

Bone forming Tumors

	Osteoid Osteoma	Osteoblastoma	Osteosarcoma
Behavior	Benign <2 cm	Benign >2cm	<ul style="list-style-type: none"> - Malignant - Excluding hematopoietic malignancies; it is the <u>most common</u> primary malignant tumor of bone - Hematogenous spread to lungs - 5-year survival reaches 60-70% - Presence of metastasis at diagnosis is a bad prognostic factor
Age & gender	Young men 10- 20	10 – 20	<ul style="list-style-type: none"> - 75% adolescents 10 -20 - another peak in older (secondary osteosarcoma) - Males > females
Common locations	Femur and tibia metaphysis of long bone	Posterior vertebrae	Metaphysis of long bones (distal femur & proximal tibia)
Morphology	nidus with surrounding bone reaction Cortical, interlacing micro trabeculae of woven bone	no rim of bone reaction Posterior elements of vertebra, histology similar to osteoid osteoma	Imaging: large destructive and infiltrative lesions with Codman triangle Extends from medulla to lift periosteum, malignant cells producing woven bone
S & S	Severe nocturnal pain (PGE2) relieved by aspirin & NSAIDS	Pain unresponsive to aspirin	Progressive pain or fractures
Treatment by	radiofrequency ablation or surgery	curetting	<ul style="list-style-type: none"> - Multimodality approach (MD Team) - 1. Neoadjuvant chemotherapy 2. Surgery 3. Chemotherapy

CARTILAGE-FORMING TUMORS

	Osteochondroma	Chondroma (Enchondroma)	Chondrosarcoma
Behavior	benign exostoses	-Benign hyaline cartilage tumors in bones with endochondral origin -medullary enchondroma or cortical chondroma	- Malignant tumors producing cartilage - 50% incidence of osteosarcoma - Px: depends on grade (grade 1 excellent px) - Excluding hematopoietic malignancies; it is the <u>third most common</u> primary malignant tumor of bone
Age & gender	10-30	20-50	40-50 M:F (2:1)
Common locations	Metaphysis of long bones	Solitary metaphyseal lesions Small bones of hands and feet	shoulder, pelvis, ribs
Morphology	- solitary (85%) -part of multiple hereditary exostoses (MHE) -Rare (<3-5%) transformation to chondrosarcoma (more common in MHE) Bony excrescence with cartilage cap	Circumscribed hyaline cartilage nodule in medulla	Large masses Extends from medulla through cortex into soft tissues, chondrocytes with increased cellularity and atypia Codman triangle
S & S			
Treatment by			surgical +/- chemotherapy



Multiple enchondromas: Ollier disease

Maffucci syndrome: multiple enchondromas + skin hemangiomatosis

Unknown origin			
	Giant cell tumor of bone	Aneurysmal bone cyst	Ewing sarcoma
Behavior	Benign Rare malignant behavior	Benign Blood filled cyst	-Malignant - 2nd most common sarcoma of bone after osteosarcoma -The most common translocation, present in about 90% of Ewing sarcoma cases, is t(11;22) (q24; q12), which generates an aberrant transcription factor through fusion of the EWSR1 gene with the FLI1 gene.
Age & gender	Locally aggressive neoplasm of adults. 20 – 40	Adults 10-20	< 20 years
Common locations	Epiphyses of long bones	Metaphysis of long bones (proximal tibia, distal femur, vertebra)	Diaphysis of long bones
Morphology	- Osteoclast-like giant cells - Cells contain high levels of RANKL - destroys medulla and cortex, sheets of osteoclasts	Vertebral body, hemorrhagic spaces separated by cellular fibrous septae	Small blue cell tumor (PNET) Codman triangle Sheets of primitive small round cells
S & S			
Treatment by	curetting		neoadjuvant CT followed by surgery; long term survival now reaches 75%

Lesions stimulating primary neoplasm:

Non-ossifying fibroma	Fibrous dysplasia (FD)
Benign lesion maybe reactive not a true neoplasm (other names: FCD, MFD)	Not a real tumor; rather a developmental abnormality of bone genesis due to mutations in GNAS1 gene (cAMP mediated osteoblast differentiation)
Metaphysis	Forms of FD: -Monostotic: affecting one bone -Polyostotic: multiple bones -Mazabraud syndrome: FD + soft tissue myxoma <u>-McCune-Albright syndrome: polyostotic FD + caféau-lait skin pigmentation + endocrine abnormalities (precocious puberty)</u>
Histology: bland fibroblastic proliferation	
May resolve spontaneously	

