Ewing sarcoma

- Second most common malignant primary bone tumors of childhood after osteosarcoma
- Typically arising from the medullary cavity with the invasion of the Haversian system.
- Ewing sarcomas usually present as moth-eaten, destructive, and permeative lucent lesions in the shaft of long bones, with a large soft tissue component and typical onion skin periostitis.
- These tumors may also involve flat bones and can appear sclerotic in up to 30% of cases.

Primary Localization

Bone (~80%)
Axial skeleton (45%)
- Pelvis (20%)
- Ribs (10%)
- Other axial bones (15%)

Distal skeleton (35%)
- Femur (12%)
- Humerus (4%)
- Other distal bones (19%)

Extraosseous location (~20%)
Mostly paravertebral and thoracic soft tissues. Nonskeletal primary cancers have been documented in the retroperitoneum, esophagus, pancreas, ileum, kidney, bladder, vagina, uterus, penis, adrenal gland, lung, breast, spinal cord, orbit, and intracranial tissue.

Primary site tumor cells

Main Metastatic Sites

- Lungs
- Bone
- Bone marrow

Metastatic site tumor cells

Radiologic Findings

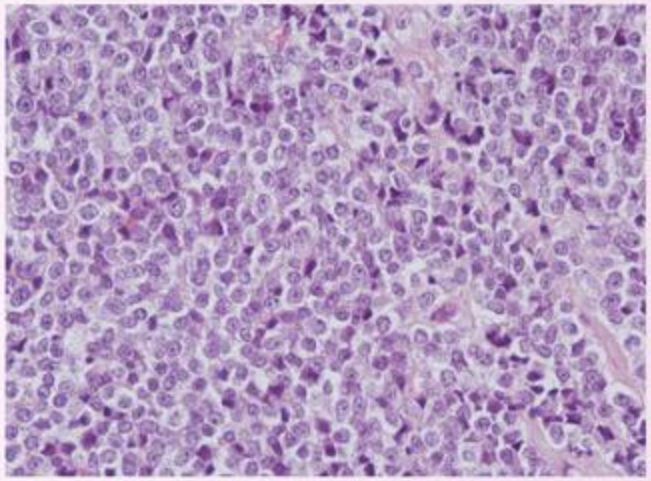
Codman triangle
New subperiostal bone growing on the tumor

"Moth eaten" pattern
Permeative destruction of bone due to multiple lytic lesions

"Onion peel" appearance
Delicate laminations constituting the periosteal layers

Histology

Poorly differentiated tissue consisting of small, round, blue cells with prominent nuclei and minimal cytoplasm



Ewing sarcoma

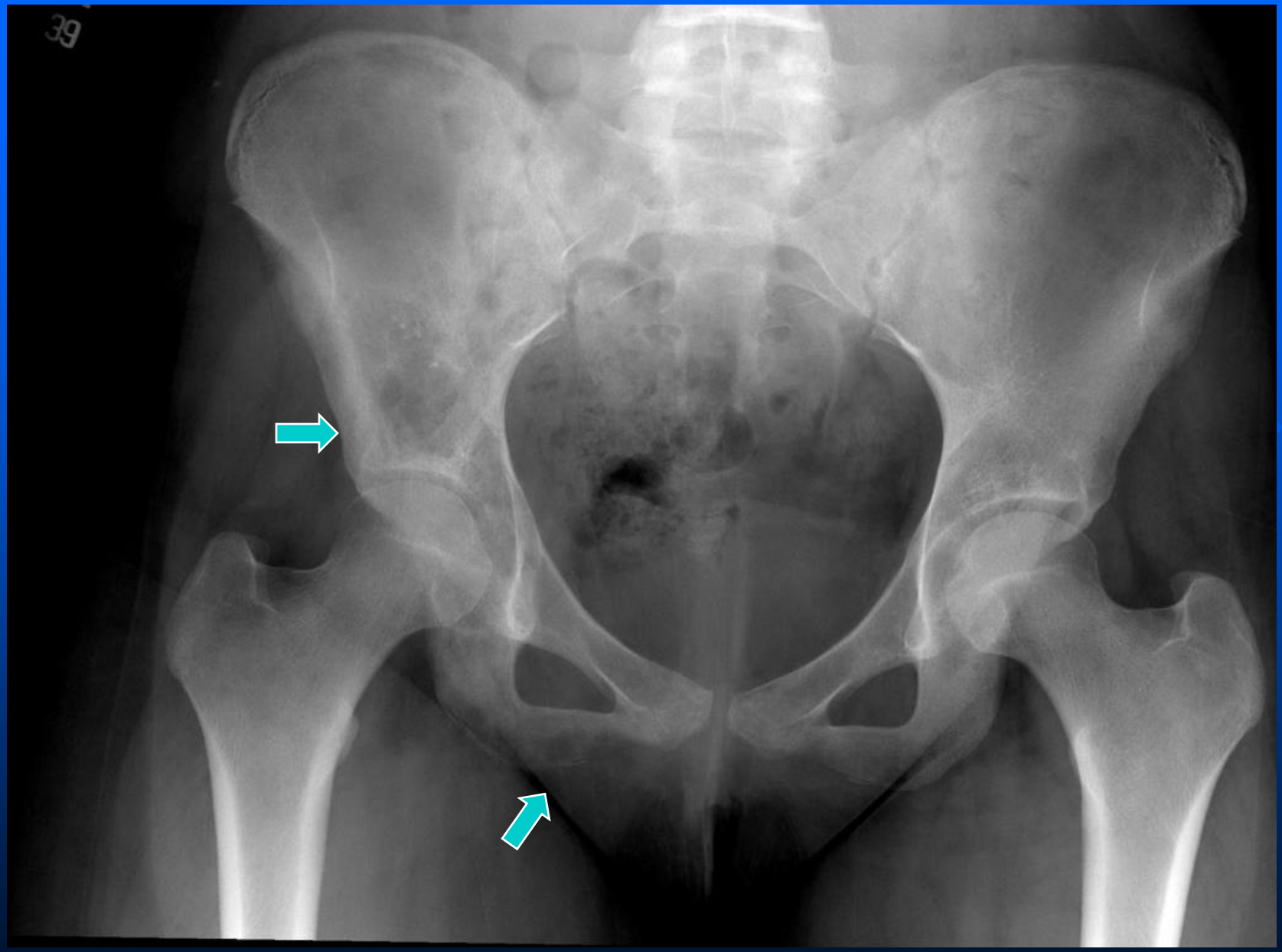
- Small round blue cell tumor with regular-sized primitive-appearing cells.
- Closely related to the soft tissue tumors
  - pPNET
  - Askin tumor
  - Neuroepithelioma
  - Collectively are referred to as Ewing sarcoma family of tumors (ESFT)

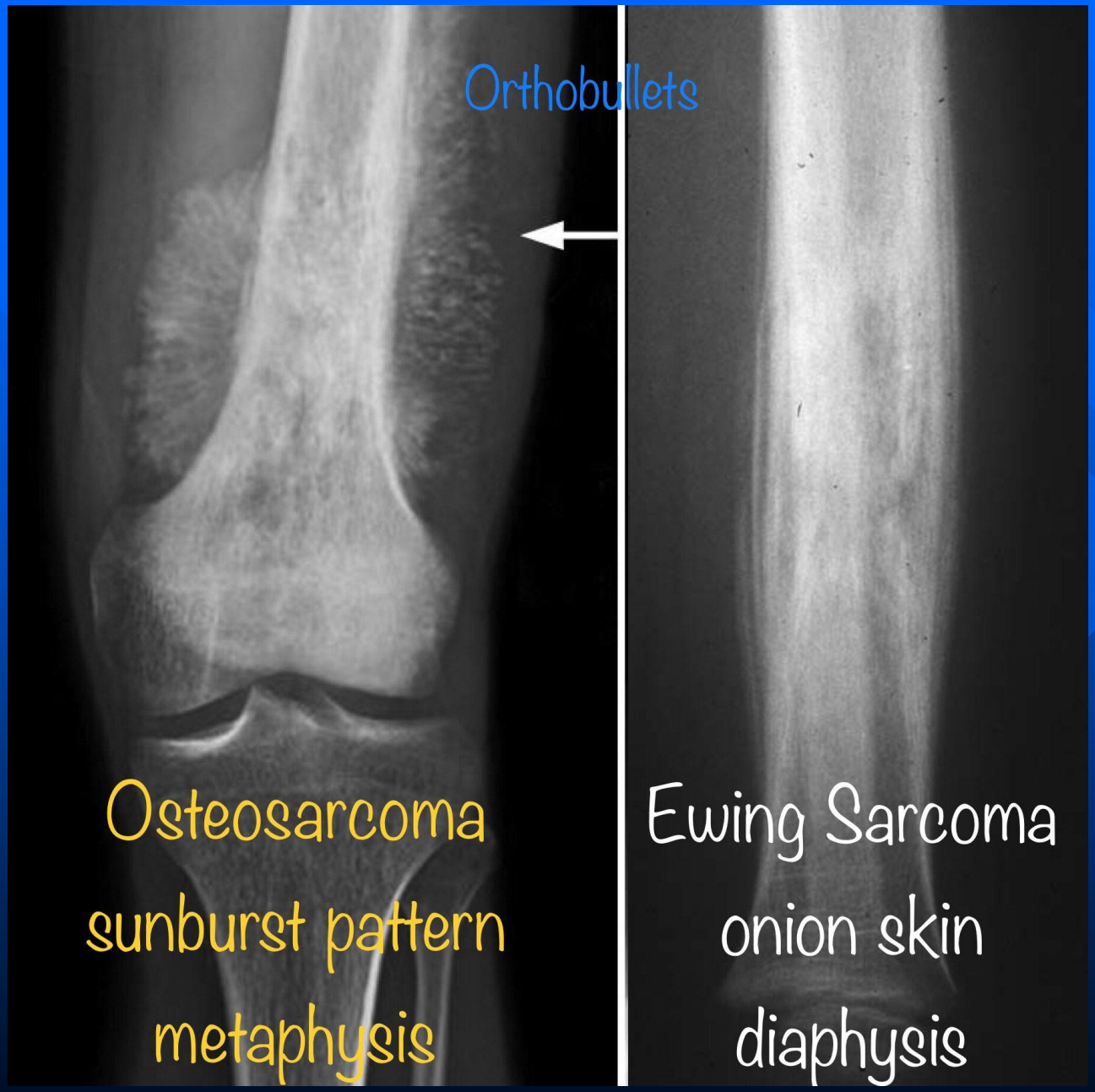
Ewing sarcoma

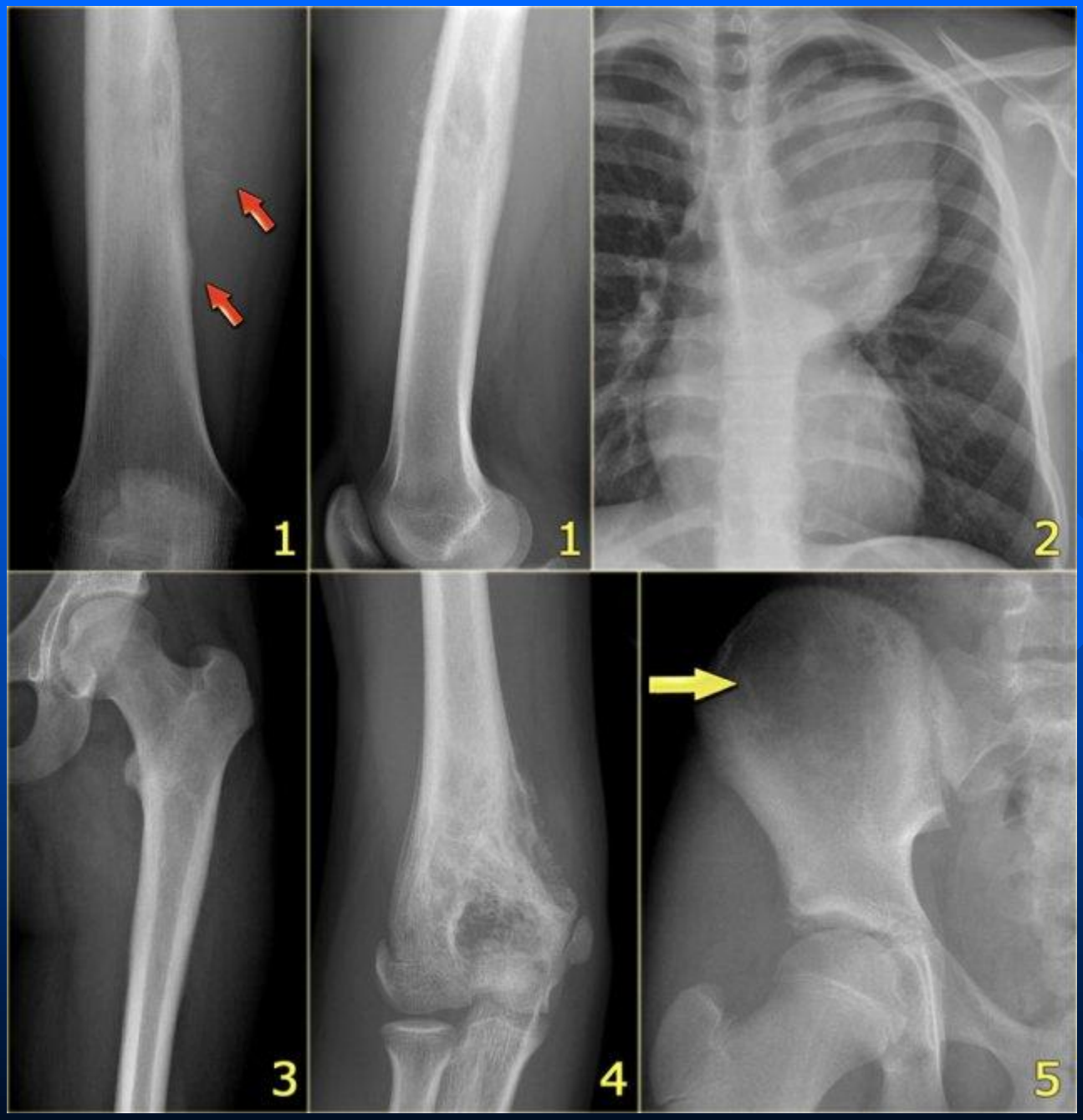
- Age
  - Range: 5-30 years old (median: 13)
  - 80% < 20 years old
- Sex
  - M > F (1.5:1)
- Ethnicity
  - Extremely rare in Black patients (0.5-2% of cases)
- Most common signs/symptoms
  - Painful (82-88%) mass (60%)
  - Other signs/symptoms
  - Fever (20-49%), anemia, leukocytosis, ↑ ESR (43%)
    - » May elevate clinical suspicion of infection
  - Pathologic fracture uncommon (5-15%)

DDx:

- Osteomyelitis
- Langerhans cell histiocytosis
- lymphoma




Orthobullets
Osteosarcoma
sunburst pattern
metaphysis
Ewing Sarcoma
onion skin
diaphysis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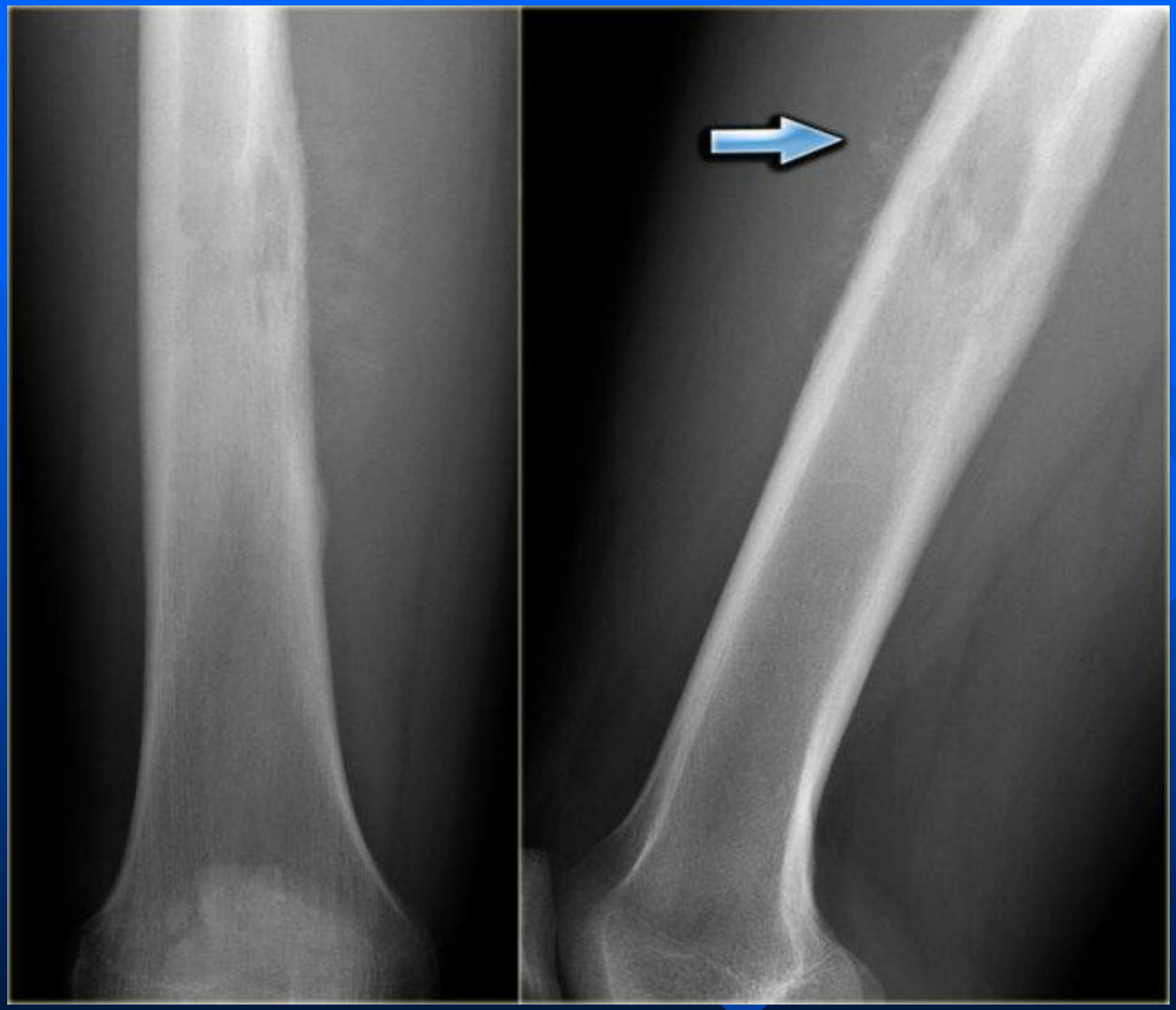

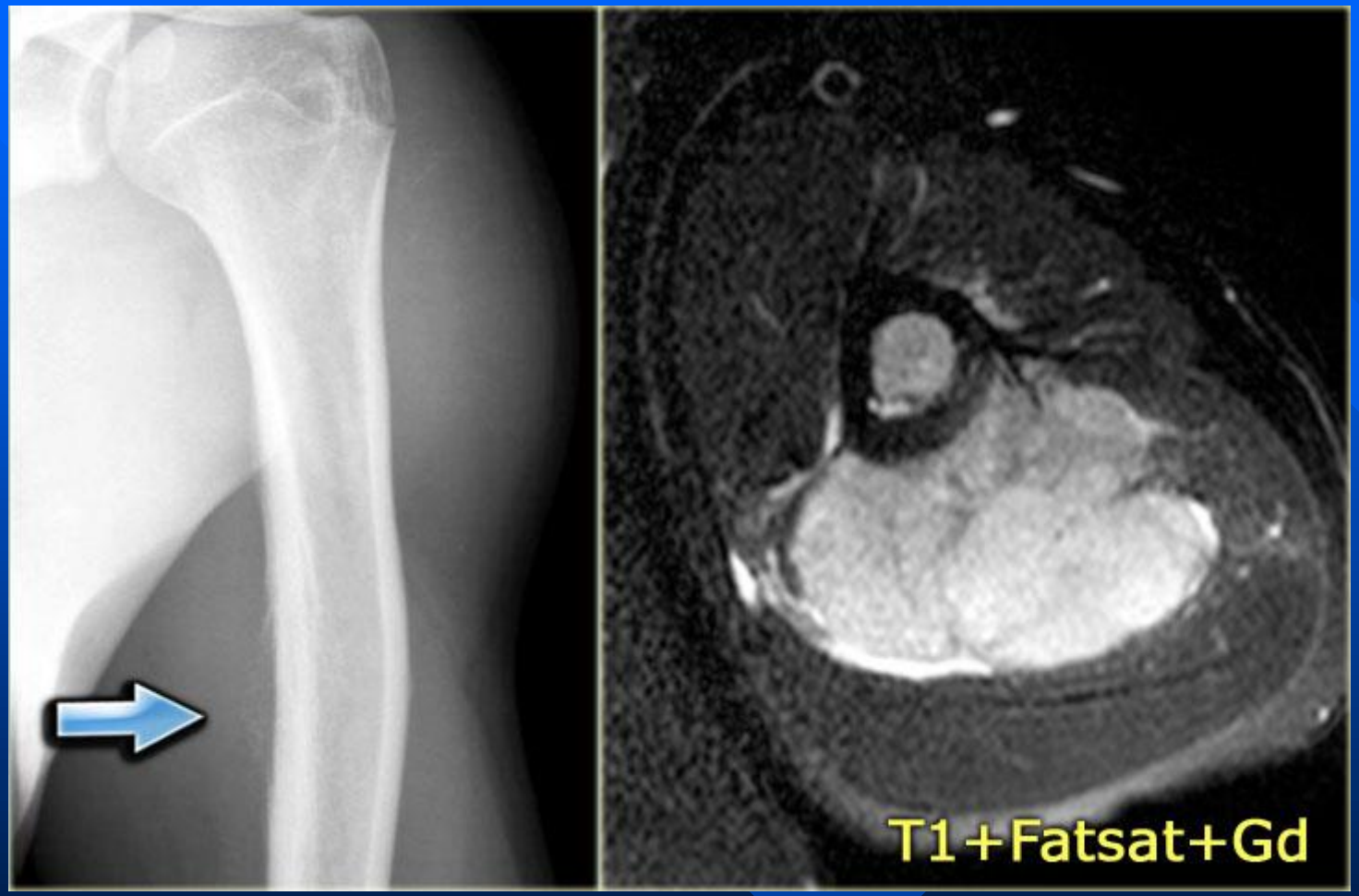
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