73M with knee pain

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AP & Lateral Radiograph Knee

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Paget's Disease of Bone

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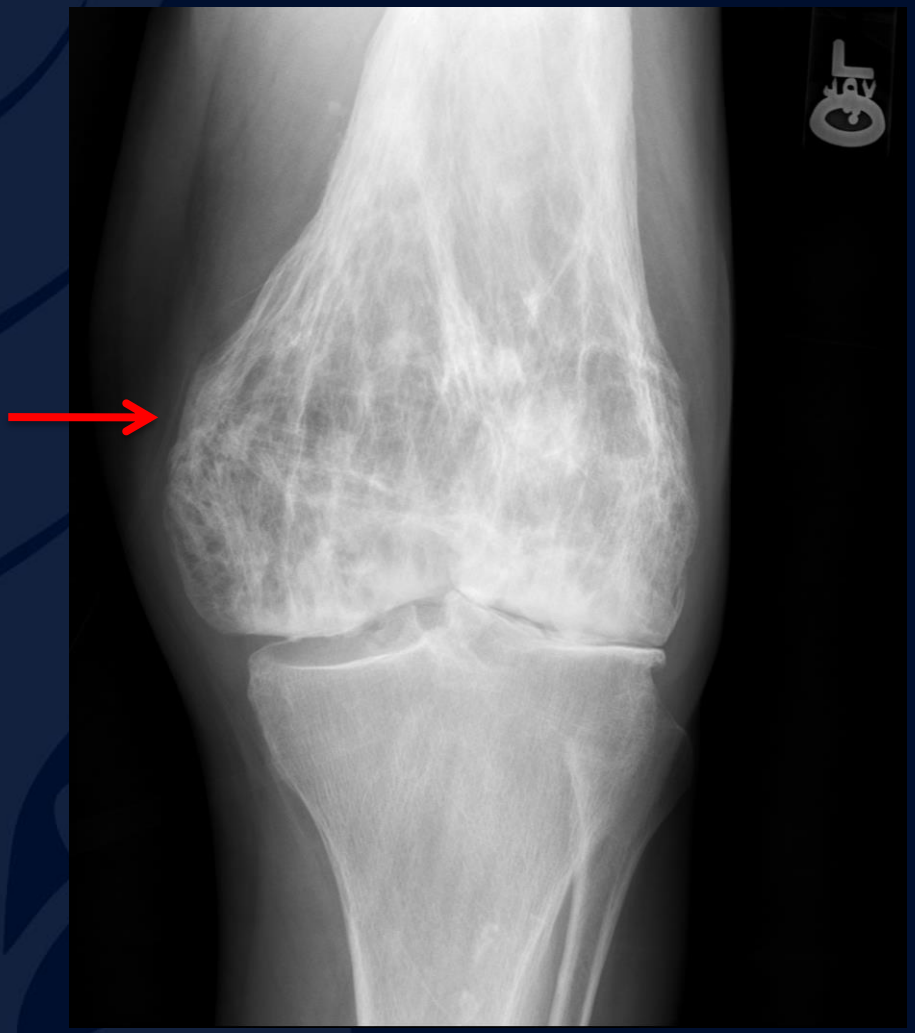
AP & Lateral Radiograph Knee



cortical thickening, osseous expansion, coarse trabecular markings

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CR Findings by Stage

- Excessive bone resorption followed by formation of structurally abnormal bone

1. Lytic stage: Active
   - Osteoclasts line trabeculae
   - Increased vascularity
2. Intermediate/ Mixed stage: Active
   - Osteoclasts and osteoblasts line seams of osteoid
3. Sclerotic stage: Late inactive
   - Osteoblasts predominate - rapid random deposition of disorganized structurally weakened new bone

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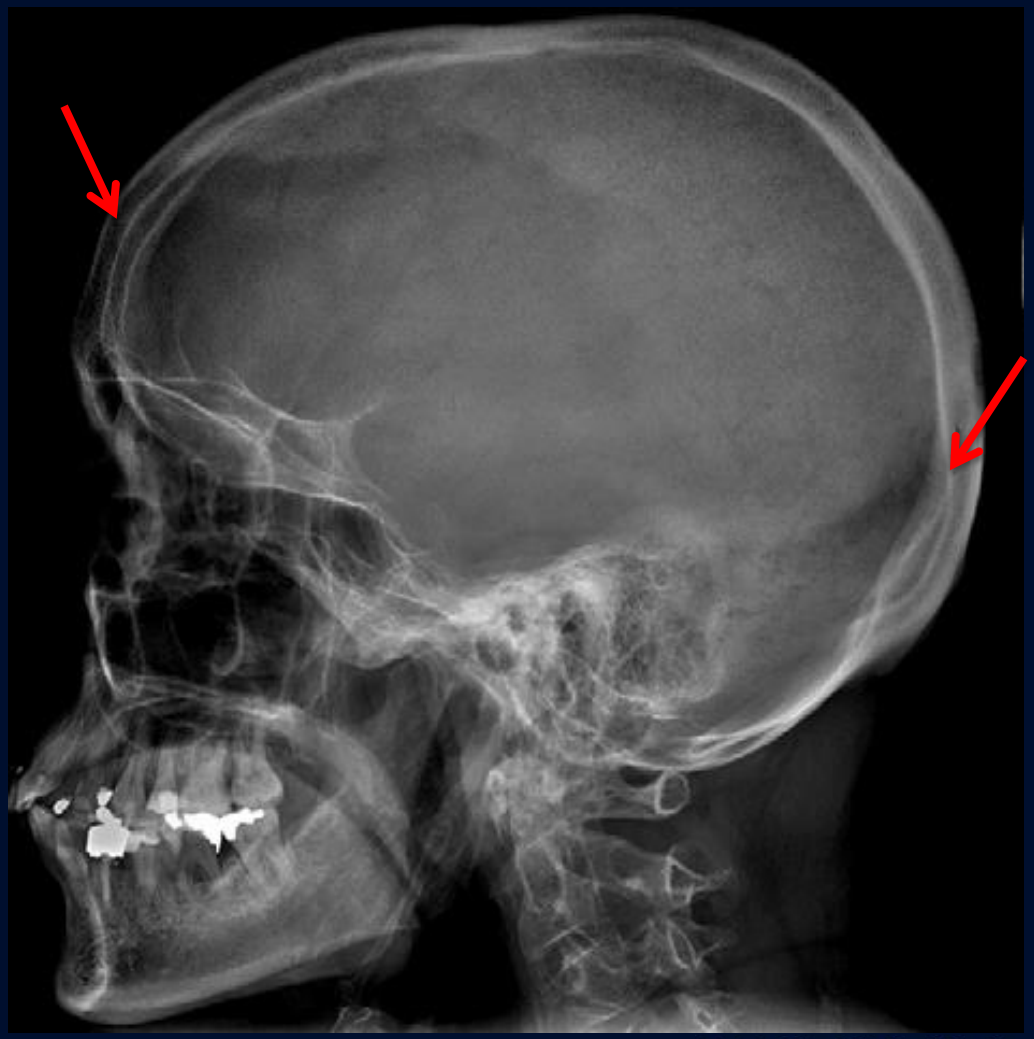
1. Osteolytic Stage

- Geographic osteolysis with an advancing edge of resorption
- long bones- a sharp wedge-shaped leading edge of osteolysis which advances from the subarticular or metaphyseal to the diaphyseal region
  - “cutting-cone”, “blade of grass”, or “candle flame sign”
- Rare exception in tibia - osteolysis may start in the diaphysis without subarticular or metaphyseal involvement
- Calvaria - same process = “osteoporosis circumscripta”
  - osteolysis assumes a well-defined, oval configuration
  - crosses sutures
  - predilection for frontal and occipital bones

1. Osteolytic Stage

“Candle Flame sign”



osteoporosis circumscripta

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2. Mixed Stage

- most commonly diagnosed stage
- cortical thickening
- osseous expansion
  - Long bones – sclerosis/expansion first seen in a subarticular or metaphyseal location, at the site of initial osteolysis
- loss of corticomedullary differentiation (due to endosteal new bone formation encroaching the medullary cavity)
- accentuated coarse trabecular markings
- advancing wedge of resorption may be seen
  - osteolytic front may be seen advancing towards the diaphysis and may be separated from bone showing signs of middle or late sclerotic phase by few cm
- The mixed lytic and blastic foci are referred to as:
  - a “cotton-wool” pattern in skull
  - “picture-frame” appearance when affecting the vertebrae

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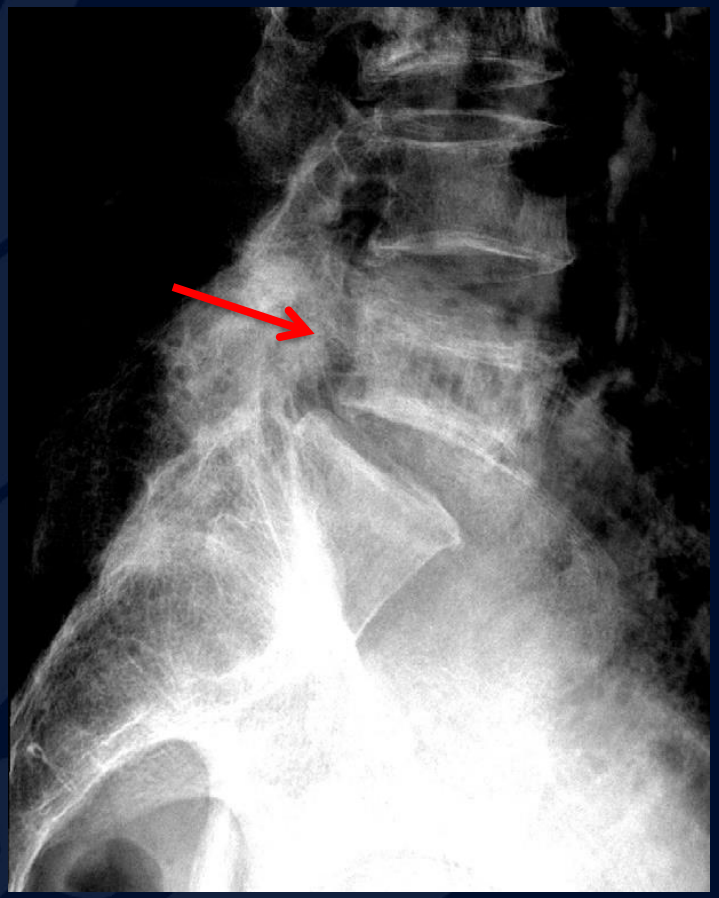
2. Mixed Stage



“Picture Frame Vertebra”
Coarse thick endplates, increased AP diameter, vertical coarse trabecular thickening

cortical thickening with encroachment of the medullary cavity, prominent coarse trabecular markings and osseous expansion in the first metatarsal

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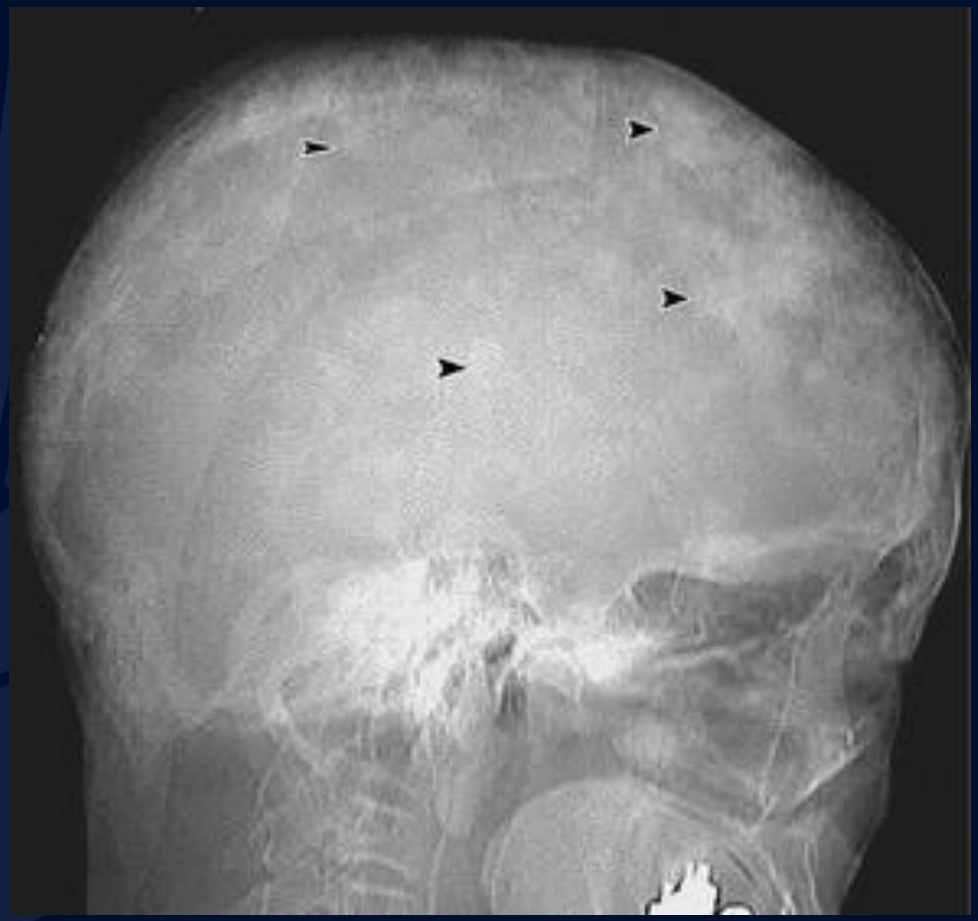
2. Mixed Stage



"Cotton Wool" – focal sclerosis

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3.Sclerotic Stage

- Considerable new bone formation
- Diffuse increase in density
- Medullary sclerosis and obliteration of medullary cavity
- Increased bone size
- Reactive sclerosis
- Thickened trabeculae
- No remodeling of incomplete insufficiency fracture
    - Contribute to abnormal lateral bowing of femur, anterior bowing of tibia

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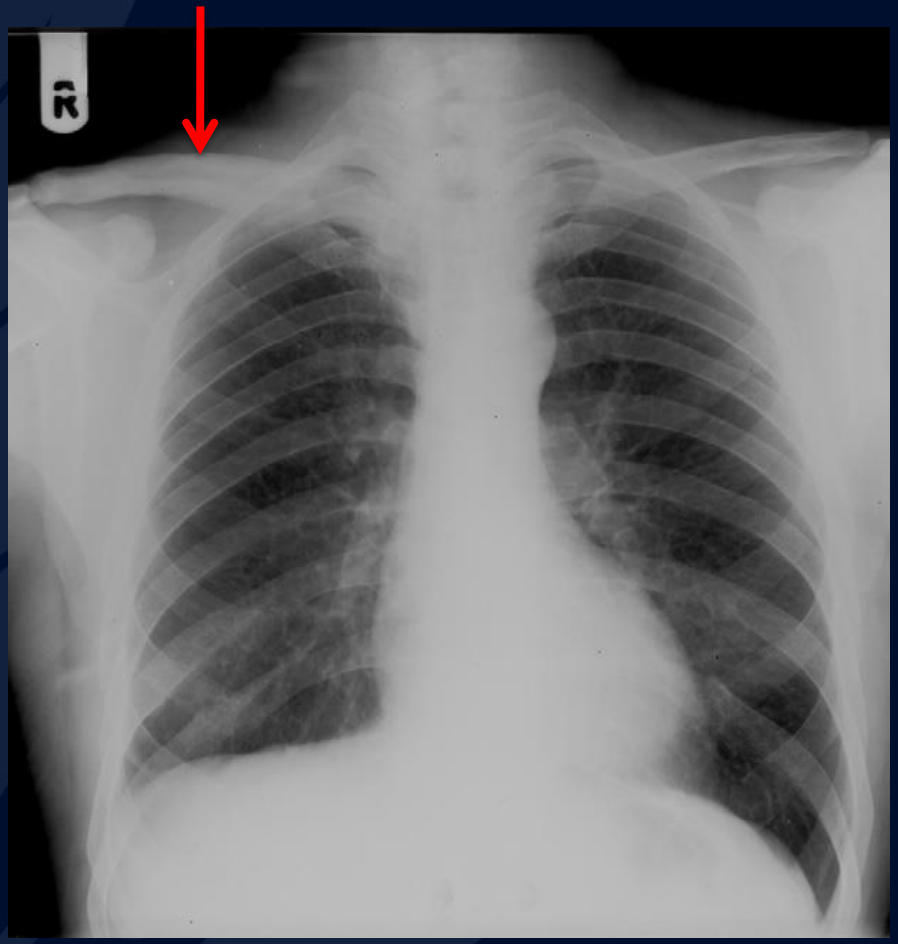
3. Sclerotic Stage



Sclerosis + expansion

"Tam O'Shanter" Sign

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Paget's Disease of Bone

- Distribution:
  - Polyostotic (65-90%)/Monostotic (10-35%)
  - Skull: 25-65%
  - Spine: 30-75%
  - Pelvis: 30-75%
  - Proximal long bones: 25-30%
- ≤4% individuals under 40yo; ≤11% over 80yo.
- M:F 3:2

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Paget's Disease of Bone Con't

- Presentation: MOST (3/4) are asymptomatic
  - Localized pain/tenderness
  - Increased focal temperature due to hypervascularity
  - Increased bone size (changing hat size)
  - Bowing deformities
  - Kyphosis
  - Decreased ROM
- Dx: ↑ALP (reflects osteoblastic activity), normal Ca/P, ↑urine hydroxyproline; CR
- Rx:
  - Second generation bisphosphonates inhibit bone resorption; promote healing of osteolytic lesions and improve bone pain
  - Mithramycin (cytotoxic antibiotic)- reserved for those resistant to other forms of medical treatment
  - Pain management with analgesics/NSAID

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Complications

- osseous weakening => deformity and pathological fractures
  - incremental stress fractures aka “banana fractures” or “pseudofractures”
- Increased risk/accelerated OA
- Malignant sarcomatous transformation (0.3% of cases)
  - Longstanding polyostotic dz
  - Osteosarcomas- femur/pelvis/humerus ; poor prognosis
  - “pseudosarcoma” – focal prolif of periosteal new bone in the context of Paget’s dz might be radiographically indistinguishable from malignant transformation – need negative bx
  - Fibrosarcoma, chondrosarcoma, malignant fibrous histiocytoma
  - Giant cell tumors- skull/facial bones

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Complications Cont

- SN/Conductive hearing loss – temporal bone involvement
- Cranial nerve palsies – skull base foramina encroached upon
- Basilar Invagination – brainstem compression, hydrocephalus
- High output CHF – due to osseous hypervascularity in polyostotic dz
  - when bone involvement >15%
- Hyperparathyroidism (~10%)

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Differential Diagnosis

- Sclerotic Mets- Blastic lesions in same distribution as Paget disease
  - No trabecular coarsening or enlargement of bone
- Fibrous Dysplasia - may enlarge bone
  - No trabecular coarsening or cortical thickening
- Multiple Myeloma - early lytic lesions like Paget's disease of bone
  - No bone/trabeculae enlargement
- Myelofibrosis – sclerotic
  - No bone enlargement

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References

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