# Langerhans cell histiocytosis

- Synonyms
  - Not recommended:
  - Eosinophilic granuloma
  - Langerhans cell granulomatosis
  - histiocytosis X
  - Hand-Schüller-Christian disease
  - Letterer-Siwe disease
- Lytic lesion(s), generally in child, with variable degrees of aggressiveness

# Langerhans cell histiocytosis

- Part of spectrum of histiocytic disorders: accumulation of macrophages, dendritic cells, or monocyte-derived cells in tissues
  - LCH: clonal neoplastic proliferation of myeloid dendritic cells with Langerhans cell phenotype
- Single system: unifocal or multifocal (typically skeletal)
- Multisystem: commonly skin, bone, liver, spleen, marrow

# Langerhans cell histiocytosis

- Can be associated with Erdheim-Chester disease (ECD)
- 20% of patients with ECD also have LCH lesions
- Either concomitantly or preceding
- Shared molecular alterations

# Prognosis

- Unifocal disease (eosinophilic granuloma): >95% survival
- Two organ involvement: 75% survival
- Langerhans cell sarcoma: 50% survival

# Chondrosarcoma

- Caution: central CS of femur or humerus is usually low grade and appears nonaggressive
- Substantial numbers of these lesions are underdiagnosed, either as EC or another lesion if no chondroid matrix is present
- Large "EC" in either femur or humerus should be viewed with suspicion
- Any thickening or prominent scalloping of cortex should heighten suspicion for CS
- Have low threshold for raising concern
- May be extremely difficult to differentiate ACT/CS1 from EC by imaging
- Scalloping of endosteal cortex over substantial length of lesion suggests ACT/CS1 on radiograph/CT
- Exception: eccentric EC arising adjacent to cortex expected to cause endosteal scalloping and even minor cortex disruption
- Increased matrix or enlargement of EC need not imply sarcomatous change; EC may show such alterations normally.

# Types

- 4 subtypes defined by location and grade
- Location
  - Central: within medullary cavity
  - Secondary peripheral: arising within cartilage cap of osteochondroma (OC)
    - » Lesions arising from enchondroma not considered secondary under WHO classification
- Grade
  - Atypical cartilaginous tumor (ACT)/CS, grade 1 (CS1): locally aggressive, hyaline cartilage-producing neoplasm
    - » ACT: lesions in appendicular skeleton (long and short tubular bones)
    - » CS1: lesions in axial skeleton (flat bones: pelvis, scapula, skull base)
  - CS2 (intermediate grade) and CS3 (high grade): malignant cartilage matrix-producing neoplasms
    - » Grouped together

# Imaging

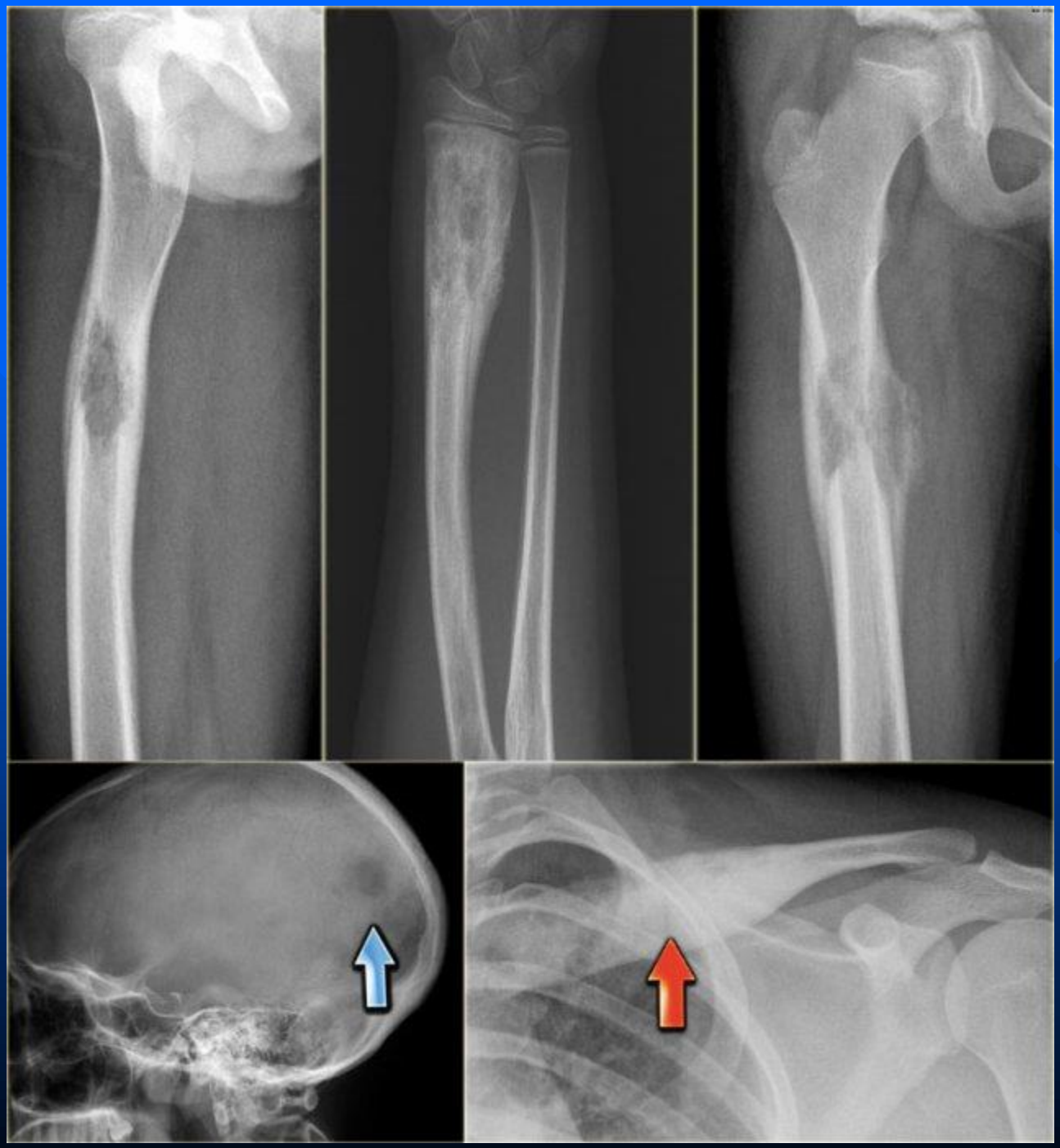
- Central ACT/CS1: can be difficult to differentiate from EC
  - Geographic with cortex scalloping without breakthrough
- Central CS2/CS3
  - Lytic lesion centrally in metaphysis > > diaphysis
  - Chondroid matrix variably present (78%) or entirely lytic
- Secondary peripheral CS2/CS3
  - Cartilage cap > 2 cm, solid enhancing areas in mass
- T1 MR: lesion fairly isointense to skeletal muscle
  - Entrapped foci of yellow marrow much less frequent in CS2/CS3 than in ACT/CS1 and EC
- Fluid-sensitive MR: variably inhomogeneous
  - ACT/CS1: lobulated high signal
  - CS2/CS3: greater inhomogeneity, ± lobulation
- C+ MR: varies with lesion grade
  - ACT/CS1: peripheral and septal enhancement with few nodular areas
  - CS2/CS3: more generalized enhancement + necrotic areas

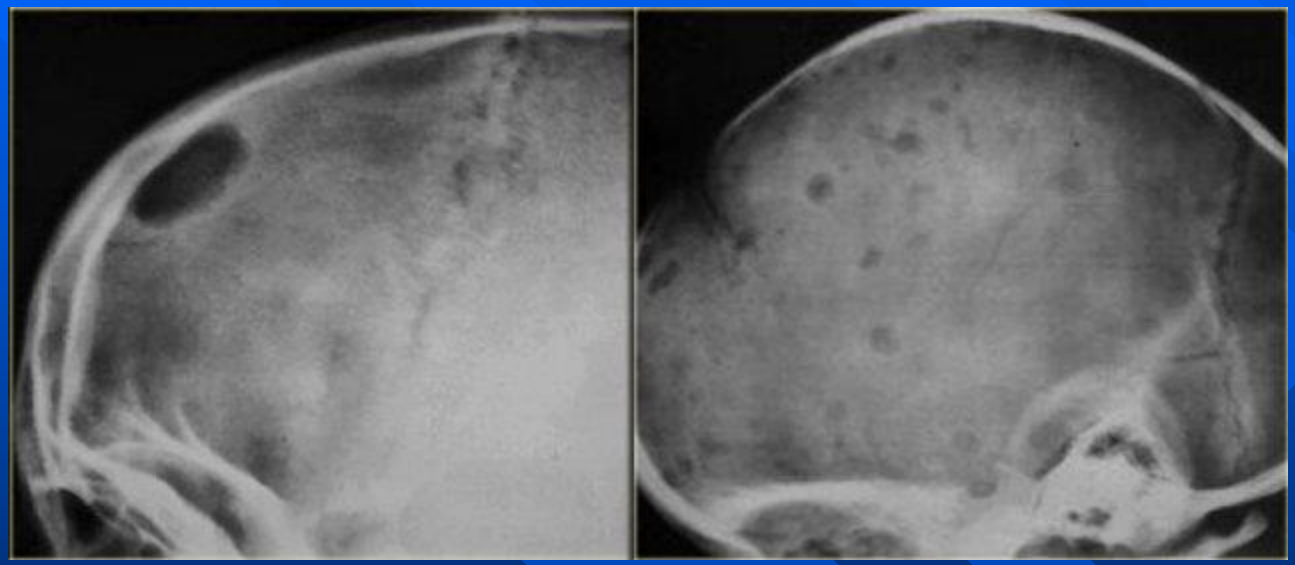
# Xray

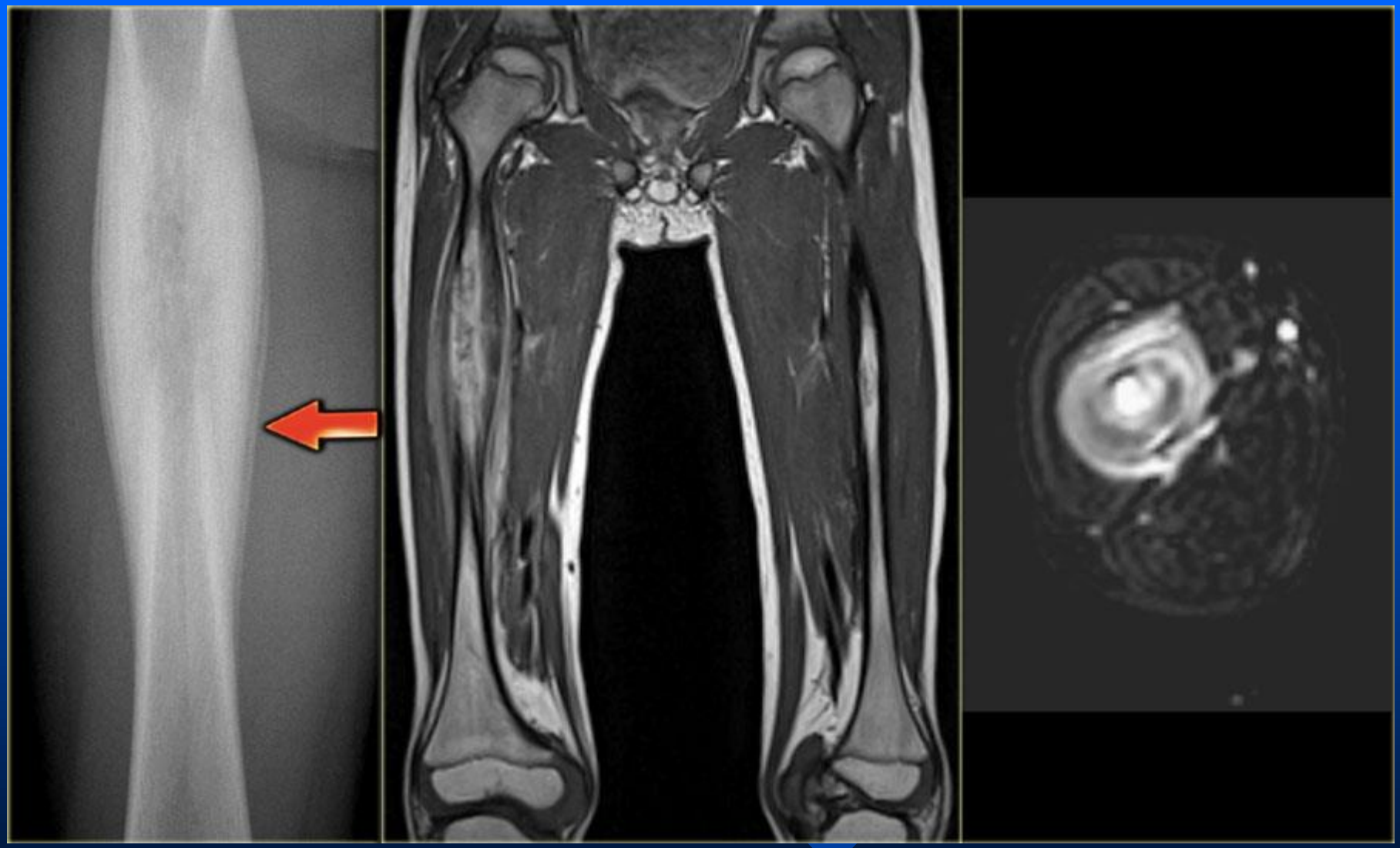
- **Size cut-off of 5 cm helpful for differentiating enchondroma (EC) from ACT/CS1**
- **If in doubt between enchondroma (EC), ACT/CS1, and CS2/CS3: follow-up**
- Lytic (50%)
- Intralesional calcifications: ~70% (rings and arcs calcification or popcorn calcification)
- Endosteal scalloping: affecting more than two-thirds of the cortical thickness (less than two-thirds in enchondromas)
- Moth-eaten appearance or permeative appearance in higher grade tumors (see chondrosarcoma grading), e.g. myxoid, dedifferentiated and mesenchymal chondrosarcomas
- Cortical remodeling, thickening and periosteal reaction are also useful in distinguishing between an enchondroma and low-grade chondrosarcoma (see enchondroma vs low-grade chondrosarcoma)

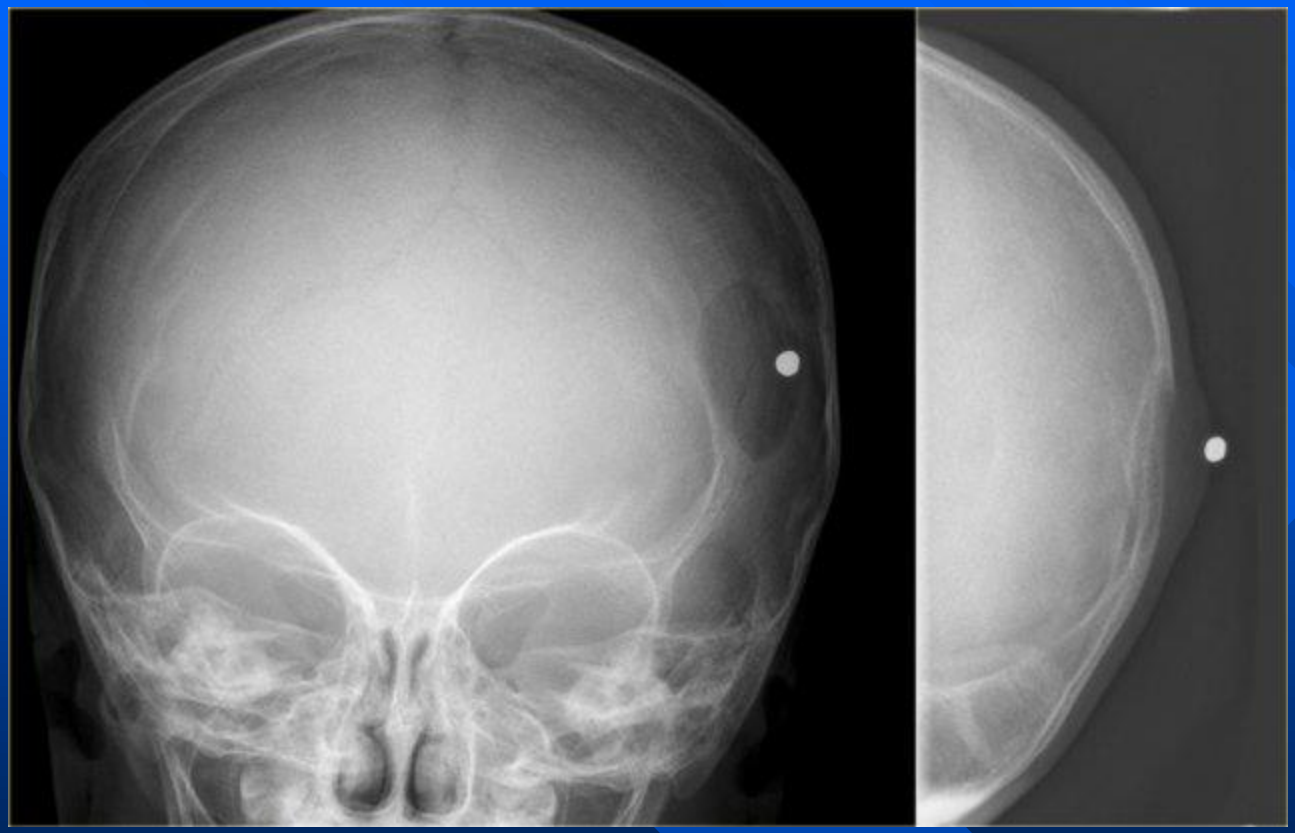
# CT

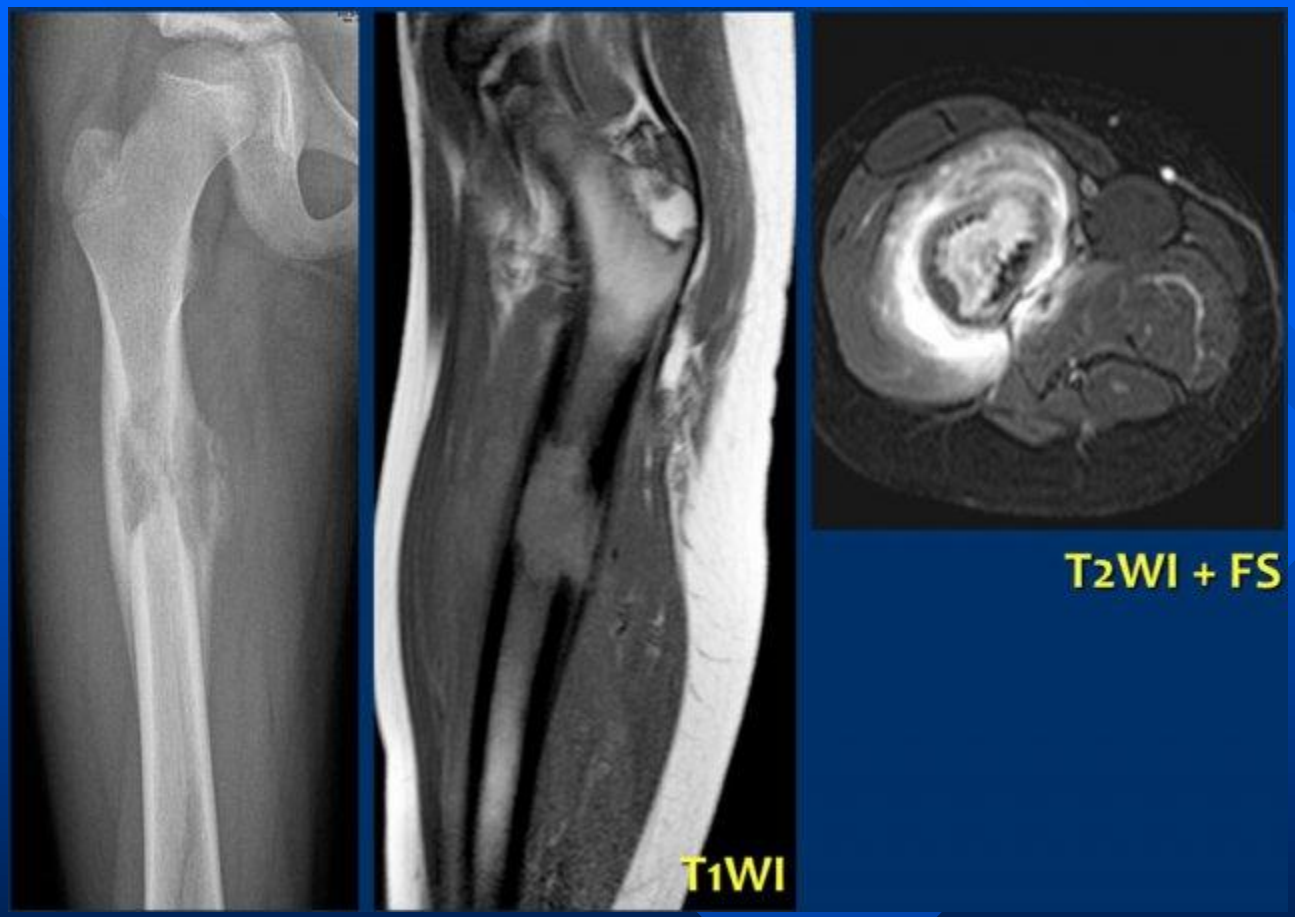
- Features seen on CT are the same as on plain film but are better seen:
- 94% of cases demonstrate matrix calcification, 60-78% on plain film
- Endosteal scalloping
- Cortical breach, seen in ~90% of long bone chondrosarcoma, only ~10% of enchondromas
- Soft tissue mass:
  - Tumor cellularity, and therefore density, increases with the increased grade of the tumor
- Heterogenous contrast enhancement







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