Malignant bone tumors

Classification (W.H.O.)

- **Bone-forming tumours**
- **Cartilage forming tumours**
- **Giant-cell tumour**
- **Marrow tumours**
- **Vascular tumours**
- **Secondary malignant tumours of bone**

Bone forming tumors

- Benign-
  - Osteoma
  - Osteoid osteomaor osteoblastoma
- Intermediate - Aggressive osteoblastoma
- Malignant –
  - Osteosarcoma Central (Medullary) and Peripheral (Surface)
    - Parosteal
    - Periosteal
    - High grade surface

Cartilage forming tumors

Benign –

- Chondroma/Enchondroma
- Osteochondroma
- Chondroblastoma
- Chondromyxoid fibroma

Malignant -

- Chondrosarcoma
- Differentiated chondrosarcoma
- Juxtacortical chondrosarcoma
- Mesenchymal chondrosarcoma
- Clear cell chondrosarcoma

Giant cell tumor

- Osteoclastoma

Marrow tumors

- Ewing’s sarcoma
- Neuroectodermal tumour
- Malignant lymphoma of bone (Primary/secondary)
- Myeloma

Vascular tumors

Benign

- Haemangioma
- Lymphangioma
- Glomus tumour

Intermediate

- Haemangio endothelioma
- Haemangio pericytoma

Malignant

- Angiosarcoma
- Malignant haemangio pericytoma

Secondary

Metastasis to bone from

- Thyroid
- Breast
- Bronchus
- Kidney
- Prostate

APPROACH TO DIAGNOSIS OF MALIGNANT BONE TUMOR

Multi phased work up

Steps

- History
- Local examination
- Laboratory test
- Radiological test
- Histopathological examination

History

- Age- some tumors are very age specific. Eg- Ewing sarcoma 10-20yrs; osteosarcoma 15-25yrs and > 45yrs (bimodal); chondrosarcoma >45yrs; multiple myeloma >50yrs.
- Non specific
- Dull aching painful lump
- Pathological fracture
- Sometimes as incidental finding
- H/o exposure to radiation and chemical carcinogens
- History of any malignancy anywhere in body or treatment history for any malignancy at present or past

Examination

- Swelling- tenderness, location, shape, consistency, fixity to skin and adjacent structure, mobility, skin over swelling , dilated or engorged veins.
- Joint range of movement limitation
- Sign of inflammation may be present
- Any other skin lesion anywhere else in the body
- Regional lymph node
- Systemic examination to diagnose primary tumor in case of metastasis

Laboratory

- Hemogram
- Erythrocyte sedimentation rate
- C- reactive protein
- Serum calcium
- Serum phosphorus
- Alkaline phosphatase
- Lactate dehydrogenase
- Parathormone
- Urinary Bence Jones protein
- Urinary 24hrs calcium
- Electrophoresis
- Bone marrow examination

Hemogram and ESR

- Anaemia may be seen
- To rule out infection, myeloma and leukemia.
- ESR is raised particularly in

  Metastasis

  Ewing's sarcoma

  Lymphoma

  Leukemia

S calcium and phosphorus

- Hypercalcemia is most common metabolic complication of metastatic bone disease.
- Increased level than normal indicates

  Metastasis

  Myeloma

  Hyperparathyroidism

S alkaline phosphatase

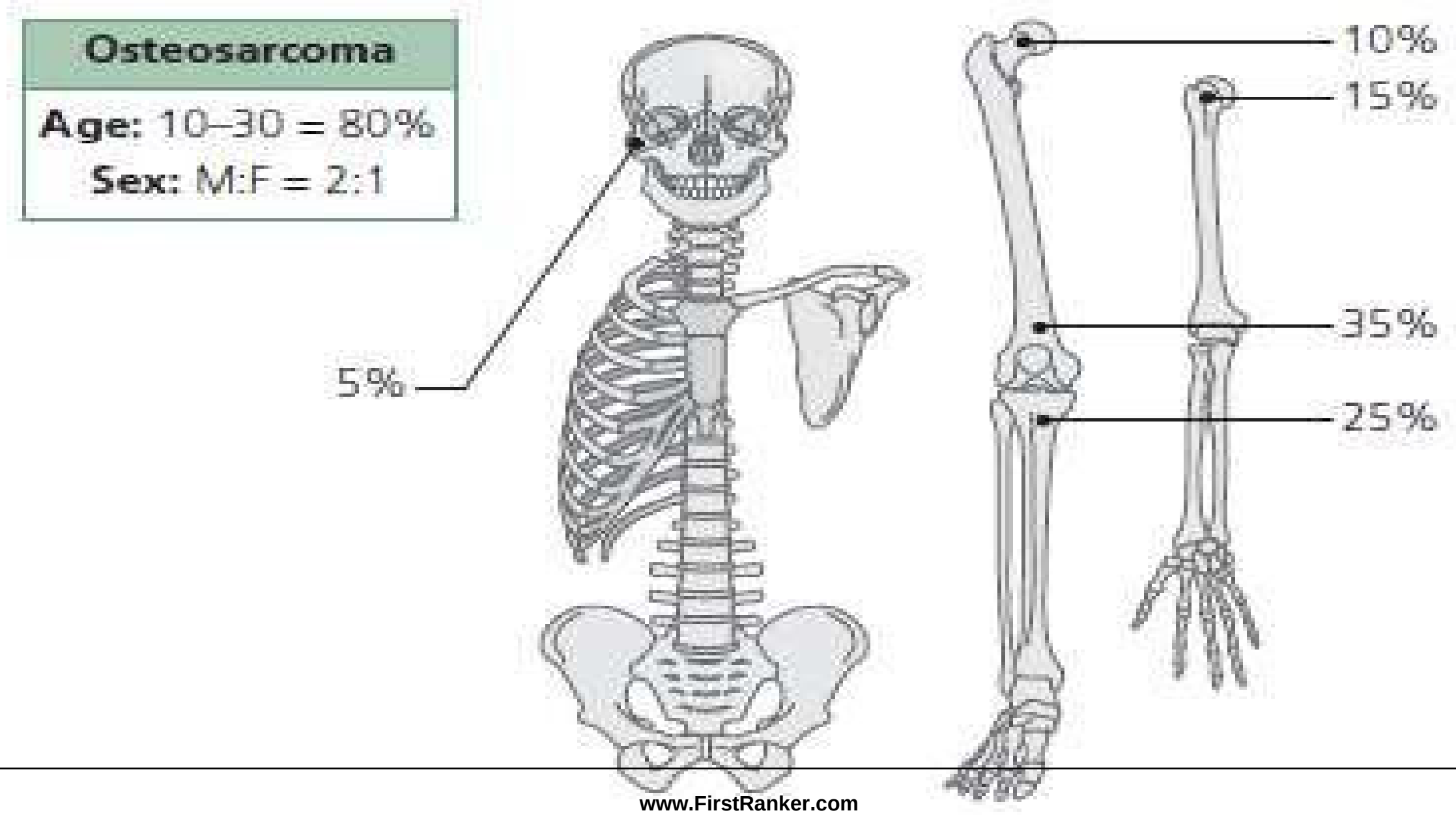
- Raised alkaline phosphatse indicates high turn over of the bone
- Raised in osteoblastic lesion
  - Blastic metastasis from prostate and breast
  - Active Paget's disease
  - Hyperparathyroidism

Radiological tests

- X-ray
- CT scan
- MRI
- Bone scan
- PET scan
- Chest X-ray
- CT thorax, abdomen and pelvis
- Mammography
- Thyroid scan
- Arteriogram

OSTEOSARCOMA

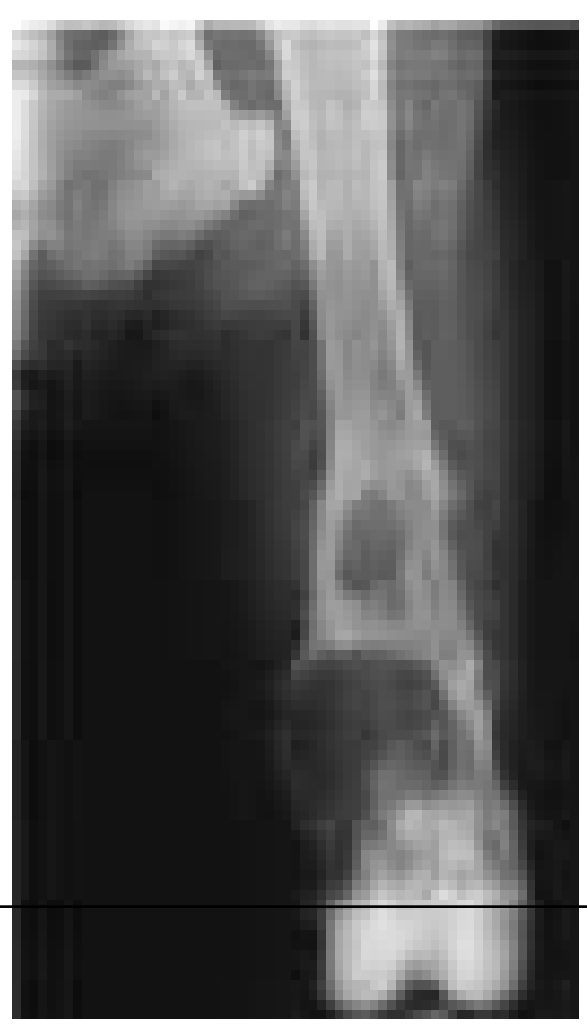
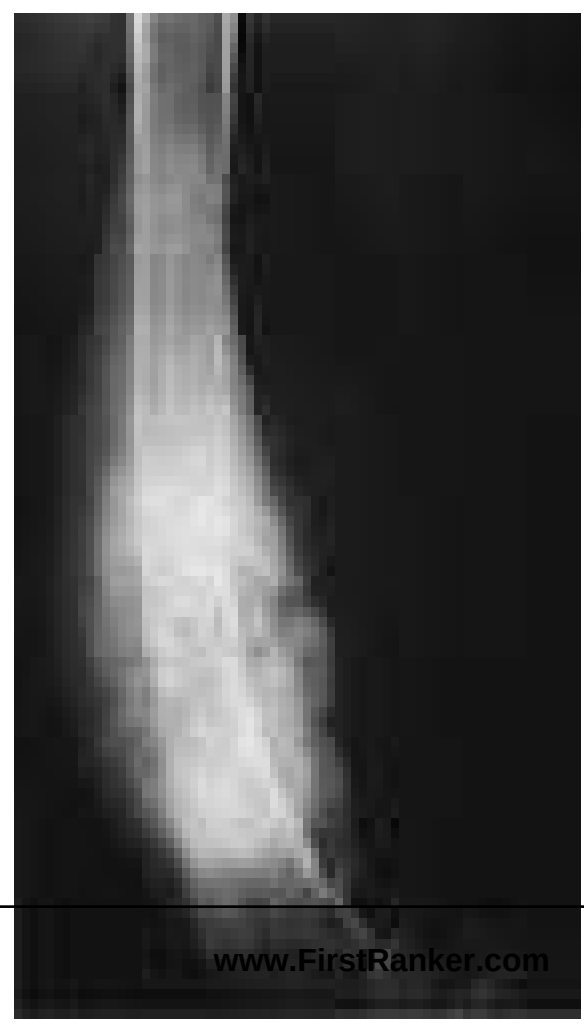
- **Characterized by the production of osteoid by malignant cells.**
- **It is the second most common primary malignant tumor of bone, accounting for approximately 20% of primary bone cancers.**
- **The most common nonhematologic primary malignancy of bone**
- **Onset can occur at any age; however, primary high-grade osteosarcoma occurs most commonly in the second decade of life.**
- **Parosteal osteosarcoma has a peak incidence in the third and fourth decades.**
- **Arise from multipotent mesenchymal cells**



Clinically

- Radiographic appearance of osteosarcoma can vary
- lesion can be either predominantly blastic or predominantly lytic
- The lesion usually is quite permeative, and the borders are ill defined.
- May take the form of a “Codman triangle,” or it may have a “sunburst” or “hair-on-end” appearance.
- Magnetic resonance imaging (MRI)
- They may be primarily osteoblastic, fibroblastic

Plain X-ray

Plain X-ray

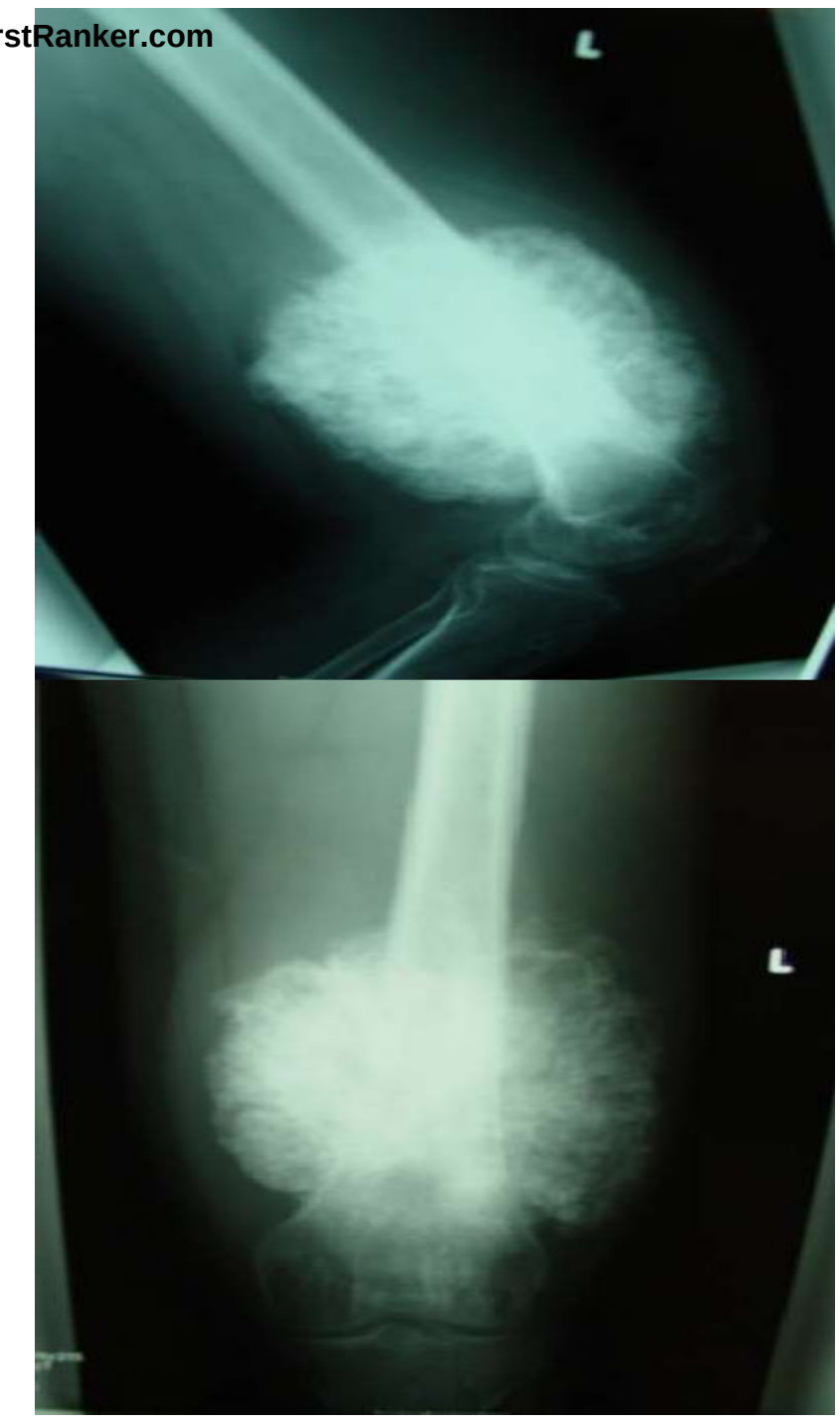
Lesions are usually permeative

Associated with destruction of the cancellous an cortical elements of the bone

Ossification within the soft tissue component, if tumour has broken through cortex

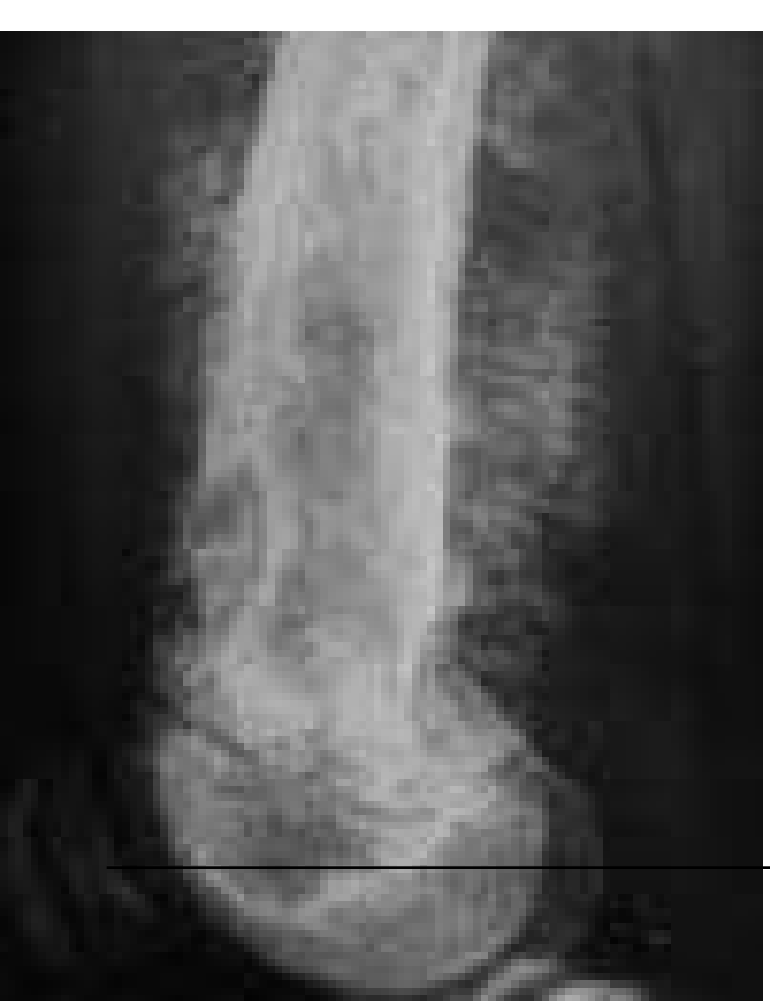
Intra medullary

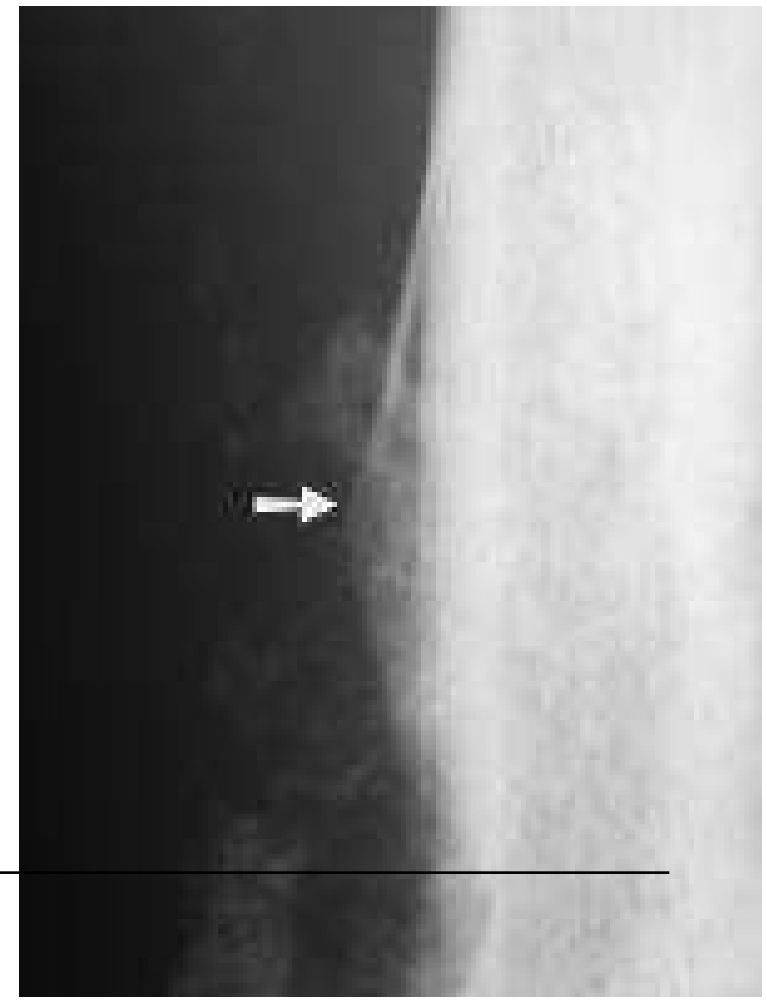
Borders are ill defined

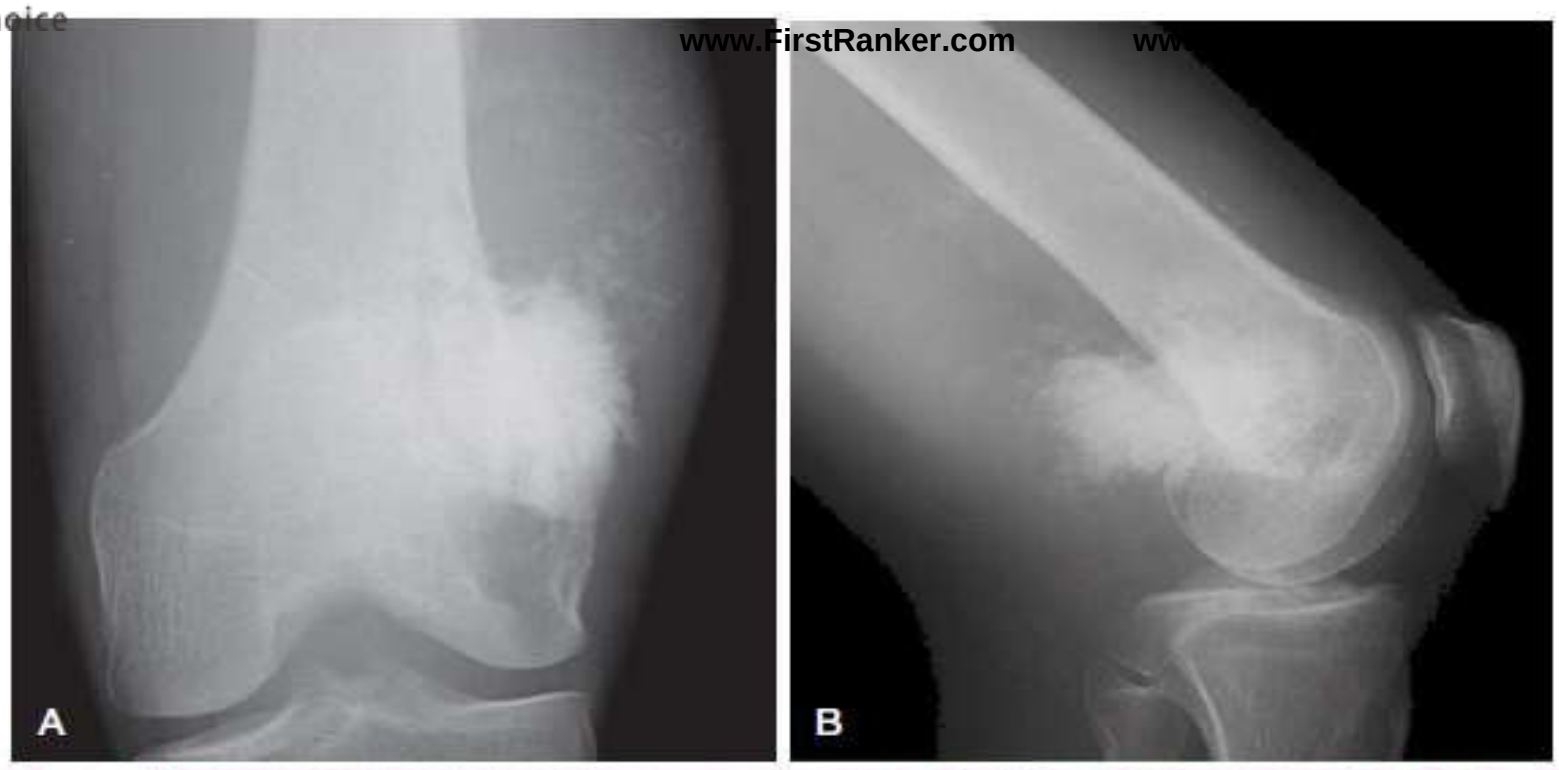


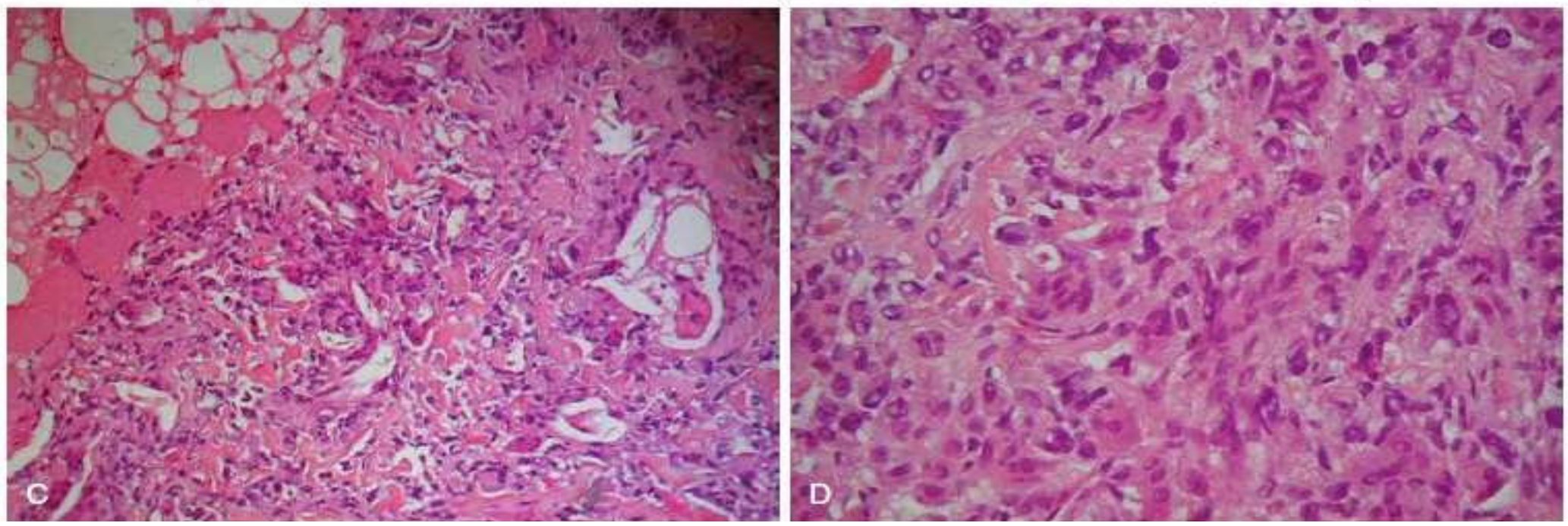
Plain X-ray

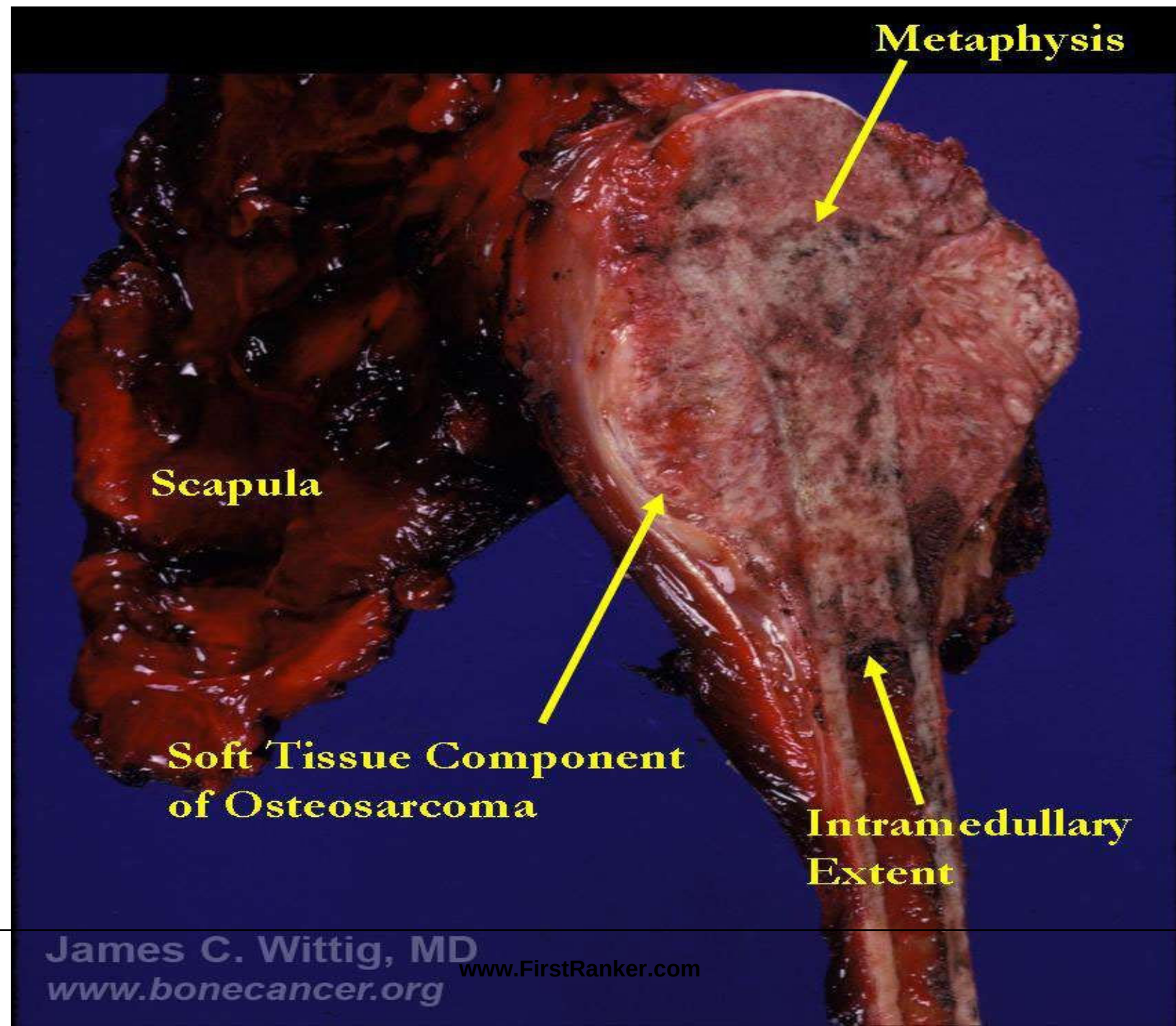
Periosteal reaction may appear as the characteristic Codman triangle.











Classification

OSTEOSARCOMAS are

- ✓ Conventional /classic osteosarcoma (high grade, intra medullary)
- ✓ Low-grade intramedullary osteosarcoma
- ✓ Parosteal osteosarcoma
- ✓ Periosteal osteosarcoma
- ✓ High-grade surface osteosarcoma
- ✓ Telangiectatic osteosarcoma, and
- ✓ Small cell osteosarcoma.

Classification:

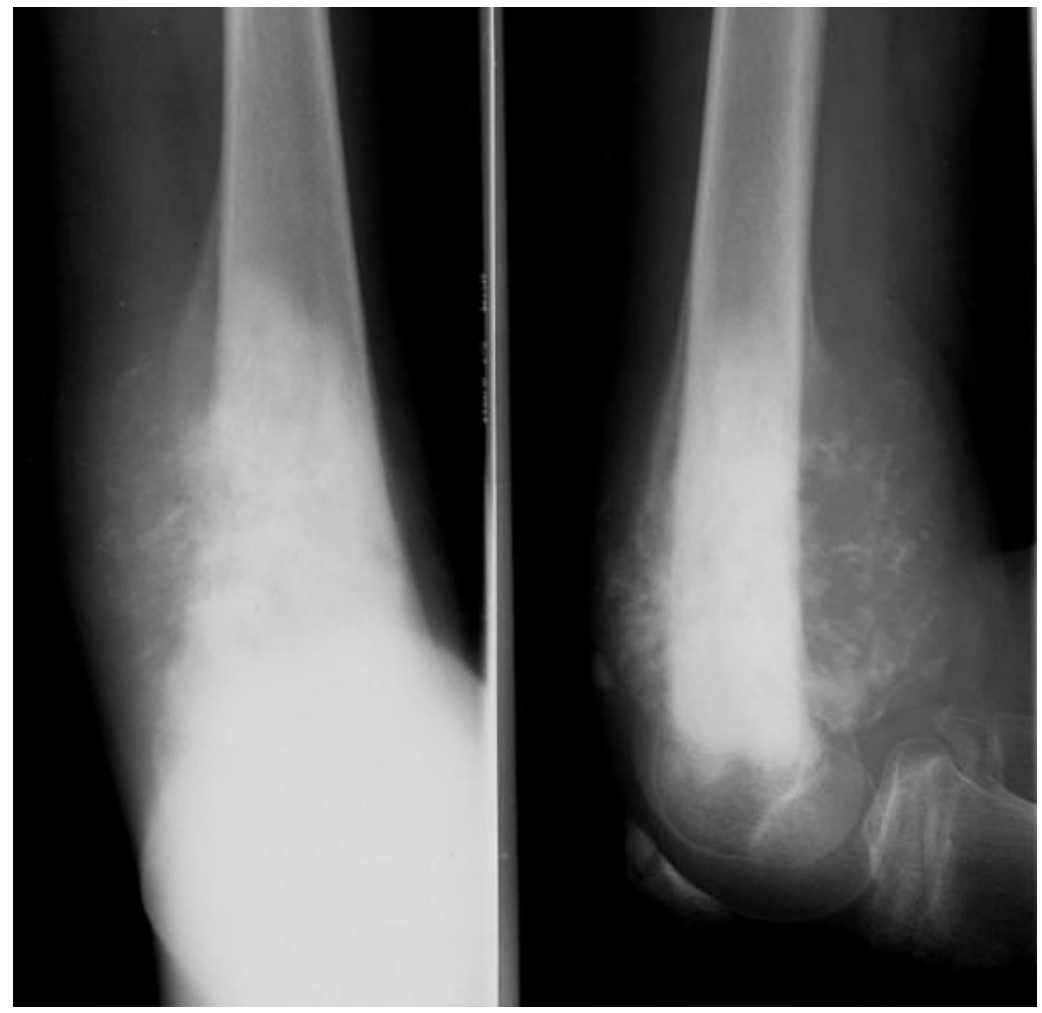
SECONDARY OSTEOSARCOMAS

Osteosarcomas occurring at the site of another disease process.
more common in >50 years of age
The most common causes are
Paget disease
Previous radiation treatment

Other associated conditions are
Fibrous dysplasia Bone infarcts
Osteochondromas Chronic
osteomyelitis
Dedifferentiated chondrosarcomas
Osteogenesis imperfecta

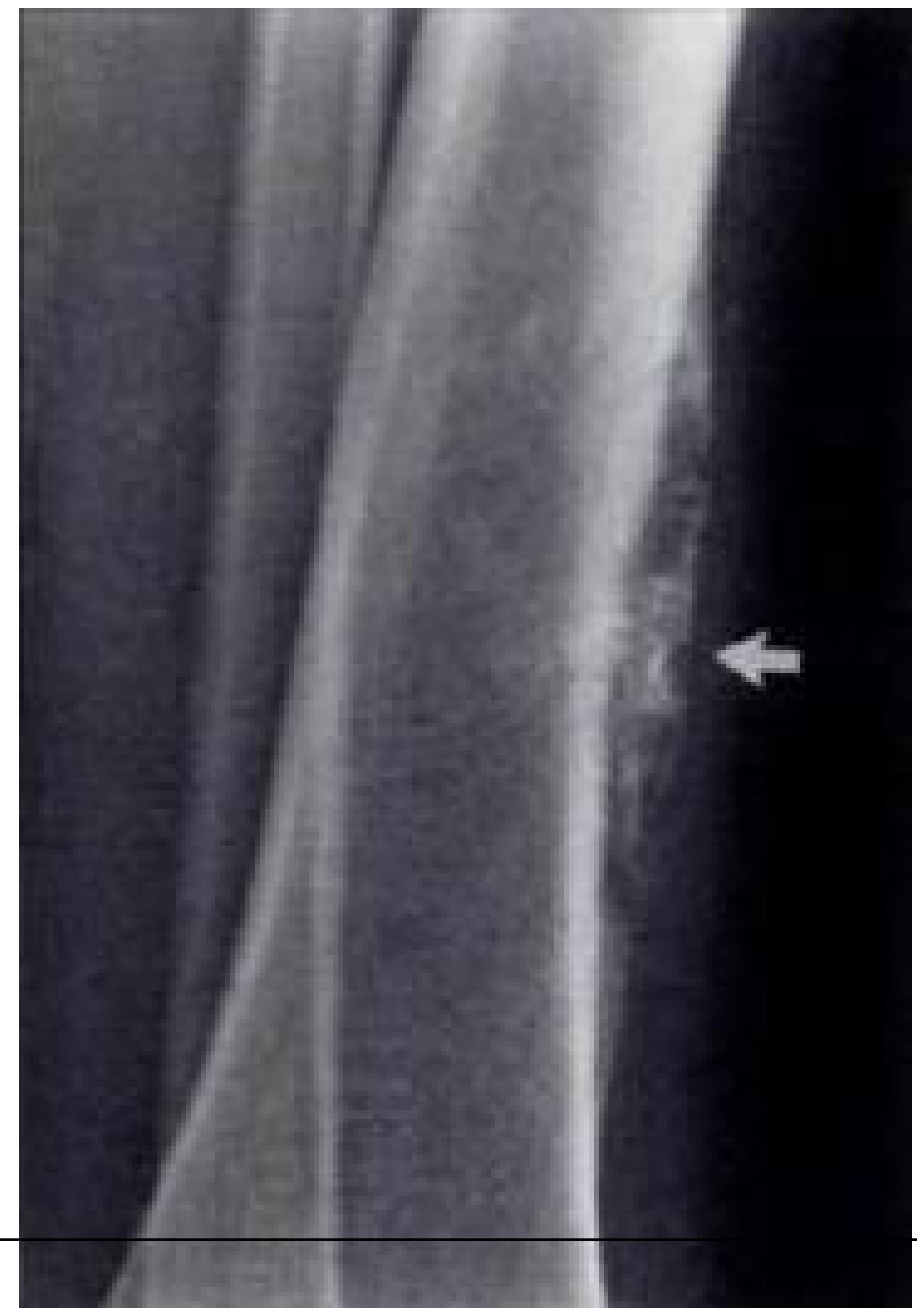
Conventional Osteosarcoma

- High grade
- Common type of Osteosarcoma
- Common radiographic appearance is aggressive lesion producing osteoid matrix
- Periosteal reaction may take the form of “codmans triangle” or “sunbrust” or “hair on end” appearence



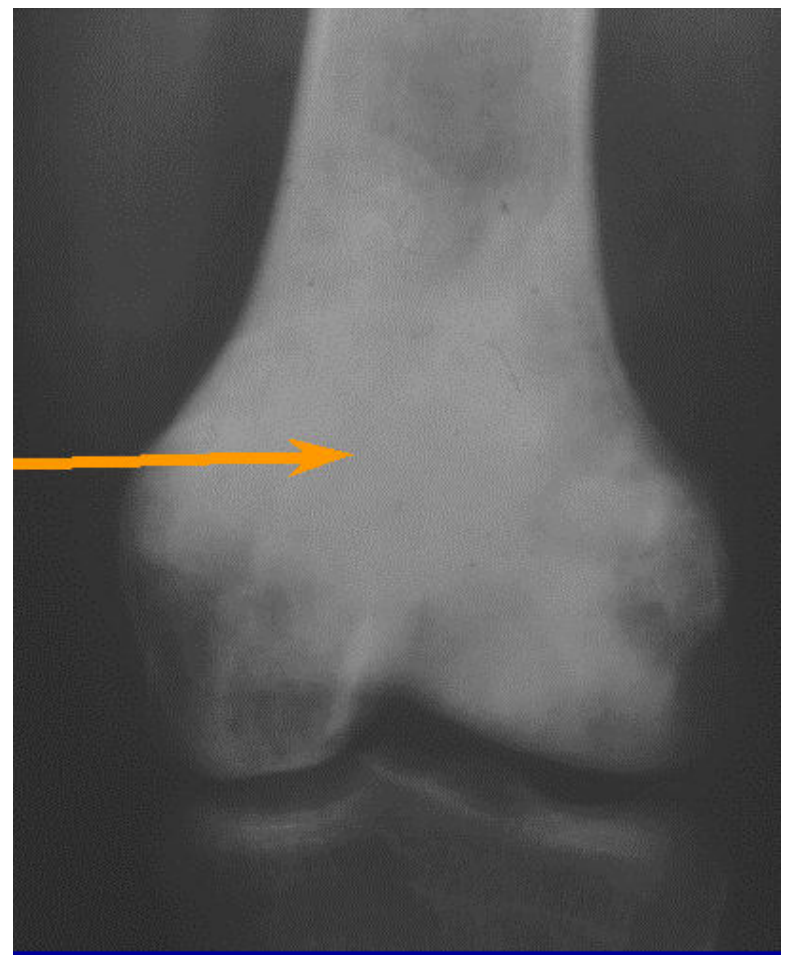
Periosteal Osteosarcoma

- Intermediate grade
- Arises from surface of bone
- Commonly on femur and tibia.



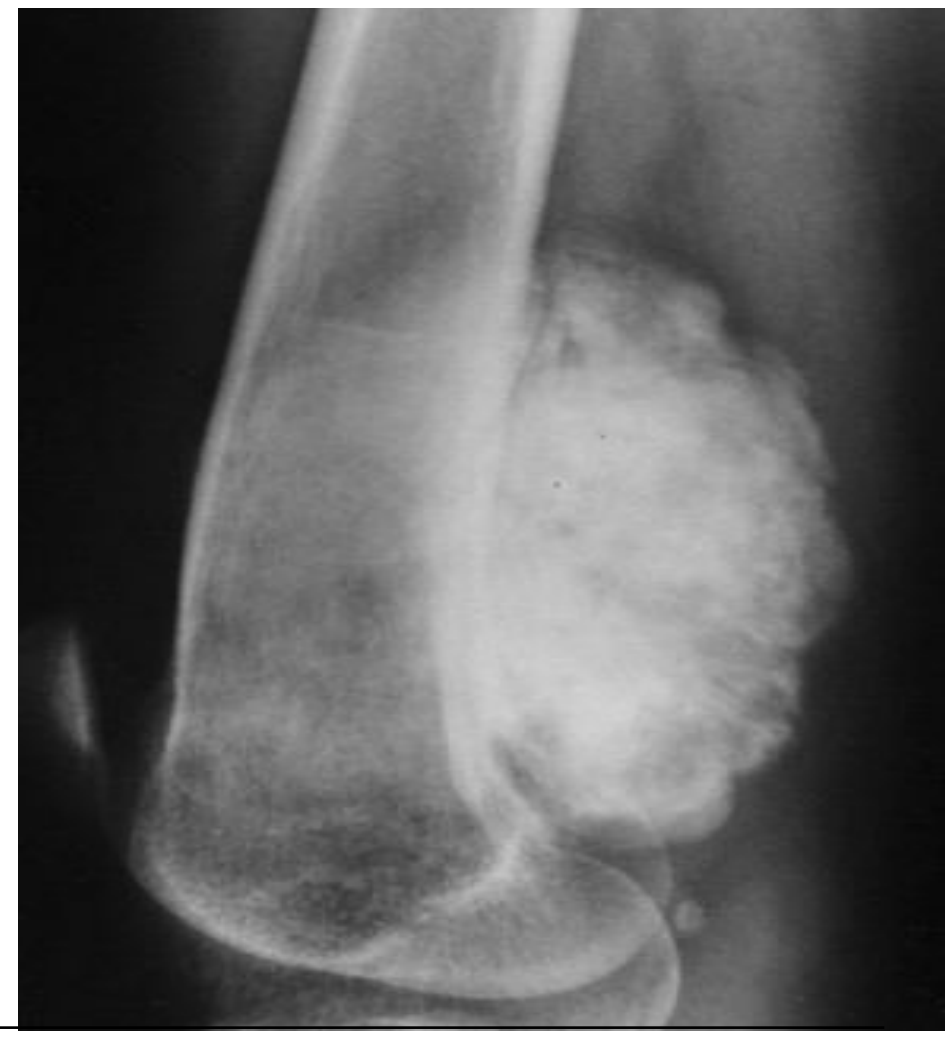
Intramedullary Osteosarcoma

- Rare
- Low grade



Parosteal Osteosarcoma

- Low grade malignancy
- Rare
- Arises on surface of bone and invades medullary cavity only at later stages.
- It has peculiar tendency to occur as a lobulated mass on the posterior aspect of femur

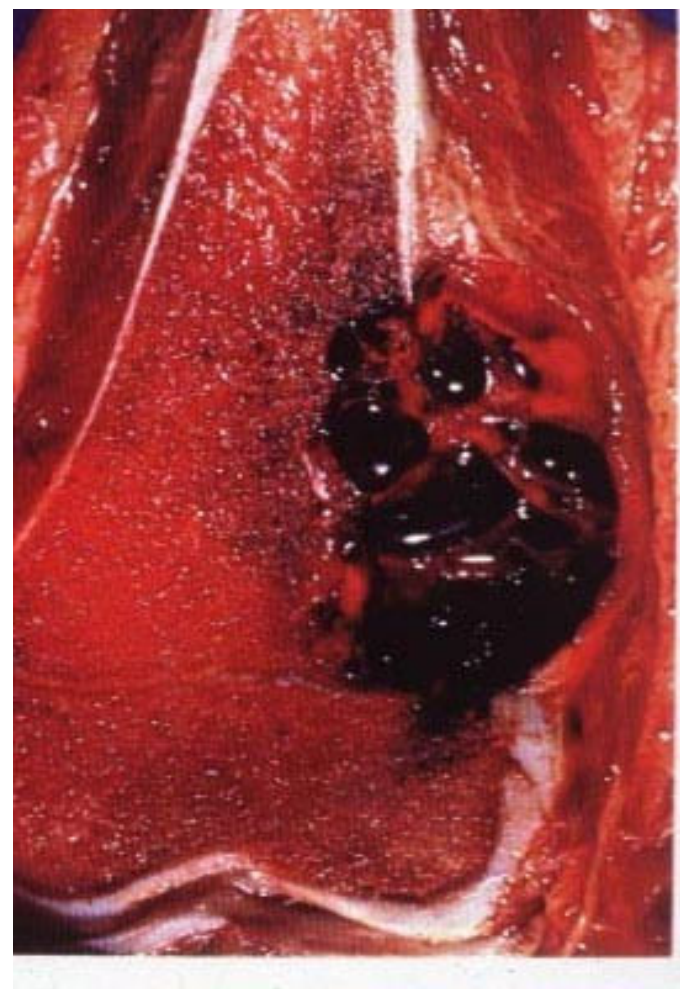


High grade surface Osteosarcoma

- High grade
- Least Common
- Radiographs show invasive lesions with ill defined borders

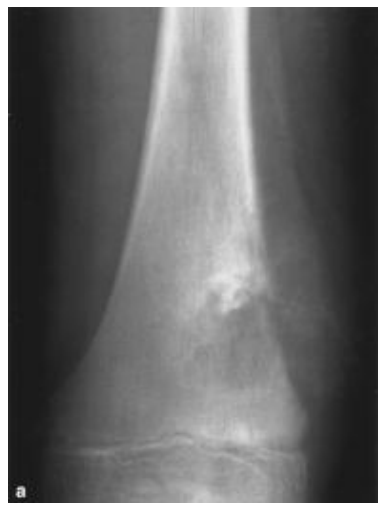
Telangectic Osteosarcoma

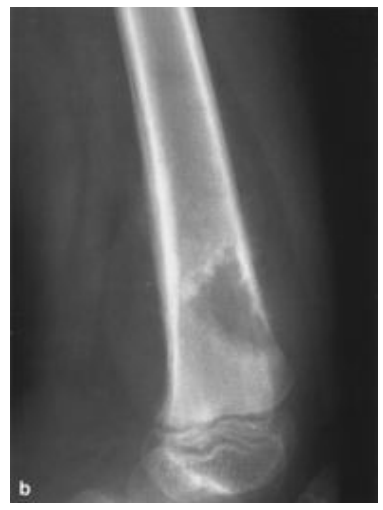
Lytic lesion

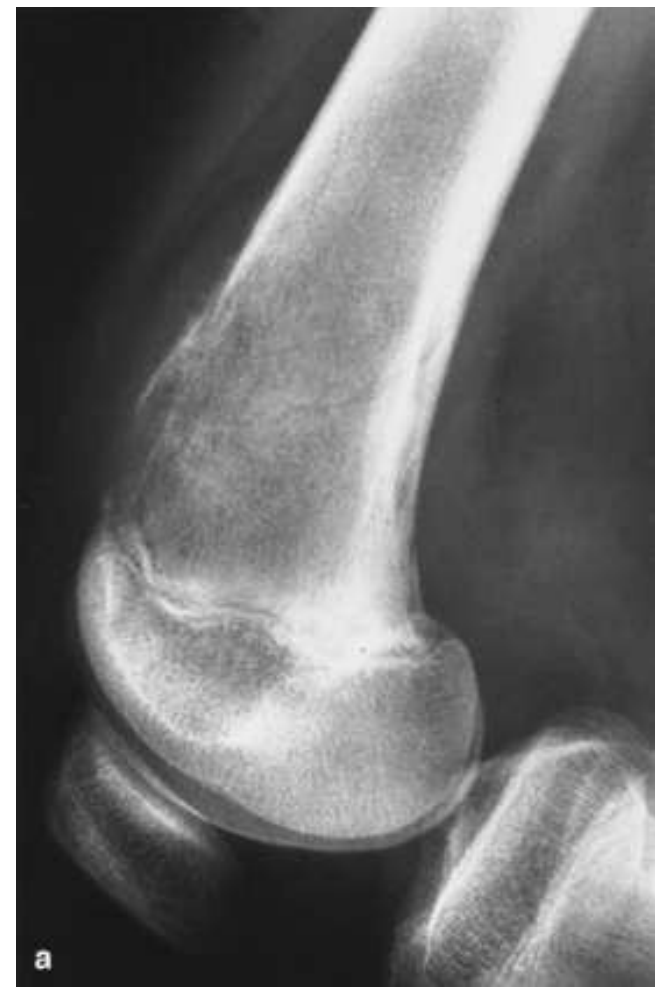


Small cell Osteosarcoma

- Rare
- High grade
- Resemble Ewing sarcoma or Lymphoma







Treatment

Radiological staging

↓

Biopsy to confirm diagnosis Preoperative

↓

chemotherapy

↓

Repeat radiological staging (access chemo response, finalize surgical treatment plan)

↓

Surgical resection with wide margin

↓

Reconstruction using one of many techniques

↓

Post op chemo based on preop response

Chondrosarcoma

- 9% of primary malignancies of bone
- Age: broad, primary chondrosarcoma peak around 40 – 60yrs, secondary chondrosarcoma 25 – 45 yrs.
- Any location but common around pelvis, proximal femur, proximal humerus.
- Most common malignancy in hand.
- Clinically: increasing pain and palpable mass.
- Pain in absence of pathological fracture is helpful to differentiate between enchondroma and low grade chondrosarcoma

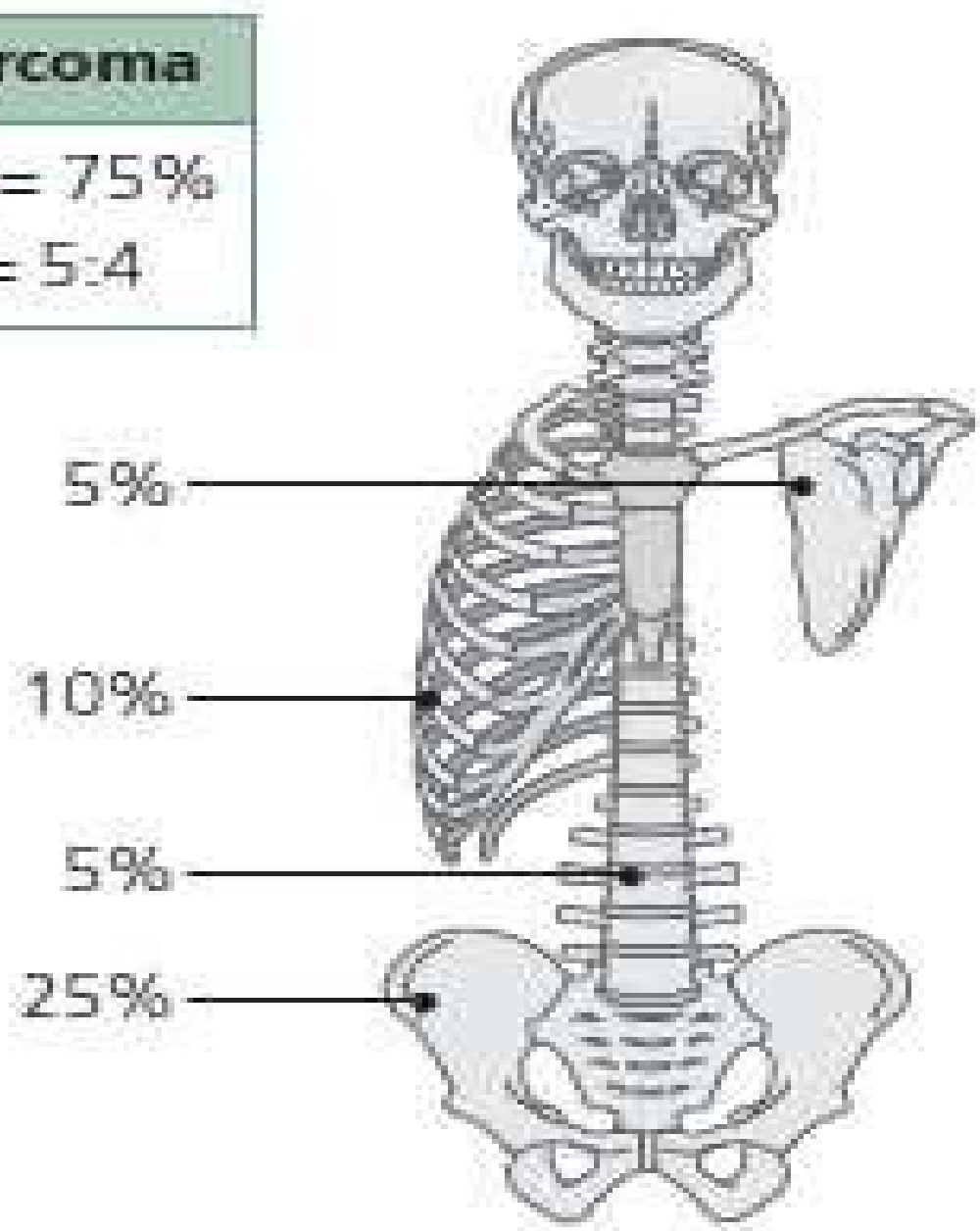
Secondary Chondrosarcoma

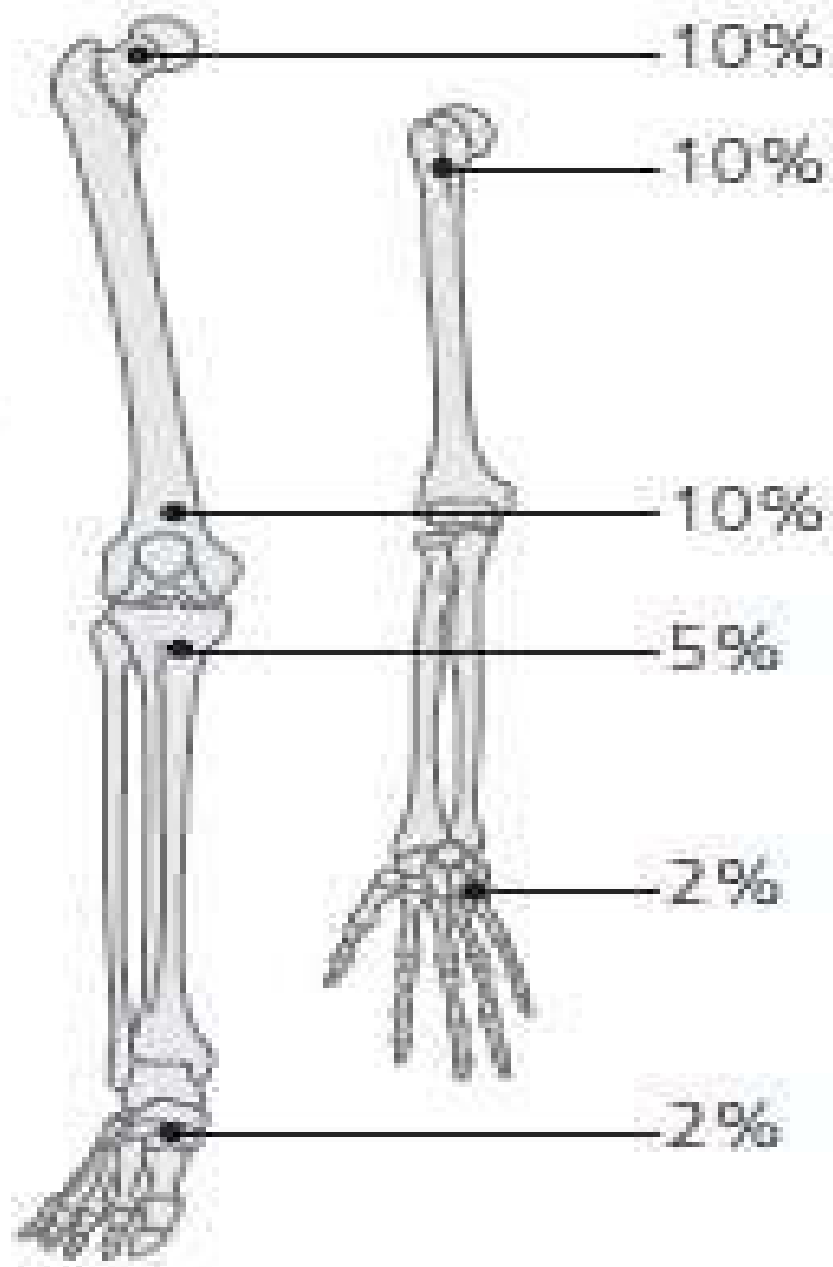
- Olliers disease (multiple enchondromatosis)
- Maffuci syndrome (multiple enchondromatosis + soft tissue haemangioma)
- Multiple hereditary exostoses
- Solitary osteochondroma
- Synovial chondromatosis
- Chondromyxoid fibroma
- Periosteal Chondroma
- Chondroblastoma
- Previous radiation treatment Firbours dysplasia

Chondrosarcoma

Age: 30–70 = 75%

Sex: M:F = 5:4





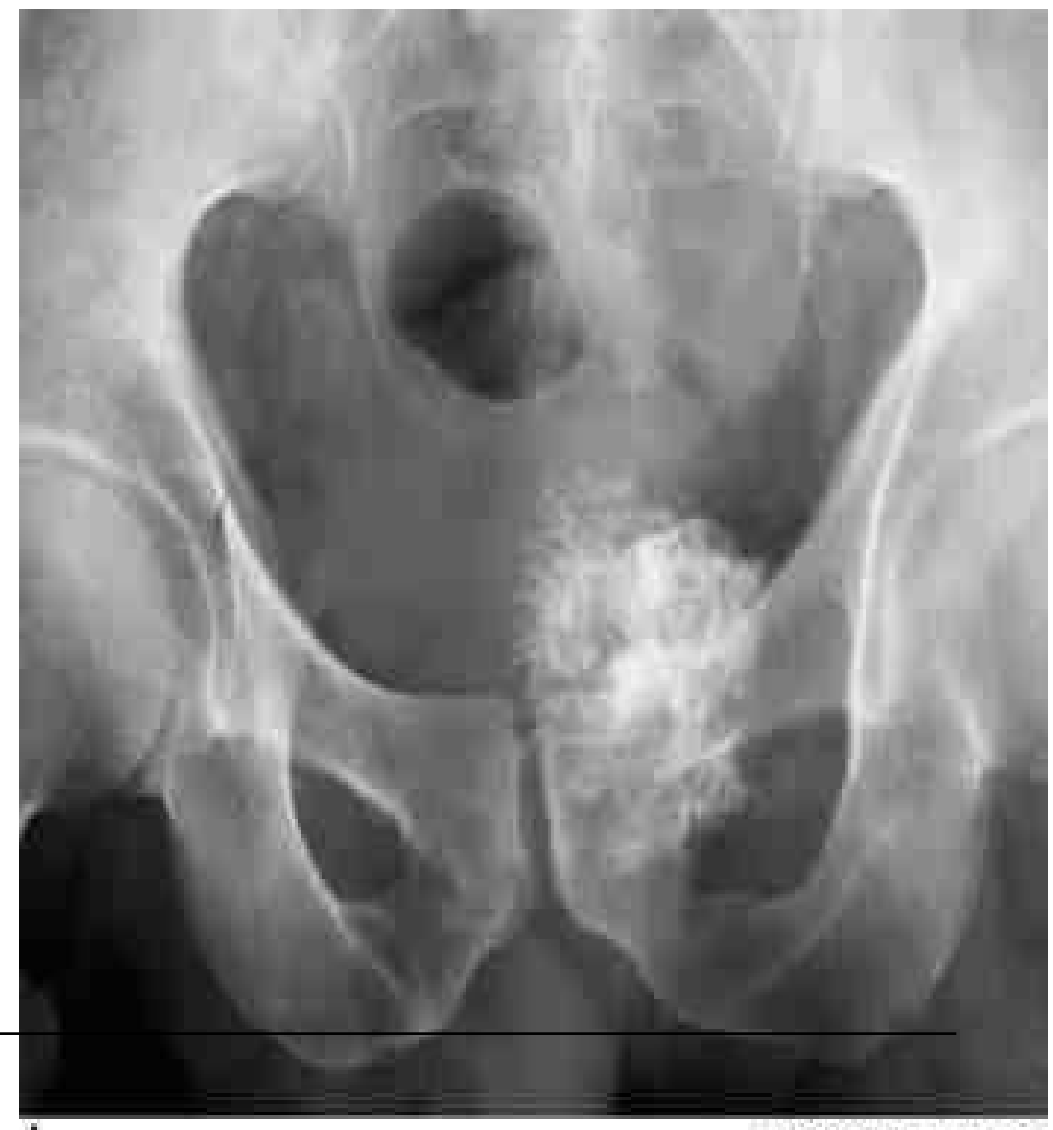
Chondrosarcoma - xrays

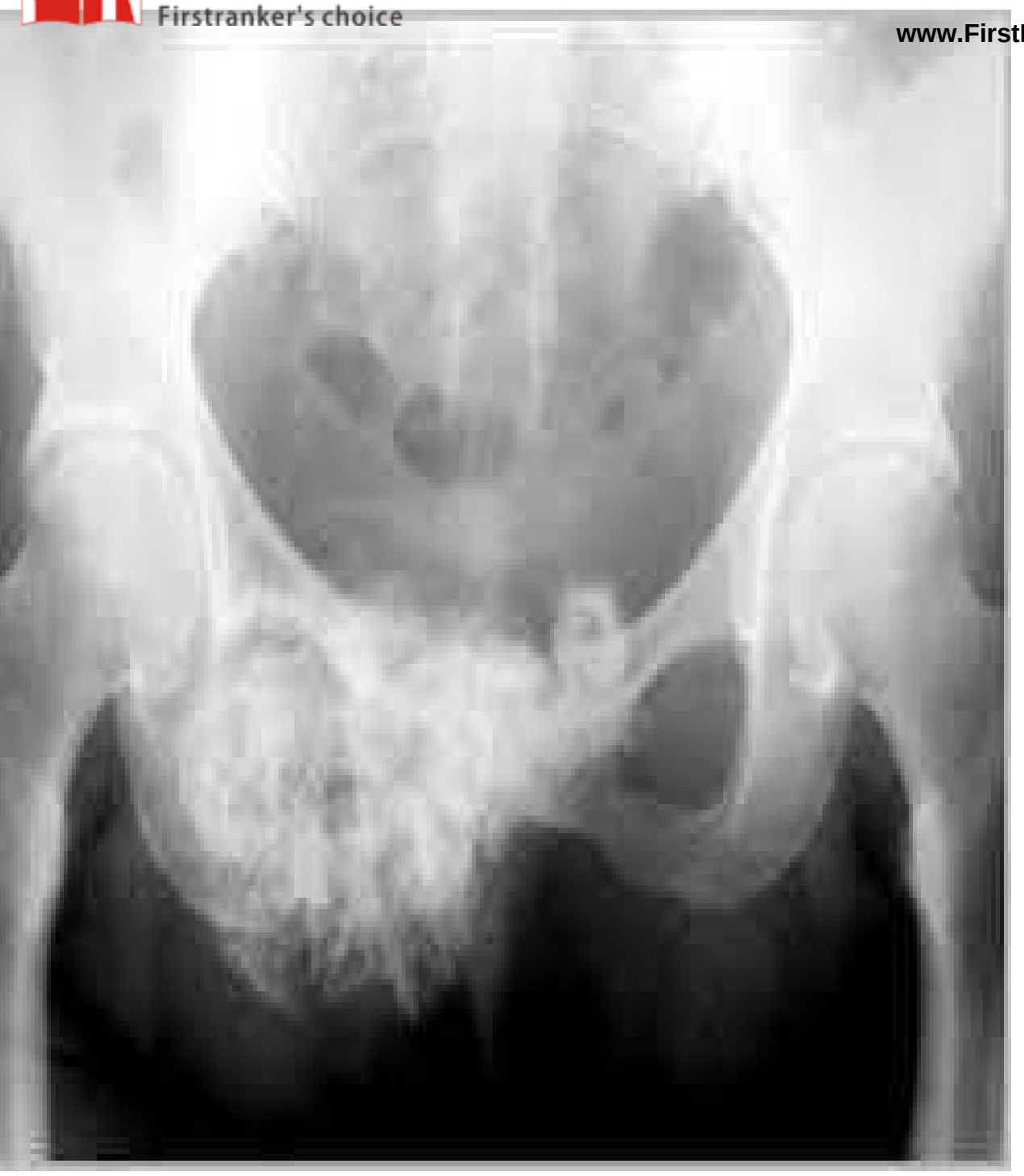
- Appearence similar to enchondroma,

it is a lesion arising in medullary cavity with irregular matrix calcification.

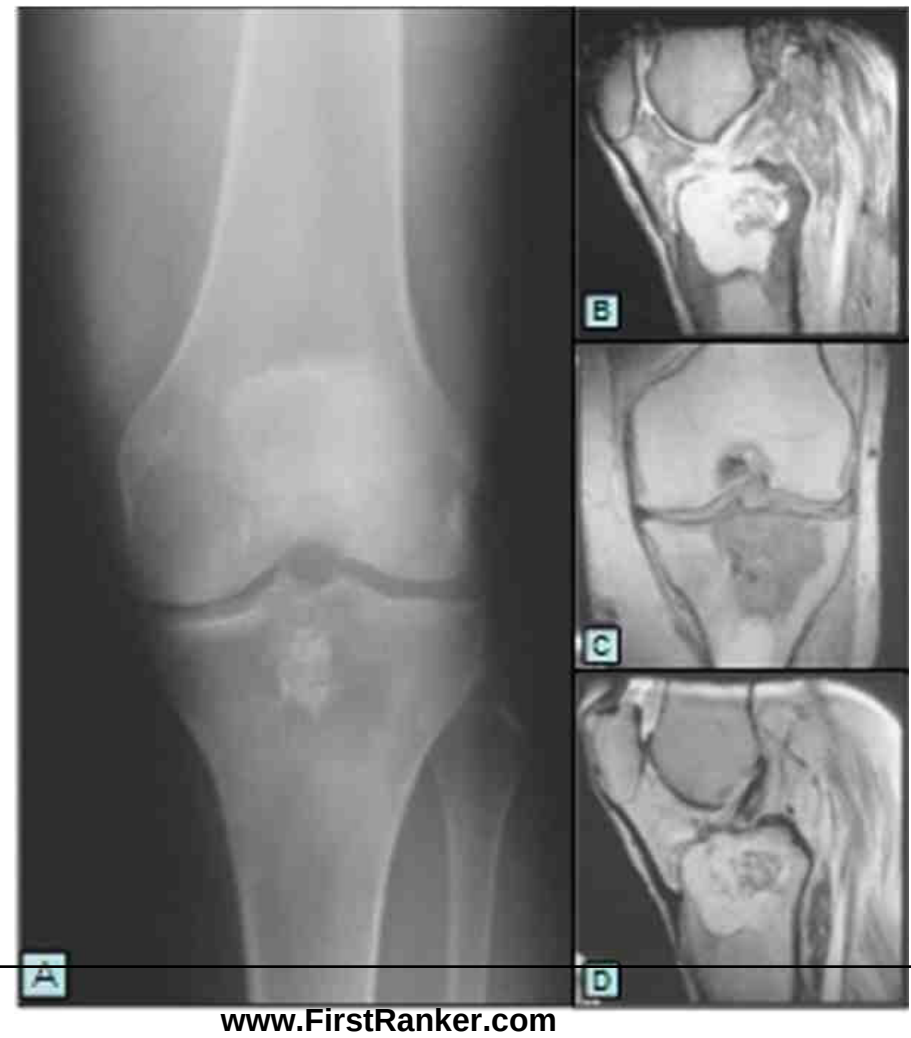
- Pattern is described as “punctate,” “popcorn,” or “comma-shaped”.
- Compared with enchondroma,

chondrosarcoma has more aggressive appearence with bone destruction and cortical erosions, periosteal reaction, and rarely soft-tissue mass.

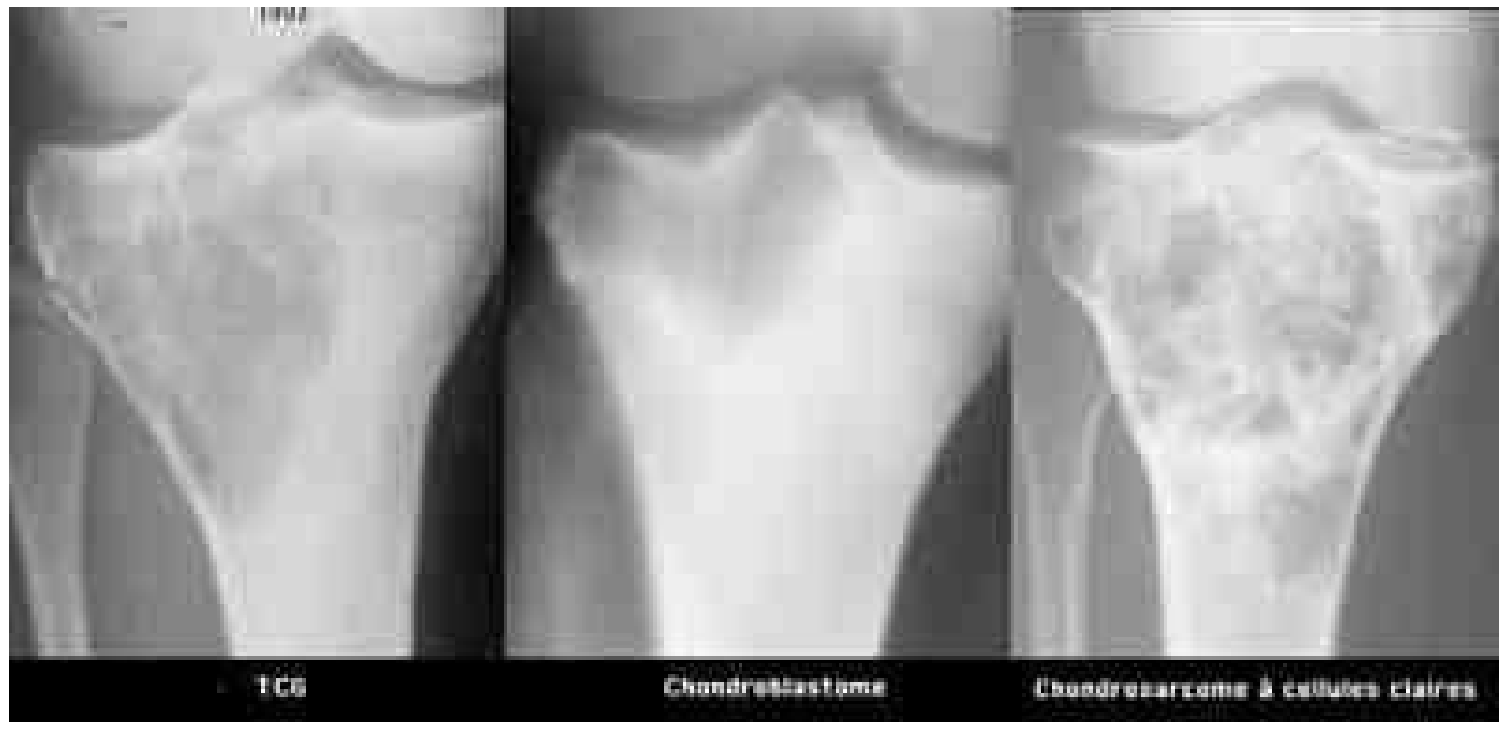




Mesenchymal chondrosarcoma

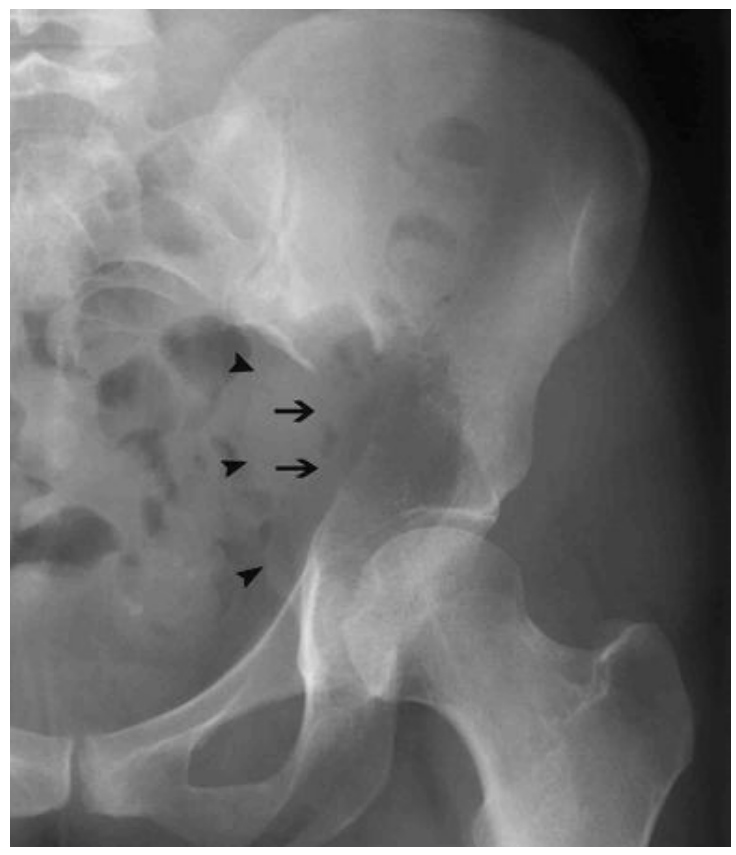


Clear cell chondrosarcoma



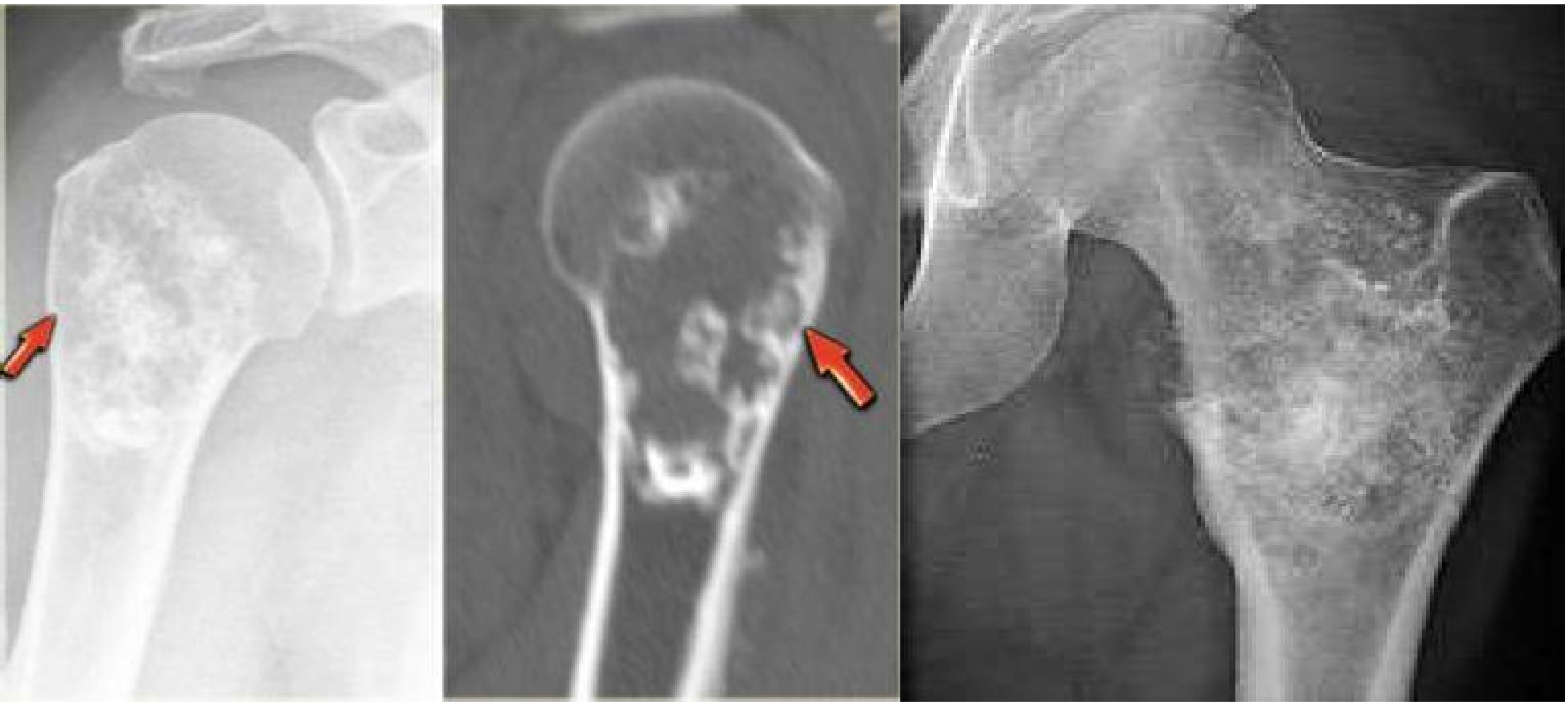
Epiphyseal Gaint Cell Tumor Chondroblastoma Clear cell chondrosarcoma

Dedifferentiated chondrosarcoma



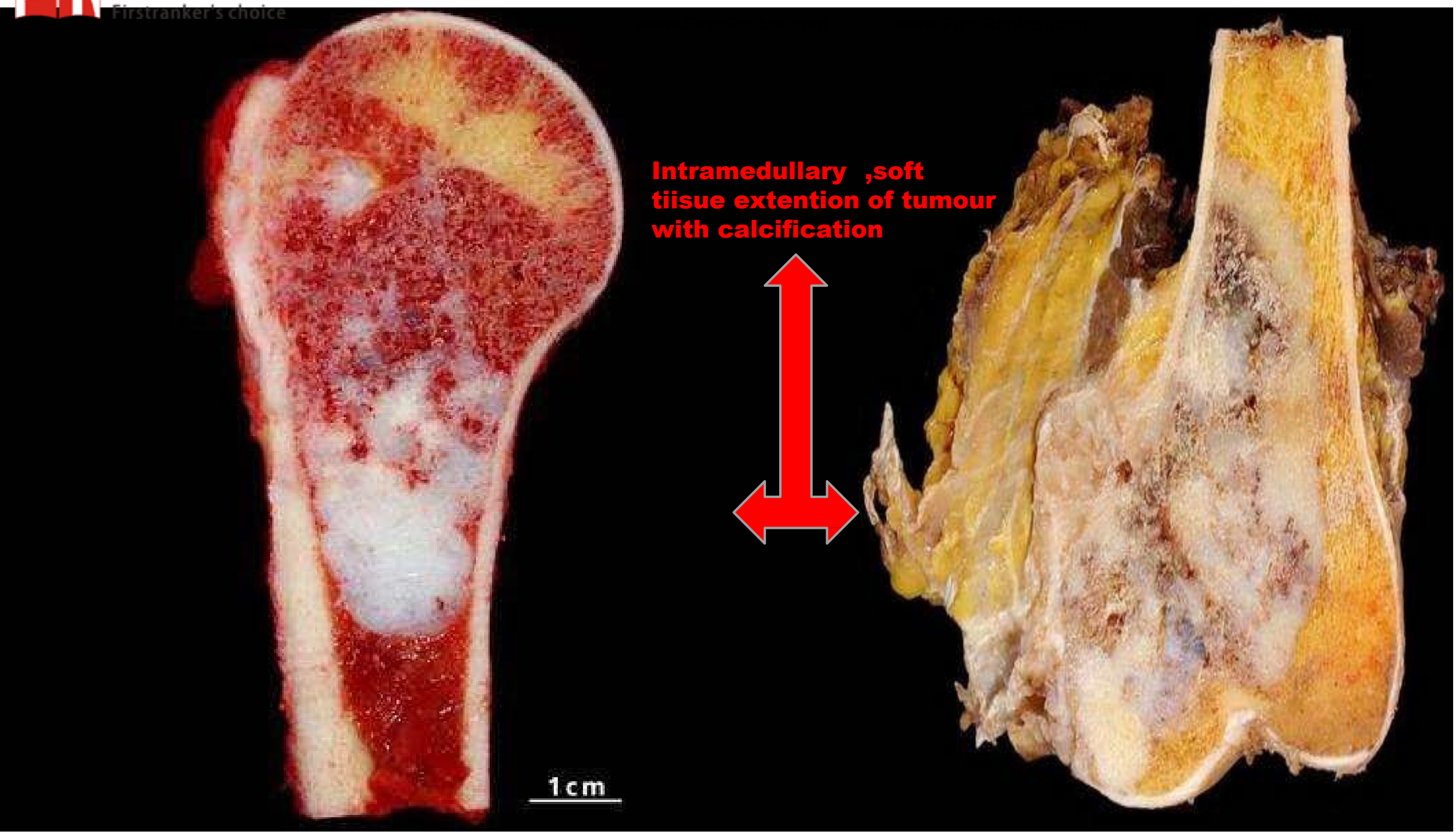
Radiographic features of dedifferentiated chondrosarcoma often show a more aggressive radiolucent area juxtaposed on a otherwise typical chondrosarcoma.

POP CORN CALCIFICATION



DIFFERENCE FROM ENCHONDROMA

- **Endosteal scalloping of more than 2/3rd of cortical thickness- chondrosarcoma**
- **Aggressive changes such as cortical erosion , bone destruction, periosteal reaction, soft tissue mass- chondrosarcoma**
- **size> 5 cm in axial skeleton- predictor of malignancy**
- **Apperance of lysis in previously calcified area- malignancy**



Chondrosarcomas Treatment

- Low grade – Extended curettage with use of intraoperative adjuvant treatment.
- High grade – Wide or radical resection or amputation.
- Radiotherapy as palliative for inaccessible lesions.

Ewing's Sarcoma

- Ewing sarcoma, a highly malignant neoplasm
- Third most common nonhematologic primary malignancy of bone
- The second most common in patients younger than 30 years of age and the most common in patients younger than 10 years of age

Ewing's sarcoma

- Approximately 90% of Ewing sarcomas occur before age 25
- Histogenesis: neurally derived small round cell malignancy very similar to the so-called primitive neuroectodermal tumor (PNET)
- Disease is extremely rare in black persons

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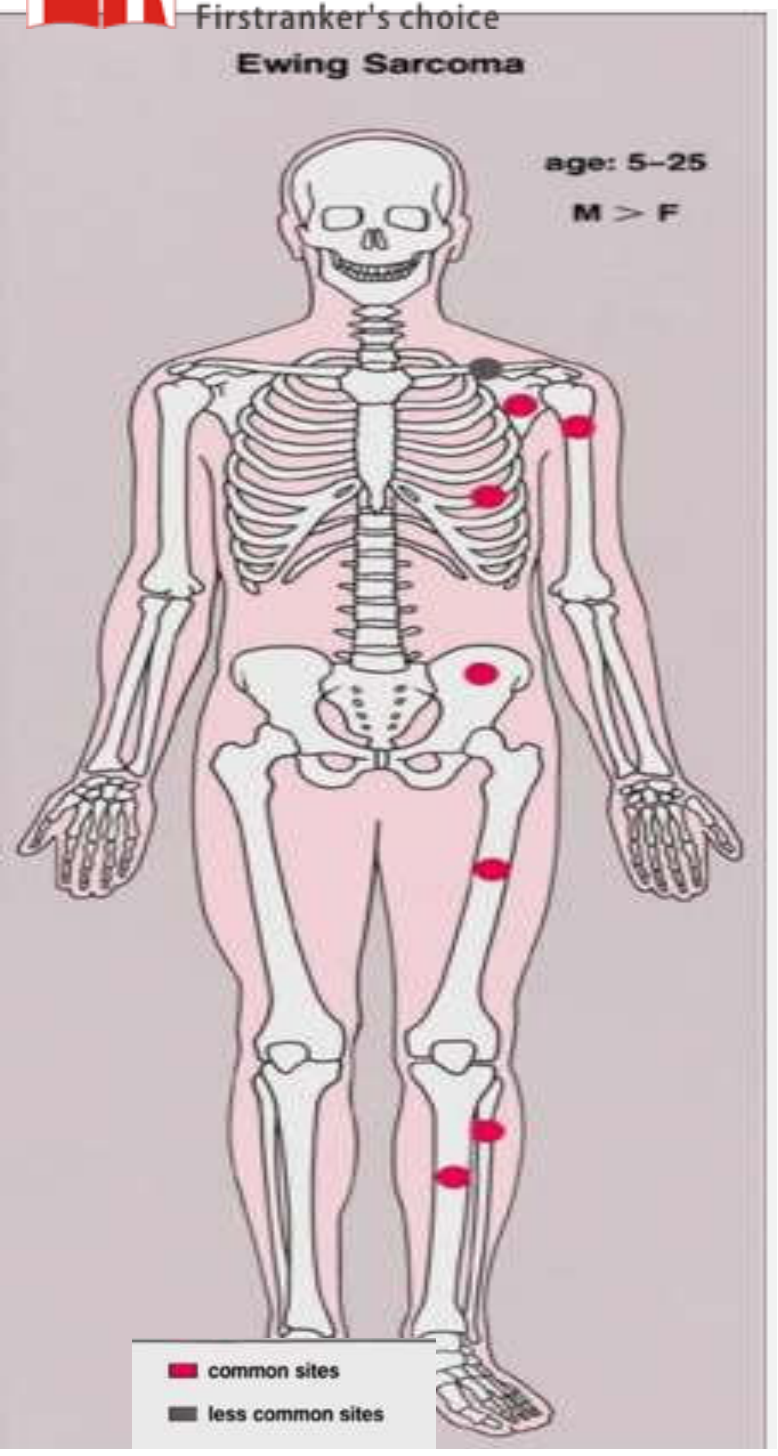
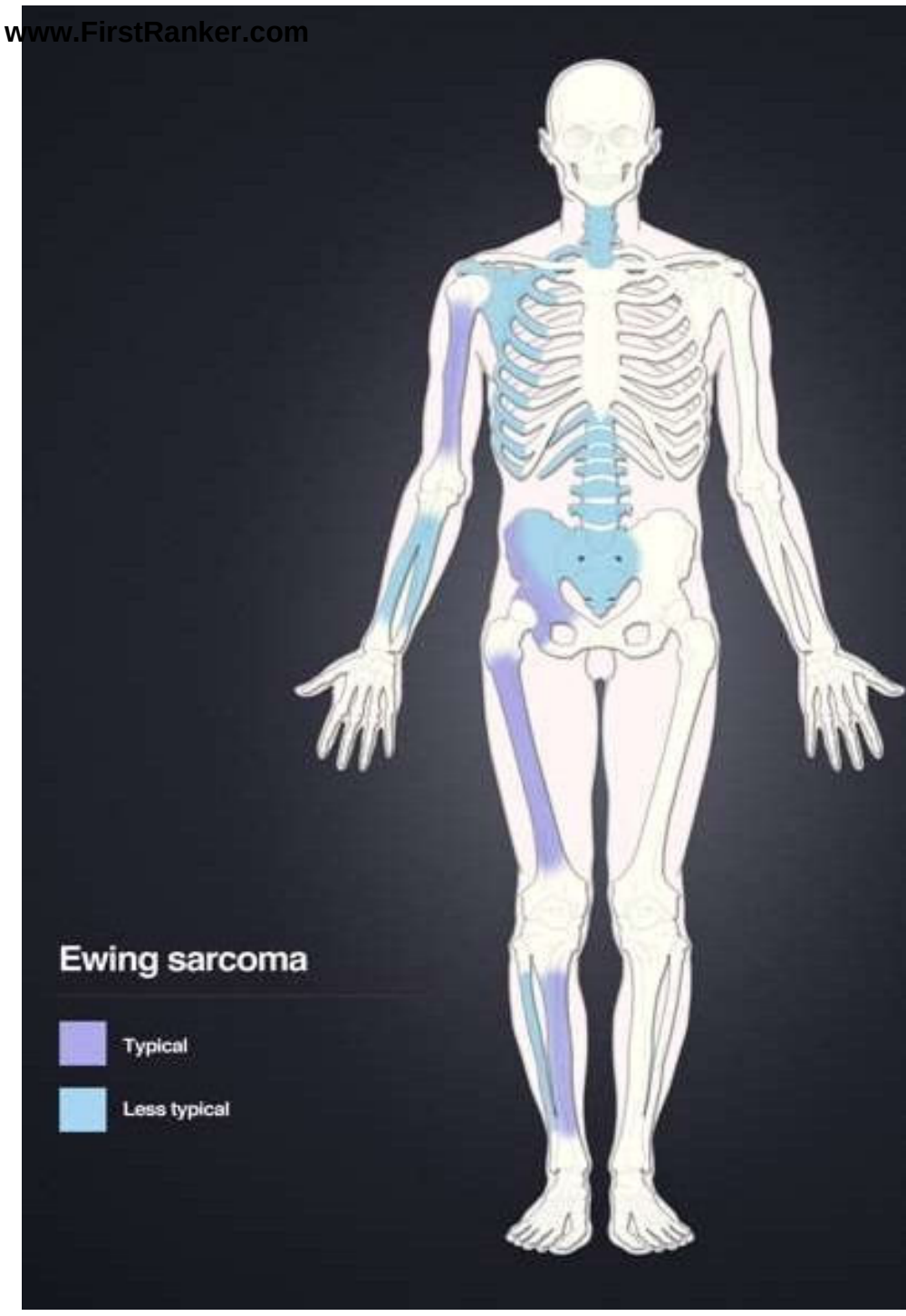


Areas of involvement

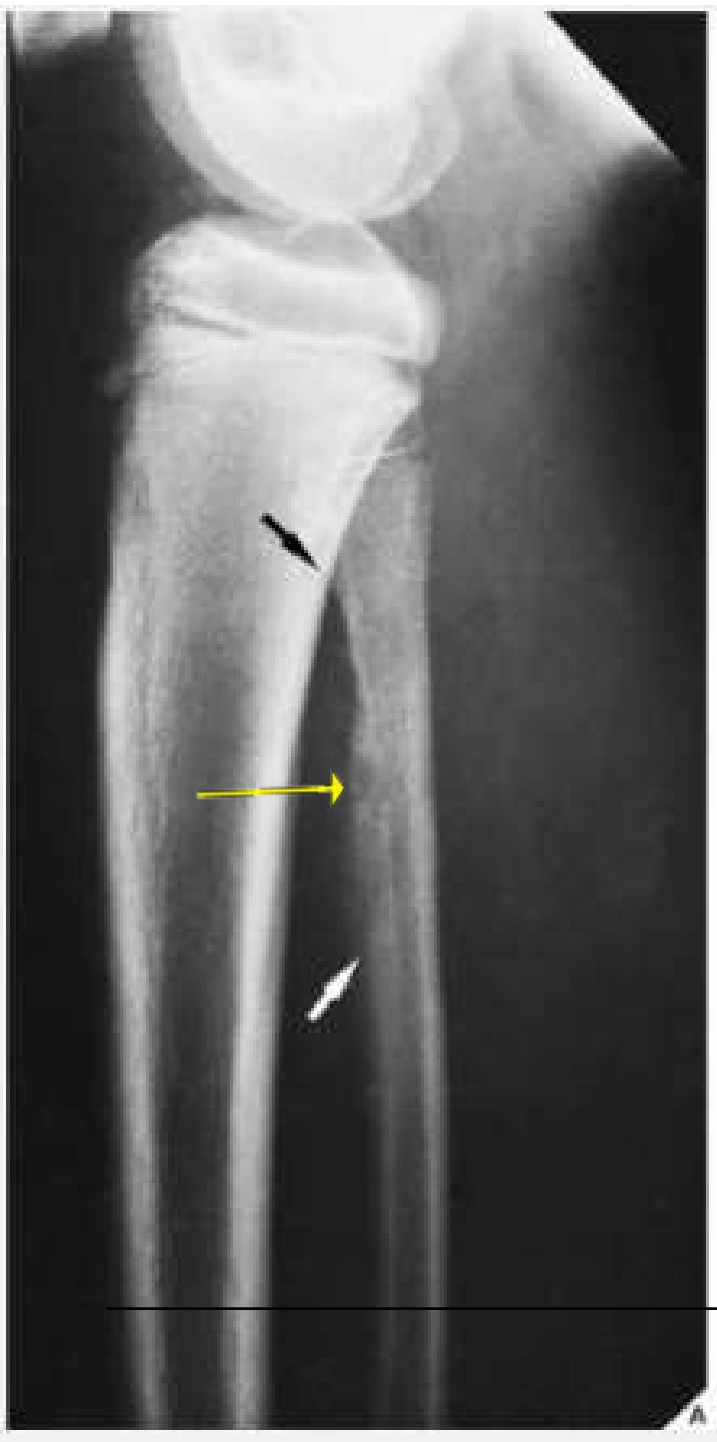


Clinical presentation

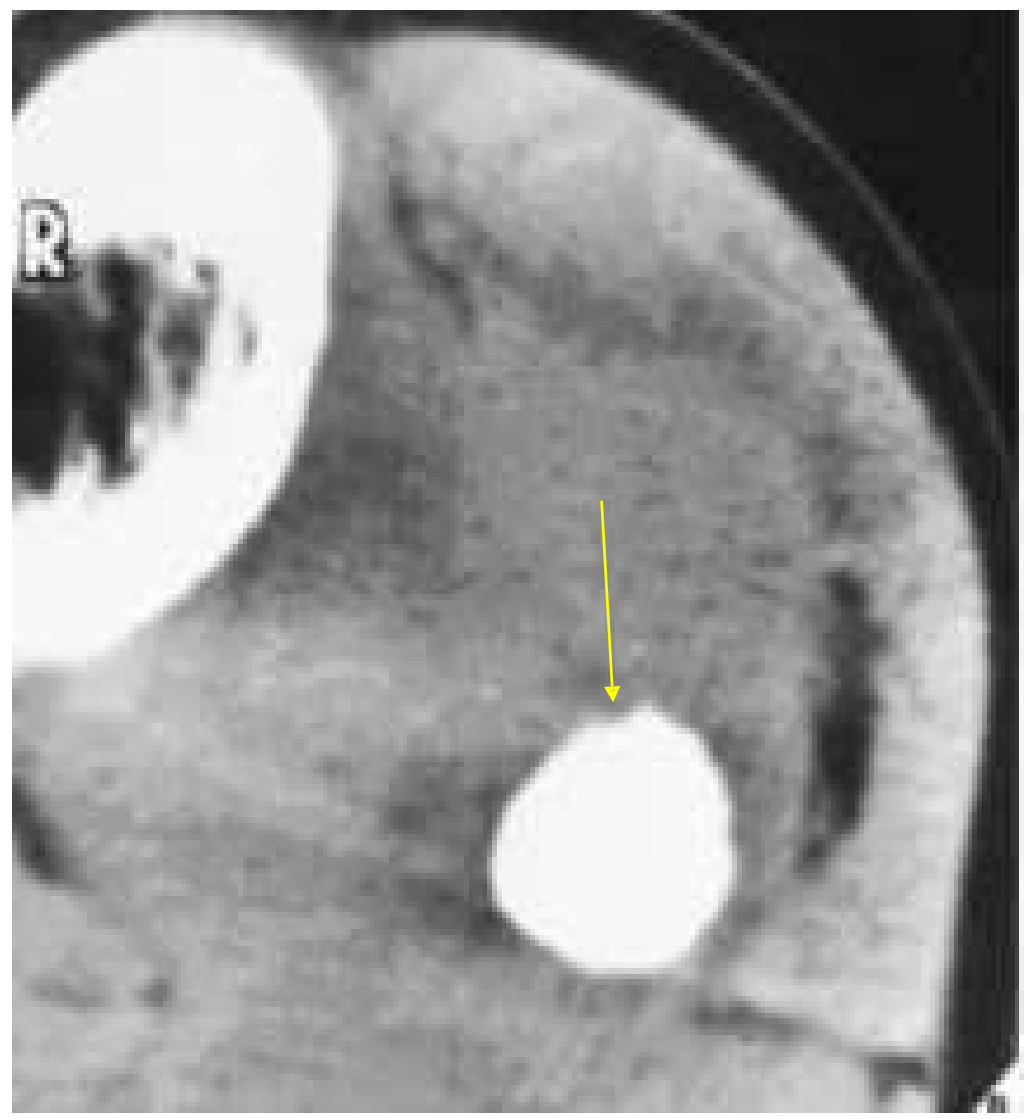
- Present as a localized painful mass
- With systemic symptoms such as fever, malaise, weight loss, and
- An **increased erythrocyte sedimentation rate**.
- These systemic symptoms may lead to an erroneous

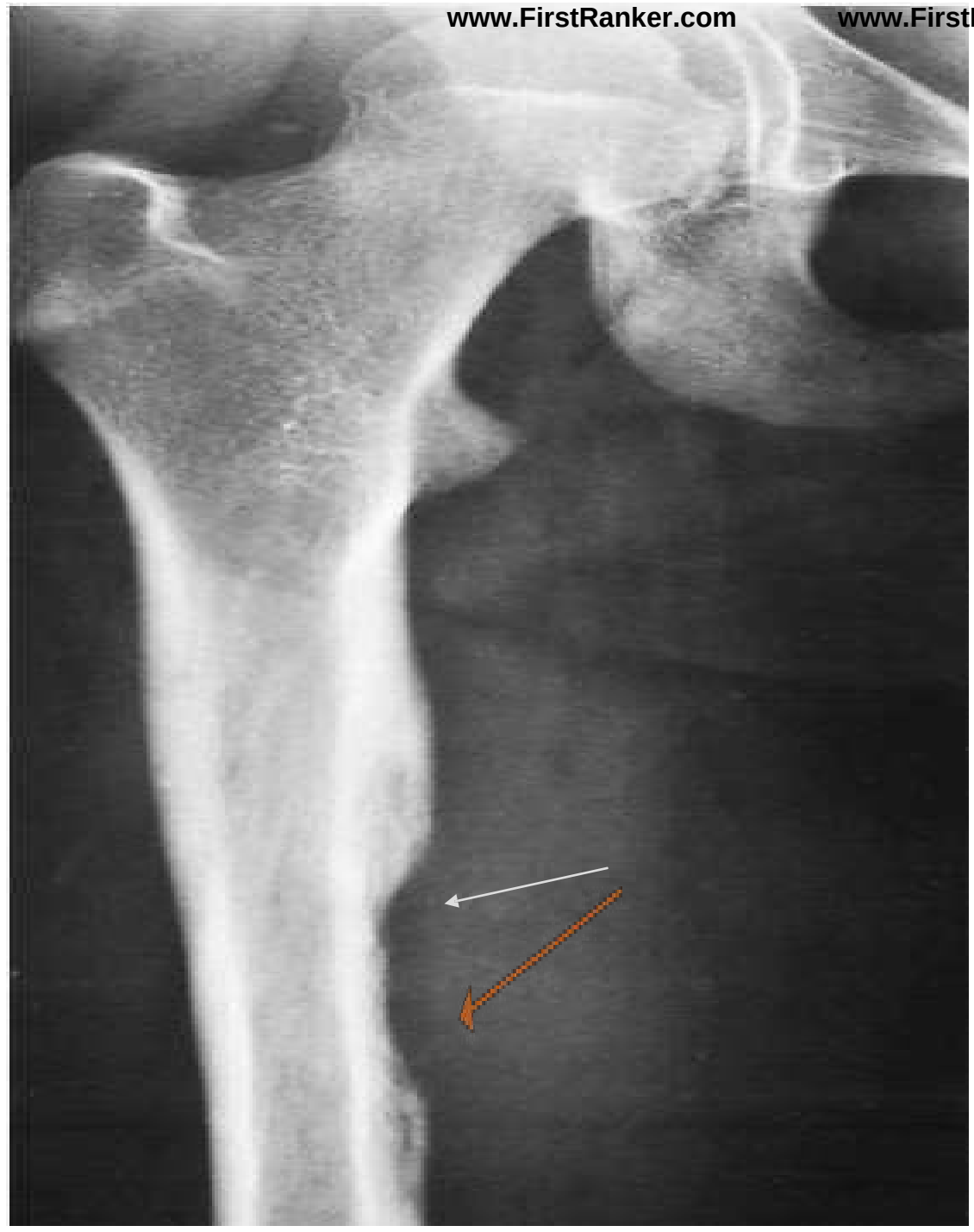
Radiological features

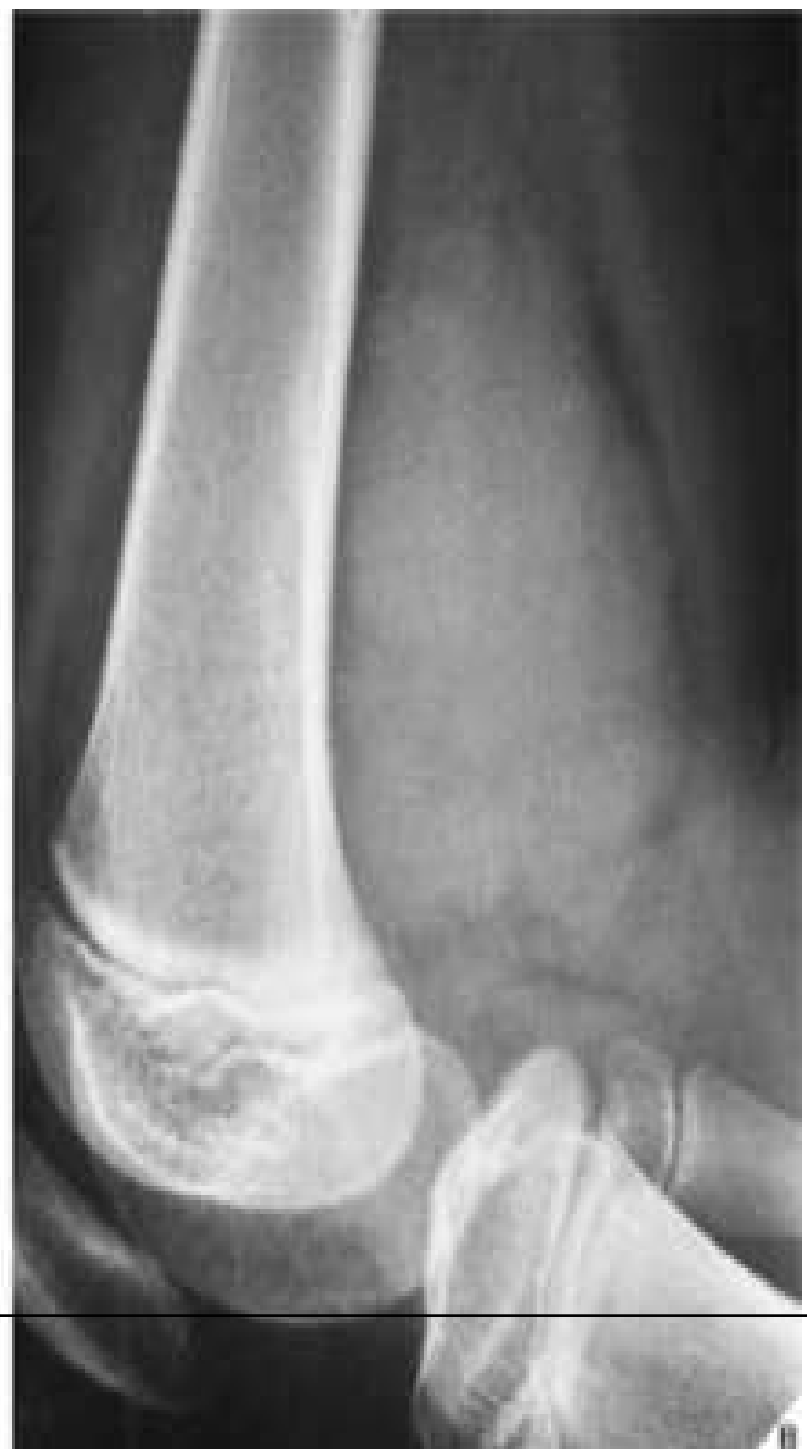
- lesion is poorly defined,
- Marked by a permeative or moth-eaten type of bone destruction,
- Associated with an aggressive periosteal response that has an onionskin (or “onion peel”)
- less commonly, a “sunburst” appearance,
- Large soft tissue mass, Occasionally, the bone lesion itself is almost imperceptible, with the soft-tissue mass being the only prominent radiographic finding

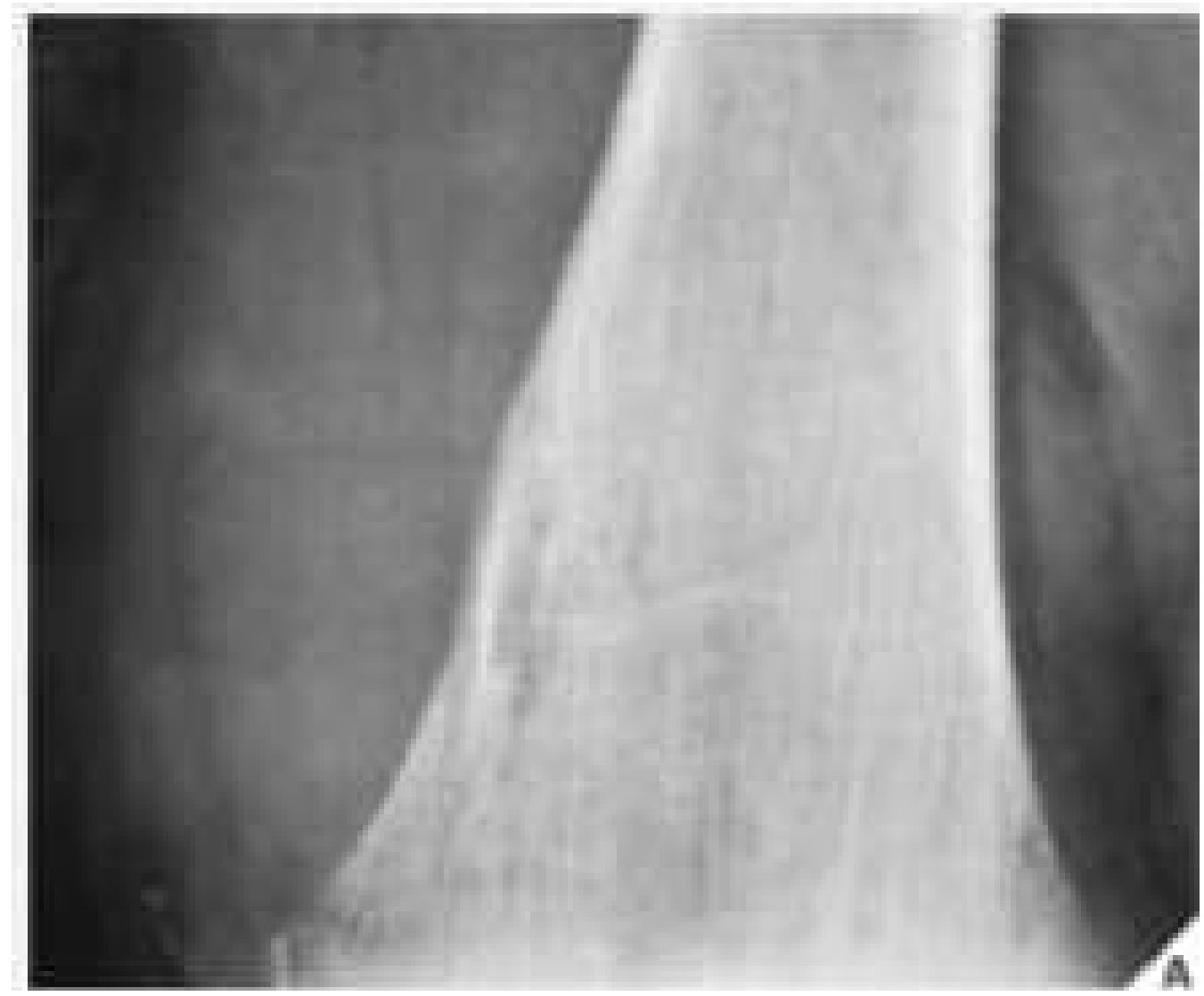


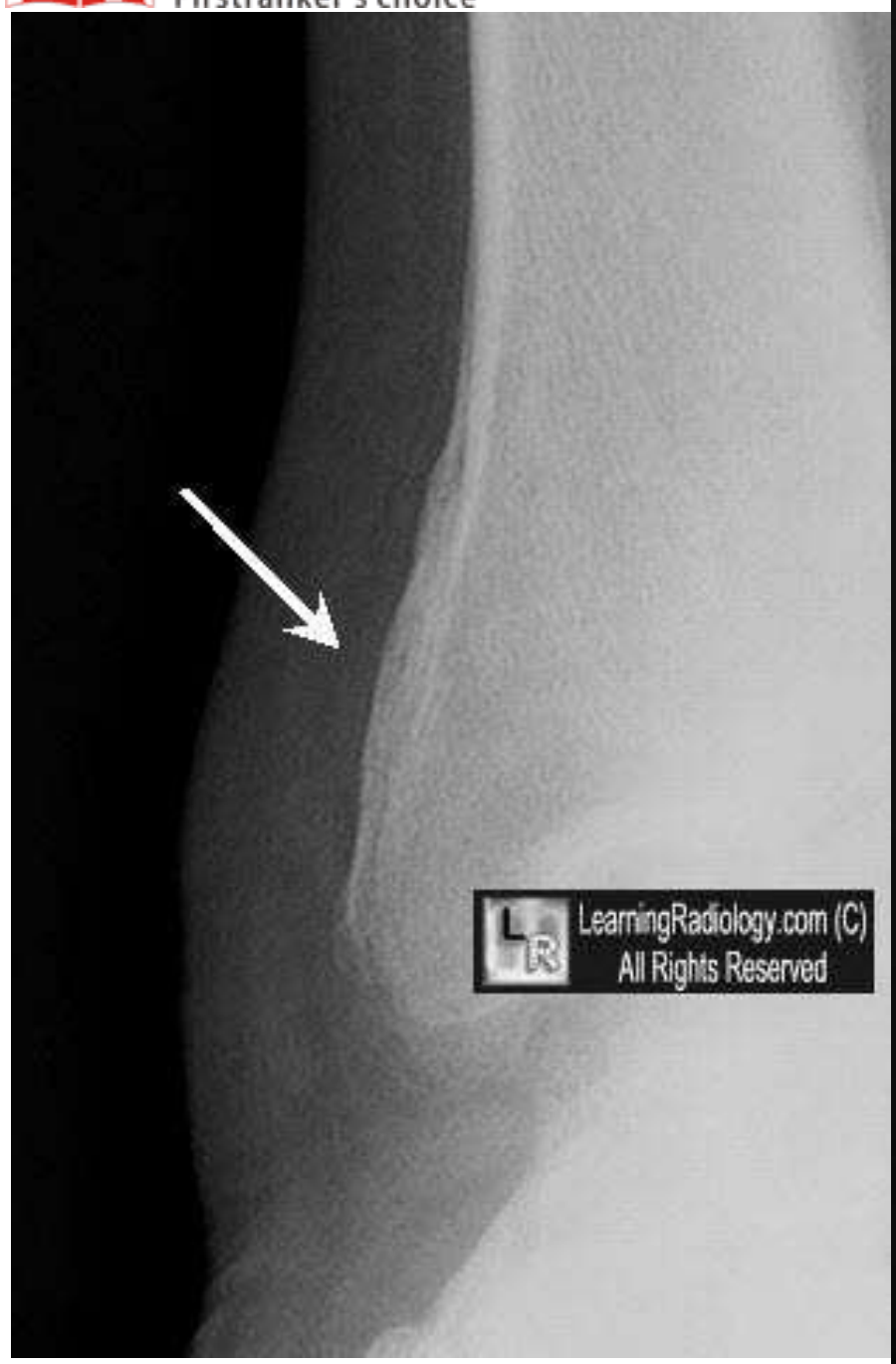
Radiological Features

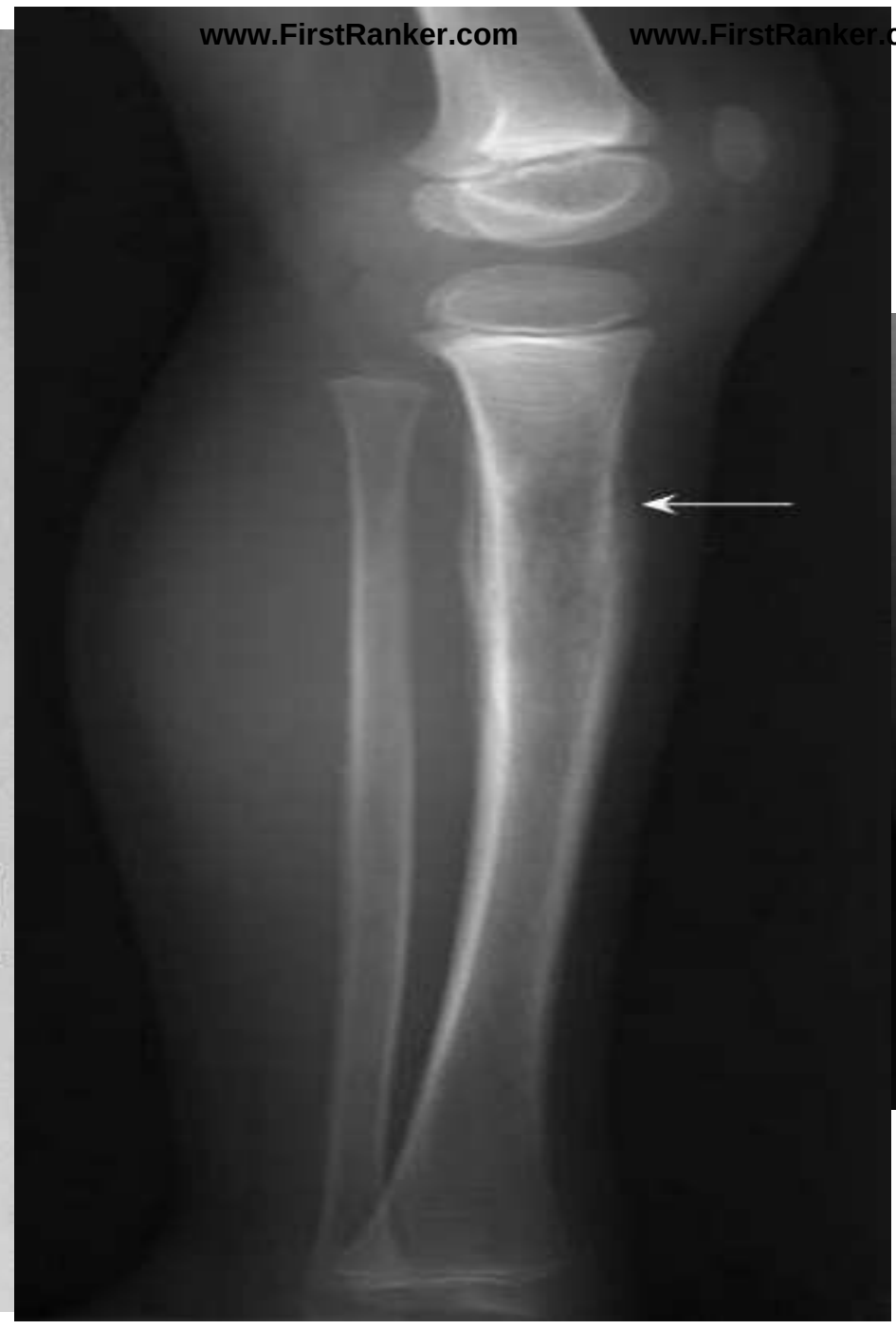


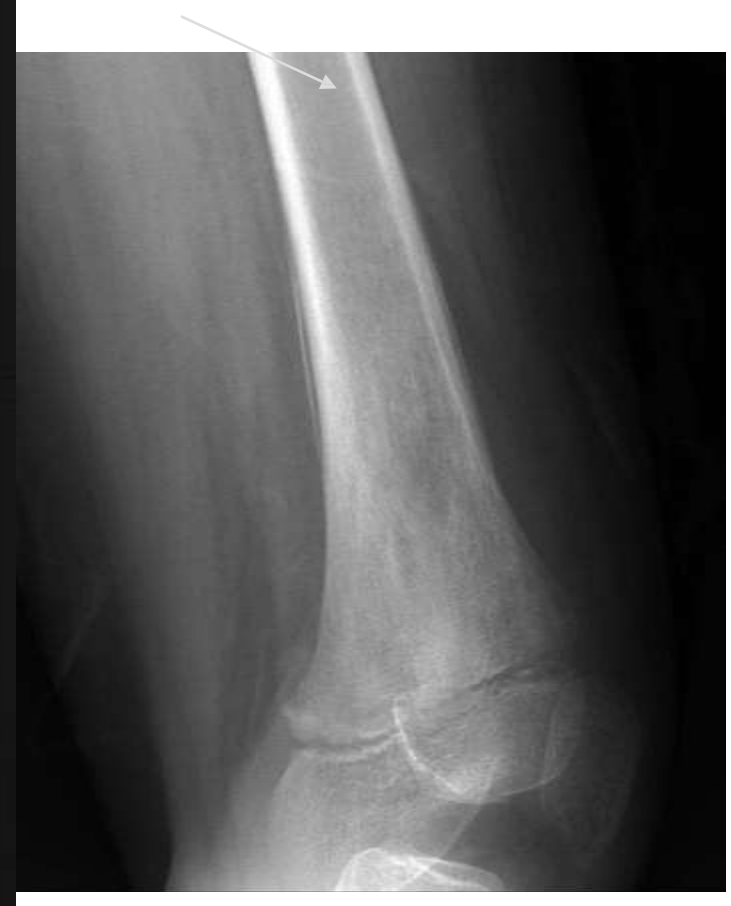






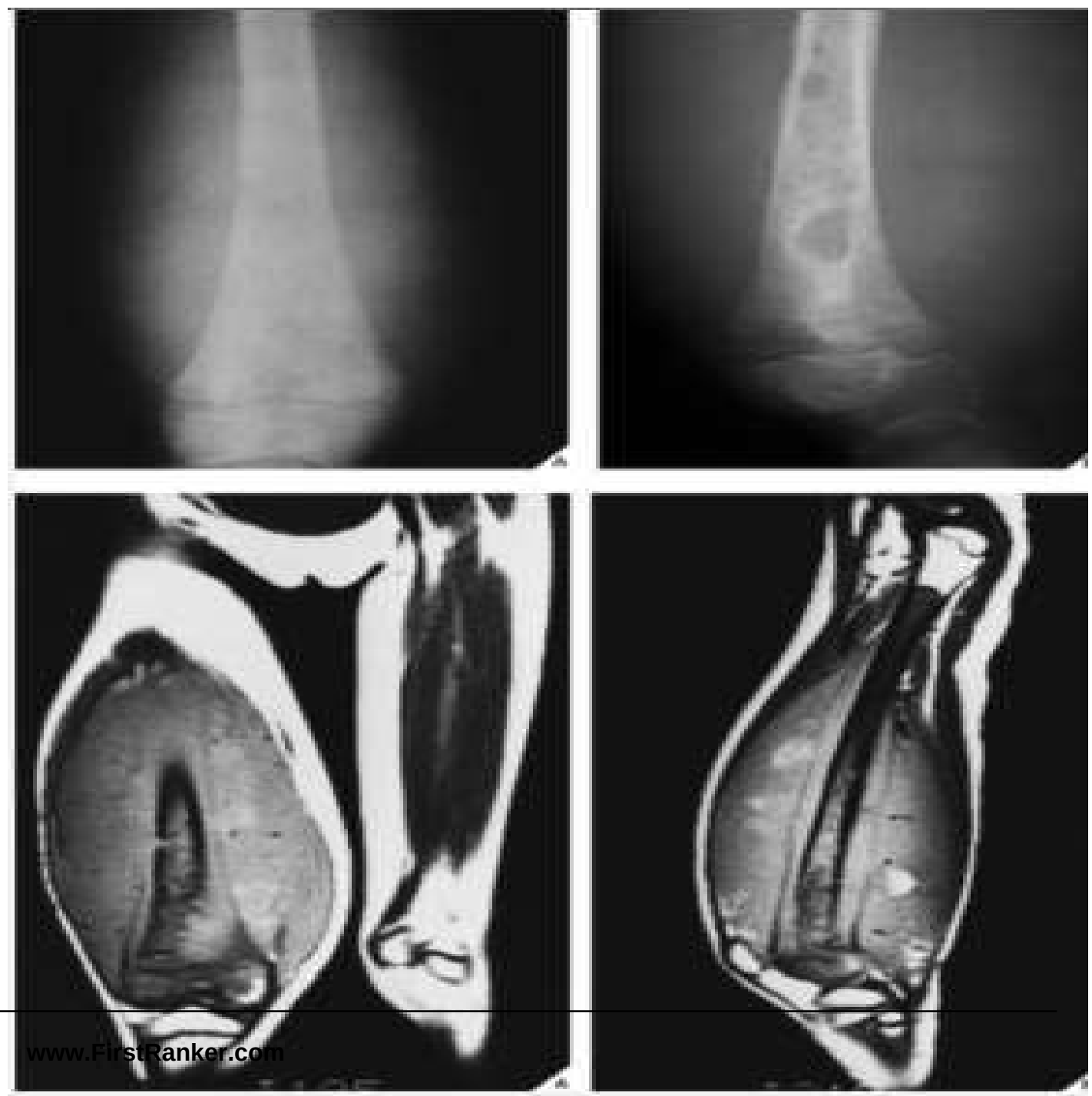


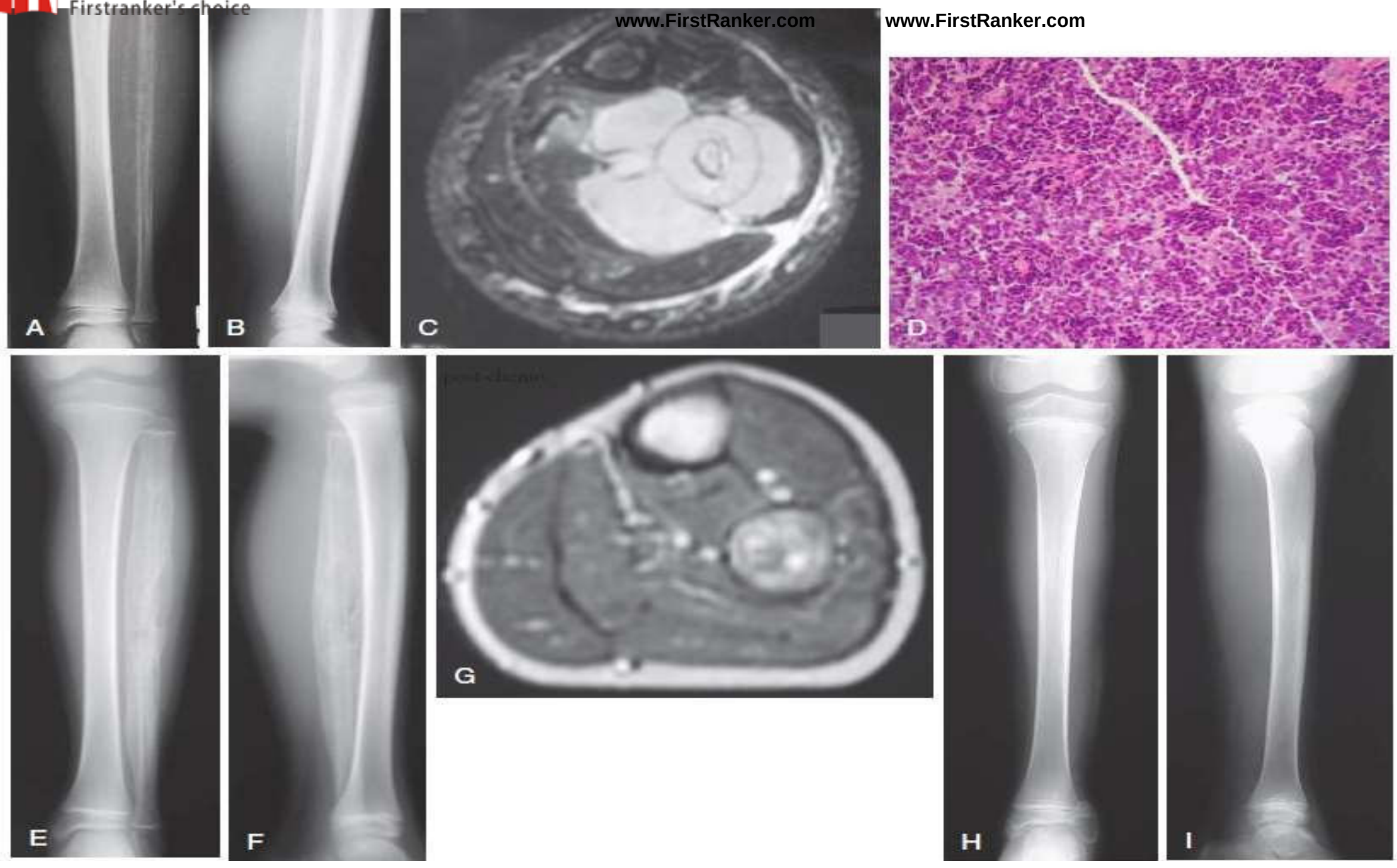




MRI

1. T1 : low to intermediate signal
2. T1 C+ (Gd) : heterogeneous but prominent enhancement
3. T2 : heterogeneously high signal, may see hair on end low signal striations

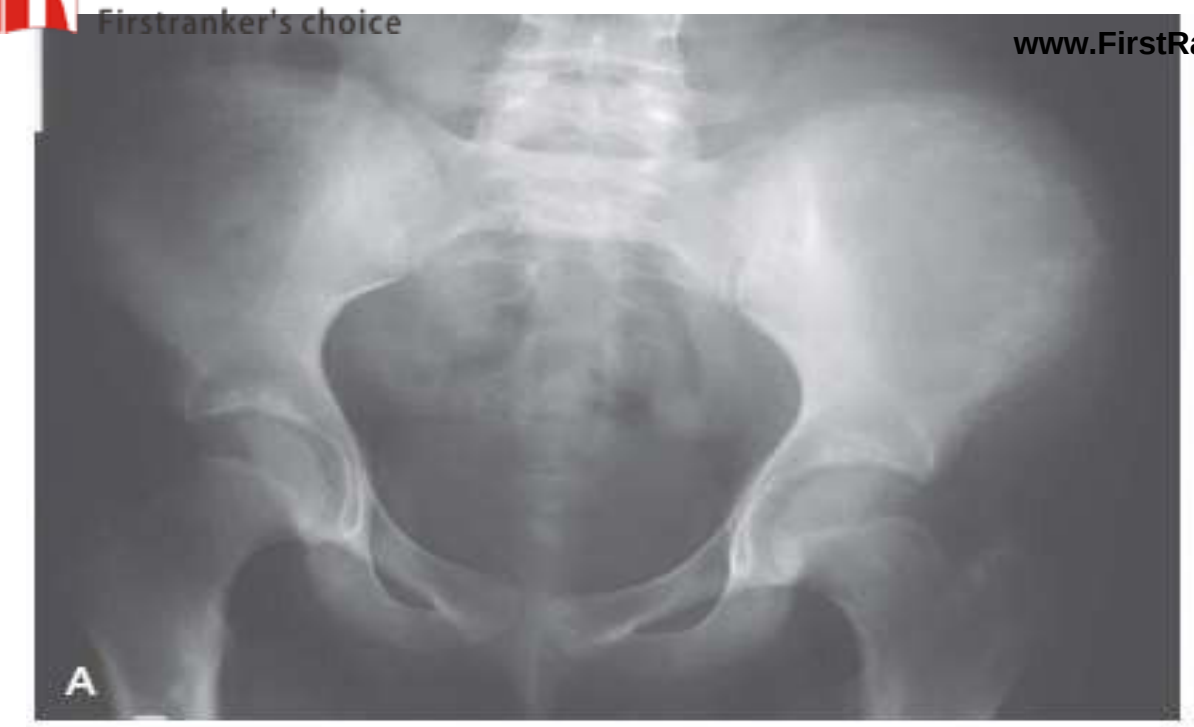


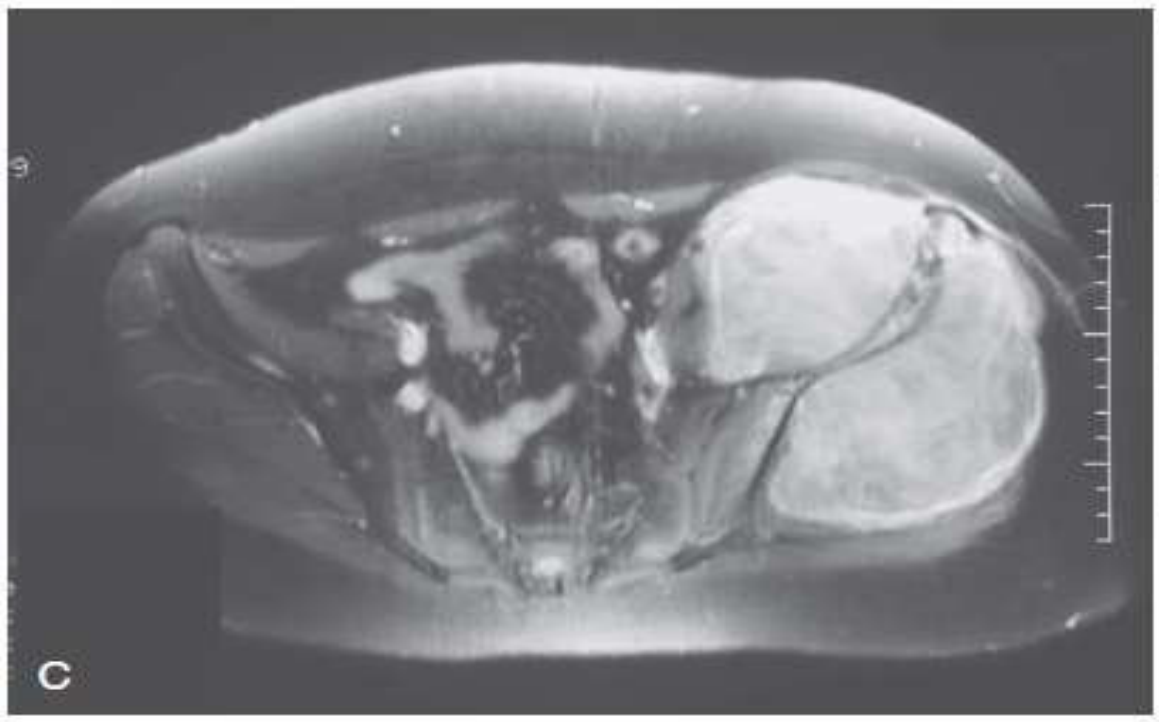


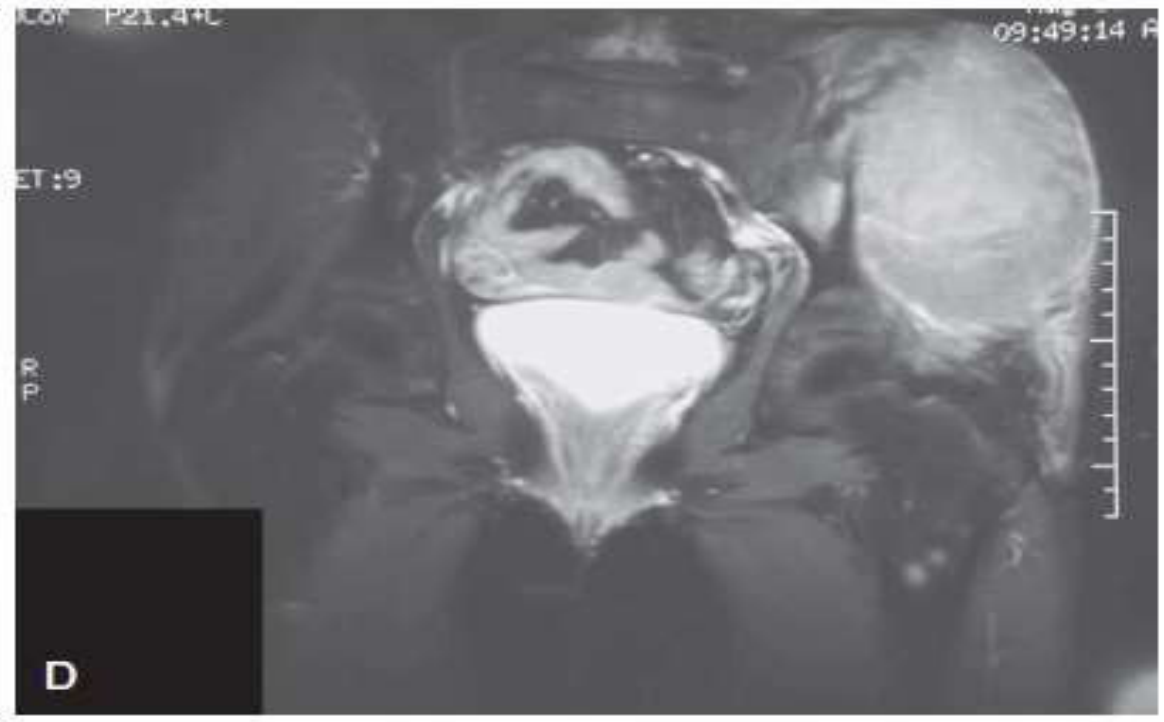
Nuclear medicine:

- Ewing sarcomas demonstrate increased uptake on both Gallium-citrate and

- Technetium99m methylene diphosphonate







Differential diagnosis

- other Ewing sarcoma family of tumours
  - pPNET : large soft tissue component with extension into bone
  - Askin tumour : chestwall
- osteosarcoma (ALP is not elevated in Ewing sarcoma)
- Leukemia
- Multiple myeloma
- Ostyeomyelitis

Ewing sarcoma – treatment

- Radiosensitive
- Large central unresectable mass – radiotherapy.
- Smaller more accessible lesions surgery.
- Neo adjuvant and adjuvant chemotherapy