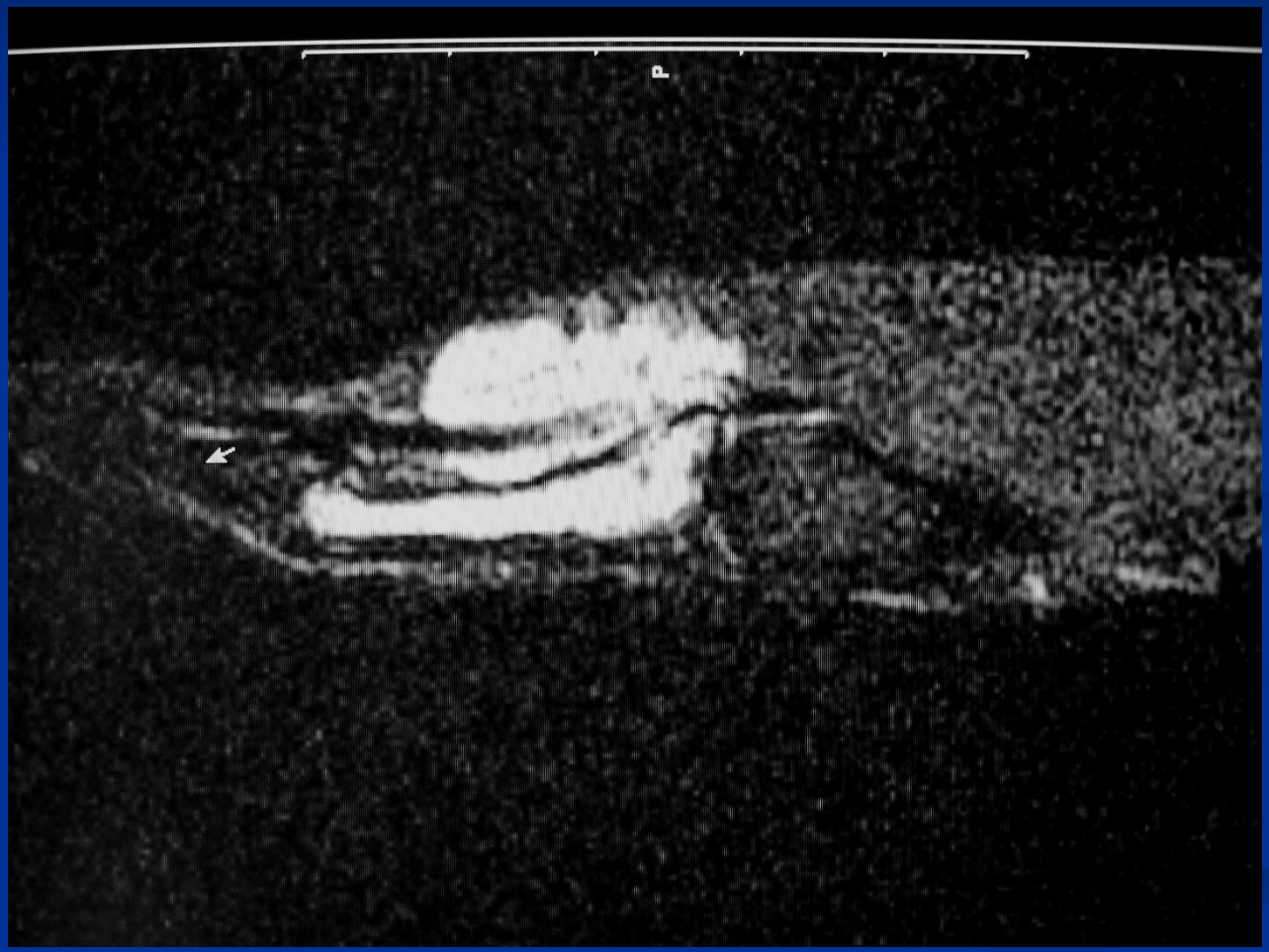
MRI T1: Giant Cell Tumor of Tendon Sheath





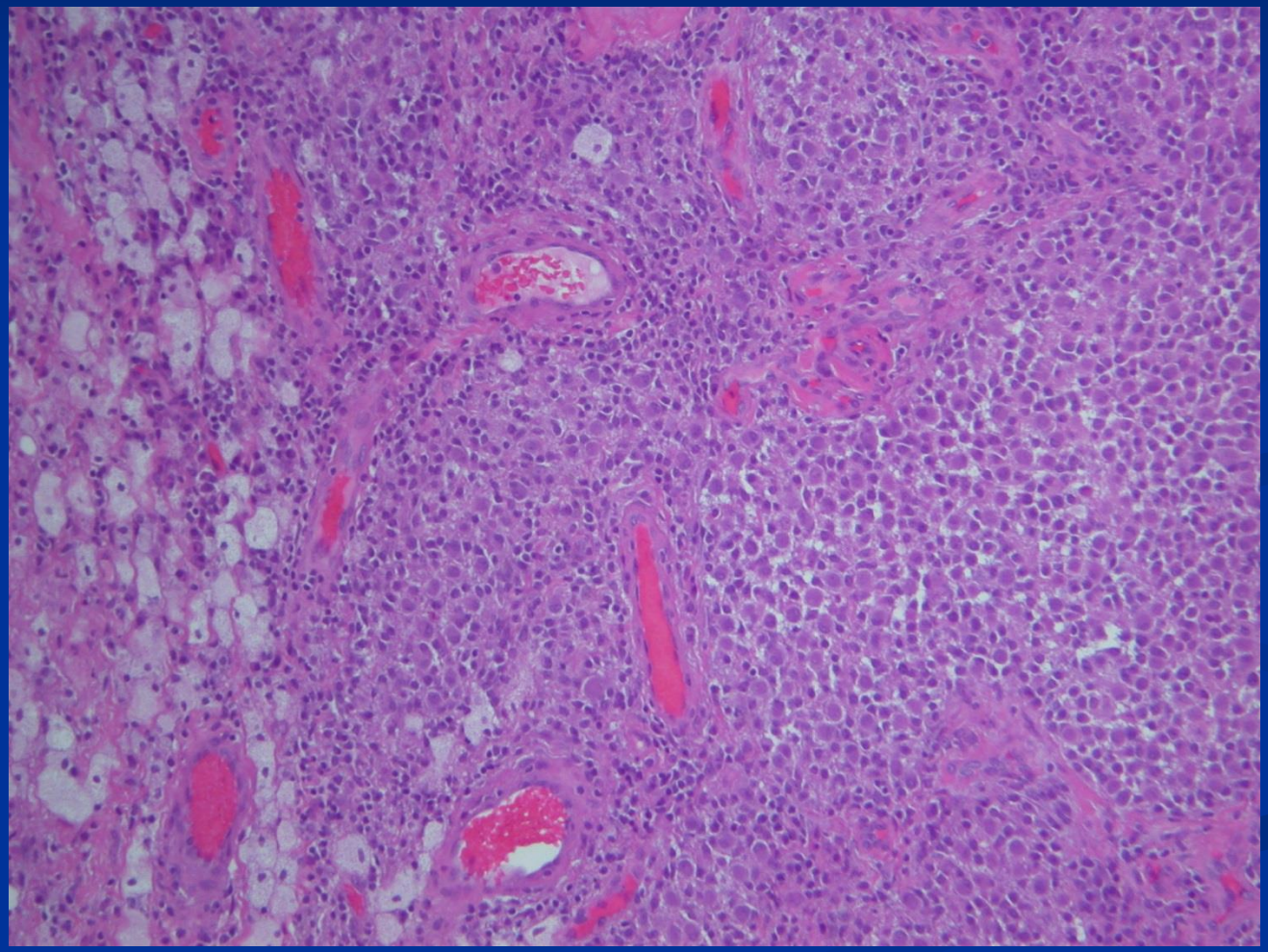
MRI T2: Giant Cell Tumor of Tendon Sheath of Hand



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



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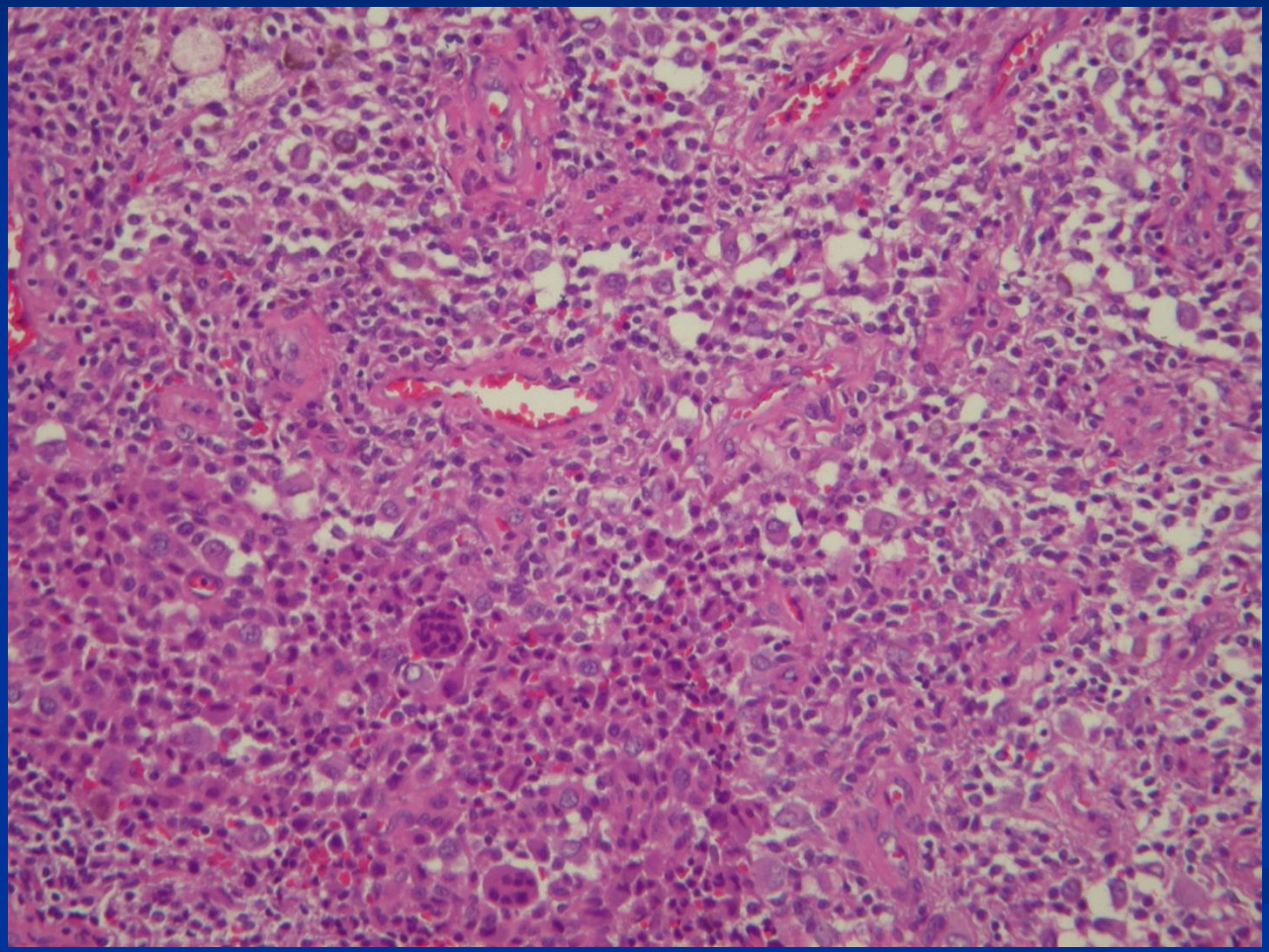
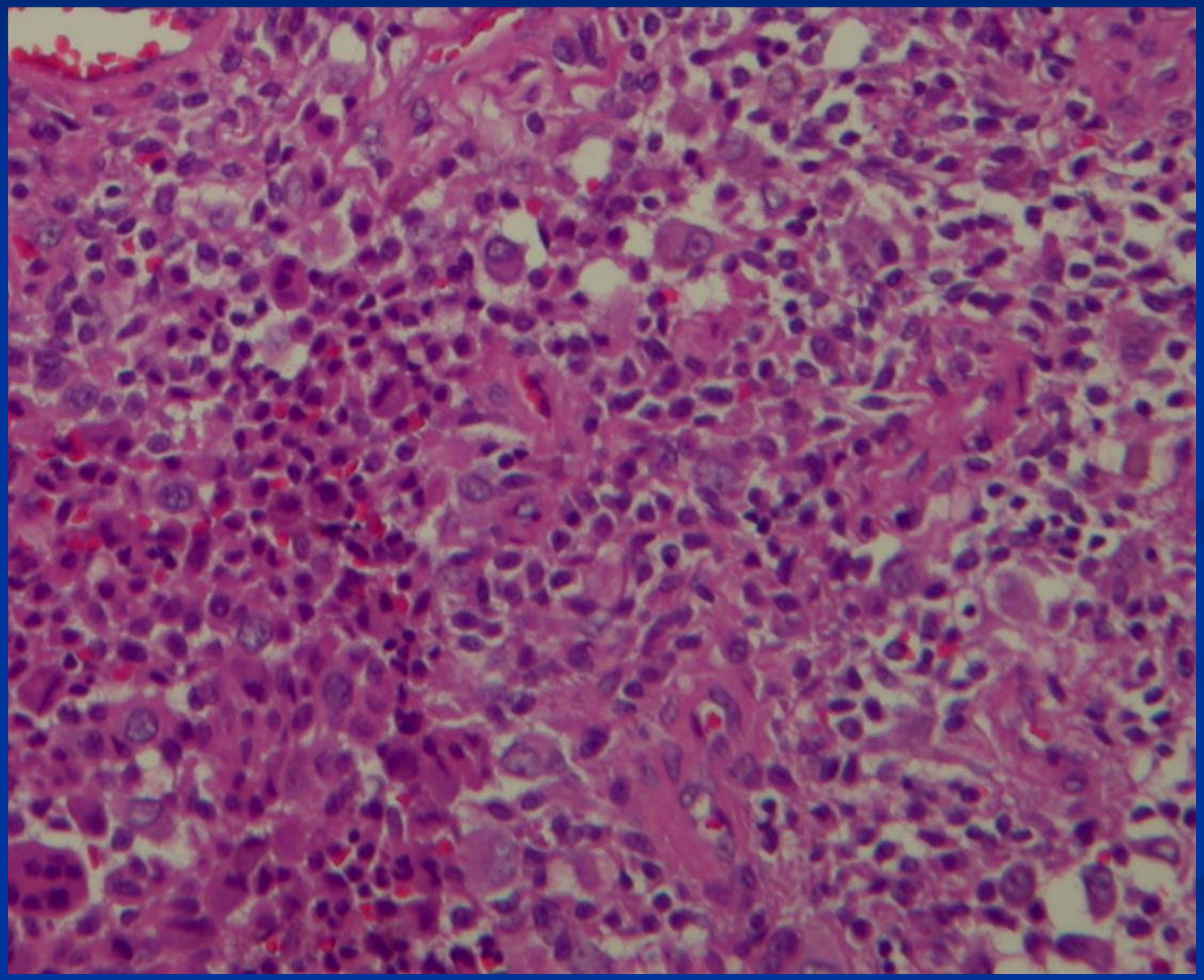
Pathology: Giant Cell Tumor of Tendon Sheath

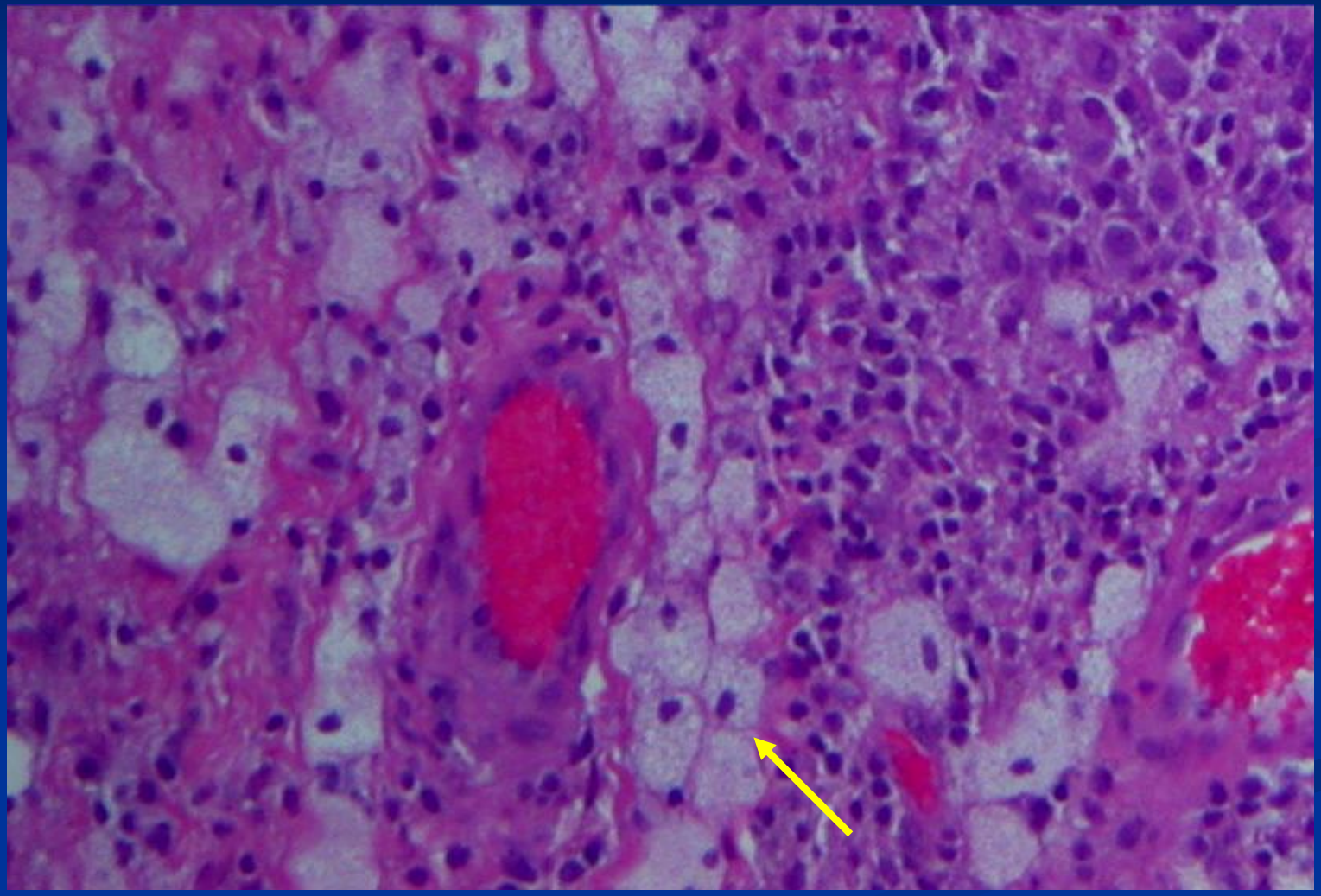


Pathology: Giant Cell Tumor of Tendon Sheath



Pathology: Giant Cell Tumor of Tendon Sheath

Foamy Histiocytes (arrow)



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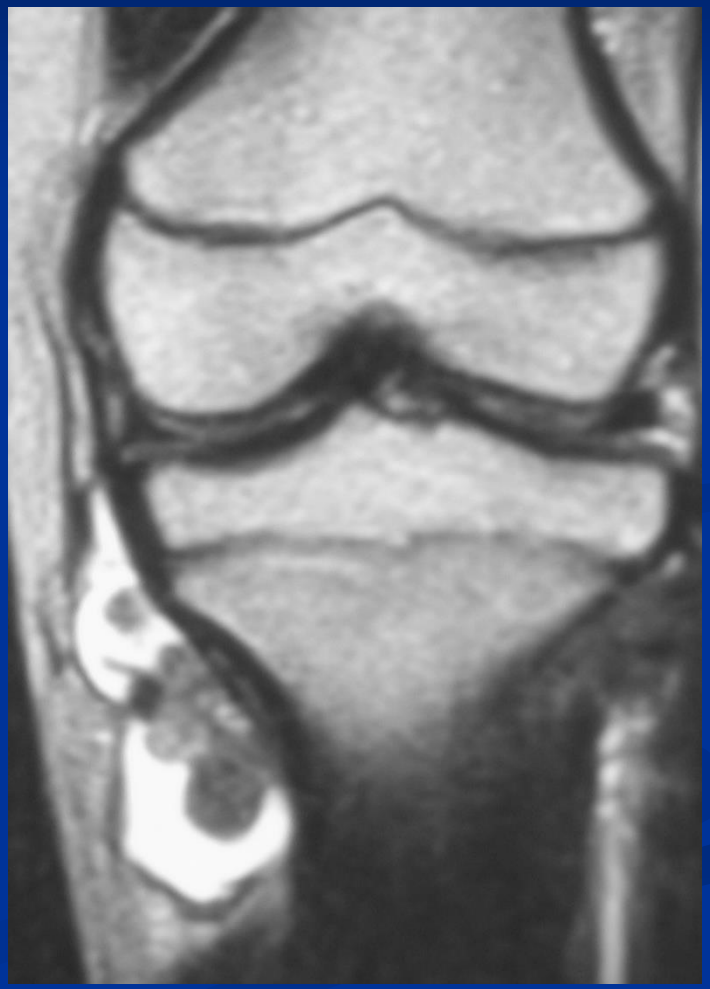
Pigmented Villonodular Synovitis

- 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



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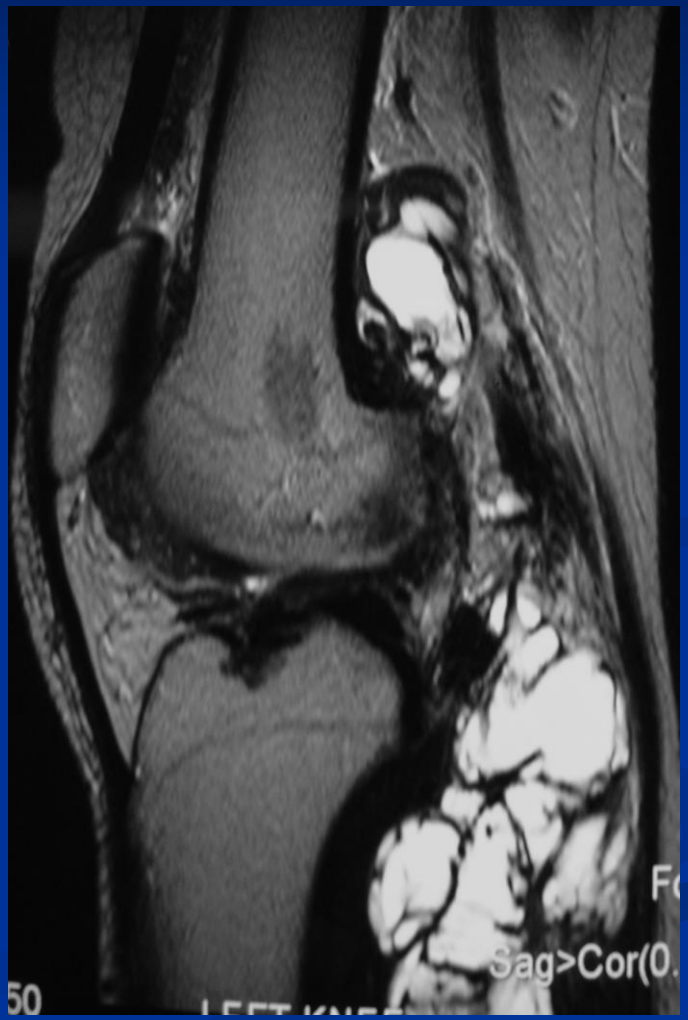
PVNS Imaging

- 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)



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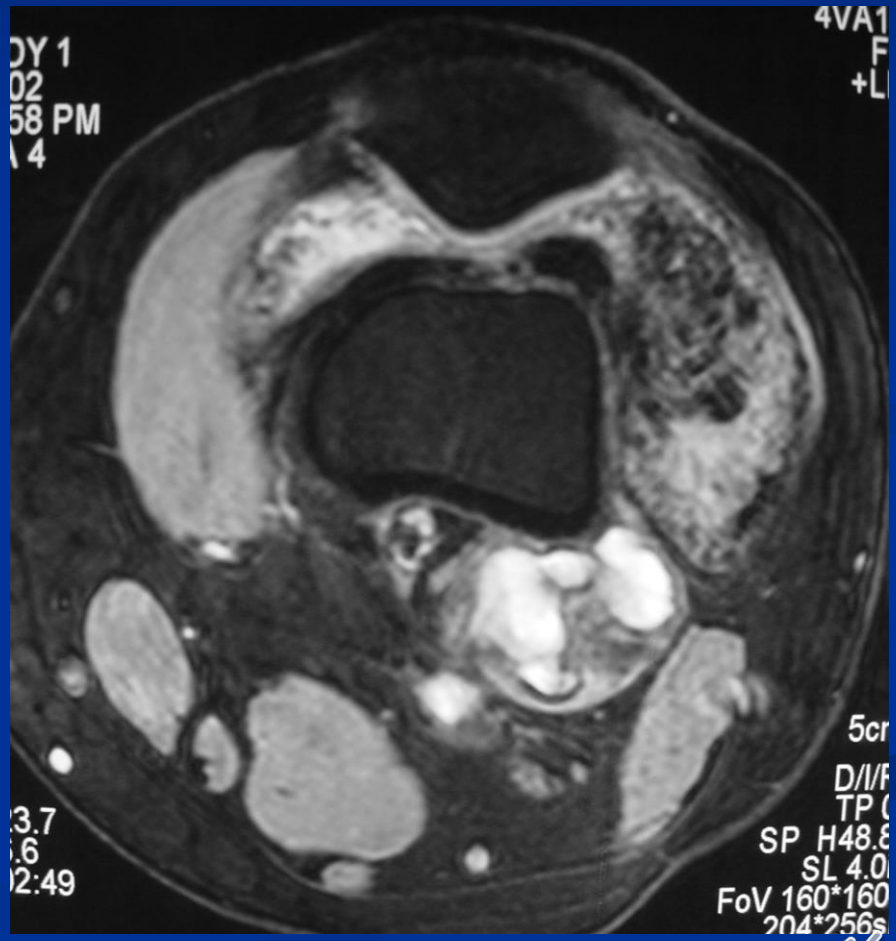
PVNS Imaging

- 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)



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Thank You!