

# Conventional Chondrosarcoma

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# General Information

- Malignant mesenchymal tumor of cartilaginous differentiation.
- Conventional Chondrosarcoma is the most common type of chondrosarcoma (malignant cartilage tumor)
- Neoplastic cells form **hyaline type cartilage or chondroid type tissue** (Chondroid Matrix) but not osteoid
- If lesion arises de novo, it is a primary chondrosarcoma
- If superimposed on a preexisting benign neoplasm, it is considered a secondary chondrosarcoma
- Central chondrosarcomas arise from an intramedullary location. They may grow, destroy the cortex and form a soft tissue component.
- Peripheral chondrosarcomas extend outward from the cortex of the bone and can invade the medullary cavity. Peripheral chondrosarcomas most commonly arise from preexisting osteochondromas.
- Juxtacortical chondrosarcomas arise from the inner layer of the periosteum on the surface of the bone. It is technically considered a peripheral chondrosarcoma.

# Chondrosarcoma

- Heterogeneous group of tumors with varying biological behavior depending on grade, size and location
  - Cartilage tumors can have similar histology and behave differently depending on location. For instance a histologically benign appearing cartilage tumor in the pelvis will behave aggressively as a low grade chondrosarcoma. Likewise, a histologically more aggressive hypercellular cartilage tumor localized in a phalanx of a digit may behave in an indolent, non aggressive or benign manner.
- There are low (grade I), intermediate (grade II) and high grade (grade III) types of conventional chondrosarcoma.
- Low grade lesions are slow growing and rarely metastasize. Low grade chondrosarcomas can be difficult to differentiate from benign tumors histologically. Clinical features and radiographic studies are important to help differentiate. Low grade chondrosarcomas can dedifferentiate into high grade sarcomas and higher grade chondrosarcomas over a prolonged period of time. These higher grade sarcomas readily metastasize and are associated with a poor prognosis.
- High grade lesions grow aggressively locally and metastasize readily. They are more easily differentiated from low grade and benign cartilage tumors.
- Chondrosarcomas most commonly arise from bone but can also arise from soft tissue. They are the most common bone sarcoma in adults and the second most common bone sarcoma overall after osteosarcoma. Most malignant cartilage tumors are low to intermediate grade tumors.

# Classification of Cartilage Tumors

## ■ Benign

- Enchondroma
- Osteochondroma
- Chondroblastoma
- Chondromyxofibroma

## ■ Malignant

- Conventional Intramedullary
  - Grade 1 (Low Grade)
  - Grade 2 (Intermediate Grade)
  - Grade 3 (High Grade)
- Dedifferentiated
- Clear Cell
- Mesenchymal
  - Intramedullary
  - Extraskeletal
- Secondary
  - Osteochondroma or Enchondroma
- Periosteal/Juxtacortical
- Extraskeletal (Soft Tissue)



# Classification Chondrosarcoma

## Primary (90%)

Arising de novo in  
normal bone

### Central Intramedullary (99%)

Conventional (85-90%)

Grade 1 (30%)

Grade 2 (40%)

Grade 3 (30%)

Dedifferentiated (8%)

Clear Cell (4%)

Mesenchymal (1%)

### Peripheral (1%)

Periosteal C.S

## Secondary (10%)

Arising from pre  
existing conditions of  
bone

Enchondroma

Osteochondroma

Ollier's, Maffucci's

Fibrous Dysplasia

Paget's

Chondroblastoma

Radiation induced

# Clinical Presentation

## Conventional Chondrosarcoma

### ■ Signs/Symptoms:

- Pain, with or without mass
- Pathological fracture is rare

### ■ Prevalence:

- 2 to 1 male predilection
- Most common bone sarcoma in adult population
- Second most common primary sarcoma of bone
- 20% of all primary malignant bone sarcomas

### ■ Age:

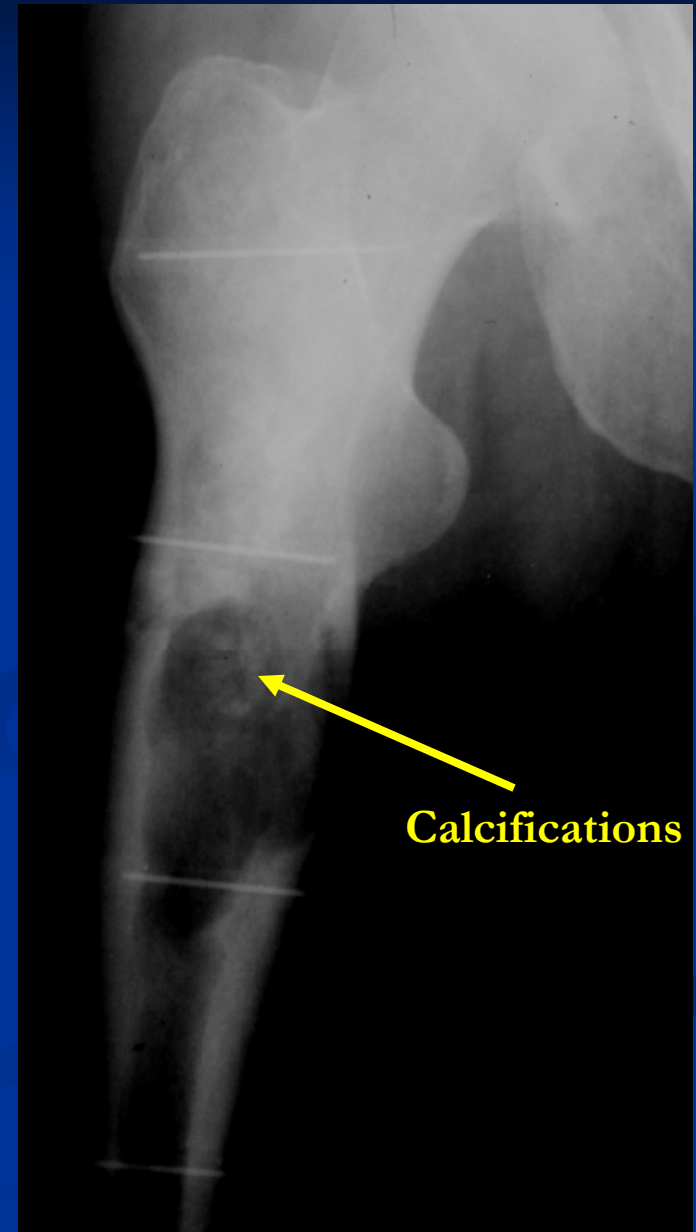
- All ages but peak incidence between 50-70 years of age
- Uncommon before the age of 40
- Primary chondrosarcoma most common between fifth and seventh decade
- Secondary chondrosarcoma most common between fourth and fifth decade

### ■ Sites:

- Most common sites: Proximal femur, Distal femur, Proximal Humerus, Pelvis, Scapula, Ribs
- Spine and craniofacial bones are rare sites

# Radiographic Presentation

- Conventional chondrosarcomas primarily occur in the metaphysis or diaphysis
  - Rarely, they arise in the epiphysis
- Peripheral chondrosarcomas form a mass that extends from surface of bone
- Calcifications have a distinctive “Ring and Arc”-like pattern
  - Due to lobular growth of cartilage and enchondral ossification around the perimeter
- Low-grade chondrosarcomas
  - Uniformly calcified
  - Well-defined margins
- High-grade chondrosarcomas
  - Large non-calcified areas
  - Irregular, ill-defined margins
  - Often extend into soft tissues



# Conventional Intramedullary Chondrosarcoma

## Radiographic Presentation

- May be Geographic to Permeative depending on Grade
- Deep endosteal scalloping
- Cortical thickening/Periosteal reaction
- Cortical destruction and soft tissue mass variable
  - Benign cartilage tumors never have a soft tissue component
- Chondroid matrix (78% by x-ray; 94% by CT)
  - Calcifications in a ring and arc-like manner
  - Stippled calcifications
  - CT is useful for evaluating subtle calcification
- MRI – similar to muscle in T1W
  - Lobulated high intensity T2W images
  - Matrix Calcification – Low intensity
  - Best test for evaluating intramedullary extent and soft tissue extension

# Conventional Intramedullary Chondrosarcoma

## Radiological Features of Malignancy

- Bone contour in the affected area may be expanded
- Cortical thickening
- Endosteal scalloping
- New areas of lysis adjacent to calcified areas
- Cortical destruction and soft tissue extension in higher grade lesions; extension into soft tissues is definitive



# Plain X-ray: Chondrosarcoma of Proximal Femur

Permeative Lesion greater than 5cm

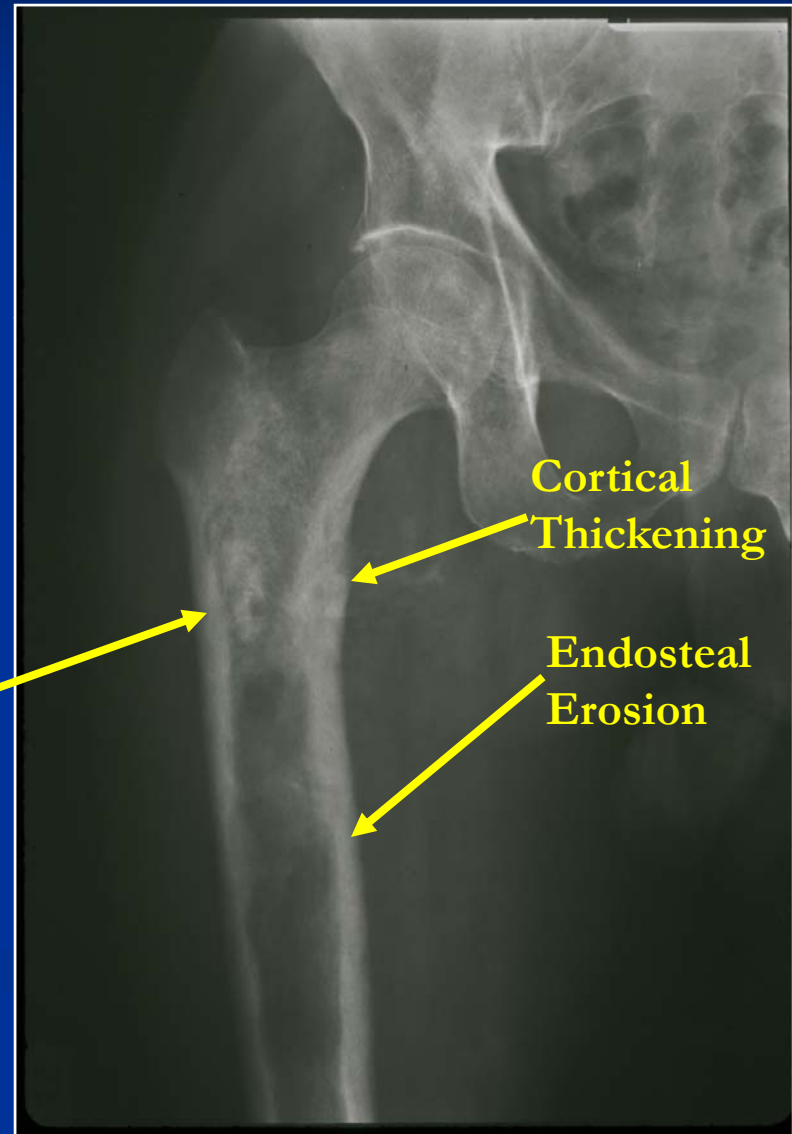
Deep Endosteal Scalloping

Cortical Thickening

Calcifications

Cortical  
Thickening

Endosteal  
Erosion

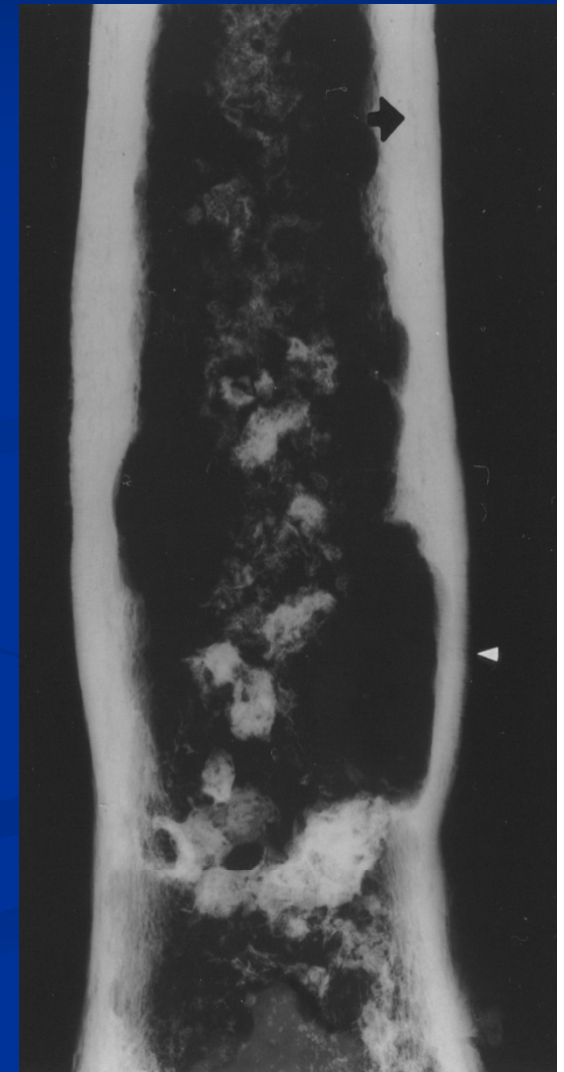




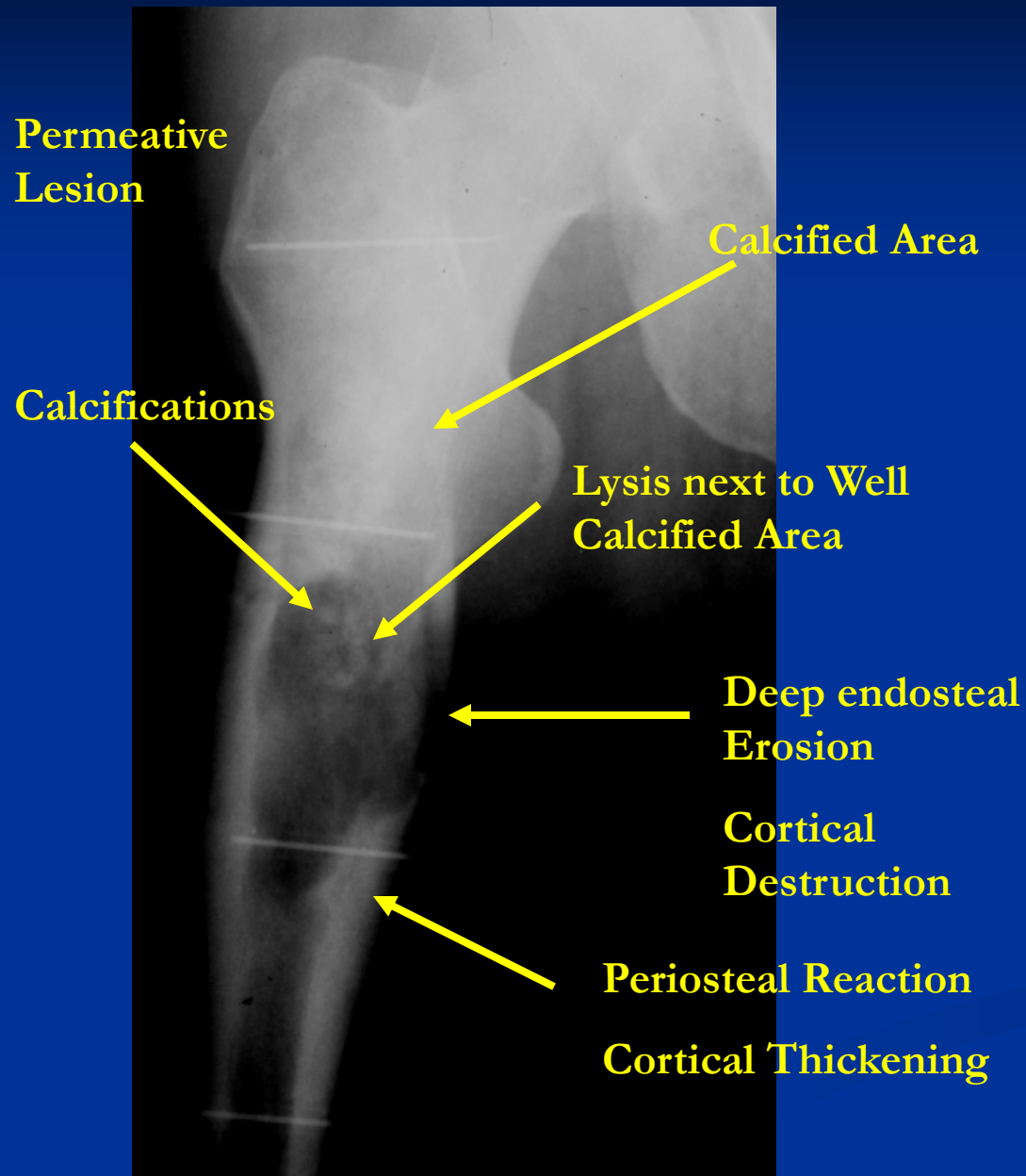
# Plain X-ray: Chondrosarcoma of Femur

Permeative  
Lesion Greater  
than 5 cm  
Calcifications in  
Lesion

Deep Endosteal  
Scalloping



# Plain X-ray: Chondrosarcoma of Proximal Femur





# Radiologic Differentiation of Chondrosarcomatous Lesions

- Aggressive chondroid lesion with soft tissue mass
  - High grade conventional chondrosarcoma
  - Dedifferentiated chondrosarcoma
  - Mesenchymal chondrosarcoma
- Large fluid component bone or soft tissue (Myxoid chondrosarcoma)
- Change in appearance or foci of more aggressive nature (Dedifferentiated chondrosarcoma)
  - Lysis or cortical destruction adjacent to well calcified area

# Diagnostic Dilemma: Long Bone

## Enchondroma vs. Chondrosarcoma

### ■ Enchondroma

- Common in hand/foot
- Common in long bones (1.7% femora)
- Rare in axial skeleton
- Rare in pelvis
- Never has an associated soft tissue component

### ■ Chondrosarcoma

- Common in axial skeleton
- Common in long bones
- Rare in hand/foot
- May or may not have an associated soft tissue mass
- Low grade chondrosarcomas do not often have an associated soft tissue mass and are most difficult to differentiate from an enchondroma

# Long Bone Enchondroma

## ■ Clinikoradiological Aspects:

- Age <50; Pain not attributable to lesion
- Size:
  - <5cm (CT/MRI)
- Bone Scan =/ $\leq$  ASIS\* 79%
- Majority in diaphysis
- Endosteal scalloping depth <2/3 cortex (90-95%)
- No cortical thickening
- No periosteal reaction
- NO cortical destruction
- NO soft tissue mass
- MRI peripheral enhancement?

■ \*AIC = Anterior Iliac Crest

# Long Bone Chondrosarcoma

## ■ Clinikoradiological Aspects:

- Age > 50; Pain attributable to lesion
- Size:
  - > 5cm (CT/MRI)
- Bone Scan = / > ASIS 82%
- Endosteal scalloping depth > 2/3 cortex (75-90%)
- Cortical Thickening (47%)
- Periosteal Reaction (51%)
- Soft Tissue Mass (Variable; May not have a soft tissue mass)
- Epiphyseal Extension (majority metaphysis)
- MRI peripheral and septal enhancement?

# Grade I Chondrosarcoma

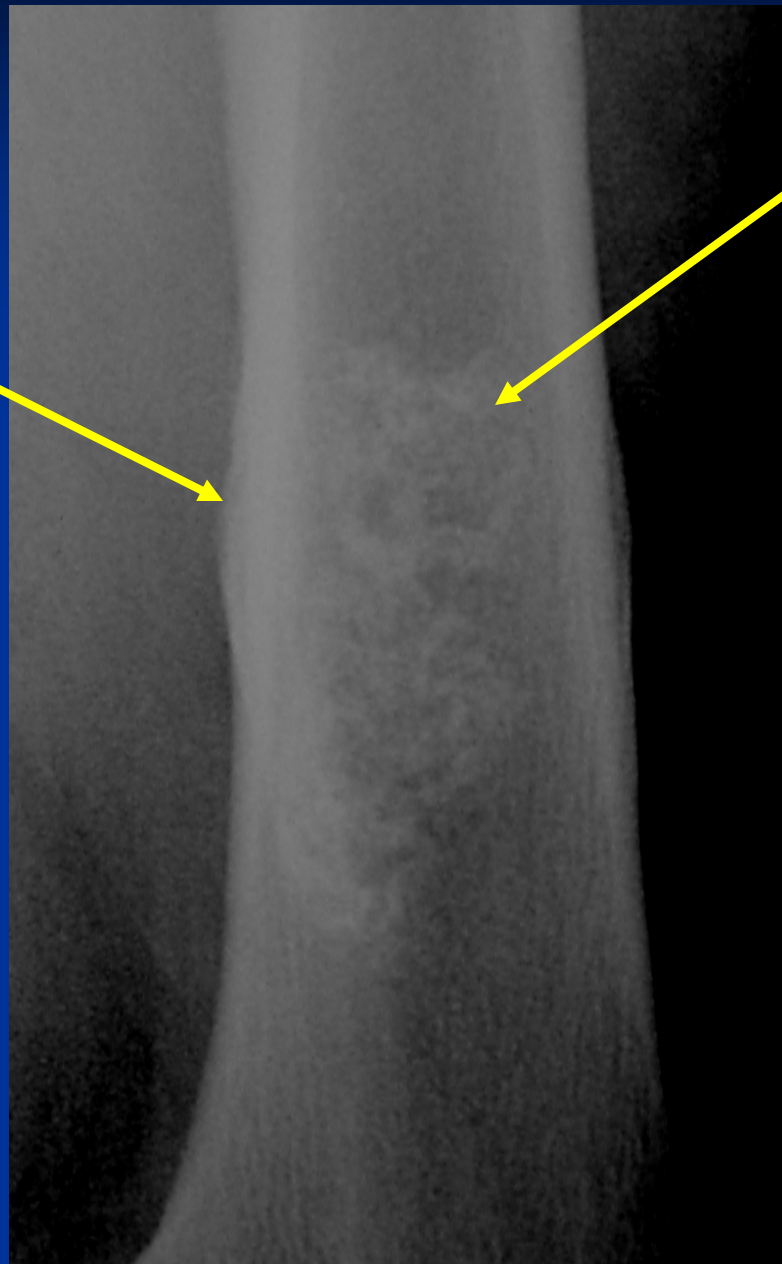
- Calcifications in ring and arc-like manner; stippled calcifications
- Mild bony expansion
- >5cm
- Endosteal scalloping > 2/3 cortical thickness



# Grade I Chondrosarcoma

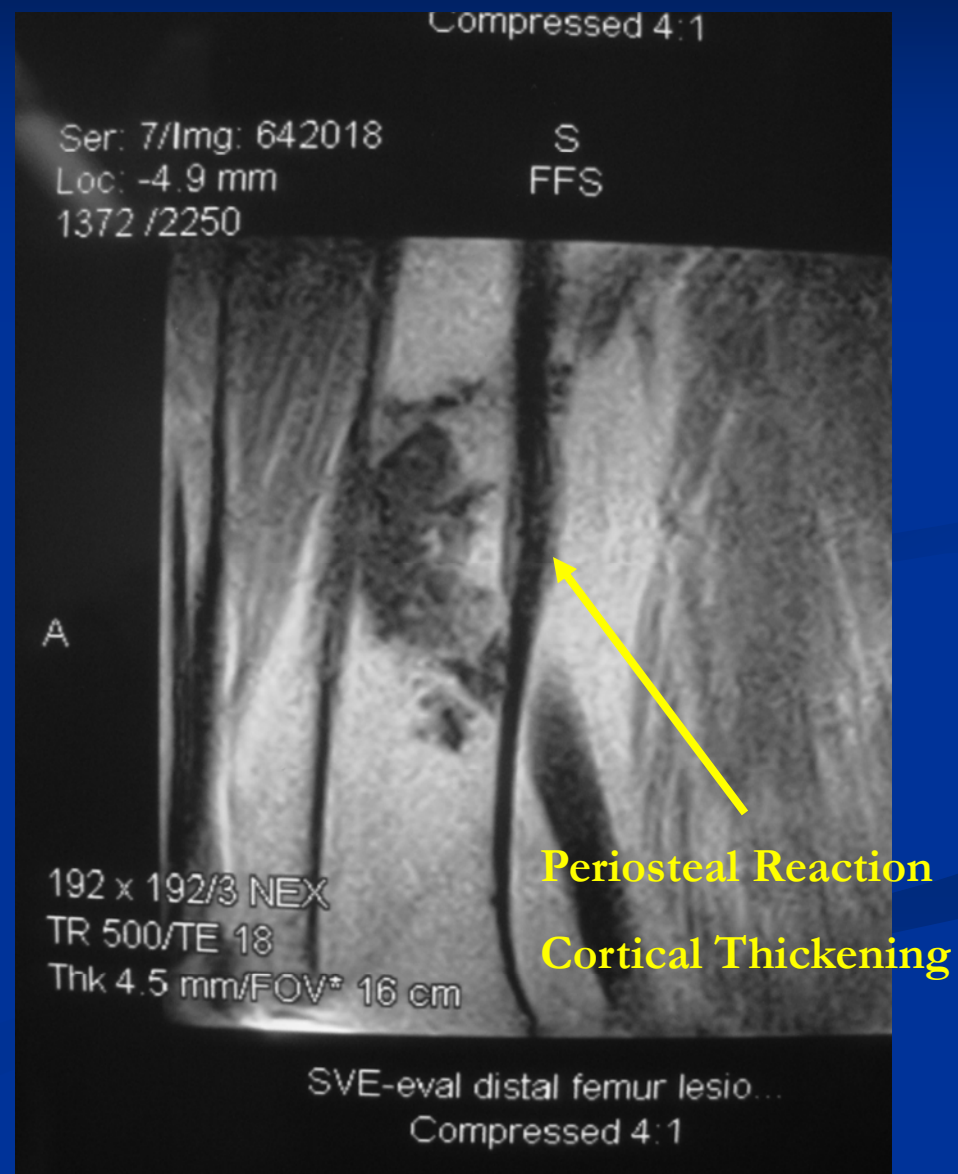
Subtle Cortical  
Thickening and  
Periosteal Reaction

Ring and Arc  
Calcifications



# Grade I Chondrosarcoma

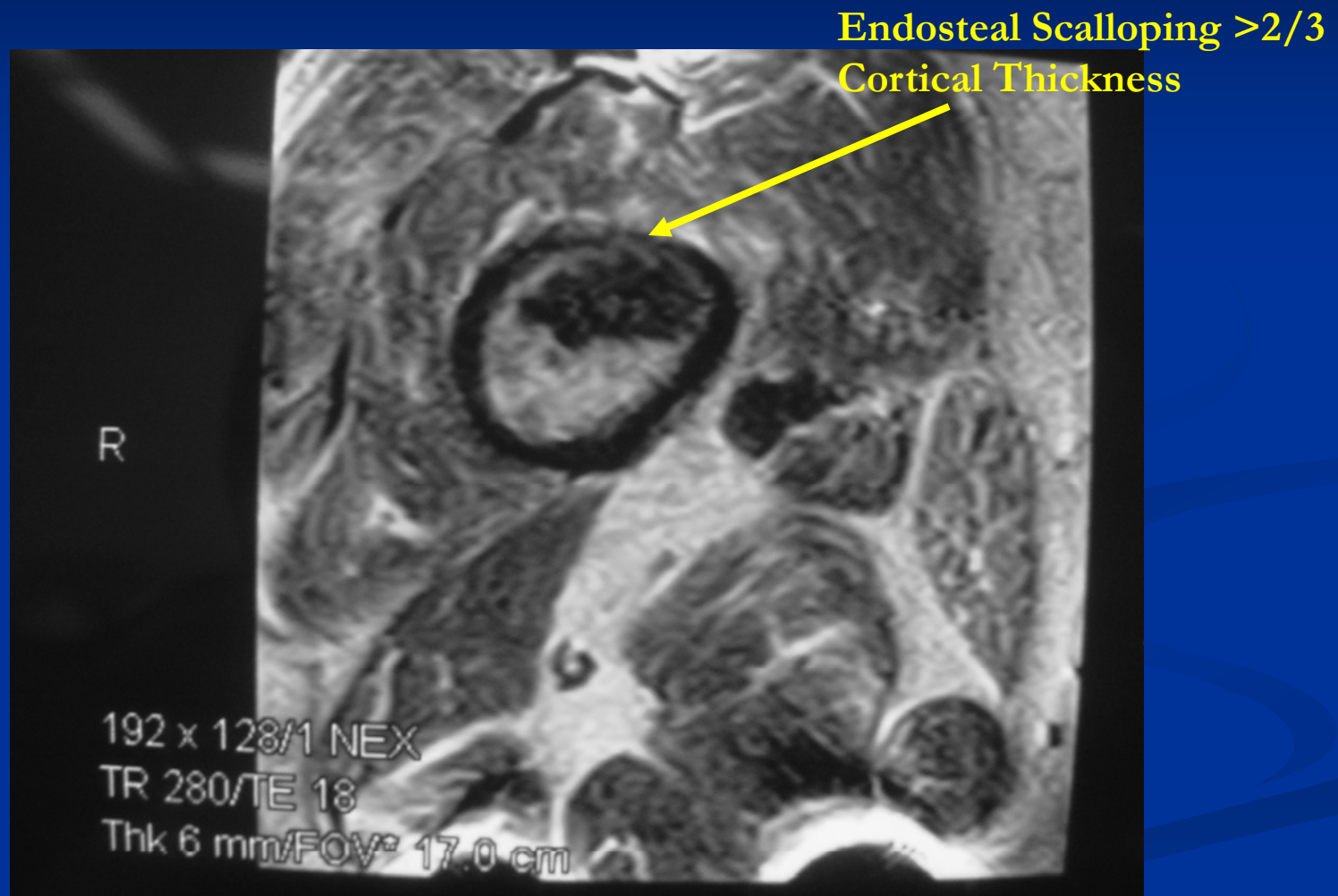
## T1 Weighted MRI





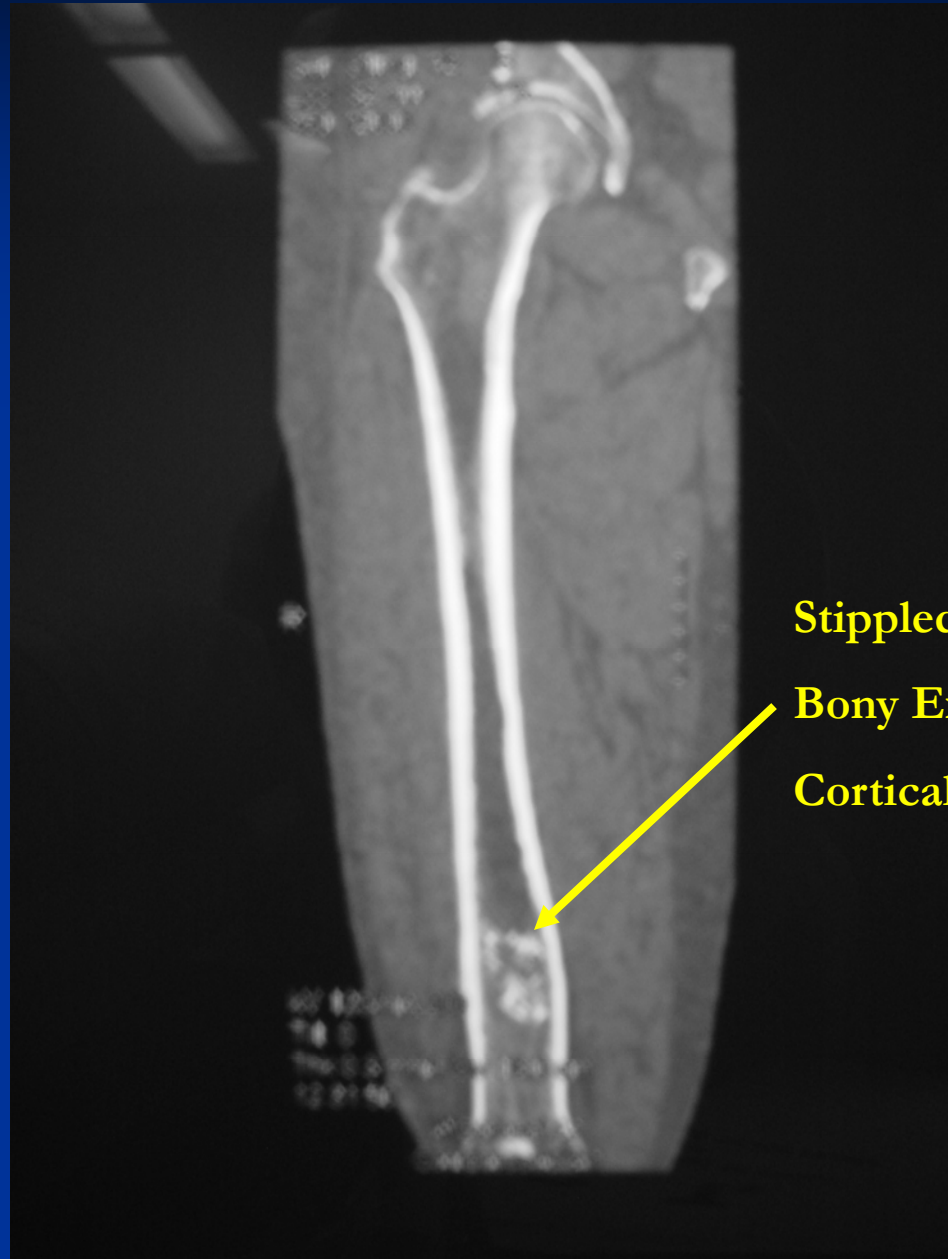
# MRI Low Grade Chondrosarcoma

## Endosteal Scalloping





# CT Scan: Grade I Chondrosarcoma



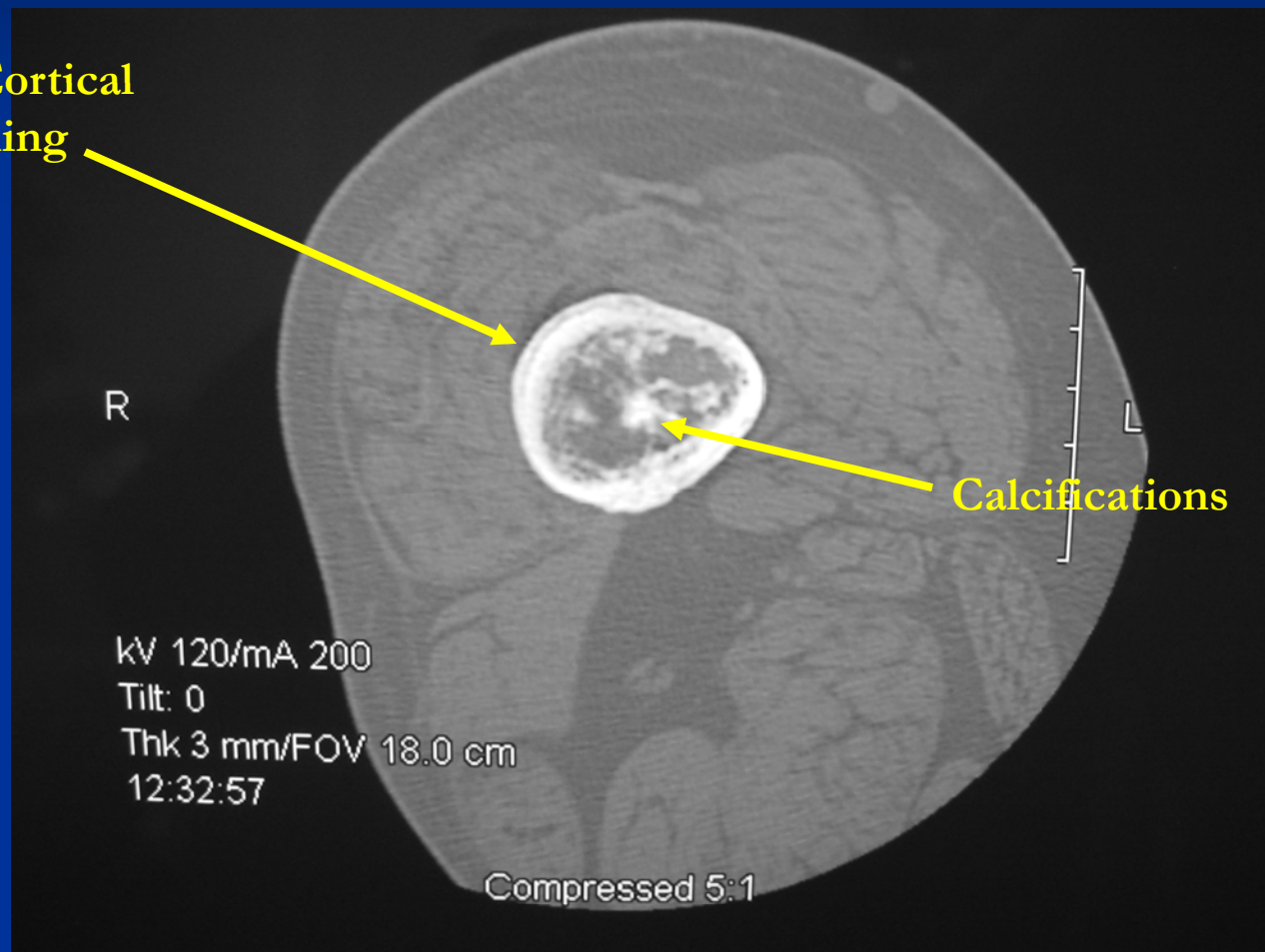
Stippled Calcifications

Bony Expansion

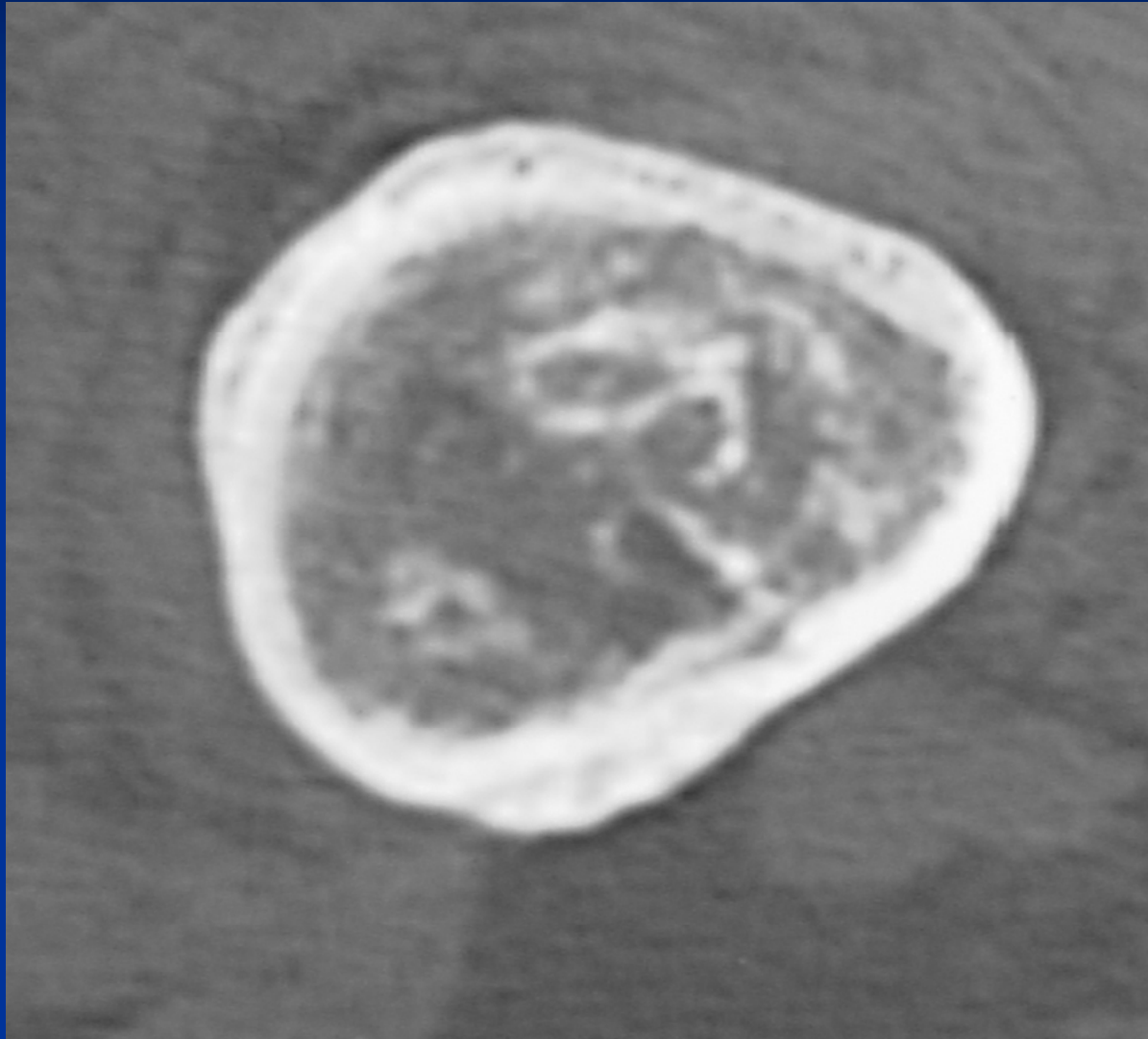
Cortical Thickening

# CT Scan Axial Section Grade I Chondrosarcoma

Subtle Cortical  
Thickening



# CT Scan: Grade I Chondrosarcoma



# Plain X-Ray/Bone Scan: Grade I Chondrosarcoma of Proximal Humerus

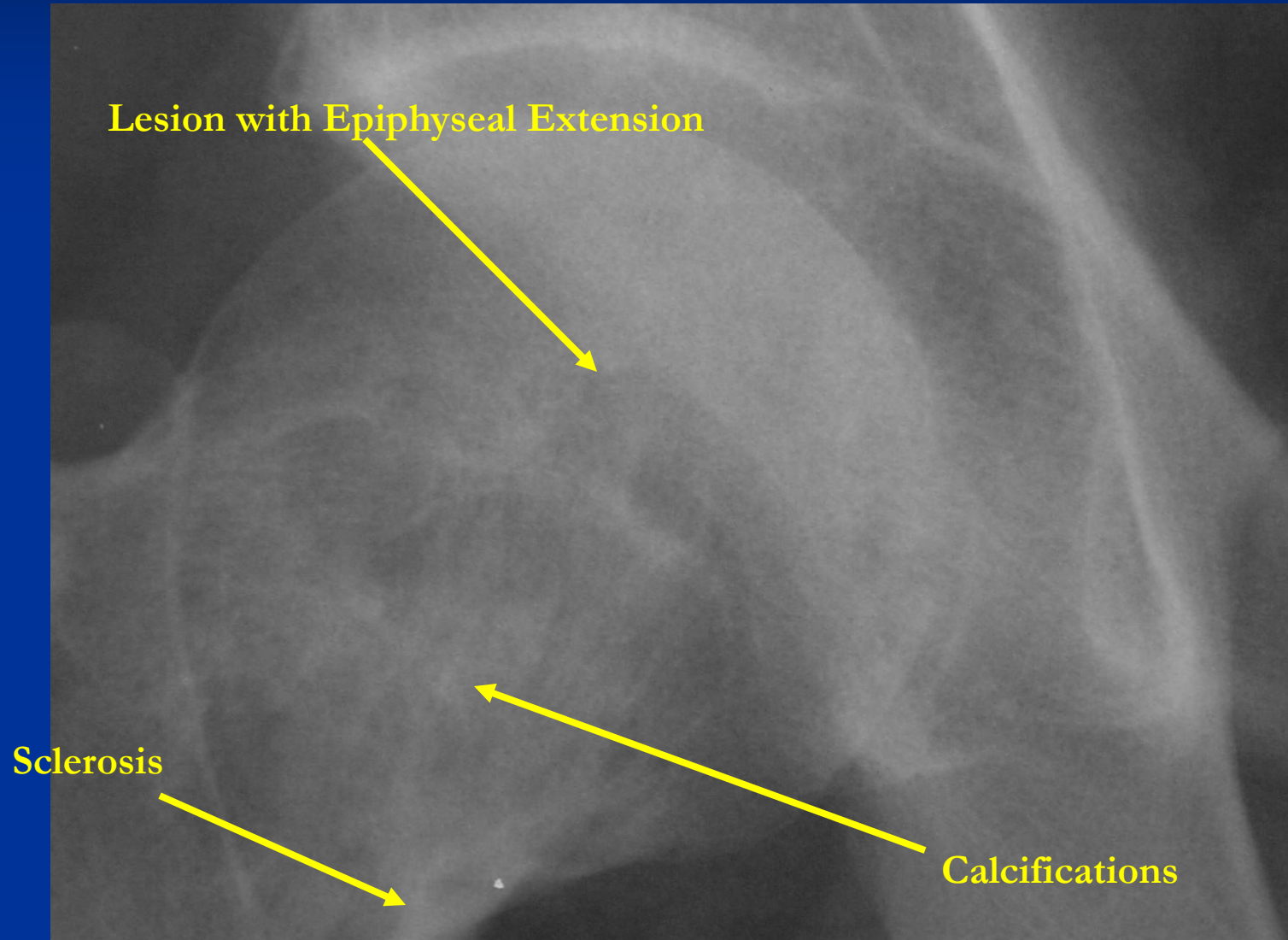
X-rays look identical to Enchondroma



Uptake Hotter than ASIS

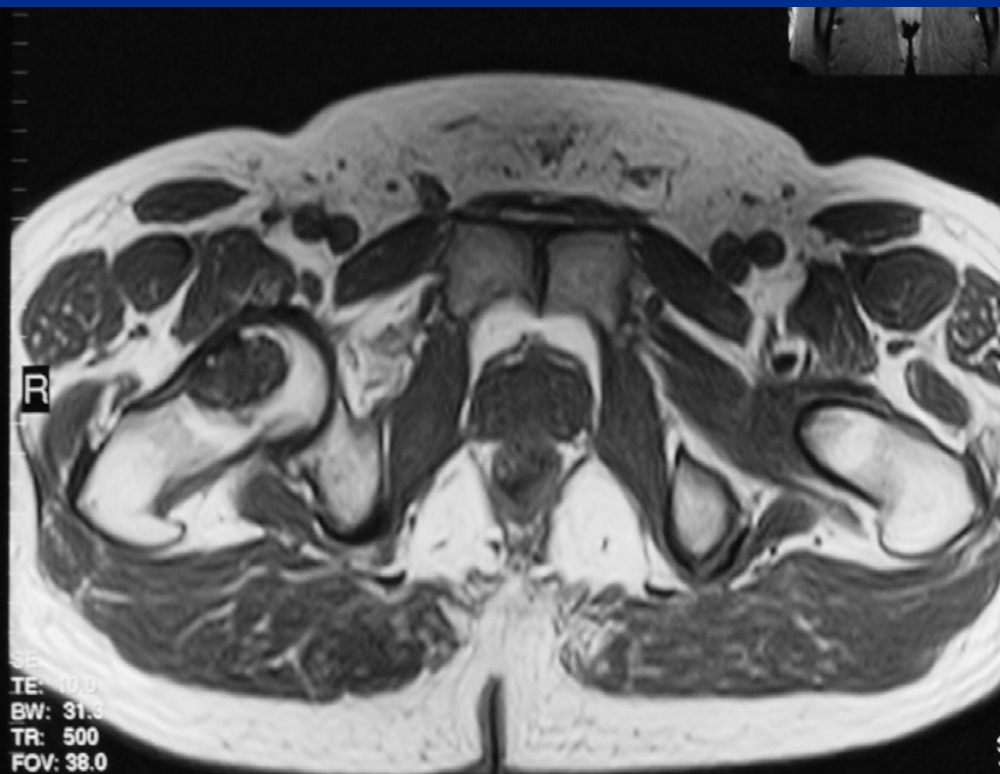


# Plain X-Ray: Grade I Chondrosarcoma of Femoral Neck

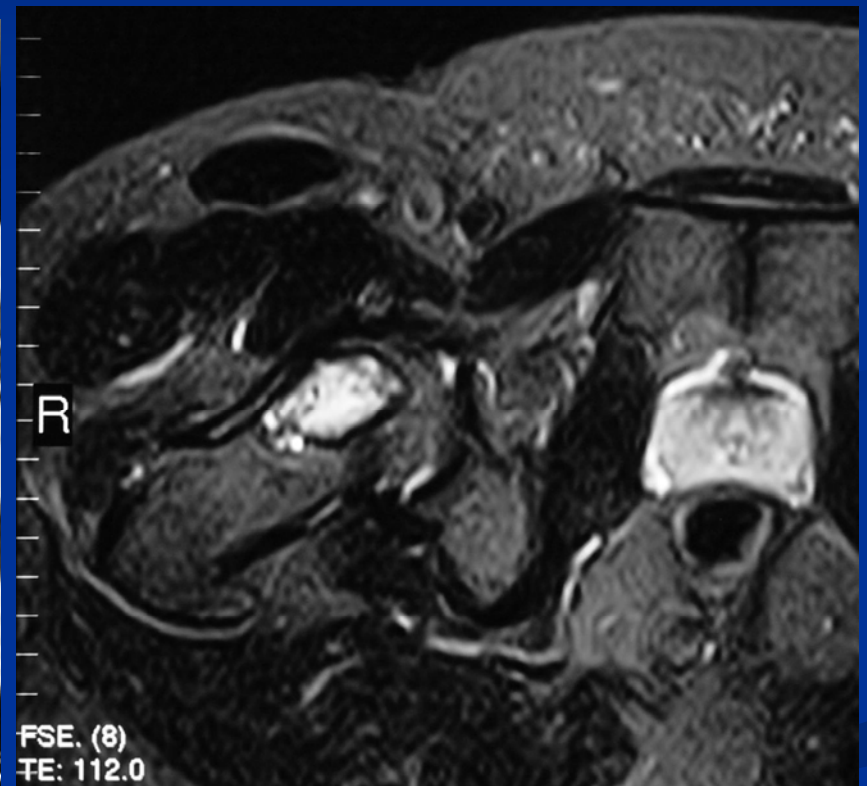




# MRI: Grade I Chondrosarcoma of Proximal Femur

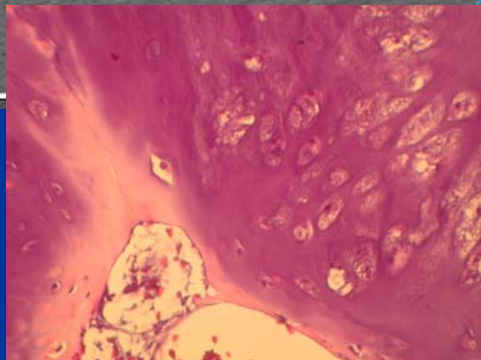
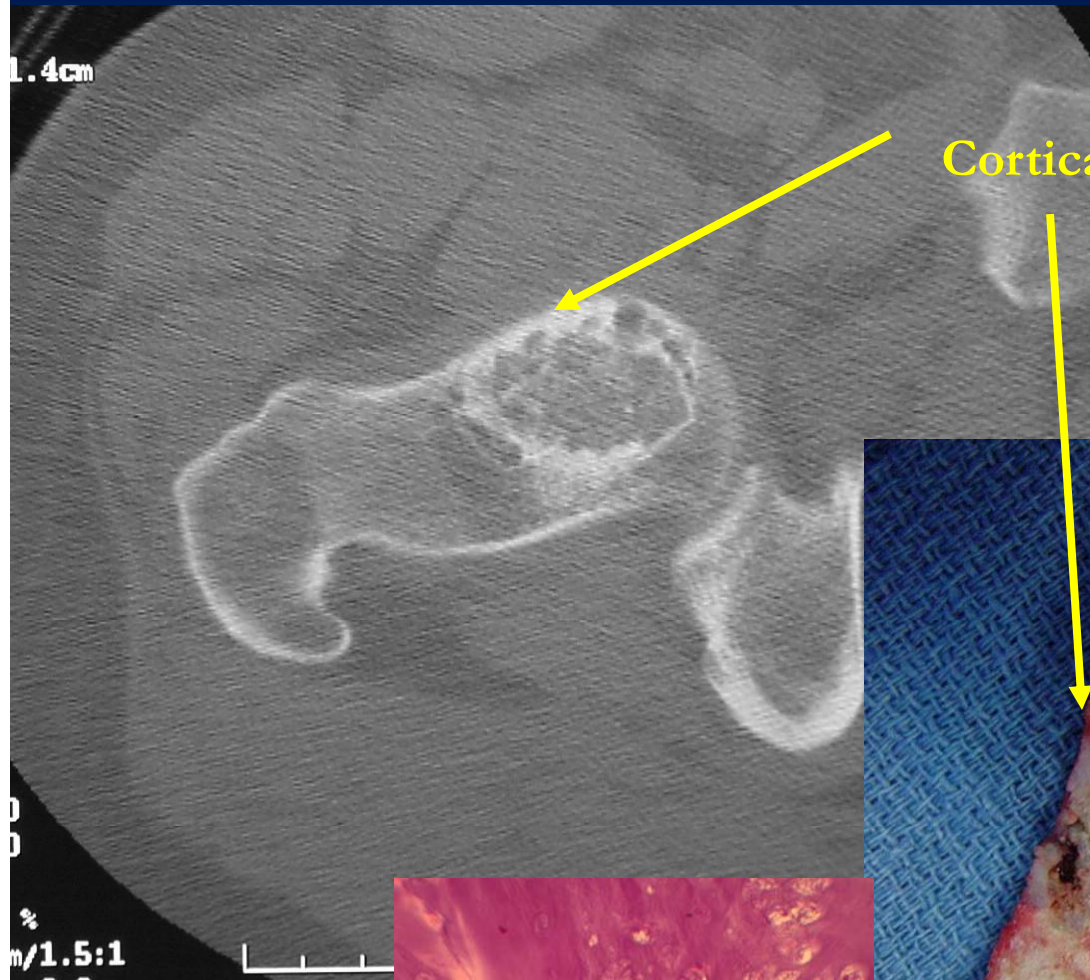


T1 Weighted Image



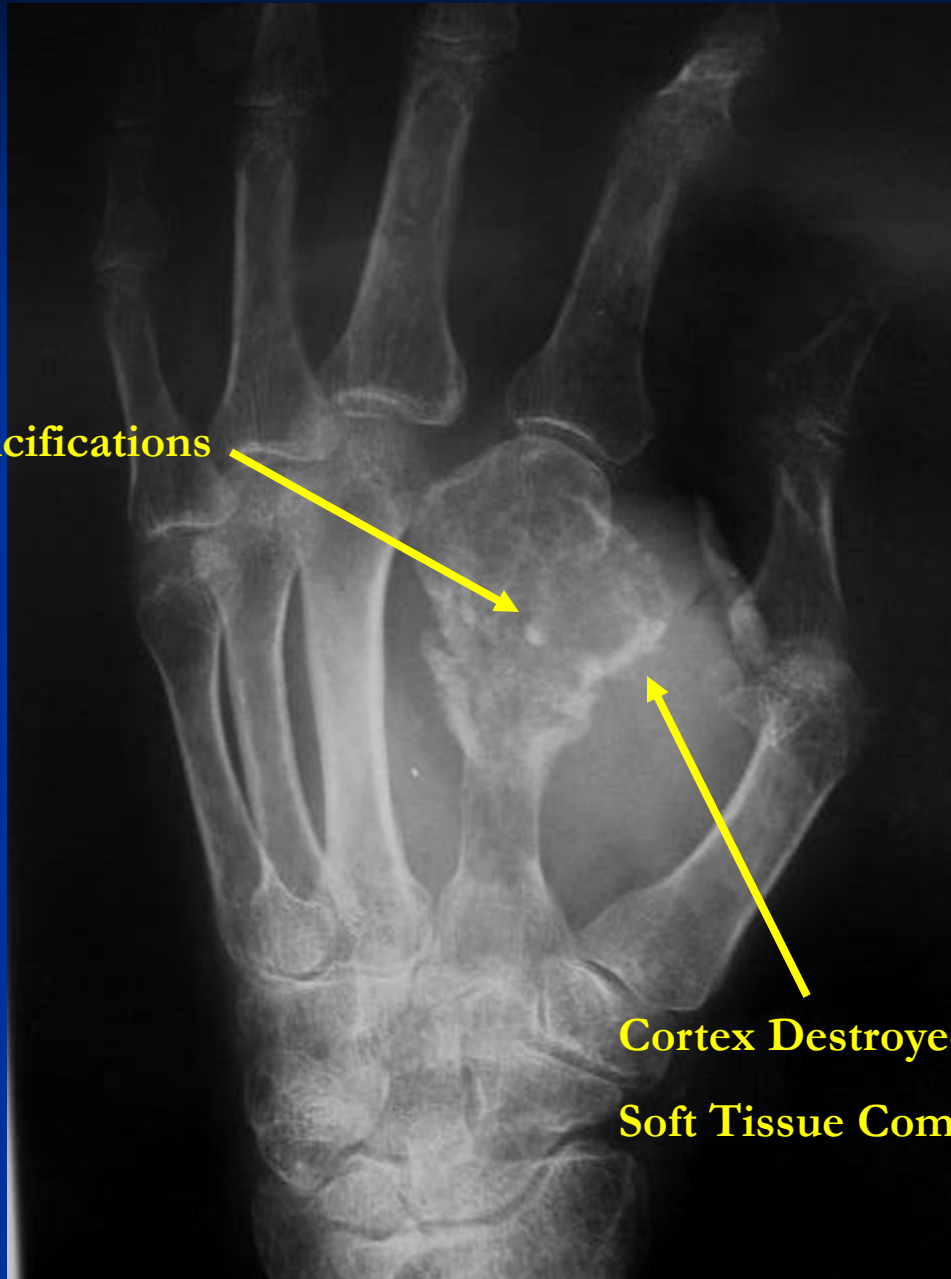
T2 Weighted Image

## CT Scan: Grade I Chondrosarcoma of Femoral Neck



## Plain X-Ray: Grade I Chondrosarcoma of Metacarpal of Hand

Stippled Calcifications



Cortex Destroyed

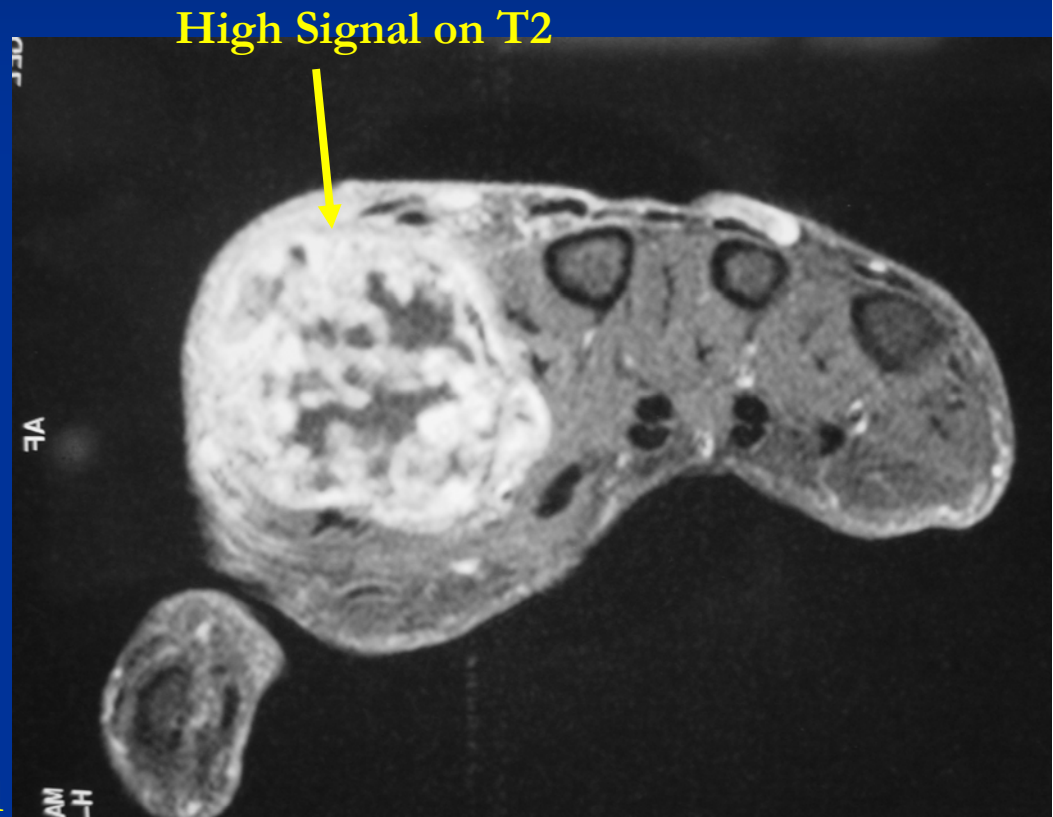
Soft Tissue Component



# MRI: Grade I Chondrosarcoma of Hand



## T1 Weighted Image

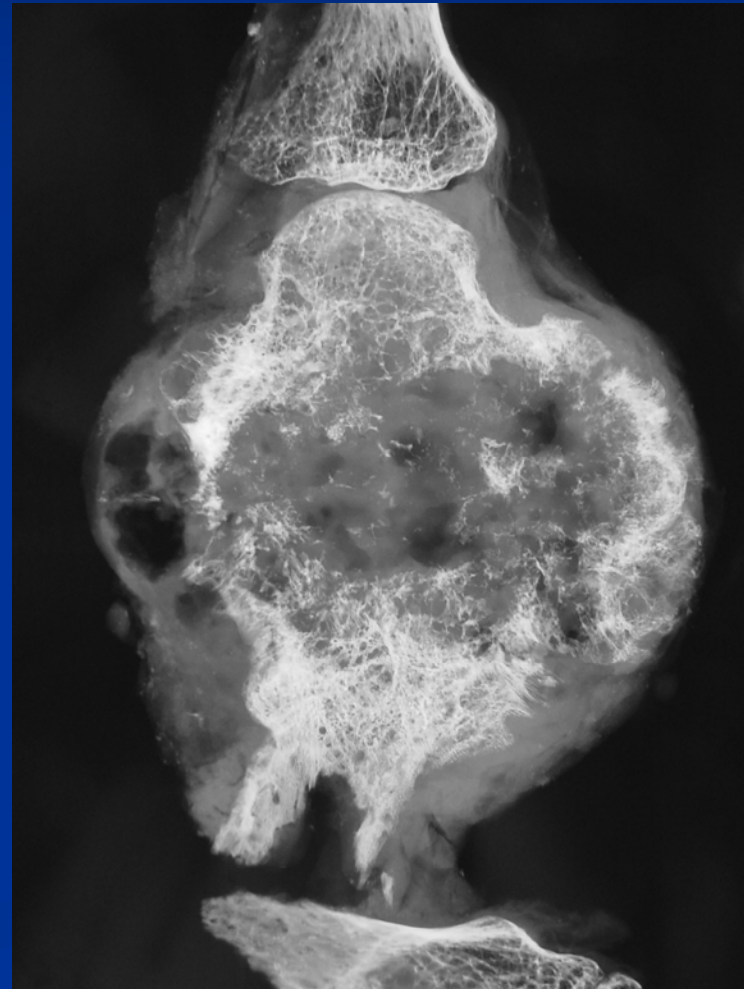
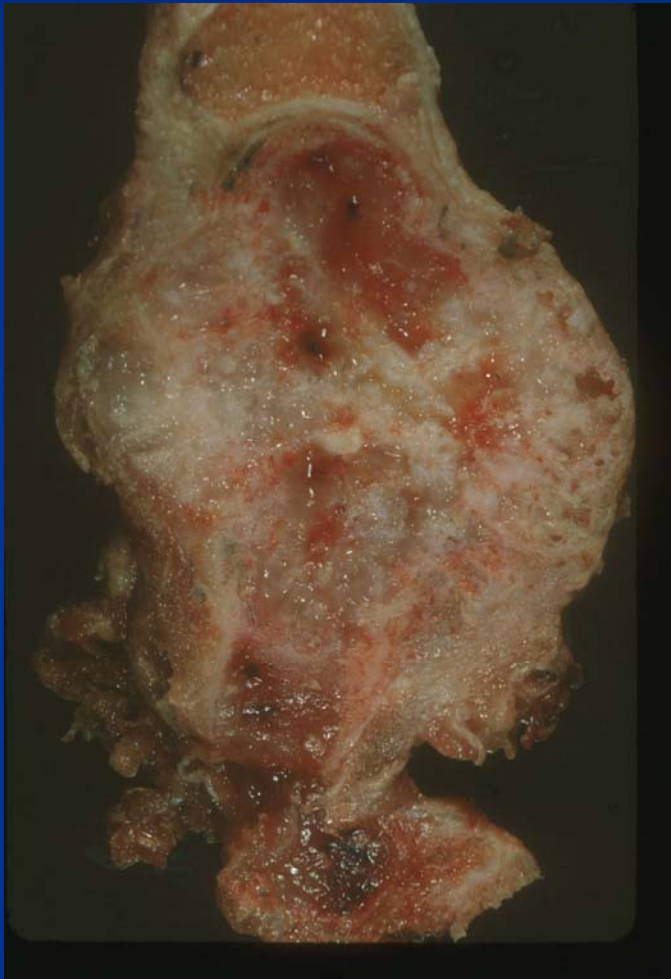


## T2 Weighted image

# Bone Scan: Chondrosarcoma of Metacarpal



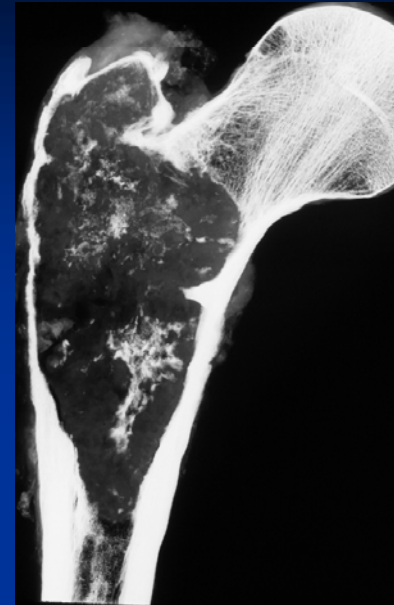
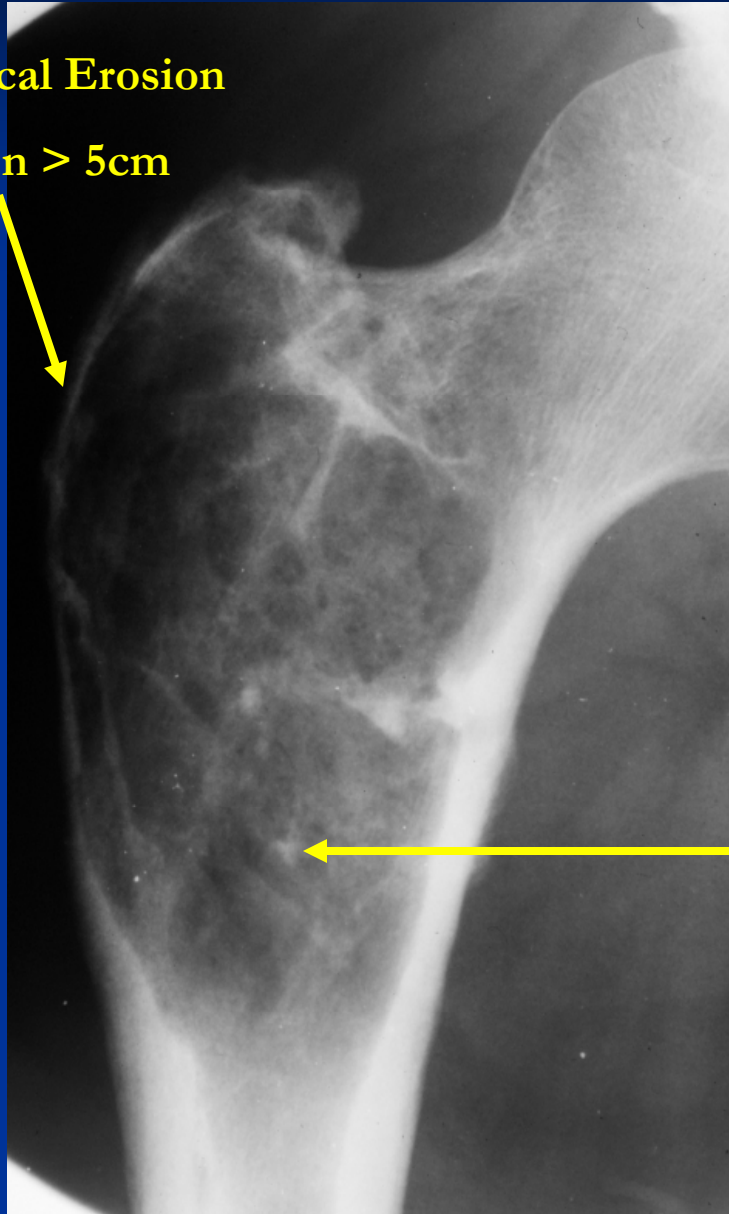
# Gross Specimen: Chondrosarcoma of Metacarpal



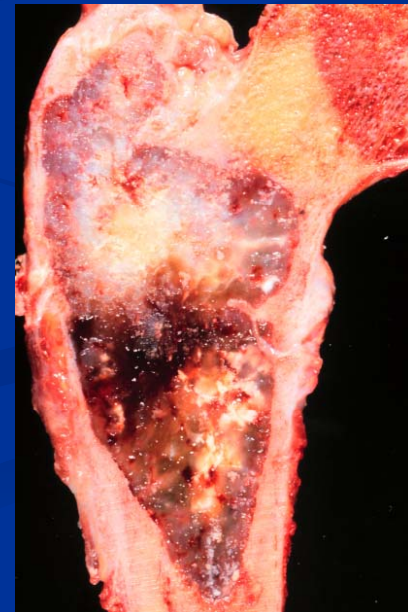
# Low Grade Chondrosarcoma

Cortical Erosion

Lesion > 5cm

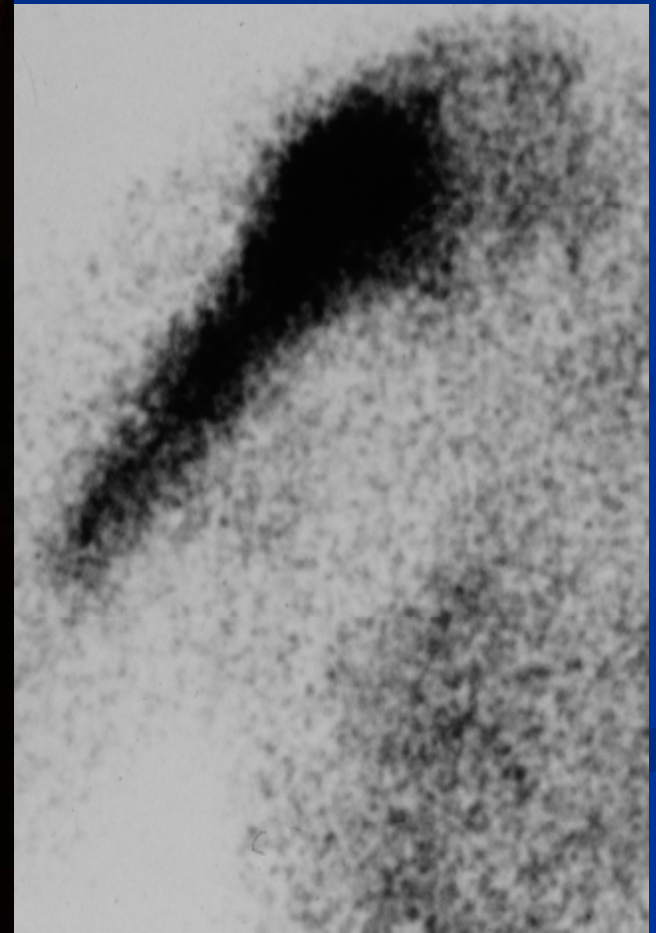
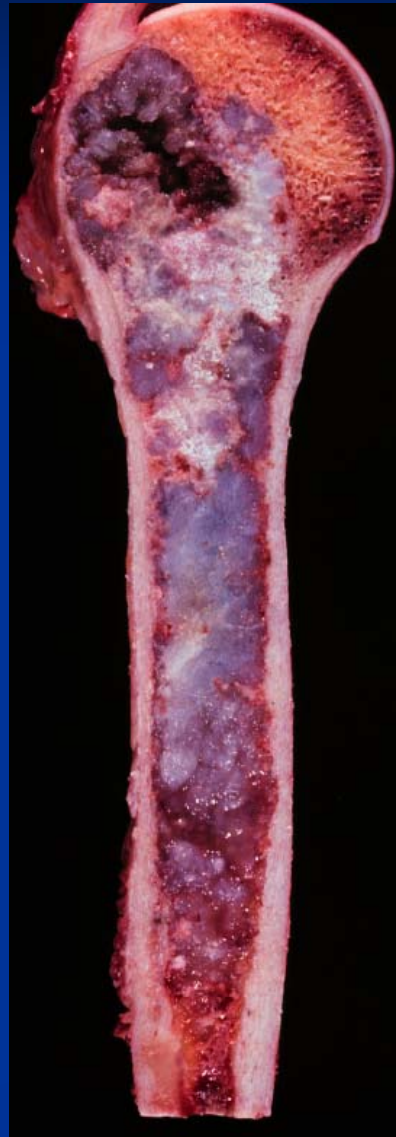
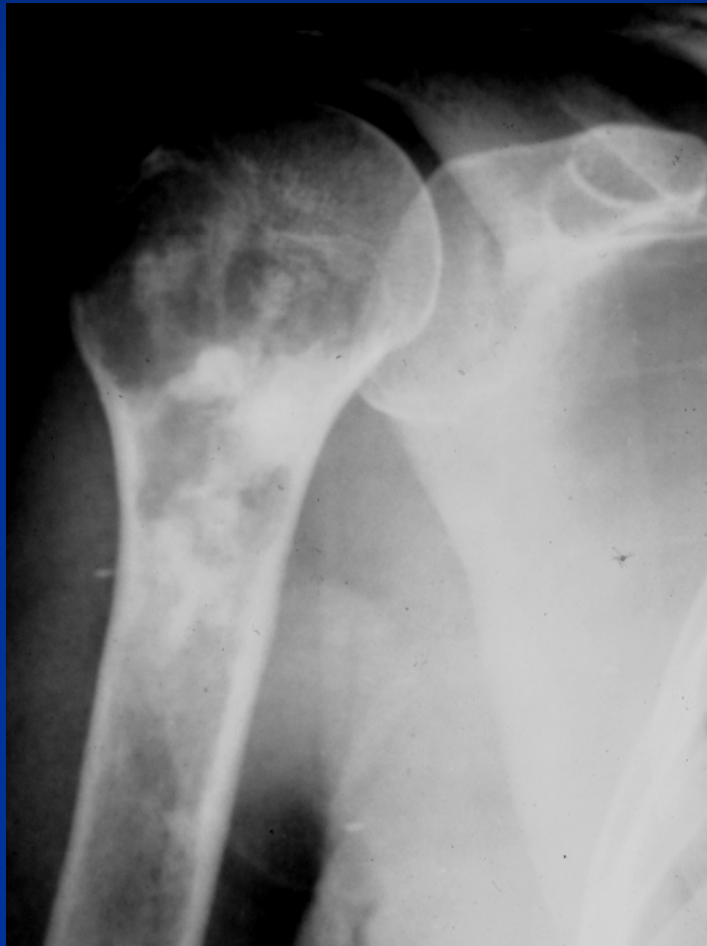


Calcifications

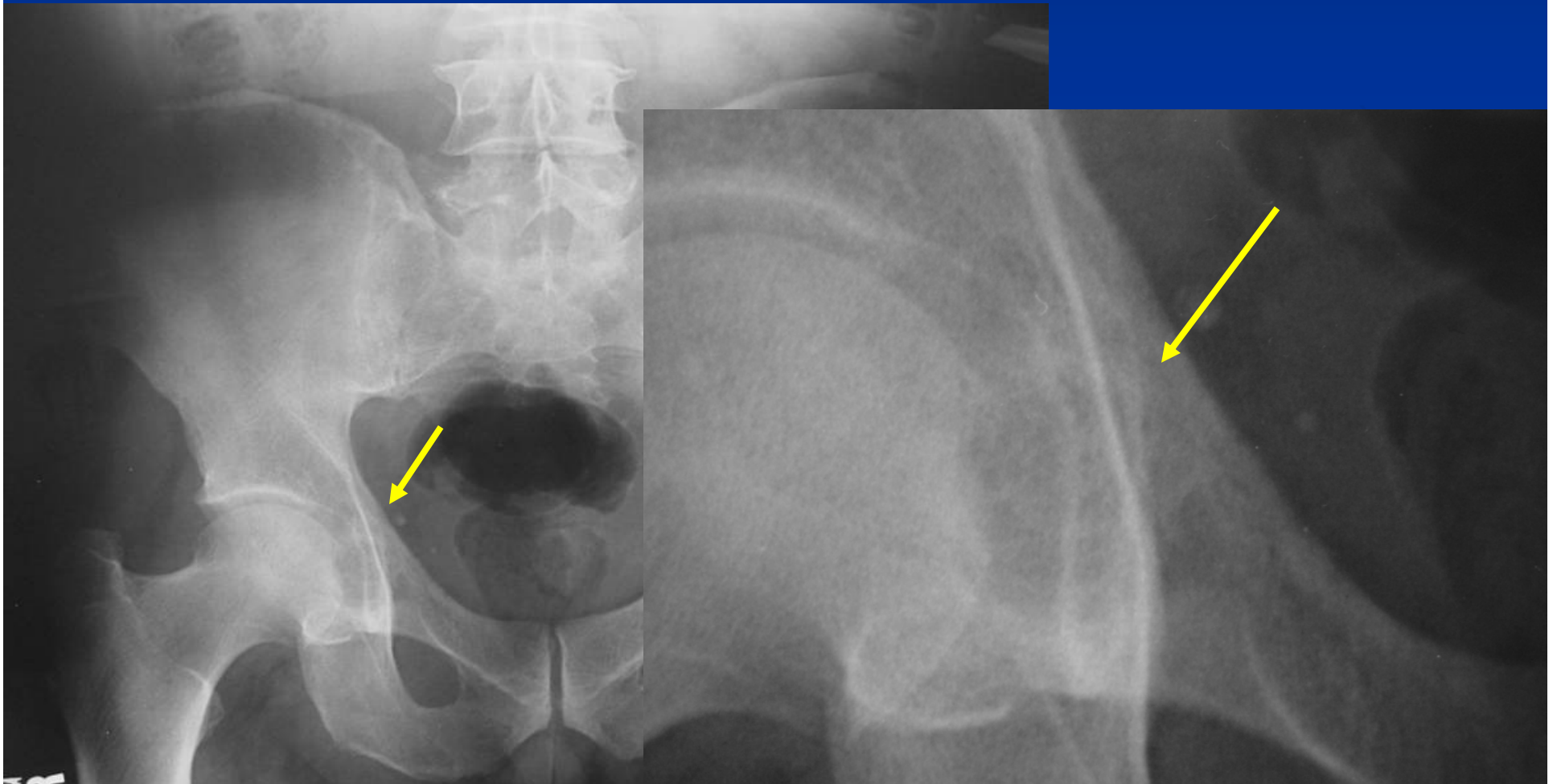




# Grade I Chondrosarcoma of Proximal Humerus



## Plain X-rays: Grade II Chondrosarcoma of Acetabulum



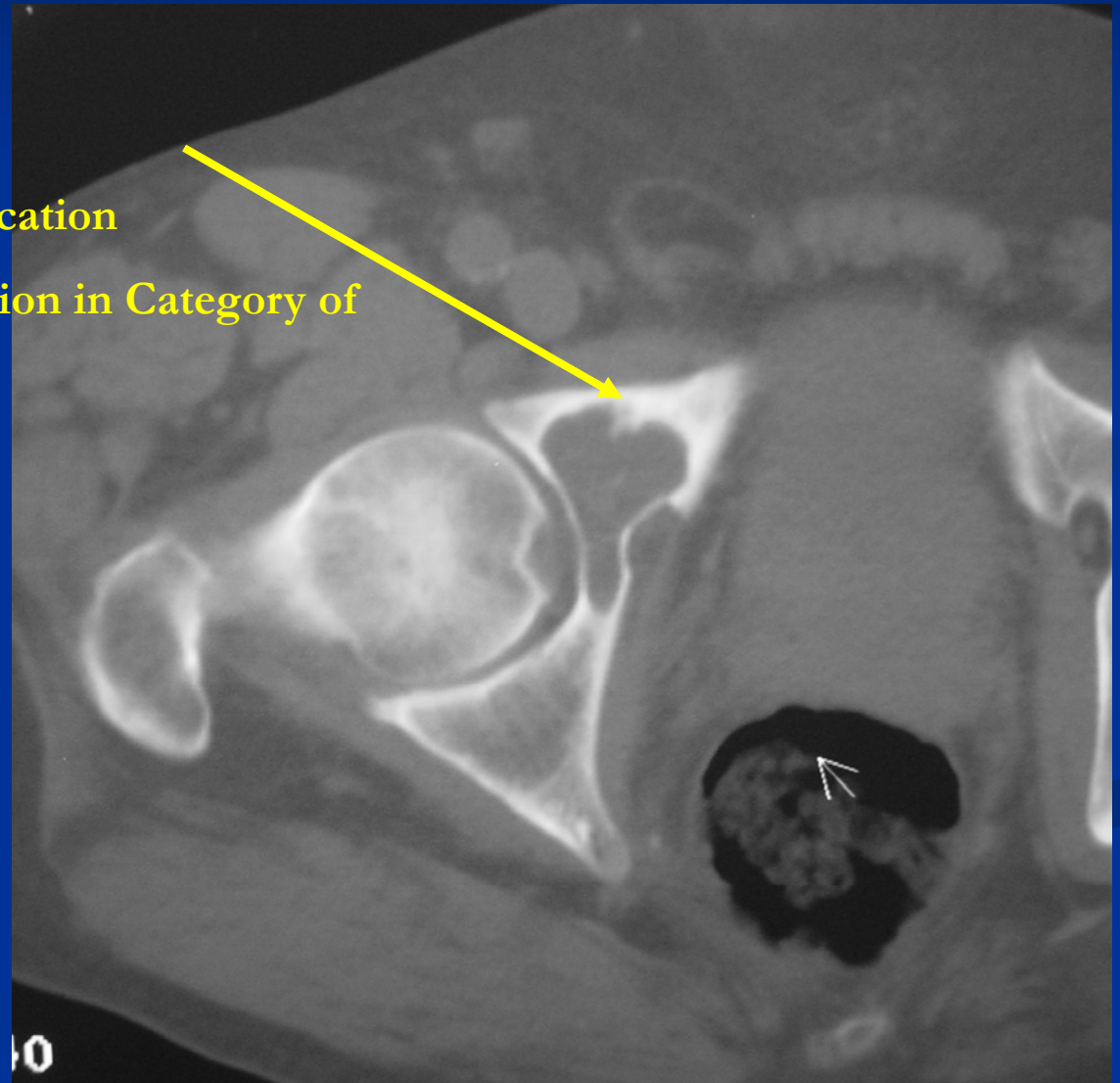
# CT Scan: Grade II Chondrosarcoma of Acetabulum

Lytic Lesion

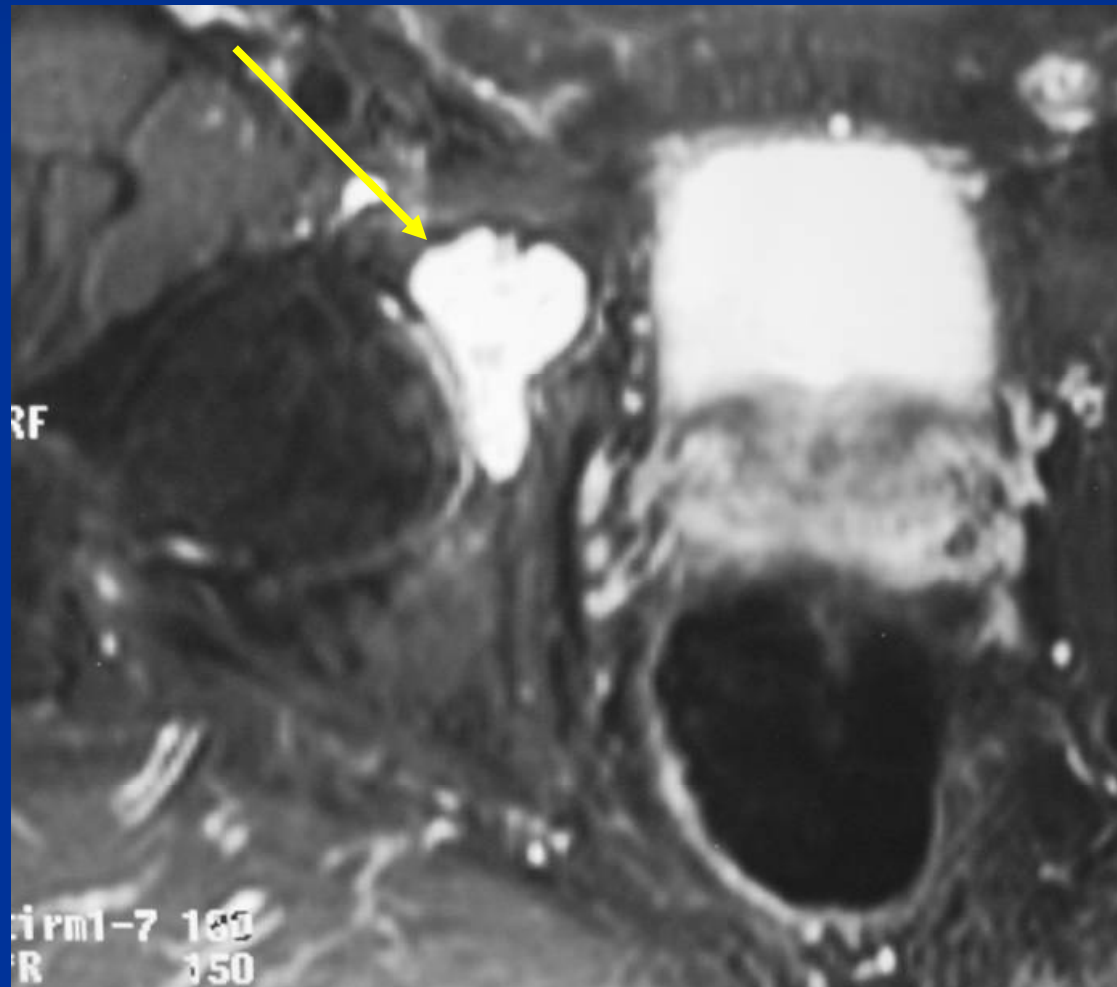
Surrounding Sclerosis

Subtle Intralesional Calcification

Pelvic Location Places Lesion in Category of  
Chondrosarcoma

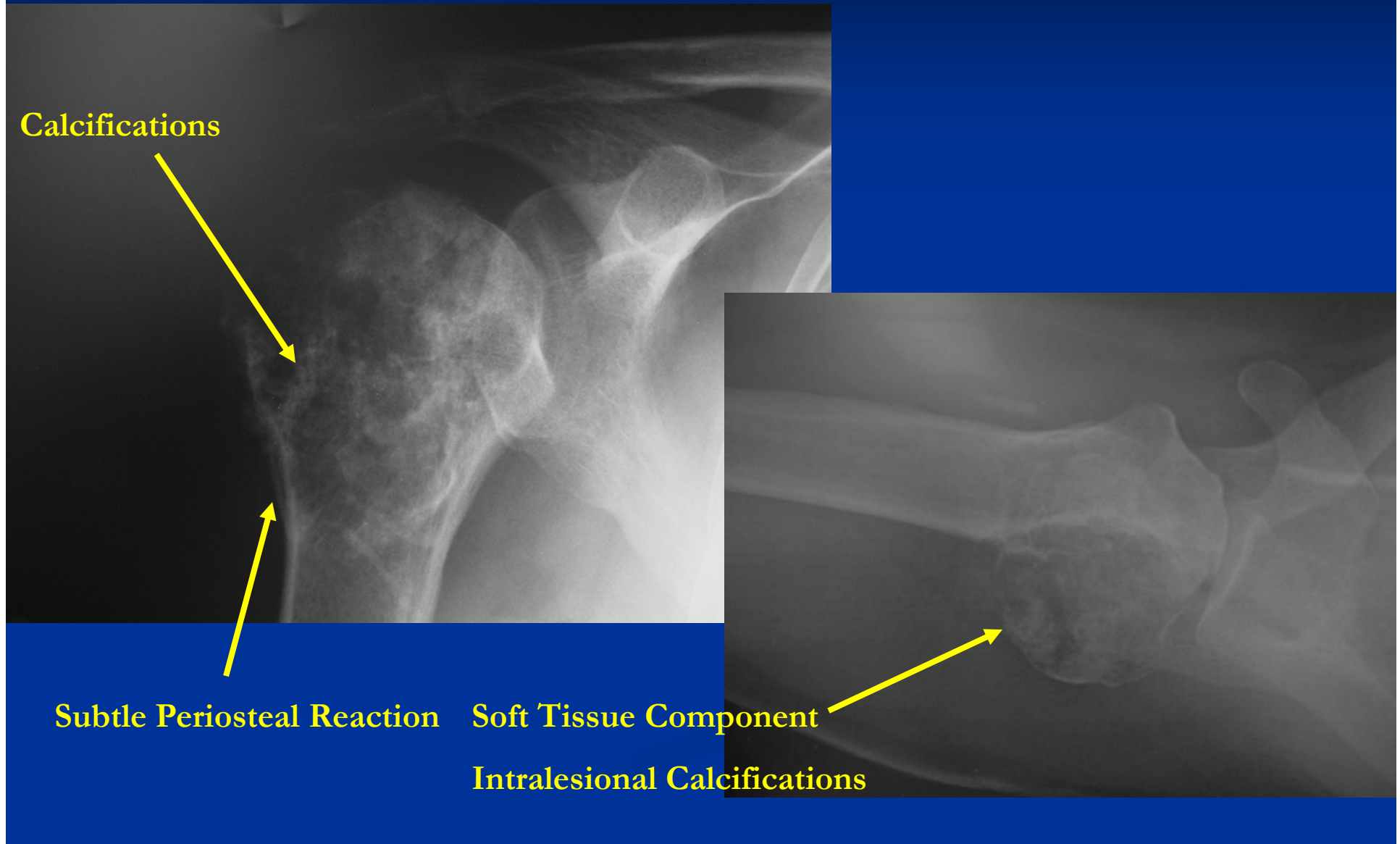


**MRI: Grade II Chondrosarcoma of Acetabulum**  
**High Signal on T2 may be misinterpreted as a cyst**

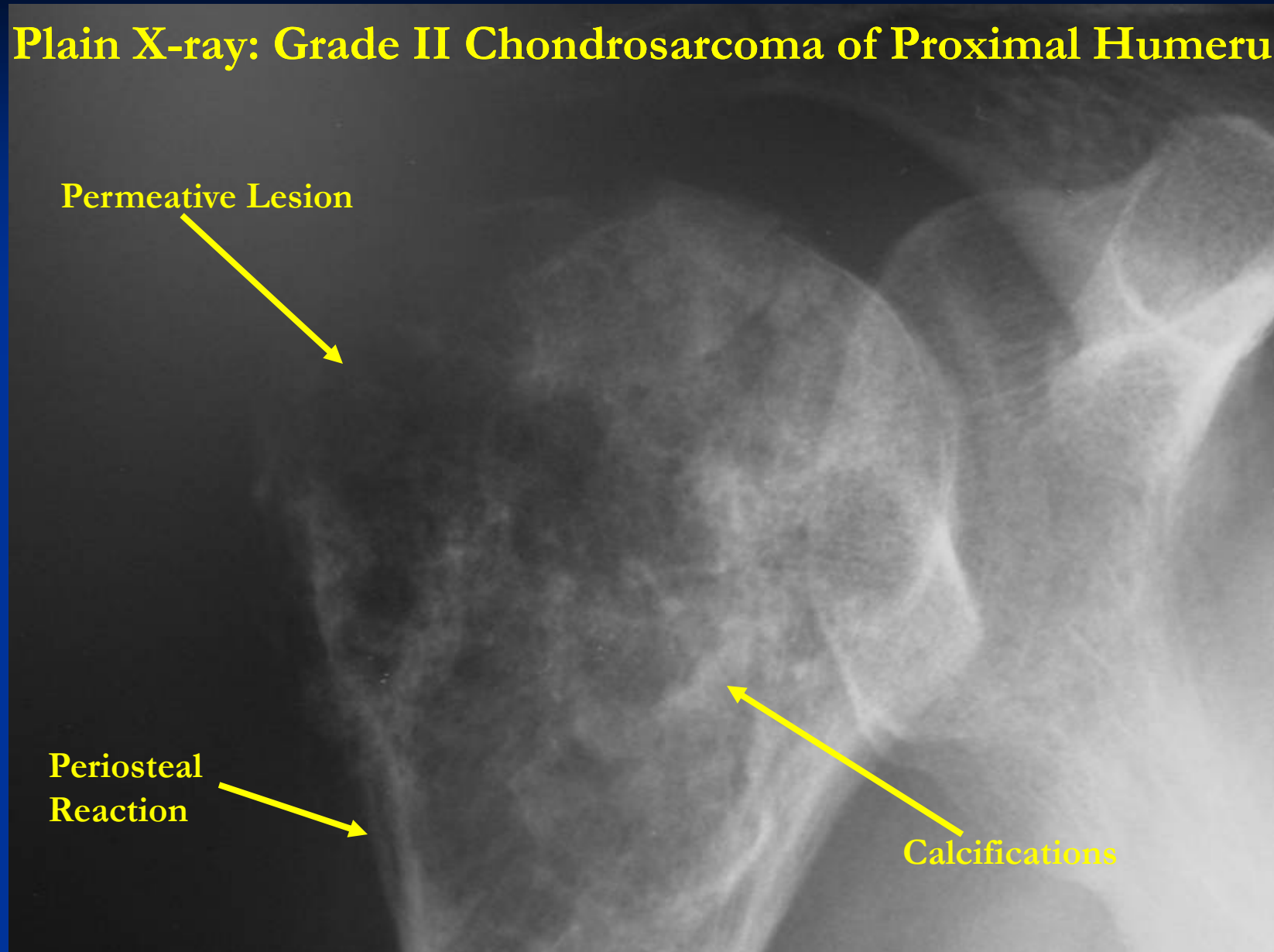




# Plain X-rays: Grade II Chondrosarcoma of Proximal Humerus



## Plain X-ray: Grade II Chondrosarcoma of Proximal Humerus

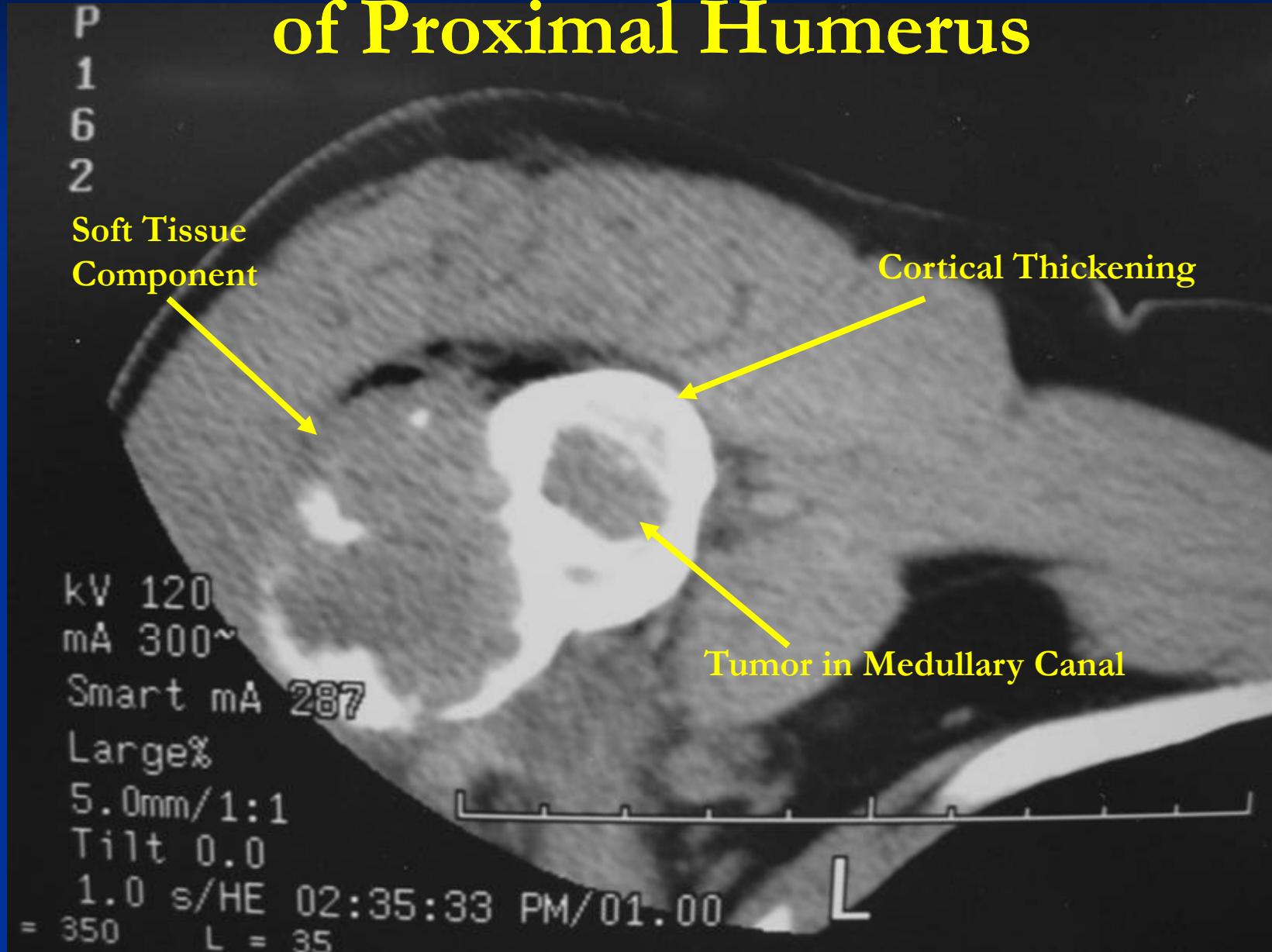


P  
1  
6  
2

kV 120  
mA 300~  
Smart mA 287  
Large%  
5.0mm/1:1  
Tilt 0.0  
1.0 s/HE 02:35:33 PM/01.00  
= 350 L = 35

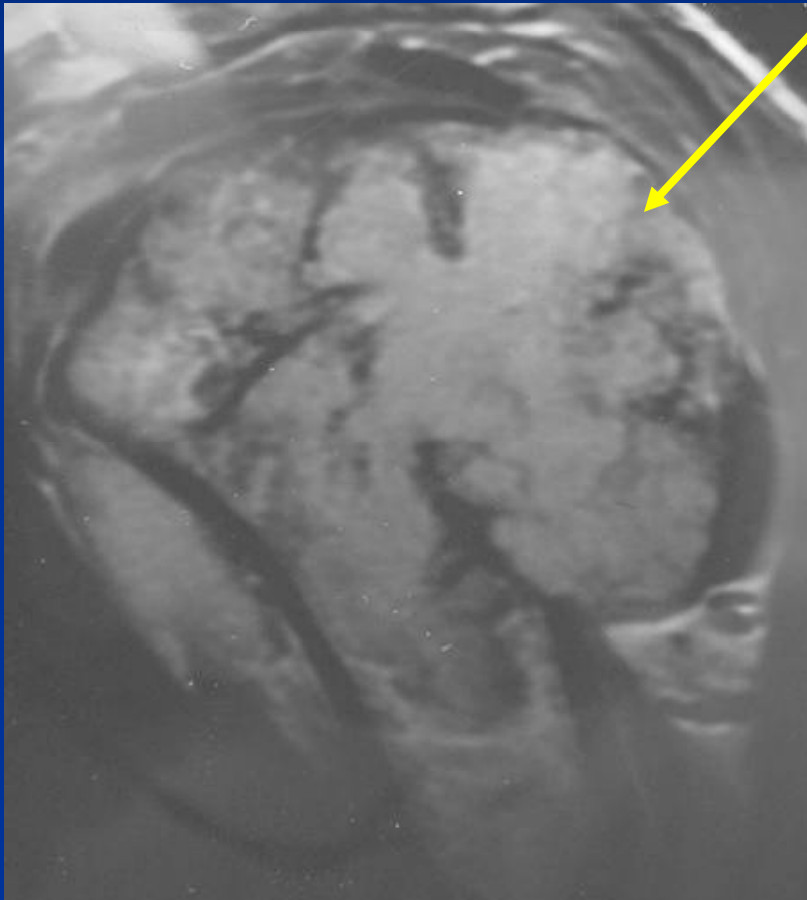


# CT Scan: Grade II Chondrosarcoma of Proximal Humerus



# MRI: Grade II Chondrosarcoma of Proximal Humerus

Soft Tissue Component Indicative of Chondrosarcoma



T1 Weighted Image

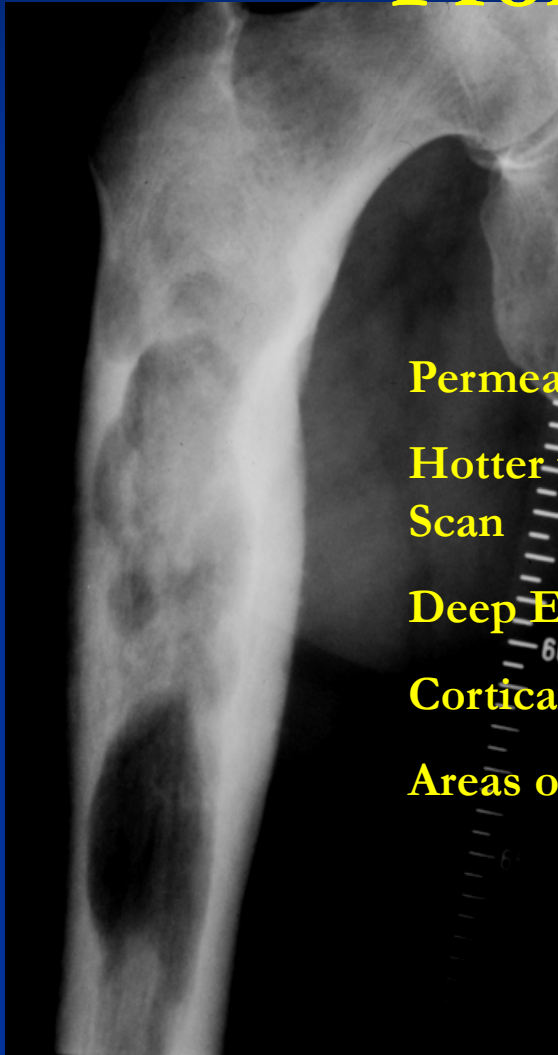
Cartilage Grows in Lobular Manner



T2 Weighted Image

Cartilage is High signal on T2

# Plain X-Ray and Bone Scan Grade II Chondrosarcoma of Proximal Femur



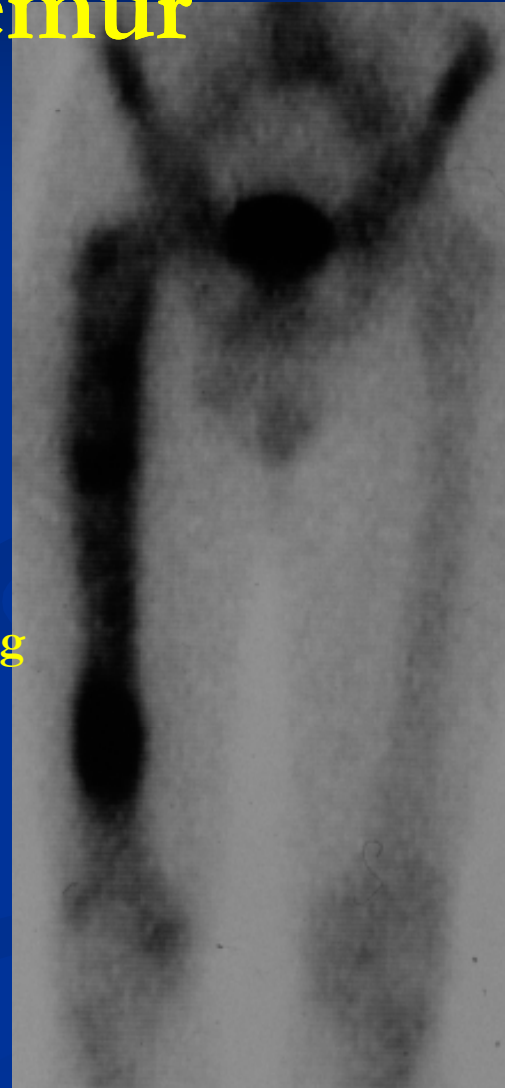
Permeative Lesion >5cm

Hotter than ASIS on bone  
Scan

Deep Endosteal Scalloping

Cortical Thickening

Areas of Lysis





# MRI: grade II Chondrosarcoma of Proximal Femur



## MRI: Grade II Chondrosarcoma of Proximal Femur



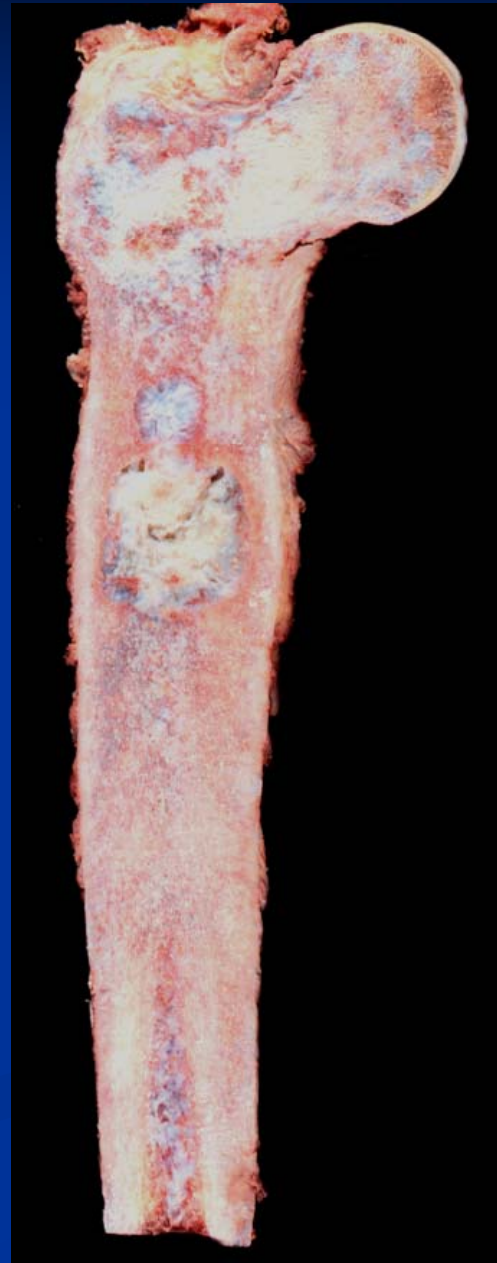
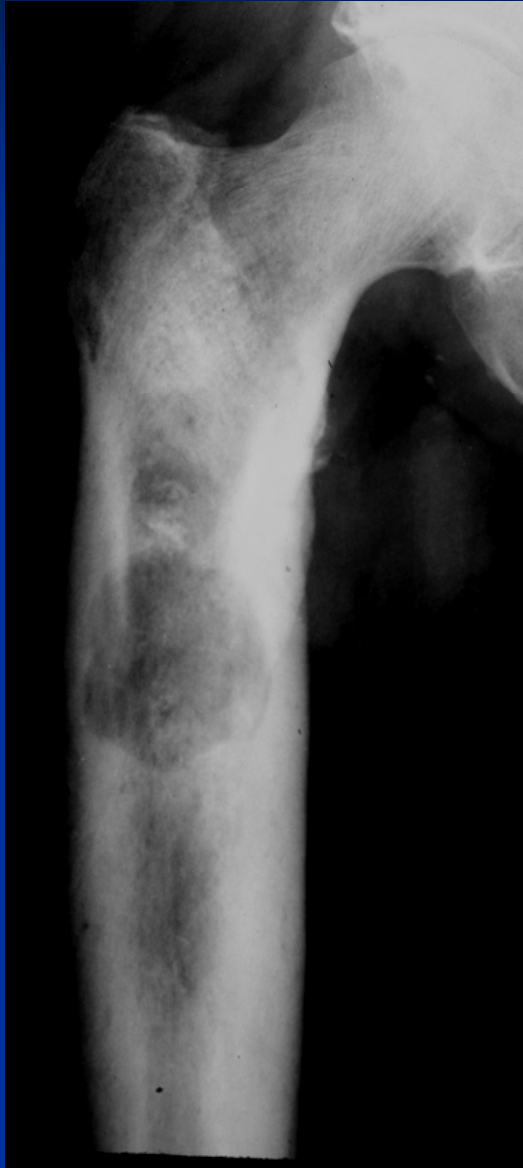
T1 Image



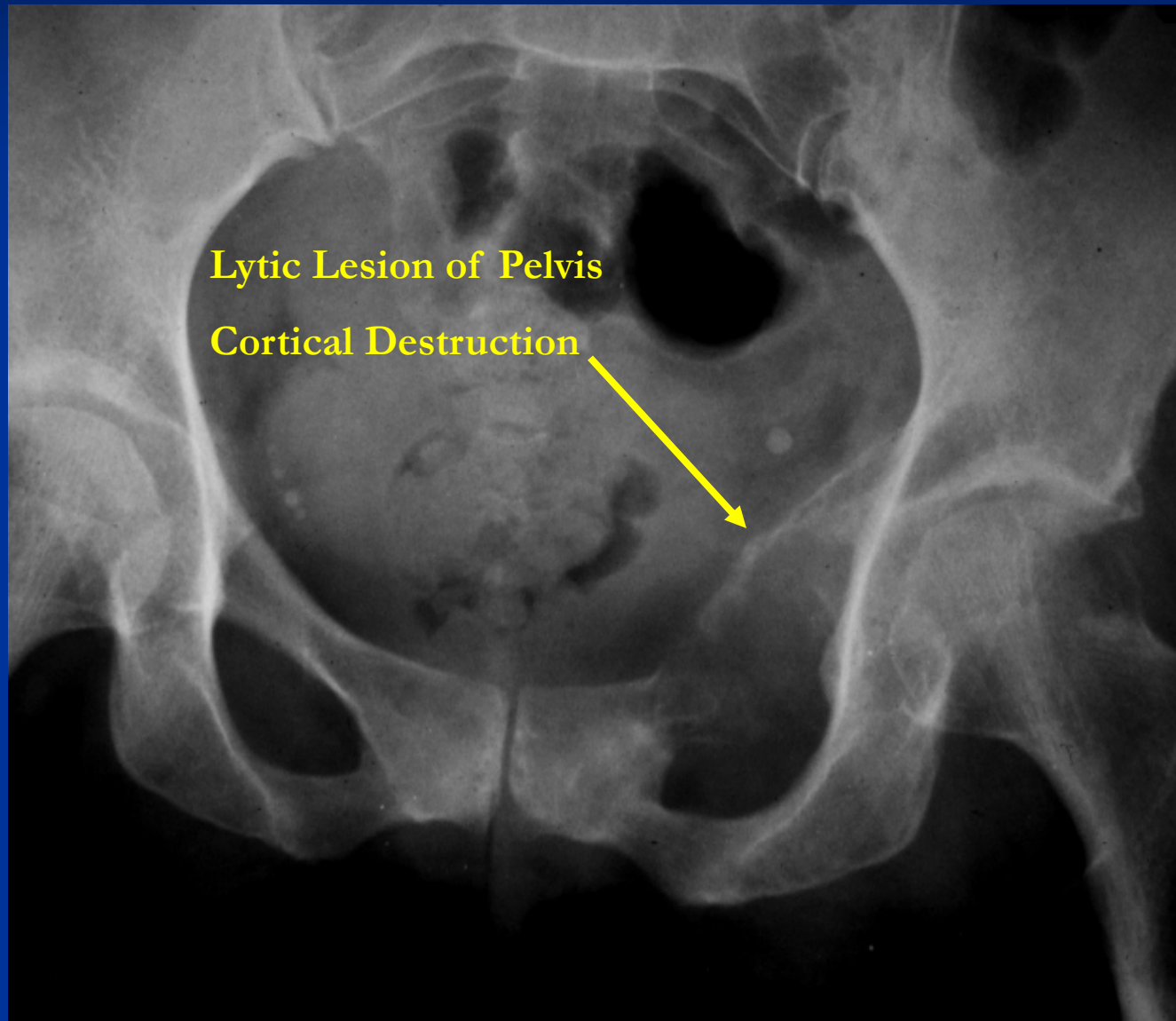
T2 Image

Soft Tissue Component not detected on X-ray

## Plain Xray: Grade II Chondrosarcoma of Proximal Femur



## Plain X-ray: Grade III Conventional Chondrosarcoma of Pelvis





# Secondary Chondrosarcoma

- Secondary Chondrosarcomas arise from a pre-existing lesion such as an osteochondroma or enchondroma
- Most commonly secondary chondrosarcomas arise from osteochondromas
- Most secondary chondrosarcomas are low grade and cured by wide excision
- Secondary chondrosarcomas can also dedifferentiate into high grade spindle cell sarcomas or become higher grade lesions if left untreated for prolonged periods of time
- Secondary chondrosarcomas are more likely to occur in setting of multiple hereditary exostoses and enchondromatosis. Osteochondromas that arise from the scapula, ribs and pelvis and proximal femur are the most likely to undergo malignant change or dedifferentiate.



# Osteochondroma vs. Secondary Chondrosarcoma

- The cartilaginous cap deserves the most attention when differentiating a benign osteochondroma from a secondary chondrosarcoma that arose from a pre-existing osteochondroma
- In adults, the cartilaginous cap regresses and becomes thin due to enchondral ossification of the majority of the cap.
- Malignant transformation is suggested by:
  - Cartilaginous cap thickness greater than 2cm
  - Cortical destruction
  - Backgrowth of the cartilaginous cap into the stalk or medullary canal
  - Lysis of calcifications in cap

# Osteochondroma: Cartilage Cap

## ■ Radiographs

- Chondroid Calcification in cap
- Increasing destruction or change in appearance is worrisome for malignancy

## ■ **MRI:** Best test for evaluating thickness of cap and surrounding bursa

- Intermediate T1W Images
- High Intensity T2W Images because of fluid content

# Osteochondroma: Cartilage Cap

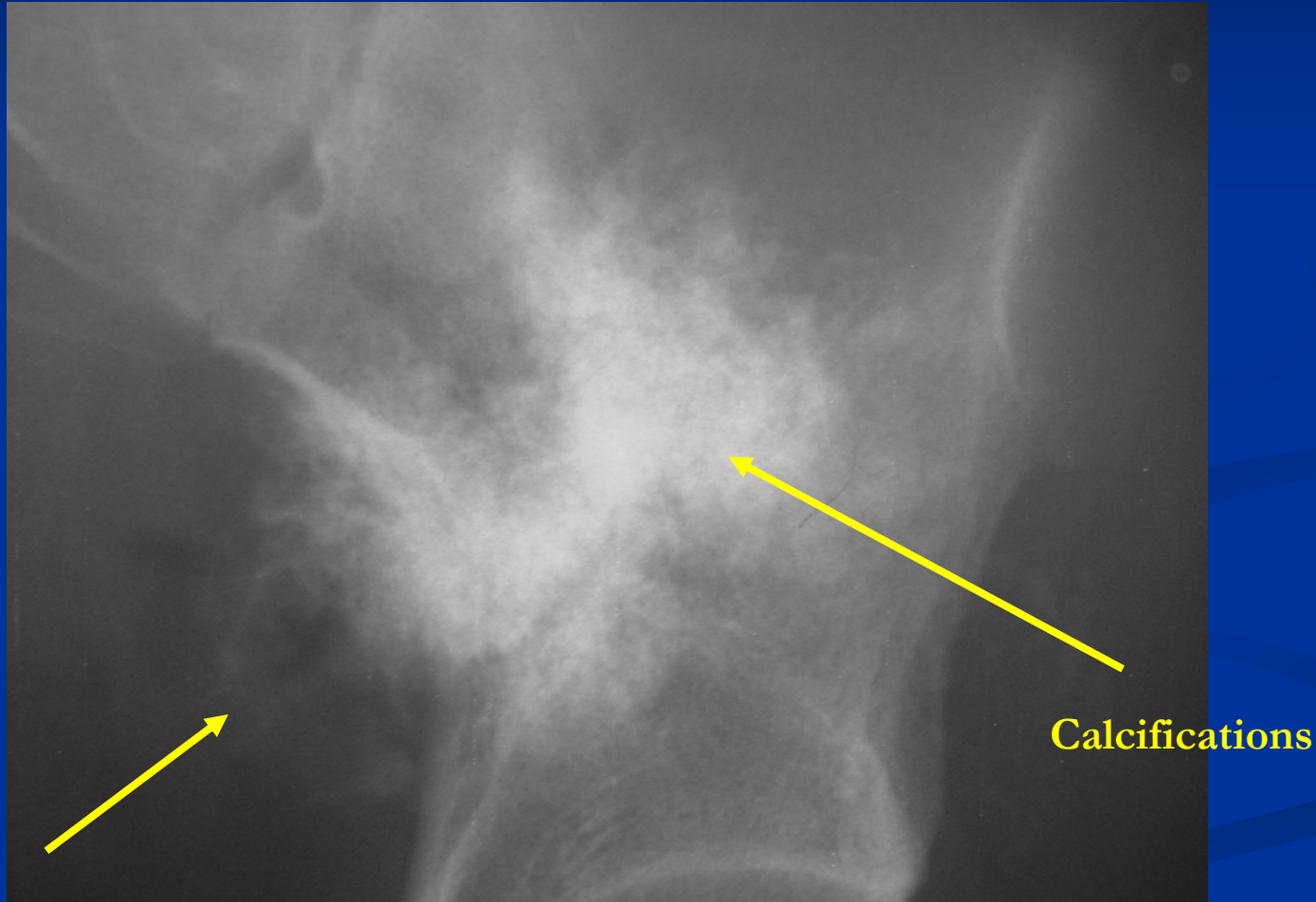
## ■ CT

- The cap will appear as soft tissue with calcification
- Peripheral stippled calcifications suggest malignancy or lysis adjacent to well calcified area
- Can be difficult to distinguish from muscle

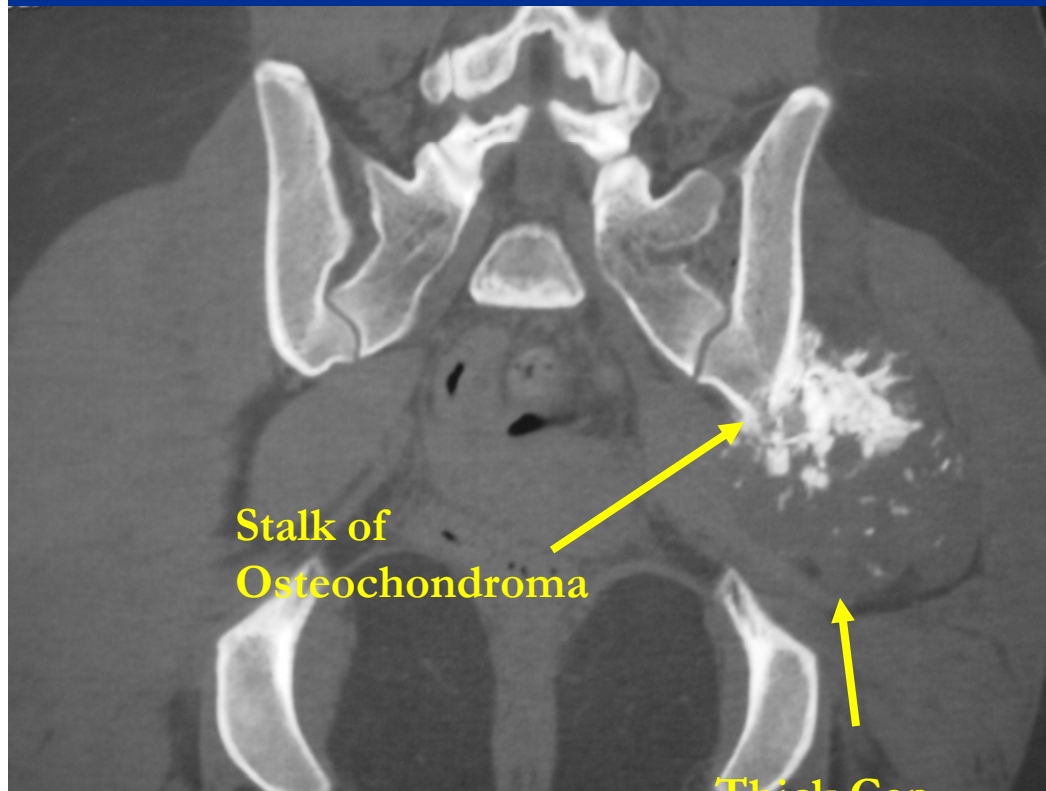
## ■ Cap thickness

- Benign  $< 1.5\text{cm}$  (0.1 – 3.0cm; Avg. 0.6 – 0.9 cm)
- Malignant  $> 1.5\text{ cm}$  (1.5 – 12cm; Ave. 6cm)

## Plain X-ray: Secondary Chondrosarcoma of Pelvis



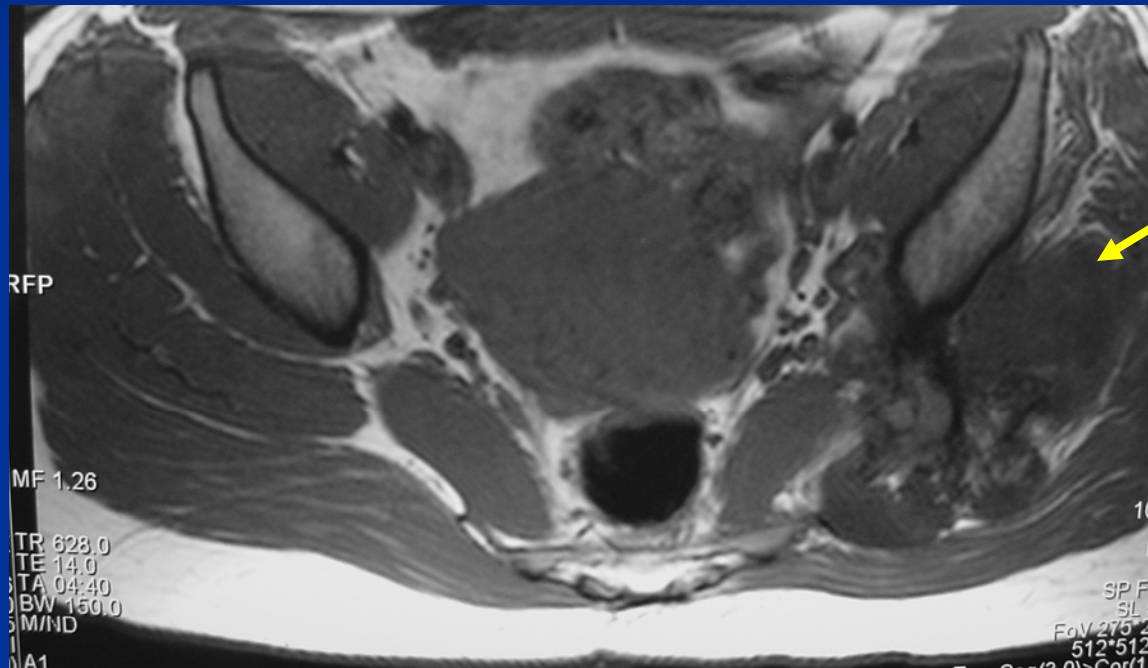
# Secondary Chondrosarcoma of Pelvis



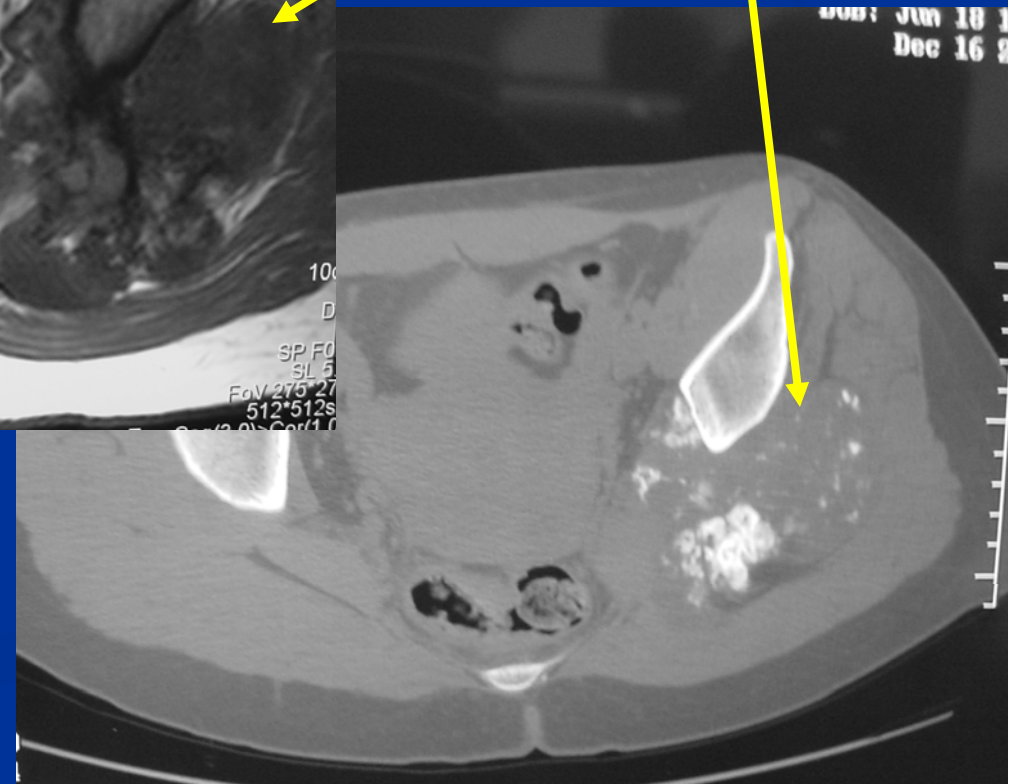
Peripheral Calcifications



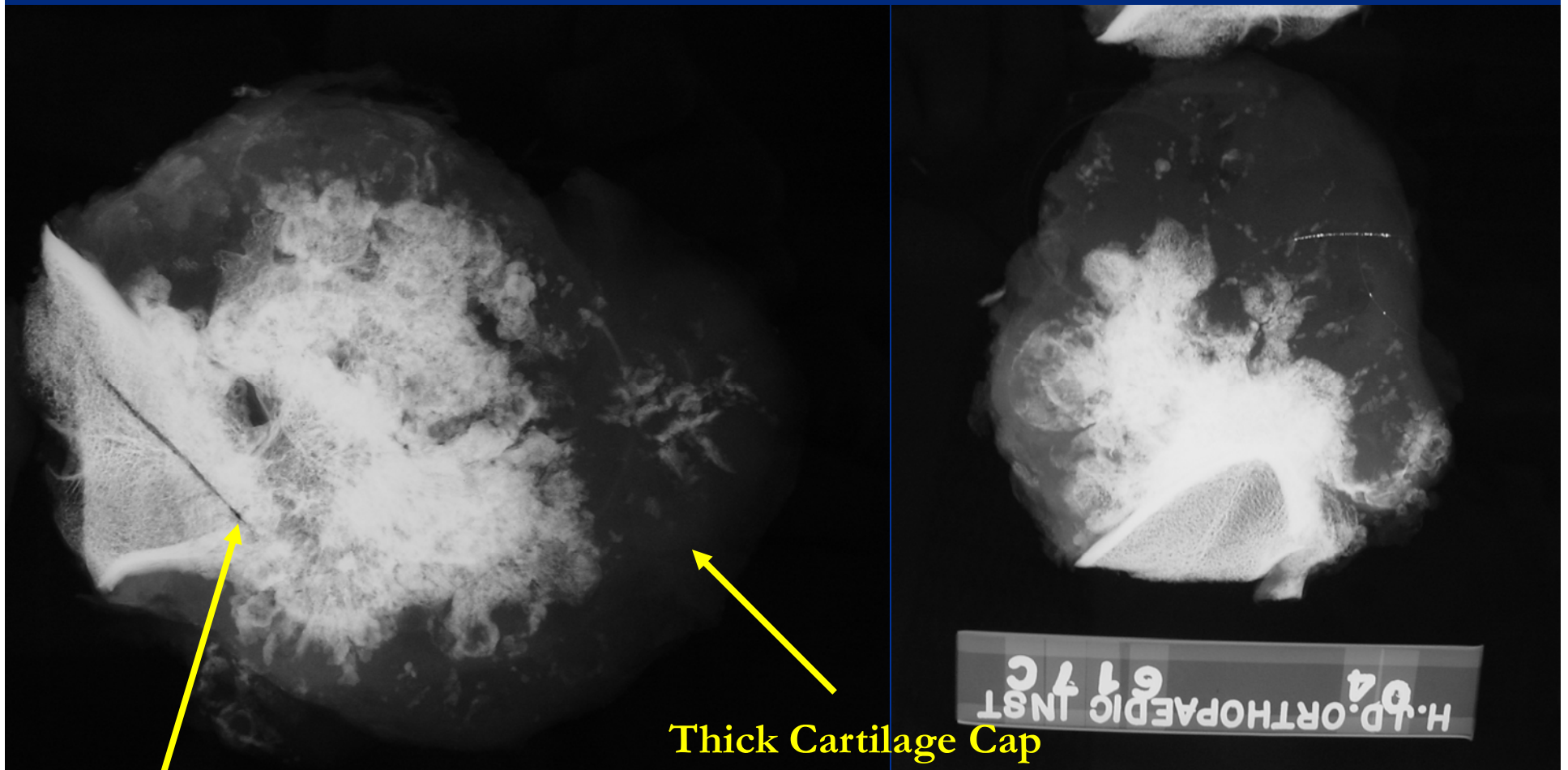
# MRI and CT: Secondary Chondrosarcoma



Thick Cartilage Cap



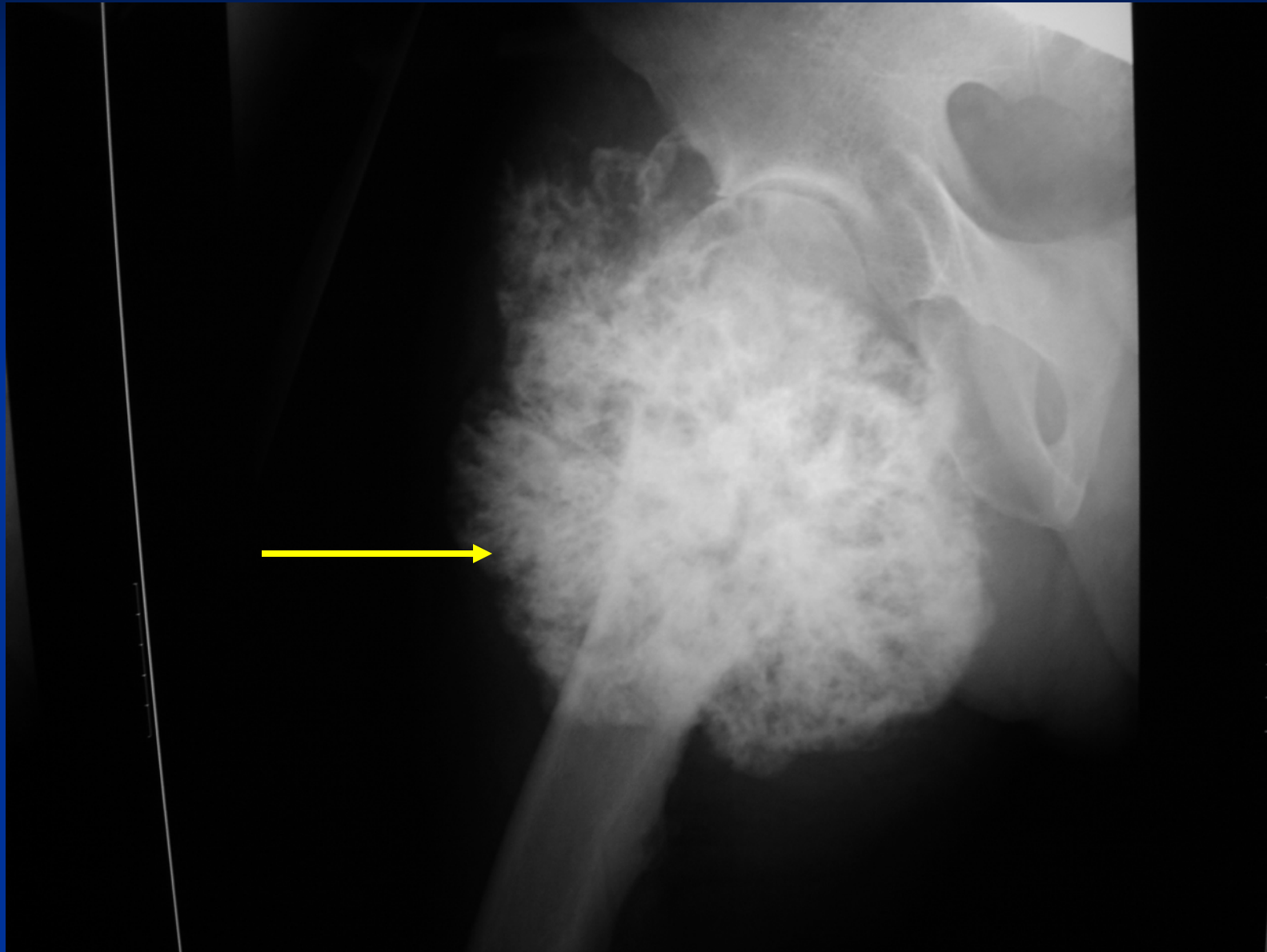
# Specimen Radiograph: Secondary Chondrosarcoma of Pelvis



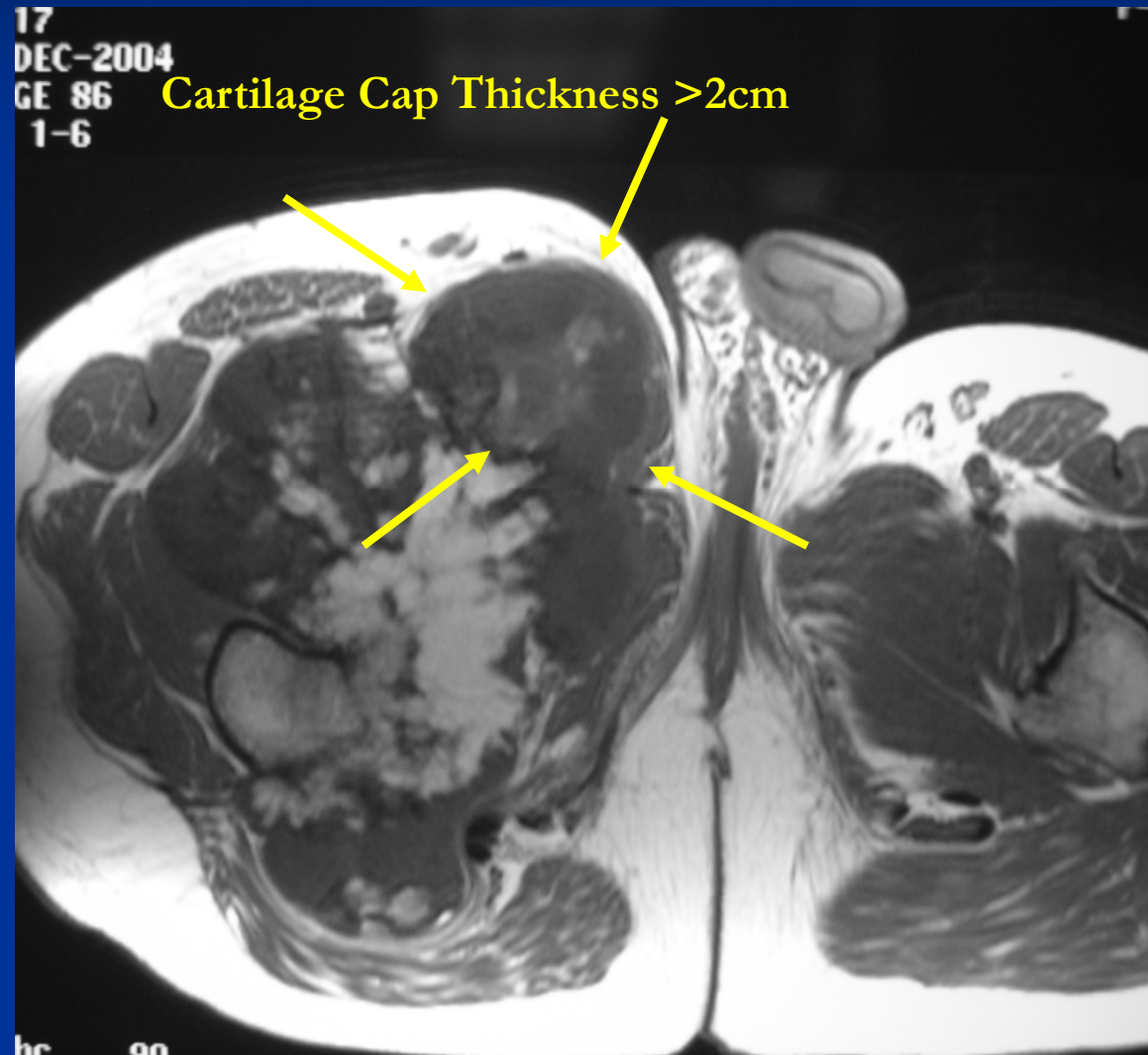
Stalk of Osteochondroma

Thick Cartilage Cap

## Plain X-ray: Secondary Chondrosarcoma of Proximal Femur

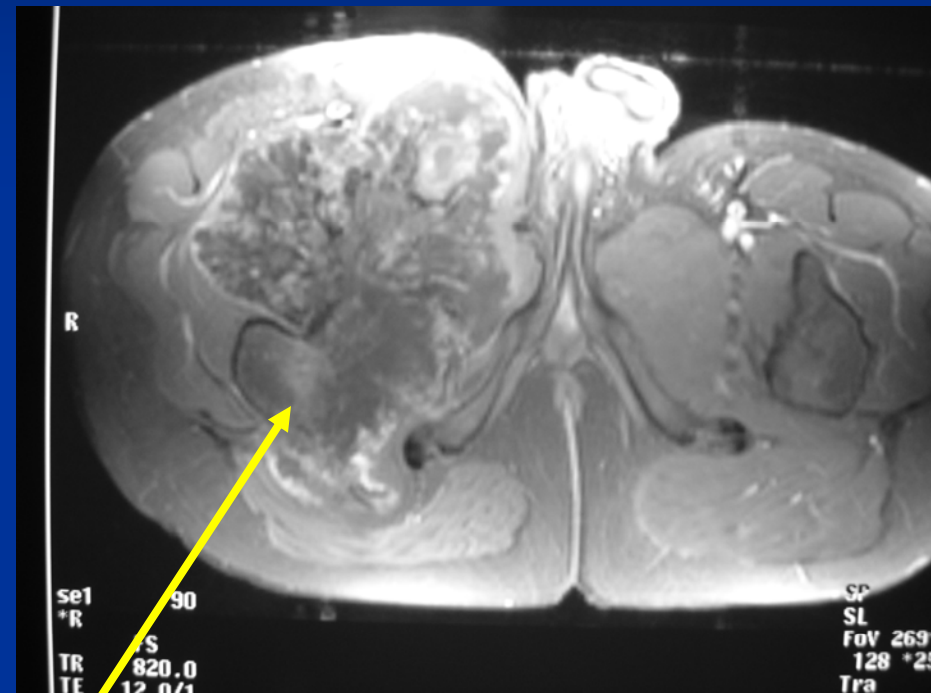
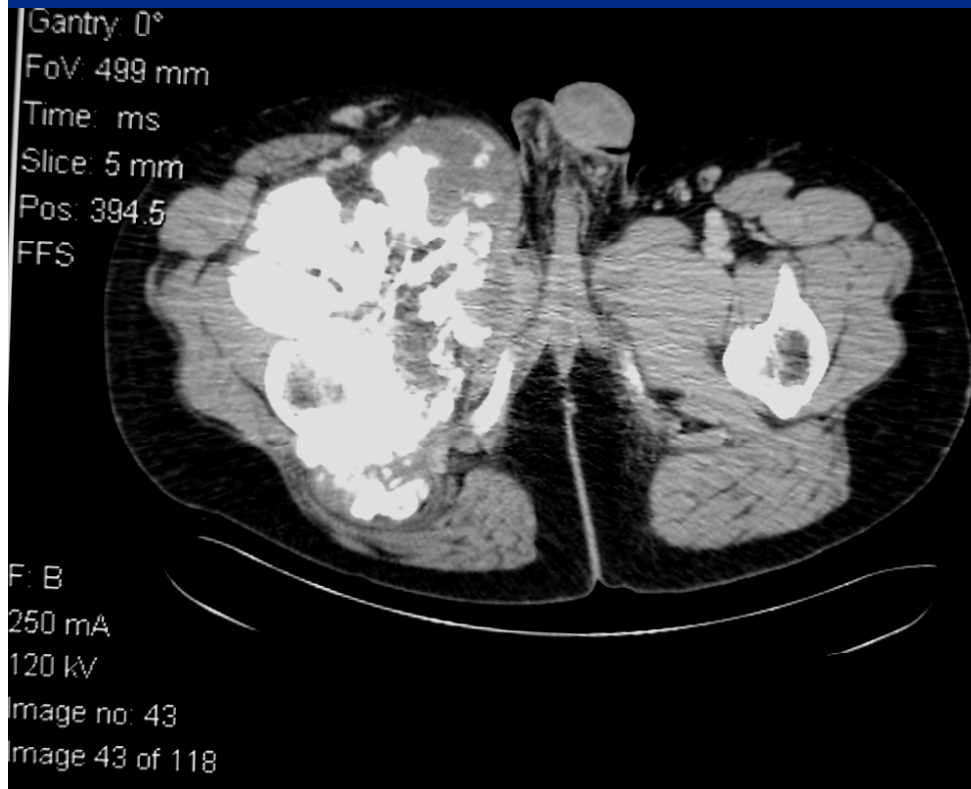


# MRI: Secondary Chondrosarcoma of Proximal Femur: Thick Cartilage Cap (>2cm)





## CT and MRI: Secondary Chondrosarcoma of Proximal Femur



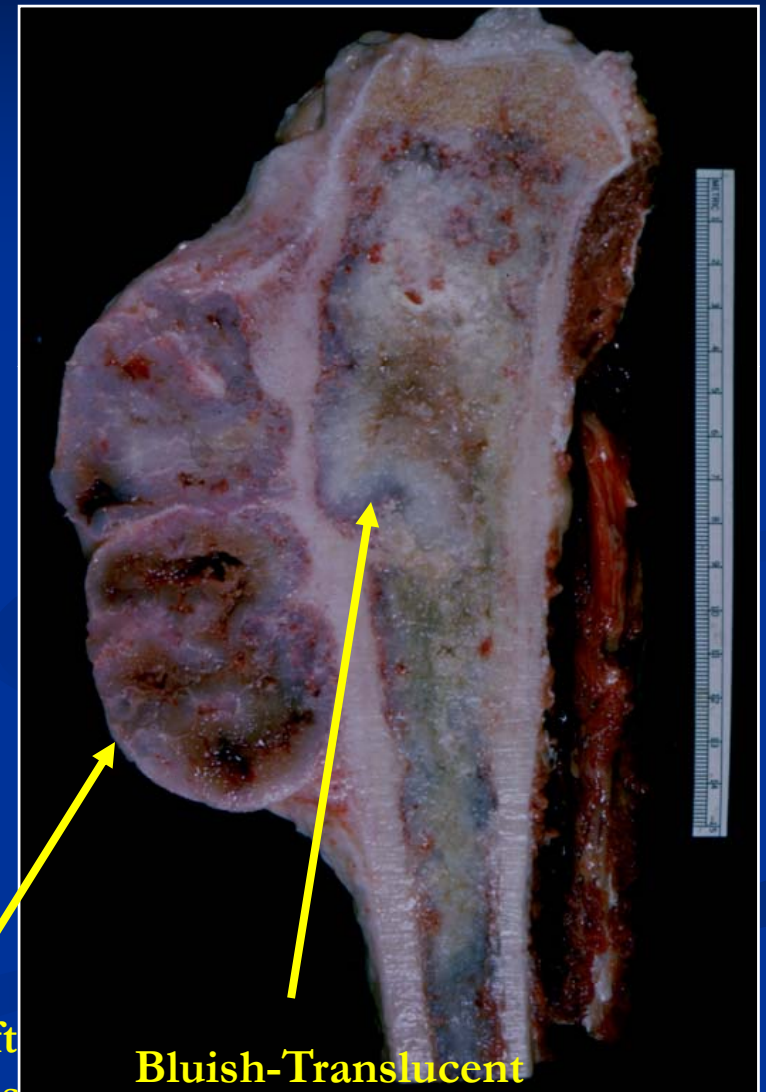
**Base of Osteochondroma**



# Gross Pathology

- Likely to have the consistency of hyaline cartilage
  - Translucent, blue-gray to white
- Lobular pattern is usually seen
- Yellow, chalky deposits of calcium
- Cystic areas with mucoid material frequent
- Hemorrhagic necrosis sometimes found especially Grade III tumors
- May have soft tissue component

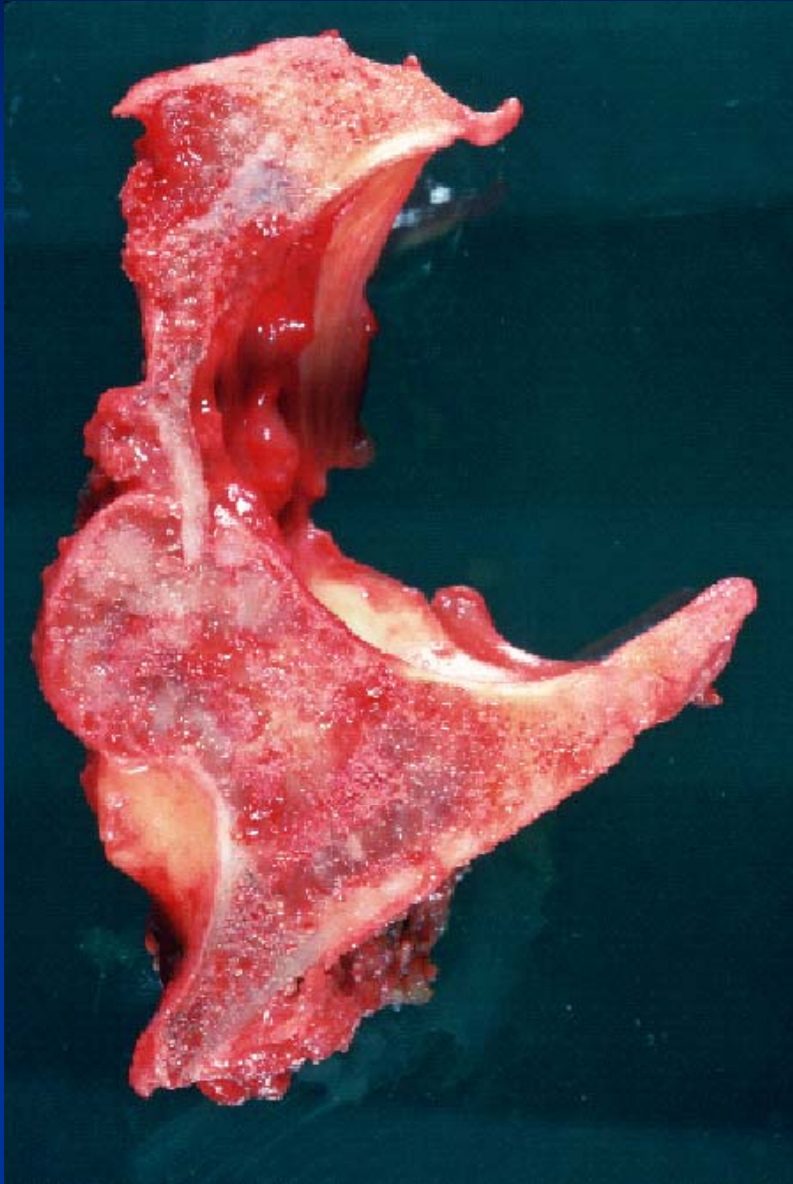
**Tibia**



**Lobular Soft  
Tissue Mass**

**Bluish-Translucent  
Cartilage Tissue**

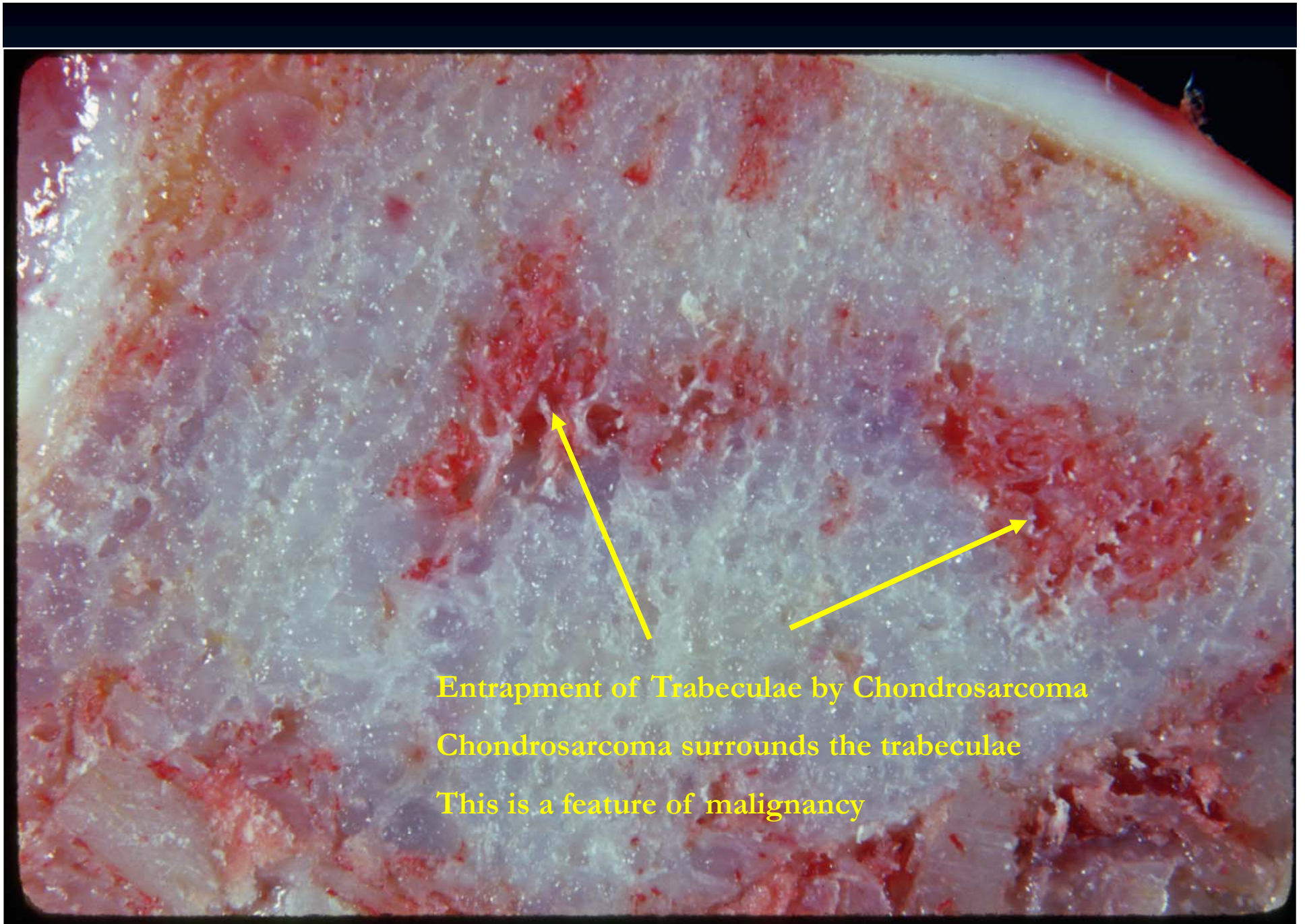
# Gross Specimen: Chondrosarcoma of Acetabulum



## Gross Specimen: Chondrosarcoma of Proximal Humerus







Entrapment of Trabeculae by Chondrosarcoma  
Chondrosarcoma surrounds the trabeculae  
This is a feature of malignancy

# Microscopic Pathology

- Broad spectrum of microscopic appearances
- General appearance
  - Irregularly shaped lobules of cartilage
    - Vary in size and abut one another
    - May be separated by fibrous bands, cleft-like spaces, or narrow bands of bone
    - Cellularity may be greater at the edges of the lobules
  - Chondrocytes often arranged in clusters
    - Normal, slightly enlarged, or overtly enlarged nuclei
    - Mononuclear or multinucleated
    - Binucleated, trinucleated cells common
    - Mitotic figures present
  - Distribution of mitotic figures is highly uneven
  - Matrix varies from mature hyaline cartilage to myxoid stroma
  - Entrapment of pre-existing trabeculae by chondrosarcoma is important for distinguishing low grade chondrosarcoma from enchondroma (The chondrosarcoma surrounds pre-existing trabeculae)



# Microscopic Pathology

## ■ Microscopic Grading

### ■ 3-Grade system

#### ■ Grade 1

- Chondrocytes with small, dense nuclei
- Few multinucleated cells
- Chondroid stroma
- Sparse myxoid area

#### ■ Grade 2

- Less matrix
- More cellular
  - Especially prominent at periphery of lobules
- Necrosis
  - Small, microscopic foci to completely necrotic lobules

#### ■ Grade 3

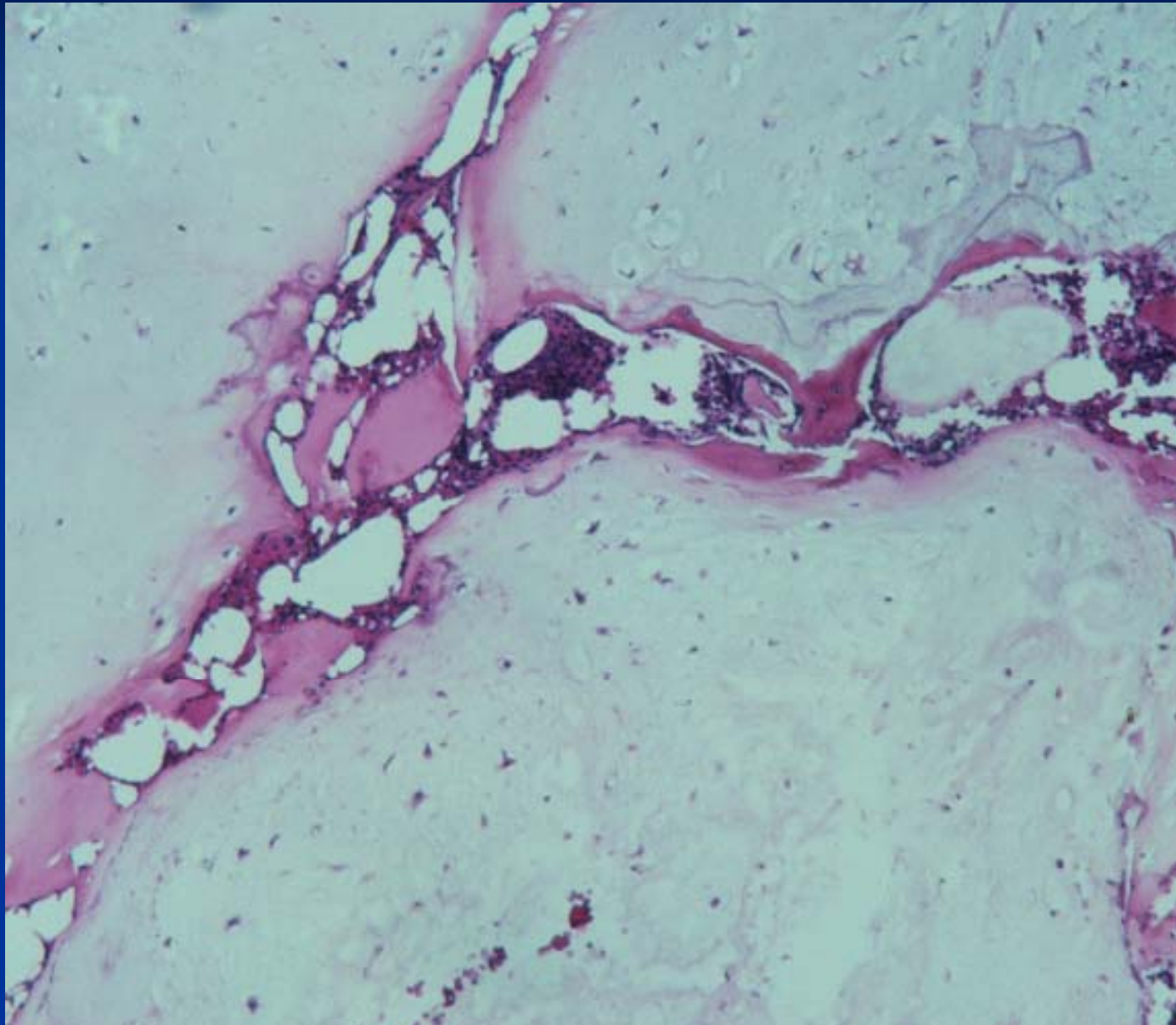
- Greater cellularity and nuclear pleomorphism
  - Chondroid matrix sparse
  - Intercellular material myxoid
  - Neoplastic chondrocytes arranged in cords and clumps
  - Individual cells are stellate or grossly irregular
  - Foci of necrosis are frequently extensive
- Individual lesions may have areas of different grades

# Conventional Chondrosarcoma

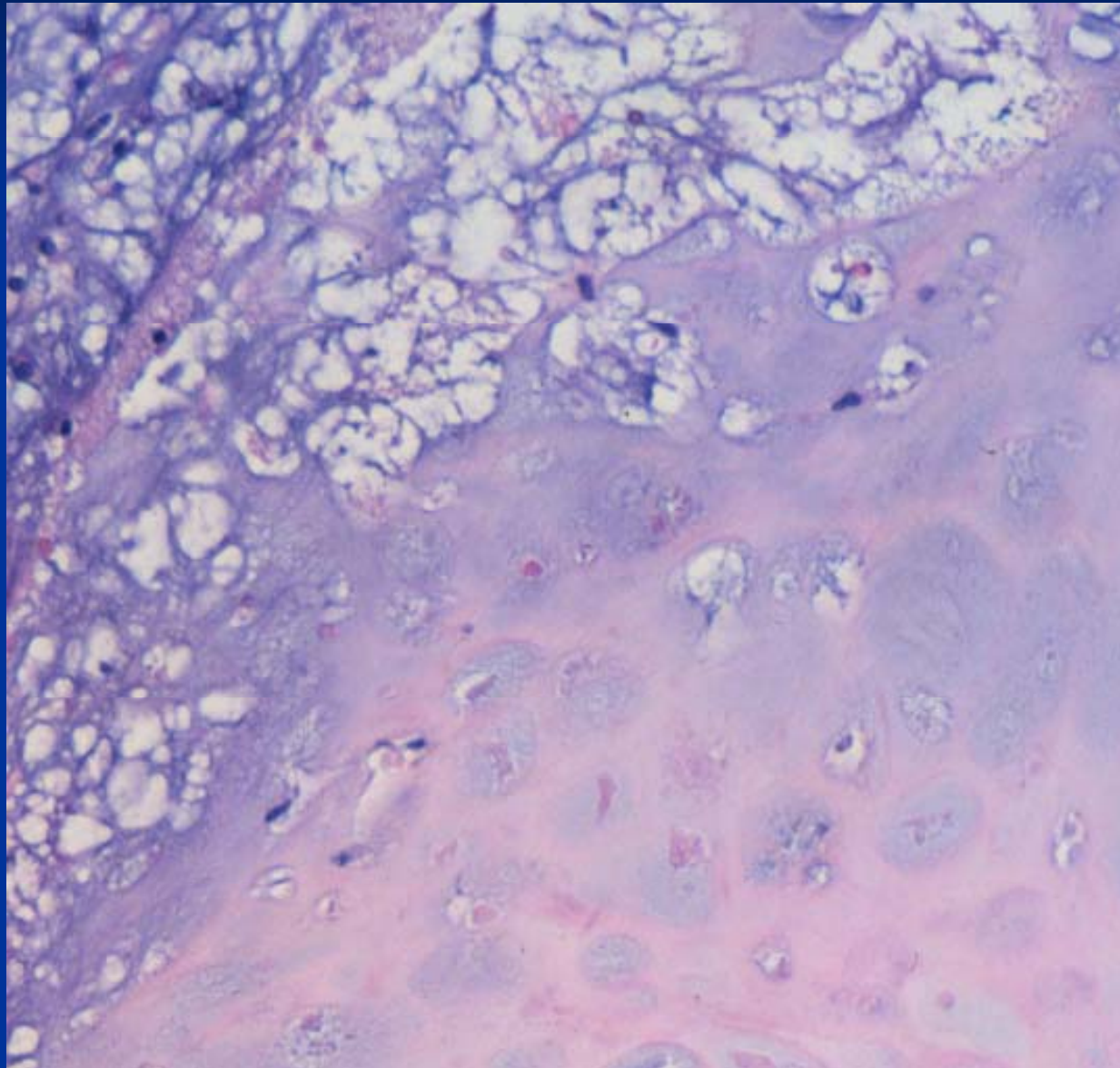
## Grade I (Low Grade Chondrosarcoma)

- Similar microscopic features to Enchondroma
- Require clinical and radiographic data to support diagnosis
- Hypercellularity, plump cells, prominent nucleoli, nuclear pleomorphism
- Mitotic figures not typically present
- **Bone Entrapment of pre-existing trabeculae is important**
- More than occasional double nuclei
- Continuous growth with infiltrative growth pattern
- Rare metastatic spread

# Microscopic Pathology: Grade I Chondrosarcoma



# Microscopic Pathology: Grade I Chondrosarcoma





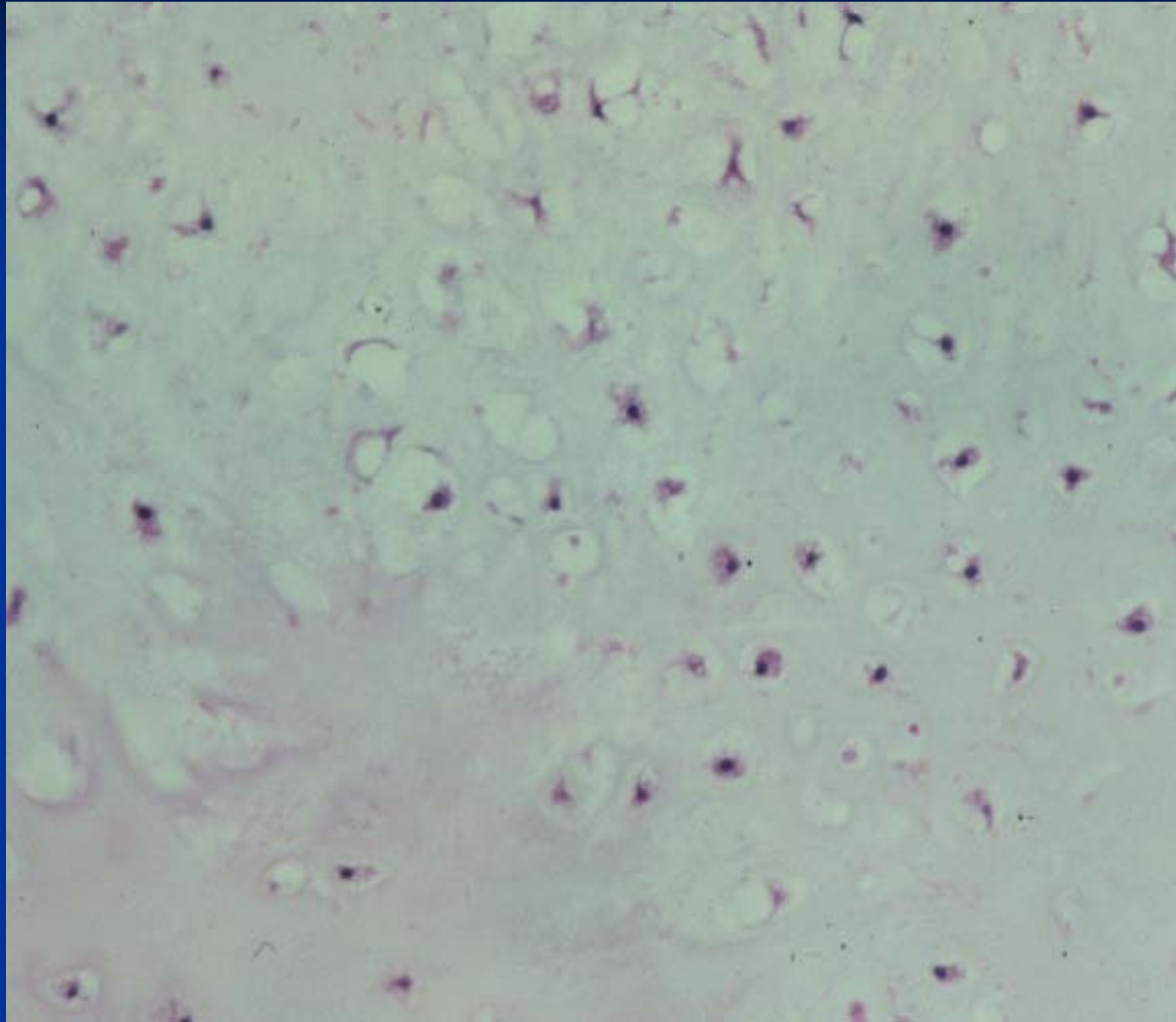
# Microscopic Pathology: Grade I Chondrosarcoma

Entrapment of Pre-existing  
Trabeculae of Bone





# Microscopic Pathology: Grade I Chondrosarcoma

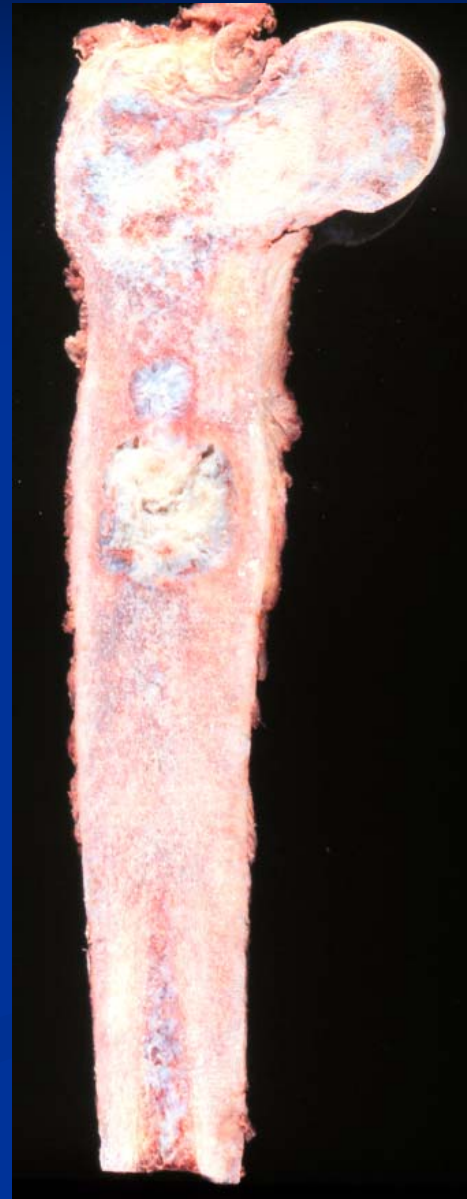
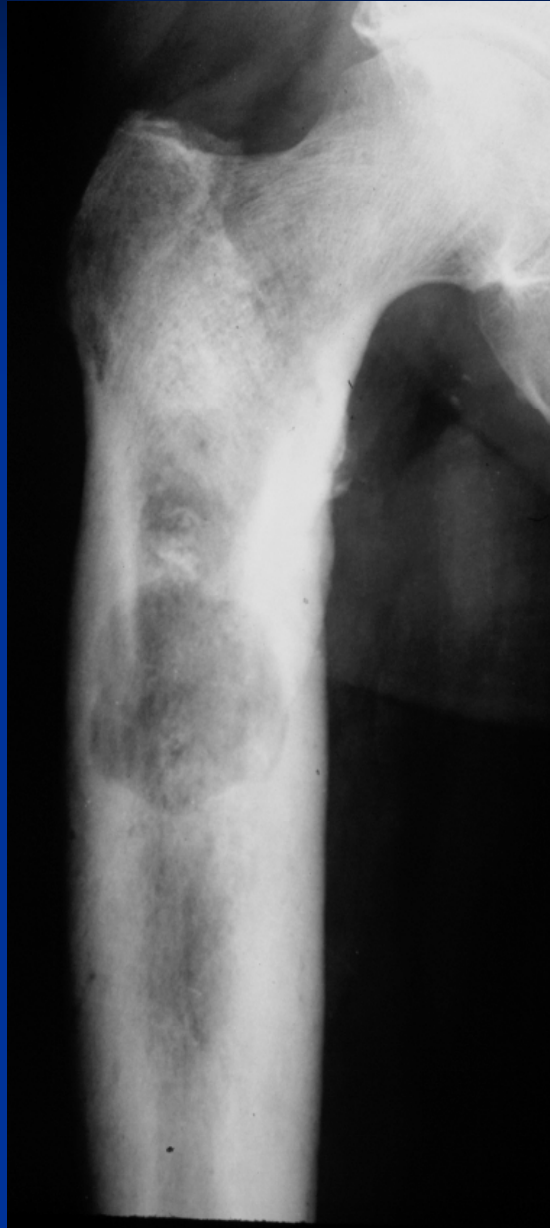


# Conventional Chondrosarcoma

## Grade II (Intermediate Grade Chondrosarcoma)

- Increased cellularity evenly distributed in a cartilaginous matrix
- Plump cartilage cells with enlarged nuclei and distinct nucleoli, greater nuclear pleomorphism
- Frequent binucleated, trinucleated cells
- Occasional mitotic figures
- Foci of Myxoid change
- Greater potential of local recurrence
- Metastatic rate 10-15%

# Grade II Chondrosarcoma

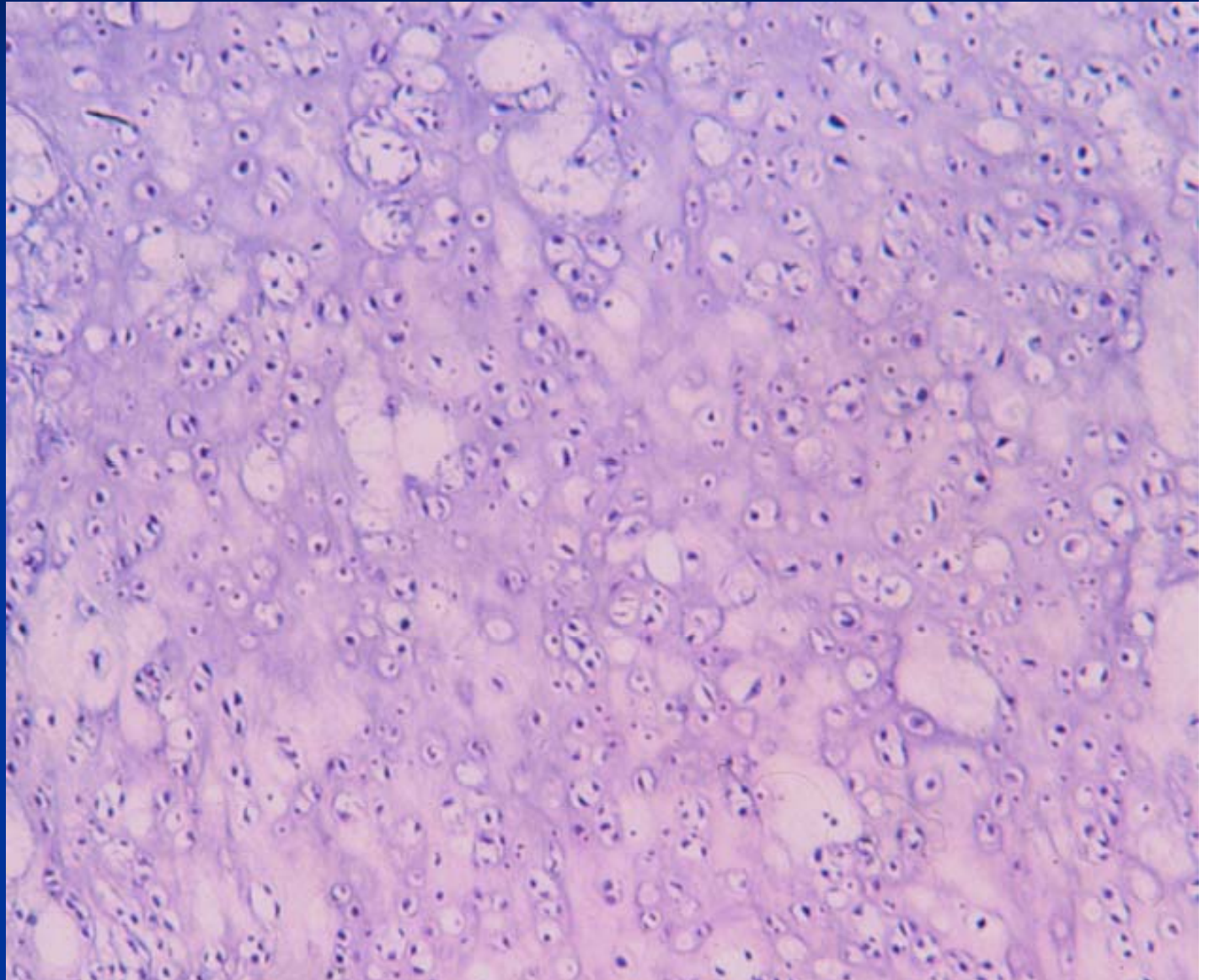


# Microscopic Pathology: Grade II Chondrosarcoma

Hypercellular

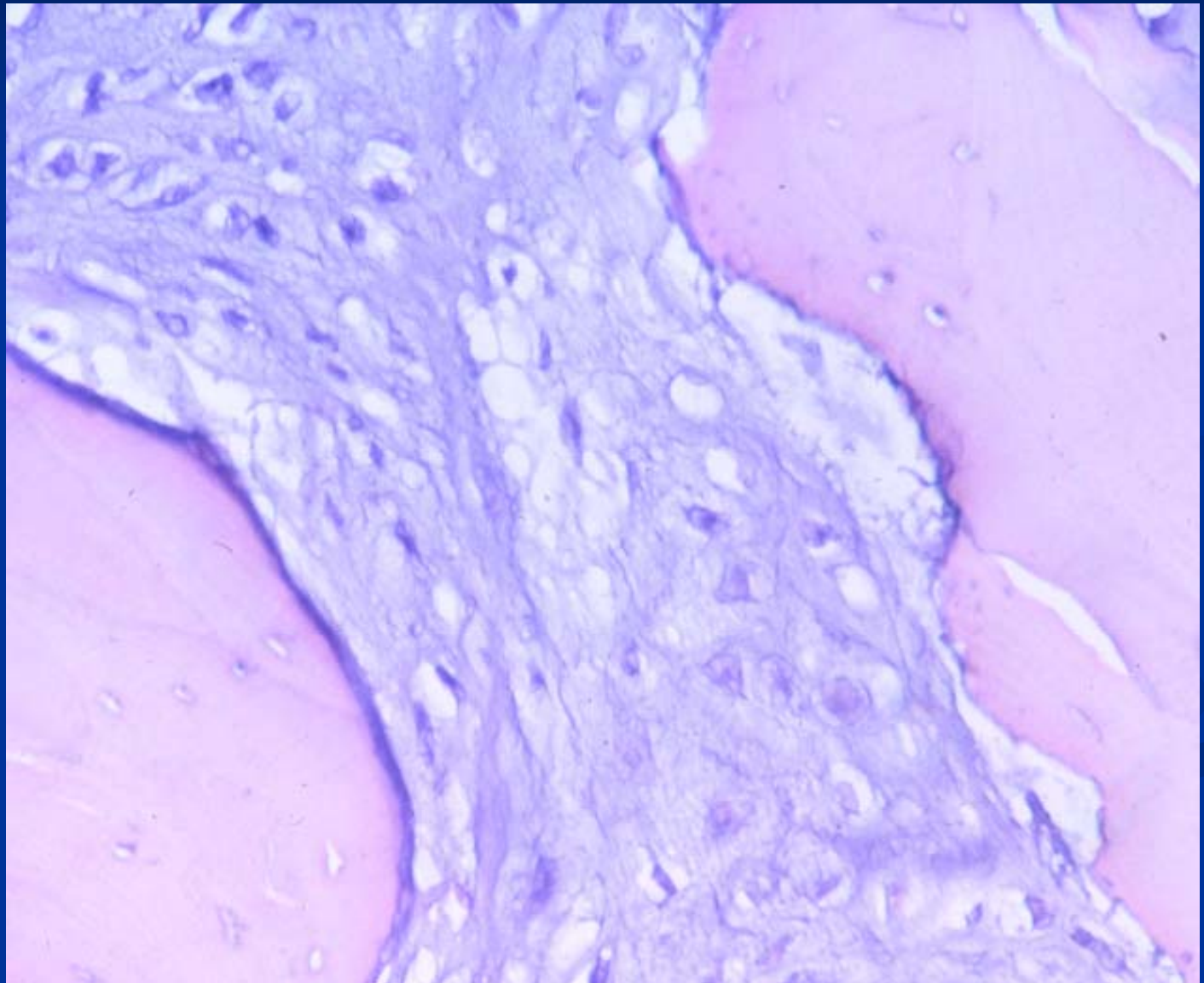
Cells are crowded

Binucleated cells  
common





# Microscopic Pathology: Grade II Chondrosarcoma

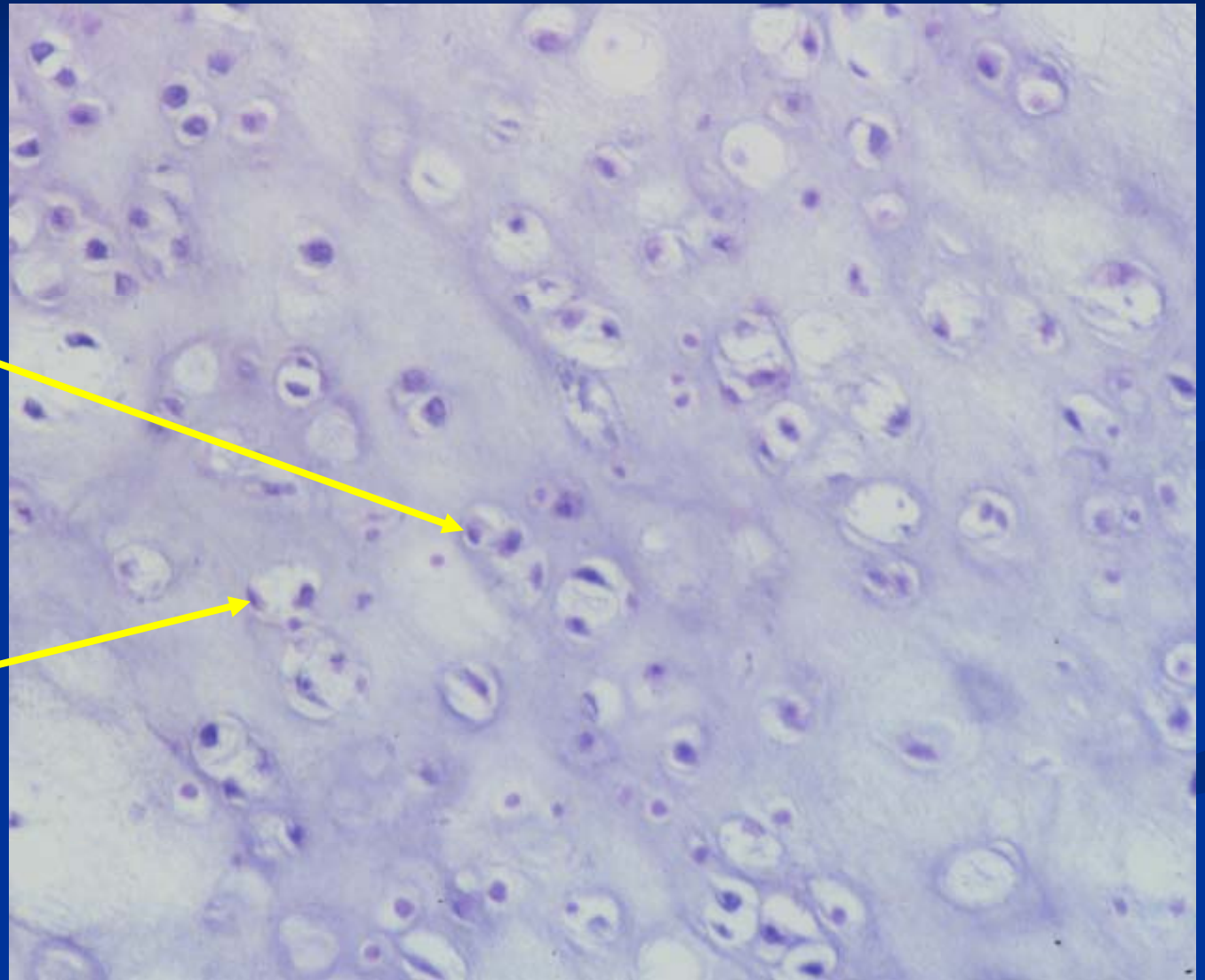




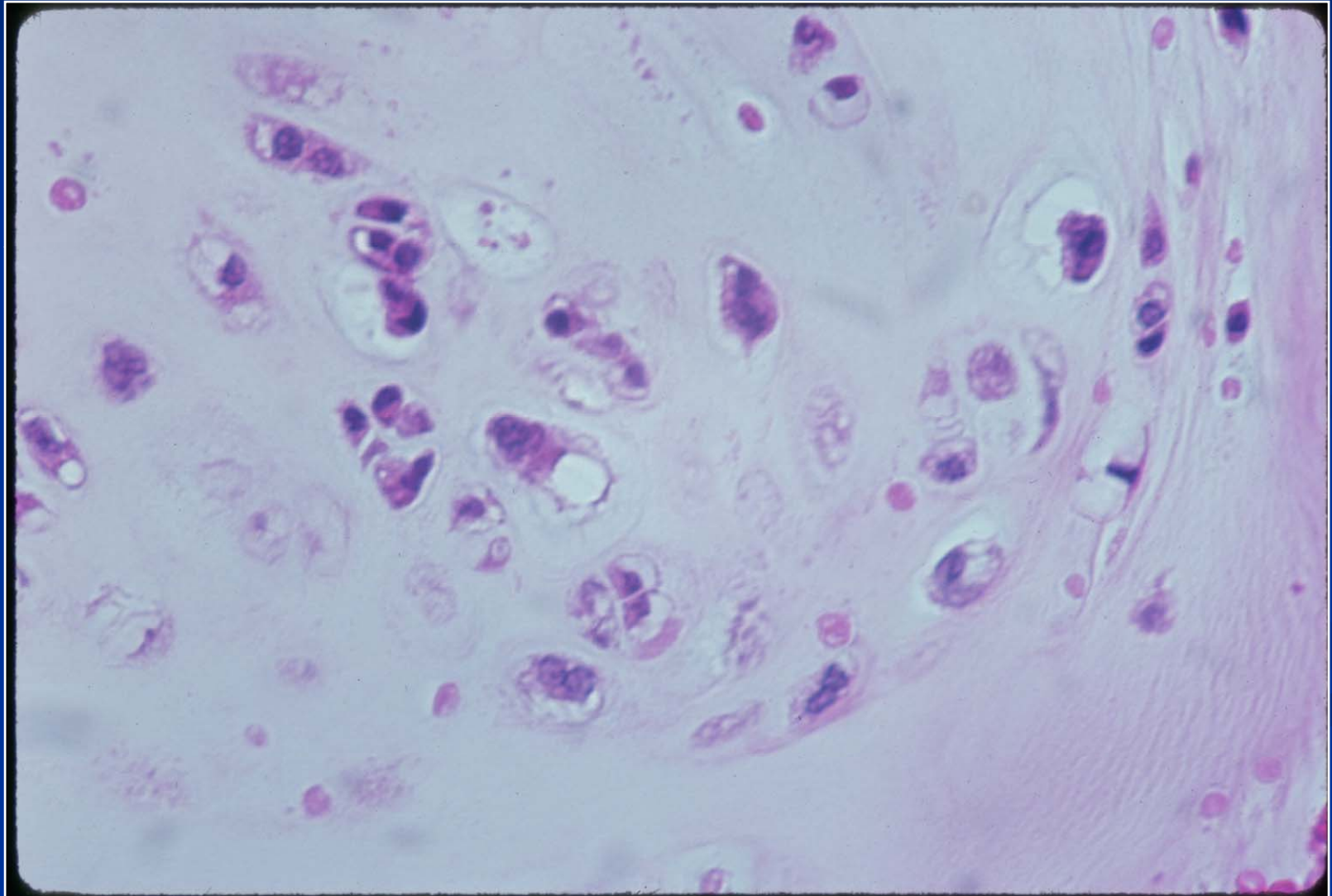
# Microscopic Pathology: Grade II Chondrosarcoma

Trinucleated Cells

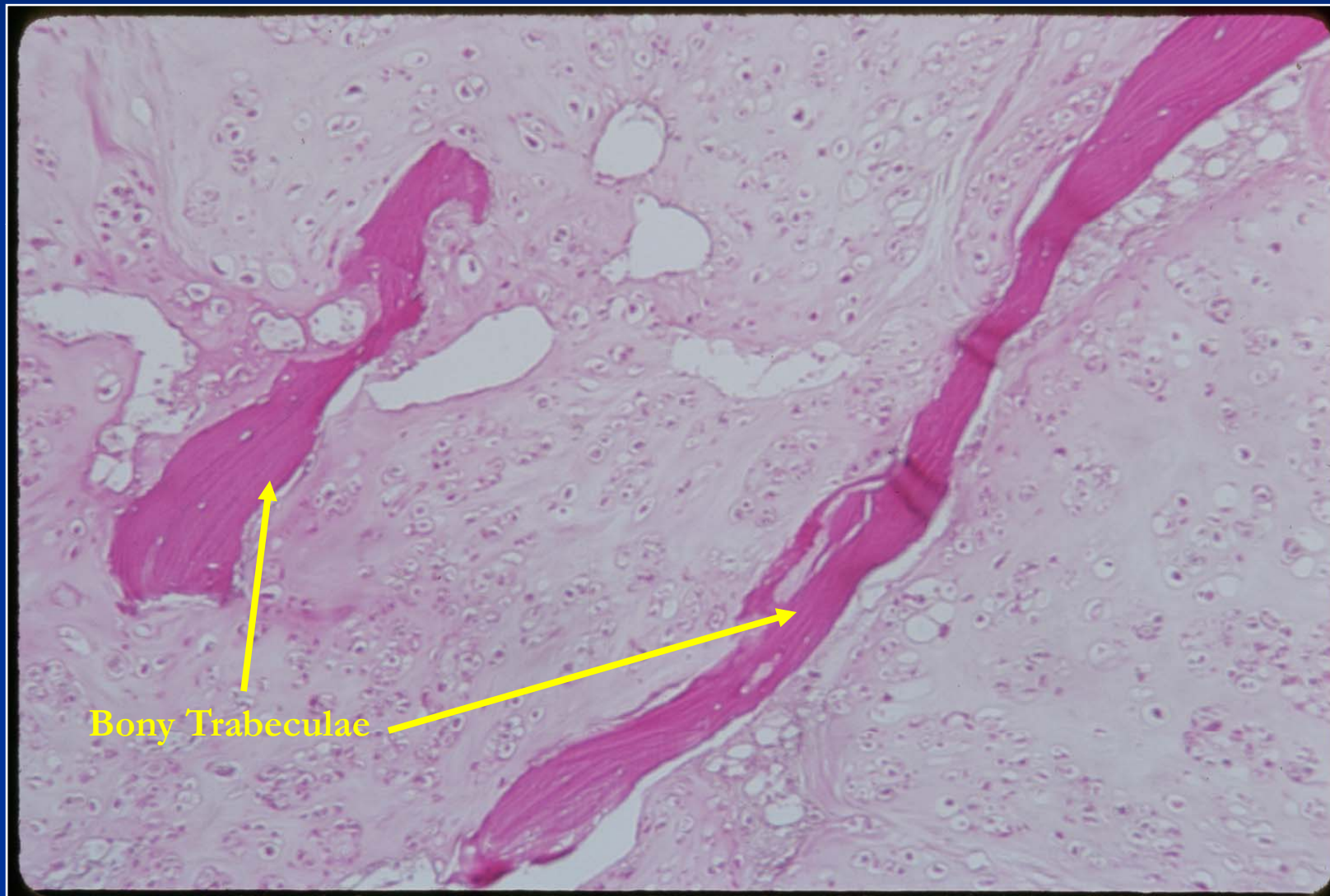
Binucleated Cells



## Microscopic Pathology: Grade II Chondrosarcoma



# Microscopic Pathology: Grade II Chondrosarcoma and Bony Entrapment



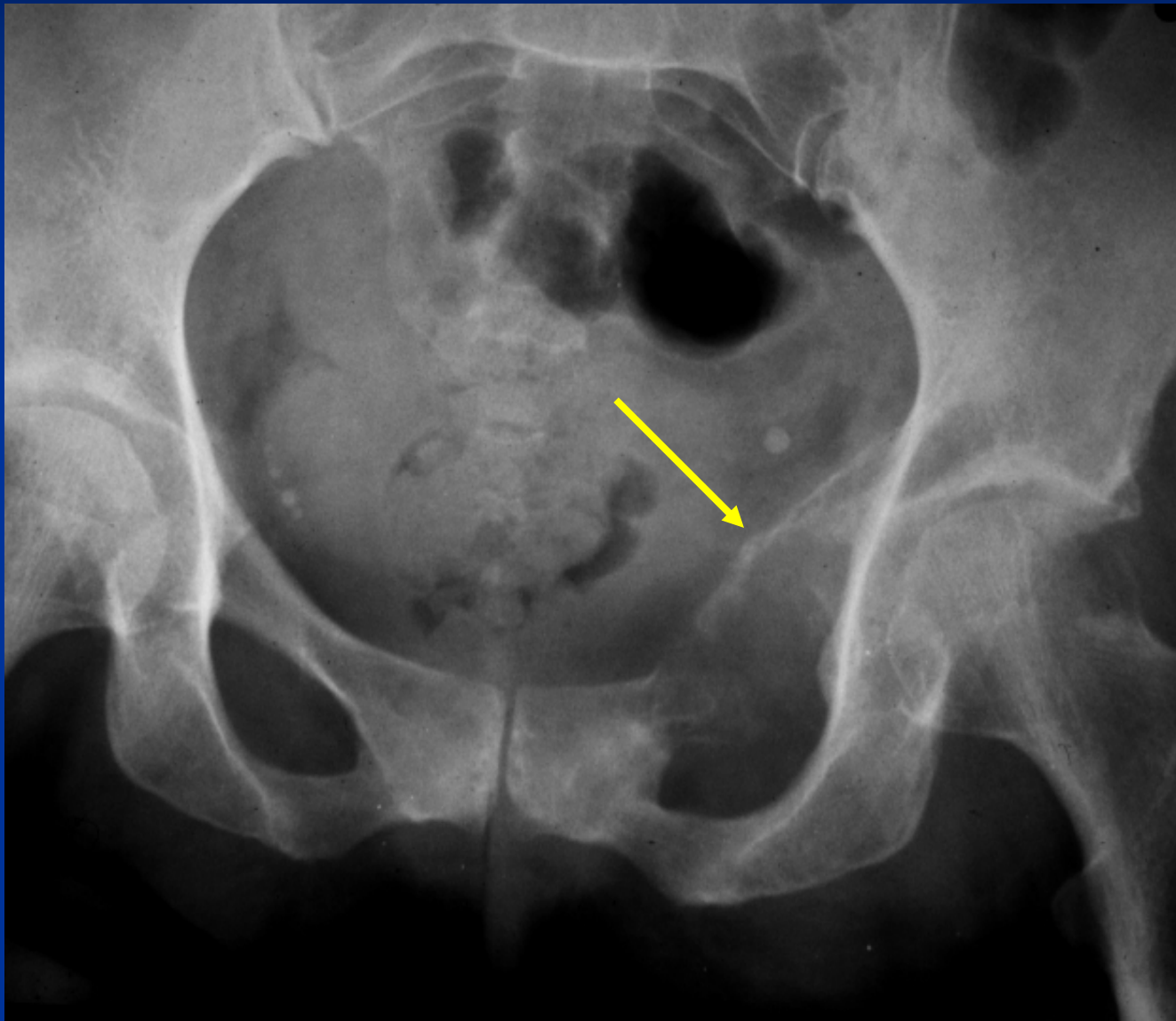


# Conventional Chondrosarcoma

## Grade III (High Grade Chondrosarcoma)

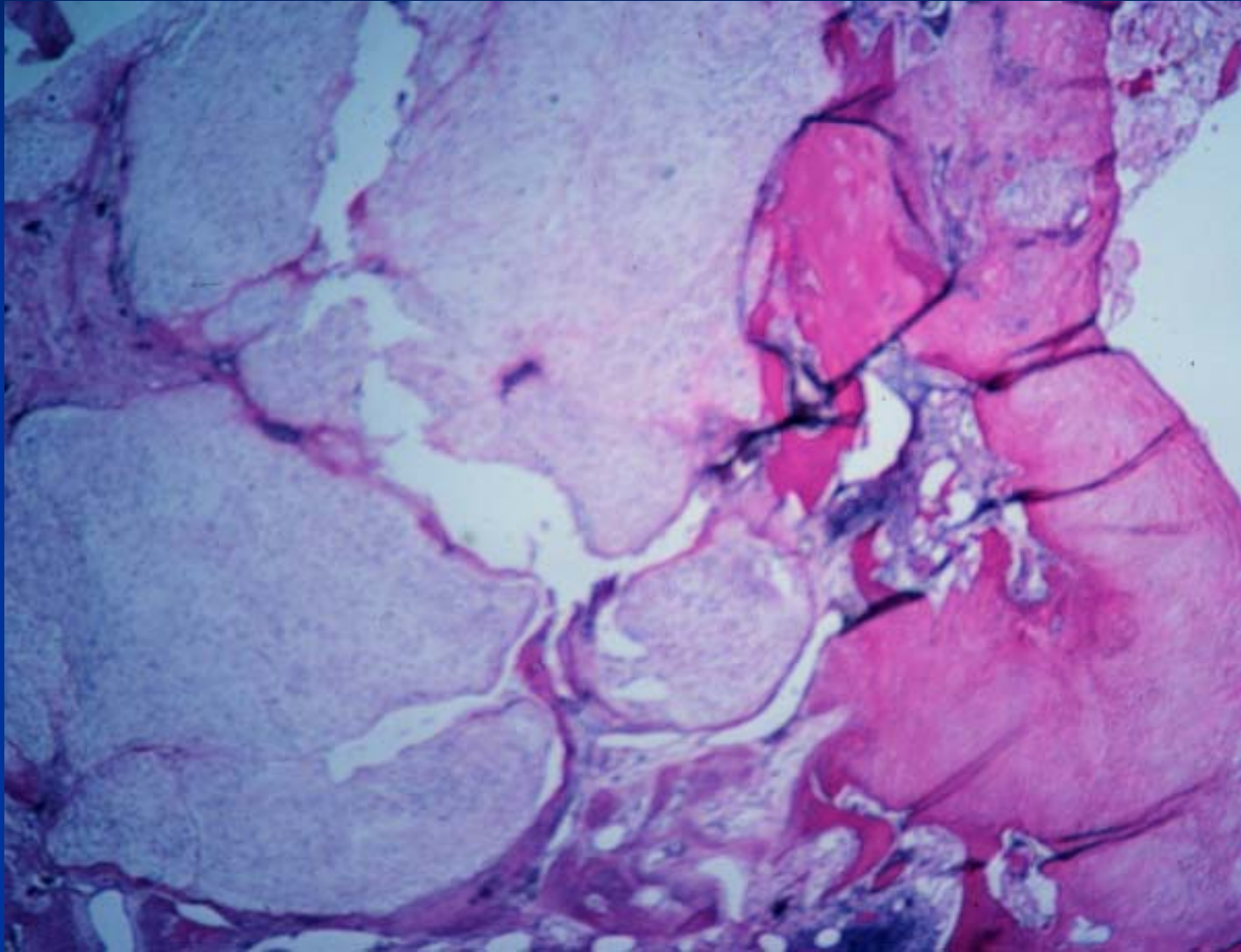
- Higher cellularity and greater degree of cellular pleomorphism
- Hyaline cartilage matrix is sparse
- Marked nuclear pleomorphism
- Cells may have stellate/spindle appearance with myxoid chondroid matrix
- May have myxoid areas
- Prominent nuclear atypia
- Presence of mitotic figures
- Locally highly aggressive
- Metastatic rate over 50%

# Grade III Chondrosarcoma of Pelvis





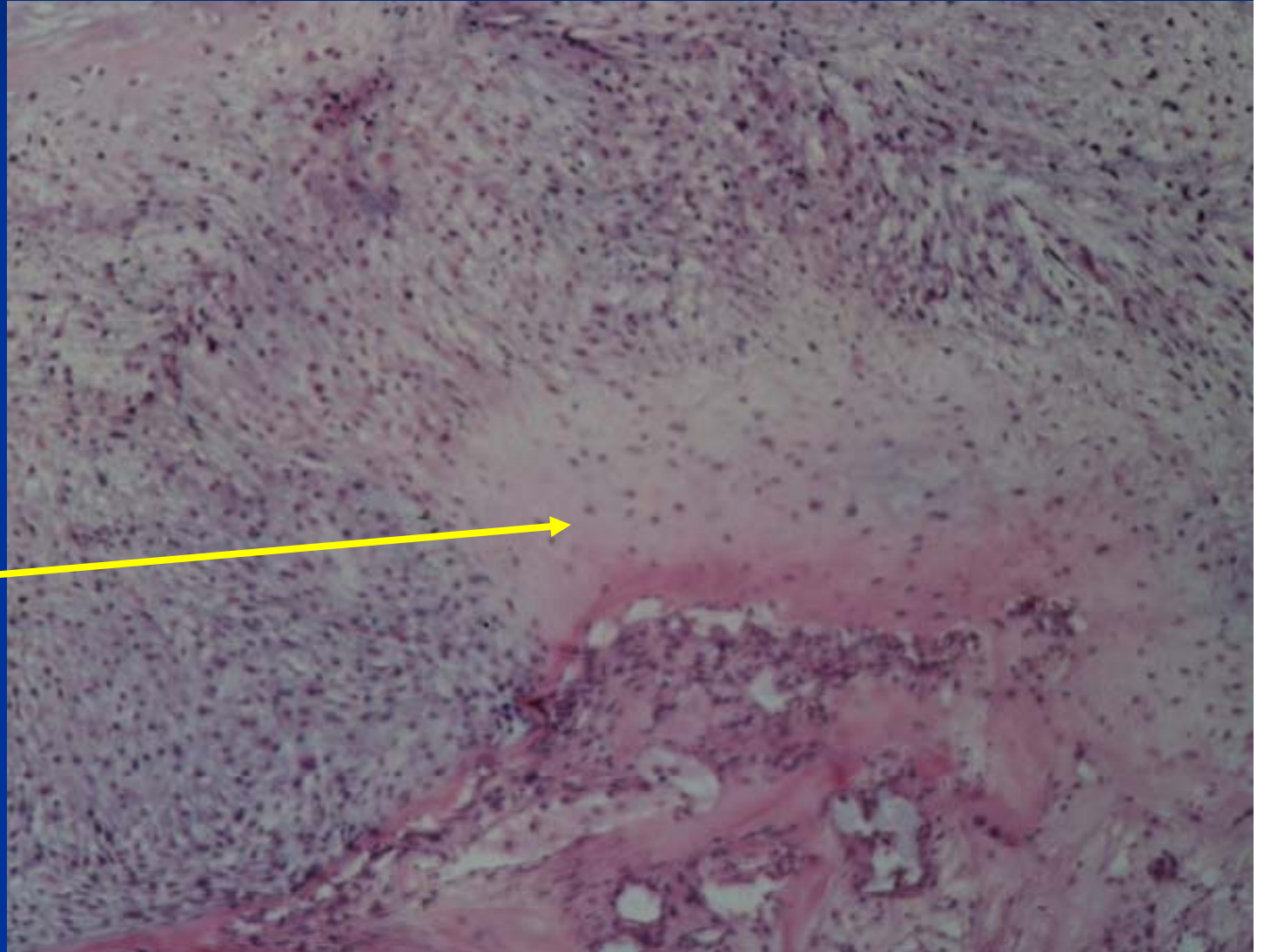
# Microscopic Pathology: Grade III Chondrosarcoma



# Microscopic Pathology: Grade III Chondrosarcoma

Hypercellular

Chondroid Area





# Microscopic Pathology: Grade III Chondrosarcoma

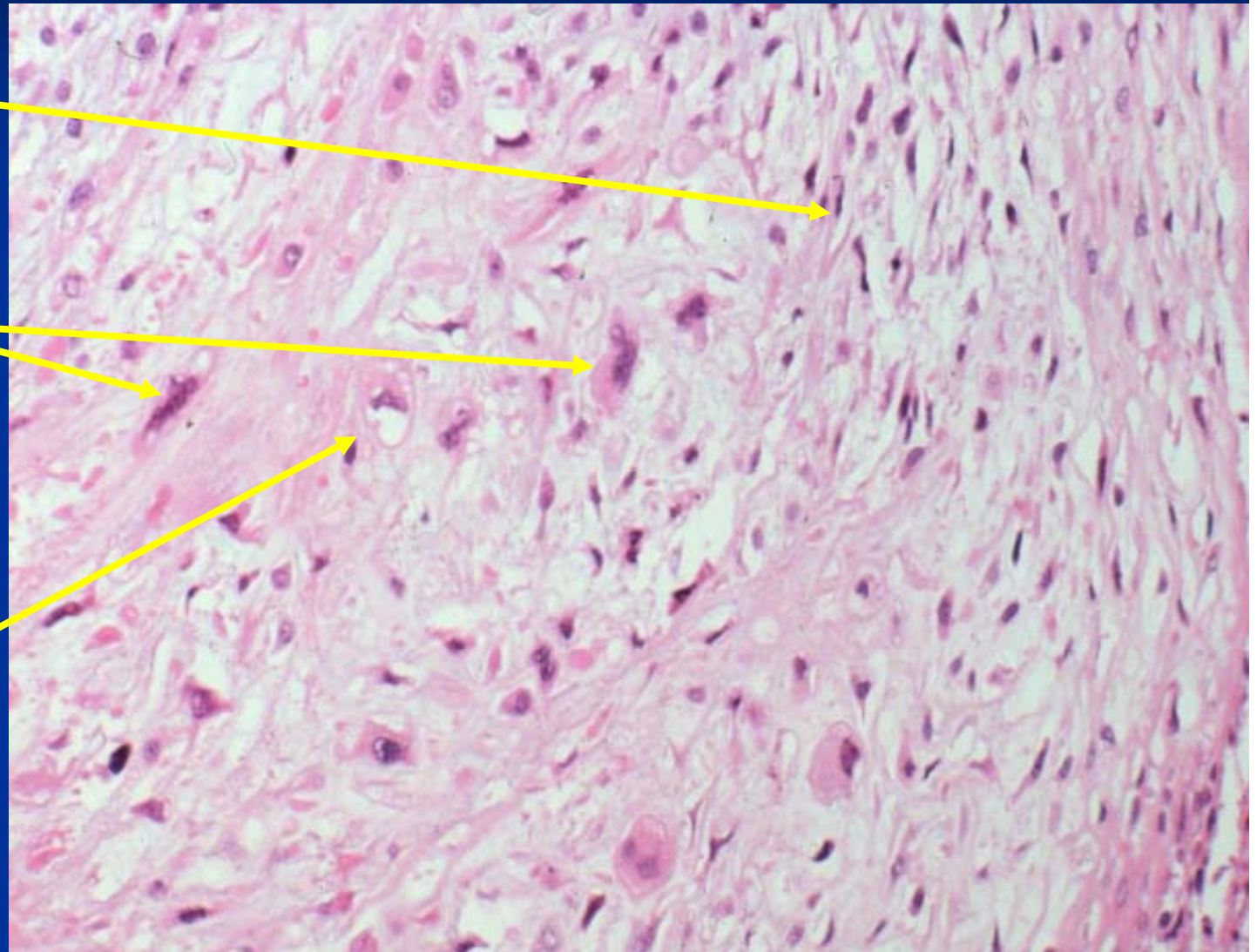
Spindle/Stellate  
Appearance to  
Cells in Areas

Mitotic Figure

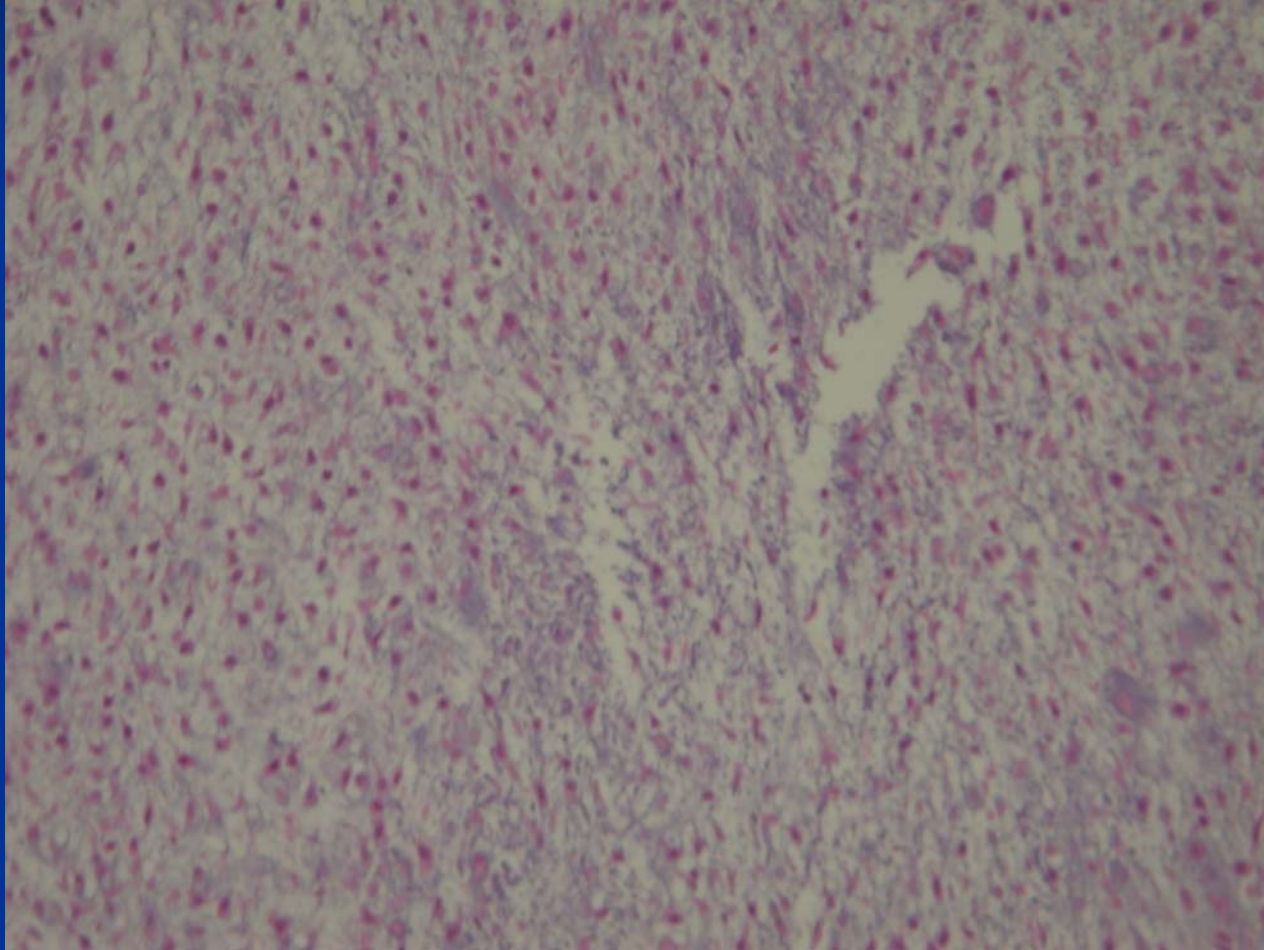
Cell in Lacunae

Signet Ring  
Configuration

Pleomorphism

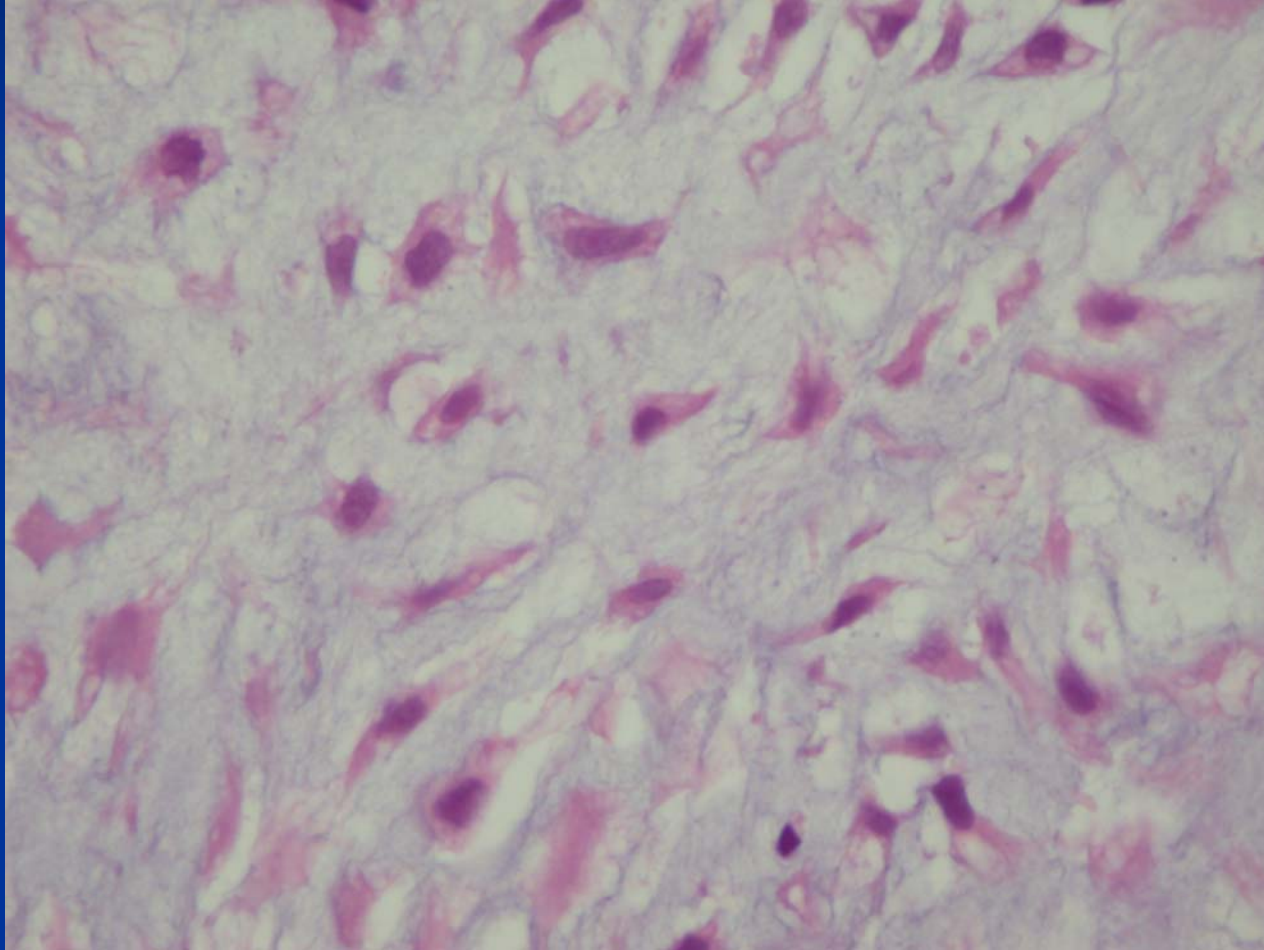


# Microscopic Pathology: Grade III Chondrosarcoma



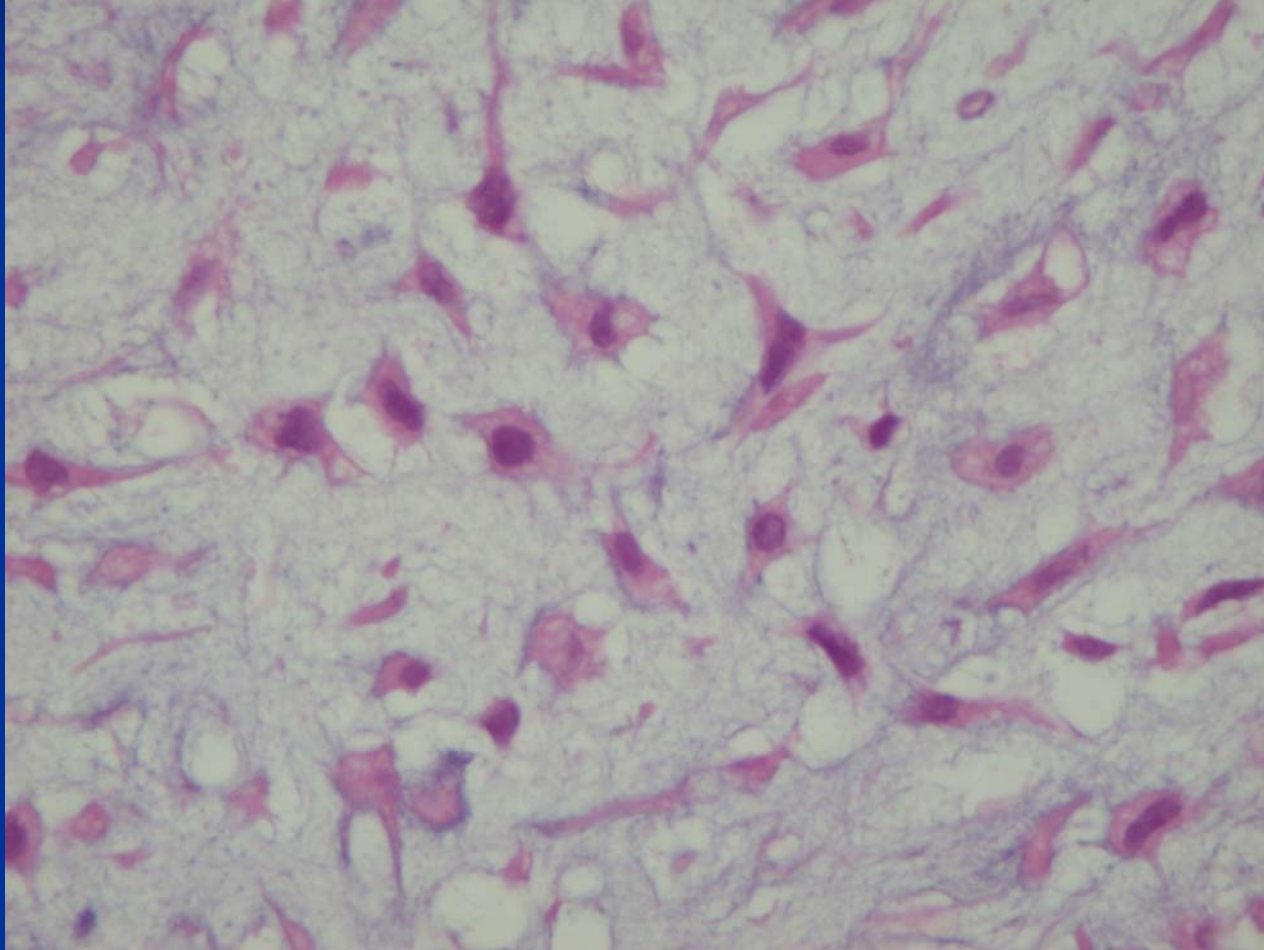


# Microscopic Pathology: Grade III Chondrosarcoma





# Microscopic Pathology: Grade III Chondrosarcoma



# Differential Diagnosis

- Grade I chondrosarcoma vs. enchondroma
- Chondroblastic osteosarcoma

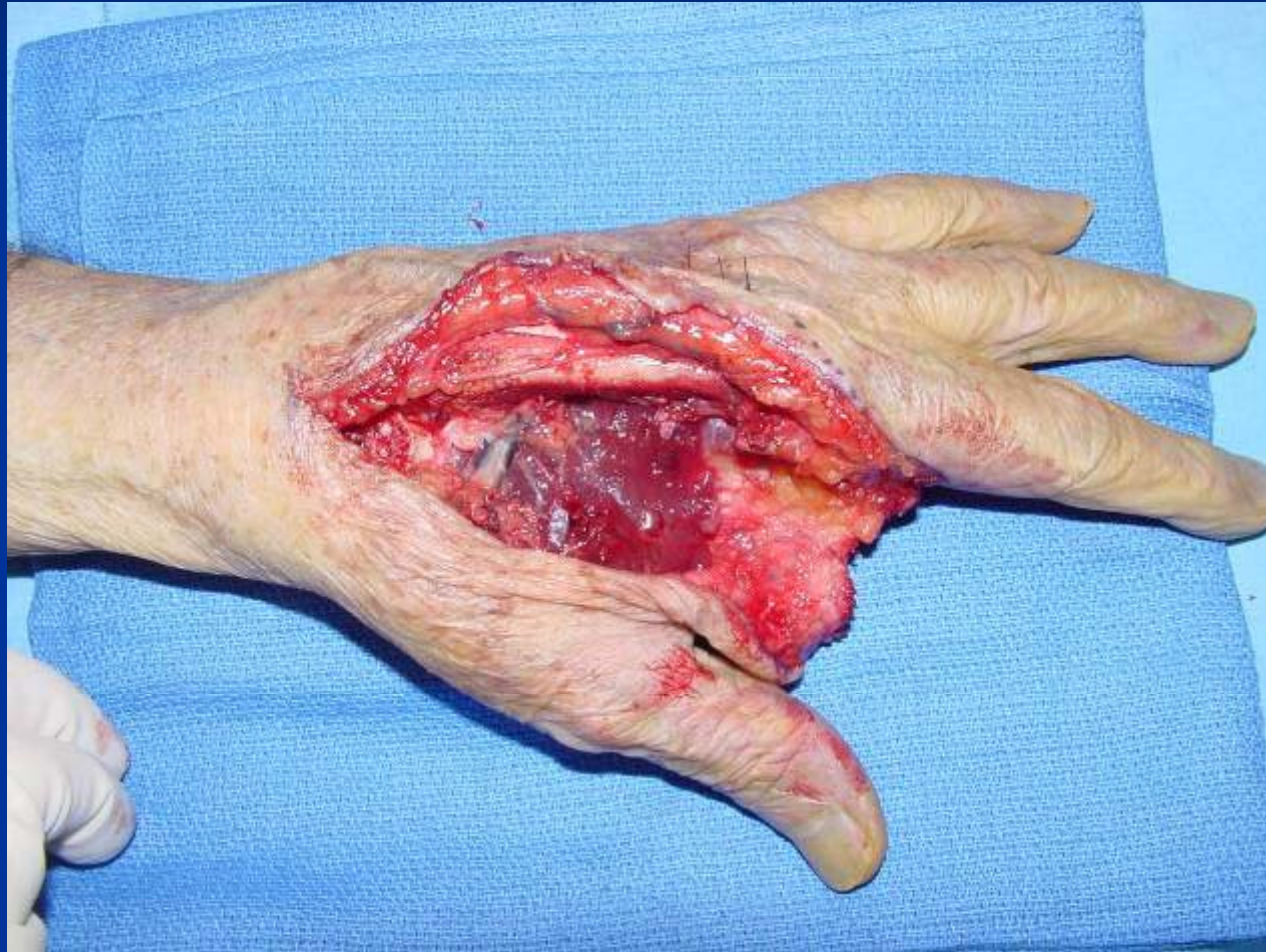
# Biological Behavior

- The biological behavior is related to grade
  - Grade I Chondrosarcoma rarely metastasize and grow slowly. They may dedifferentiate to high grade sarcomas such as osteosarcoma, MFH and fibrosarcoma
  - Grade II Chondrosarcomas grow locally in an aggressive manner. They metastasize in up to 33% of cases. Most commonly metastasize to the lungs
  - Grade III Chondrosarcomas grow locally in an aggressive manner and metastasize in up to 70% of cases. Most commonly metastasize to the lungs.

# Treatment

- Surgery is the main treatment. Most patients can be treated with a limb sparing resection although amputation may be needed for large tumors. Chemotherapy has little role in treatment of chondrosarcomas. It may be considered for grade III and dedifferentiated chondrosarcomas although its use is controversial. Radiation has little role in treatment of chondrosarcomas. It may be considered for treating microscopic disease following surgical removal of large tumors especially of the pelvis and spine when wide resection is not feasible.
- Wide/Radical En bloc Resection
  - Preferred treatment for Grade II and III Tumors
- Curettage can be considered for grade I chondrosarcomas
  - High recurrence rate for grade II and III chondrosarcomas

# Amputation for a Large Chondrosarcoma of the Hand Metacarpal





# Prognosis

- Depends on 2 major factors
  - Stage
  - Histological grade
- ~70% of grade 3 tumors metastasize even if a wide surgical margin is achieved
  - Metastases are uncommon in grade 1 tumors
  - 10-33% of grade 2 tumors metastasize
- Prognosis of grade 1 and 2 tumors governed by resectability and location
- Patients who present with metastatic disease have a dismal prognosis
- Local recurrences or metastases usually occur within 5 years
- Death rate is very high
  - Especially if tumor is in difficult location or grade 3