Lecture 5

BONE TUMORS AND TUMORLIKE CONDITIONS:

- Primary bone tumors are rare
- Benign >>>> malignant tumors
- First 3 decades (benign); adults more to be malignant
- Trx: aims to optimize survival while maintaining function
- Age & location help narrow ddx
- S&S: asymptomatic, pain, path #

Category	Behavior	Tumor Type	Common Locations	Age (yr)	Morphology
Cartilage forming	Benign	Osteochondroma	Metaphysis of long bones	10–30	Bony excrescence with cartilage cap
—	—	Chondroma	Small bones of hands and feet	30–50	Circumscribed hyaline cartilage nodule in medulla
—	Malignant	Chondrosarcoma (conventional)	Pelvis, shoulder	40–60	Extends from medulla through cortex into soft tissue, chondrocytes with increased cellularity and atypia
Bone forming	Benign	Osteoid osteoma	Metaphysis of long bones	10–20	Cortical, interlacing microtrabeculae of woven bone
—	—	Osteoblastoma	Vertebral column	10–20	Posterior elements of vertebra, histology similar to osteoid osteoma
—	Malignant	Osteosarcoma	Metaphysis of distal femur, proximal tibia	10–20	Extends from medulla to lift periosteum, malignant cells producing woven bone
Unknown origin	Benign	Giant cell tumor	Epiphysis of long bones	20–40	Destroys medulla and cortex, sheets of osteoclasts
—	—	Aneurysmal bone cyst	Proximal tibia, distal femur, vertebra	10–20	Vertebral body, hemorrhagic spaces separated by cellular, fibrous septae
—	Malignant	Ewing sarcoma	Diaphysis of long bones	10–20	Sheets of primitive small round cells

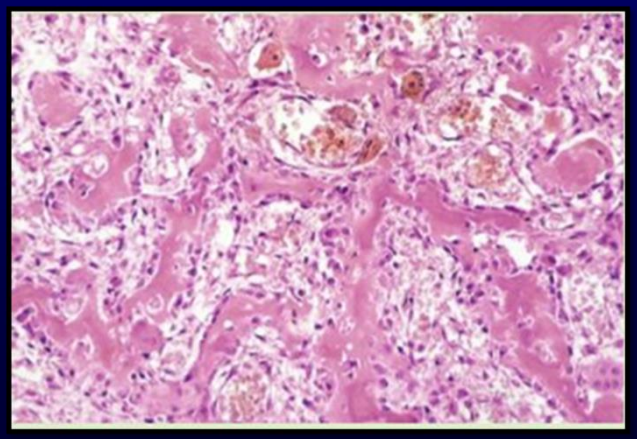
BONE-FORMING TUMORS

OSTEOID OSTEOMA

- < 2 cm
- Young men
- Femur & tibia; nidus with surrounding bone reaction
- Severe nocturnal pain (PGE2) relieved by aspirin & NSAIDS
- Treated by: radiofrequency ablation or surgery

OSTEOBLASTOMA

- > 2 cm
- Posterior vertebrae; no rim of bone reaction
- Pain unresponsive to aspirin
- Treated by curetting



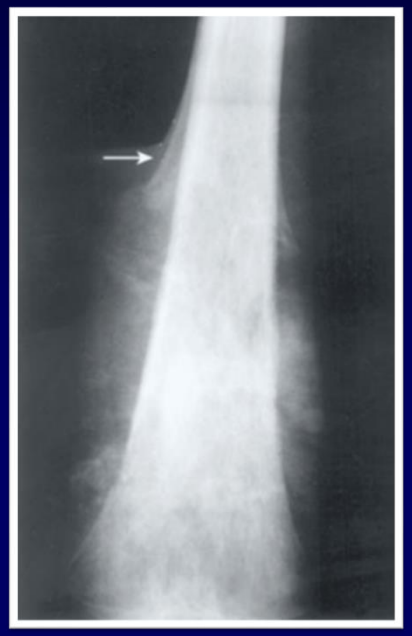
OSTEOSARCOMA:

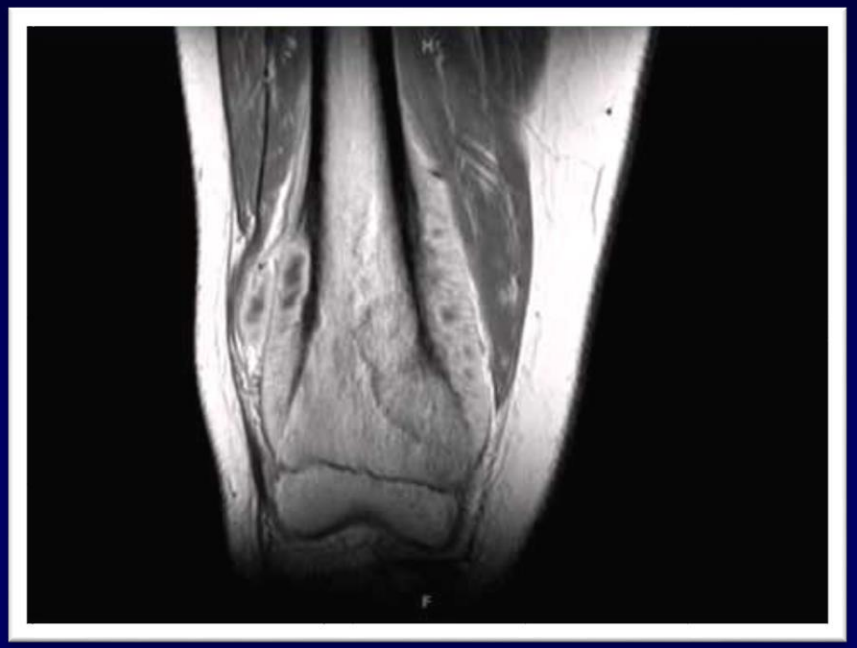
- Malignant osteogenic tumor
- Excluding hematopoietic malignancies; it is the most common primary malignant tumor of bone
- 75% adolescents; another peak in older (secondary osteosarcoma)
- Males > females (1.6:1.0)
- Metaphysis of long bones (distal femur & proximal tibia)

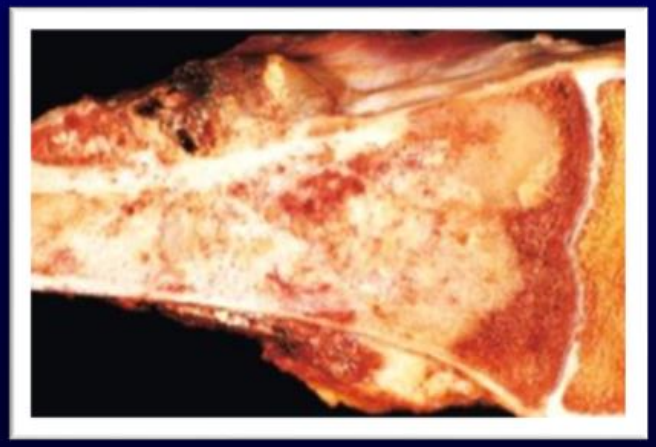
OSTEOSARCOMA:

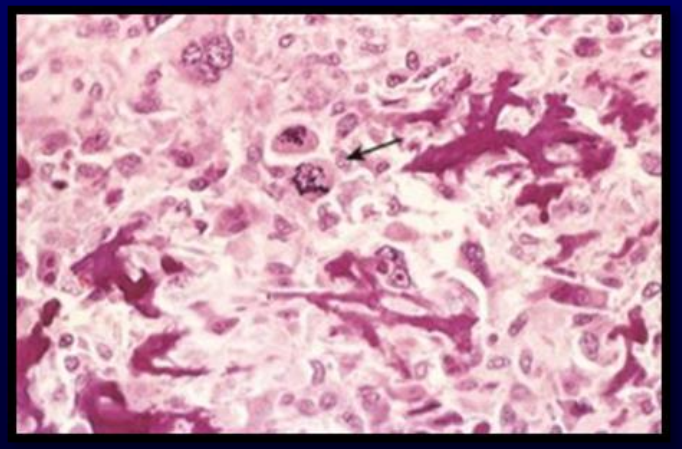
- **Progressive pain or #**
- **Imaging: large destructive and infiltrative lesions with Codman triangle**
- **Genetic abnormalities: mutations in RB gene, TP53 gene, CDKN2A (p16 & p14), MDM2 & CDK2**

OSTEOSARCOMA FEATURES:









OSTEOSARCOMA TREATMENT:

- Multimodality approach (MDTeam)
- 1. Neoadjuvant chemotherapy 2. Surgery 3. Chemotherapy
- Hematogenous spread to lungs
- 5 year survival reaches 60-70%
- Presence of mets at diagnosis is a bad prognostic factor

CARTILAGE-FORMING TUMORS:

- **Osteochondroma (benign exostoses): solitary (85%); part of multiple hereditary exostoses (MHE): EXT1, EXT2 gene mutations**
- **Rare (<3-5%) transformation to chondrosarcoma (more common in MHE)**

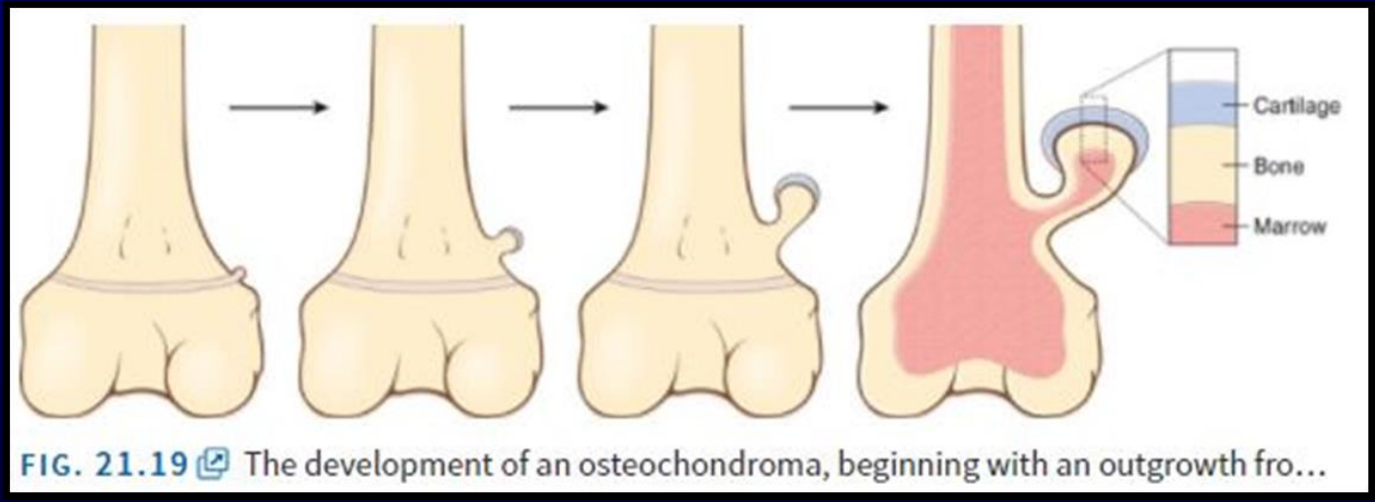
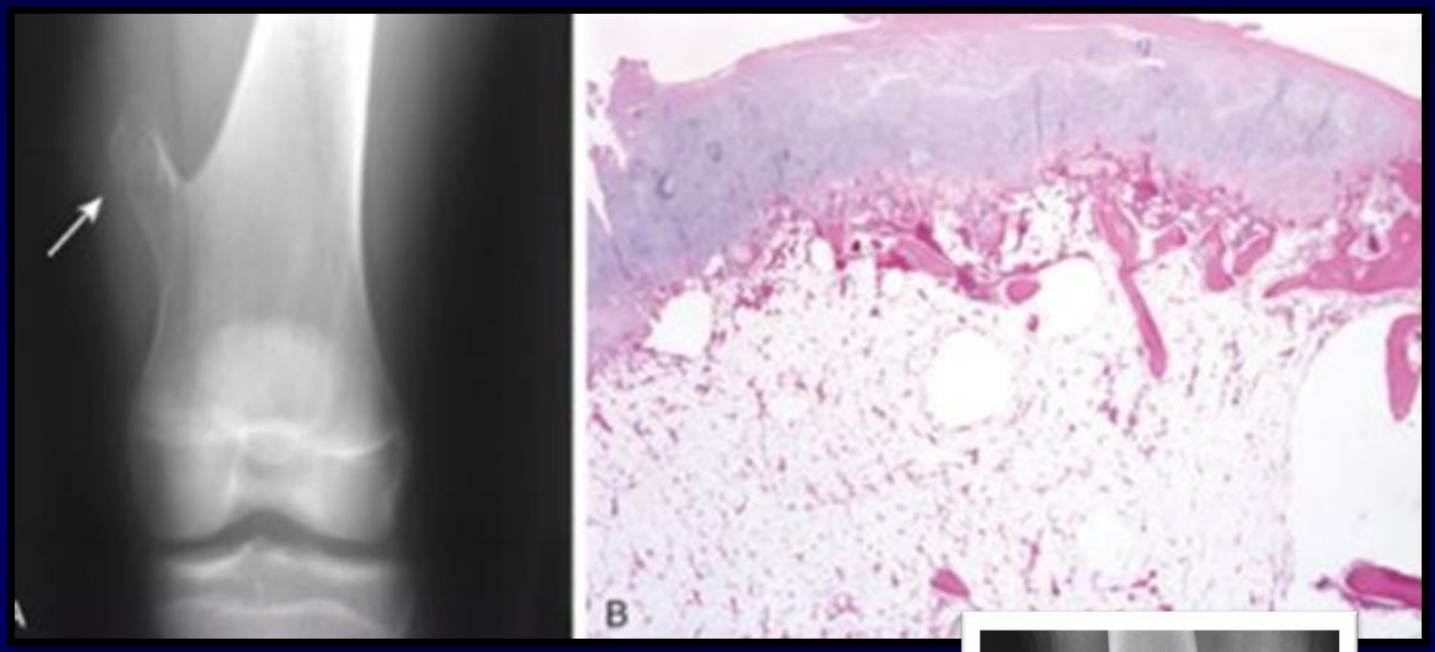
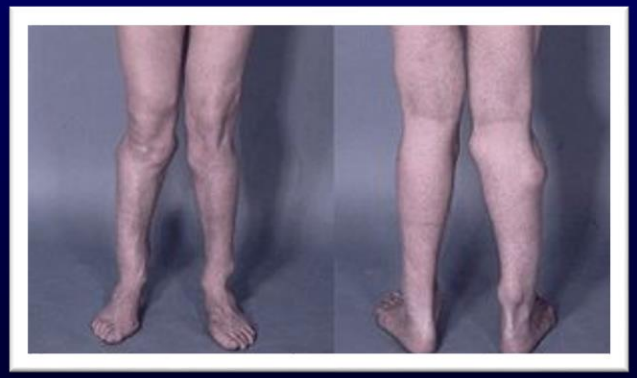
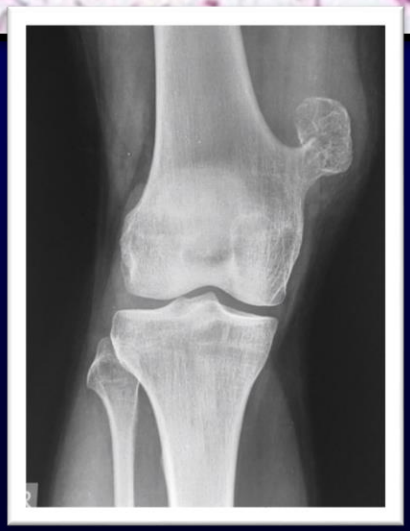


FIG. 21.19 The development of an osteochondroma, beginning with an outgrowth fro…

OSTEOCHONDROMA:

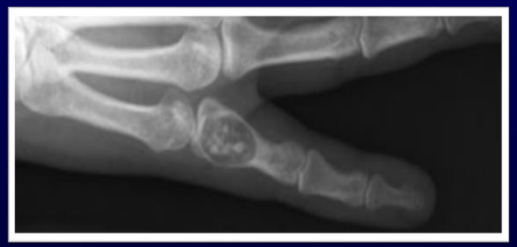


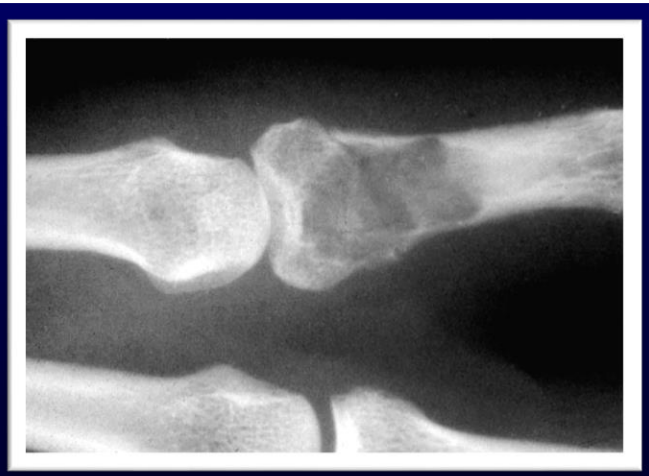


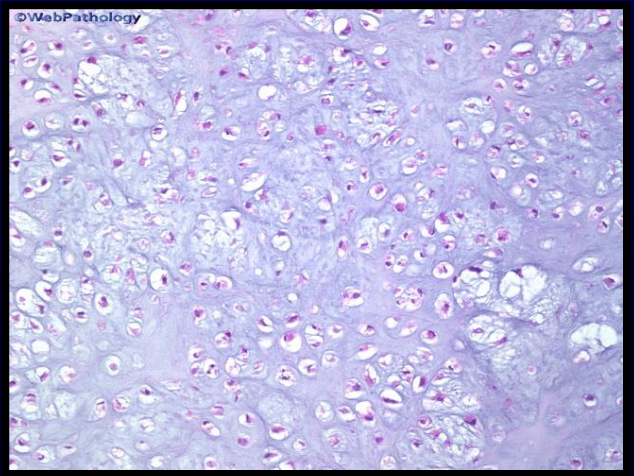


CHONDROMA (ENCHONDROMA):

- Benign hyaline cartilage tumors in bones with endochondral origin; medullary enchondroma or cortical chondroma
- Solitary metaphyseal lesions; 20-50 years
- Multiple enchondromas: Ollier disease
- Maffucci syndrome: multiple enchondromas + skin hemangiomatosis
- IDH1 & IDH2 gene mutations







CHONDROSARCOMA:

- **Malignant tumors producing cartilage**
- **50% incidence of osteosarcoma**
- **40-50 years of age; M:F (2:1)**
- **Large masses; shoulder, pelvis, ribs**
- **Genes: *EXT, IDH1, IDH2, COL2A1, CDKN2A***
- **Px: depends on grade (grade 1 excellent px)**
- **Trx: surgical +/- chemotherapy**

CHONDROSARCOMA FEATURES: