Radiolucent Lesions of Bone (GCT, ABC, UBC, EG, NOF)

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Giant Cell Tumor of Bone (GCT) aka Osteoclastomas

- Definition: Benign aggressive bone tumor composed primarily of "osteoclast-like" giant cells in a mostly vascular background of mononuclear to spindled stromal cells
- The mononuclear cells coalesce to form the giant cells
- Clinical features:
  - ~5% of all biopsied primary bone tumors
  - Symptoms: pain and swelling often relieved by decreased activity
  - Pathologic fracture in 10-35% of patients

Giant Cell Tumor

- Affects Skeletally Mature Patients (90%)
  - **Age: 20-50 yrs old**
  - **Rare in children 1-2%**
- Approximately equal sex distribution

Giant Cell Tumor (GCT)

- **Location**
  - Metaphysis and usually grow to the subchondral bone in the epiphysis
    - **Distal Femur or Proximal Tibia—most common**
    - **Distal Radius (3rd Most Common Site: 10%)**
    - **Sacrum 7%**
    - **Humerus 6%**
    - **Pelvis 4%**
    - **Hands/Feet 5%**
    - **Mutifocal 0.5-1%**

Giant Cell Tumor (GCT)

- **Staging: (Arabic Numerals)**
  - Stage 1: Slowly Growing
    - Tumor is entirely intraosseous
    - Thin sclerotic rim around the tumor
  - Stage 2: Active
    - Tumor is entirely intraosseous but growing more rapidly
    - No sclerotic rim
    - Bone may be expanded or scallopped
  - Stage 3: Aggressive
    - Tumor has destroyed the cortex
    - Formed a soft tissue mass

Giant Cell Tumor (GCT)

- **ALWAYS CHECK FOR HYPERPARATHYROIDISM especially if the GCT is occurring in an unusual location**
- **Brown tumors of hyperparathyroidism can look similar histologically as a GCT**

Giant Cell Tumor (GCT)

- **Radiographic Findings:**
  - Solitary eccentric geographic lytic lesion arising from metaphysis and extending into epiphysis usually to the subchondral plate
  - Usually no margin or a faint margin of sclerosis surrounding the tumor
  - No matrix mineralization

Giant Cell Tumor (GCT)

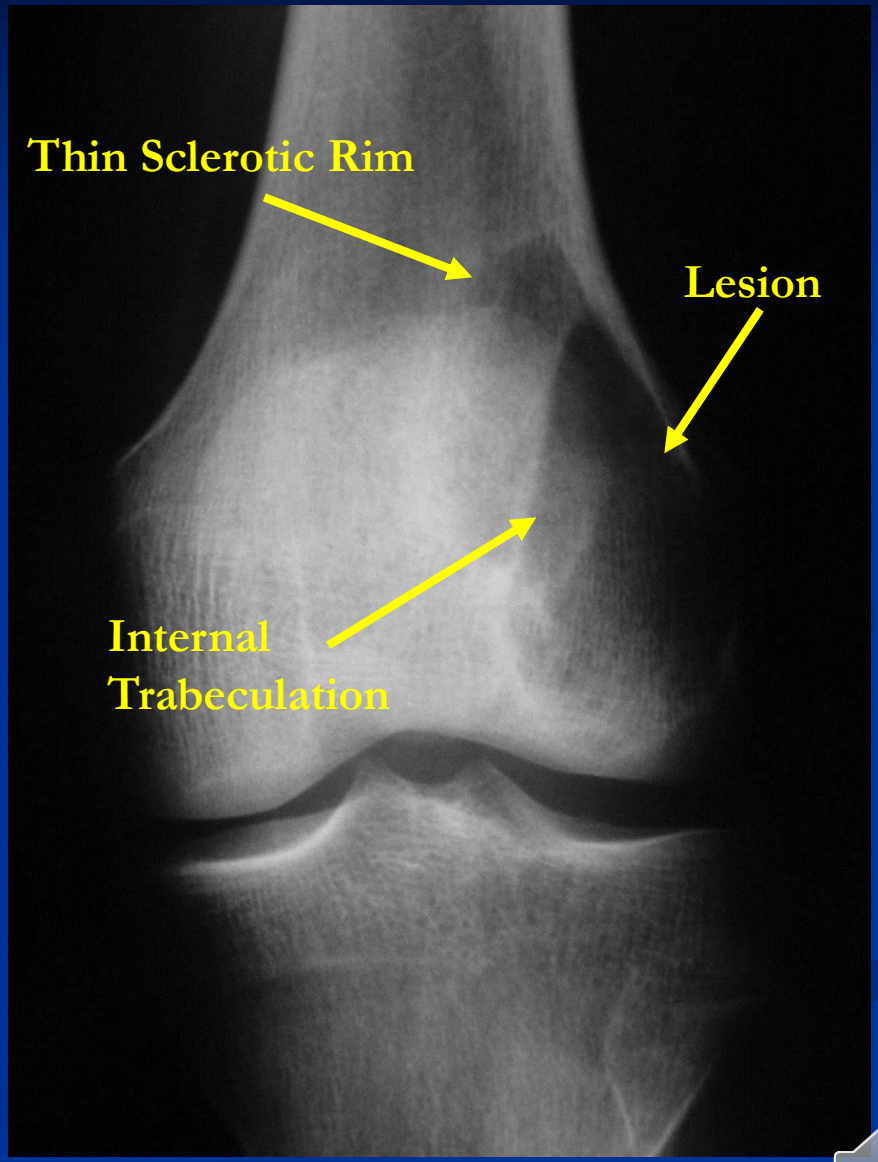
- **Radiographic Findings:**
  - Expansile lesion
  - Lesions that extend through cortex are usually encased by a delicate shell of **periosteal new bone** often only detectable on a CT scan
  - Internal Trabeculations may be present
    - **Reactive, Thickened Residual Trabeculae of Bone**
  - **Unusual periosteal reactions—rarely occur**

Giant Cell Tumor (GCT)

- **Radiographic Findings:**
  - Bone scan – hot on bone scan
  - MRI > CT for evaluation of bone and soft tissue extent
    - T1: Intermediate Signal similar to muscle
    - T2: Heterogeneous: Low to intermediate signal intensity usually predominates mixed with high signal areas
    - Fluid-Fluid Levels: **Secondary ABC changes**
  - CT:
    - Absence of mineralization
    - Internal trabeculations
    - Subtle periosteal reactions around soft tissue component

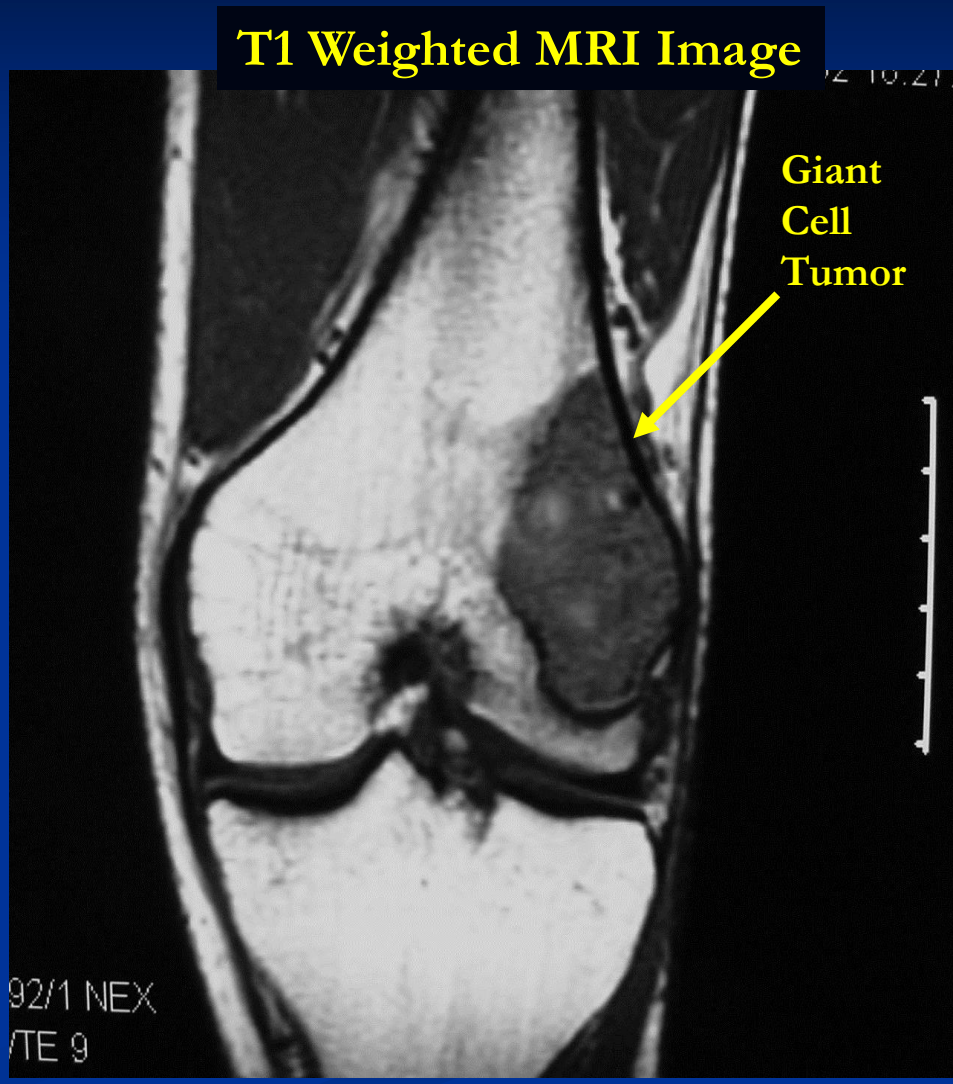
Giant Cell Tumor (GCT) of Distal Femur

- Eccentric Lytic Lesion
- Metaphyseal extending into Epiphysis
- Geographic (Well Circumscribed)
- Thin Rim of Surrounding Sclerosis
- No Mineralization
- Internal Trabeculations



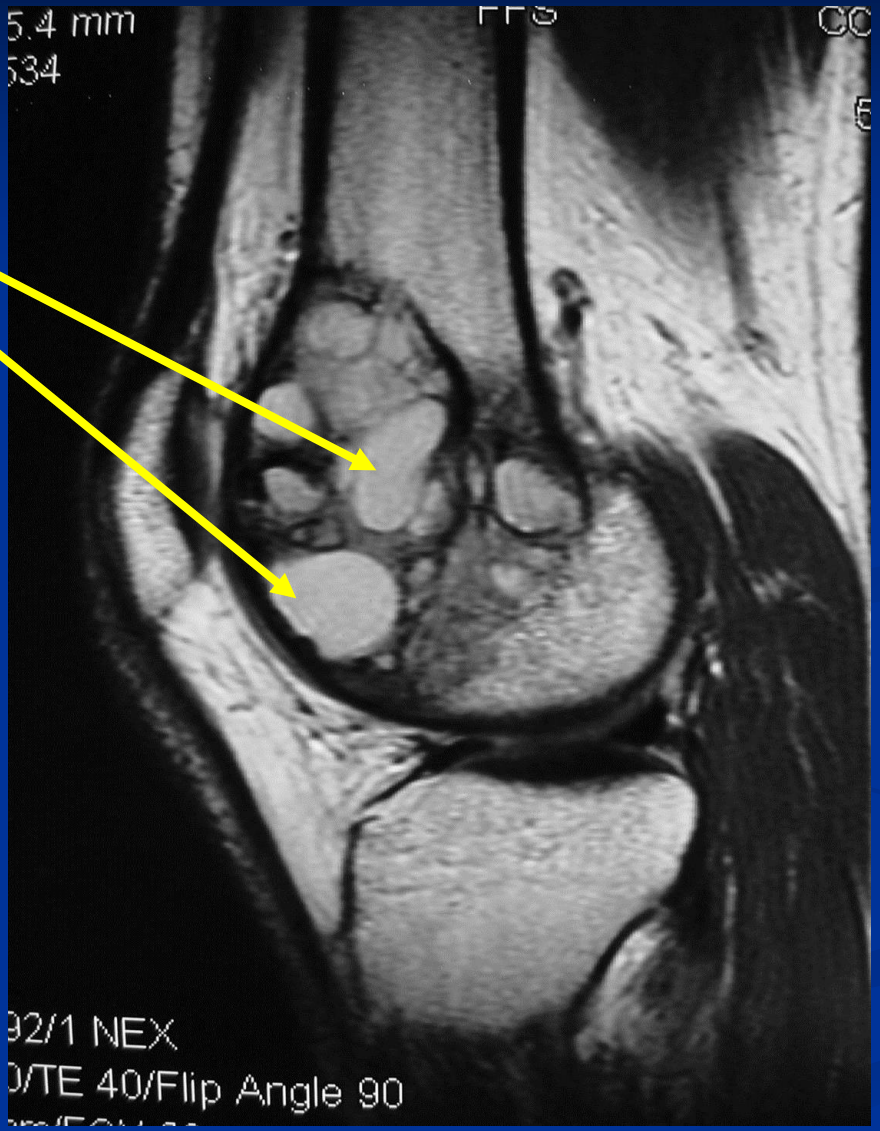
MRI Confirms Geographic Eccentric Lesion

- MRI: usually intermediate SI on T1
- High signal areas on T1 represent areas of hemorrhage
- MRI is most useful for demonstrating true extent of lesion

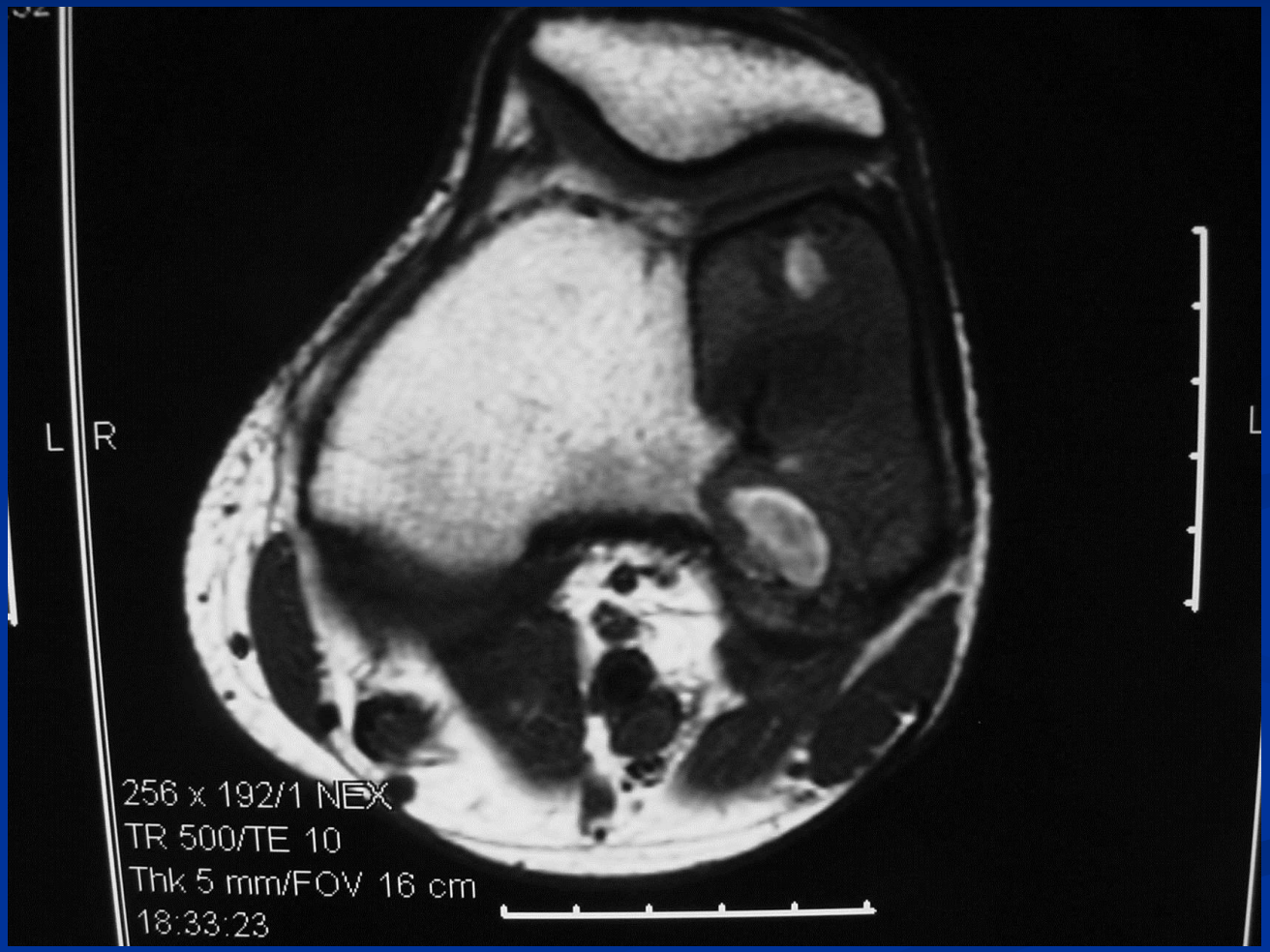


Sagittal Proton Density MRI

Cystic Areas

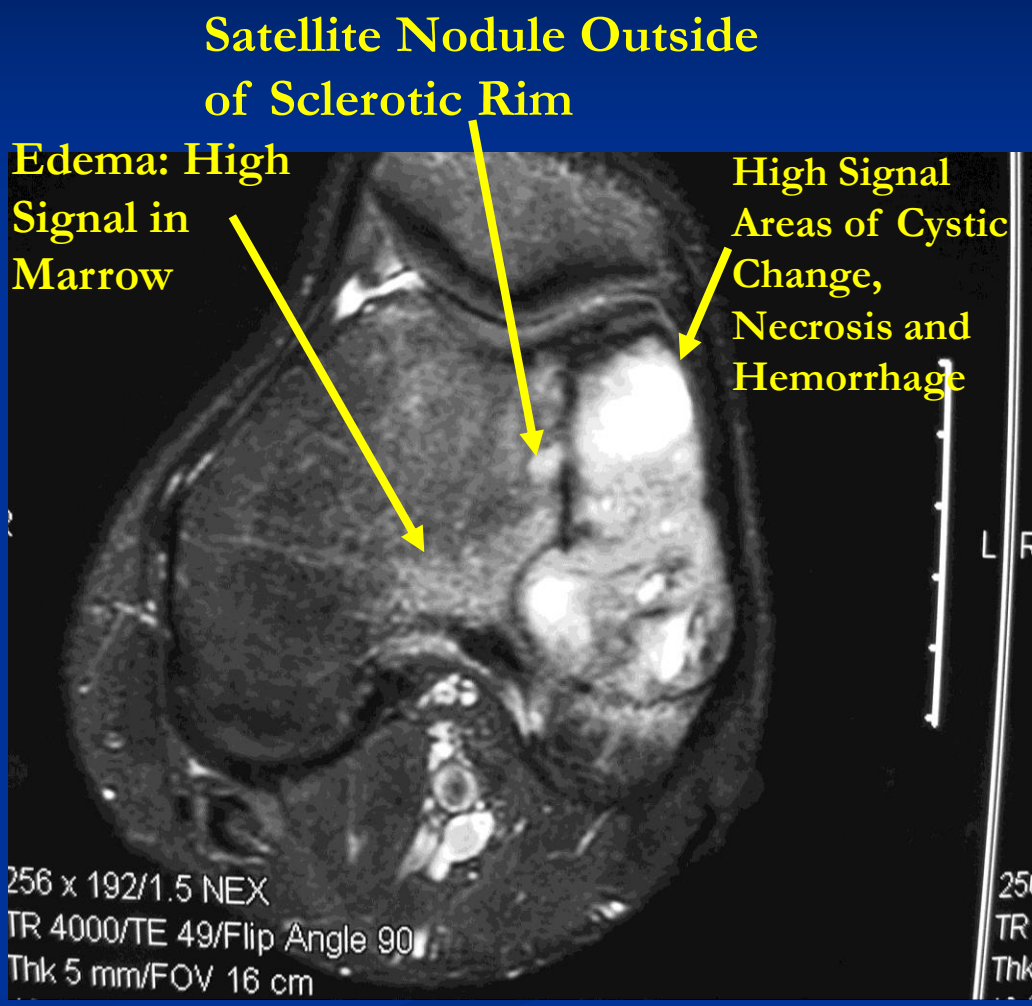


Axial T1 Weighted MRI: Stage 2 Giant Cell Tumor; No Extraosseous Extension



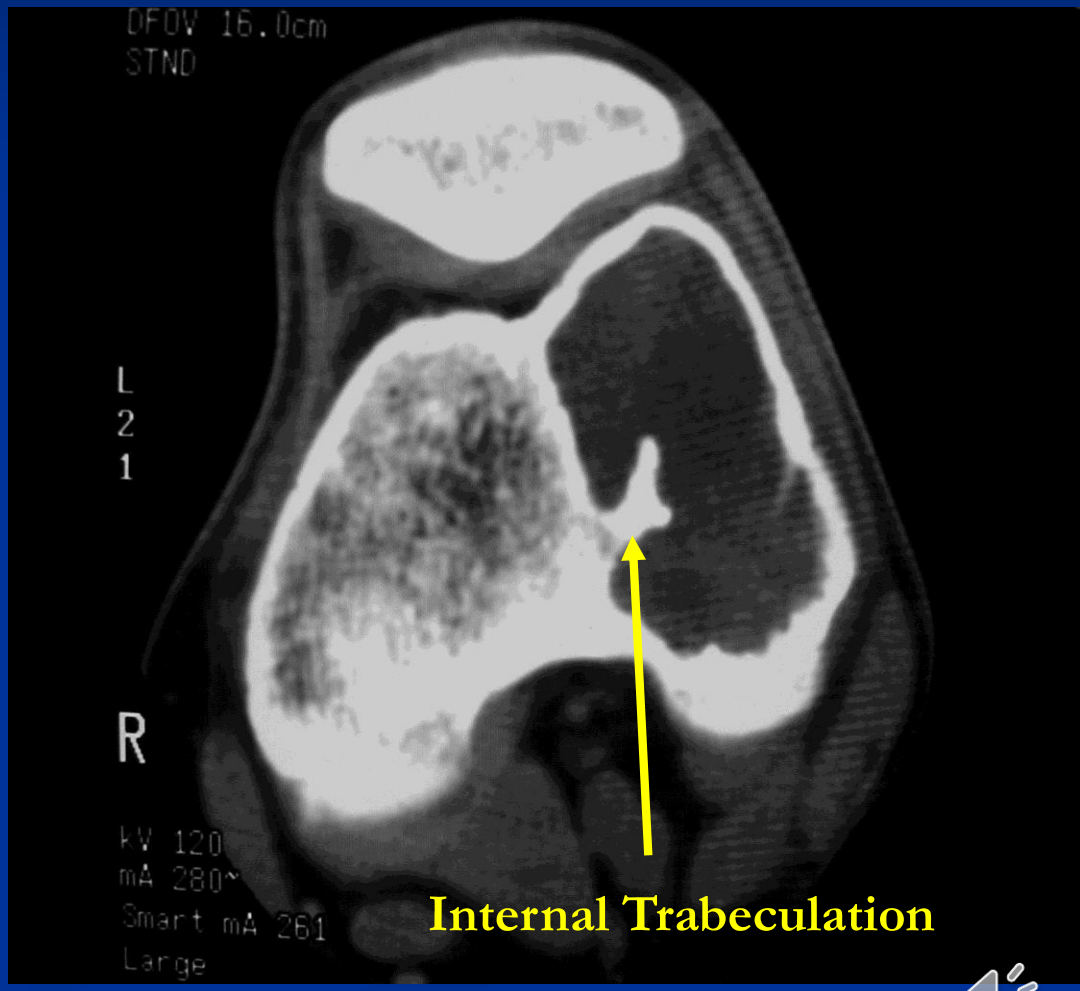
MRI: T2 Weighted Fat Suppressed Axial Image of a Giant Cell Tumor of the Distal Femur

- T2: heterogeneous lesion
- Predominantly low signal mixed with high signal cystic areas
- High signal areas:cystic areas of hemorrhage, necrosis and edema
- Edema (high signal in marrow) surrounding the lesion



CT Scan of Giant Cell Tumor (GCT) of Distal Femur (Axial Image)

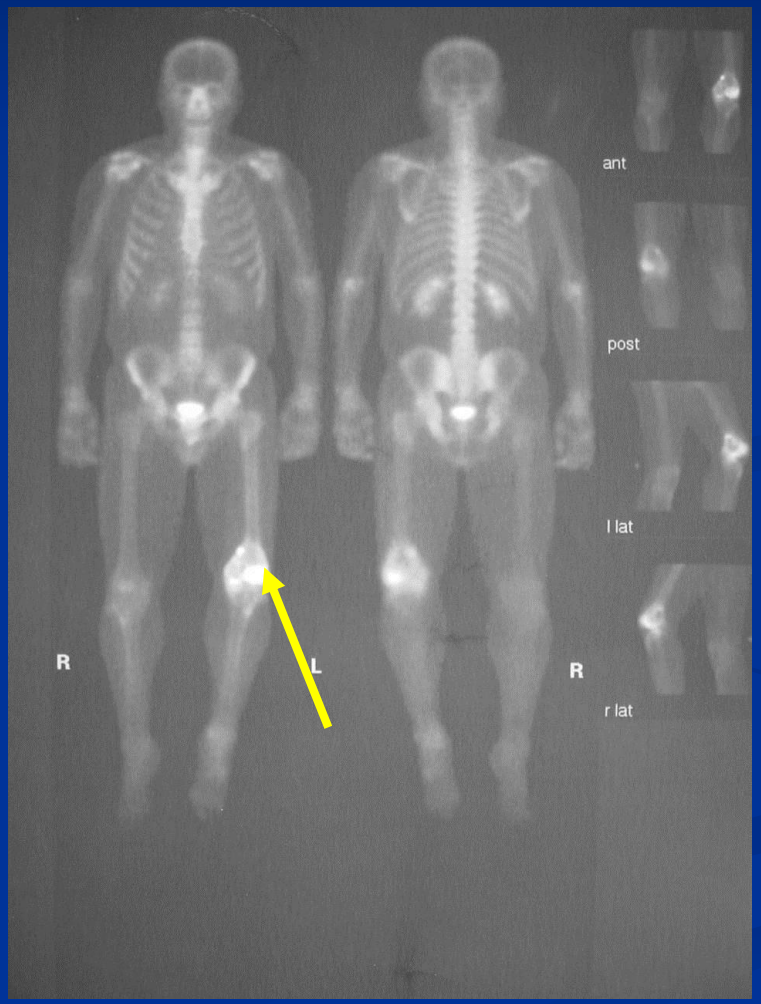
- Eccentric Lesion
- Geographic
- Well Circumscribed
- Cortex is Intact
- No Subtle Mineralization
- Internal Trabeculation Identified



Internal Trabeculations

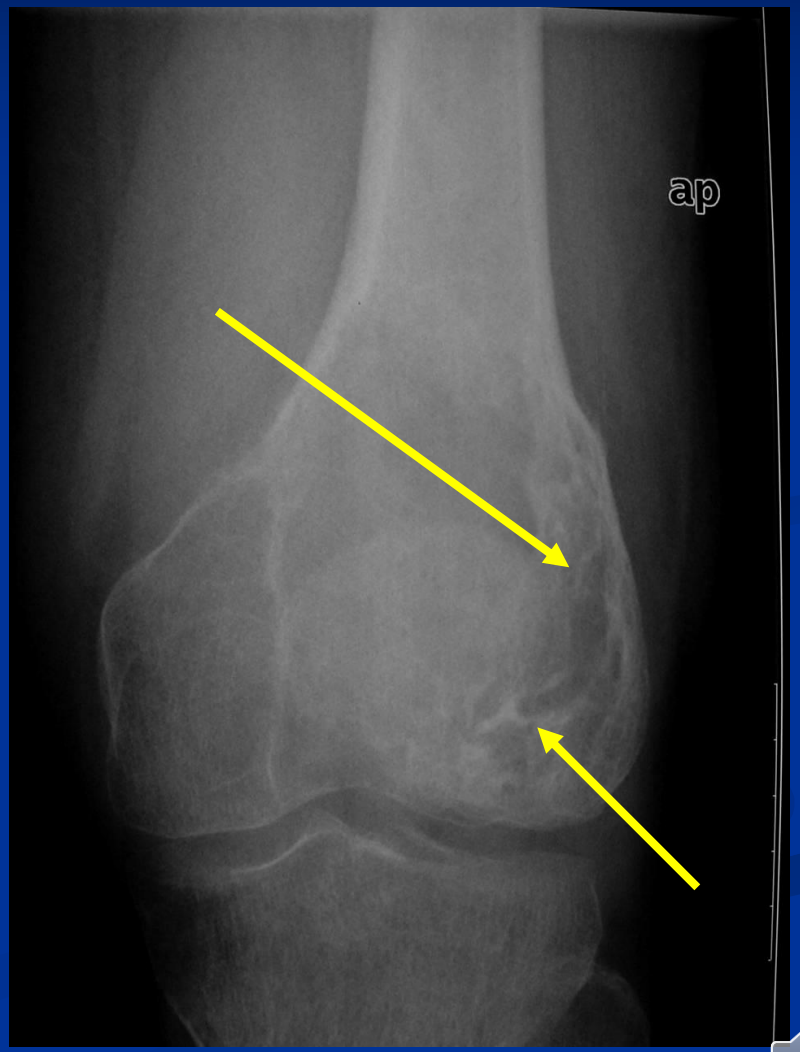
- Thickened Reactive Trabeculae of Bone within a Lesion
- Differential Diagnosis: (DCHANG)
  - Desmoplastic Fibroma
  - Chondromyxofibroma
  - Hemangioma
  - Aneurysmal Bone Cyst
  - Nonossifying Fibroma
  - Giant Cell Tumor
  - UBC—Not Really IT but looks like it

Bone Scan Demostrates Increased Uptake in Area of Giant Cell Tumor of Distal Femur

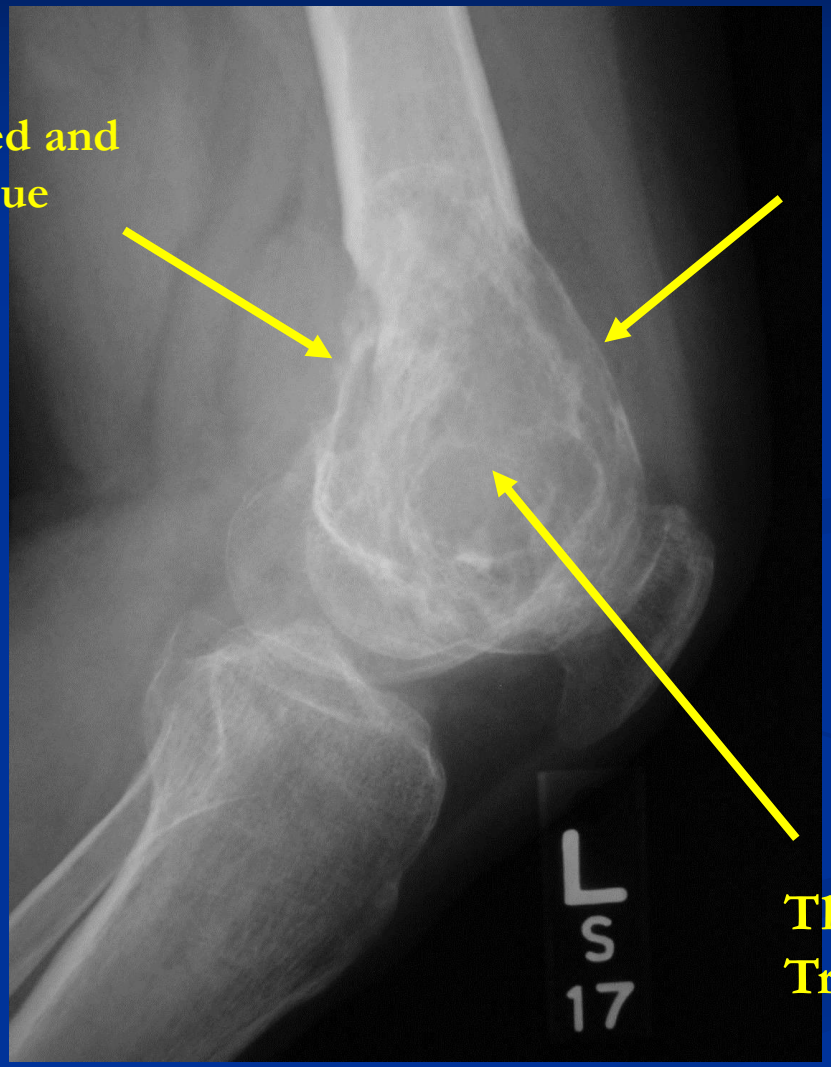


X-Ray: Giant Cell Tumor of Distal Femur

- Aggressive Stage 3 Giant Cell Tumor of Distal Femur
- Many Internal Trabeculations (arrows)



X-Ray: Giant Cell Tumor of Distal Femur (Stage 3)



Cortex Destroyed and Soft Tissue Mass

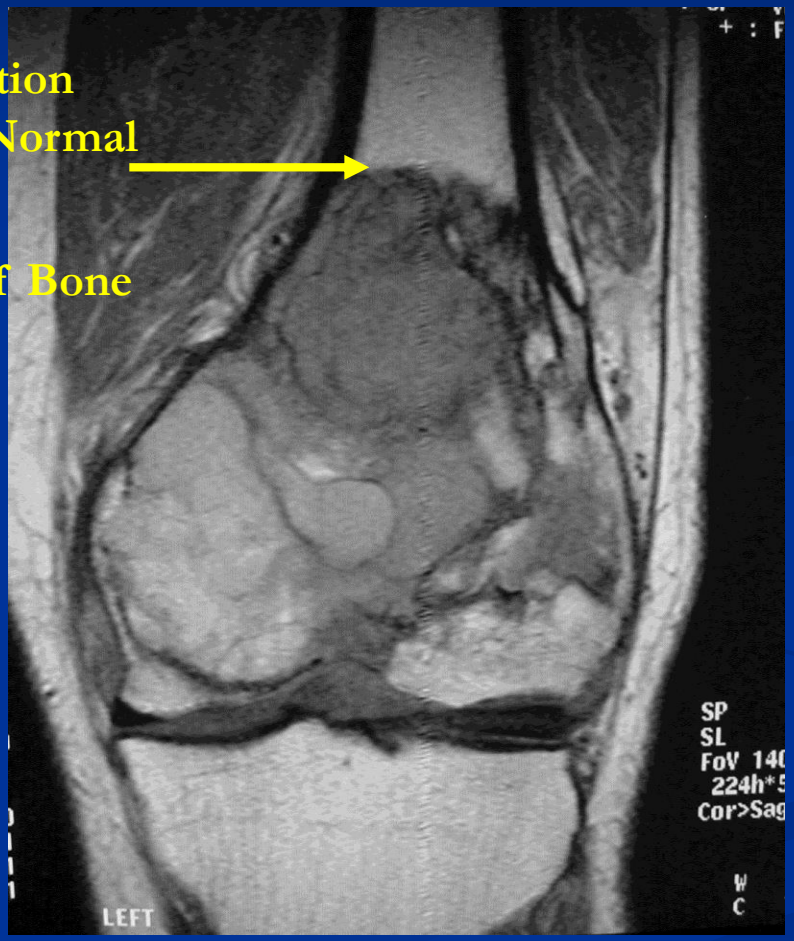
Cortex Destroyed Anteriorly; Soft Tissue Component Encased by Egg Shell Rim of Periosteum

Thick Internal Trabeculations

MRI of Giant Cell Tumor of Distal Femur Demonstrating Cystic Changes and Confirms Geographic Pattern of Bone Destruction

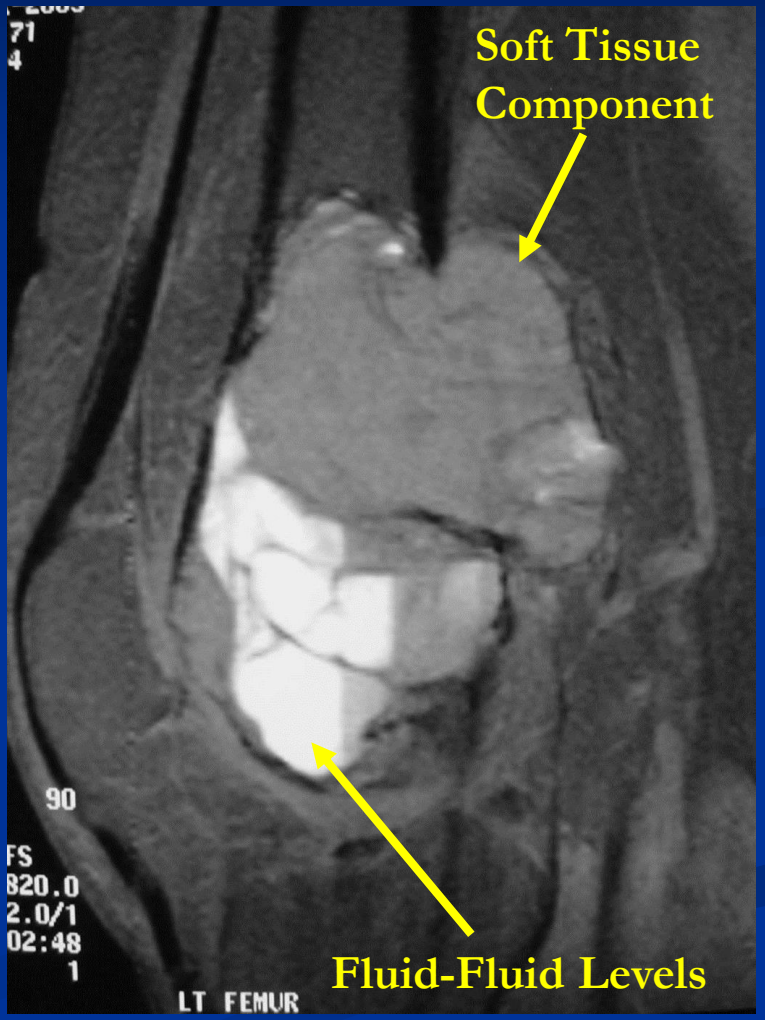
Sharp Zone of Transition between Tumor and Normal Bone

Geographic Pattern of Bone Destruction

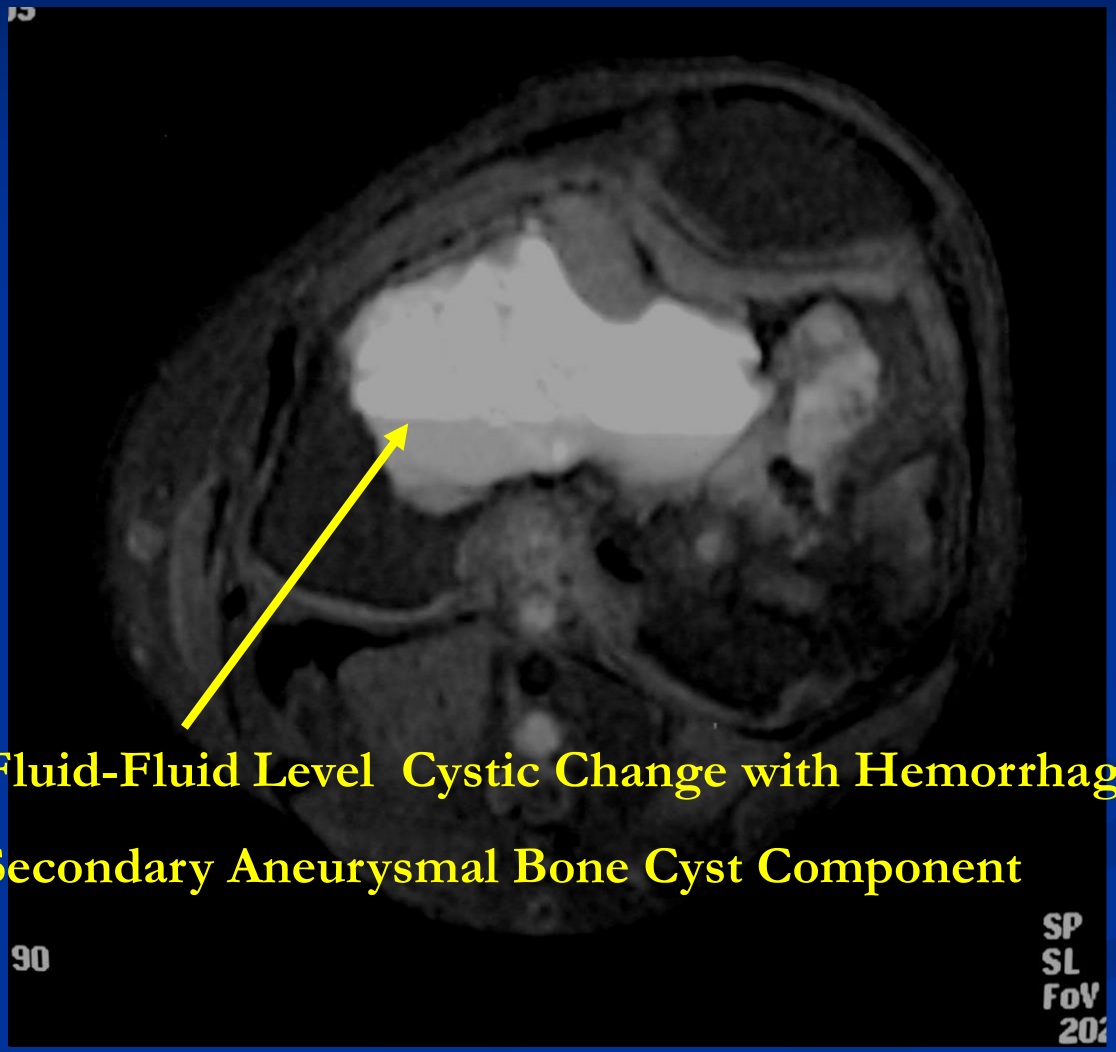


MRI T2 Weighted of Giant Cell Tumor of Distal Femur

- Soft Tissue Extension (Stage 3 Tumor)
- Fluid-Fluid Levels Detected indicative of Aneurysmal Bone Cyst Changes or a Secondary ABC Component

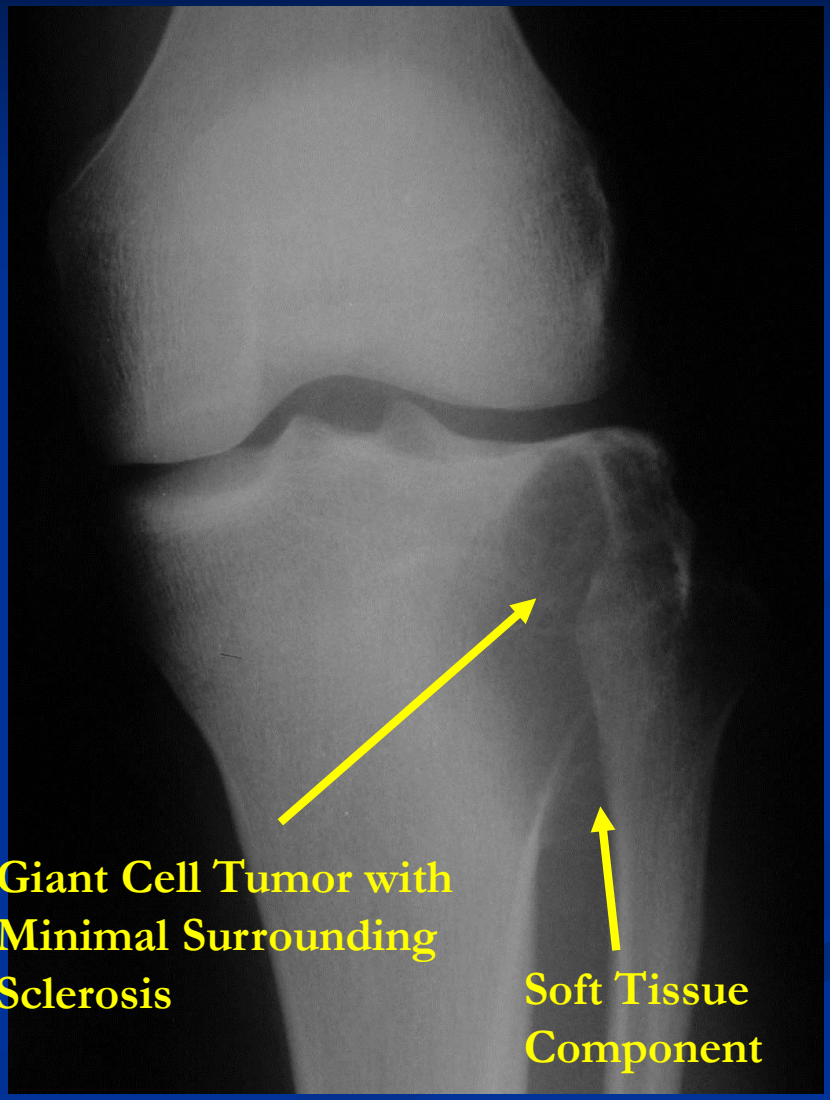


MRI: Giant Cell Tumor of Distal Femur with Secondary Aneurysmal Bone Cyst Component



X-Ray: Giant Cell Tumor of Proximal Tibia

- Eccentric, Lytic Lesion
- Metaphyseal with Extension into Epiphysis
- Geographic Pattern
- Minimal Surrounding Sclerosis
- No Matrix Mineralization

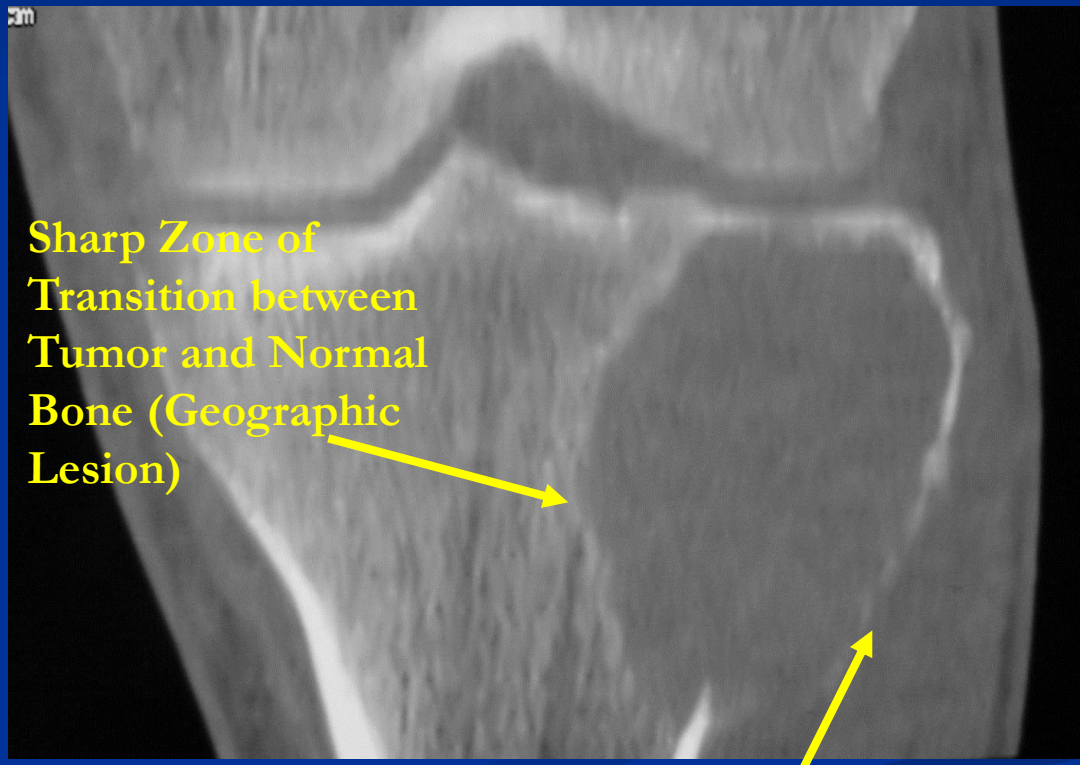


X-Ray: Lateral of Giant Cell Tumor of Proximal Tibia

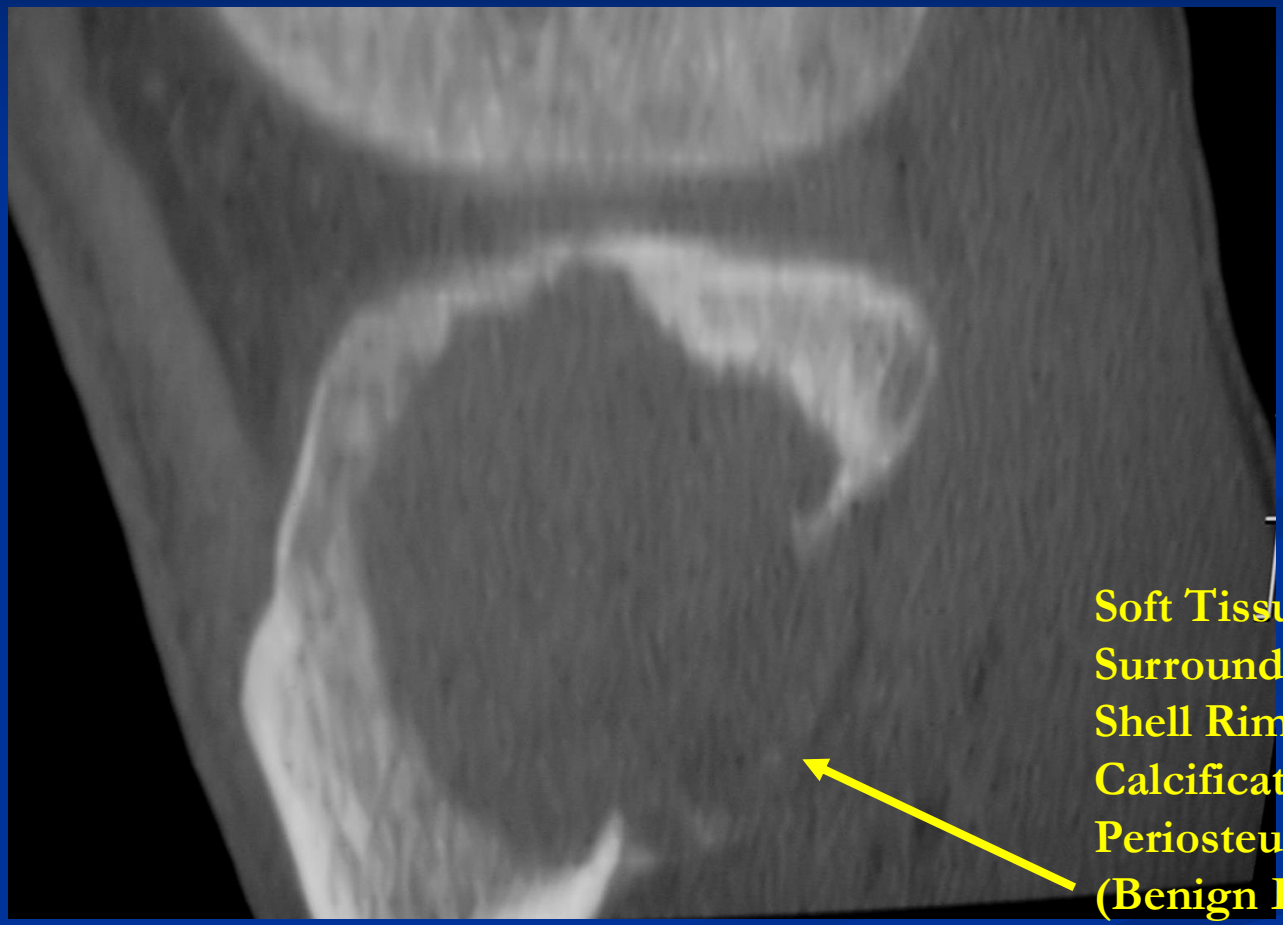
Giant Cell Tumor of Proximal Tibia

CT Scan of Giant Cell Tumor of Proximal Tibia

- Bone is expanded where tumor is eroding cortex
- The periosteum is intact where tumor destroys cortex and extends into soft tissue
- Eccentric Lesion
- Geographic/Well Circumscribed
- No Matrix Mineralization

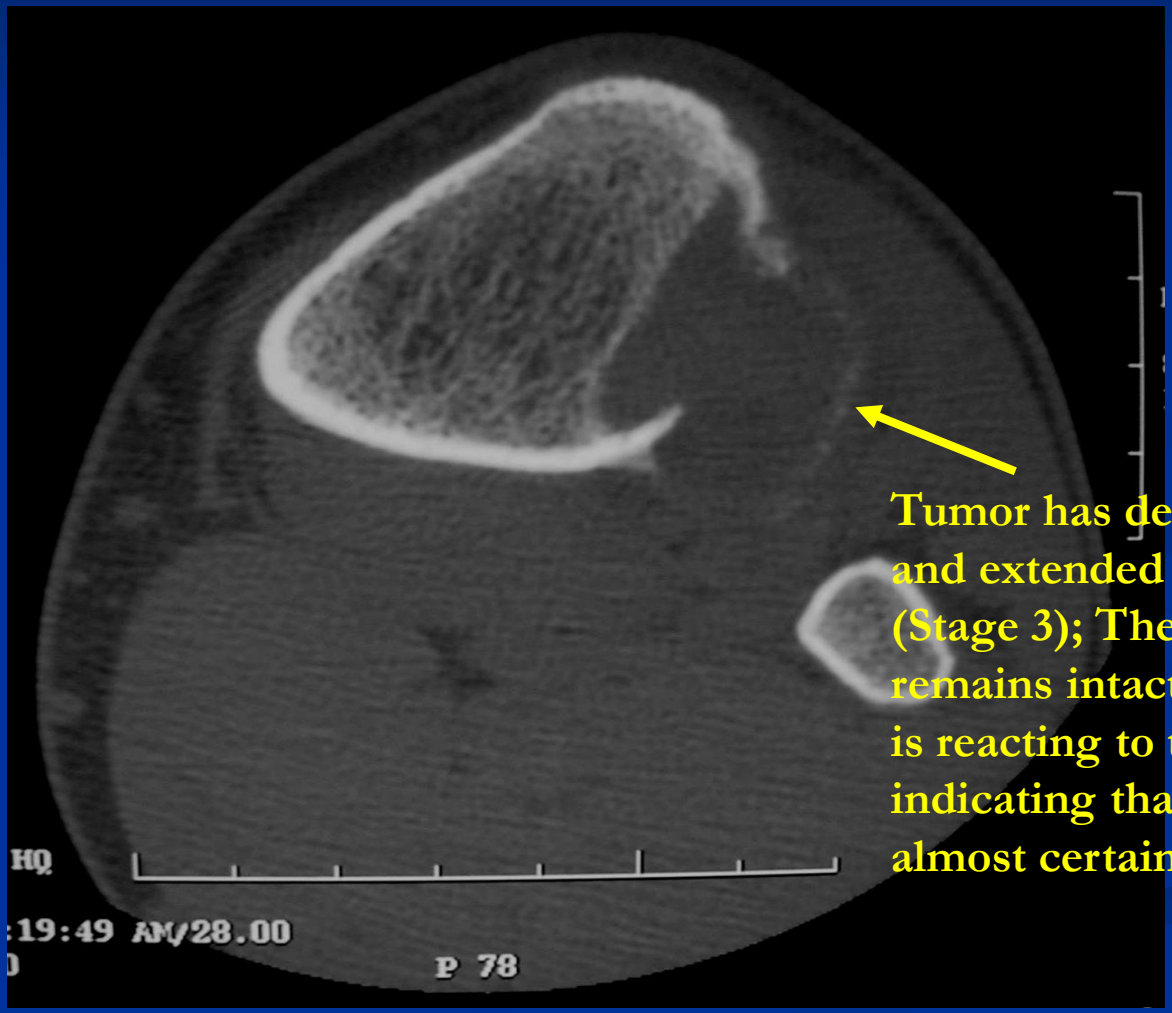


CT Scan: Sagittal Image of GCT of Proximal Tibia Stage 3



Soft Tissue Component Surrounded by Egg Shell Rim of Calcification indicating Periosteum is Intact (Benign Lesion)

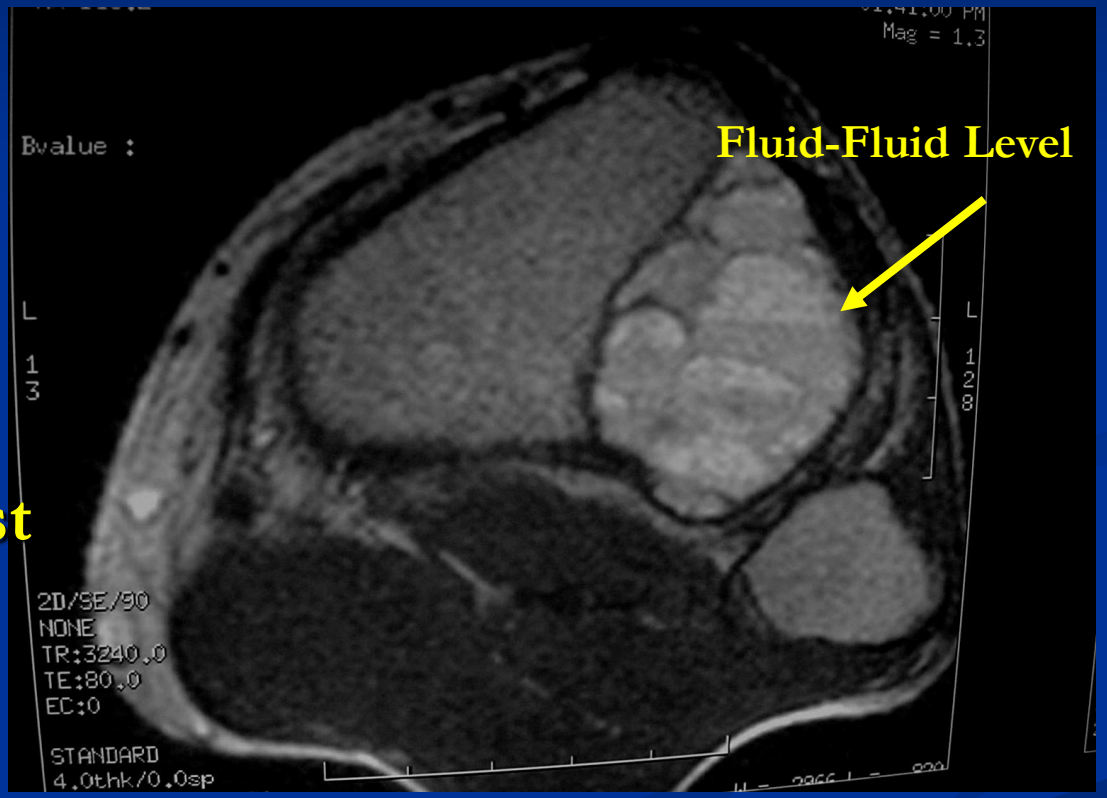
CT Scan: Axial Section of Giant Cell Tumor of Proximal Tibia



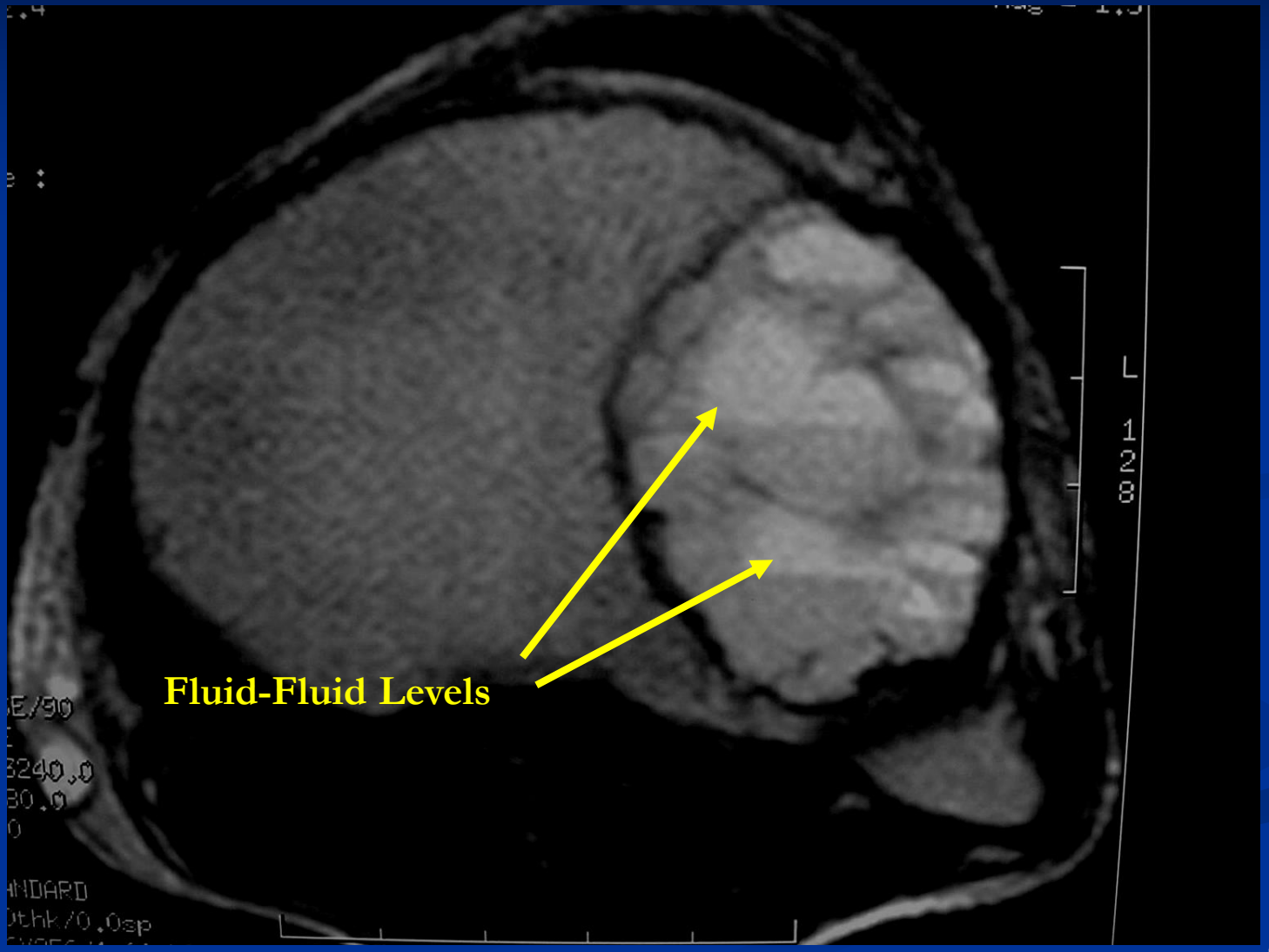
Tumor has destroyed cortex and extended into soft tissue (Stage 3); The periosteum remains intact (arrow) and is reacting to the lesion indicating that the lesion is almost certainly benign

MRI of Proximal Tibia Giant Cell Tumor

- There are multiple fluid-fluid levels indicative of a **secondary aneurysmal bone cyst component**



MRI T2 Weighted Image Demonstrating Multiple Fluid-Fluid Levels (Secondary ABC Changes)

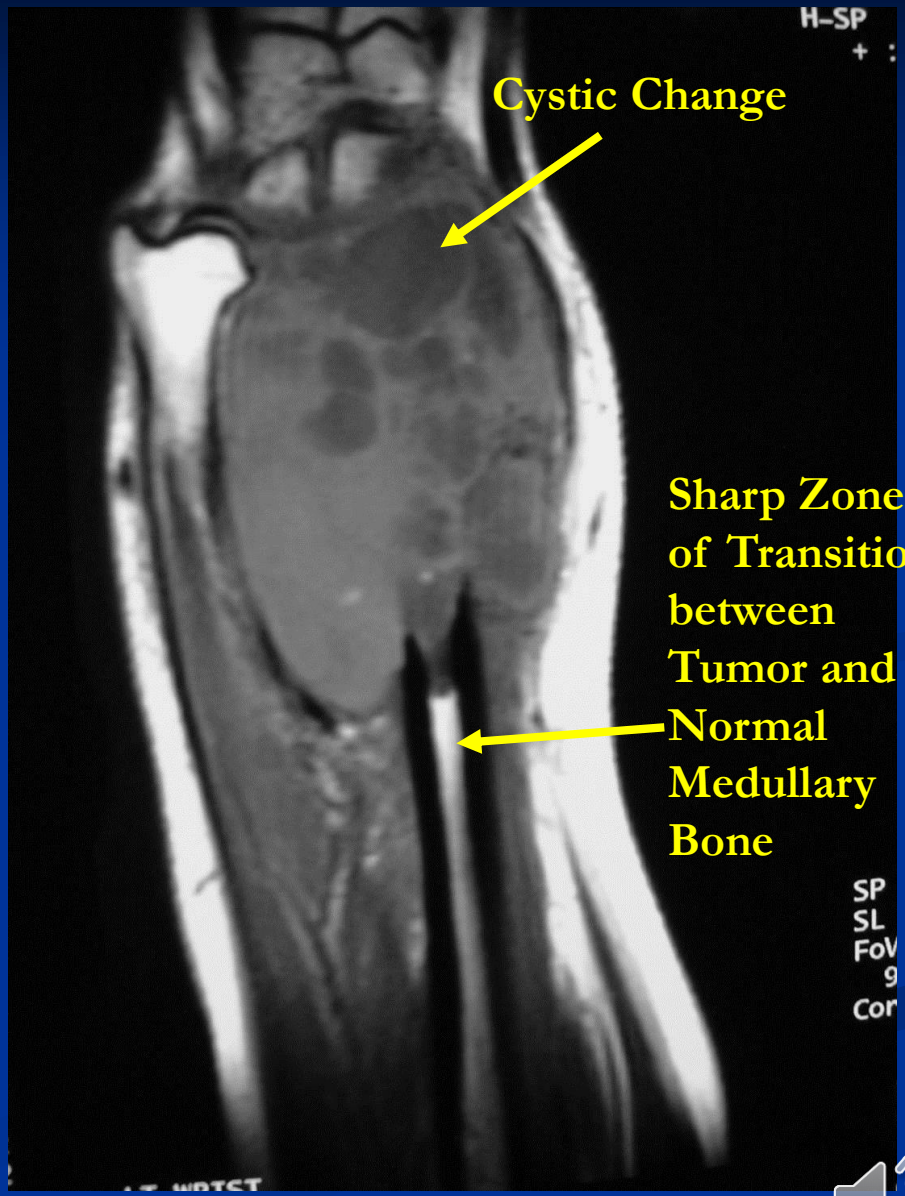


X-Ray: Giant Cell Tumor of Distal Radius (Stage 3)

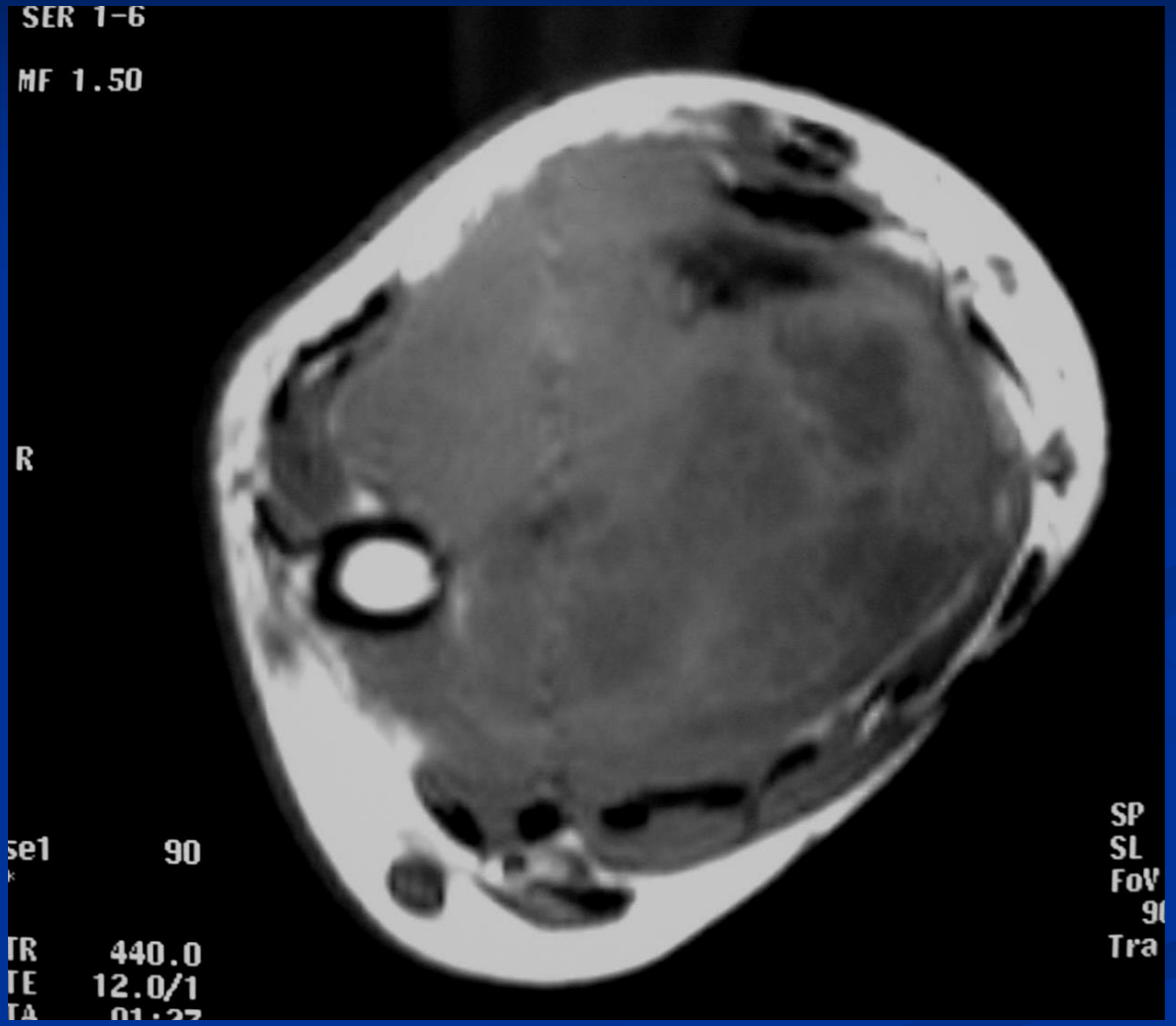
- Expansile Tumor Destroying Distal Radius
- Metaphyseal Origin with Destruction of Metaphysis and Epiphysis
- Multiple Internal Trabeculations

MRI T1 of Giant Cell Tumor of Distal Radius

- Large Destructive Stage 3 Giant Cell Tumor
- Cystic Changes
- Sharp Zone of Transition between Tumor and Normal Medullary Bone



MRI: Large Stage 3 Giant Cell tumor of Distal Radius

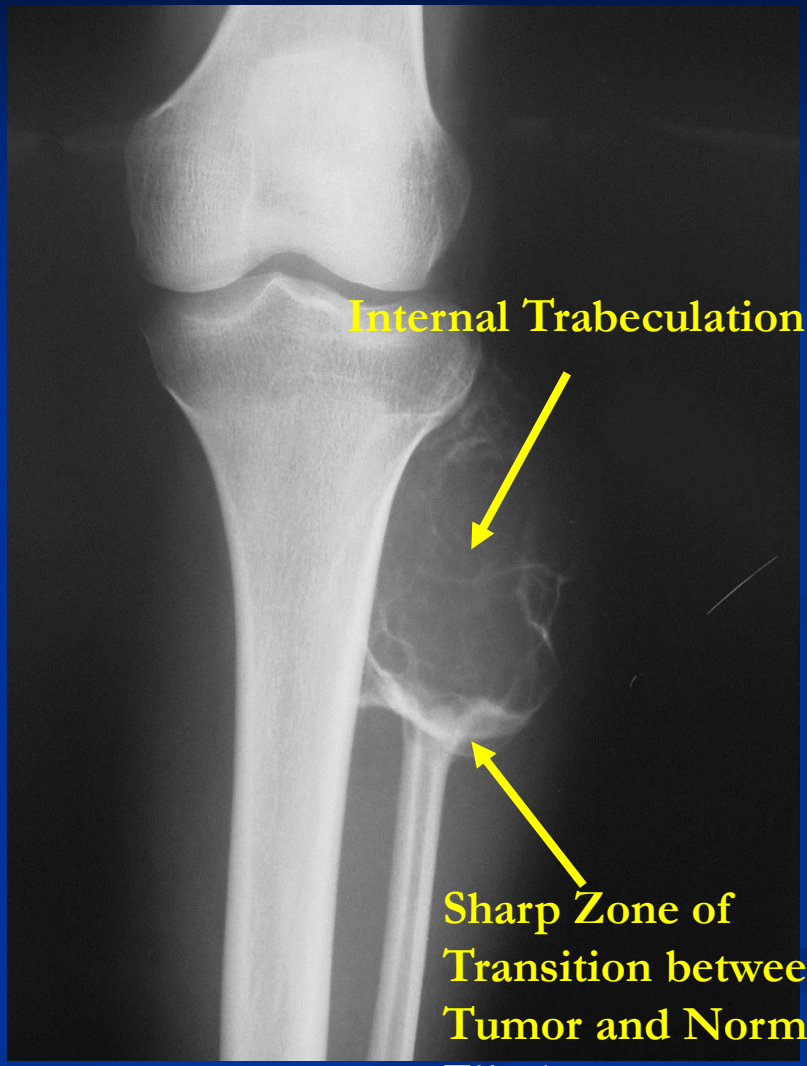


X-Ray: Giant Cell Tumor of 3rd Metacarpal of Hand

Giant Cell Tumor with Indistinct Margin Inferiorly on Xray

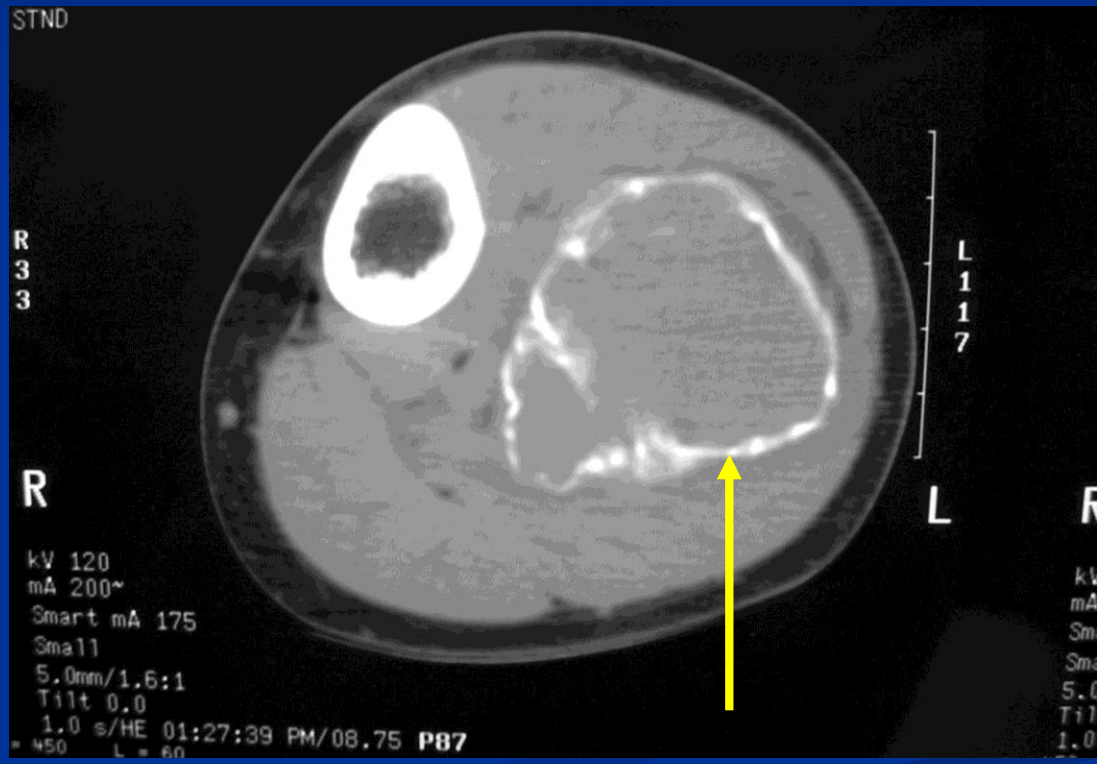
X-Ray: Giant Cell Tumor of Proximal Fibula (Stage 3)

- Large Expansile Lesion Destroyed Proximal Fibula
- Internal Trabeculations
- Sharp Zone of Transition between Tumor and Normal Fibula



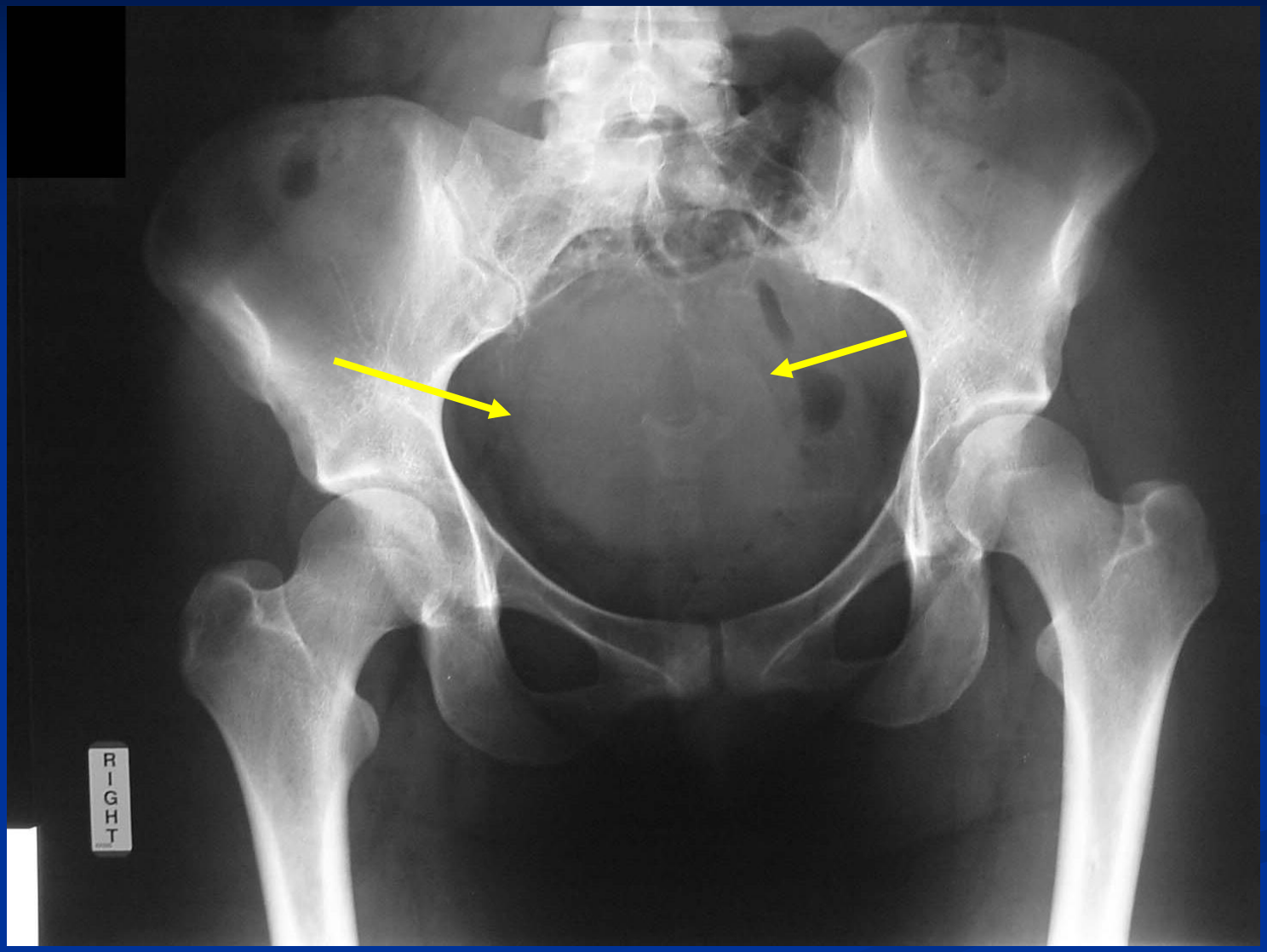
CT Scan of Giant Cell Tumor of Proximal Fibula

- Periosteum intact
- Reactive shell of bone; thin rim of calcification (reactive periosteum)
- This feature is consistent with a benign neoplasm

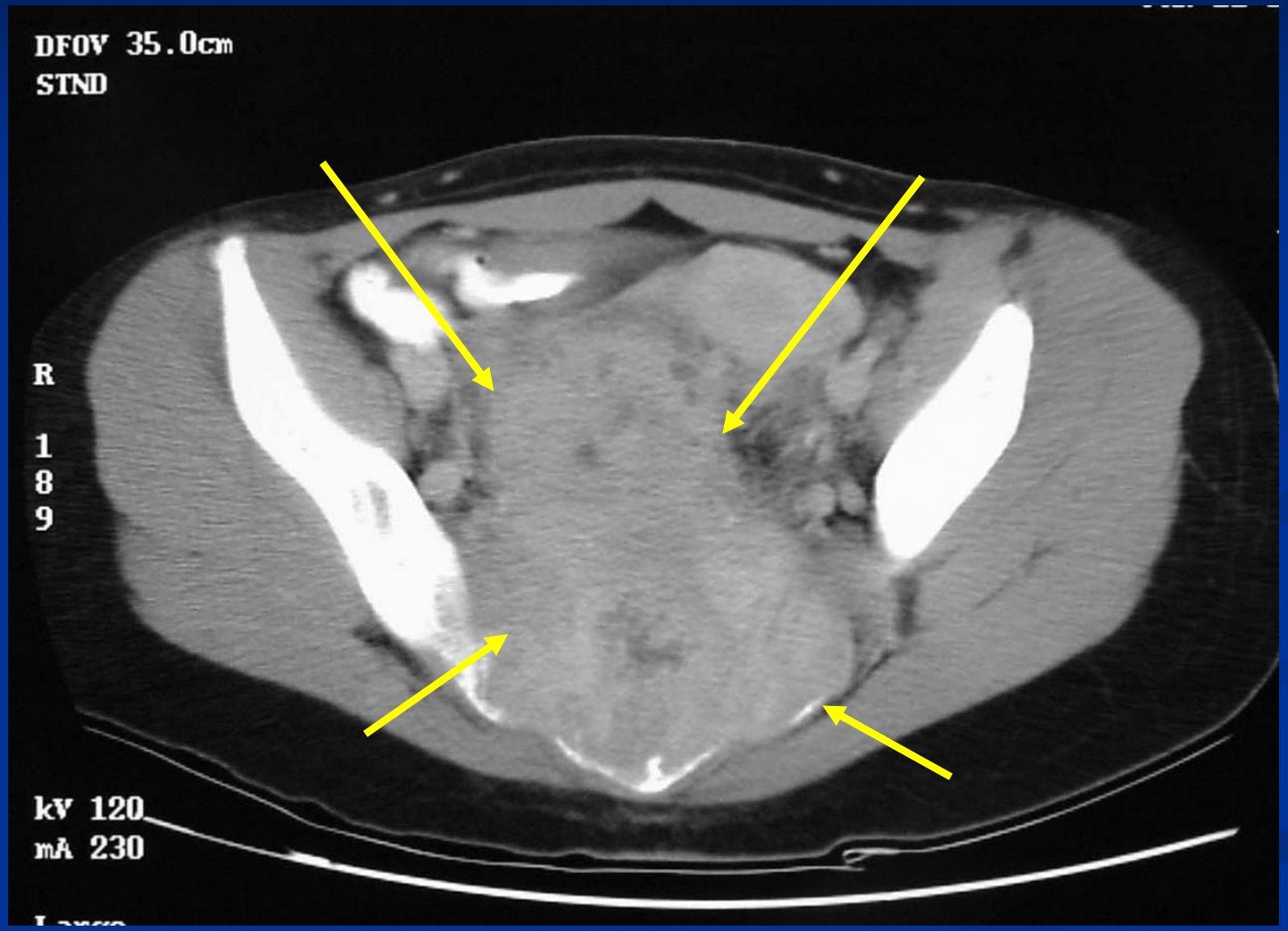


Reactive Periosteum Intact

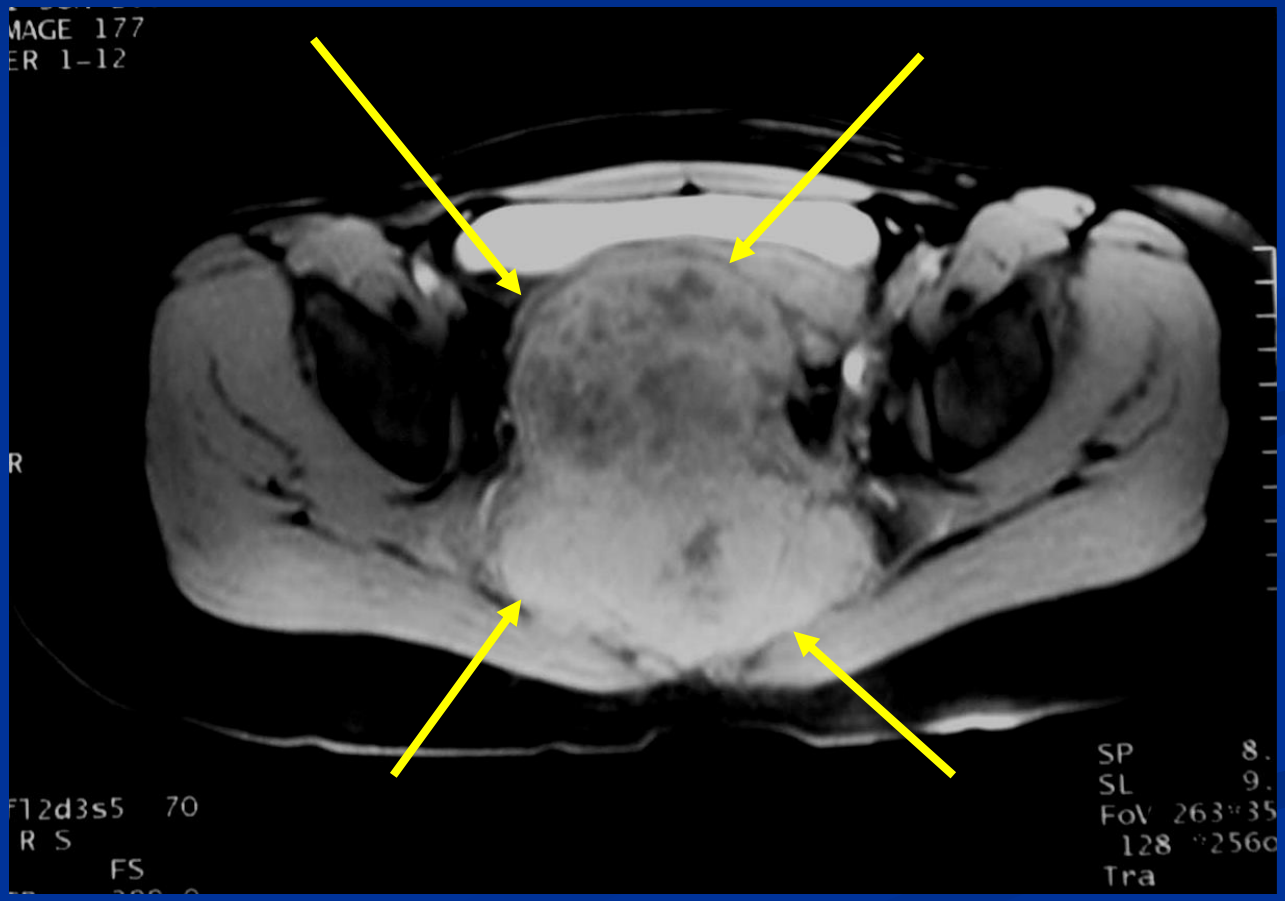
Xray of Giant Cell Tumor of Sacrum



CT Scan Showing a Giant Cell Tumor of the Sacrum



MRI: Giant Cell Tumor of the Sacrum

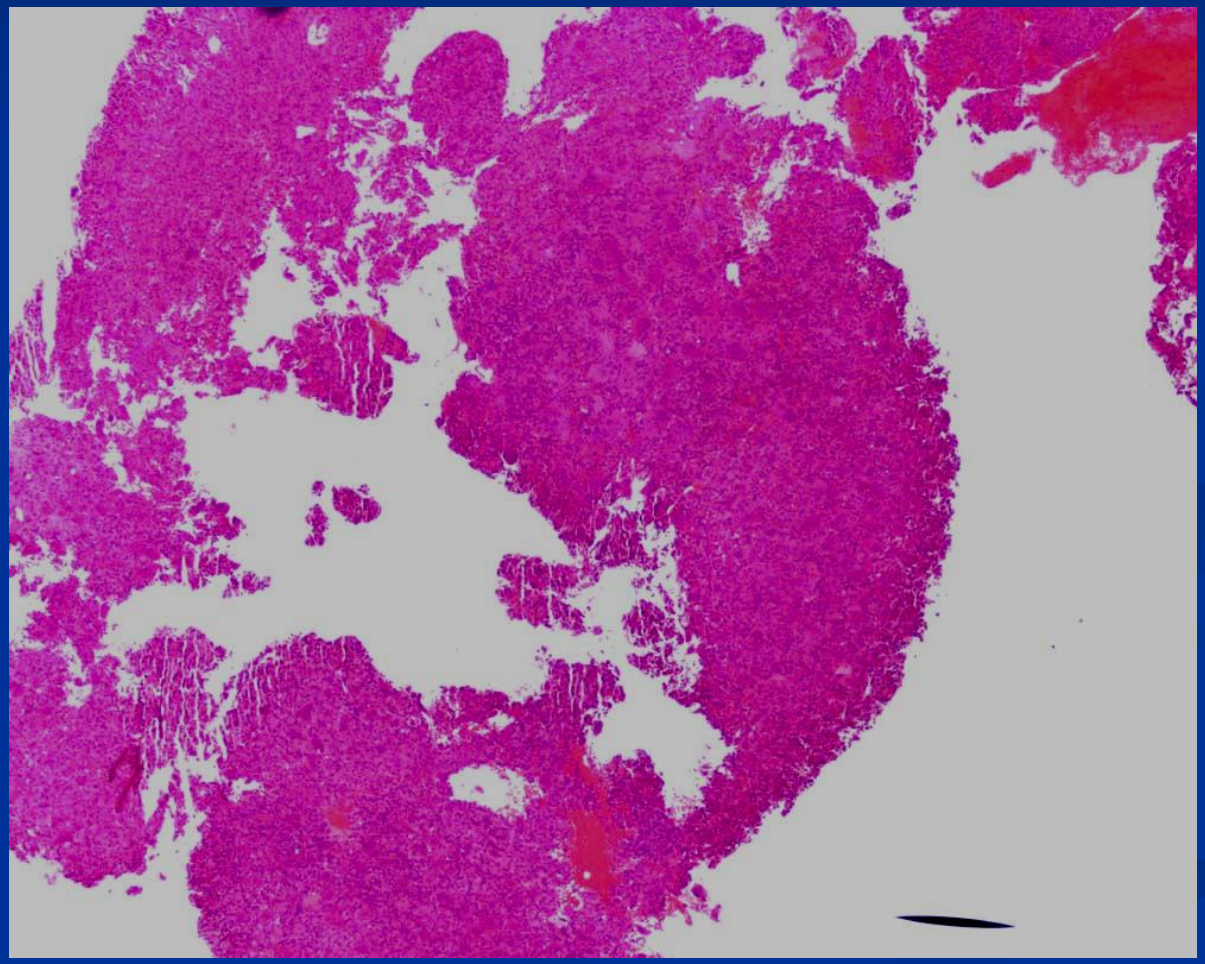


GCT

Pathology:

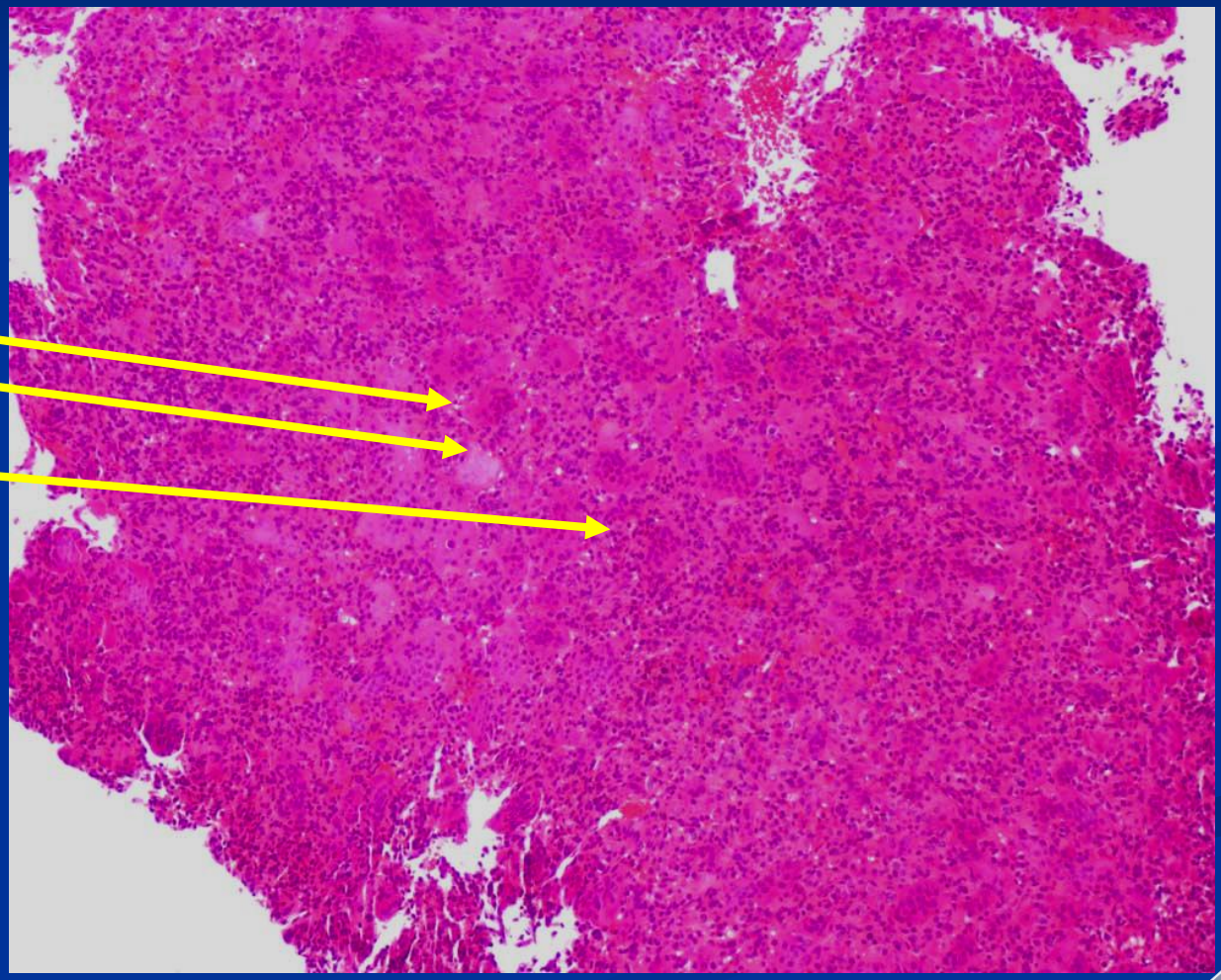
- Osteoclast like giant cells (90%)
- Spindle cell stromal component
- The stromal cell nuclei are identical to the giant cell nuclei; they coalesce to form the giant cells
- Hemorrage, necrosis and hemosiderin deposition are often present
- ABC like areas are present in 10-15%

Pathology: Microscopic Low Power View of Giant Cell Tumor



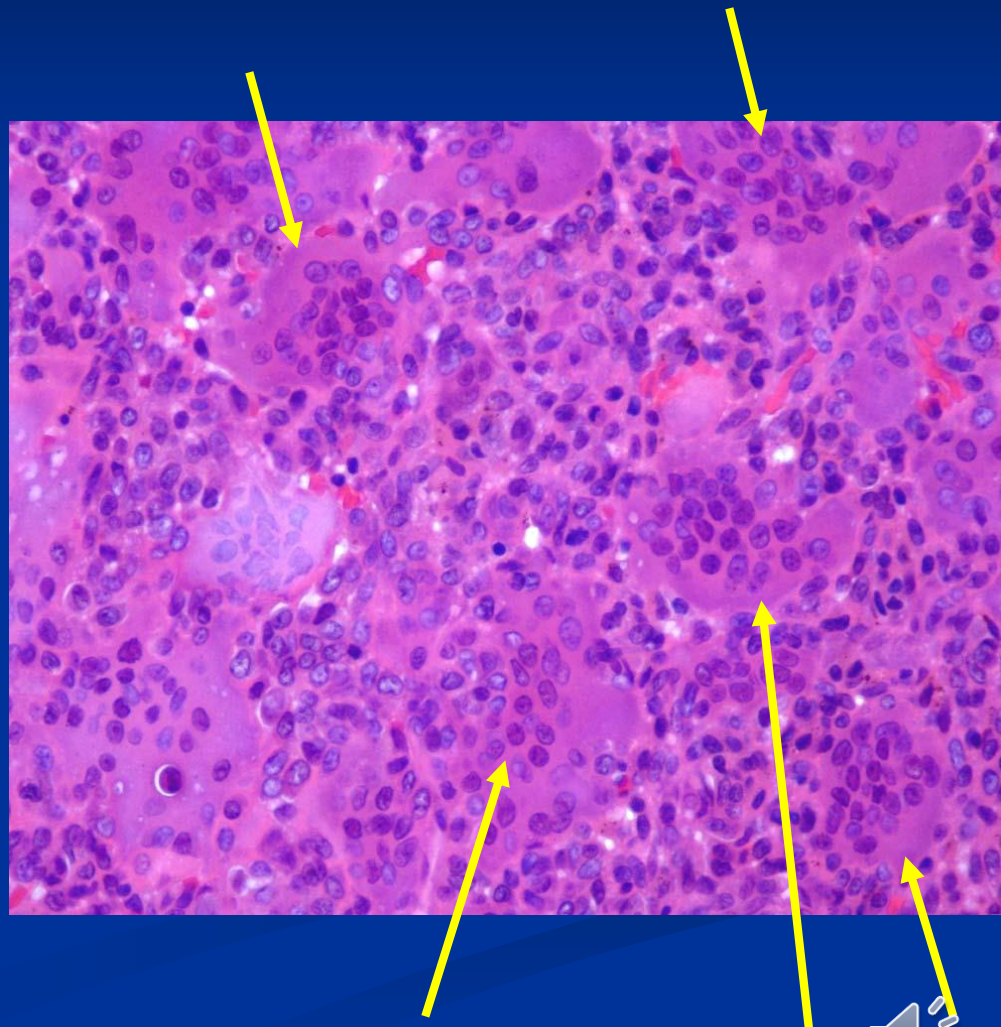
Pathology: Giant Cell Tumor Microscopic View: Intermediate Power

Multiple Multinucleated Giant Cells in a Sea of Mononuclear Cells



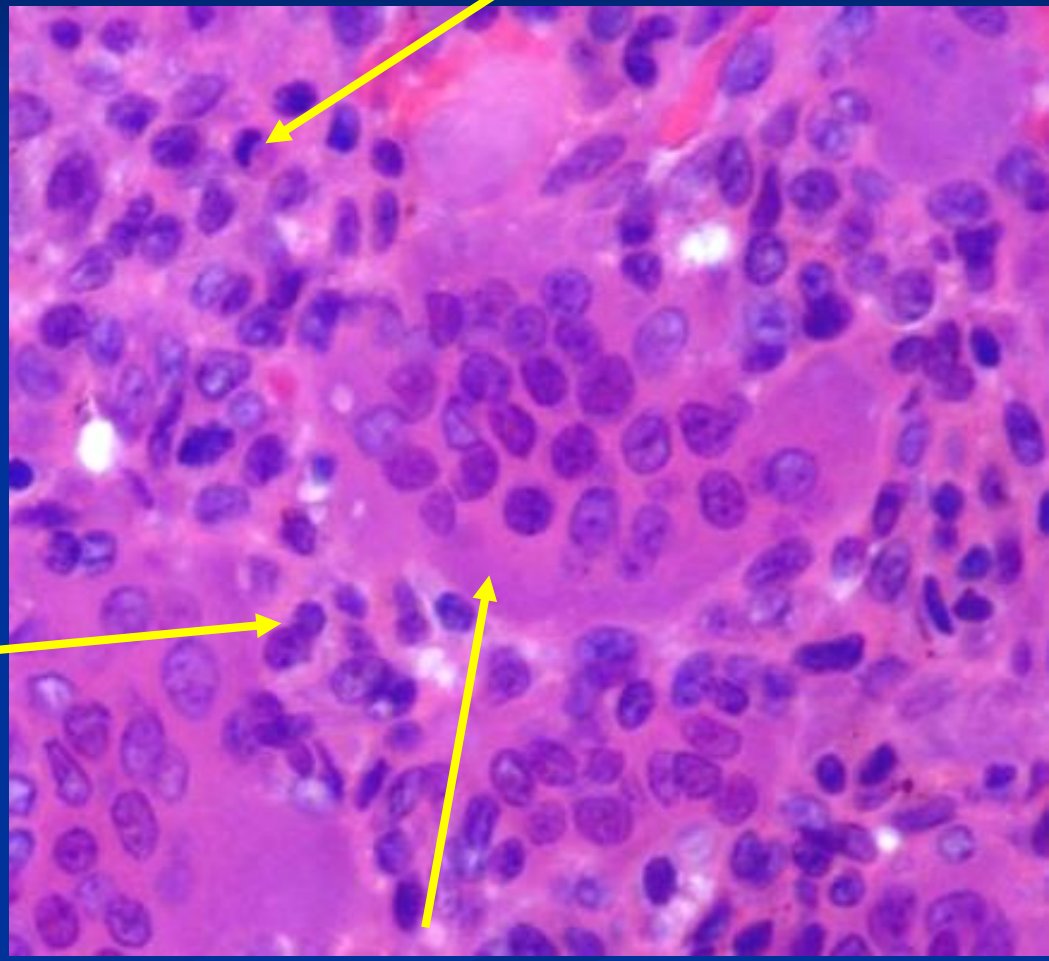
Pathology: Giant Cell Tumor
Microscopic High Power View

- Multiple Multinucleated Giant Cells (Arrows) in a Sea of Mononuclear Cells
- The nuclei of the Mononuclear Cells look identical to the nuclei within the Giant Cells



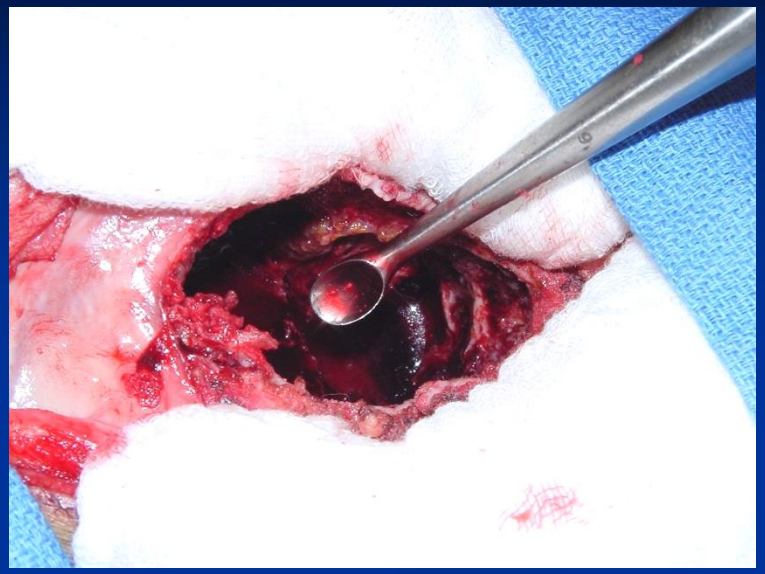
Pathology: High Power of a Giant Cell Tumor

- The nuclei of the cells in between the giant cells look very similar to the nuclei within the giant cells



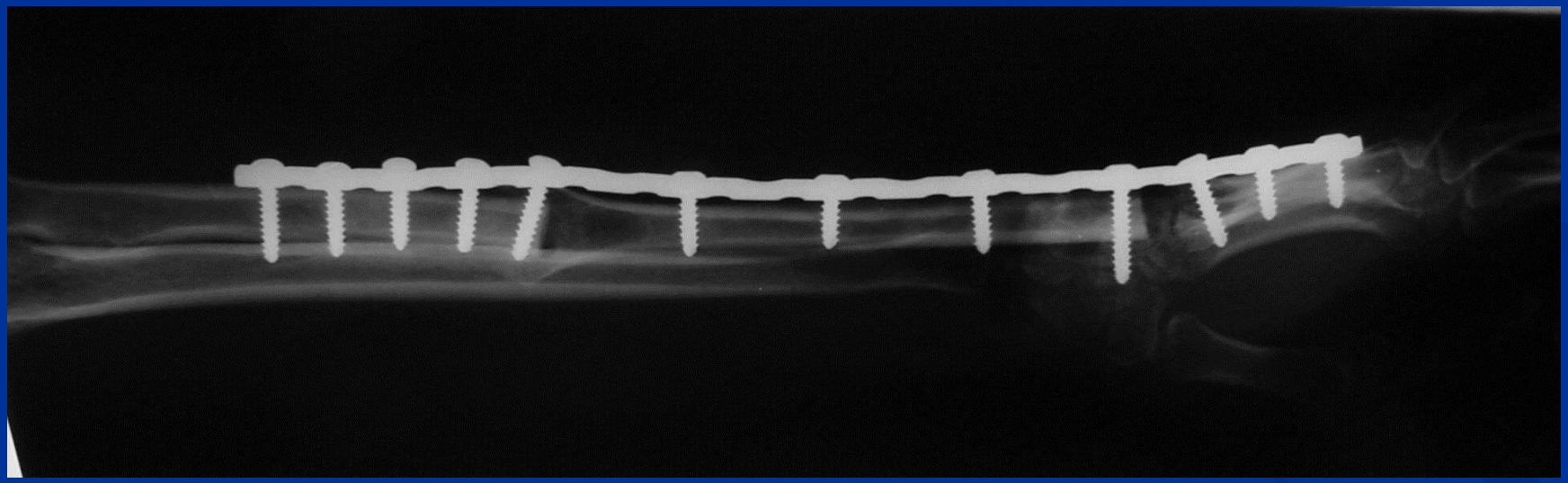
Treatment: GCT

- **GCT: Benign aggressive; They grow and destroy the bone and often the adjacent joint**
- **Surgery: Intralesional Curettage-Resection and Cement** plus adjuvant liquid nitrogen, phenol, etc
- **En-bloc resection** for some Stage 3 lesions if there will be insufficient bone stock remaining for reconstruction following a curettage



Treatment of a Stage 3 Giant Cell Tumor of the Distal Radius with a Wide/Radical Resection and Reconstruction with a Free Nonvascularized Fibula Autogenous Bone Graft and Internal Fixation; The Wrist was Fused

Treatment of a Stage 3 Giant Cell Tumor of the Distal Radius with a Wide/Radical Resection and Reconstruction with a Free Nonvascularized Fibula Autogenous Bone Graft and Internal Fixation; The Wrist was Fused



Prognosis GCT

- Osseous recurrence – new bone destruction; area of lysis adjacent to the cement
- Soft tissue recurrence – mass and may **calcify**
- Metastatic rate – 3%
  - Lungs—most common site
  - Controversy: are mets really retrospectively from a malignant GCT; Do GCTs metastasize from surgical procedure forcing tumor emboli into venous system?
- Malignant GCT –rare entity (more common after radiation)

Aneurysmal Bone Cyst (ABC)

- **Definition:** Benign aggressive lesion of bone with cystic blood filled cavities. It is locally destructive. The cystic cavities are blood filled and the walls contain spindle cells, reactive osteoid and multinucleated giant cells.
- 50% arise secondary to a pre-existing lesion
  - Secondary ABC
- Debate: Is ABC a cyst vs neoplasm vs a periosteal to intraosseous arteriovenous malformation?

Aneurysmal Bone Cyst (ABC)

- Clinical features:
  - 2% of all biopsied primary osseous neoplasms (1/2 as common as GCT of bone)
  - Usually<20 years of age (80%)
  - Pain, swelling, pathologic fracture (10-20%)
  - May be associated with trauma
  - Slightly more common in women

Secondary ABC
Secondary Lesion from Underlying Condition or Tumor

- Benign lesions:
  - Chondroblastoma, CMF, NOF GCT, Fibrous dysplasia, UBC, Brown Tumor, Hemangioma, Giant Cell Reparative Granuloma

Aneurysmal Bone Cyst (ABC)

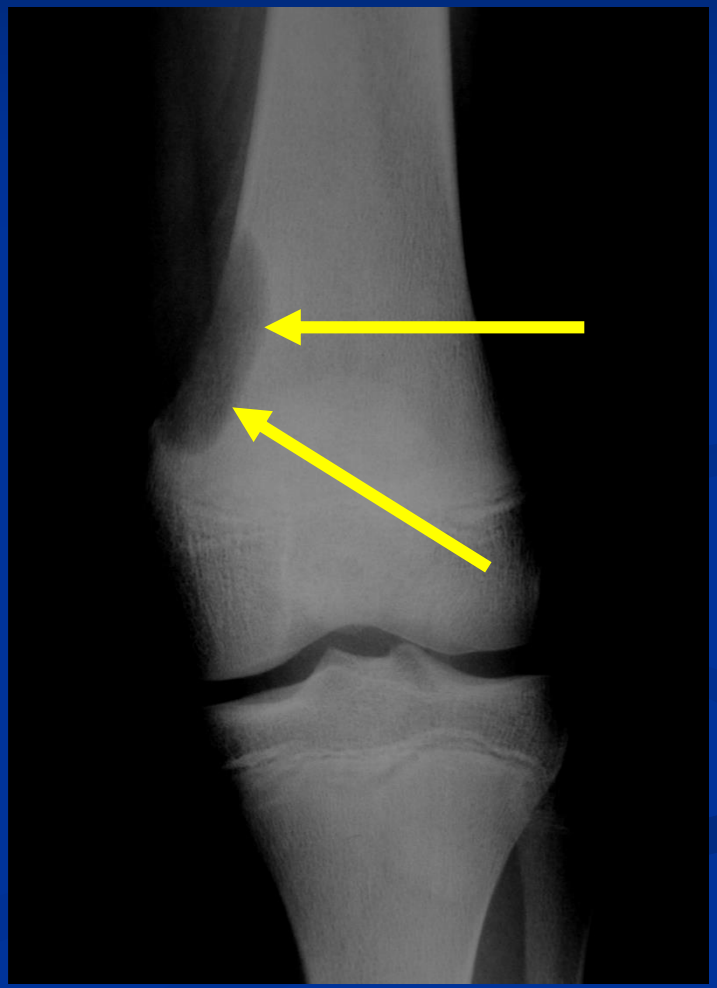
- Location
  - Metaphysis Long Bone 70-80%
    - **Distal Femur**
    - **Proximal Tibia**
  - Spine: posterior elements – 15% (thoracic, lumbar, cervical, sacral); In spine 50% may affect multiple spinous processes
  - Hands (10-15%)
  - Pelvis (5-10%)

Aneurysmal Bone Cyst (ABC)

- Radiology:
  - Eccentric, Parosteal or Central Geographic Lytic Lesion (Eccentric most common)
  - Metaphysis (80-90%), Diaphysis (10-20%)
  - Expansile Remodeling
  - Periosteal membrane usually intact CT/MRI
  - Bone scan – peripheral activity (65%)
  - **Fluid-fluid levels (CT/MRI) – nonspecific representing sedimentation of blood**

ABC Distal Femur
Eccentric, Geographic, Metaphyseal

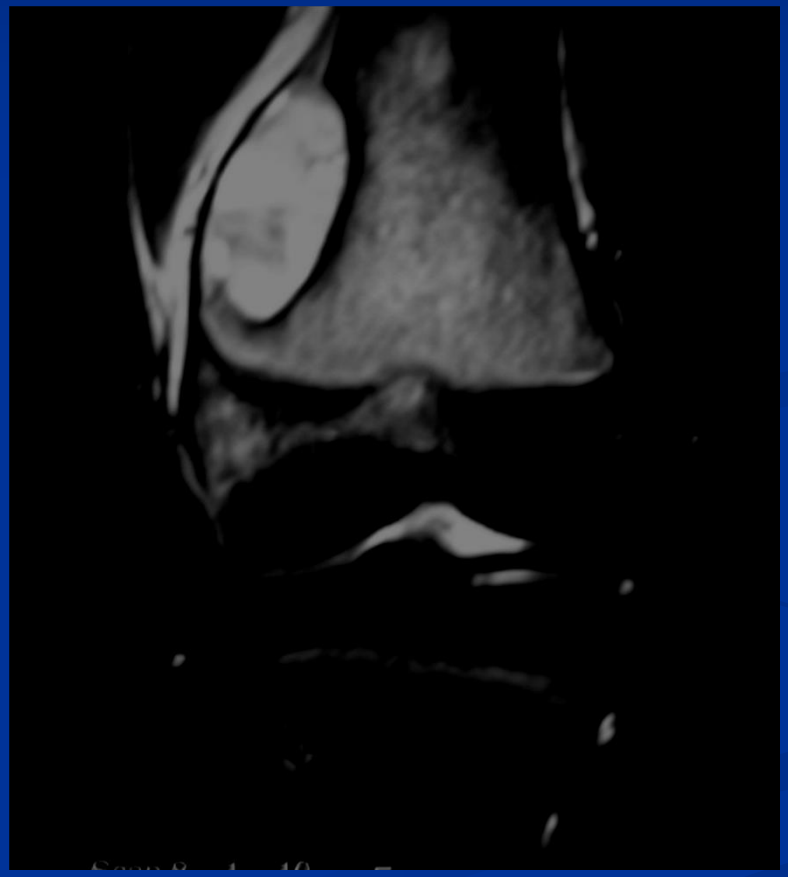
- Eccentric
- Geographic
- Metaphyseal
- Well Circumscribed
- Sclerotic Margin
- Skeletally Immature



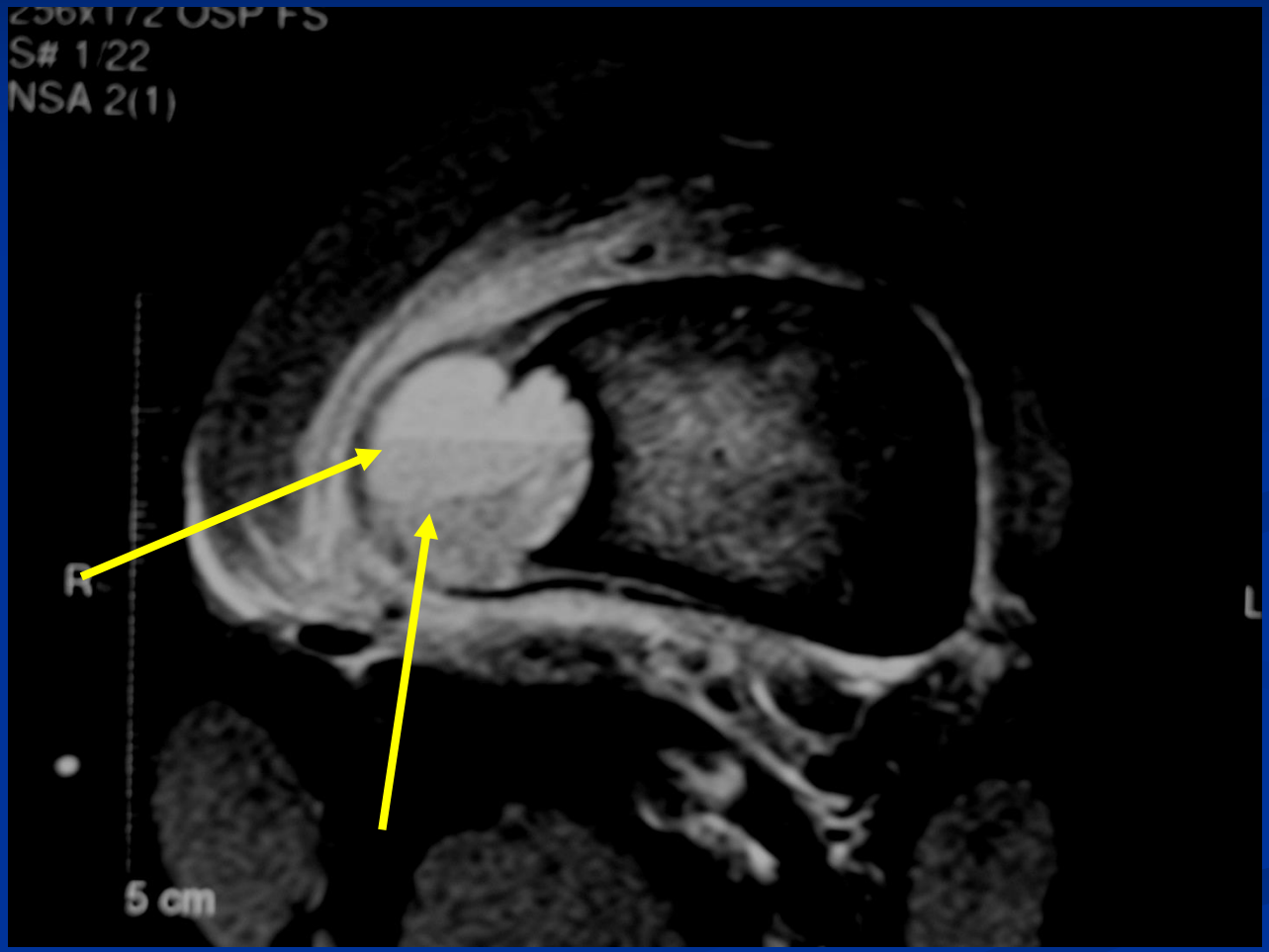
ABC: CT Scan

- Reactive Shell of Periosteal New Bone
- Encases Soft Tissue Component
- No Internal Mineralization

MRI T2 Weighted Image Surrounding Edema



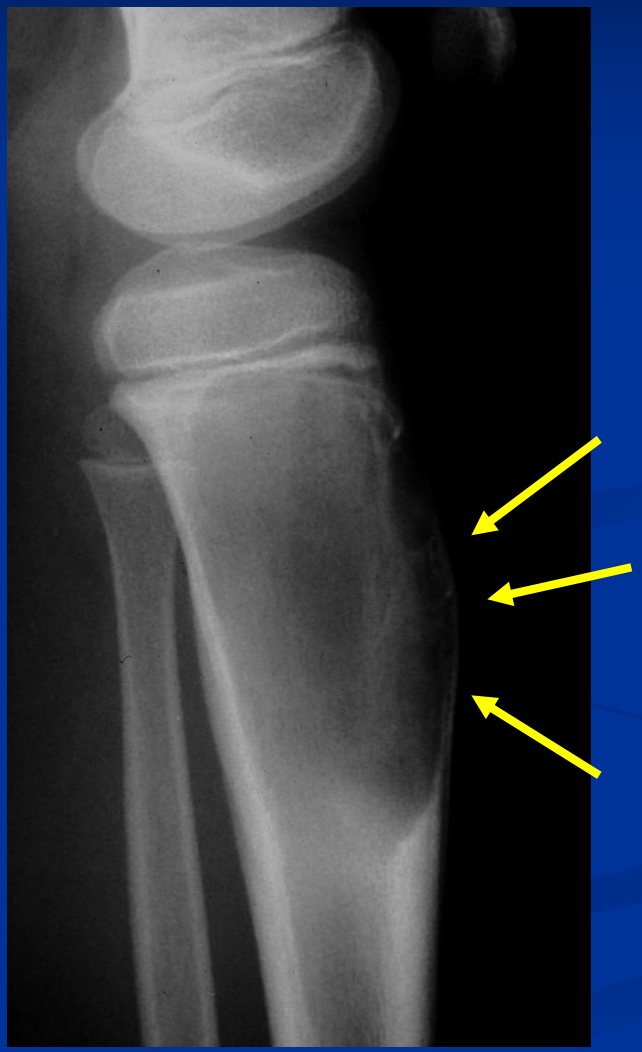
MRI T2 Weighted
Fluid-Fluid Level



ABC Proximal Tibia

- Central Lesion
- Geographic
- Expansile
- Metaphyseal
- Radiolucent
- Skeletally Immature

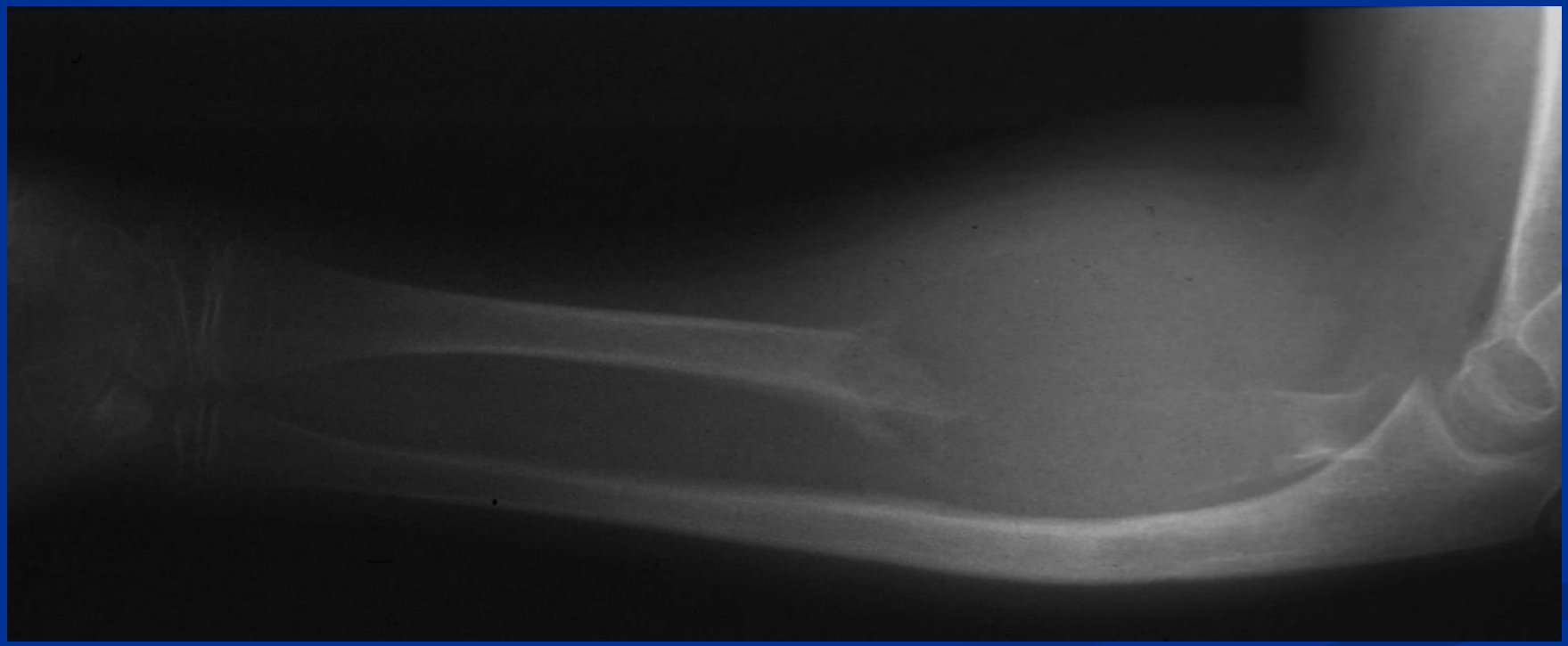
ABC Proximal Tibia
Eccentric Expansion of Bone



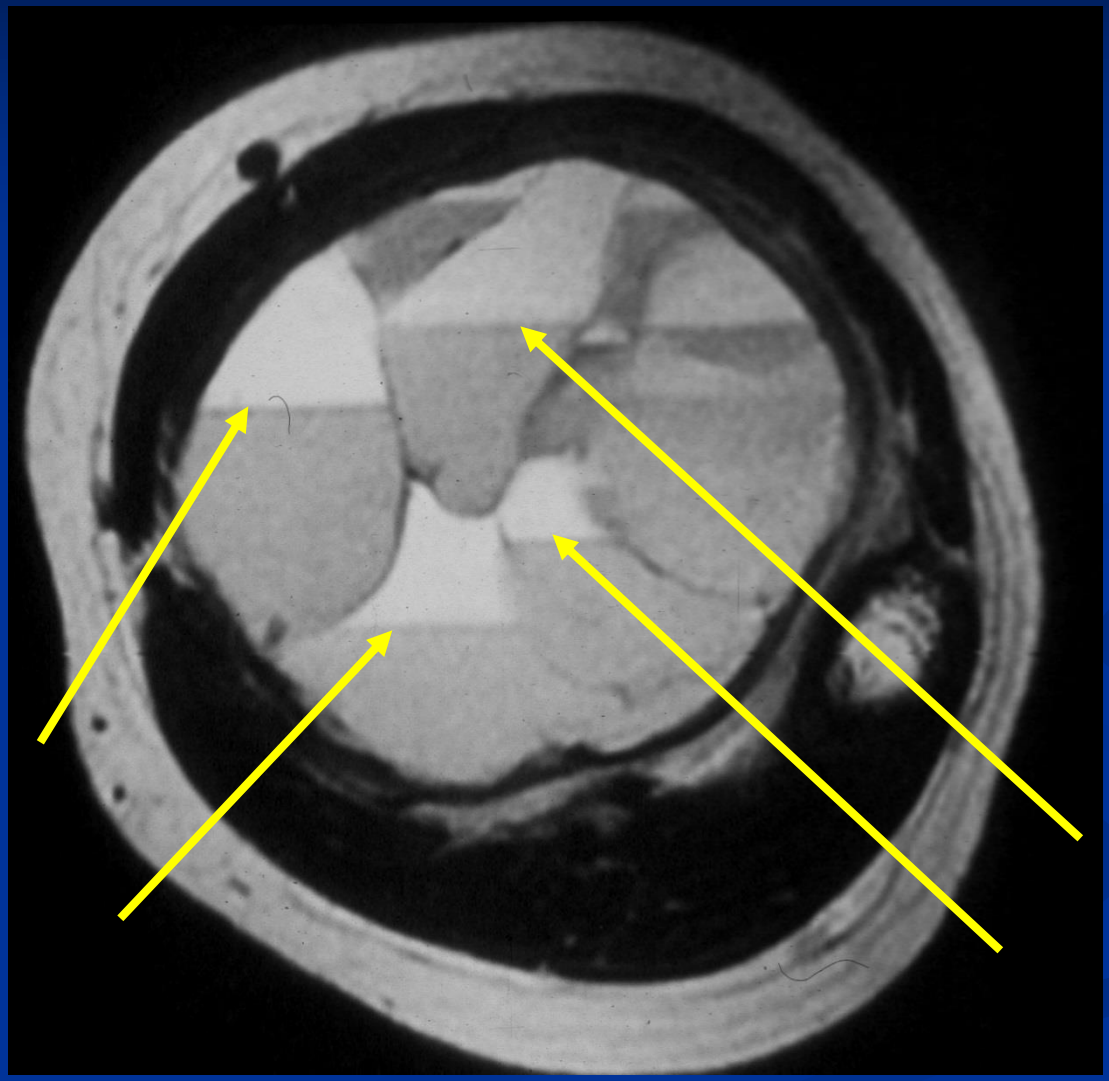
MRI: Fluid-Fluid Levels

ABC Proximal Radius
Geographic, Central, Expansile, Internal Trabeculations

ABC: Proximal Radius
Aggressive Growth



ABC: MRI Fluid-Fluid Levels

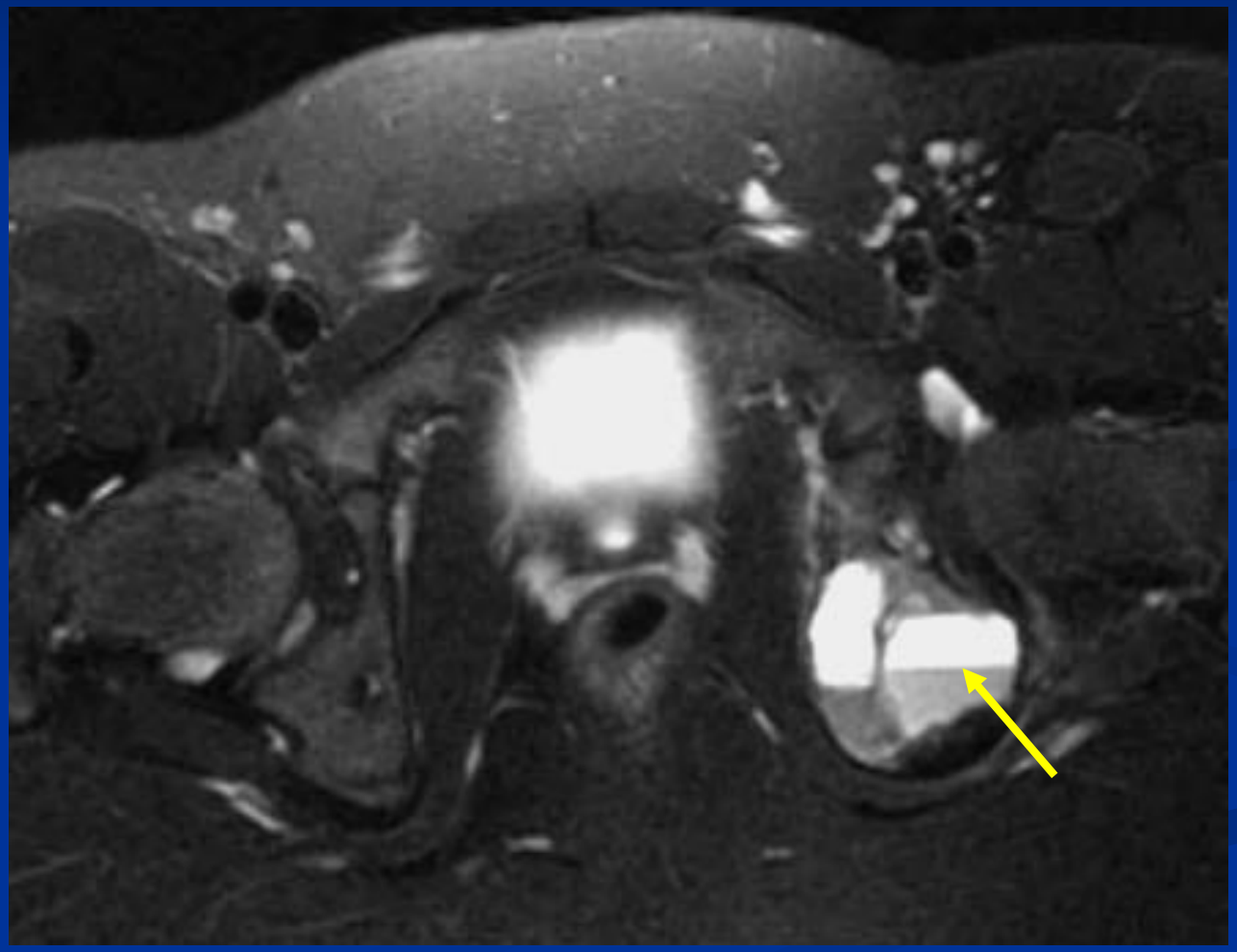


ABC of Left Ischium

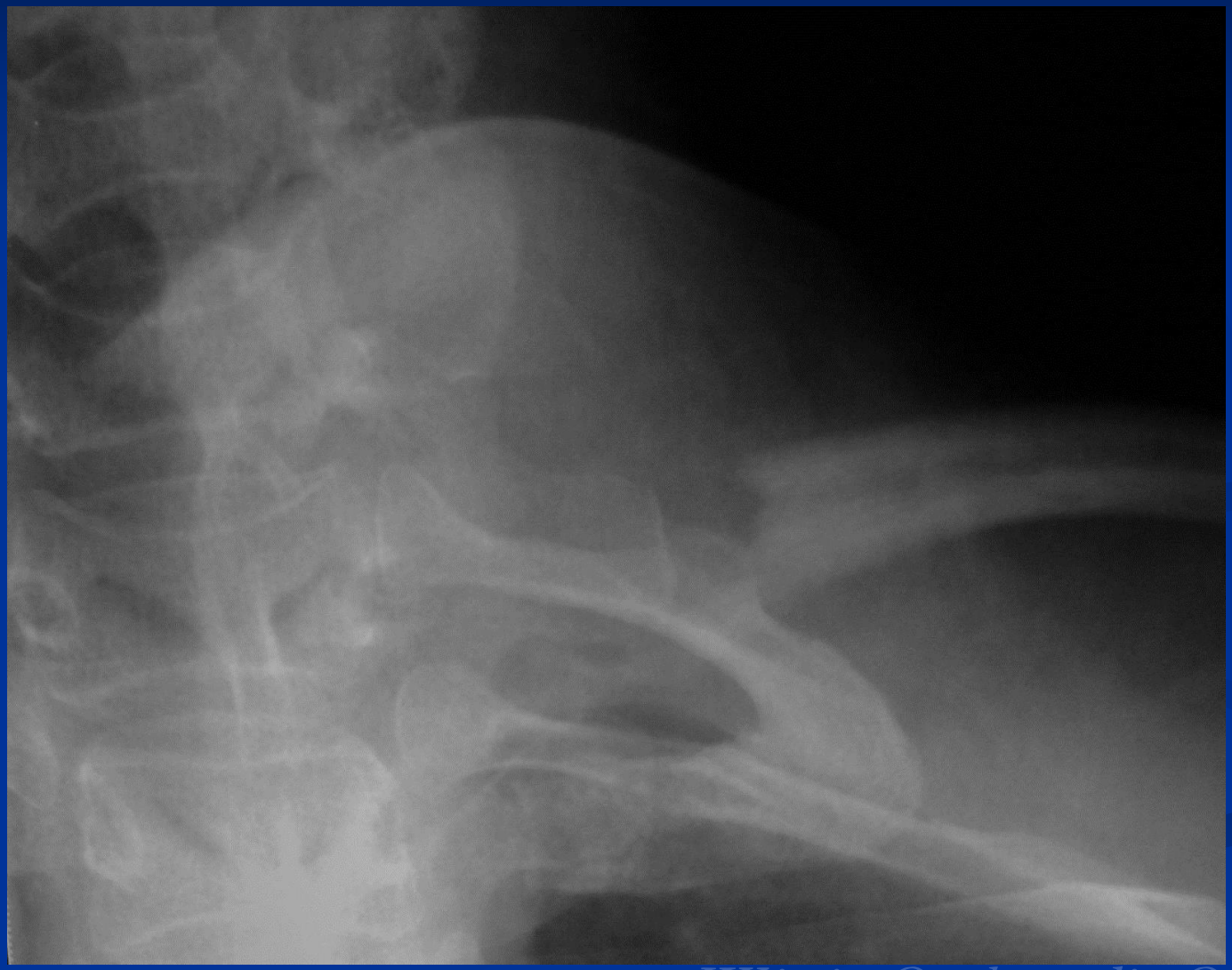
ABC of Left Ischium

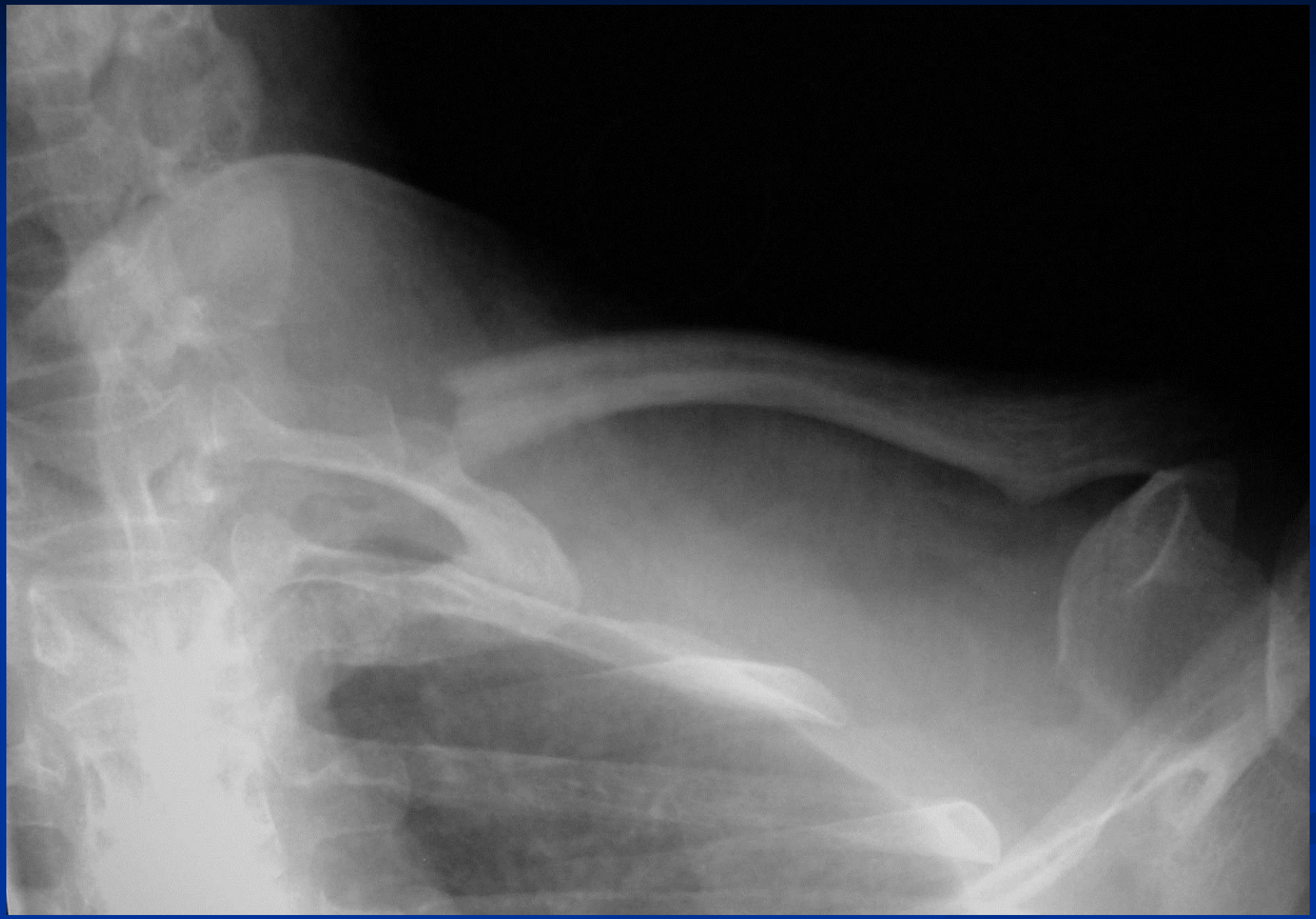
Expansile, Radiolucent, Skeletally Immature, Internal Trabeculations

MRI T2 Weighted: Fluid-Fluid Levels

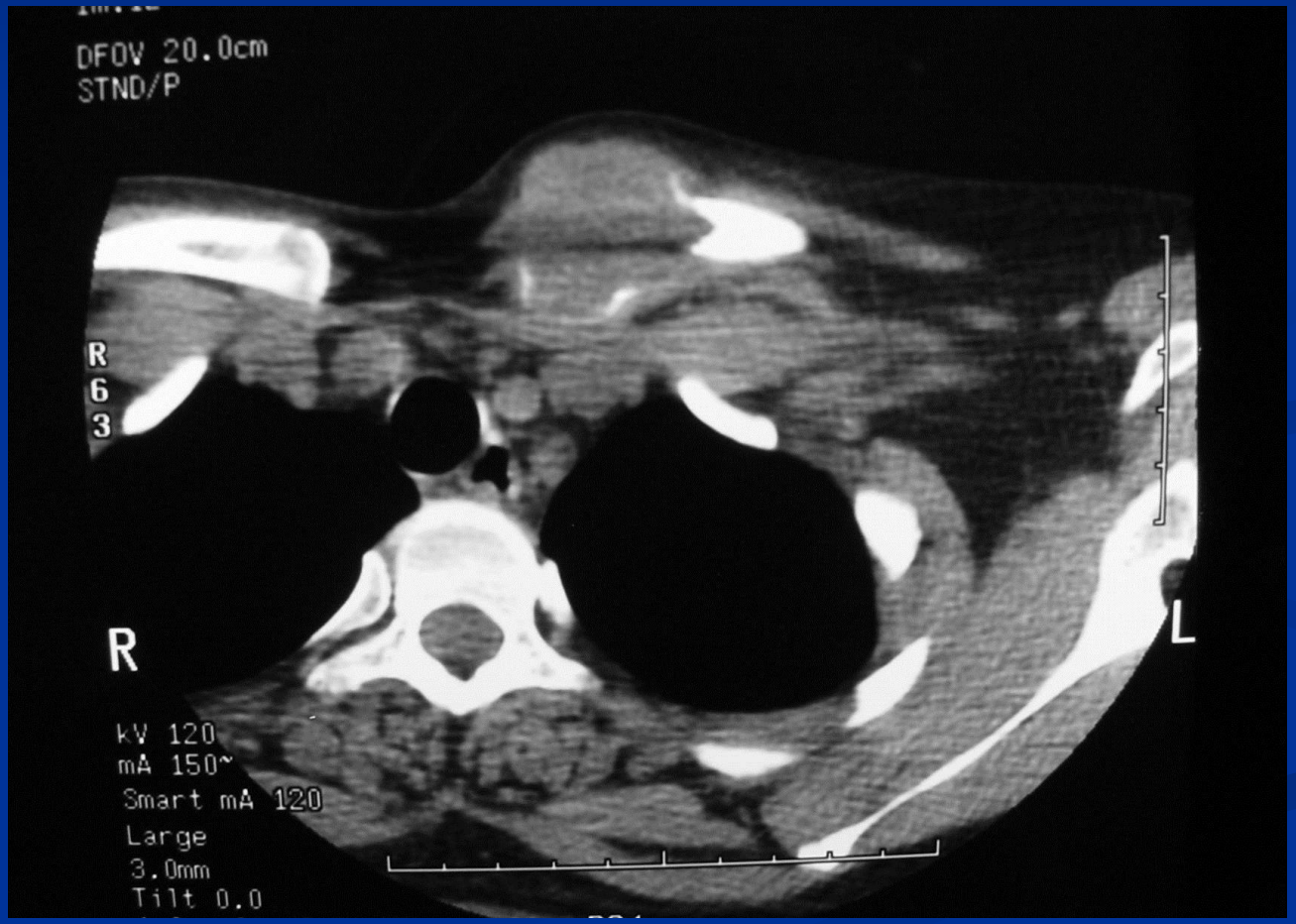


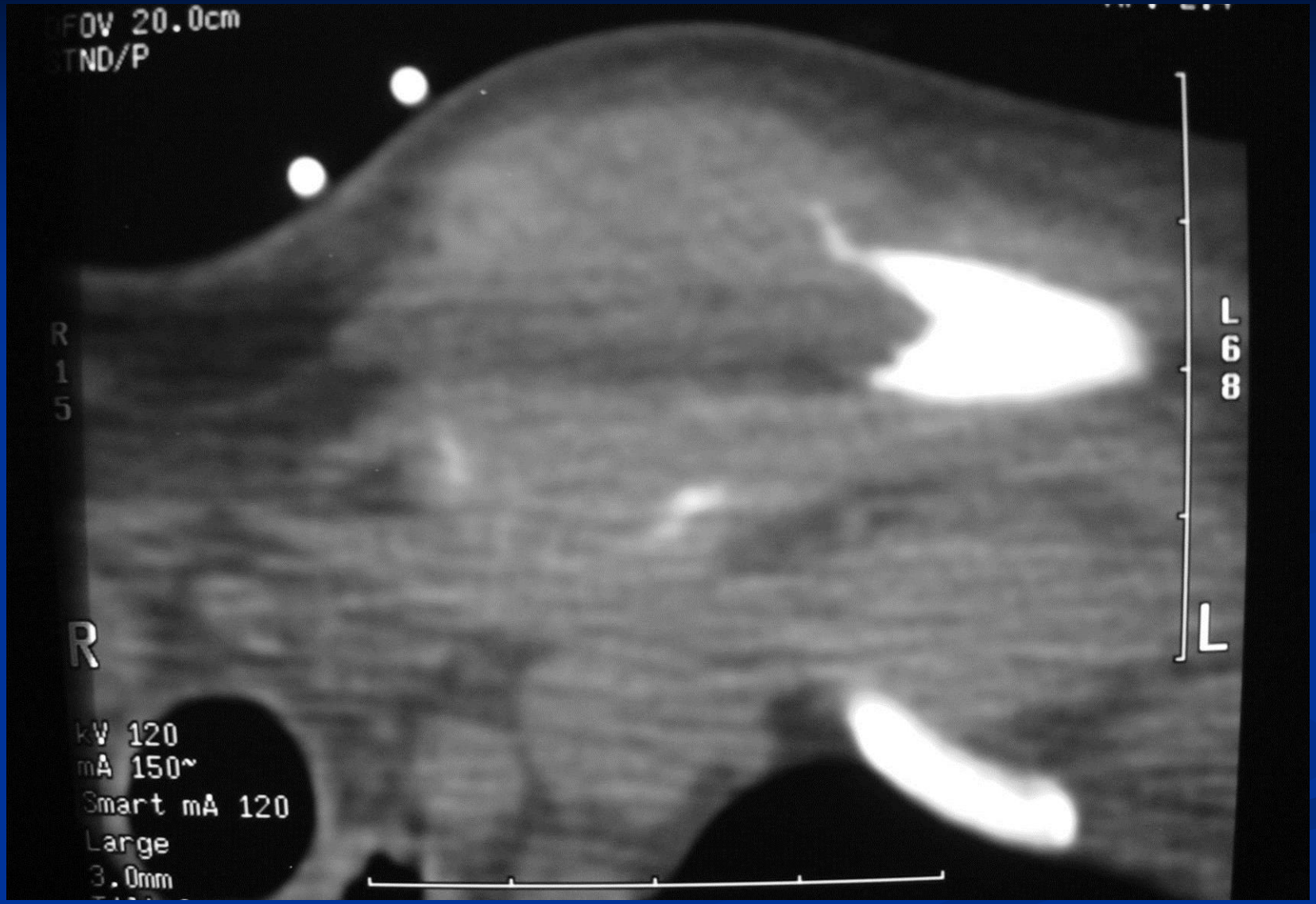
ABC of Medial End of Clavicle



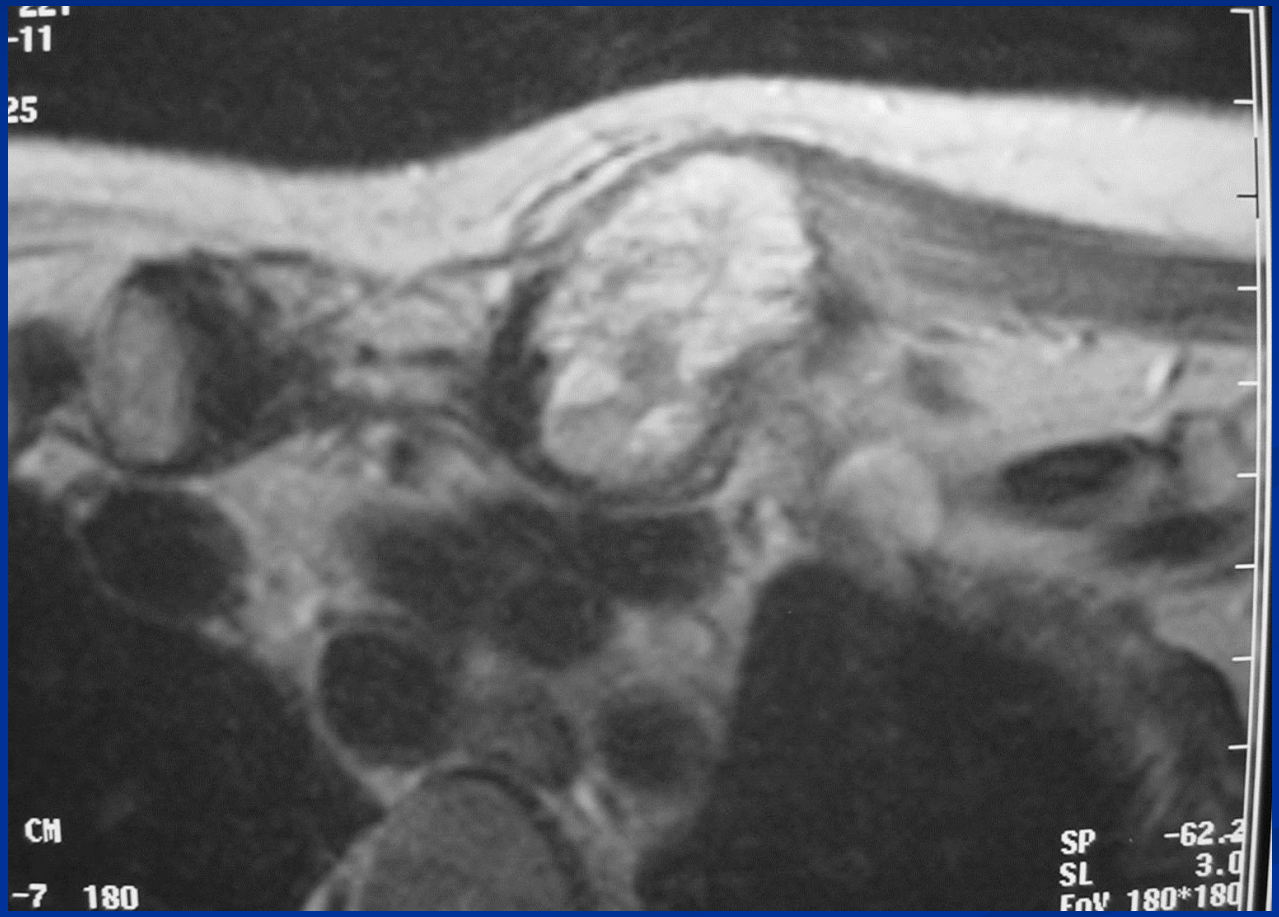


CT Scan: ABC of Medial End of Clavicle Geographic, Expansile, No Mineralization, Reactive Periosteum Surrounding Lesion




FOV 20.0cm
TND/P
R
1
5
L
6
8
L
R
kV 120
mA 150~
Smart mA 120
Large
3.0mm

MRI: Fluid-Fluid Levels



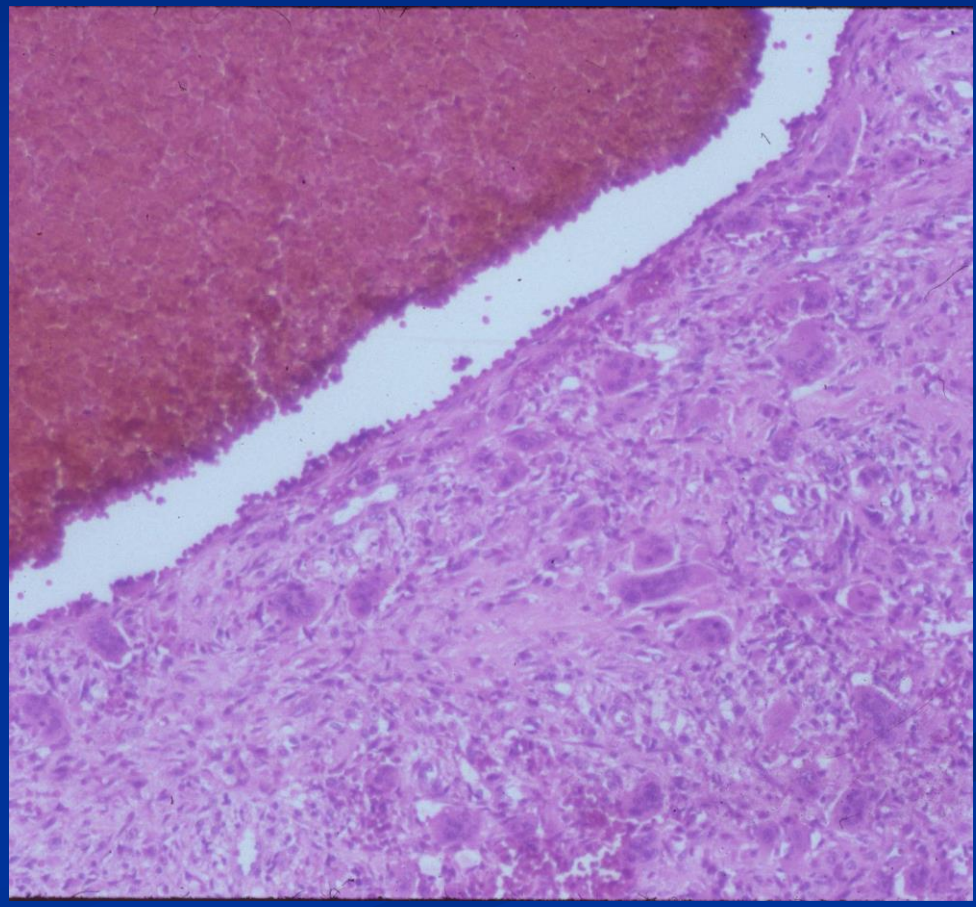
ABC Pathology

- **Pathology:**
  - Gross – "blood filled sponge"
  - Cavernous blood filled spaces lined by fibrous walls
  - Walls Contain Spindle Cells (fibroblast like cells), and fibrous tissue admixed with reactive Giant Cells
  - In an ABC, the cells in between the giant cells are spindled; the nuclei appear different than the nuclei within the giant cells
  - May see reactive osteoid in walls

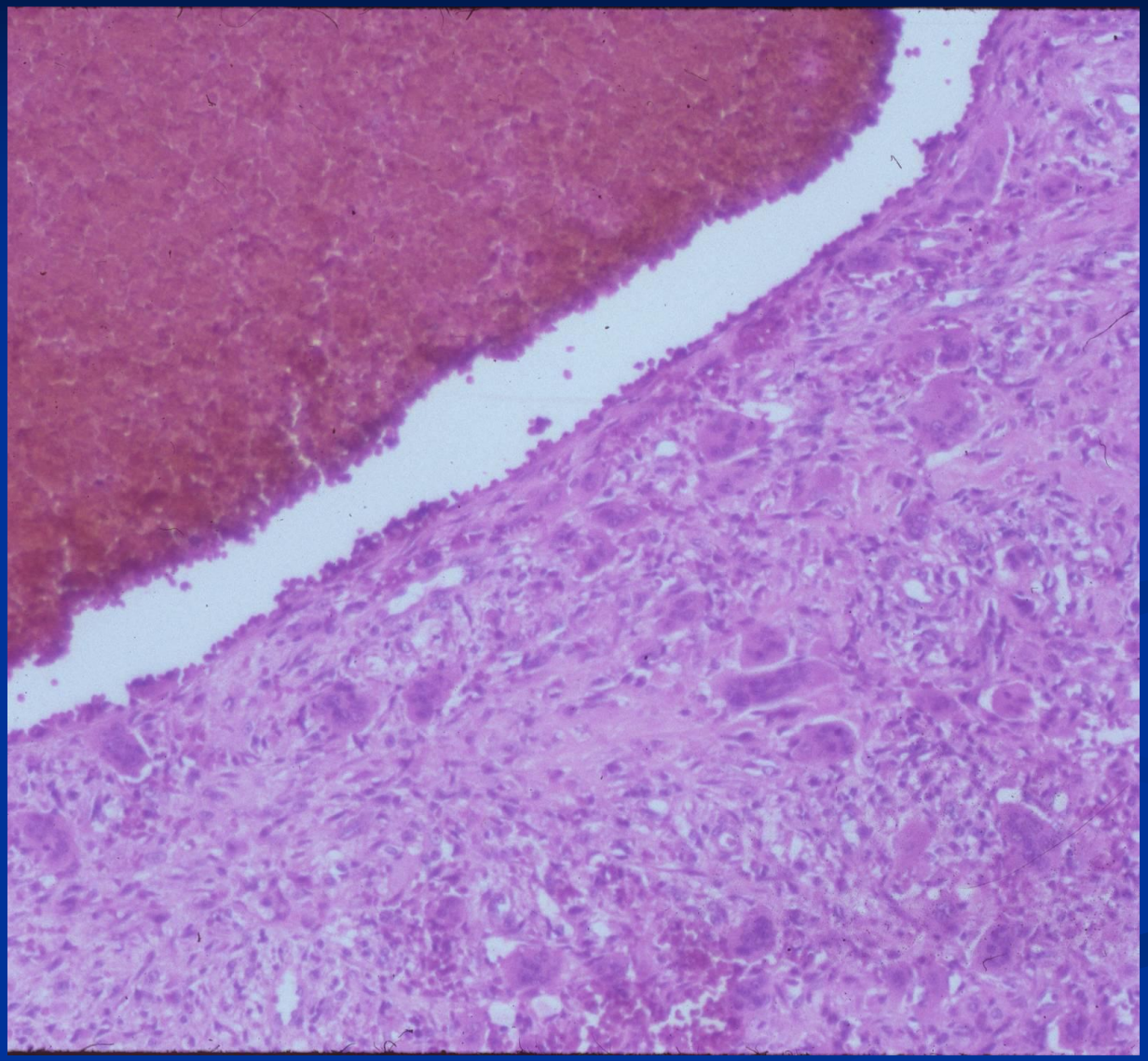
Gross Pathology: ABC of Medial End of Clavicle Expansile Lesion with Large Cystic/Cavernous Spaces and Thickened Septae

Microscopic Pathology ABC

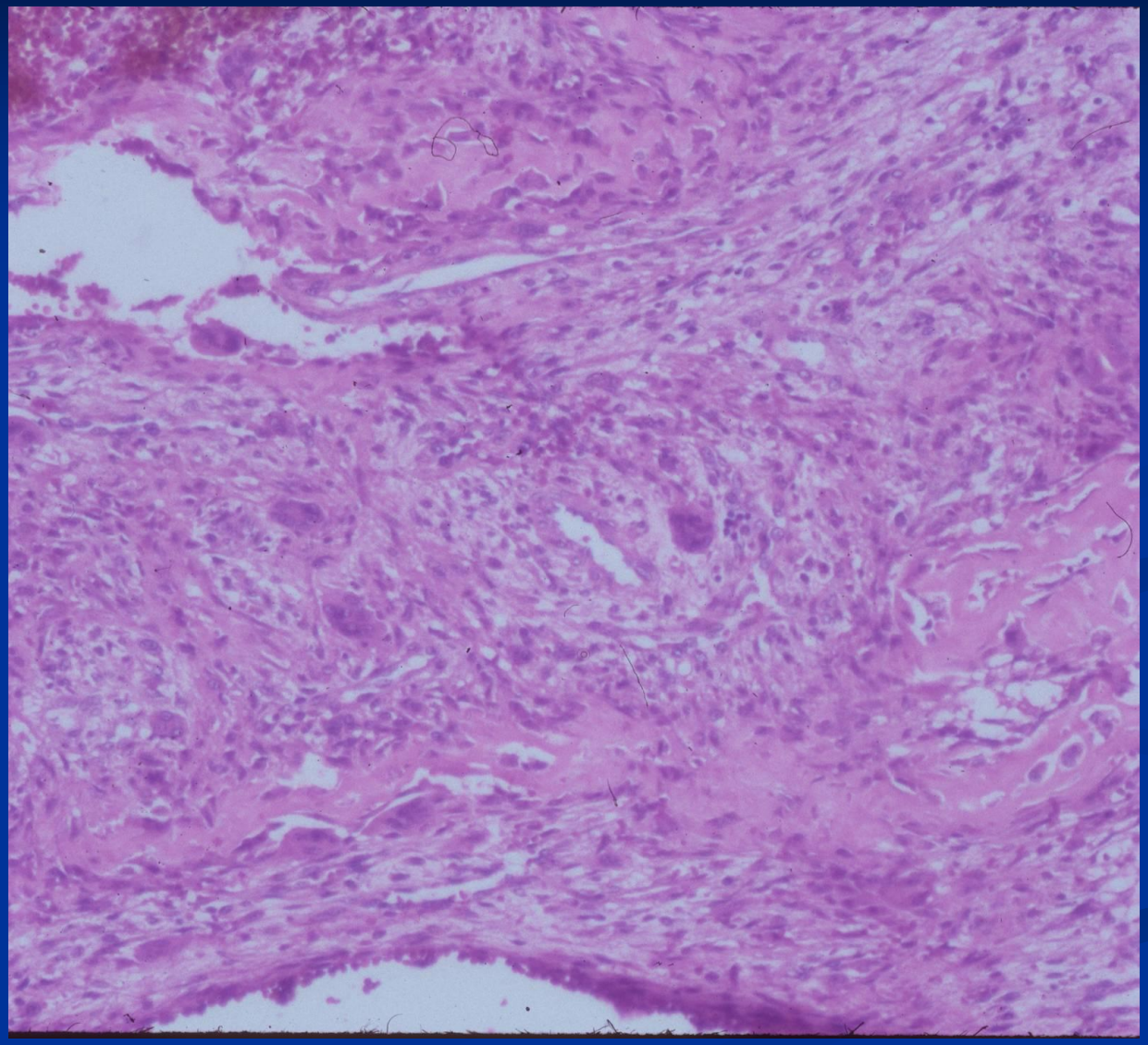
- Large Cystic, Blood Filled Cavernous Space
- No Epithelial Lining around Space
- Thick Wall with Fibrous Tissue, Spindle Cells and Giant Cells



Microscopic Pathology: ABC

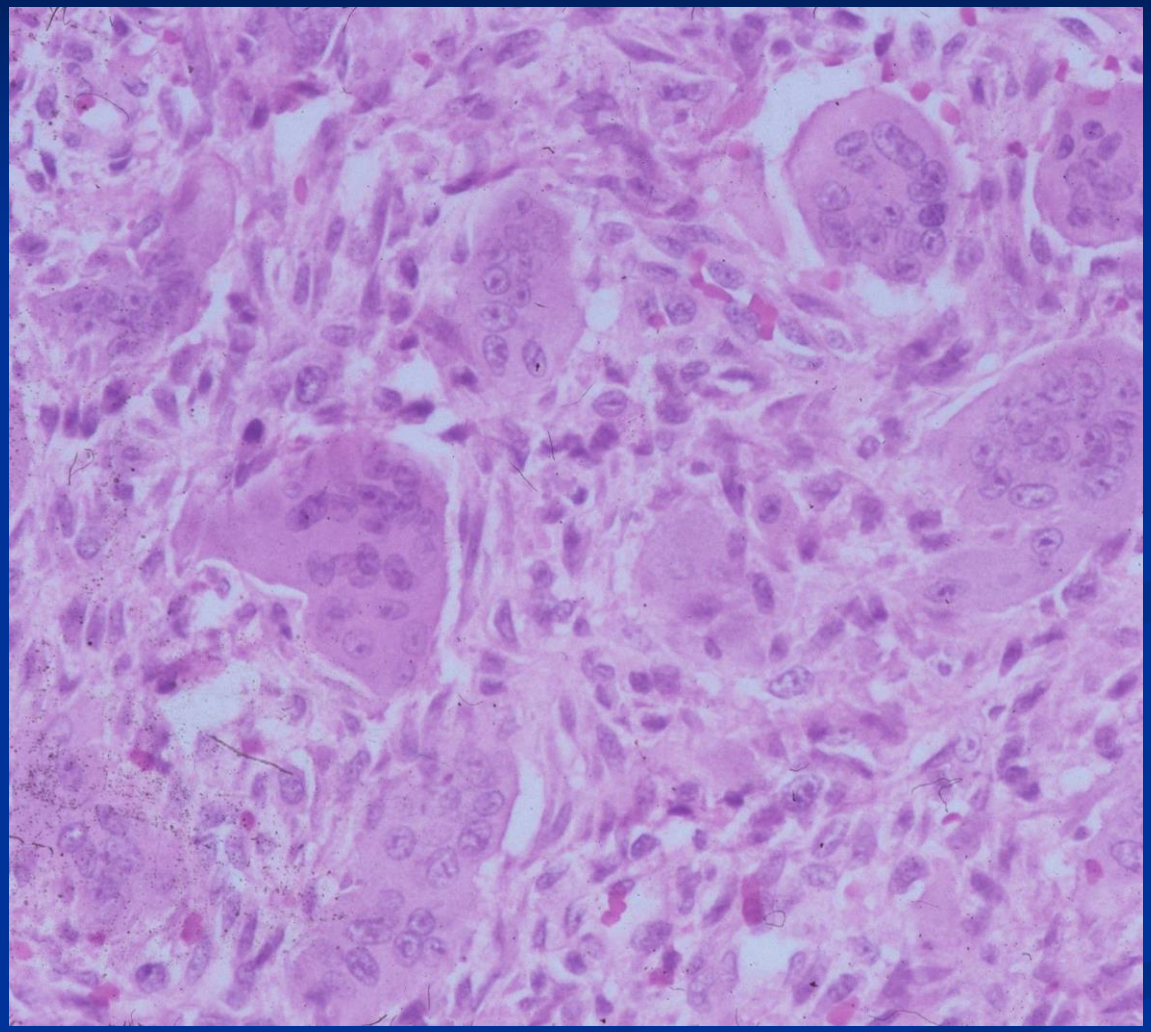


Microscopic Pathology: Wall of ABC



Micropscopic Pathology: Wall of ABC

Giant Cells admixed with Spindle Cells/Fibrous Tissue



Aneurysmal Bone Cyst (ABC)

- Treatment and prognosis:
  - Rarely spontaneous regression
  - Intralesional Curettage and Bone Grafting
  - Enbloc resection for lesions that have destroyed the entire bone
  - Recurrence 10-20%

Unicameral Bone Cyst (UBC)
Simple Bone Cyst

- Definition: A fluid containing lesion lined by thin fibroconnective tissue membrane usually arising central in metaphysis of long bone adjacent to physis
- 3% of all biopsied primary osseous neoplasms
- Young patients < 20 yr. old (85%)
- Male>Female, 3:1
- Pathologic fracture 50%
- Etiology: Lymphatic/Venous Obstruction vs Synovial Origin

Unicameral/Simple Bone Cyst

- Location:
  - Proximal Humerus
  - Proximal Femur
  - Proximal Tibia
  - Pelvis, Calcaneus—More common over 20 years of age
- Clinical:
  - Incidental finding
  - Mild pain, swelling, stiffness in adjacent joint
  - Sudden pain secondary to pathological fracture

Unicameral/Simple Bone Cyst

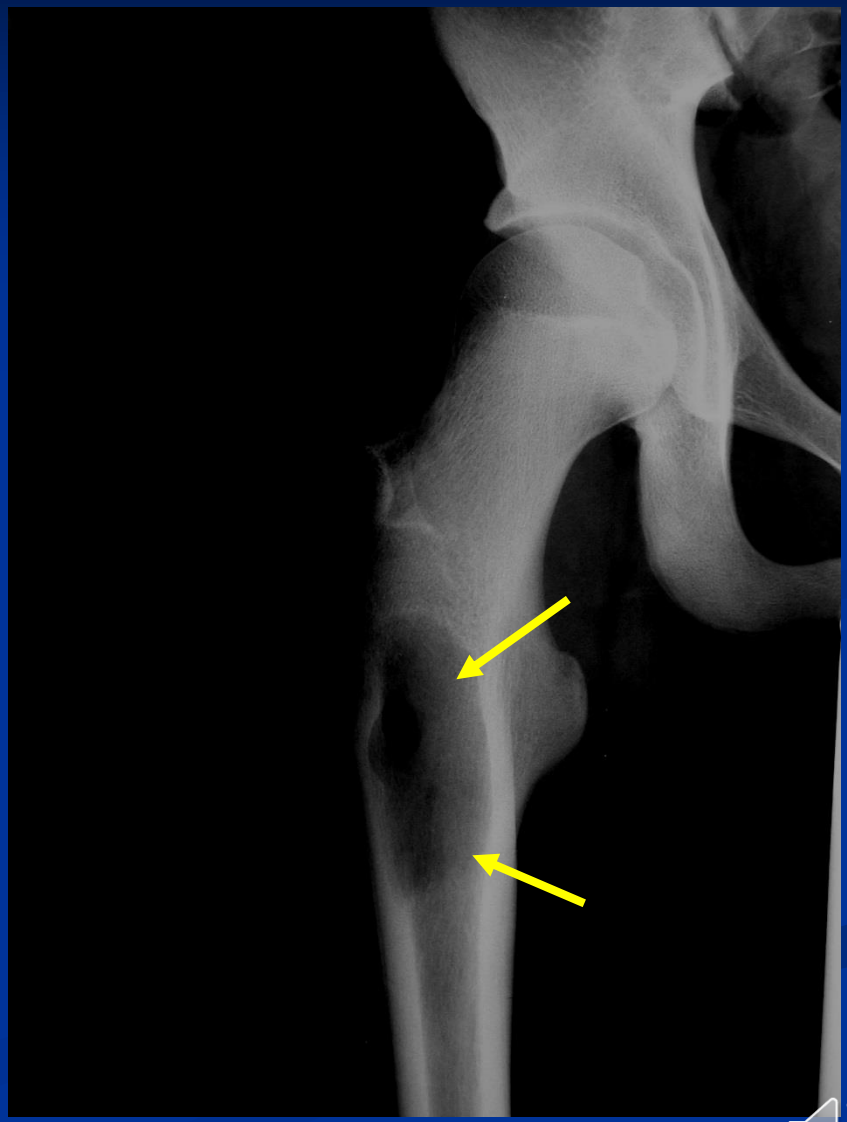
- **Radiology:**
  - Geographic lesion – central metaphyseal (active) adjacent to physis
  - Thin rim of surrounding sclerosis
  - Can be diaphyseal (latent); lesion migrates distally with growth
  - Mild expansile remodeling (typically, not as expansile as an ABC) Overlying cortex is thin but never penetrated
  - May be **multilocular/trabeculated** due to osseous ridges on the inner wall of the cyst

Unicameral/Simple Bone Cyst

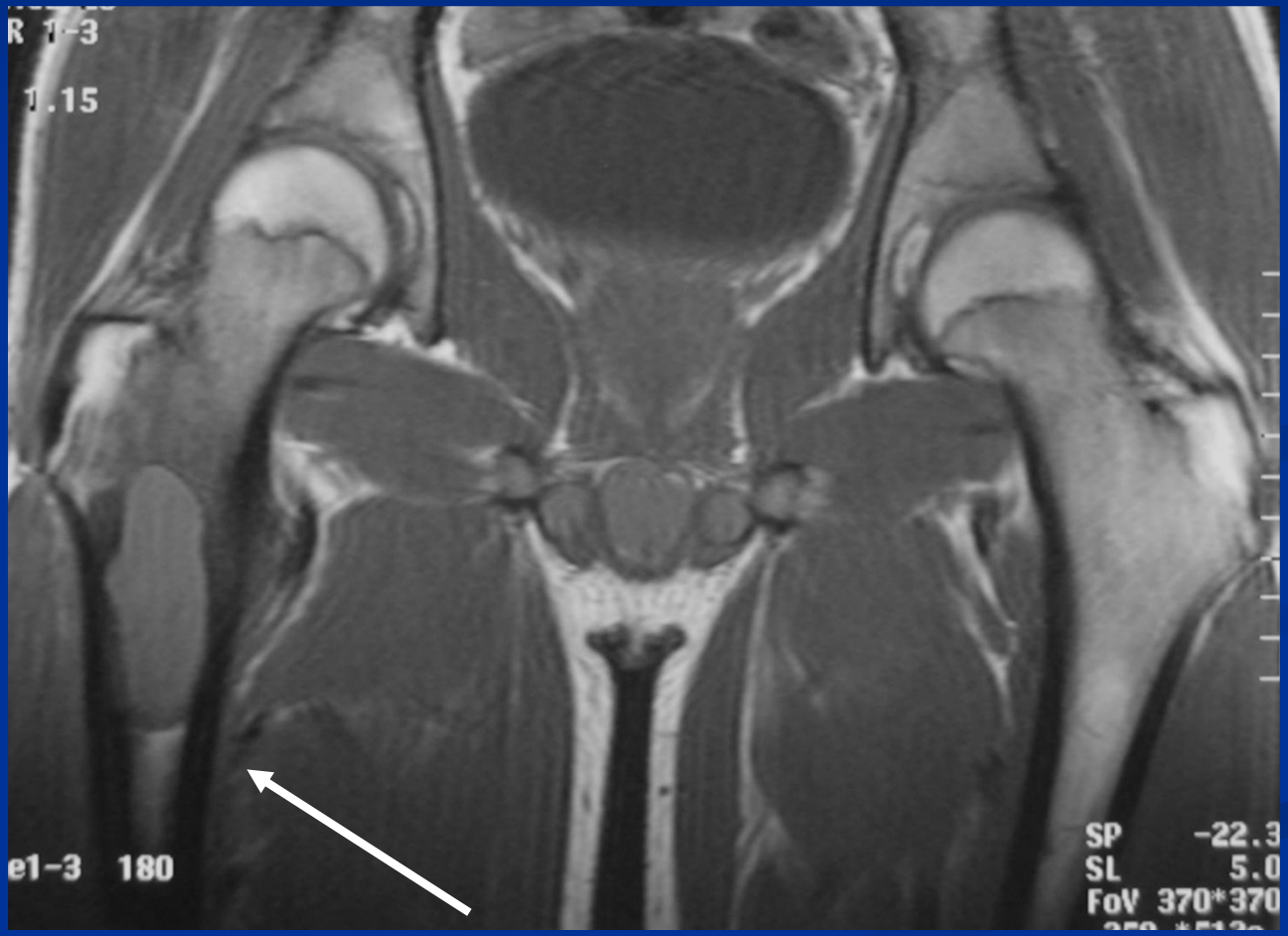
- **Radiology:**
  - Pathologic fracture – "fallen fragment" sign
  - May contain calcified granules/reactive osteoid/new bone formation
  - CT/MR- simple fluid in noncomplicated case
  - CT/MR- complicated case
    - Soft tissue with unusual thick membrane
    - Fluid-fluid/gas-fluid levels: possible with Pathological Fracture

X-Ray: UBC of Proximal Femur

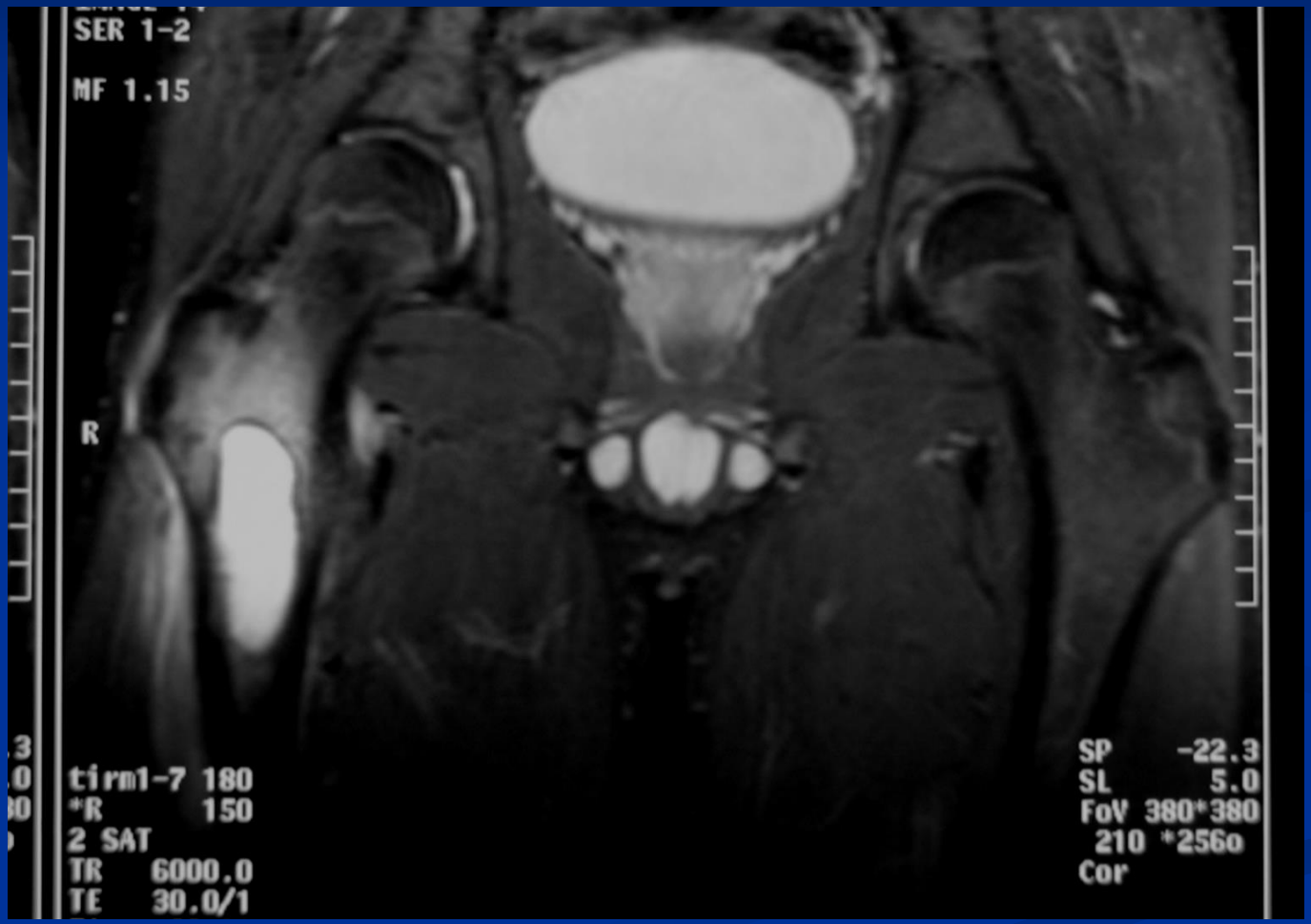
- Central lesion
- Geographic
- Thin rim of sclerosis
- Mildly expansile
- No mineralization



MRI T1: Homogeneous Lesion Low to Intermediate Signal

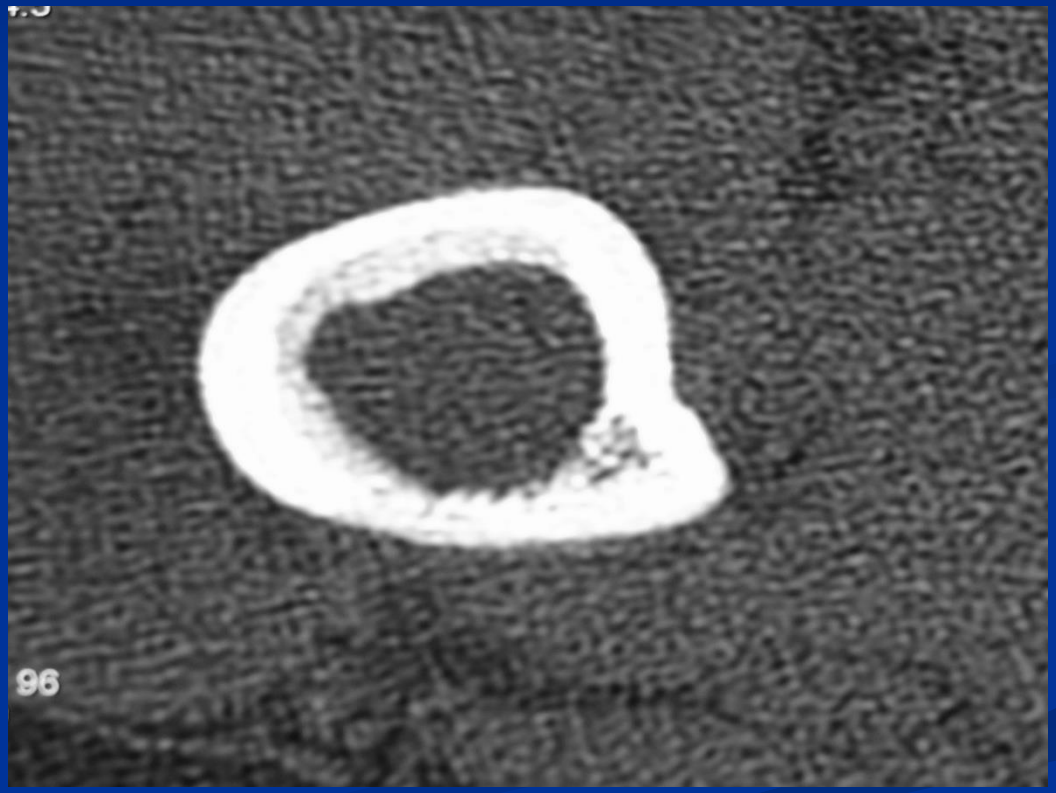


MRI T2: UBC Proximal Femur Homogeneous Fluid-Filled Lesion



CT Scan: UBC of Proximal Femur
Well Circumscribed, Thin Rim of Sclerosis, No Mineralization

- Well Circumscribed
- Thin Rim of Surrounding Sclerosis
- No Mineralization
- Fluid Attenuation within Lesion

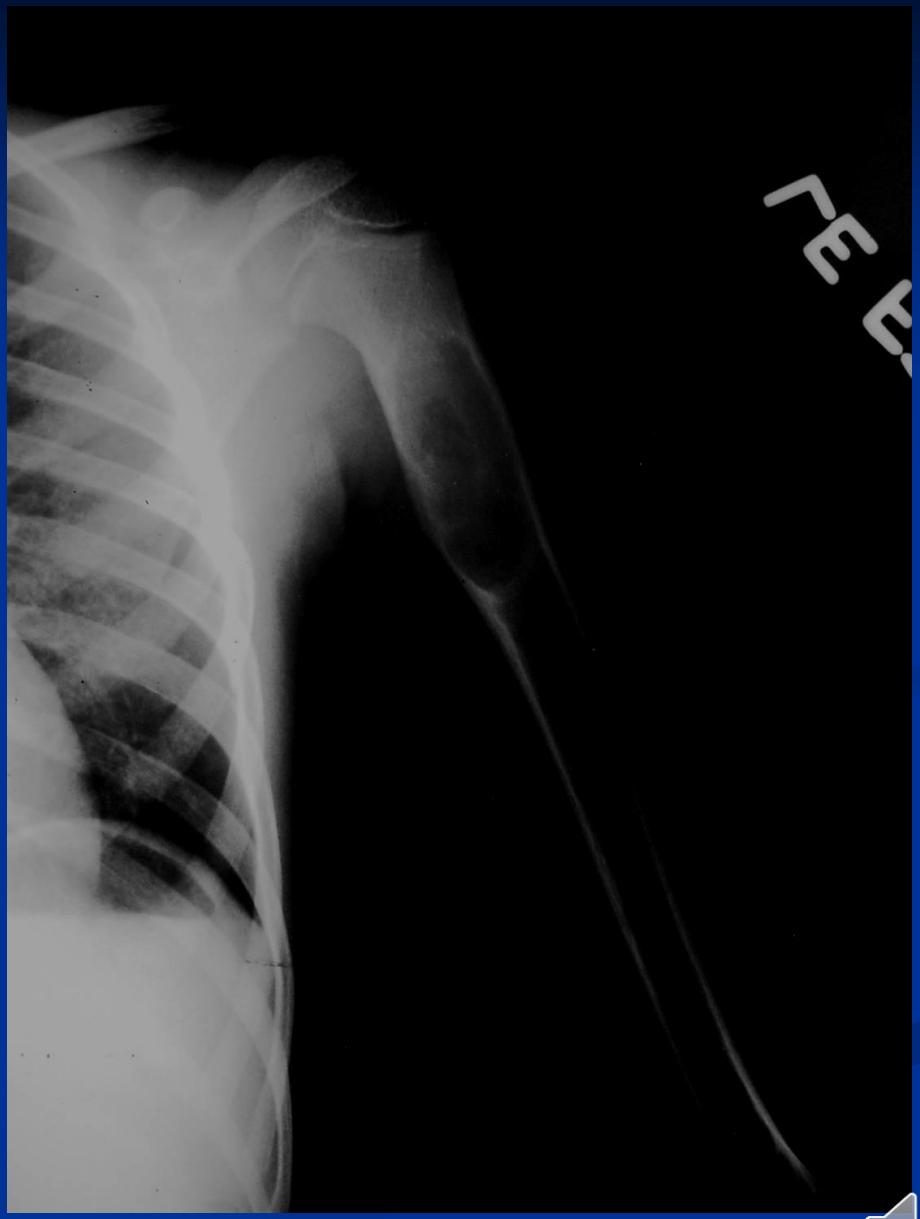


CT: UBC Proximal Femur

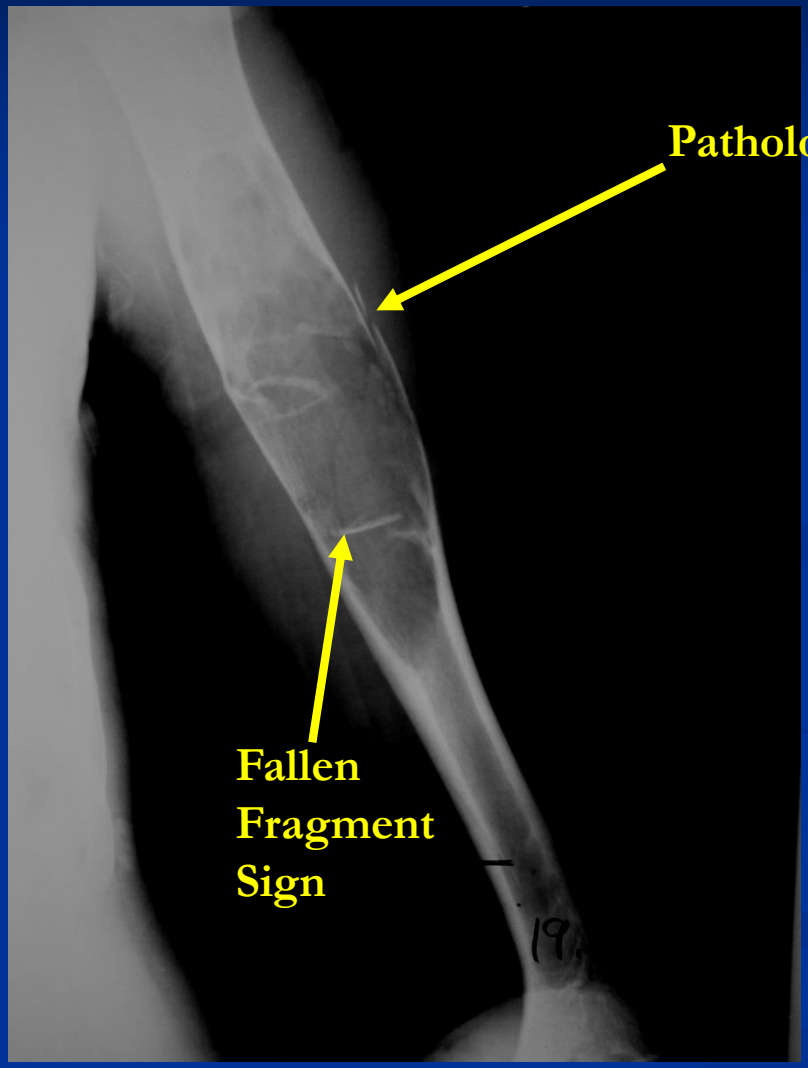
Xray: UBC Proximal Femur

X-Ray: UBC Proximal Humerus

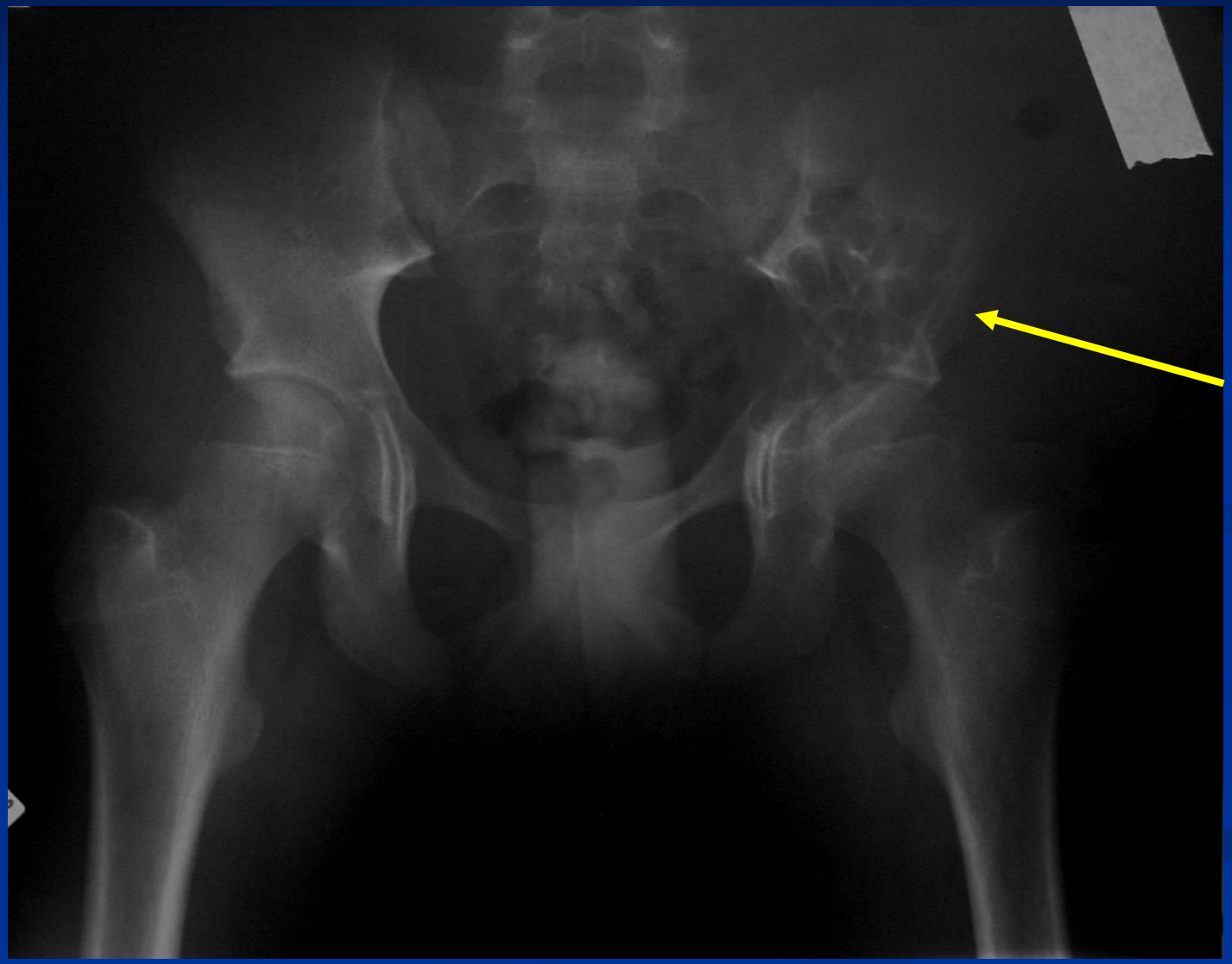
- Central Lesion
- Geographic
- Mildly Expansile
- No Mineralization



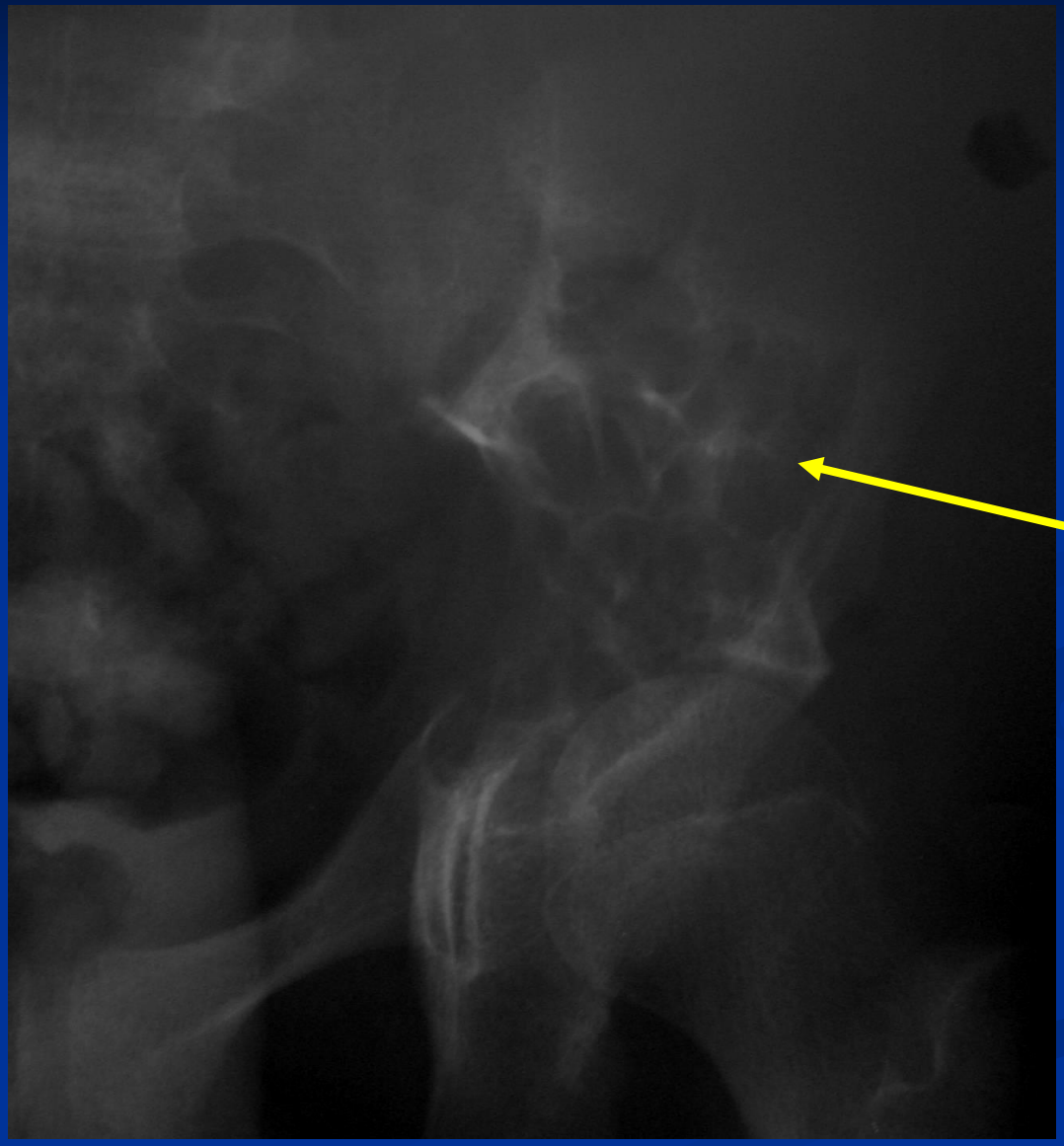
Xray: UBC of Humerus
Pathological Fracture and Fallen Fragment Sign
Central, Mildly Expansile, Radiolucent Lesion



Xray: Multiloculated UBC of Pelvis (Ilium)

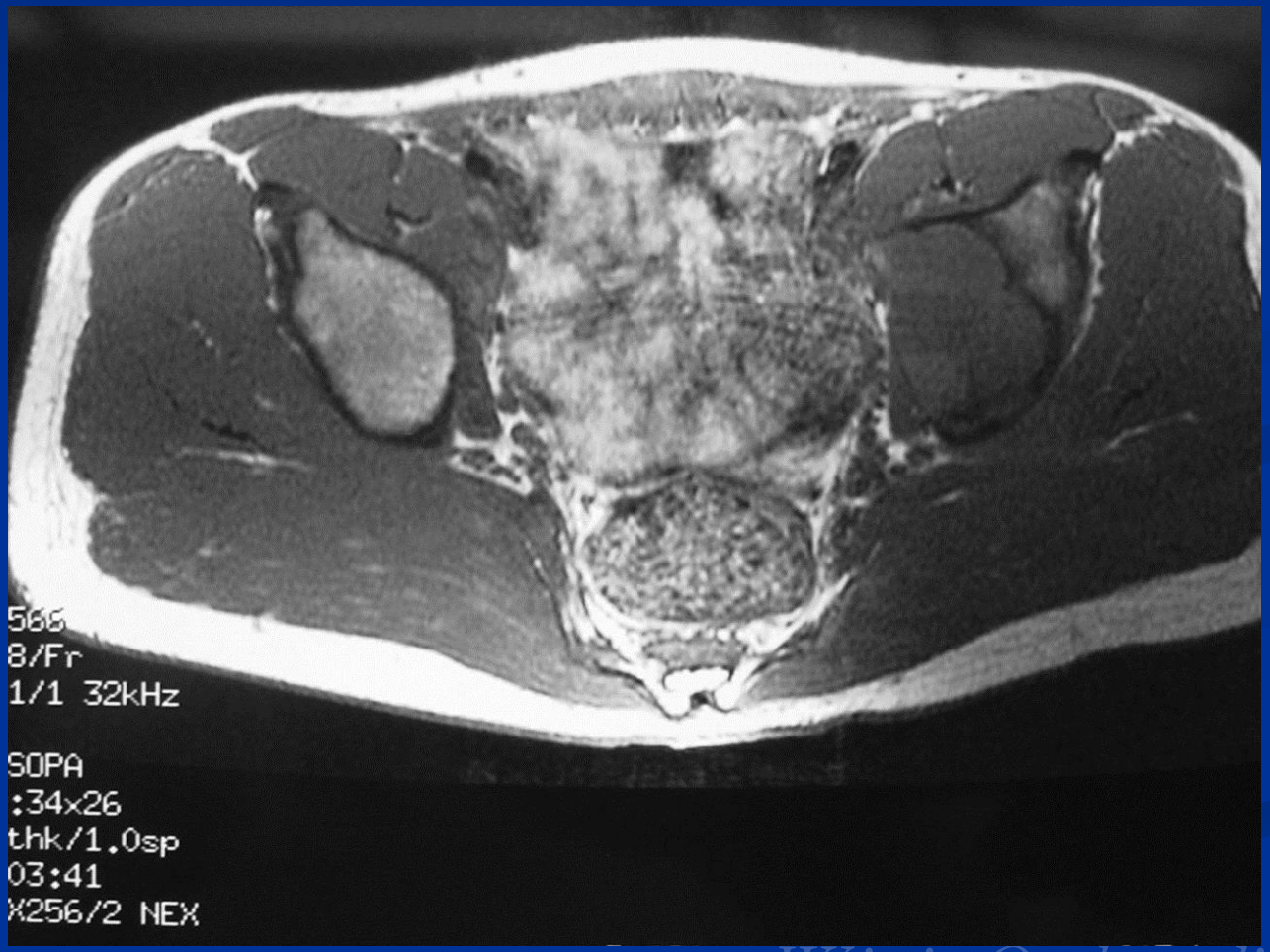


Xray: Multiloculated UBC of Pelvis



CT Scan of Multiloculated UBC of Pelvis

MRI T1: UBC of Pelvis Homogeneous Fluid Signal (Intermediate SI)



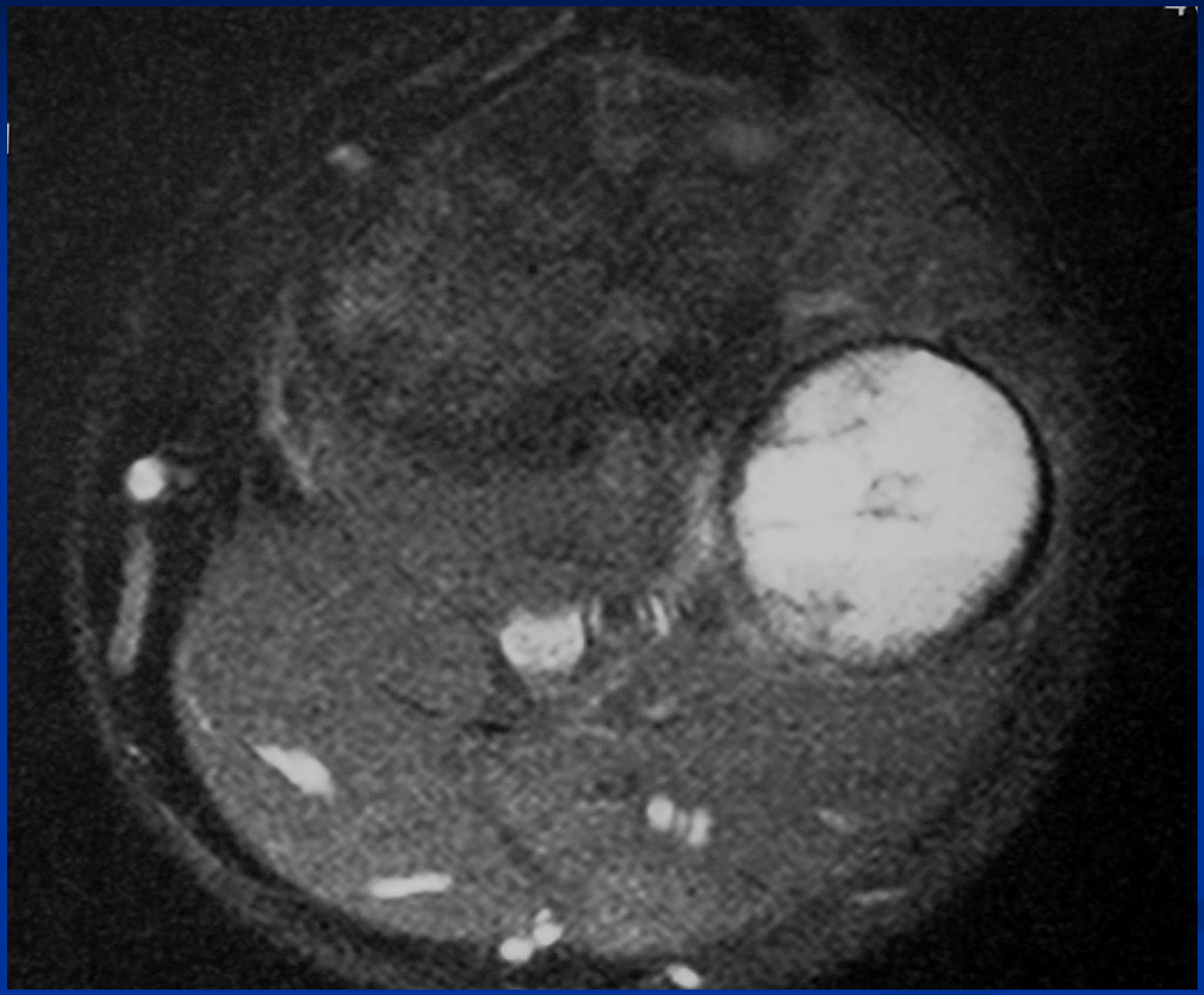
Xray of Proximal Fibula UBC with Unusual Reactive Bone Formation

CT Scan of Proximal Fibula UBC with Unusual Reactive Bone Formation

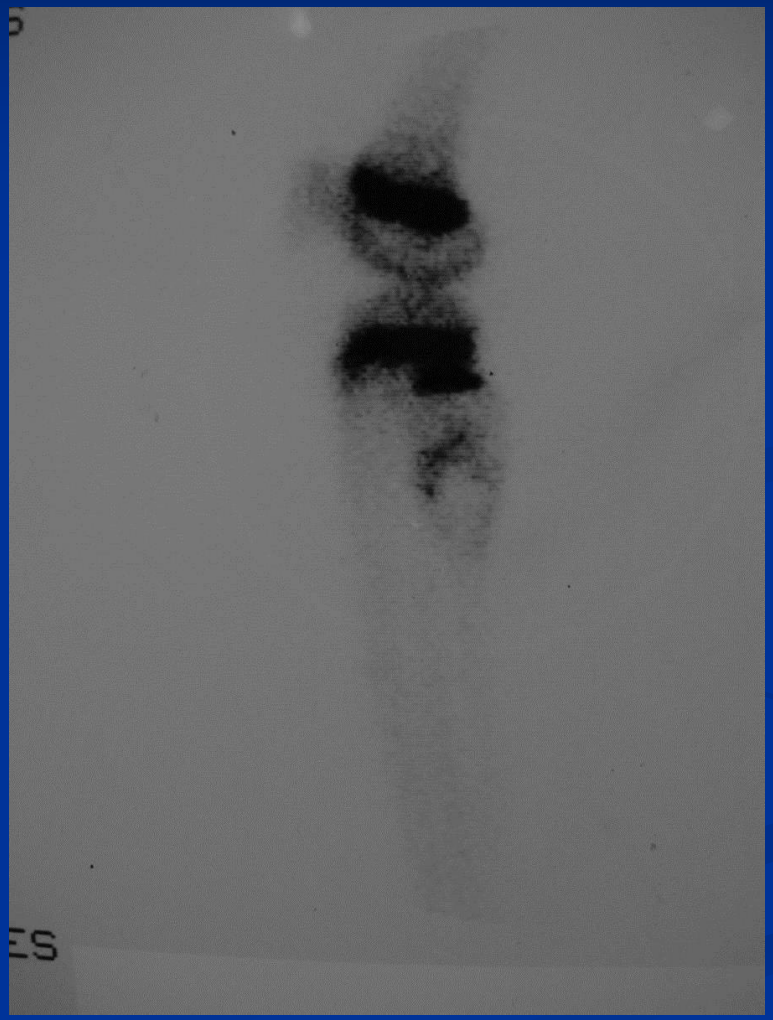
MRI T1: Proximal Fibula UBC

MRI T2: Proximal Fibula UBC Homogeneous Fluid Signal

MRI T2 Axial: Proximal Fibula UBC



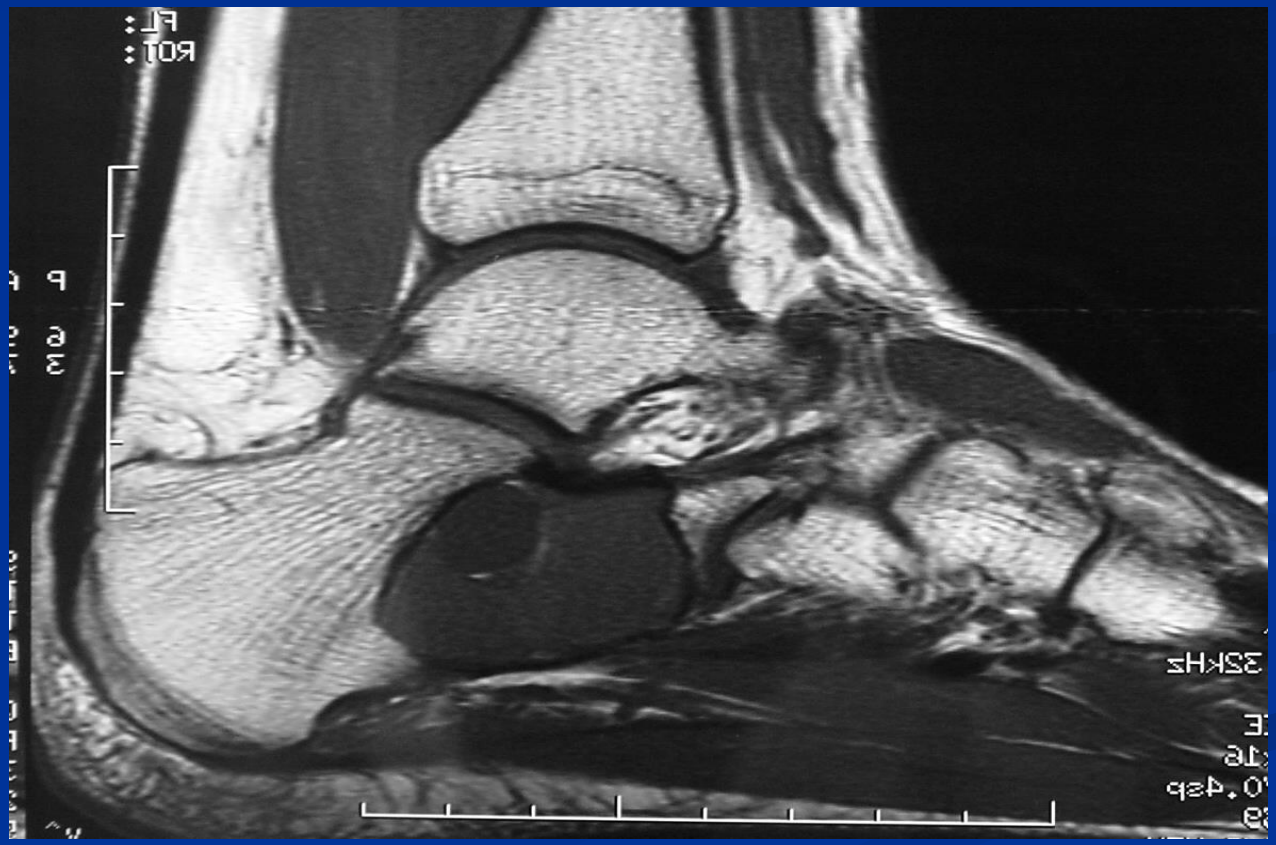
Bone Scan: Proximal Fibula UBC No/Minimal Uptake



Xray: UBC Calcaneus

Xray: UBC of Calcaneus

MRI T1: UBC Calcaneus Homogeneous Fluid Signal

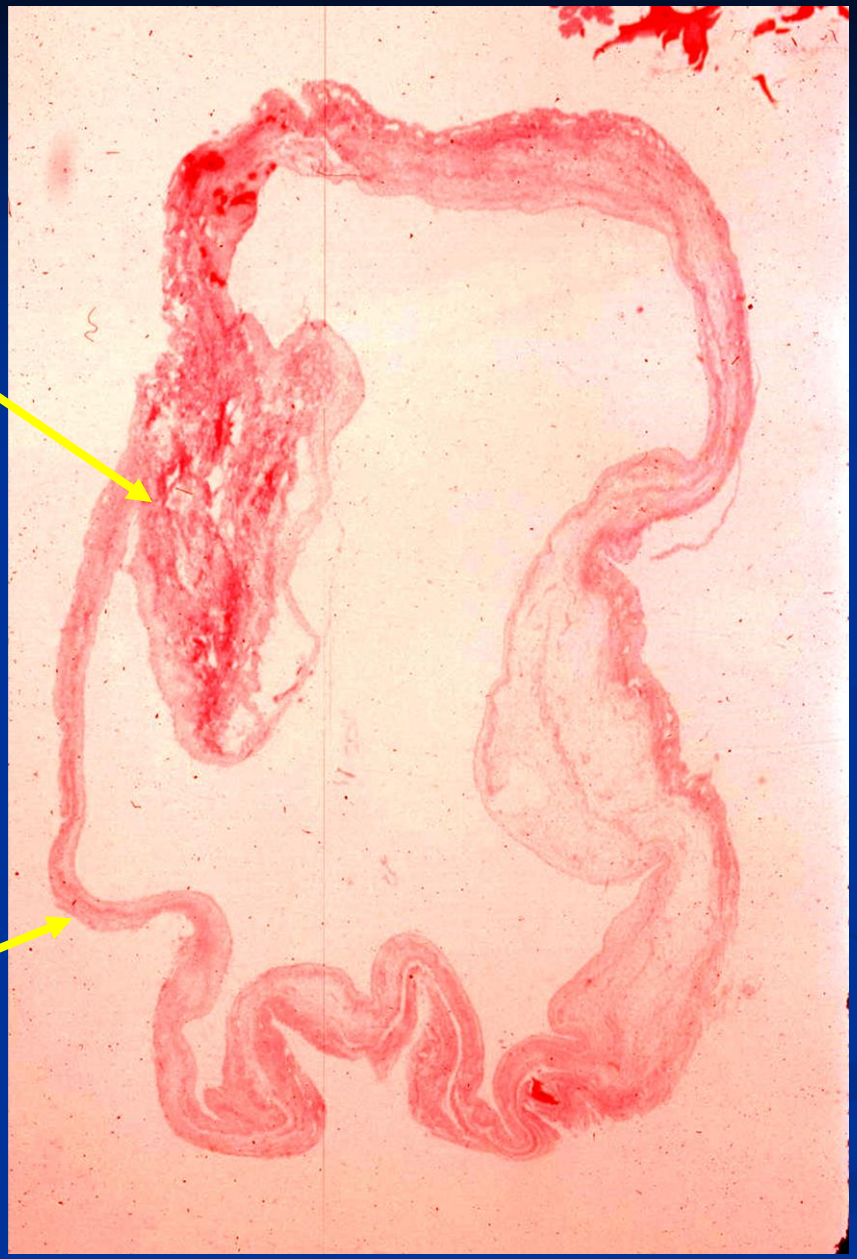


MRI T2: UBC Calcaneus
Homogeneous Fluid Signal

Unicameral/Simple Bone Cyst

- Pathology:
  - Clear, straw-colored fluid filled cyst (serous/serosanguinous fluid)
  - Thin fibroconnective tissue lining (1mm thick)
  - Thicker walls may contain small arteries and veins
  - May be new bone formation even without fracture
  - May contain spherical calcified structures in loose fibrous stroma (calcospherites)

May Have Areas with Blood Vessels and Hemorrhage



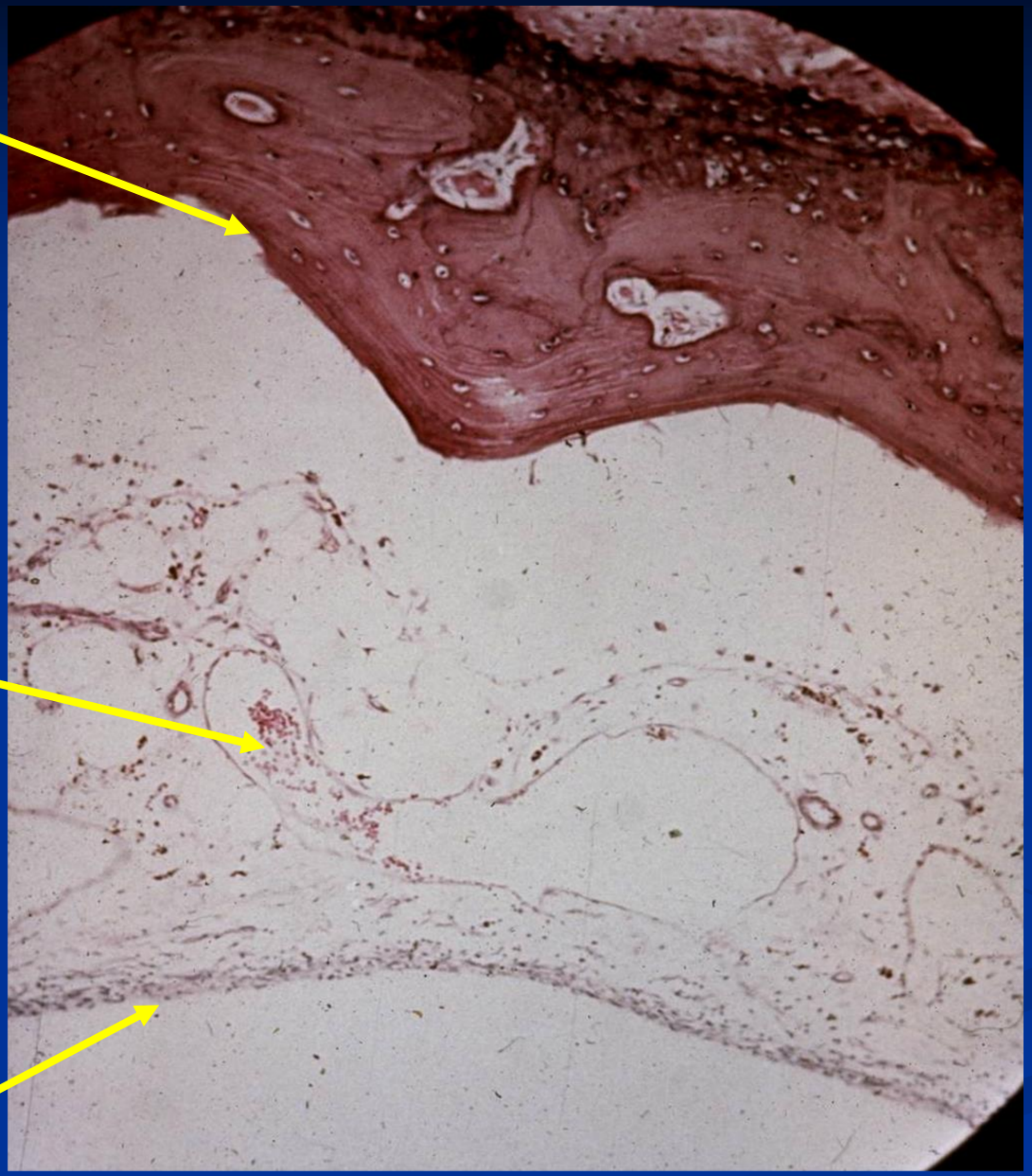
Very Thin/Scant Lining of Cystic Cavity

May Have
Reactive Bone in
Lining

Thin Scant
Mesothelial
Lining

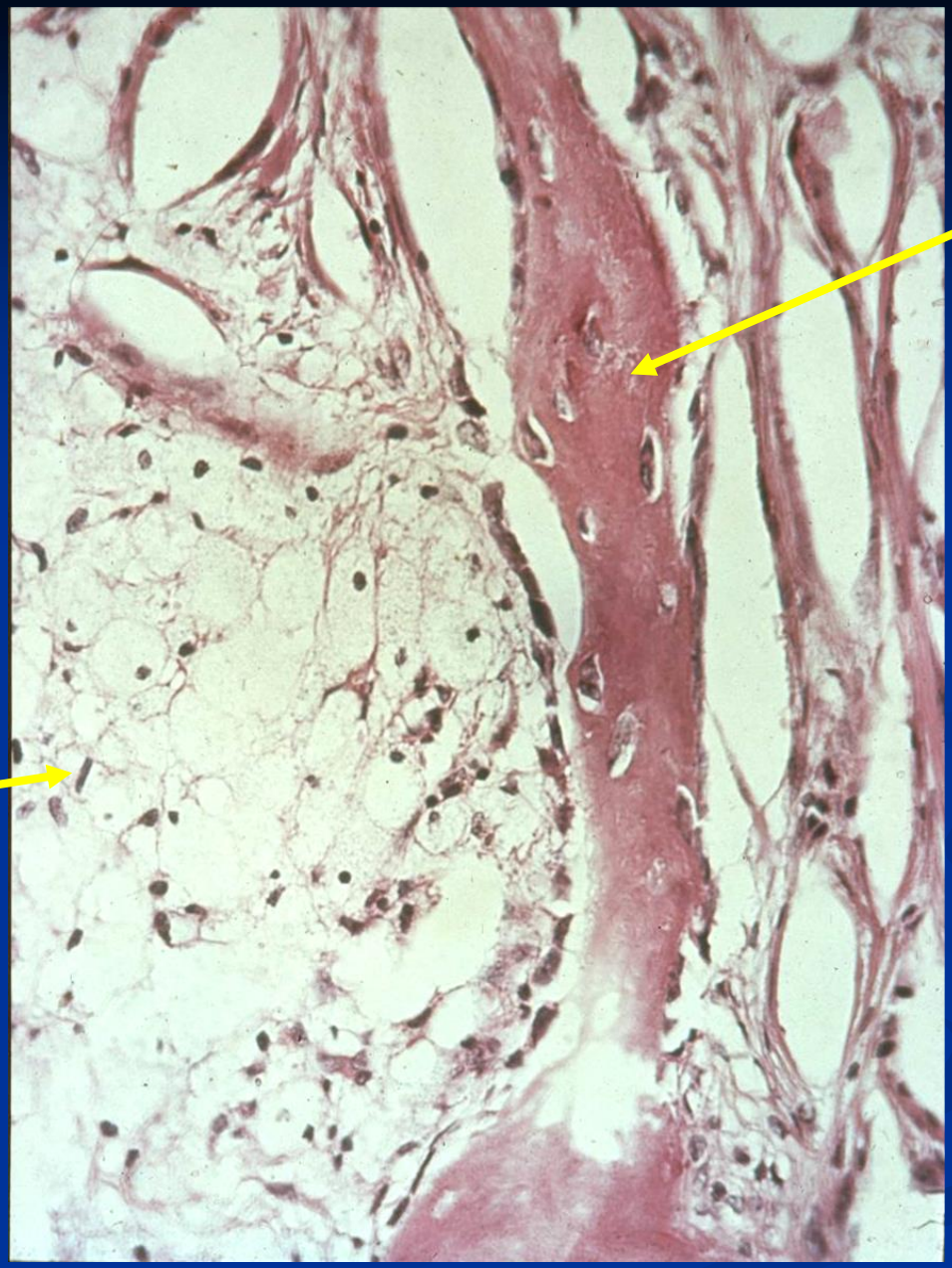
Normal
Bone

Normal Cortical Bone



Vascular Spaces

Thin Lining

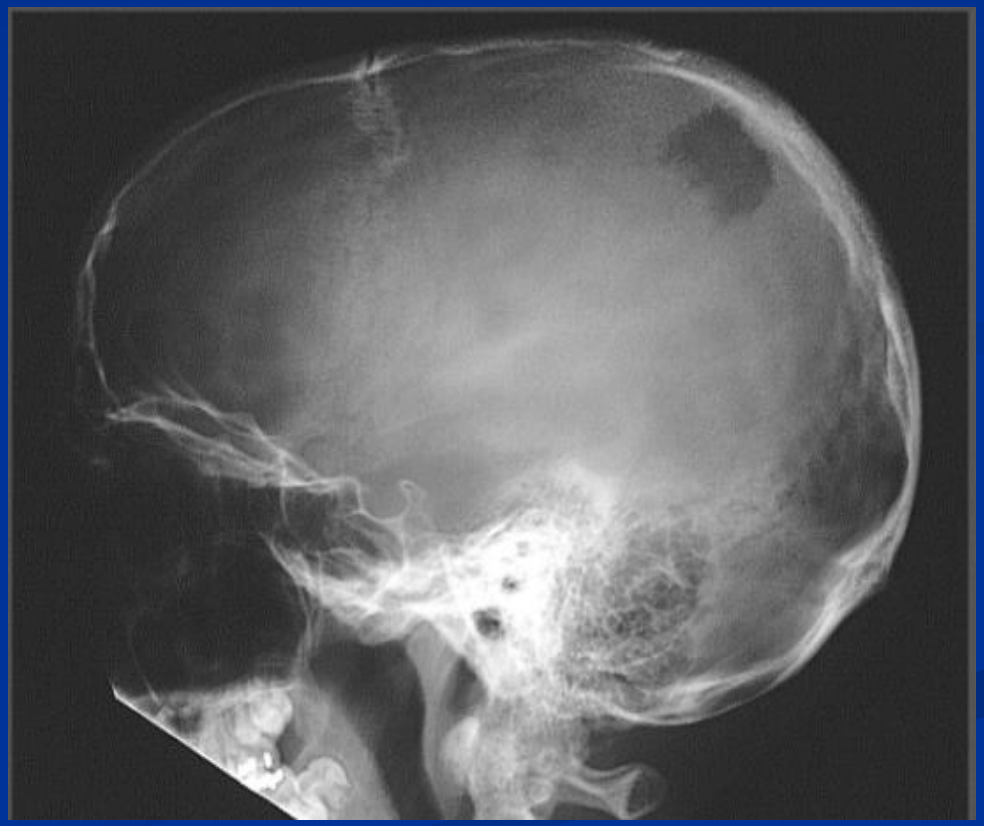


Unicameral/Simple Bone Cyst

- **Treatment and Course**
  - Spontaneous regression or heal after fracture (rare)
  - Curettage and bone grafting (20% recurrence rate)
  - Intralesional steroids (70-95% effective)
  - Extremely rare – malignant transformation (secondary sarcoma)

Langerhans Cell Histiocytosis/ Eosinophilic Granuloma

- Spectrum of Diseases
- Localized form (EG) accounts for 70% of cases
- Least aggressive form
- Age: 5-15 years
- 95% Caucasian
- Pain, tenderness, mass, systemic sx mimicking infxn
- 10% go on to develop multifocal disease



Hand-Schuller-Christian Disease

- **Triad:**
- Destructive skeletal lesions
- Exophthalmos
- Diabetes Insipidus
- 10% of patients with unifocal EG develop multifocal and extraskeletal disease
- Usually <5 years old
- Hepatosplenomegaly, adenopathy, anemia, fever, neurological complaints
- Fatal in 15%
- Any bone but 90% have skull involvement

Letterer-Siwe Disease

- Develops in 1st year of life
- Disseminated disease and small bone lesions
- Fatal in 95% who develop before 1 year of life

Eosinophilic Granuloma

- **Sites:**
  - Flat Bones (most common—70%)
    - Skull, Mandible, Ribs
    - Pelvis
  - Femur
  - Humerus
  - Tibia
  - Hands and Feet are rare in solitary disease

Eosinophilic Granuloma

- Age: 5-15 years (85% of patients less than 30 years; 60 % less than age 10)
- 95% are caucasian
- Pain, tenderness and fever
- Mild peripheral eosinophilia in 5-10% of patients

Eosinophilic Granuloma

- **Radiology:**
- Variable radiological appearance (Benign to Malignant)
  - Geographic or Permeative
  - Onion Skin/Lamellated Periosteal Reaction
- Any bone and any portion of the bone (epiphyseal, metaphyseal, diaphyseal)
- Lytic, Radiolucent Lesion, No Mineralization
- May have rind of sclerosis
- 5-10% of patients have an associated soft tissue mass
- Sequestrum (button-like); Hole in a Hole appearance

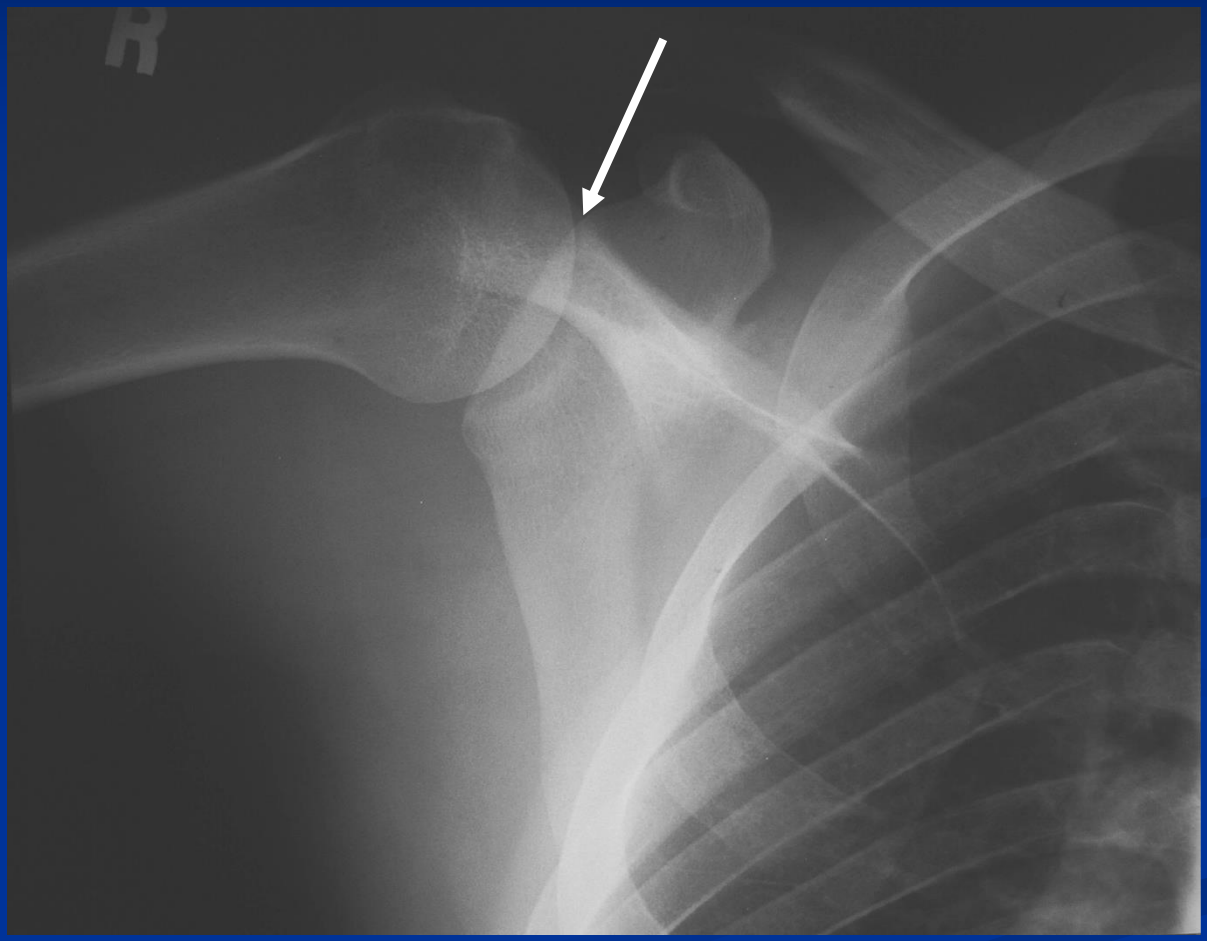
Eosinophilic Granuloma

- Skull: Beveled Edge; Button Sequestrum
- Flat Bone: Hole in a Hole
- Spine: Vertebra Plana

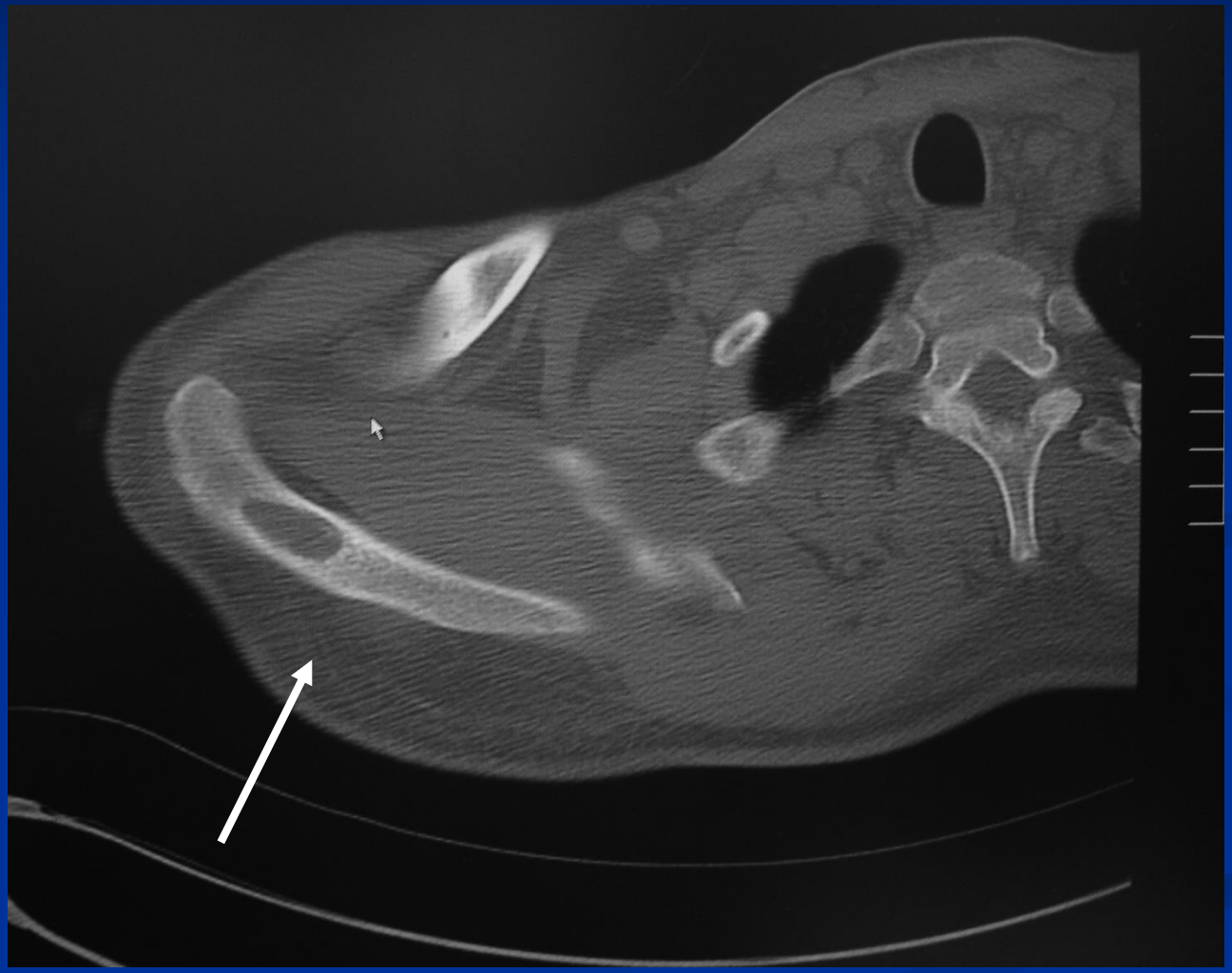
X-Ray: EG of Skull

X-Ray: EG of Spine (Vertebra Plana)

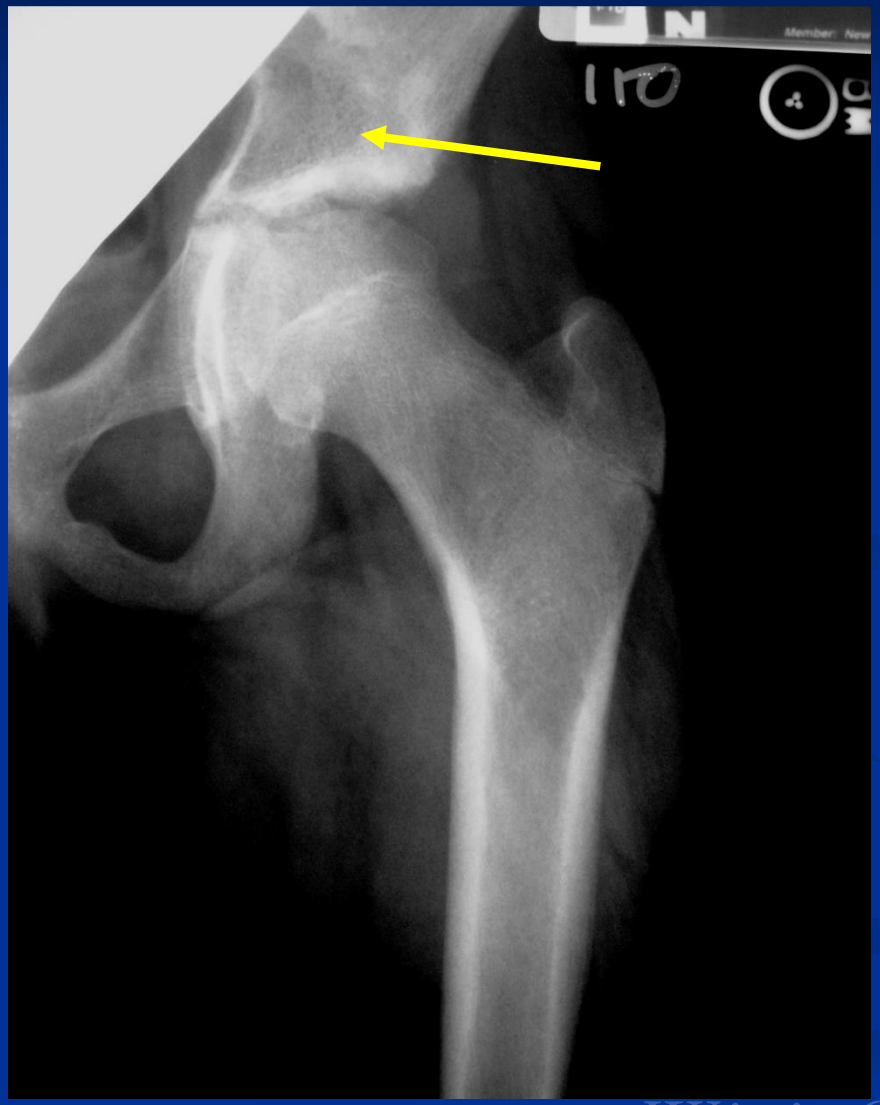
Xray Scapula: EG of Scapula Spine



CT Scan: EG of Scapula Spine

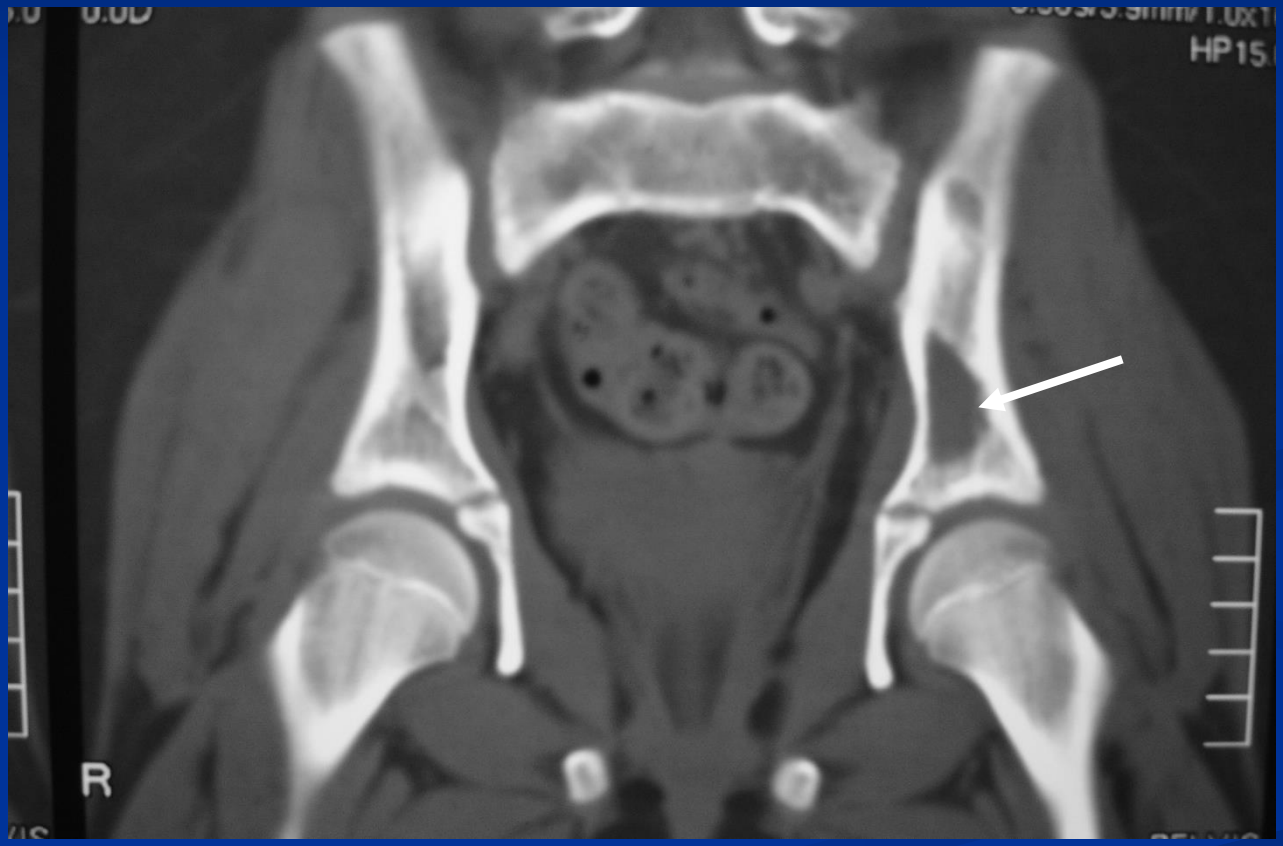


Xray of Hip: EG of Left Acetabulum

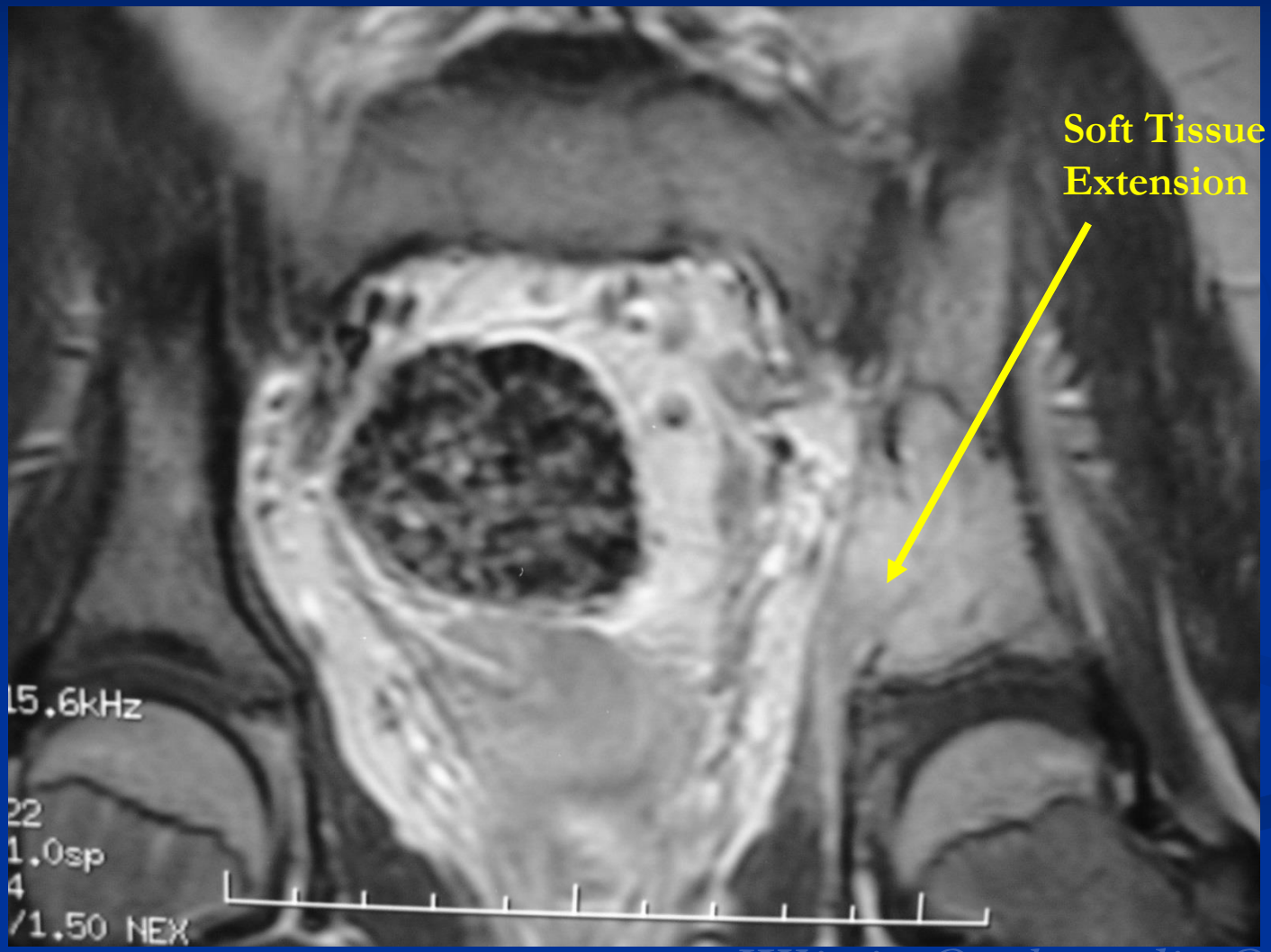


Xray of Hip: EG of Acetabulum

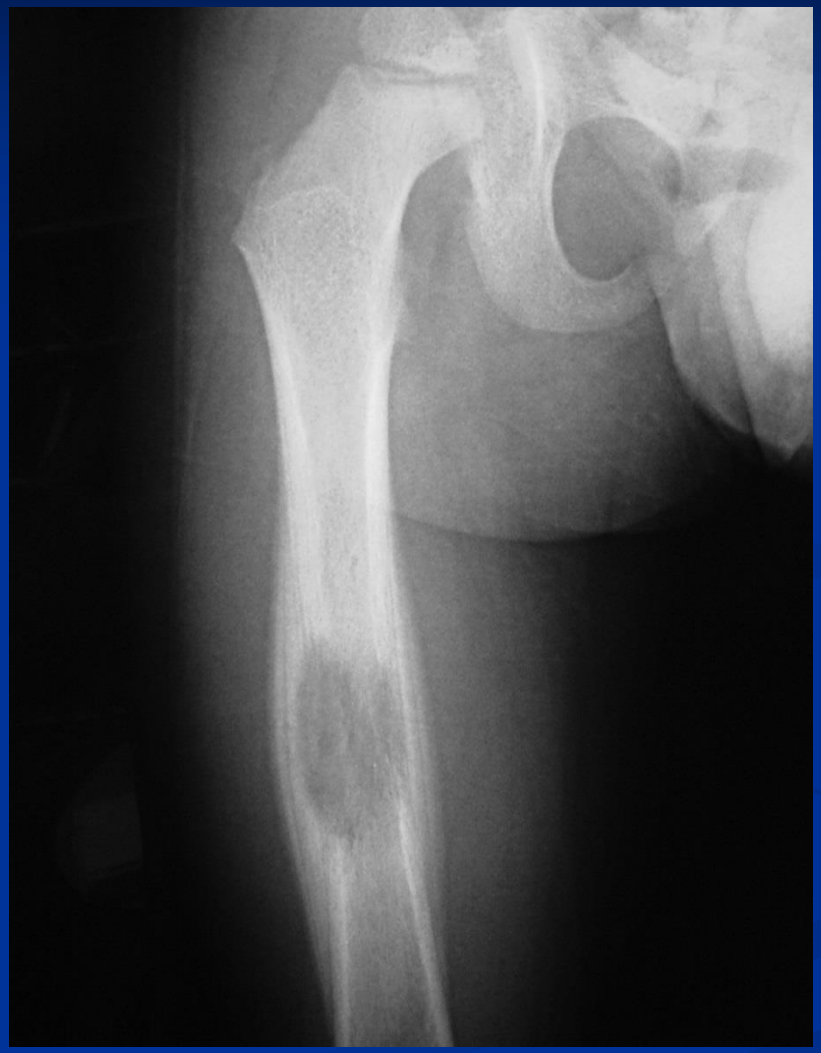
CT Scan: EG of Left Acetabulum



MRI: EG of Left Acetabulum Soft Tissue Extension/Mass

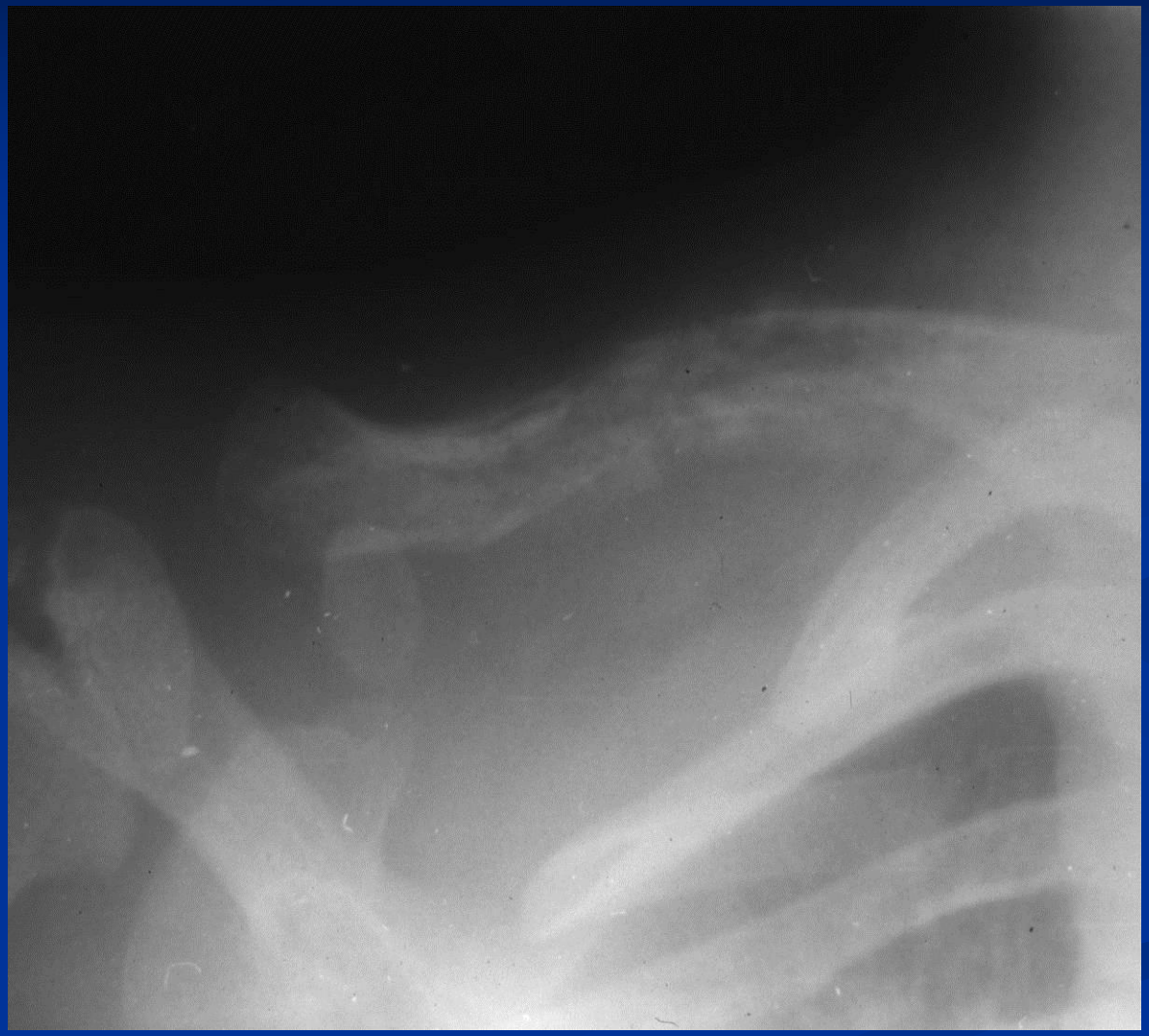


Xray of Femur: EG



Xray of Femur: EG

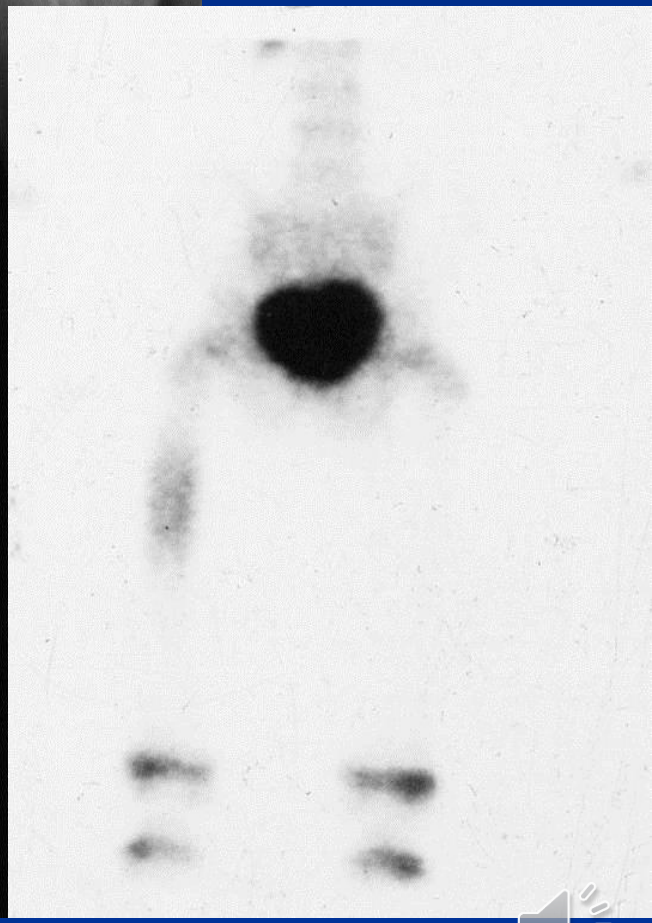
Xray of Clavicle: EG



Xray of Humerus: EG

Bone Scan: EG

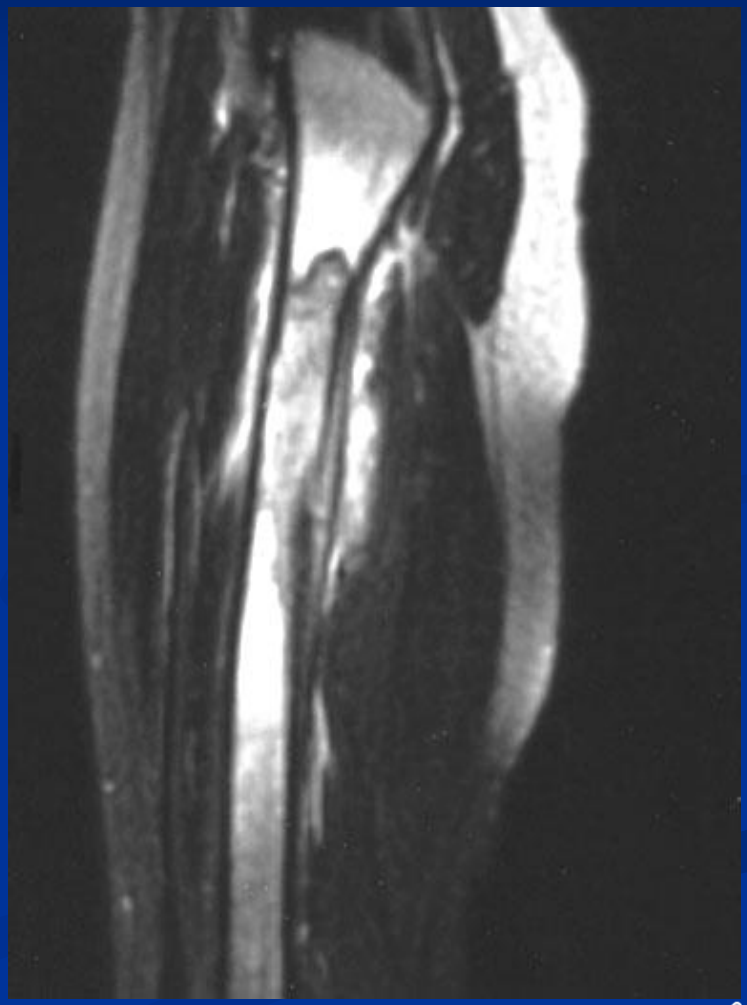
- Increased activity on BS, but activity may be less than expected from XR appearance, and may even produce a cold defect



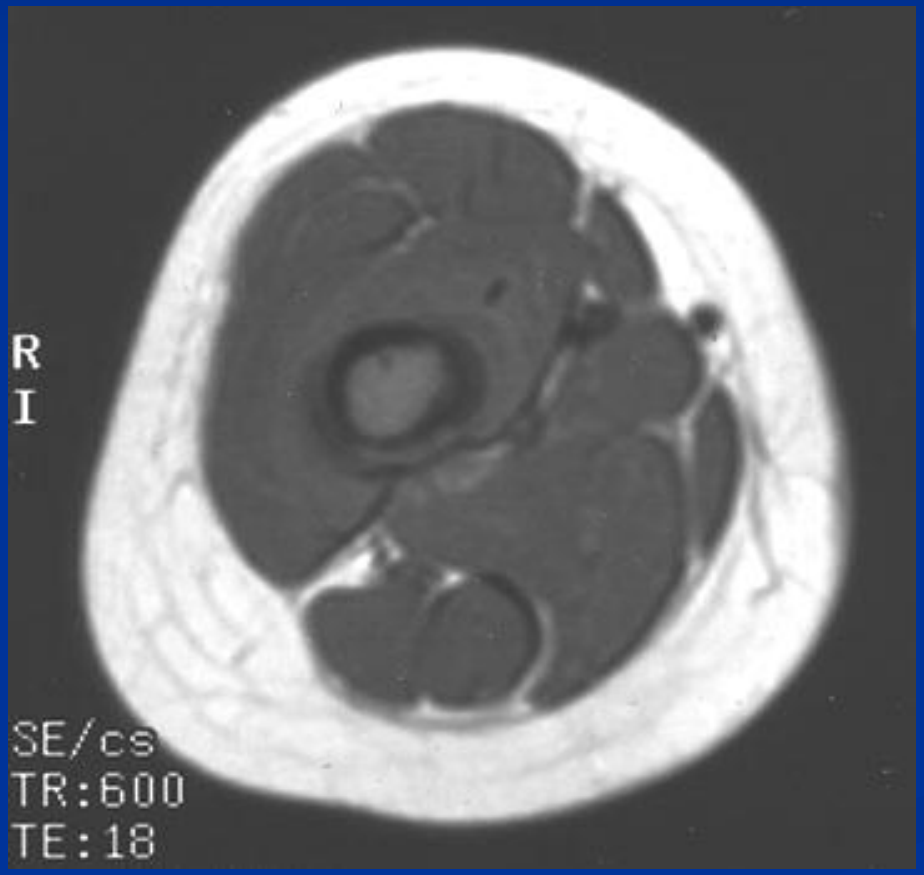
MRI: Eosinophilic Granuloma

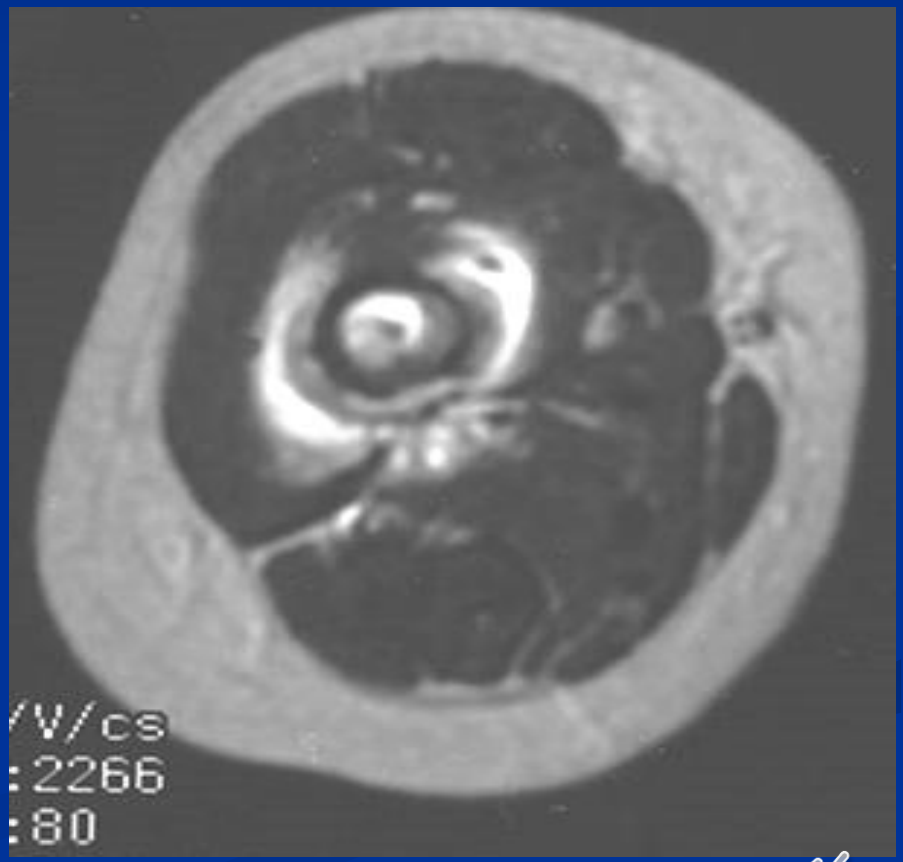
- Marrow Replacement Intermediate Signal on T1
- High Signal on T2
- May have surrounding edema
- Soft Tissue mass possible

MRI: Eosinophilic Granuloma



MRI: Eosinophilic Granuloma

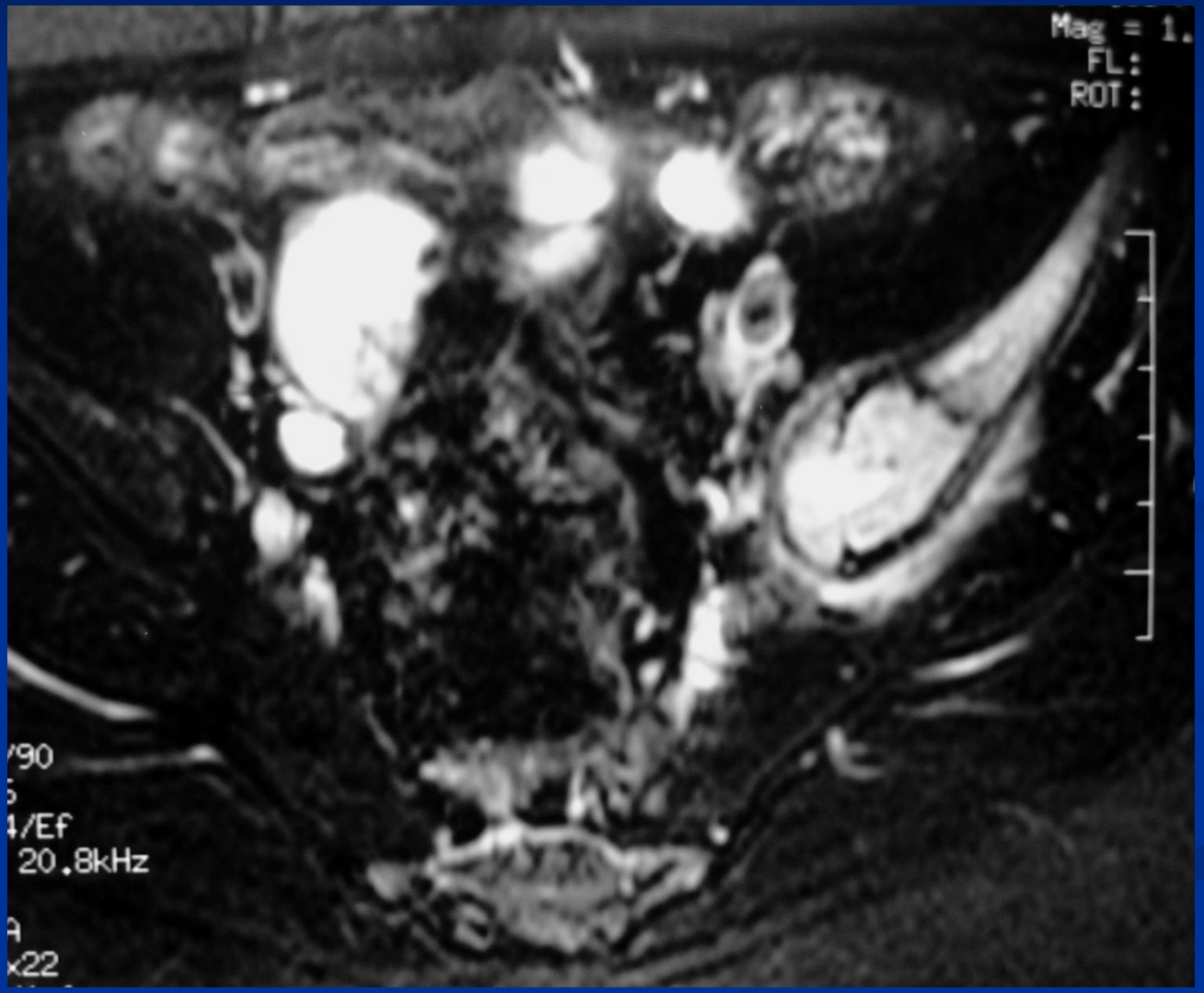




Xray of Hip: EG of Acetabulum

MRI T1: EG of Left Acetabulum

MRI T2: EG of Left Acetabulum

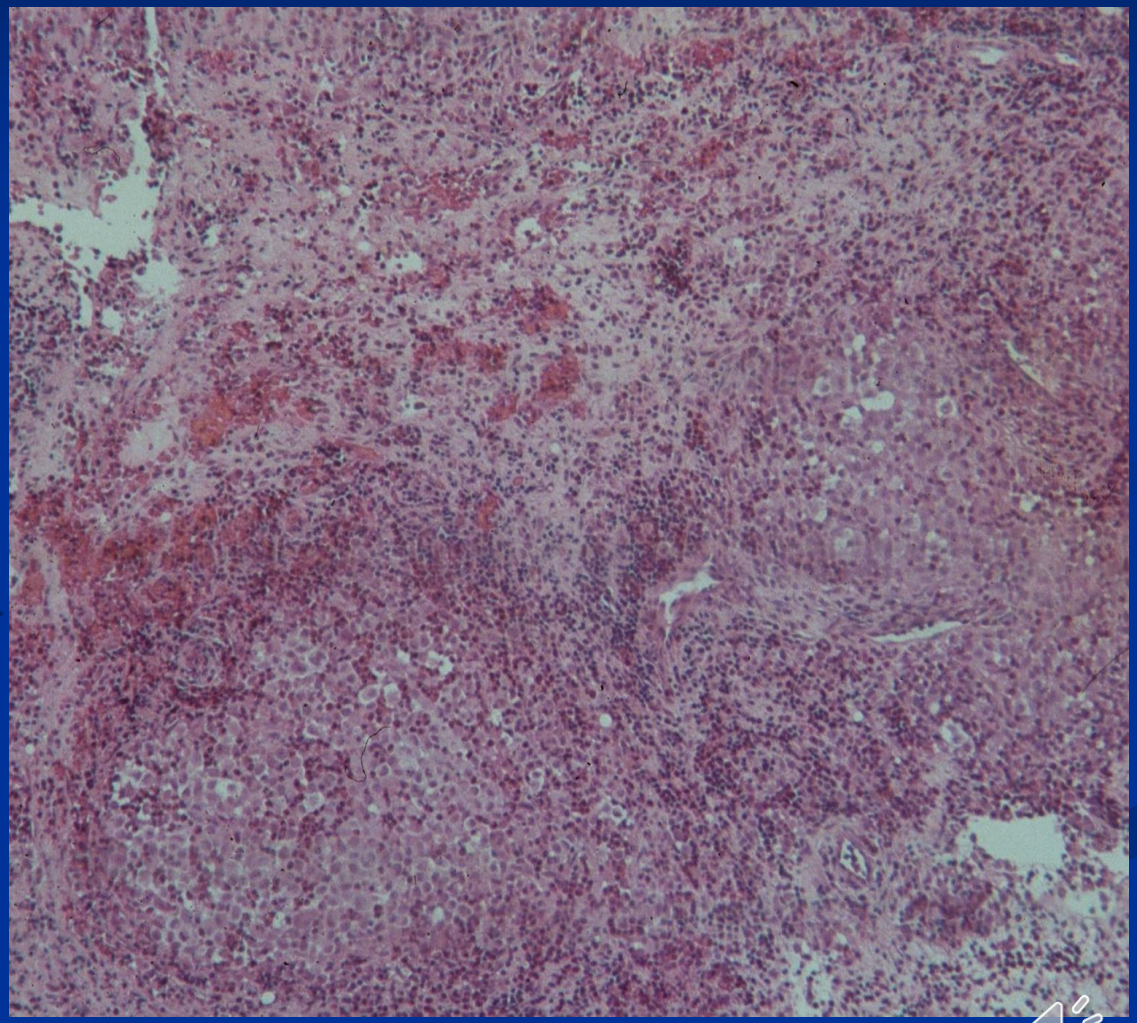


Pathology: Eosinophilic Granuloma

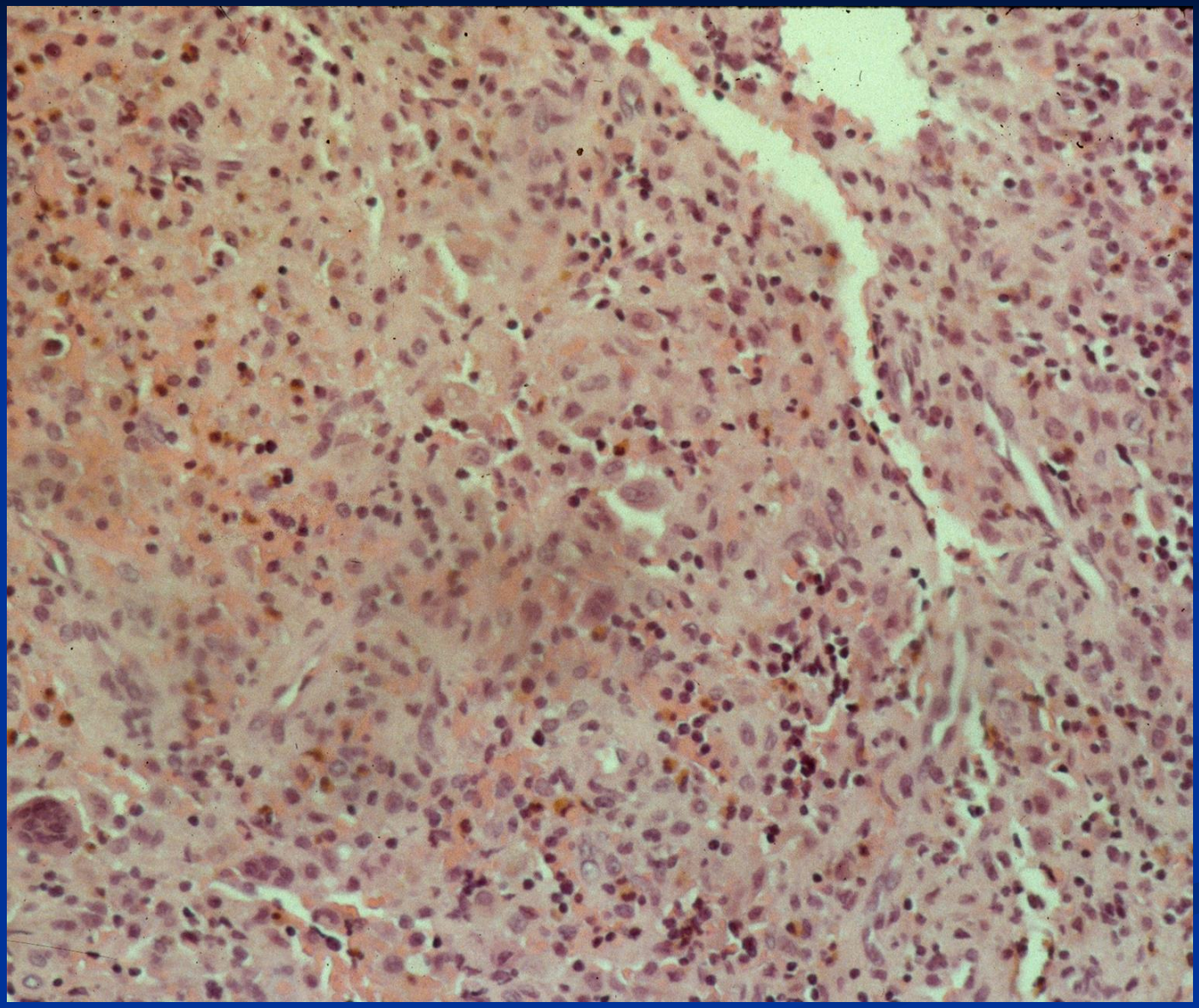
- Small Round Blue Cell Tumor (No Matrix)
- Variety of cell types (Inflammatory cells)
- Langerhans cell is diagnostic
  - Abundant eosinophilic cytoplasm
  - Bean shaped nucleus with convolutued nuclear grooves and indentations
- Eosinophils may predominate but not diagnostic
  - Small percentage of EGs do not have eosinophils.
- Lymphocytes and plasma cells can predominate and create confusion with osteomyelitis

Pathology: Eosinophilic Granuloma

- Small Round Blue Cell Tumor
- Cells without Matrix production
- Mixed Inflammatory Cells



Pathology: Eosinophilic Granuloma

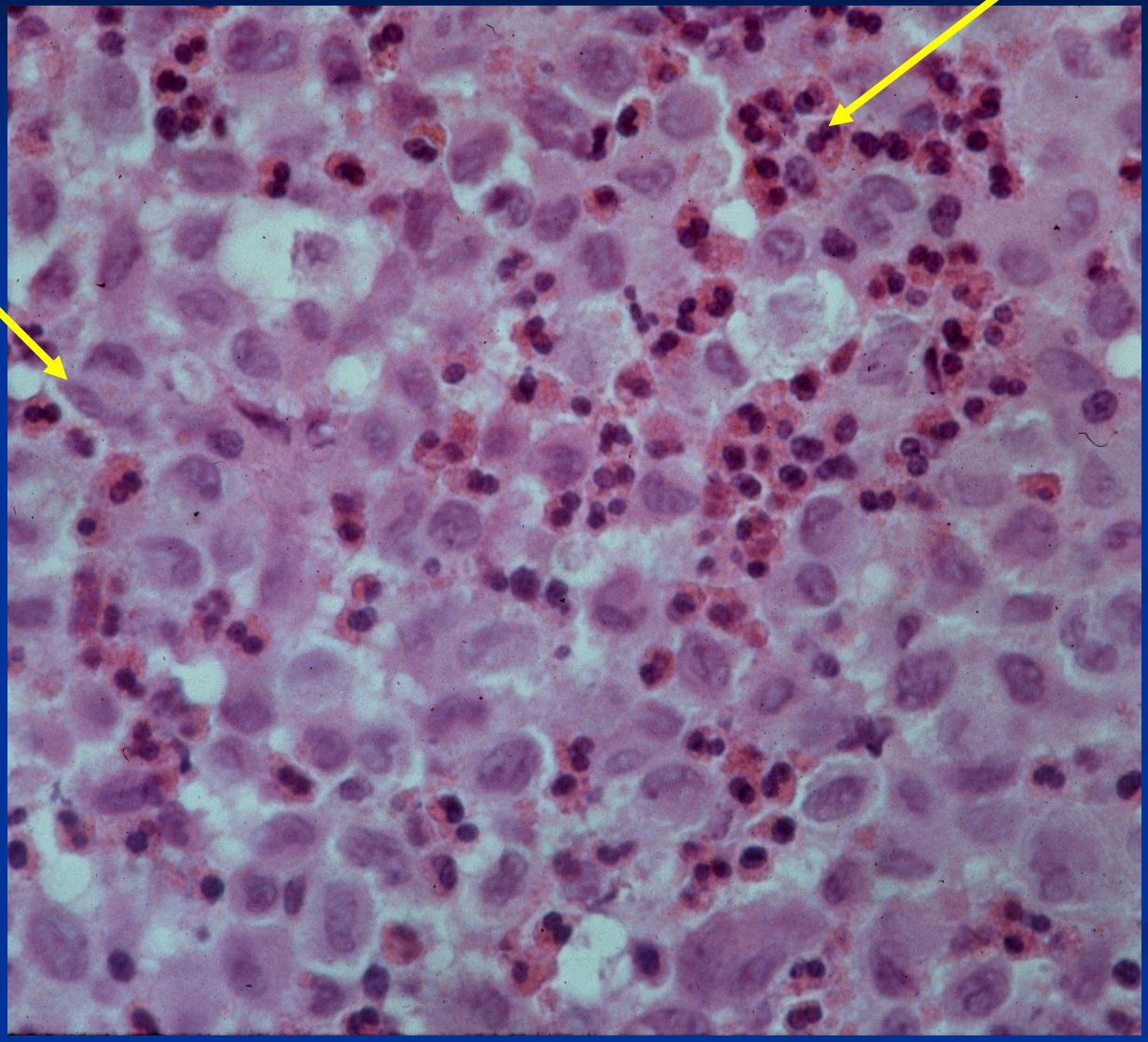


Pathology: Eosinophilic Granuloma

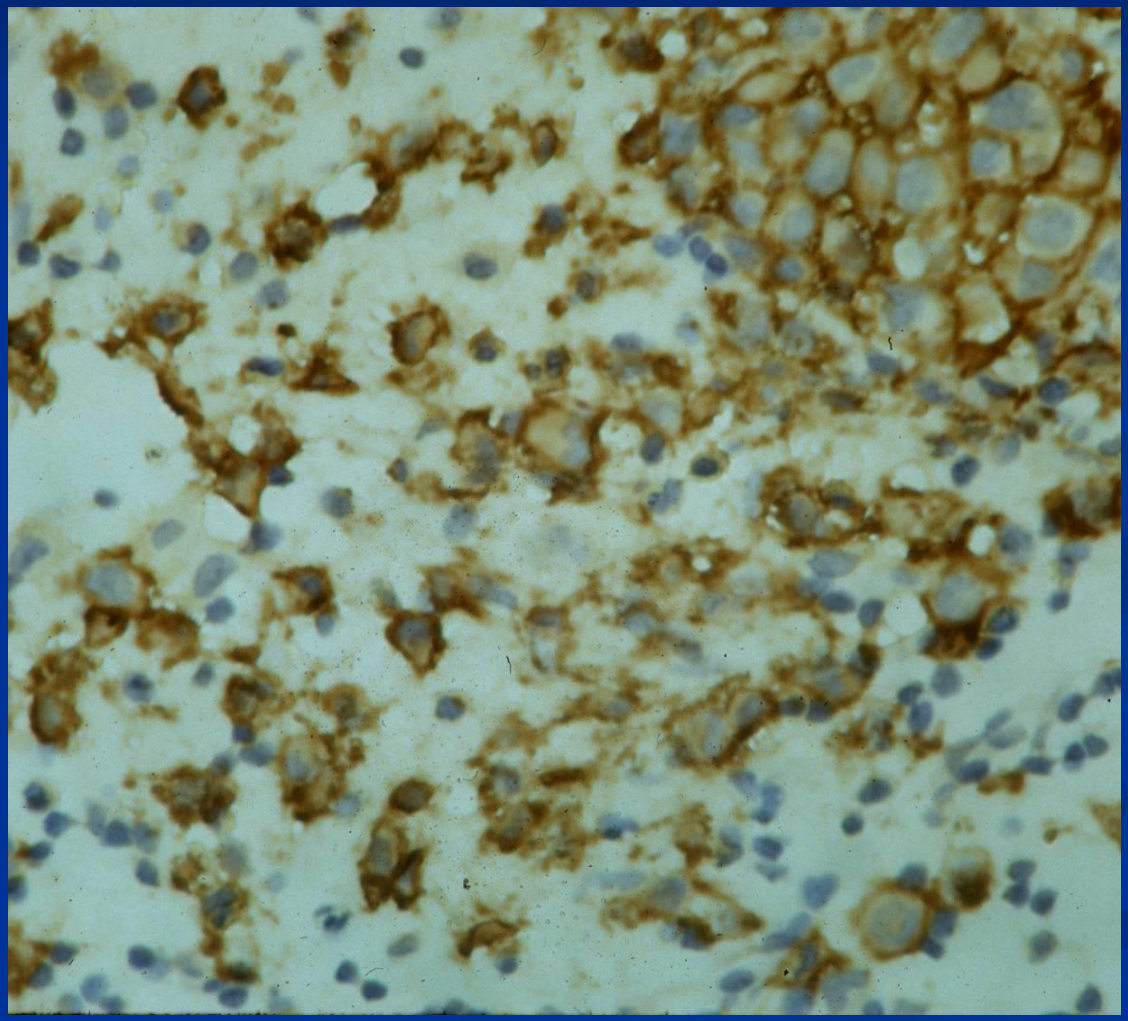
Eosinophils

Langerhans Cells

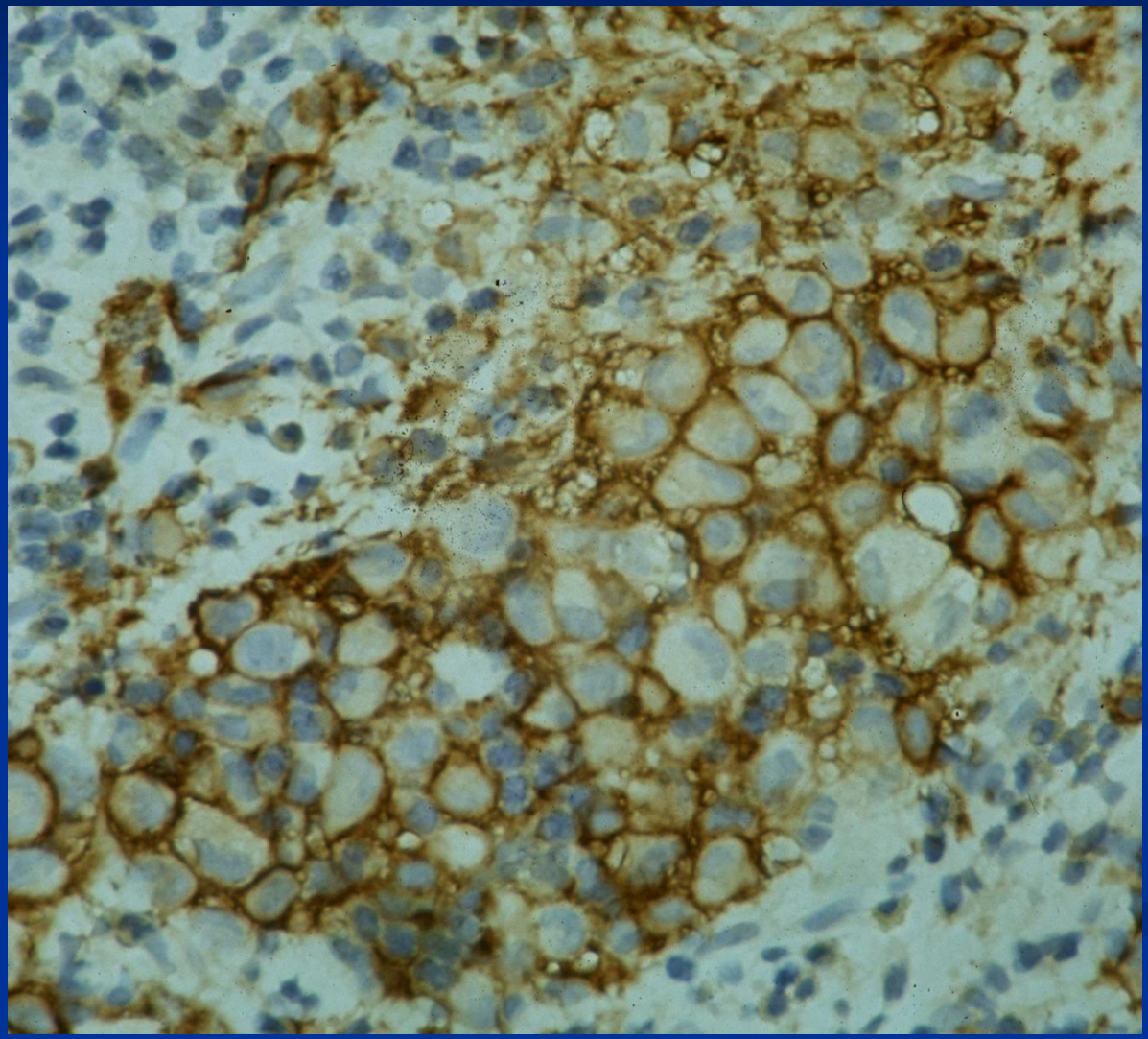
Coffee Bean Shaped Nucleus



Pathology Eosinophilic Granuloma S-100 Positive (helps distinguish from osteomyelitis)

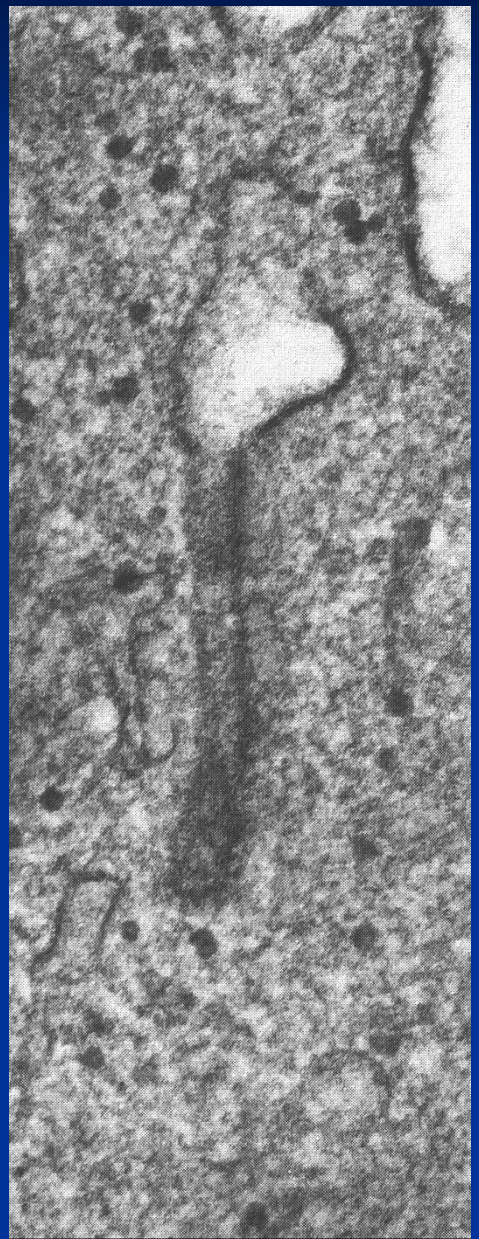


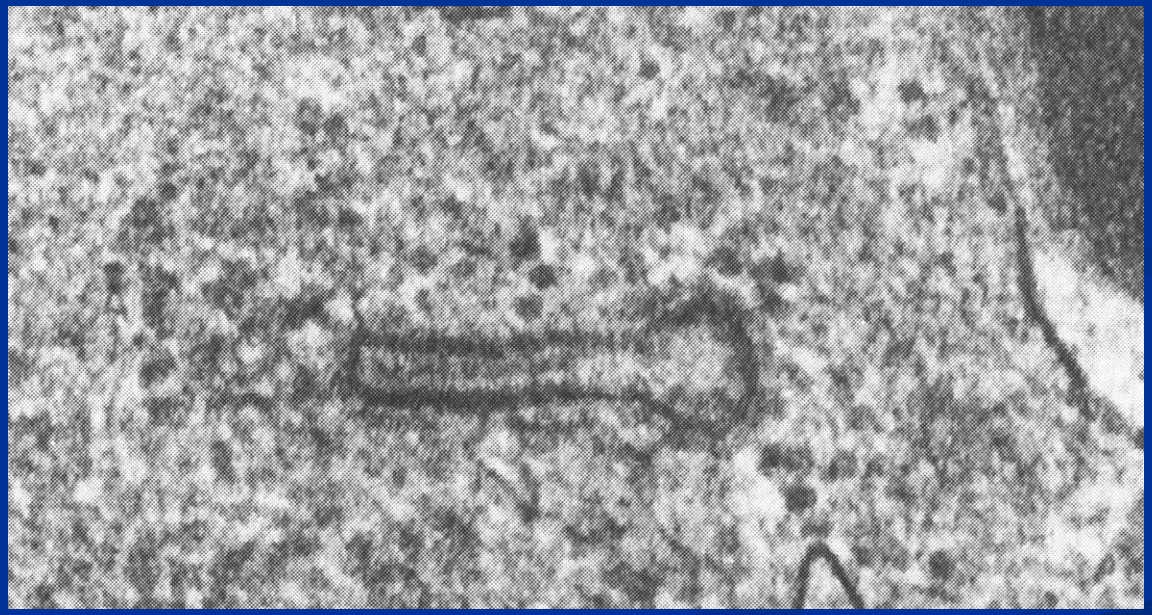
Pathology Eosinophilic Granuloma
CD-10 Positive



Electron Microscopy: EG Birbeck's Granule

Electron Microscopy: EG Birbeck's Granule (Tennis Racquet Shaped)





Eosinophilic Granuloma

- **Treatment:**
  - Curettage and bone graft
  - Observation of spine lesion—usually spontaneously regress
  - Intralesional prednisone
  - Low Dose XRT (300-1000 rads) for inaccessible lesions

Nonossifying Fibroma (NOF)

- Definition: Intracortical proliferation of fibrous tissue and histiocytes that extends into the intramedullary canal
- Fibrous Cortical Defect: Small lesion (up to 1cm) that involve only the cortex
- Jaffe-Campanacci Syndrome: Multiple NOFs with café au lait spots
- Usually found incidentally on a radiograph
- Large lesions may cause pain from pathological fracture or stress injury from weakened bone (microscopic fractures)

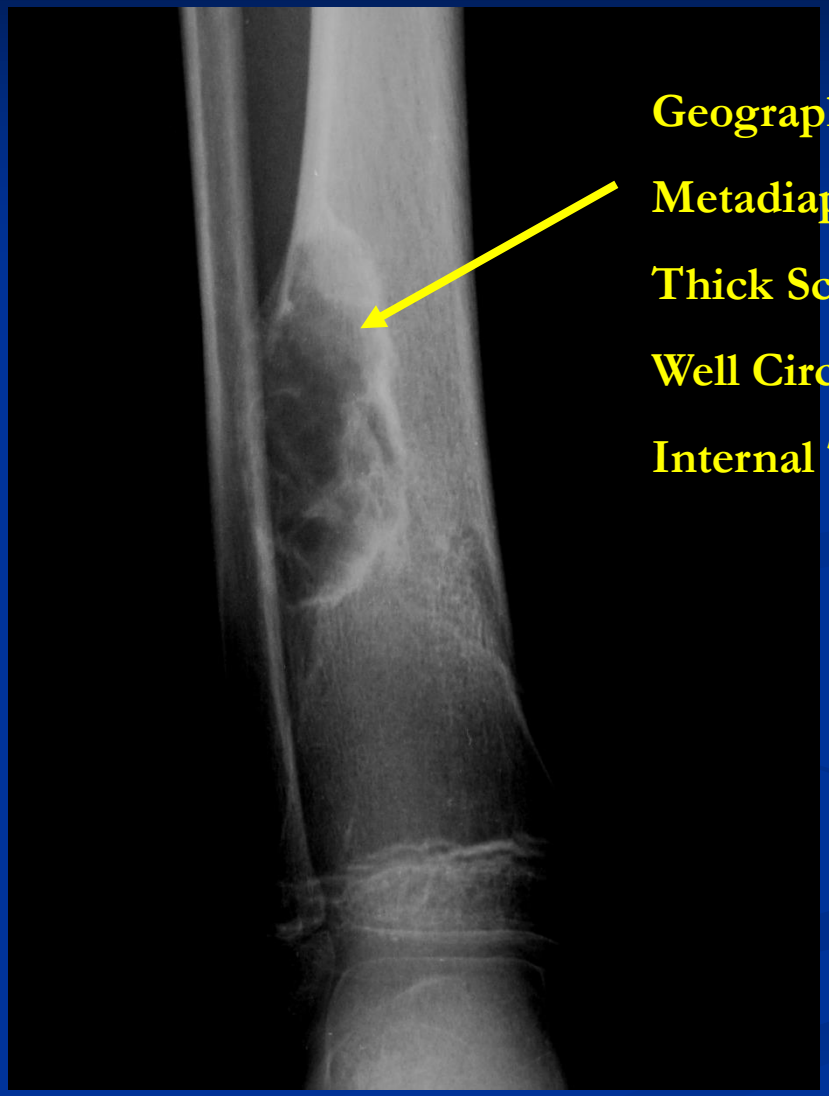
Nonossifying Fibroma (NOF)

- Many believe that NOFs start as fibrous cortical defects that enlarge
- Can be a precursor to an ABC
- NOFs: Teenage years <25 years; Usually heal spontaneously in a patient's 20s
- Males>Females 2:1
- Sites: Distal Tibia, Distal Femur, Proximal Tibia, Fibula (90% of lesions)

Radiology: Nonossifying Fibroma (NOF)

- Lytic, Geographic, Radiolucent Lesion
- Metadiaphyseal
- Sharply circumscribed with **thick rind of sclerosis**
- Internal Trabeculations
- Intracortical with growth into the intramedullary canal
- May expand slightly into soft tissue
- Heal from diaphysis to epiphysis (fill in with bone)
- Multiple NOFs usually more expansile and larger than solitary NOFs

Xray Tibia: NOF with Pathological Fracture



Geographic, Eccentric

Metadiaphyseal

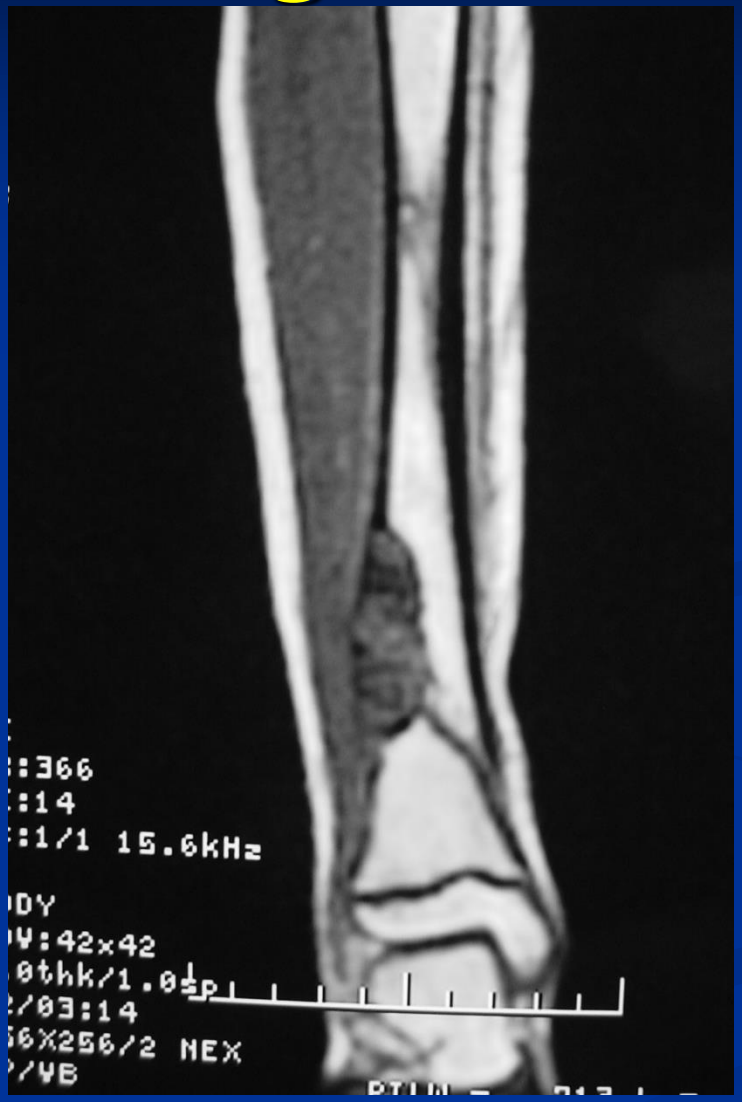
Thick Sclerotic Rim

Well Circumscribed

Internal Trabeculations

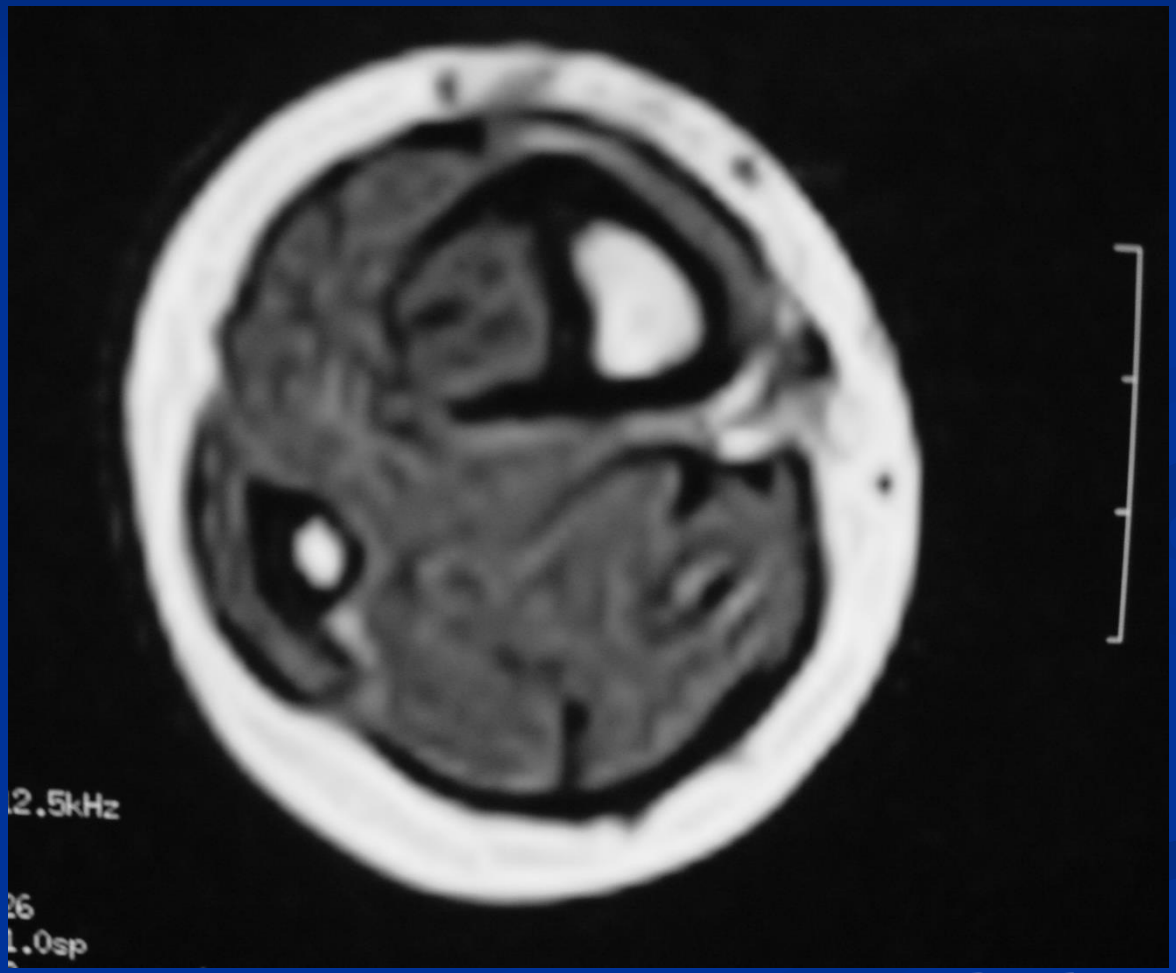
Xray Tibia: NOF with Pathological Fracture

MRI Tibia T1: NOF with Pathological Fracture

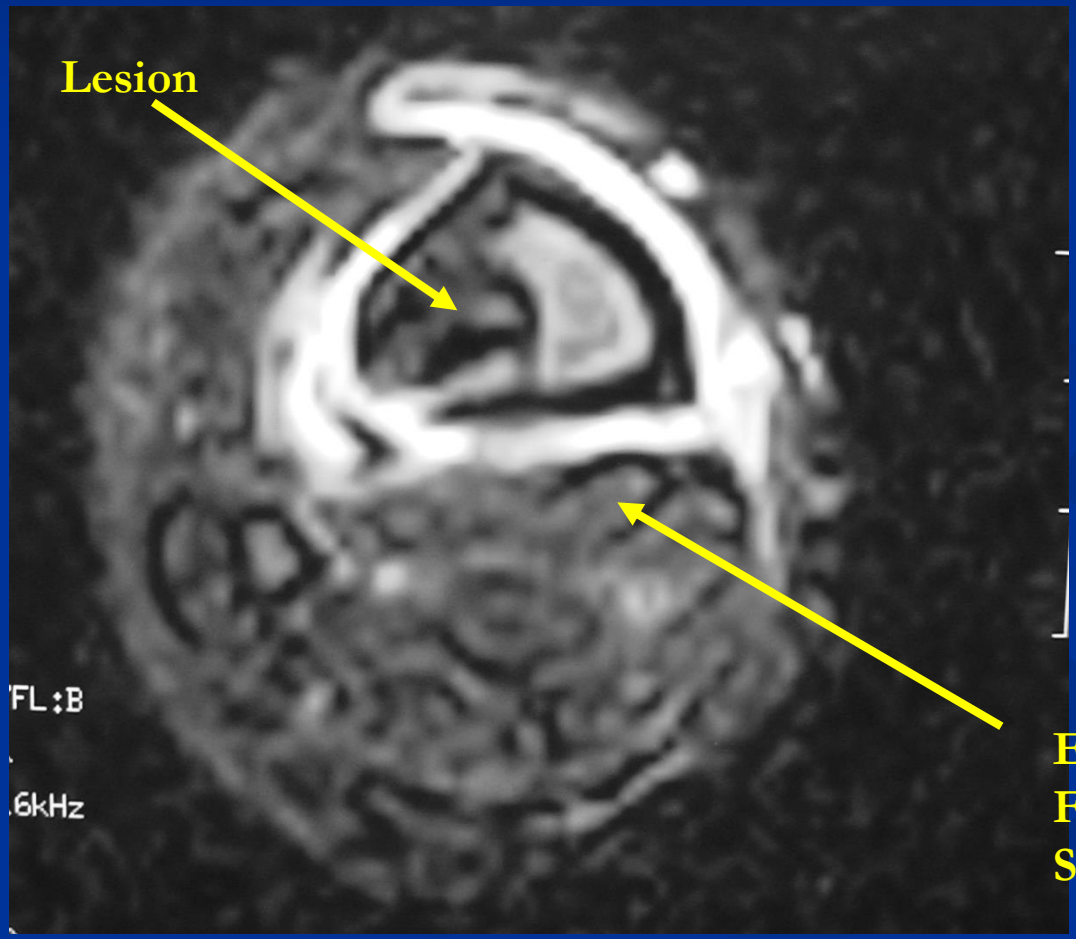


MRI Tibia T2: NOF with Pathological Fracture

MRI Tibia T1: NOF
Intermediate Signal Similar to Muscle



MRI Tibia T2: NOF with Fracture
Low to Intermediate Signal– Fibrous Tissue
(Not all NOFs are Low to Intermediate Signal on T2)



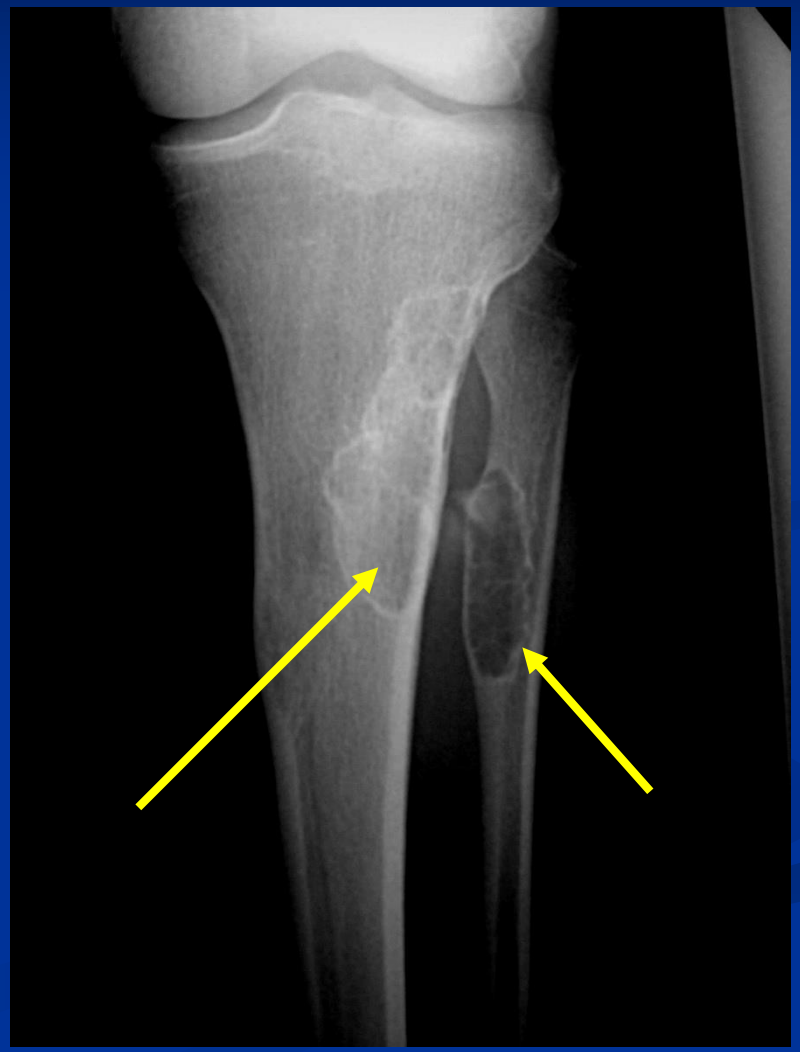
Xray Femur: NOF
Jaffe-Campanacci Syndrome

- Geographic
- Metadiaphyseal
- Eccentric
- Thick Sclerotic rim
- Internal Trabeculations
- No Mineralization
- May be expansile especially in multiple NOFs

Xray Femur: NOF
Jaffe-Campanacci Syndrome

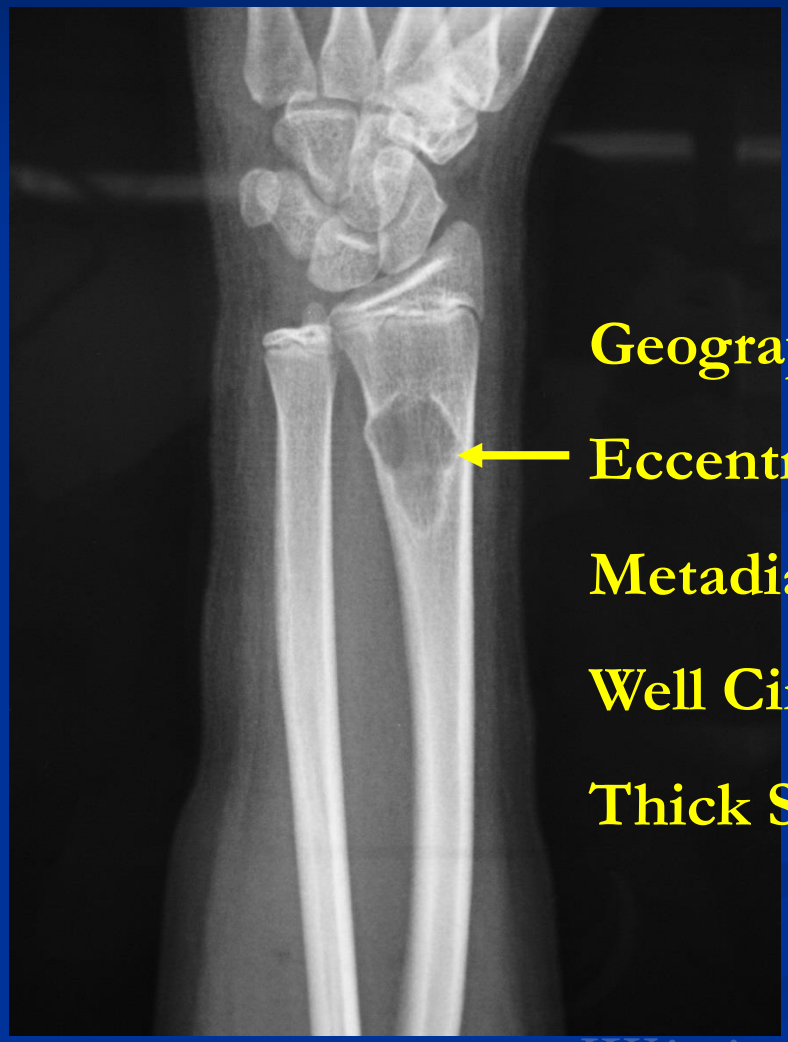
Xray Tibia/Fibula: Multiple NOFs

- Lesion in Proximal Tibia Almost Completely Healed
- Geographic Lesion
- Eccentric
- Metadiaphyseal
- Well Circumscribed
- Sclerotic rim
- Fibula lesions may be more expansile than in other larger long bones



Xray Distal Radius: Fibrous Cortical Defect

Xray Radius One Year Later
Growth into NOF (Atypical Location)



Geographic, Radiolucent

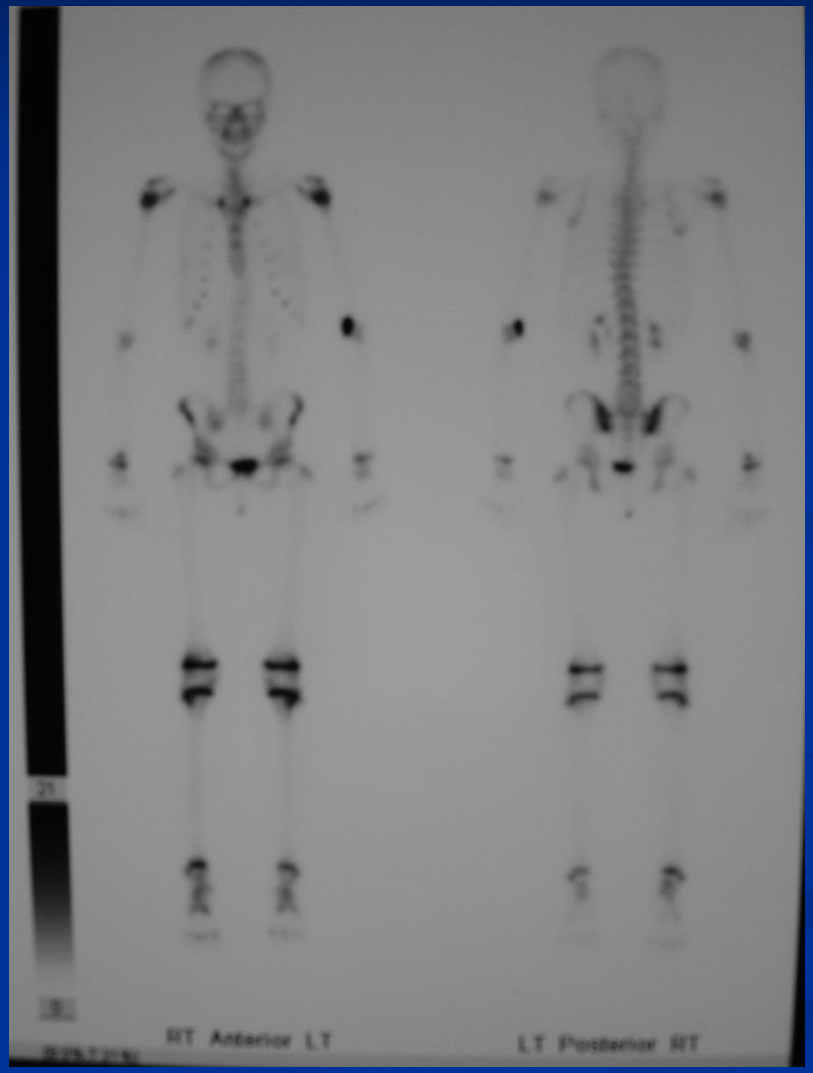
Eccentric

Metadiaphyseal

Well Circumscribed

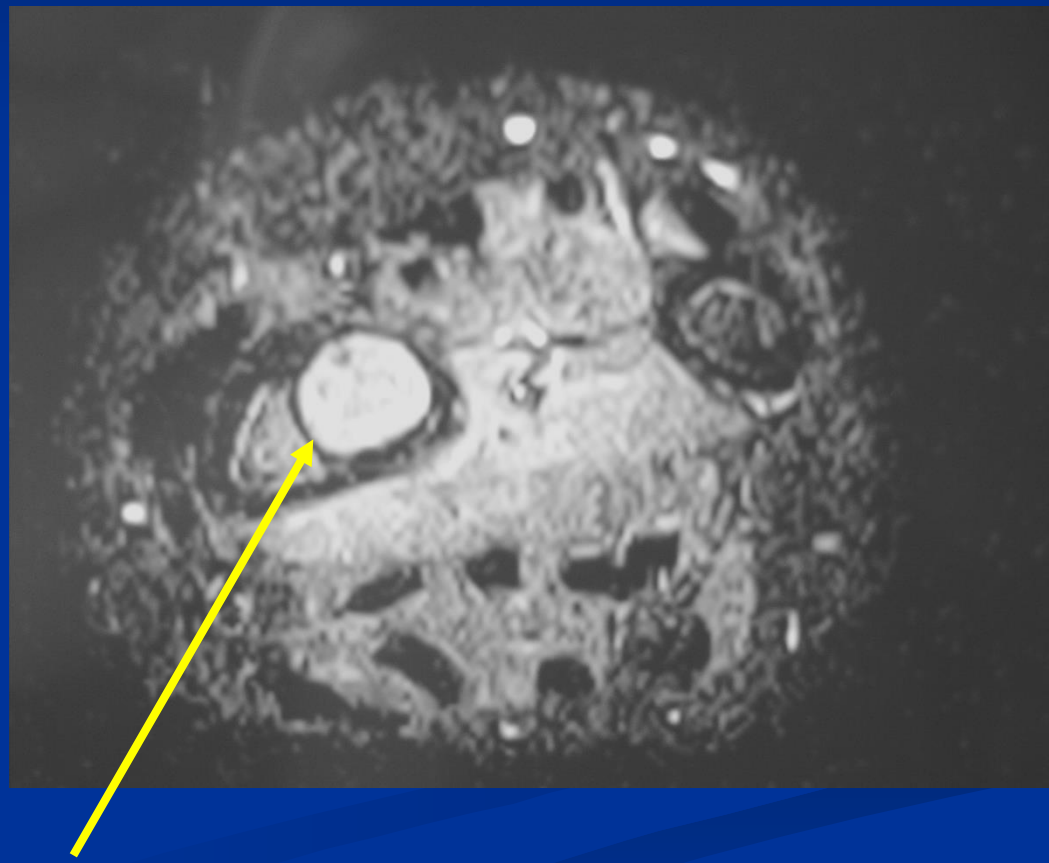
Thick Sclerotic Rim

Bone Scan: NOF Right Distal Radius Mild Uptake



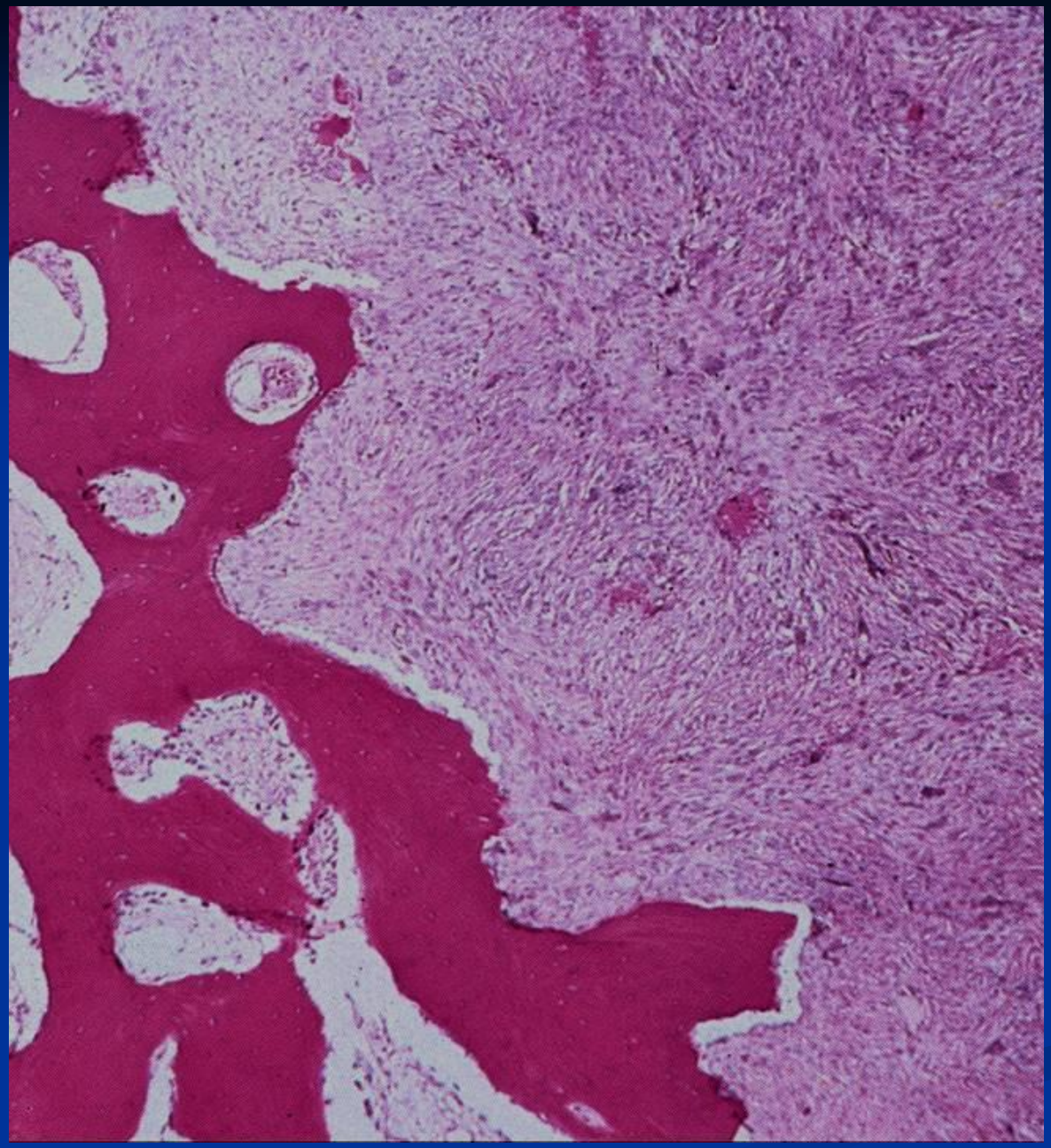
MRI T2: NOF Distal Radius

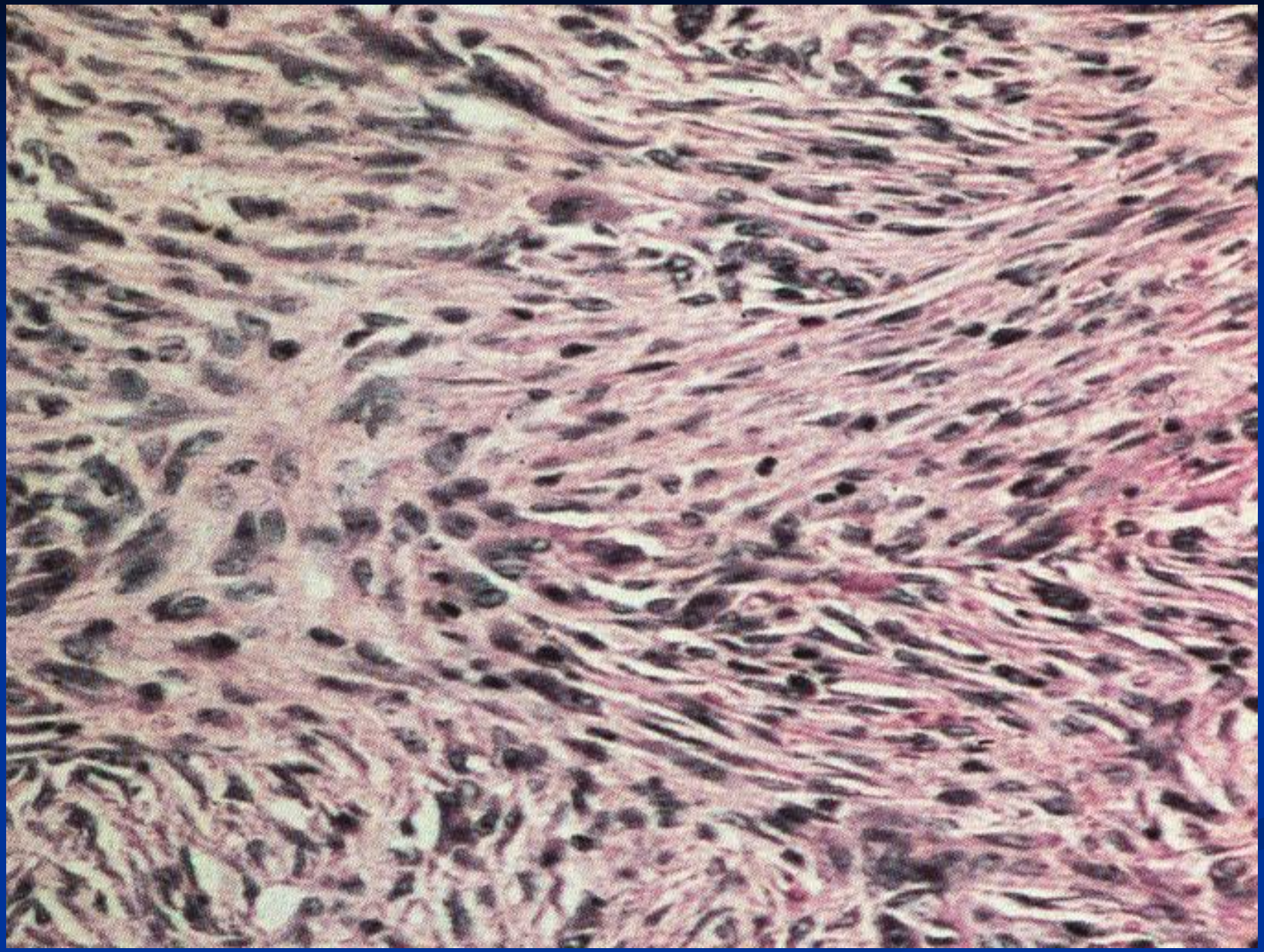
- T2 Signal Variable
- Does not always follow signal intensity of fibrous tissue (low to intermediate)
- This NOF is High Signal on the T2 weighted image

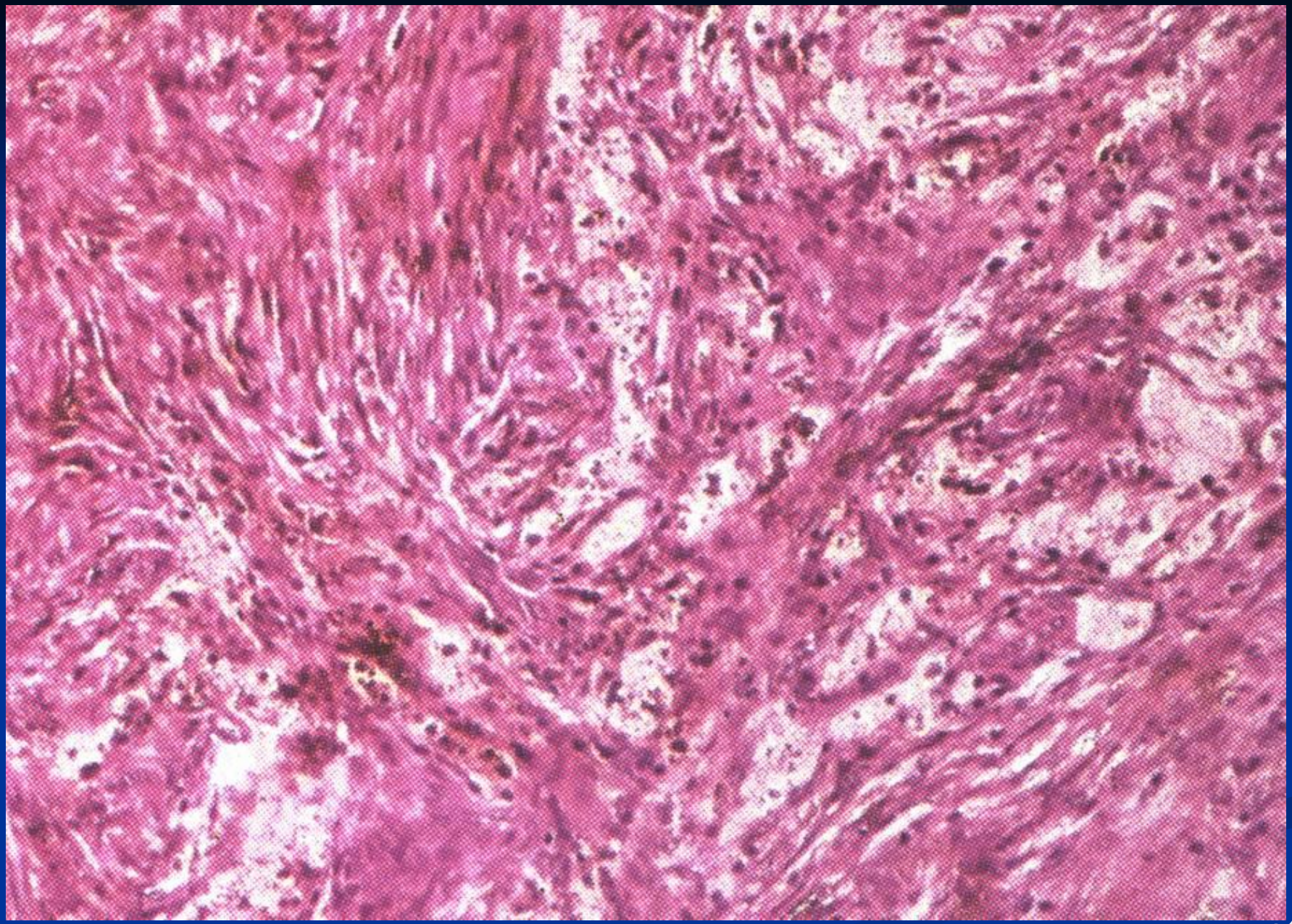


Pathology Nonossifying Fibroma (NOF)

- Benign, bland appearing spindle cells with fibrous tissue stroma
- Usually a storiform or swirling irregular arrangement of the fibrous tissue
- Contains foamy histiocytes, hemosiderin laden histiocytes and giant cells in variable portions







Treatment Nonossifying Fibroma (NOF)

- Incidental Finding: Observe
- Intralesional Curettage and Bone Graft:
  - Symptomatic Lesions
  - Fractured Lesions (may also require internal fixation)
  - Prophylactic treatment of lesions greater than 50% diameter of the bone

Thank You!

- www.TumorSurgery.org