Journal Club
Topic: Osteosarcoma

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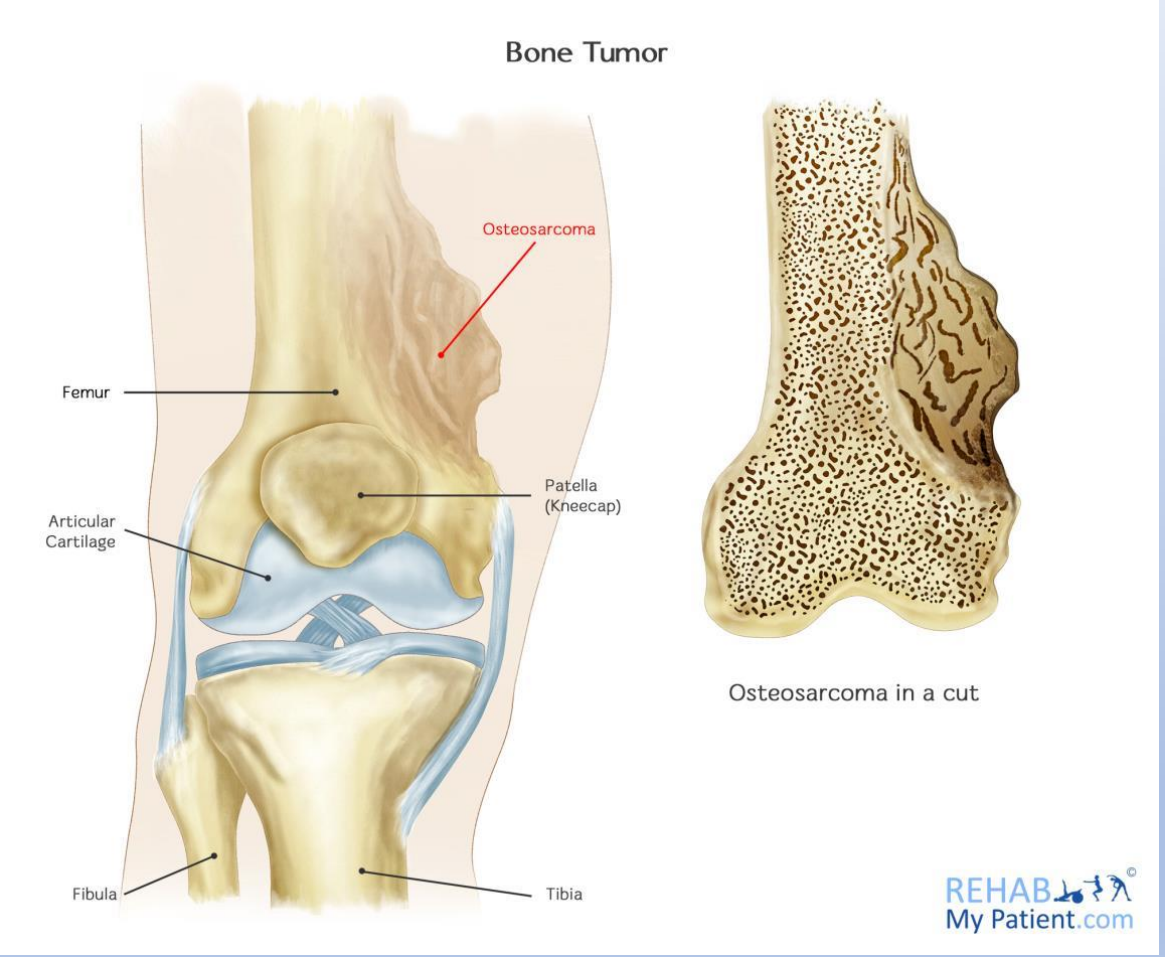
Introduction

- Osteosarcoma (also known as osteogenic sarcoma) is malignant bone forming tumor.
- It is the second most common primary bone tumor after multiple myeloma, accounting for approximately 20% of all primary bone tumors.



Classification

- Primary Osteosarcoma

➢Originate from bone cells.

➢Typically occurs **in young patients (10-20 years)** because the growth centers of the bone are more active during puberty/adolescence.

➢Slight male predominance.

- Secondary Osteosarcoma

➢Secondary to other conditions, including Paget disease, fibrous dysplasia, extensive bone infarcts, post-radiotherapy, osteochondroma, and osteoblastoma.

➢**Occurs among the elderly.**

Classification

The **WHO classification of bone tumors** lists the histologic types of osteosarcoma.

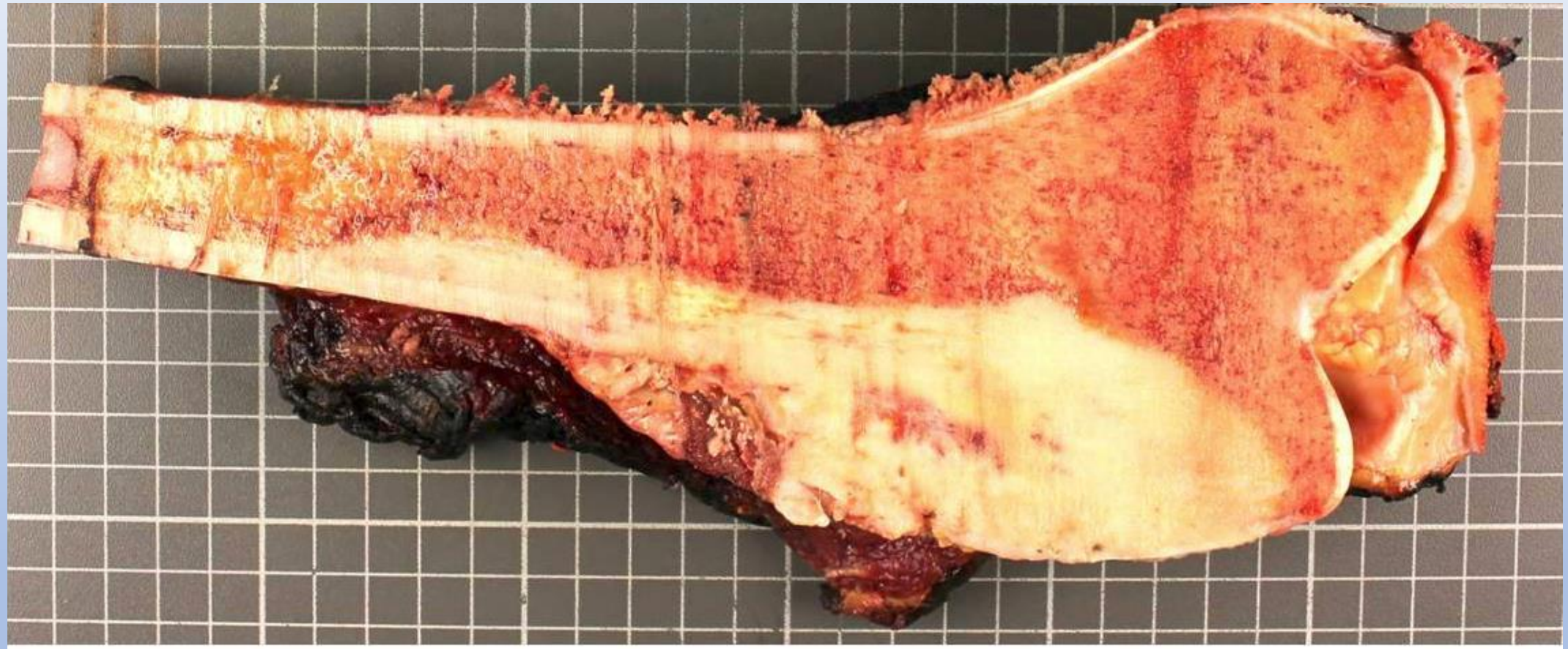
Primary osteosarcoma can be further divided to:

- Intramedullary/central
  - conventional osteosarcoma: most common (75-80%)
  - low-grade central osteosarcoma
  - telangiectatic osteosarcoma
  - small cell osteosarcoma
- Surface
  - parosteal osteosarcoma
  - periosteal osteosarcoma
  - high-grade surface osteosarcoma

Macroscopic Appearance

- Bulky tumors where a heterogeneous cut surface demonstrates areas of hemorrhage, fibrosis and cystic degeneration.
- Extension into intramedullary canal and subperiosteal region.
- Varying degree of new bone formation.

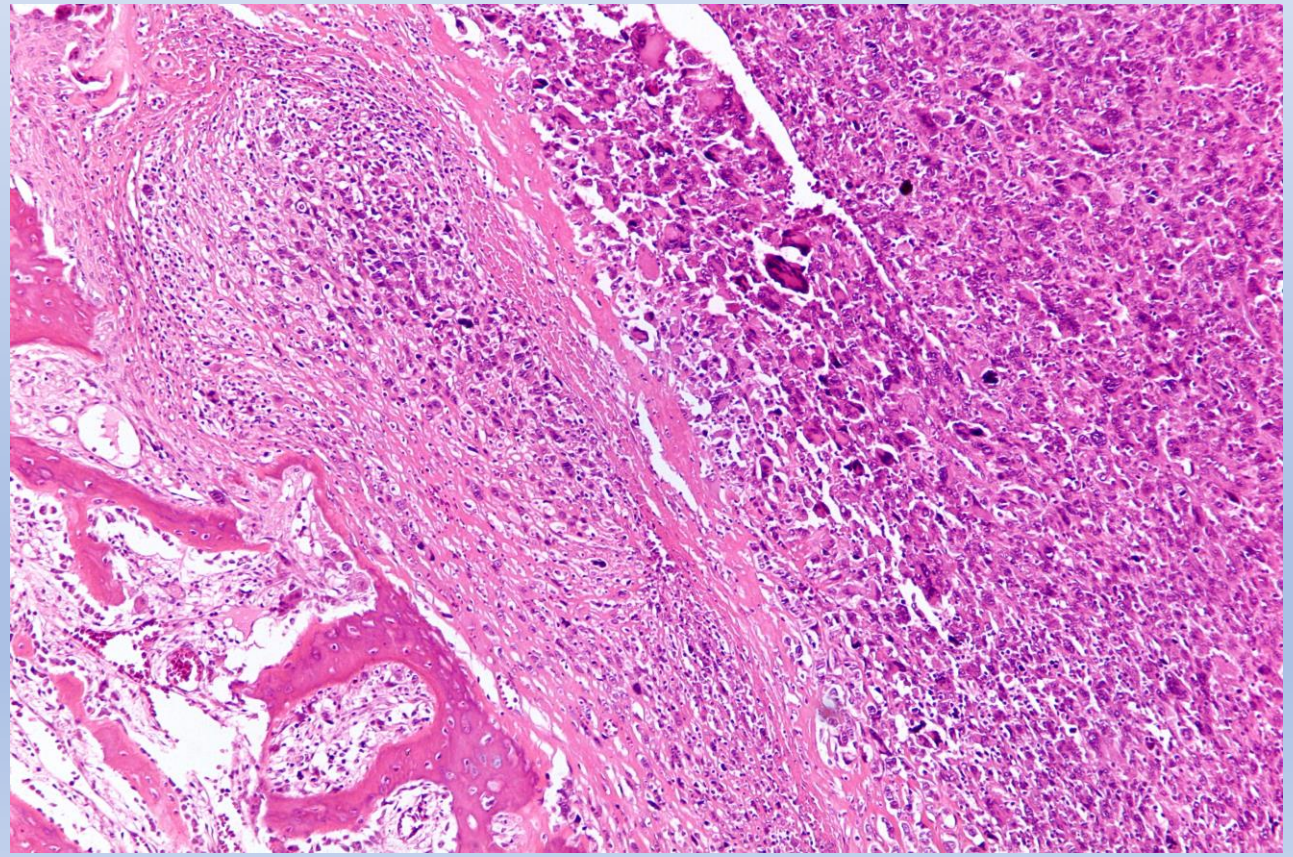
Macroscopic Appearance



This low grade central osteosarcoma arose in the distal femoral metaphysis and invaded through the cortex and into the adjacent soft tissue

Microscopic Appearance

Poorly formed trabecular bone is seen with cellular pleomorphism and mitoses. Variable amounts of fibrocystic and chondroblastic appearing cells may also be encountered.



Intermediate magnification micrograph of an osteosarcoma (center and right of image) adjacent to non-malignant bone (left-bottom of image)

Location

Primary osteosarcomas typically occur at the **metadiaphysis of long bones** in the appendicular skeleton, most commonly at the following sites:

- femur: ~40% (especially distal femur)
- tibia: ~16% (especially proximal tibia)
- humerus: ~15%

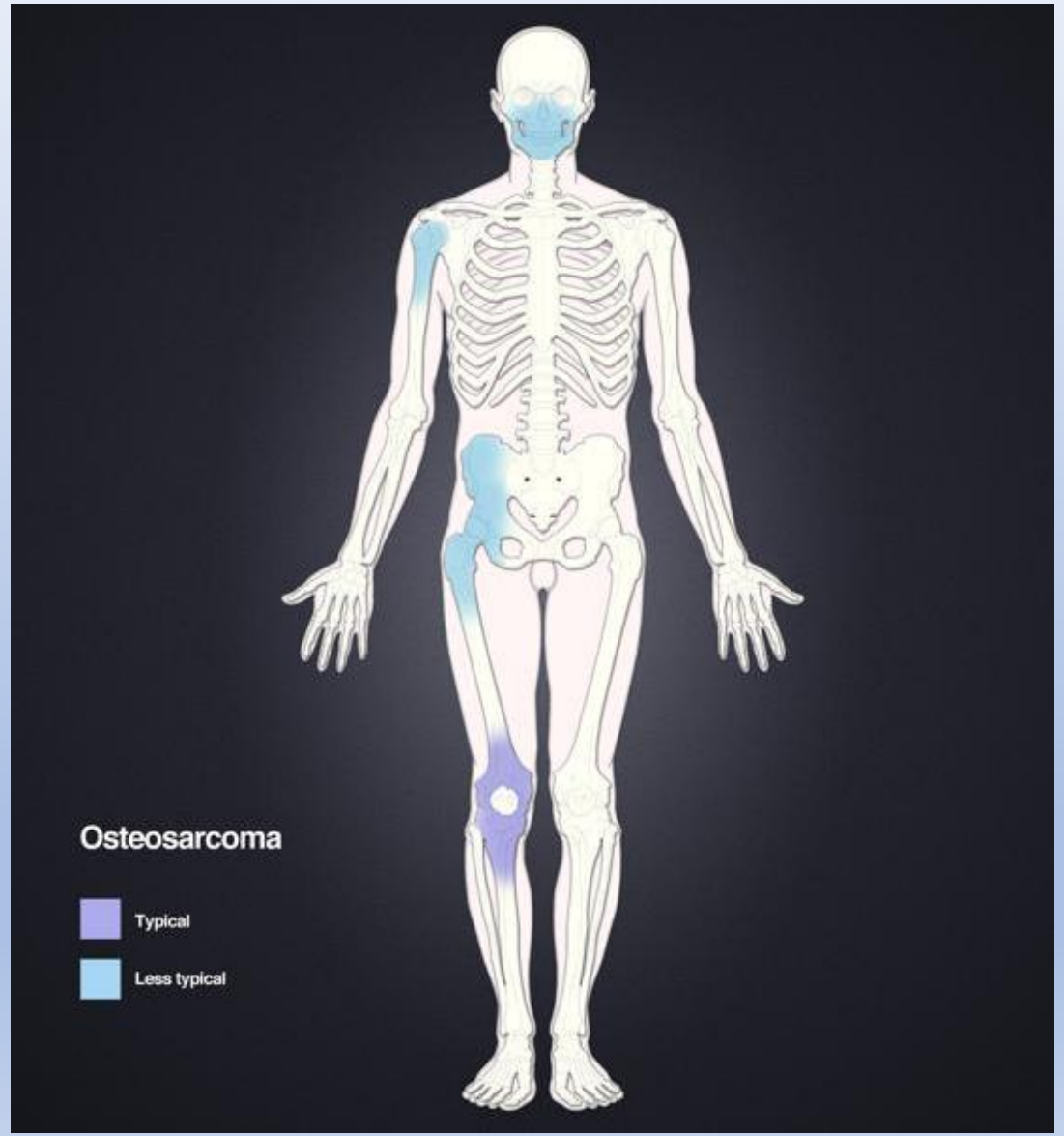
Location

Other less common sites of primary osteosarcoma:

- fibula
- innominate bone (i.e. os coxae)
- mandible
- maxilla
- vertebrae

Secondary osteosarcoma, on the other hand, have a much wider distribution, largely mirroring the combined incidence of their underlying conditions, and thus much have a **higher incidence in flat bones**, especially the pelvis (a favorite site of Paget disease).

Location



Radiographic Features

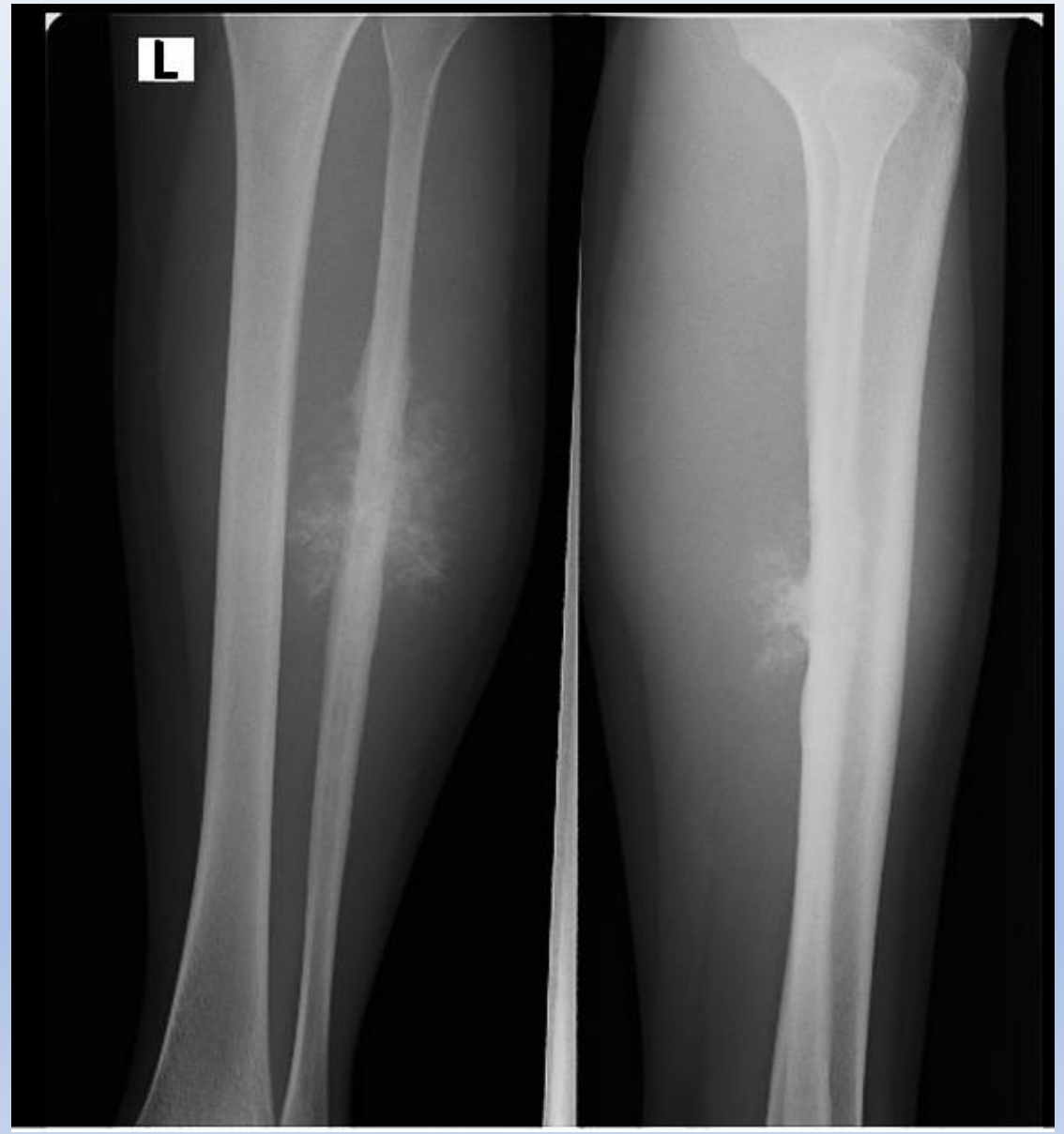
- **Conventional radiography** continues to play an important role in diagnosis.
- **MRI** is used for local staging by assessing intraosseous tumor extension (e.g. growth plate/epiphysis, skip lesions) and soft-tissue involvement.
- **CT and bone scanning** have a role in distant staging.

Plain Radiograph

- Medullary and cortical bone destruction
- Wide zone of transition, permeative or moth-eaten appearance
- Aggressive periosteal reaction
  - sunburst type
  - Codman triangle
  - lamellated (onion skin) reaction: less frequently seen
- Soft-tissue mass
- Tumor matrix ossification/calcification

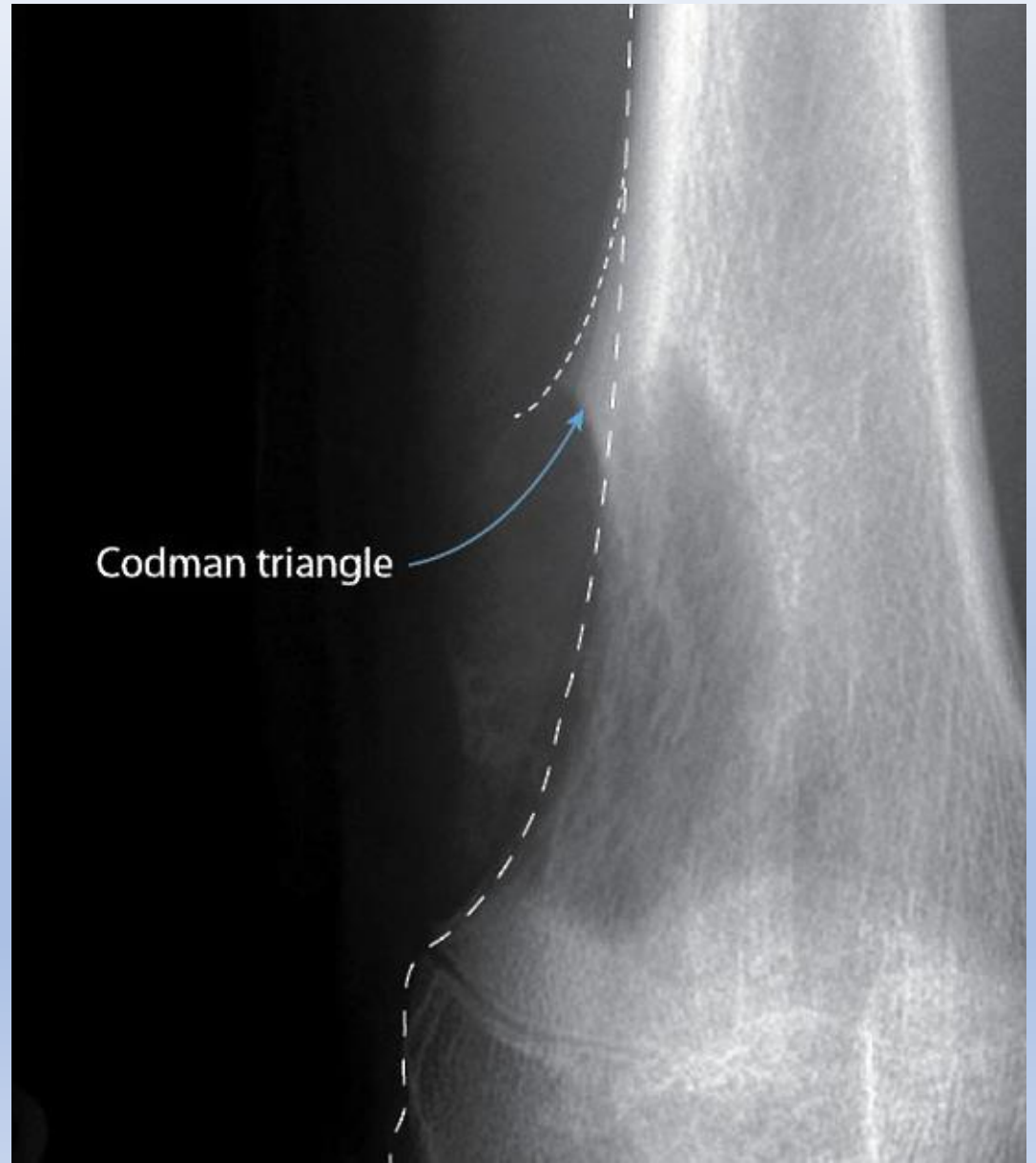
Sunburst Appearance

A type of periosteal reaction which occurs when the lesion grows too fast and the periosteum does not have enough time to lay down a new layer and instead the Sharpey's fibers stretch out perpendicular to the bone.



Codman Triangle

A type of periosteal reaction seen when the periosteum does not have time to ossify with shells of new bone in aggressive lesions, so only the edge of the raised periosteum will ossify.



Lamellated (Onion Skin) Reaction

Periosteal reaction which demonstrates multiple concentric parallel layers of new bone adjacent to the cortex, reminiscent of the layers on an onion. The layers are thought to be the result of periods of variable growth.

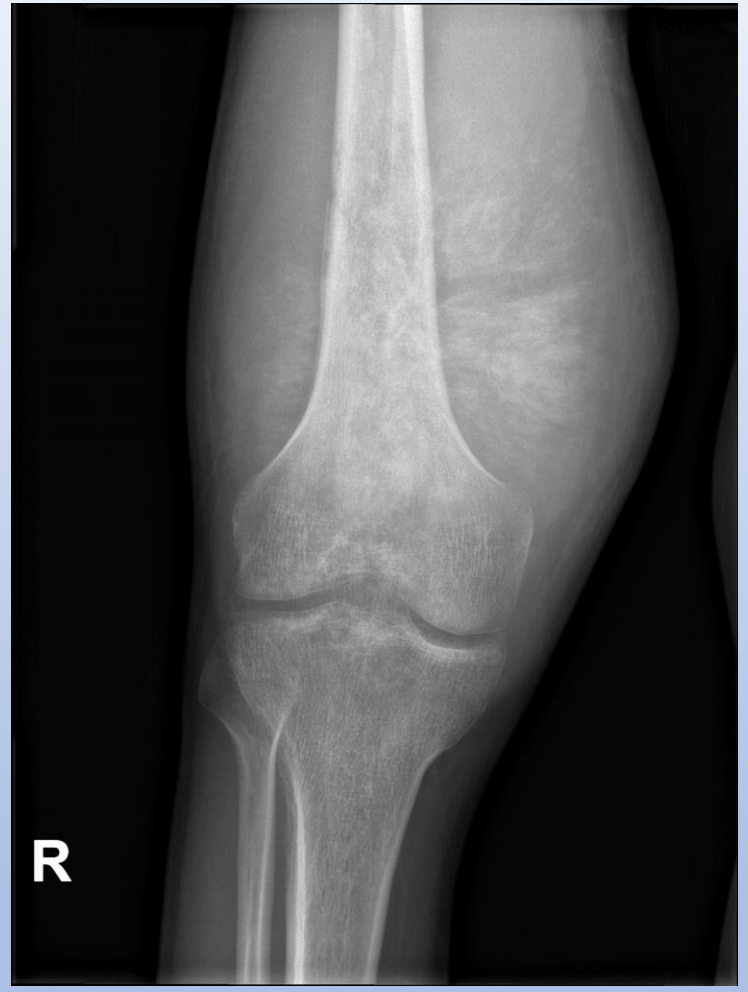
Radiographic Appearance

The distal half of the femur is occupied and expanded with a heterogeneous mass with areas of bone formation. Posterior the periosteum is elevated (Codman's triangle).

Radiographic Appearance

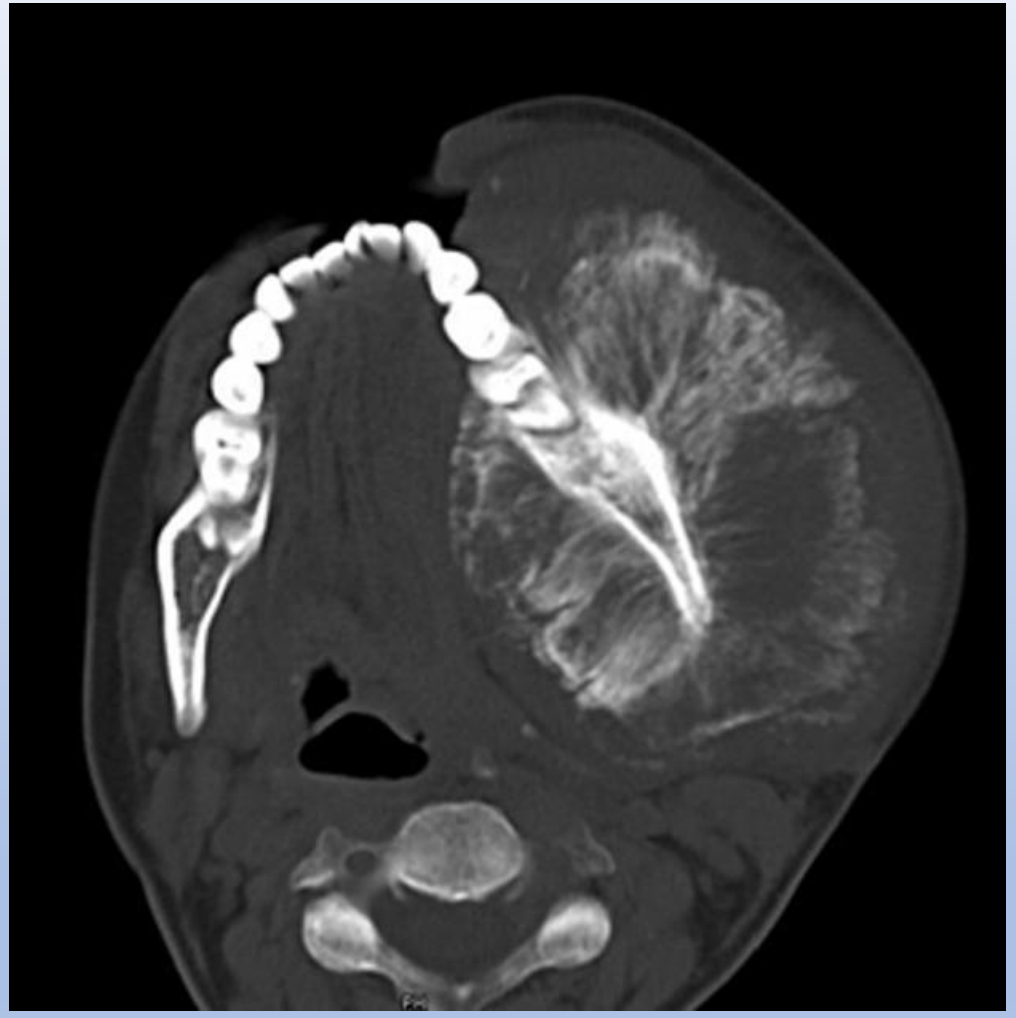
Radiographs showing sclerotic lesion involving dia-metaphyseal region of the tibia with a wide zone of transition, osteoid matrix, periosteal elevation (Codman Triangle) and characteristic " Sunburst " type of periosteal reaction.

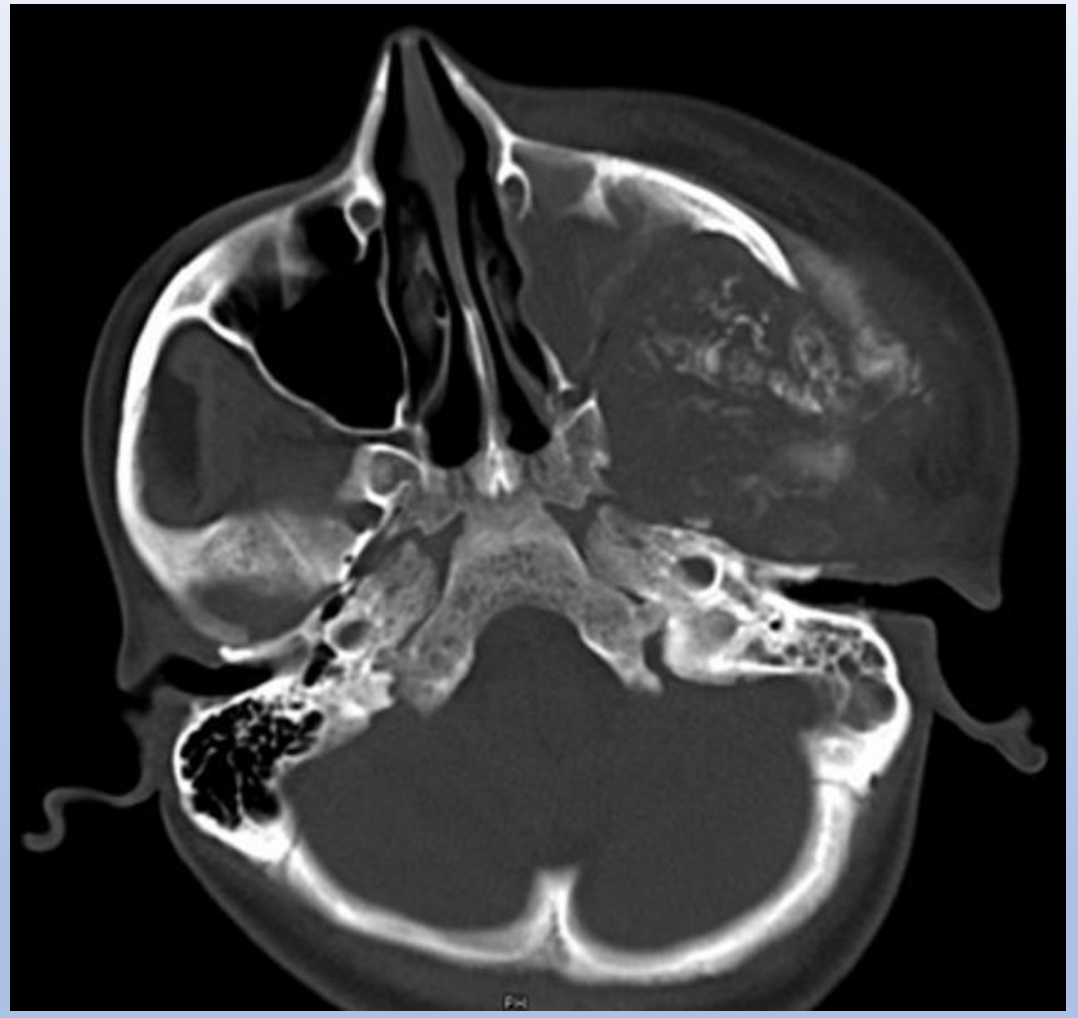
Radiographic Appearance



An ill-defined sclerotic area affecting the right distal femoral shaft with an aggressive sunburst type of periosteal reaction. The mass is associated with significant swelling of the adjacent soft tissues. Findings are suggestive of osteosarcoma.

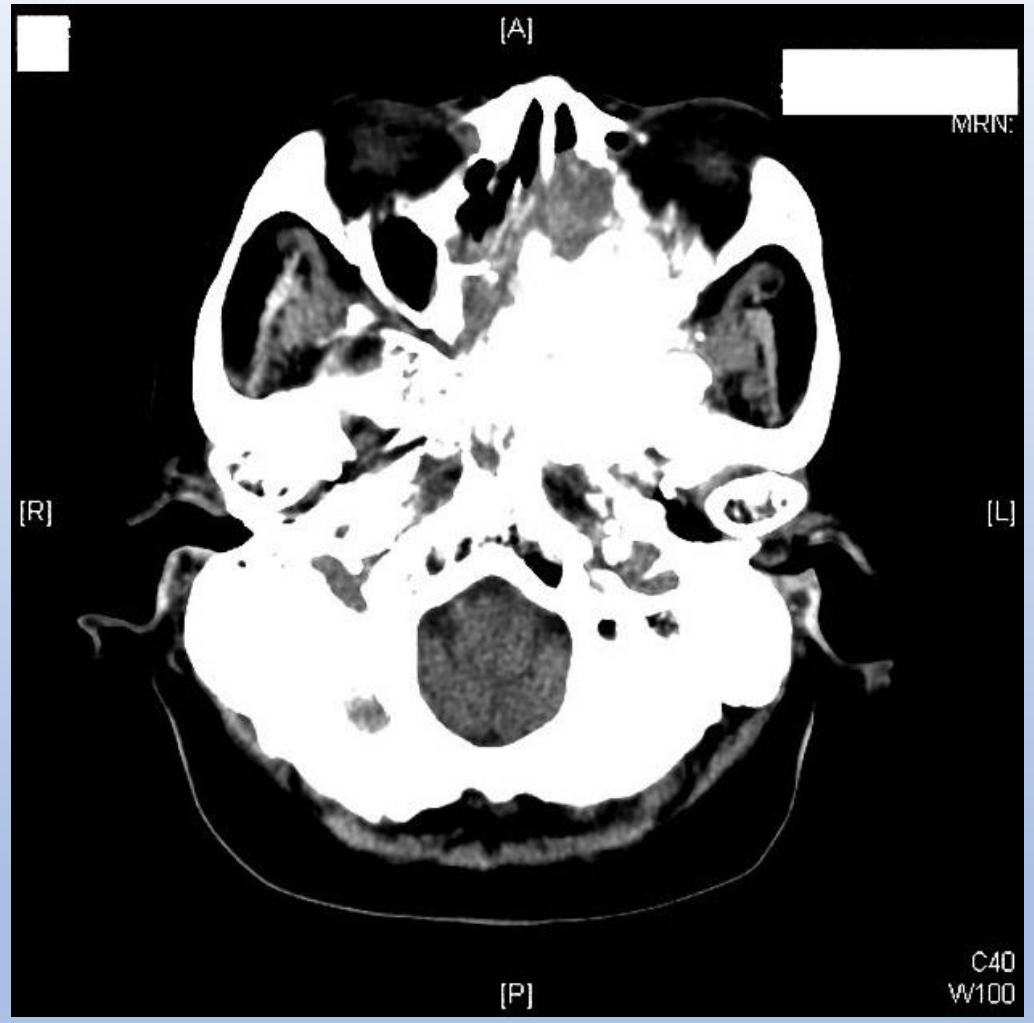
CT Appearance – Case 1

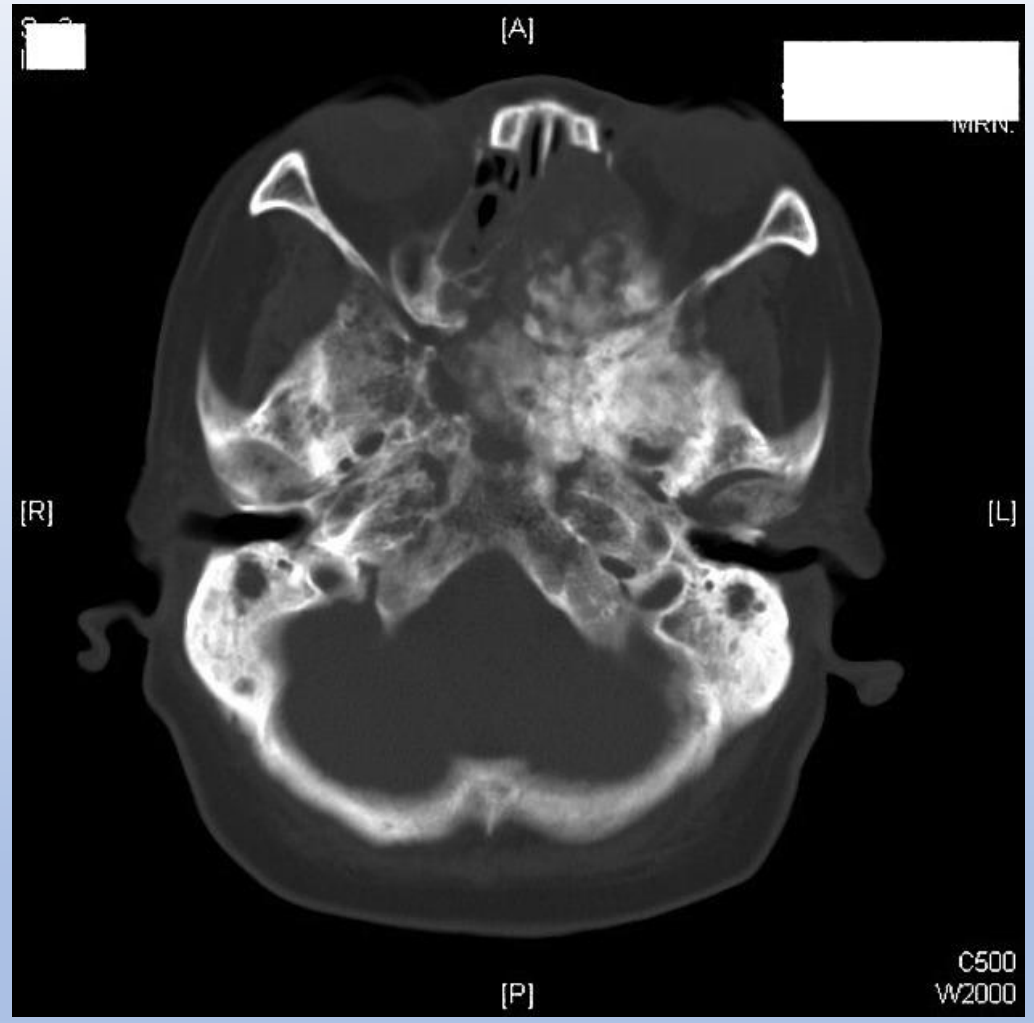




Gnathic osteosarcoma: Left maxillary fullness, pterygoid plate involvement and mandibular destruction with sunburst appearance. Associated soft tissue swelling.

CT Appearance – Case 2





Post radiation osteosarcoma: Large sclerotic bone lesion at skull base, associated with soft tissue mass and interval growth.

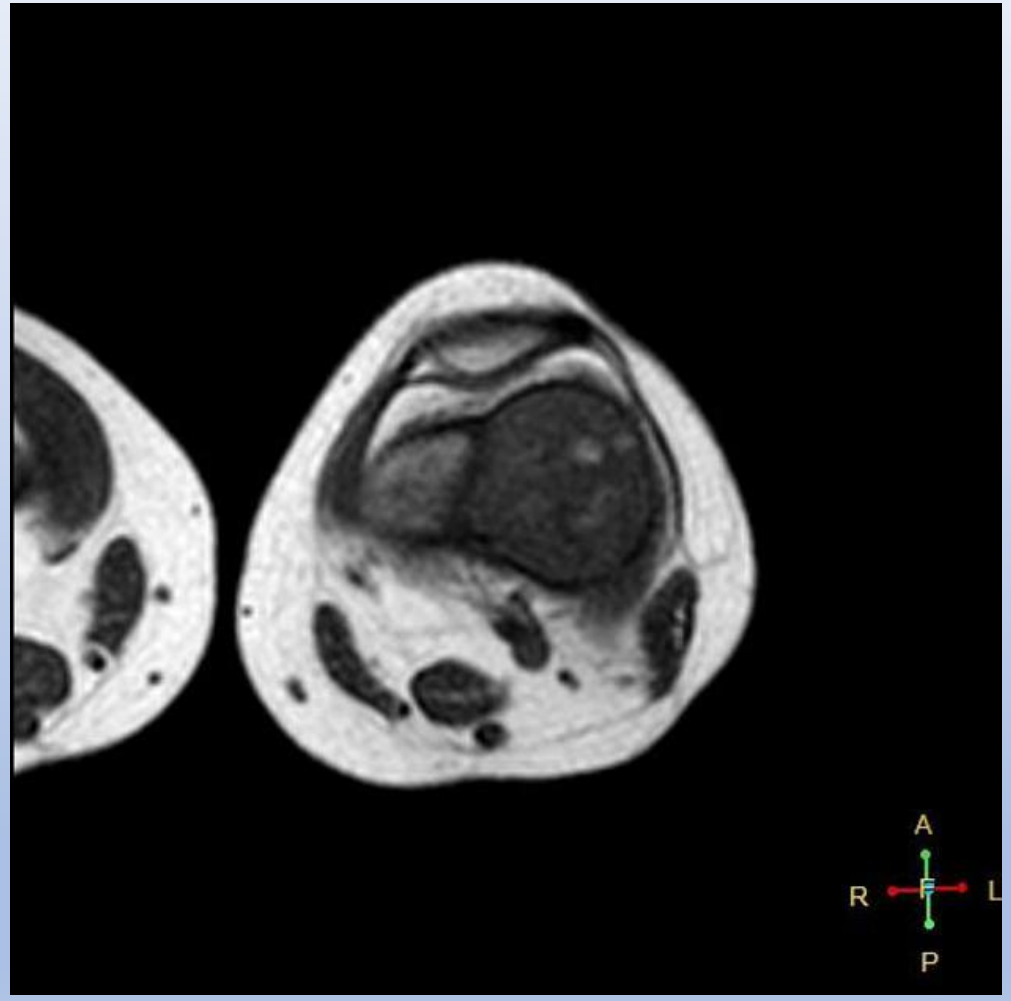
MRI

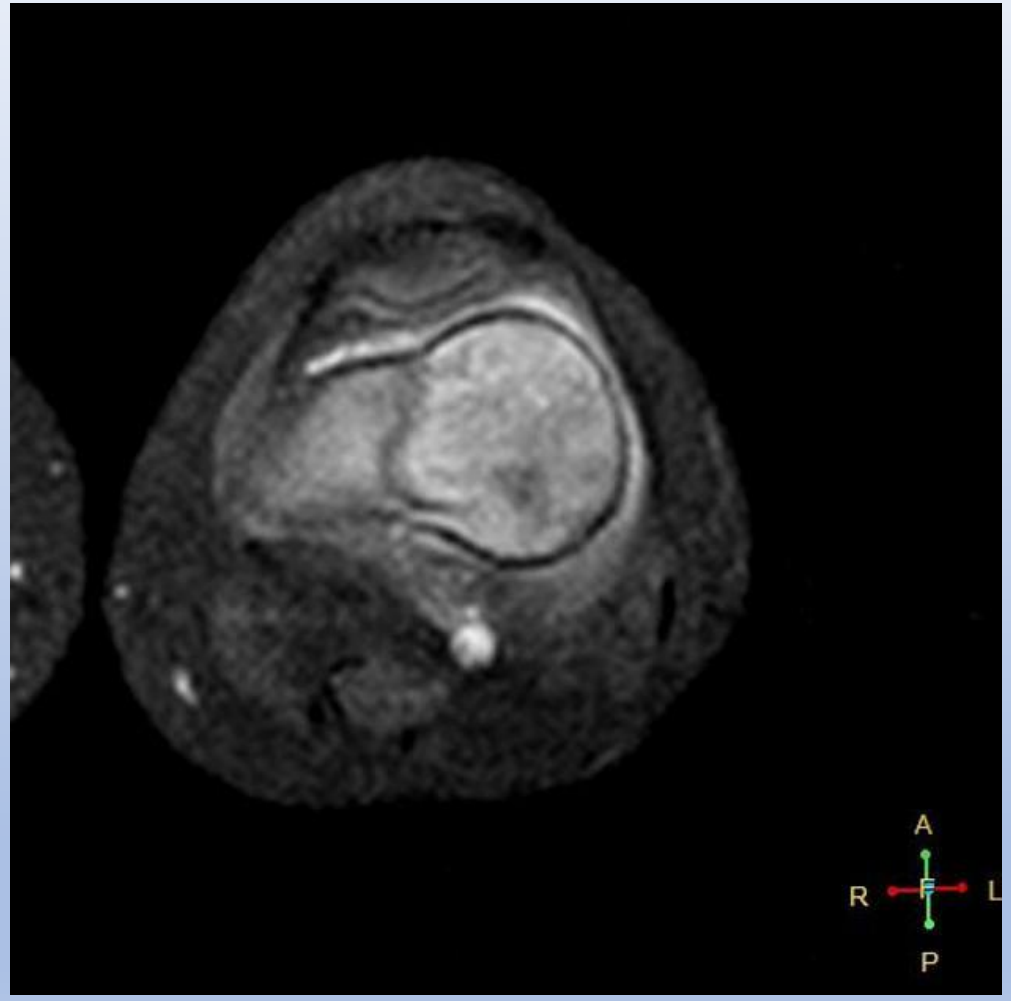
- An essential tool to determine accurate local staging and assessment for limb-sparing resection, particularly for evaluation of intraosseous tumor extension and soft-tissue involvement.
- Evaluation of the growth plate is also essential as up to 75-88% of metaphyseal tumors do cross the growth plate into the epiphysis.

MRI – Case 1

Distal femoral metaphyseal eccenteric aggressive marrow lesion that displays low signal in T1 and heterogenous signal in T2, high signal in STIR. There is associated cortical destruction, extraosseous soft tissue extension, periosteal infiltration and marrow edema.

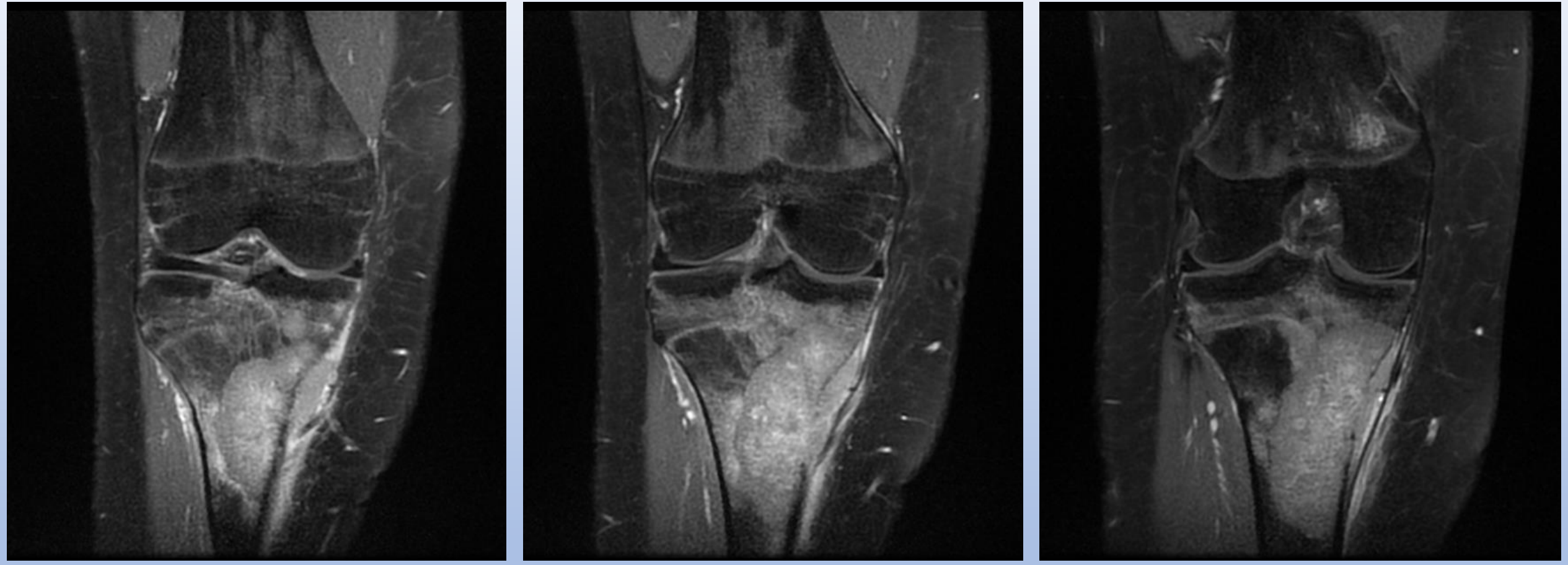
MRI – Case 1 (Continuation)





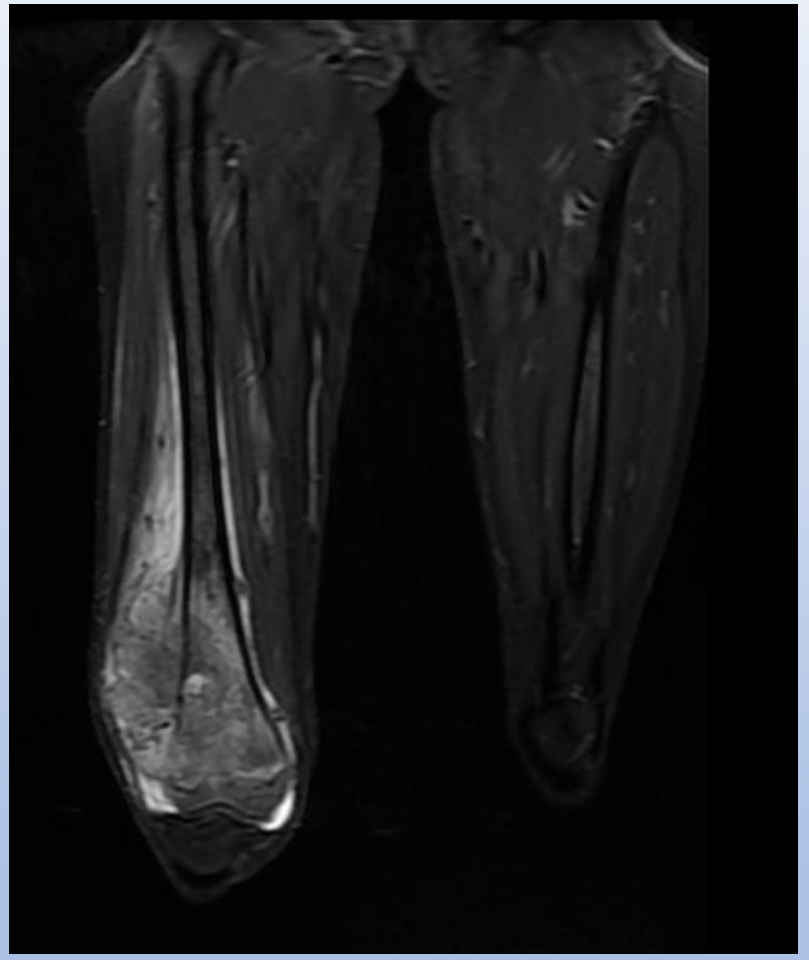
Pre-contrast axial T1W image (left) and avid enhancement in post-contrast T1W FS image (right)

MRI – Case 2



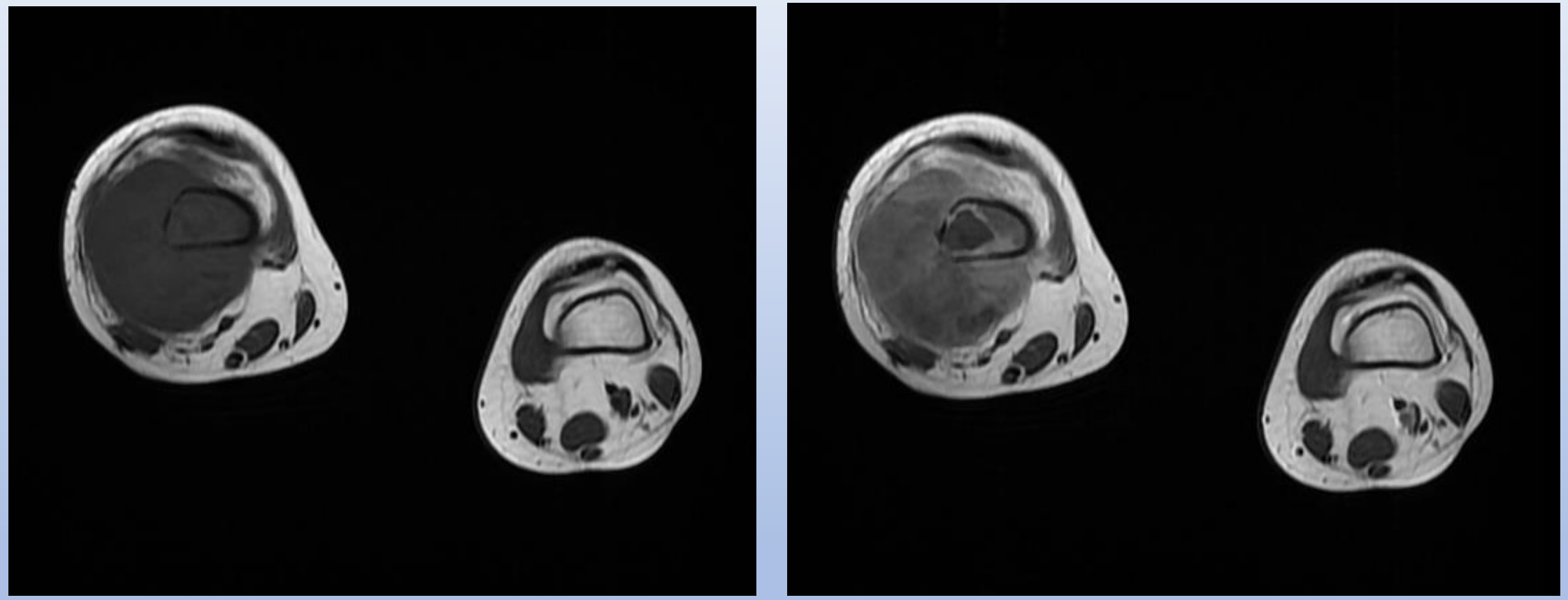
Post contrast coronal T1W FS: Heterogenous enhancing mass in the proximal medial tibial metaphysis eroding the cortex and raising the periosteum. Multiple additional satellite masses are in the nearby bone.

MRI – Case 3



A lesion at postero-lateral aspect of distal right femoral shaft. It elicits isointense signal on T1, heterogeneous signal on T2 and high STIR signals with internal break-down foci of fluid signal. The lesion causes cortical destruction and extra-osseous eccentric soft tissue component.

MRI – Case 3 (Continuation)



Pre-contrast axial T1W (left) and post-contrast axial T1W (right). The lesion and the soft tissue component show heterogeneous contrast enhancement.

Treatment

- Cure, if achievable, requires aggressive surgical resection often with amputation followed by chemotherapy.
- If a limb-salvage procedure is feasible, a course of multidrug chemotherapy precedes surgery to downstage the tumor, followed by wide resection of the bone and insertion of an endoprosthesis.

Outcome

- Currently, the 5-year survival rate after adequate therapy is approximately 60-80%.
- The most frequent complications of conventional osteosarcoma are pathologic fracture and the development of metastatic disease.
- Common sites of metastasis include lungs (around 50%), bones and regional lymph nodes.

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Thank you