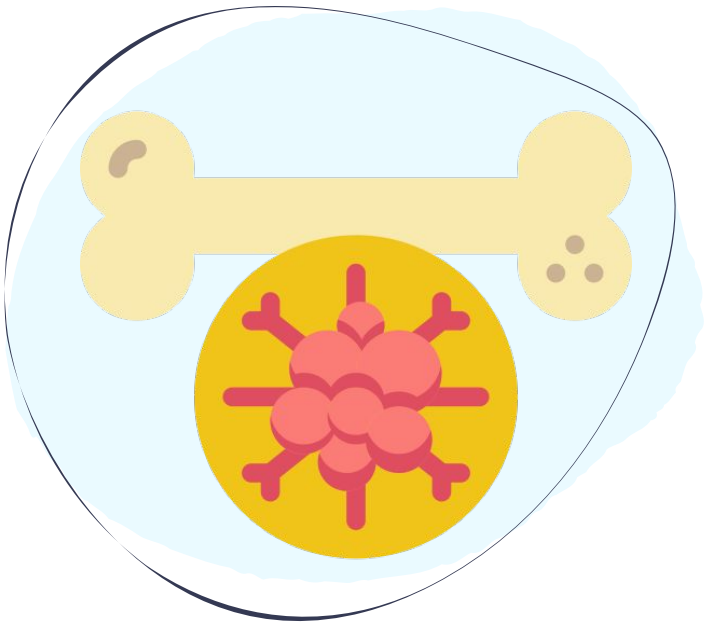
16

Orthopedic 441 Team

MED441 KING SAUD UNIVERSITY

Editing File

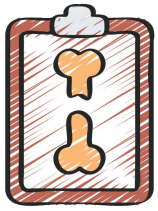


MSK Tumors

Dr. Ibrahim Alshaygy

Color Index:

- Main Text
- Important
- 441 Notes
- Old Notes
- Extra



Objectives

Discuss presenting history and physical examination features of bone tumors.

Discuss imaging characteristics of bone tumors.

Discuss biopsy principles and techniques for bone tumors.

Resources

Classification of bone tumors

- Benign[1]
- Malignant[2]
  - Primary
  - Secondary (Malignant transformation of benign process)
  - Metastatic[3]

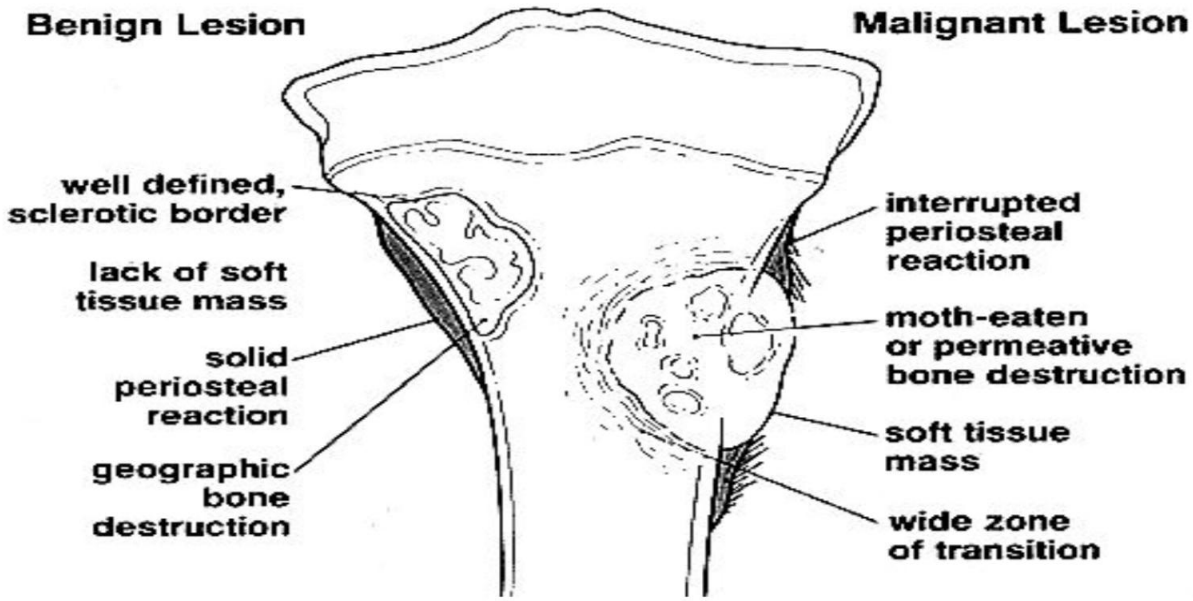


Table 25. Distinguishing Benign from Malignant Bone Lesions on X-Ray

Benign	Malignant
No periosteal reaction or benign appearing reaction (e.g. uniform smooth periosteal thickening as seen in a healing fracture)	Acute periosteal reaction • Codman's triangle • "Onion skin" • "Sunburst"
Sharp, well-demarcated borders, narrow zone of transition (between lesion and normal bone, suggesting slow-growing lesion)	Poorly defined borders, with a wide zone of transition, or infiltrative (suggesting fast-growing lesion)
Well-developed bone formation Intraosseous and even calcification	Varied bone formation Extraosseous and irregular calcification
No soft tissue mass	Soft tissue mass present
No cortical destruction or uniform cortical destruction in some low grade and locally aggressive benign lesions	Aggressive cortical destruction or tumour infiltration without cortical destruction

Adapted from: Buckholtz RW, Heckman JD. Rockwood and Green's Fractures in Adults. Volume 1. Philadelphia: Lippincott Williams & Wilkins, 2001. p558

1-Most bone tumors are benign. Common in pediatrics.
2-Most malignant tumors are metastatic. Primary is very rare.
3-paired organs (such as breasts, lungs, kidneys) and organs with 2 lobes (such as thyroids, prostates) are the most common primary carcinomas which might metastasise to the bone and cause secondary lesions in the bone.
-Embryological origins:

- Ectoderm-nerves, skin
- Endoderm-viscera
- Meso-bone, cartilage, muscle, nerves, blood vessels

Classification of bone tumors

The red color is based on previous teams not 441

Histologic type	Benign	Malignant
Bone marrow Hematopoietic and lymphatic	Giant cell tumor (osteoclastoma), Eosinophilic granuloma, Lymphangioma	Myeloma (plasmacytoma), Malignant giant cell tumor, Histiocytic lymphoma, Ewing's sarcoma, Leukemia
Cartilage-forming Chondrogenic	Enchondroma (chondroma), Periosteal (juxtacortical) chondroma, Enchondromatosis (Oiller's disease), Osteochondroma (osteocartilaginous exostosis, single or multiple) Chondroblastoma, Chondromyxoid fibroma	Chondrosarcoma (central), Conventional, Mesenchymal, Clear cell, Dedifferentiated, Chondrosarcoma (peripheral), Periosteal (juxtacortical)
Bone-forming Osteogenic	Osteoma, Osteoid osteoma, Osteoblastoma	Osteosarcoma (and variants), Juxtacortical osteosarcoma (and variants)
Histiocytic origin	Fibrous histiocytoma	Malignant fibrous histiocytoma
Fibrous and fibrohistiocytic Fibrogenic	Fibrous cortical defect (metaphyseal fibrous defect), Nonossifying fibroma, Benign fibrous histiocytoma, Fibrous dysplasia (mono- and _polyostotic), Periosteal desmoid, Desmoplastic fibroma, Osteofibrous dysplasia (Kempson- Campanacci lesion), Ossifying fibroma (Sissons' lesion)	Fibrosarcoma, Malignant fibrous histiocytoma
Neural Neurogenic	Neurofibroma	Malignant schwannoma
Vascular	Hemangioma, Glomus tumor, Cystic angiomatosis	Hemangioendothelioma, angiosarcoma, Hemangiopericytoma
Fat Lipogenic	Lipoma	Liposarcoma
Notochordal	Neurilemmoma	Chordoma
Unknown origin	Simple bone cyst, aneurysmal bone cyst, Intraosseous ganglion	Adamantinoma

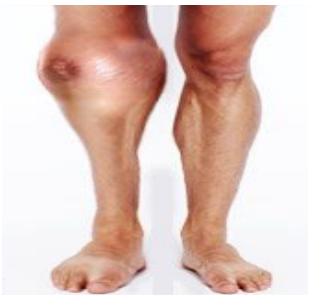
★ The diagnosis of tumors is made by History, physical examination, and investigations.

Key History Questions:

- Onset of pain: traumatic vs. atraumatic (Ask the pts if he/she has this pain prior to the trauma?)
- Progressive pain: indicates metabolic activity in the bone.
- Rest pain (to exclude mechanical pain), Night pain (night pain that fades with NSAIDs is **osteoid osteoma**).
- Relieving factors (NSAIDs)
- Family history.
- History of radiation (radiotherapy), Paget's disease, other cancers, systemic diseases.
- Constitutional symptoms (Weight loss, fever, night sweat, loss of appetite), not the primary presentation, as they present late (metastasis).
- DDx of any tumor is infection.

Clinical Features ★

- Pain
  Not just mechanical, usually even at rest
- Pathologic fractures
  Disease eating the bone —->bone become weak—->easily break
- Mass[1]
- Incidental findings on x-ray



Physical Exam

- Lump/mass physical exam:
  - 2 Ts: Temperature and Tenderness.
  - 3 Ss: Site, Size and Shape.
  - Margins (demarcated or not), consistency (solid or not), pounding pulse (e.g. Pulse proximal to tumor), decreased pulse distal to tumor?
  - Fixed or mobile[2].
- Lymphadenopathy: To check if there is metastasis or not.
- Neurovascular examination.

Investigations

- **Lab**: CBC (infection or anemia), ESR, CRP, Acute phase reactive molecules, hematological investigations
- **Imaging**:
  1. Local: X-ray (for the entire segment, cause some tumors have skip feature), MRI, CT.
  2. Systemic: chest CT, Bone scan.
- Biopsy: biopsy should be done by who will do the definitive surgery, ideally orthopedic oncologist or interventional radiologist who works with the orthopedic oncologist.

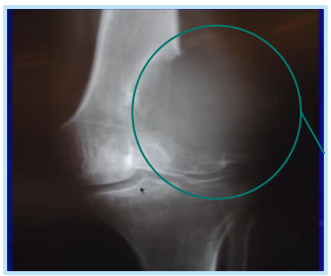
1-Progressive, mostly painless..pain if it compressed muscle, nerve ...etc.
2-how do we differentiate?
- Mass is fixed during relaxation and Contraction = fixed to the bone.
- Mass is mobile during relaxation and fixed during Contraction = deep to the superficial fascia (in the muscle).
- Mass is mobile in both contraction and relaxation = above the superficial fascia.

MSK bone tumors

Nine questions of Tumor Staging

1- Where is the lesion?

- Which bone? Which side?
- Which part of the bone? Epiphyseal (such as chondroblastoma), metaphyseal (such as osteosarcoma), diaphyseal (such as Ewing).
- Surface.
- Periarticular.
- Central or eccentric.
- The pic shows a tumor in the epiphyseal-metaphyseal area, eccentric, periarticular with cortical erosion, no periosteal reaction. There is a pathological fracture.

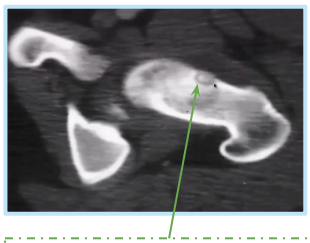


No clear demarcation

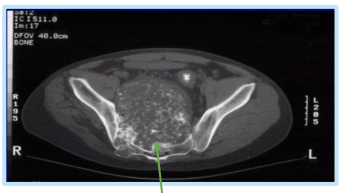
2- How big is the lesion

Size of mass is directly correlated to its ability to metastasize (size of army, # of cells)
-track how fast it grows over time
-compression of surrounding structures (localization is a mere important for that though)

osteoid osteoma (Benign)



Neck lesion 1x0.5 cm

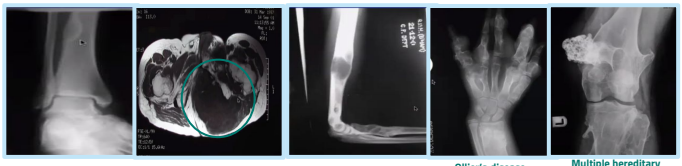


15x8 cm (Huge mass)

3- Is it solitary or multifocal?

The only way to know this is by getting the entire segment of the tumour and sometime we do skeletal survey to upper and lower extremities if we are suspecting it has metastasized to other sides.

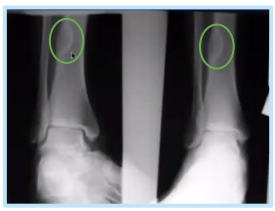
Solitary | Multifocal



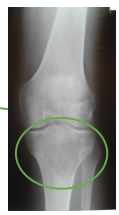
Note[1] | Ollier's disease (Aggressive) | Multiple hereditary exostosis (Benign)

4- What is the interface between the bone and the lesion?

Don't say it's a malignant say aggressive, cause aggressive either benign or malignant.



Good demarcated sclerotic lesion



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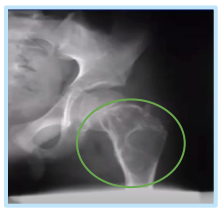
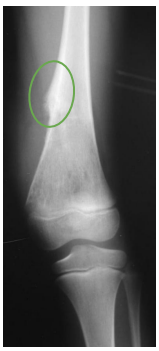


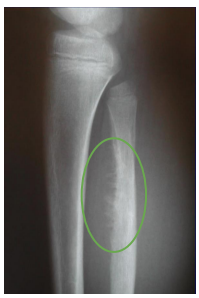
Well defined lytic lesion



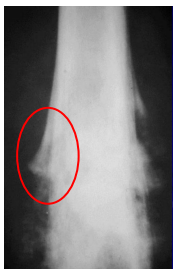
Ill defined lesion having Periosteal reaction (codman triangle) = bone try to form periosteal but مايلحق

5- Is there a periosteal reaction? (Thin layer surrounding the bone.)

Body's way of trying to contain it. Not a part of the tumor. "Sclerotic rim"



Sunburst
usually in Ewing Sarcoma



Cloud-like = sign of Bone-forming tumor

Codman triangle
(Commonly with osteosarcoma)

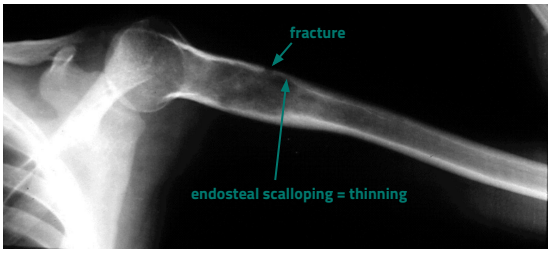
1-Right pic: Here you focused on the big tumor in the humerus so you told me that's a solitary tumor, but there is a pathological fracture in forearm, its thin and not easy to detect but if you look carefully it does not not look normal, so it's not actually solitary. That's why we emphasize to do x-ray for the entire segment.

MSK bone tumors

Nine questions of Tumor Staging

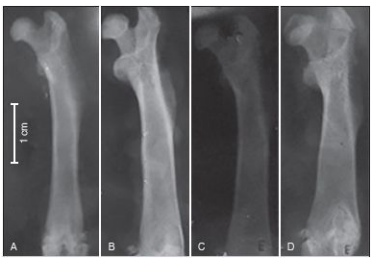
6- Is the cortex eroded (thinning)?

-The more aggressive the tumor is, the more it preserves the cortex.

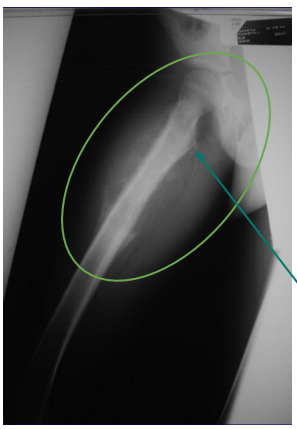


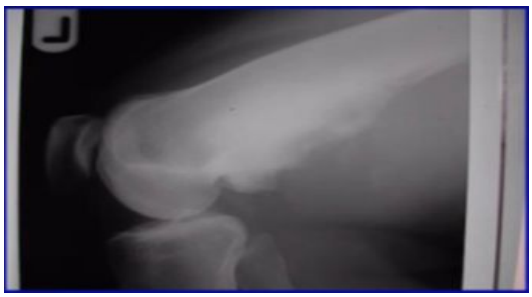
7-Is there bony remodeling?

-The normal bone try to contain lesion (and this usually happens in benign lesion).

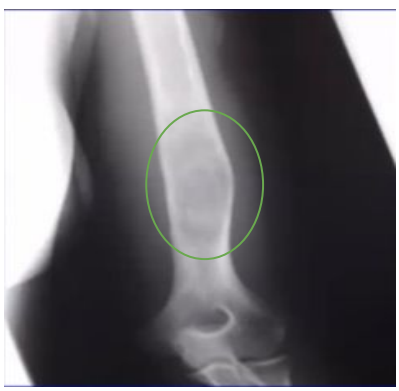
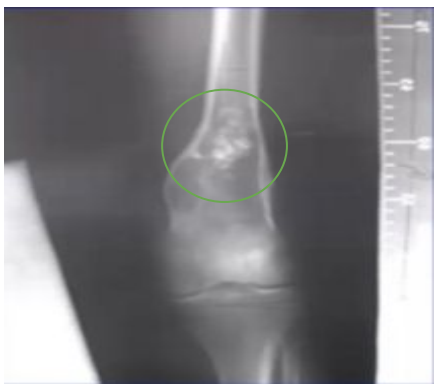
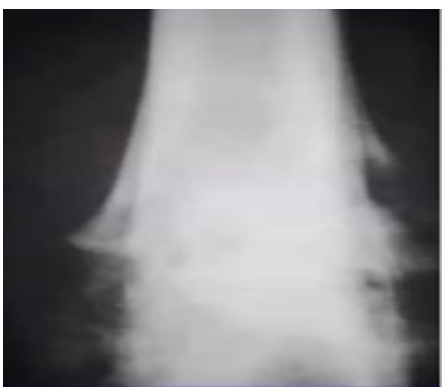


8- Is there a soft tissue mass?





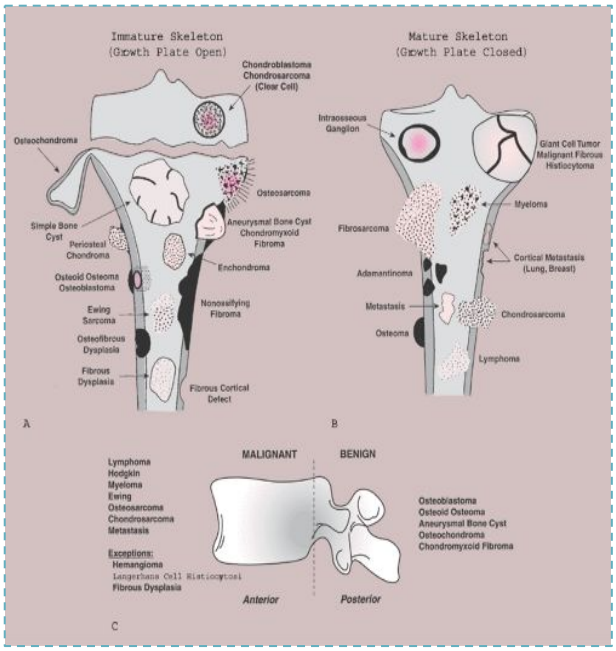
9- Is there any matrix?

	Lytic		Sclerotic
Types of matrix	**Fibrous**	**Cartilaginous**	**Osseous bone**
Picture			
Features	Fibrous: ground glass	Cartilage: popcorn like /arcs and rings appearance كأنه واحد راسم نقاط	Bone forming (Cloud-like)

X-Ray lesion description should include:

Number:

- Is the lesion solitary or are there multiple lesions? [1]



Site:

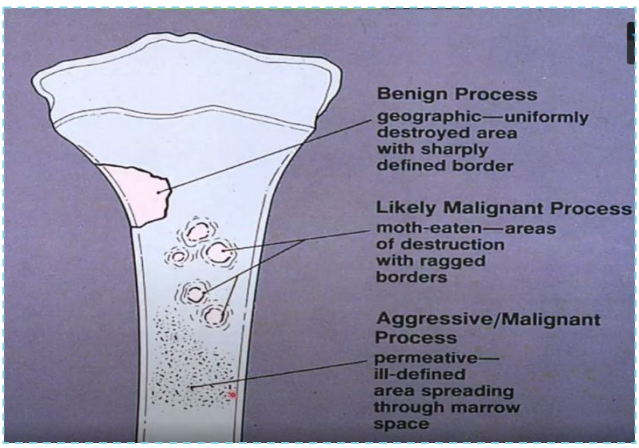
What bone is involved & where is the lesion in the bone?

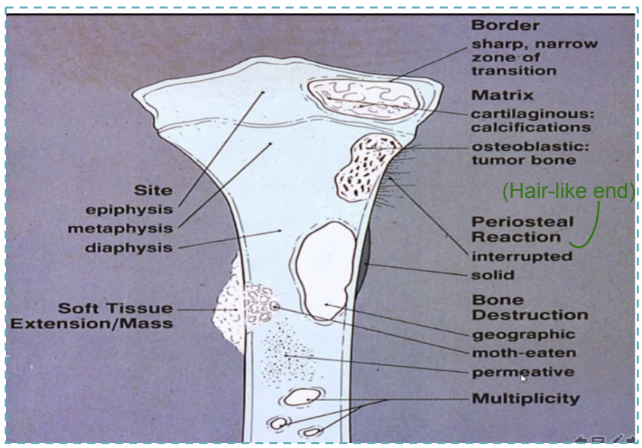
1. Epiphysis.
2. Metaphysis. (most common)
3. Diaphysis.
4. Centric (in the middle of the bone).
5. Eccentric (in the bone border).

Distribution of various lesions in a vertebra:
1- Malignant lesions are seen predominantly in its anterior part (body).
"Most likely" exceptions: hemangioma, Langerhans cells, fibrous dysplasia.
2- Benign lesions predominate in its posterior elements.

Type of bone destruction (morphology):

- Geographic (Organized uniformly destroyed area with sharply defined border): benign.
- Moth eaten كأنها ماكلتها العثة (areas of destruction with ragged/disorganized border): likely malignant.
- Permeative "همجية" (ill-defined areas spreading through bone marrow): aggressive/ malignant.





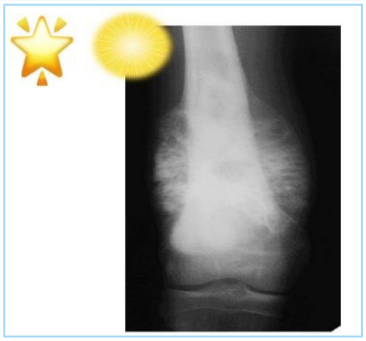
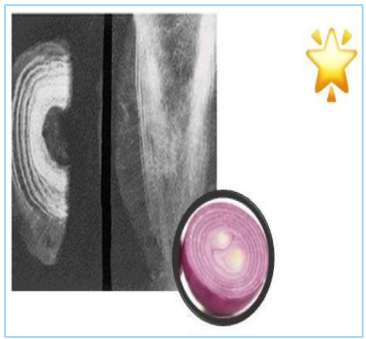
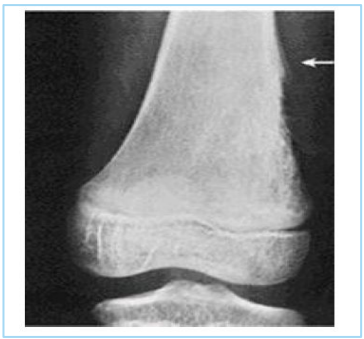
1- DDx for Multiplicity: metastases, myeloma, lymphoma, fibrous dysplasia, enchondromatosis.

Border or zone of transition:

- Well defined & sharp borders (either sharp sclerotic or sharp lytic), narrow zone of transition → benign (grow slowly > encapsulate by bone).
- Ill-defined borders, wide zone of transition → malignant tumors (fast, the bone didn't have the time to encapsulate it).

Periosteal reaction:

- ❖ Uninterrupted periosteal reaction usually indicates a benign process (solid clear buttress).
- ❖ Interrupted/large (teeth like) reaction indicates a malignant or aggressive nonmalignant process, and we have 3 types of these:

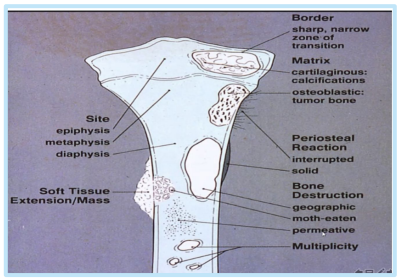
Sunburst pattern[1] in osteosarcoma and in Ewing Sarcoma	**Lamellated or onion-skin type in Ewing Sarcoma**	**Codman triangle (arrow) in Ewing Sarcoma and usually appears in Osteosarcoma**
		

Matrix of the tumor:

- **Sclerotic**: it means bone forming (osseous). So, on X-ray → opaque/white.
- **Lytic**: it means forming tissue other than bone "fluid" (it may be cartilage, fibrou tissue, or cystic), on X-ray → translucent/ black (e.g. popcorn calcification = chondroid).

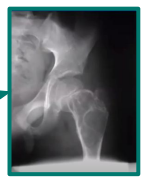
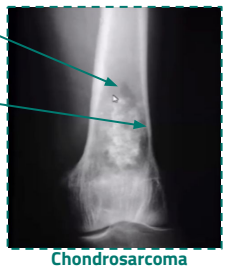
Soft tissue extension (swelling/ shadow):

- Mostly with malignant tumors (MRI is ideally used).



1-when the periosteum does not have enough time to lay down a new layer and instead the Sharpey's fibres stretch perpendicular to the periosteum.

The spectrum:

Types	characteristics
Benign latent[2] Watch. Follow up in 1 yr.	• Asymptomatic. (if there is pain it may be from trauma, not the tumor) • Well defined. • Narrow zone of transition (geographic). • No soft tissue mass. • No periosteal reaction. • No fracture. • May or may not have matrix. • Examples: enchondroma[1], non-ossifying fibroma
Benign Active[2] Follow up, images. Grows with child.	• **Symptomatic.** (pain not attributed to trauma) • Geographic. • Well-ordered periosteal reaction. • No soft tissue mass. • May or may not have matrix. • e.g: **osteoid osteoma**, UBC, eosinophilic granuloma, fibrous dysplasia, osteochondroma.  Unicameral bone cyst
Benign Aggressive -Destroying bone. -Jump on them! follow up .	• Symptomatic. • Geographic or permeative. • Usually lytic, cortical erosion. • May have soft tissue mass. • Periosteal neocorticalization. (It mean the tumour might go beyond of the cortex, eat the cortex and the bone try to build new cortex) • e.g: **GCT, ABC, osteoblastoma**, chondroblastoma, chondromyxoid fibroma, periosteal chondroma.
Low Grade Malignant	• Usually permeative. • May have matrix. • Cortical erosion. • May have soft tissue mass in continuity with cortical erosion. • Low-grade CSA[1], Adamantinoma (only in ant. tibia), Parosteal OSA, Chordoma (in the sacrum). • Image shows suspicious lytic lesions with popcorn like matrix.  Gas Cortical thinning Chondrosarcoma (CSA)
High Grade Malignant	• Permeative. • Usually has soft tissue mass. • Cortex usually intact. (tumor penetrates the cortex) or destroyed • Malignant periosteal reaction (onion skinning, sunburst, codman's triangle) • May or may not have matrix. • **Osteosarcoma, Ewing's sarcoma**, high grade chondrosarcoma, non-osteogenic spindle cell sarcoma (eg. MFH).

1-Enchondroma vs Chondrosarcoma: both will show popcorn pattern but CSA will have gas and cortical thinning.
2-Benign latent is distinguished from active by the presence of pain.

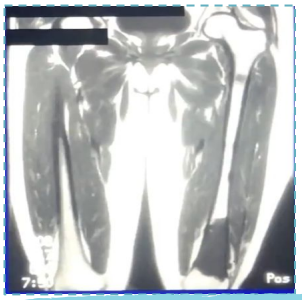
What's next? Staging:

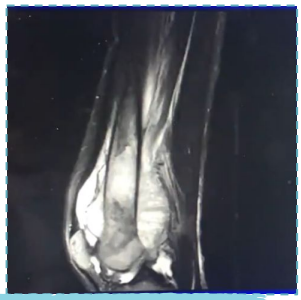
Staging:

- If lesion is **benign** latent, no further investigation is usually necessary, may follow up after 6m. (Benign active vs benign latent > pain)
- If **benign active or aggressive**, requires further local imaging, perhaps systemic staging.
- If **malignant**, requires further investigation including local and systemic staging.

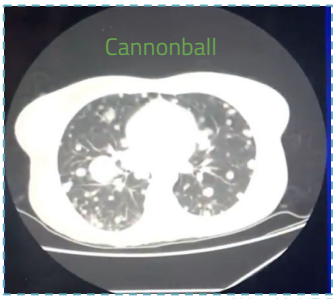
Local and systemic staging:

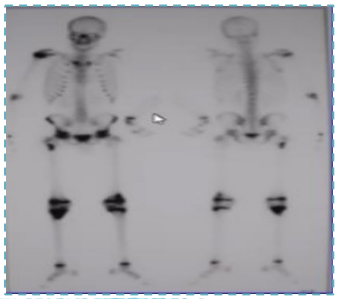
- Blood Work – CBC, ESR, CRP, serum calcium, **Alkaline phosphatase , LDH (latter 2 are prognostic in sarcoma)** (Ca, ALP and LDH indicate metabolic activity (bone turnover))
- **Local x-ray =** whole bone (Because some tumours have something called skip lesions (which is mass completely separated from the primary location of the tumour in the same bone)., chest x-ray (To rule out lung* metastasis)
- MRI of local site: must cover entire bone (full length MRI) may find lesions in proximal which is called skip lesions (occasionally CT = if there is soft tissue) (whats the **only** tumor that requires a full body MRI? **Myxoid liposarcoma**)
- CT chest to rule out metastasis.
- Total body bone scan for other lesions.
- For Ewing sarcoma we should order a gallium scan.
- ★ We should take a bone marrow aspiration for lymphoma, ewing sarcoma and MM.





Skip lesions (full length MRI)





Systemic staging

Metastasis?

>40 = metastasis | <40 = primary tumor

- Bloodwork – same bloodwork plus PSA prostate surface antigen, serum immunoelectrophoresis (in case of lymphoma and multiple myeloma for M band).
- CT chest and abdomen and pelvis to look for the source (lungs most commonly affected)
- Mammogram.
- Bone scan* other area are involved.

How to stage tumors?[1]

- Benign Latent/Active: Local - X ray +/- CT/MRI +/- TBBS = total body bone scan.
- Benign Aggressive: Local - X Ray/CT/MRI /Systemic - TBBS, CXR.
- Malignant: Systemic - CT Chest (<40) and bone scan , TBBS.
- Special: Gallium scan, CT Abd + Pelvis, Bone marrow biopsy.

1-Because lungs receive large venous supply which drains tumor .
2-PET Scan (how biologically active the tumor is).
-There is only one case in which we do total body MRI which is mexoid liposarcoma. The doctor told us it is coming in the exam but it did not :(

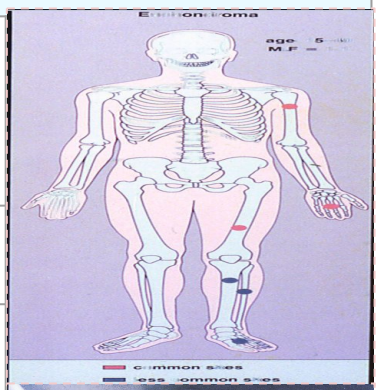
Primary Benign Bone Tumors:

Benign Latent Tumors

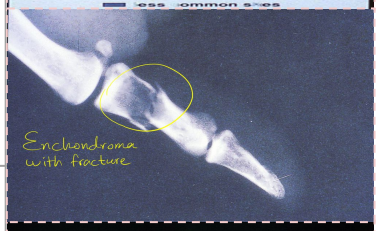
(1) Enchondroma

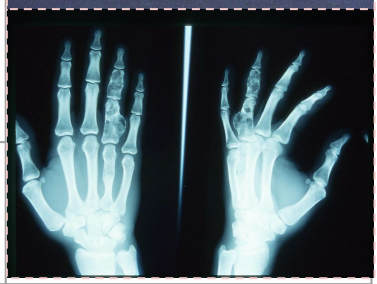
"441: ماني سائلكم فيها"

- Middle aged patients (15-50 age group).
- It is composed of translucent hyaline cartilage (chondroid).
- **Mostly found in the fingers.**



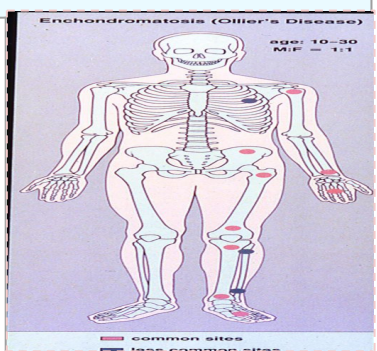
Site	Mainly small bone e.g. phalanges in hand & foot...etc.
Presentation	• **Asymptomatic**: Usually found incidentally. • Rings may become tight due to the swelling. • Pathological fracture which may cause pain, affect one side and prolonged healing.
Radiographic features	• **Popcorn matrix is characteristic for cartilaginous content.** • Benign features. • Can be seen on CT.
Treatment	• If symptomatic (pain, bulky, fracture) → Curettage + bone graft ± fixation.



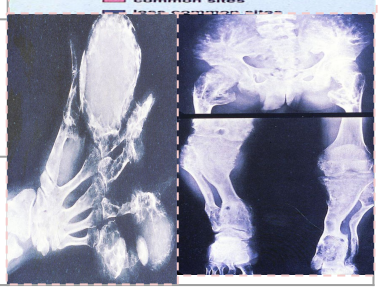


Enchondromatosis (Ollier's Disease)

- Multiple sites of enchondroma in the body most common sites are proximal humerus/tibia, which are premalignant.
- Autosomal recessive
- Benign, **affects both sides**. Usually seen in children. Rare.
- The difference between single enchondroma and multiple enchondromatosis is the high risk of malignant transformation (10-15% transforms into **chondrosarcoma,** which is chemo- and radio- resistant).



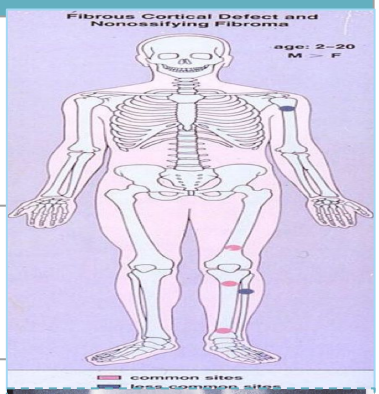
Presentation	• Not painful. • Very disabling deformity → restricts movement.
Treatment	• Only remove painful lesions.

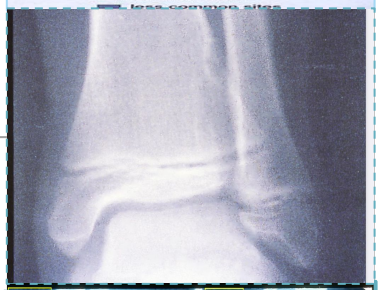


Primary Benign Bone Tumors:

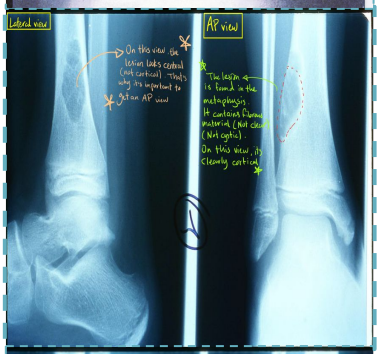
(2) Fibrous Cortical Defect and Non-ossifying Fibroma (NOF)

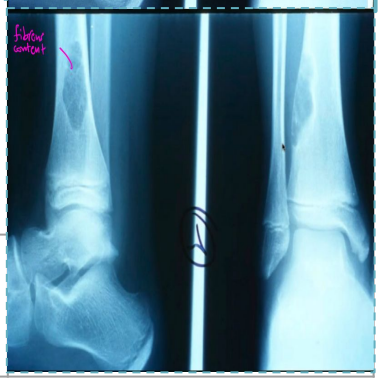
- A benign incidental finding, it is benign latent (never transfers to malignant).
- Found in children and it's usually an incidental finding.





Site	• Around the knee (lower femur and proximal tibia) • Lower tibia • May appear on proximal humerus
Presentation	• **Asymptomatic.**
Radiographic features	• On x-ray, always found on the cortex (eccentric). Contains fibers and not clear material (ground glass appearance). • **FIBROUS COMPONENT!!!** • Metaphyseal Mixed lesion (cystic + sclerotic components) • Lesion that appears to be 'central' is actually adjacent to or within the cortex, cortex intact, hence the alternative name 'fibrous cortical defect'. • Benign features: No periosteal reaction, No Soft tissue swelling. • Well-defined, sclerotic margin. X ray description: (bottom image) - X ray of a child, growth palate is seen. - Cloud shaped, defect in the metaphyseal area, geographic shape and fibrous material. - This x-ray shouldn't be worrisome and could be observed with no Tx.
Treatment	• **Self-limited (Reassurance).** • If large or fractured → curettage + bone graft.



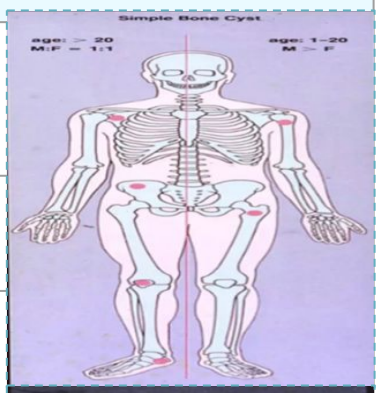


Primary Benign Bone Tumors:

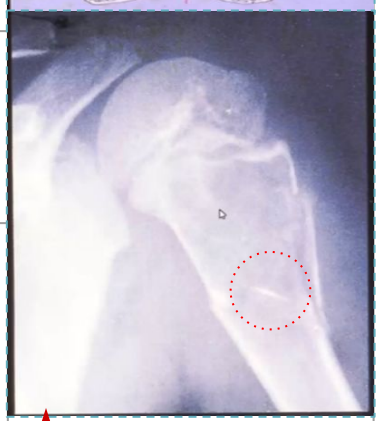
Benign Active Tumors

(1) Simple Cyst or Unicameral Bone Cyst (UBC)

- A common benign tumor, it is benign latent except if it ruptures.
- Usually seen in young patients.
- Less than 20 year olds.



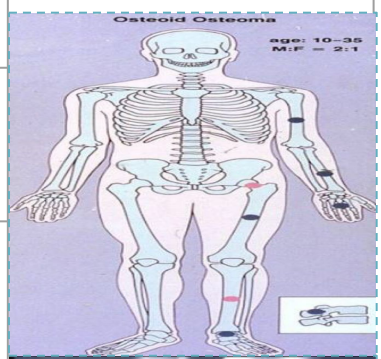
Site	• Pelvis, calcaneus, scapula and around the knee • Proximal part of long bones (e.g. proximal humerus, femur...)
Presentation	• Most commonly as incidental finding (asymptomatic) or pathological fracture.
Radiographic features	• **Metaphysic lytic lesion** (it contains fluid like suntop juice) • Well defined sharp border • No periosteal reaction • Fallen leaf sign indicates intra-cystic fracture (pathognomonic)
Treatment	• Observation unless there's a stress fracture • Often supportive, lesions will regress following skeletal maturity • Curettage and bone grafting may be required in areas at risk of fracture



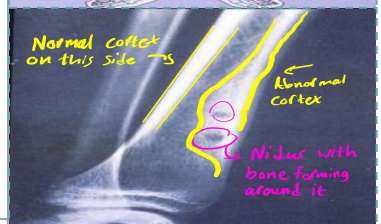
★ Fallen Leaf Sign

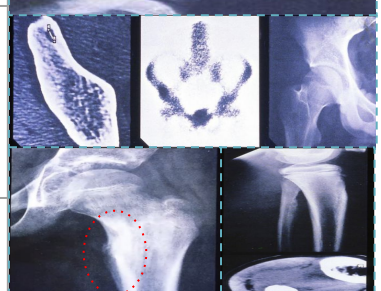
(2) Osteoid Osteoma (*night pain that improves with NSAIDs* احفظوها بيجي عنها سؤال)

- Bone-forming tumor



Site	• Found in the diaphysis and neck of the femur, tibia, humerus and posterior element of the spine • May arise in the cortex of long bones and rarely in the talus
Presentation	• Main presentation with **pain.** • Well localized pain (in the back or groin area) • Worse at night and prevents patient from sleep. ➔ Because of prostaglandin release. • Responds well to NSAIDs, leading to complete resolution • Painful scoliosis if affecting the spine. While idiopathic scoliosis is painless. • Nothing on physical exam.
Radiographic features	• Lytic lesion (central nidus), surrounded by a reactive zone of dense sclerotic new bone formation • Fine cut CT scan → nidus (modality of choice) • Cortical thickening on X ray
Treatment	• NSAIDs challenge (aspirin) • **Radiofrequency ablation**

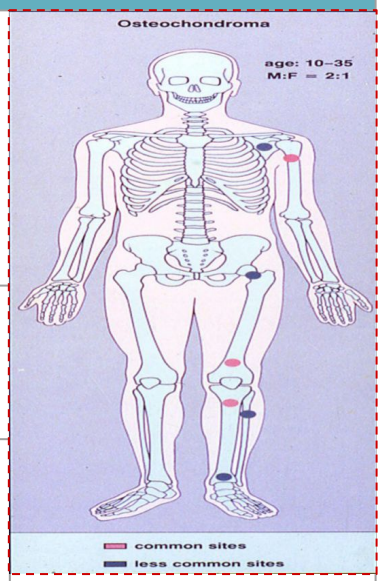




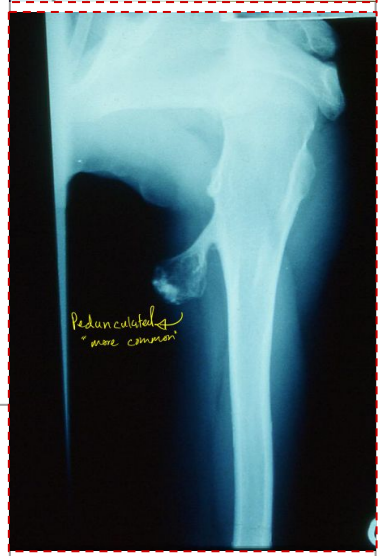
Primary Benign Bone Tumors:

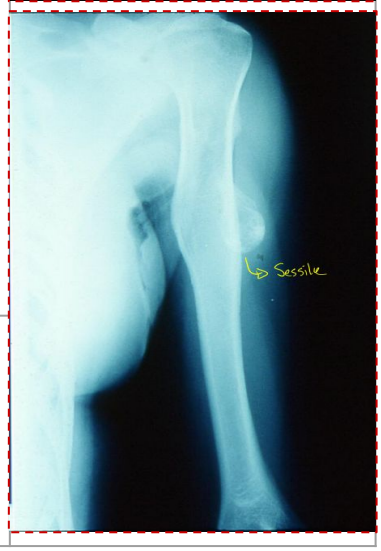
★ (3) Osteochondroma

- Exostosis
- The only surface tumor (outside the bone)
- It is a combination of both bone & cartilage.
- Starts from the growth plate and Stops growing when patient stops to grow usually at 18 yrs
- > 2 cm masses have a risk of malignant transformation



Site	• The commonest are around the knee (**distal femur** & proximal tibia) • Proximal humerus, scapula and neck of femur
Presentation	• Patients usually present for cosmetic purposes or pain • Painless progressive swelling • Painful in children due to growth plate compression and pressure effects • On adjacent nerve or vascular structures (obese patients won't notice it) Complications: 1. Pressure symptoms: - Pseudoaneurysm → artery - Hypoesthesia/ paresthesia → nerve - Limited ROM → tendon - Formation of an overlying bursa due to friction 2. Fractures especially in the pedunculated type
Radiographic features	• Exostosis (fungated): mushroom-like stalk of the bony tumor (connected to the bone) • Benign features. • Metaphyseal lesion. • MRI: cartilaginous cap • **A distinctive feature is the continuity of the medullary canal content with tumor** • According to the shape of the neck, we divide it into: A. **Pedunculated type** (more common): long & thin neck. Directed away from the bone. B. **Sessile type**: has short & thick neck.
Treatment	• We usually resect the lesion even if it's benign • Surgery is indicated for: **(NEVER for cosmetic reasons)** 1. If the lesion is obstructing a nerve, vessel or tendon. 2. If the lesion limits movement. 3. If the lesion is quickly increasing in size



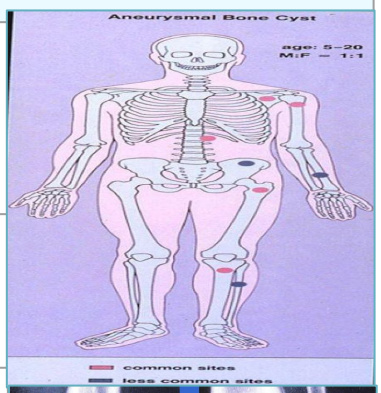


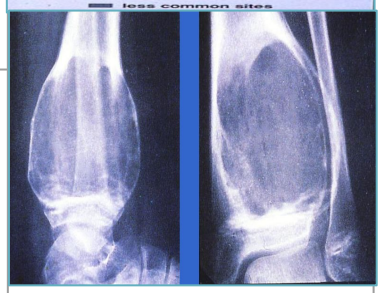
Primary Benign Bone Tumors:

Benign Aggressive Tumors

(1) Aneurysmal Bone Cyst (ABC)

- They can recur and may transfer to osteosarcoma
- Blood-filled cystic spaces, bigger than simple cysts
- Simple cyst is not wider than the growth plate and has fallen leaf sign, aneurysm bone cyst is wider than growth plate



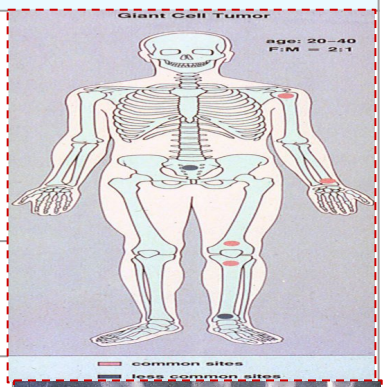


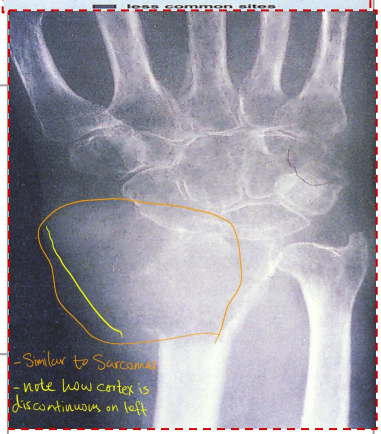
Site	• Around joints: upper humerus, femur, and tibia • Spine (neural arch: lamina and pedicle) • Flat bones: scapula and pelvis
Presentation	• Asymptomatic
Radiographic features	• **Balloon like lesion** no periosteal and soft tissue swelling • Metaphyseal, septated lytic lesion • Well defined and sclerotic border • Expansile with thinned cortex (egg-shell) → • MRI shows fluid fluid level

(2) Giant Cell Tumor (GCT) very high recurrence rate (18%)

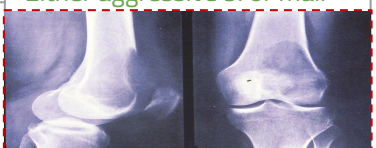
مهمة جدا:441

- Benign but very aggressive tumor (most aggressive benign one)
- The only benign that can metastasize (to the lung → get chest x-ray)
- GCTs can occasionally be seen with Paget's disease of bone and can arise in association with focal dermal hypoplasia (Goltz syndrome).
- In middle age group, In 20-40 years





AP x-ray with a lesion on metaphysis or epiphysis w soft tissue involvement, cortical destruction
Either aggressive b. or mal.



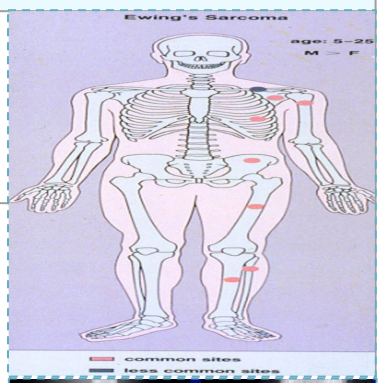
Site	• Proximal humerus, distal femur, proximal tibia. • Most common: distal radial Metaphyseal-juxta articular.
Presentation	• Presents with: pain, swelling & later pathological fracture
Radiographic features	• **Aggressive features:** - Permeative destruction - Ill defined borders - Huge soft tissue component • No new bone formation because this tumor is made up of osteoclasts (little or no periosteal reaction)
Investigation	• Soft tissue extension → MRI • Joint extension: the articular surface prevents extension • Bone extension → CT scan • Bone scan → metastasis • **Biopsy** → rule out malignant transformation
Treatment	• No place for conservative treatment. • Excision (Curettage) followed by either bone graft or prosthesis depending on the site of the tumor.

Primary Malignant Bone Tumors:

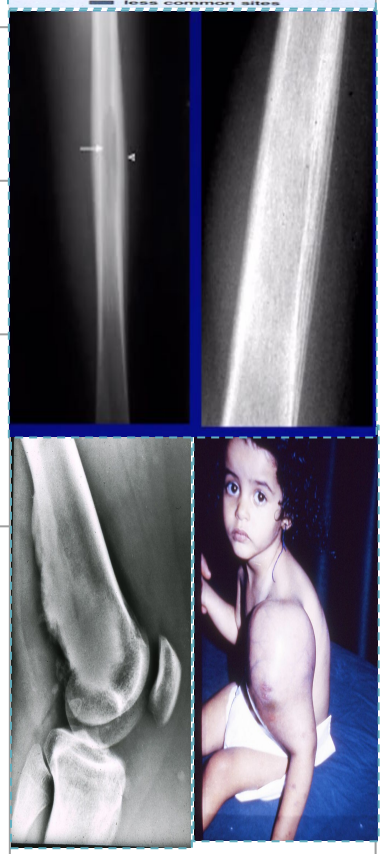
Malignant Tumors

(1) Ewing Sarcoma

- **One** of the most common tumors in **children (3-5y)**
- Most Ewing's sarcomas are misdiagnosed as acute osteomyelitis because of systemic symptoms, elevated ESR, elevated CRP, high WBCs. Even with biopsy some **pus** comes out



Site	• Around the growth plate → femur (most common), tibia, and humerus. • It is the only bone tumor which takes origin from **diaphysis,** Iliac flat bone, and ribs.
Presentation	• Pain • Huge Swelling • Lump with constitutional symptoms
Radiographic features	• Onion-skin periosteal reaction. • Very significant soft tissue component (characteristic of Ewing sarcoma)
Investigation	• MRI and biopsy make definitive diagnosis • N.B. you can't differentiate from osteosarcoma unless in biopsy. But hints are age group, radiological location (diaphysis in Ewing and metaphysis in osteosarcoma)
Treatment	For both Ewing and osteosarcoma: • Neoadjuvant chemotherapy followed by tumor excision and reconstruction. Why do we give neoadjuvant chemotherapy prior to surgical resection? 1. **To shrink the tumor for easier removal** 2. **To kill micrometastasis** 3. **Hints on post surgical chemotherapeutic agent**

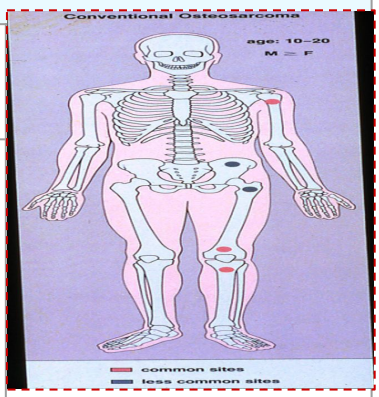


Primary Malignant Bone Tumors:

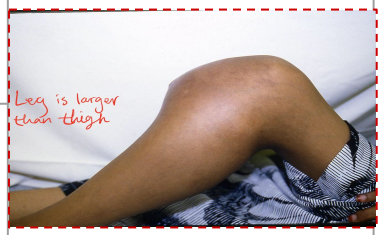
Malignant Tumors

(2) Osteosarcoma

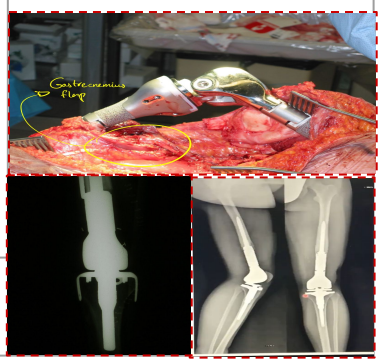
- More common than Ewing's sarcoma. **(age 10-20)**
- **History of radiation and paget's disease.**



Types	1. Primary (conventional, low-grade central, telangiectatic, multicentric (multifocal), juxtacortical; no need to know types of primary sarcoma) 2. Secondary (malignant transformation of benign process): A. Paget's disease → Paget's sarcoma **B. Post radiation sarcoma** 3. Metastatic from breast, lungs or bones.
Site	• Around the growth plate → around the shoulder, knee. • Distal femur, proximal tibia/Humerus
Presentation	• Pain, pathological fracture (typical presentation of malignant tumor) • Swelling (palpable mass in the site of the tumor "metaphysis commonly" but not as huge as Ewing's) • There might be prominent veins overlying the skin over the swelling • Overlying skin is warm due to high vascularity • The patient may look cachectic (in late and advanced cases)
Radiographic features	• Codman triangle • Irregular medullary and cortical destruction of metaphysis • Cortex is intact
Investigation	• CT scan → bone extension, lung metastasis. • MRI → soft tissue extension, neurovascular bundle involvement • Bone scan → metastasis to other bones *characteristic feature*. • Fever, elevated alkaline phosphatase (ALP) and lactate dehydrogenase (LDH) • Definite diagnosis made by **biopsy**
Treatment	• Similar to Ewing's Sarcoma



Bone producing, cortex is intact

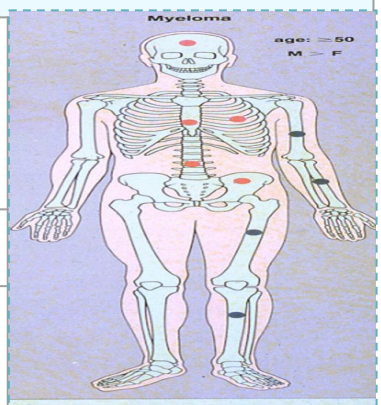


Primary Malignant Bone Tumors:

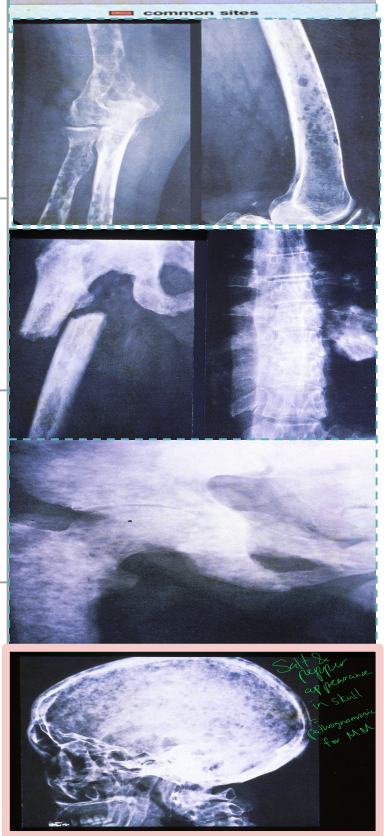
Malignant Tumors

(3) Multiple Myeloma

- Systemic disease.
- Arise from plasma cells in the bone marrow.
- The most common primary malignant tumor in elderly > 50.
- Some consider it a bone tumor, and some consider it hematological



Site	• Central bones (axial skeleton): skull, ribs, pelvic girdle. & spine
Presentation	• Pain, spinal cord compression, Osteoporosis, Pathological fractures. • Medical complications include anaemia, hypercalcaemia, hyperviscosity, • Immunosuppression and renal dysfunction. • CRAB: C = Calcium (elevated), R = Renal failure, A = Anemia, B = Bone lesions (bone pain)
Radiographic features	• Radiological features: Multiple Lytic or sclerotic lesions. "Moth-eaten appearance" • In skull, there will be pepper (lytic) & salt (sclerotic) appearance (pepper-pot). You have to do skull x-ray.
Investigation	• Bence jones proteins test found in 24h urine collection (highly suggestive)(protein electrophoresis) (urine & serum electrophoresis) UPEP and SPEP • Only definitive diagnosis is by bone marrow aspiration
Treatment	• Radiotherapy, Chemotherapy (mainly medical) • Bone marrow transplant: Success rate is 30% and it costs millions • The role of orthopedic surgeon is only fix pathological fractures, do internal fixation.

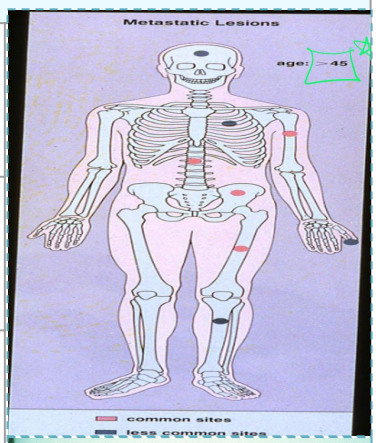


Secondary Metastatic Bone Tumors:

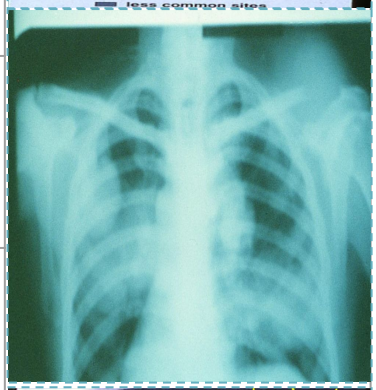
Malignant Tumors

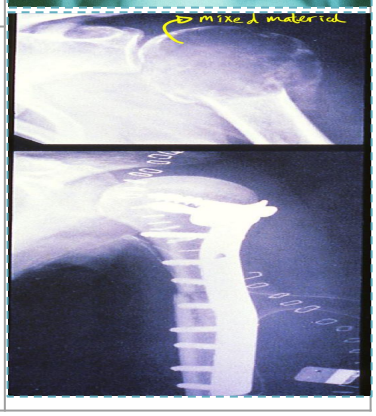
Metastasis

- More common than all primary tumors. Common in adults (>45 y)
- The most common tumors are: thyroid, lung, breast, colorectal, prostate and kidney



Site	• Usually metastasis occurs in the **highly vascular bones** e.g. vertebral body, hip, ribs, pelvis, upper end of femur, and humerus
Presentation	• Patient may present with known primary tumor. • May present with secondary metastasis. So, we must identify the primary sit and treat it
Radiographic features	• X-ray shows lytic lesions (bone eaten away) with thinning of the cortex. • Resembles bone cysts but the **age group** directs the diagnosis
Investigation	• CT scan of the chest. • Take biopsy to know where is the primary. • You can do bone scan to check metastasis in other areas
Treatment	• Palliative chemotherapy (not surgical). • The new trend is to downgrade the tumor then allograft it Surgery indications: ❖ Severe pain ❖ Mechanical instability: pain in the spine with neurological symptoms (spinal cord compression) ❖ Prophylactic (preventive fixation): in weight bearing bones with involvement of >60% of the cortex ❖ Fixate a fracture (curettage, grafts and plates)



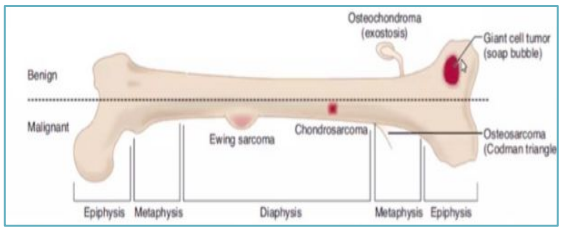


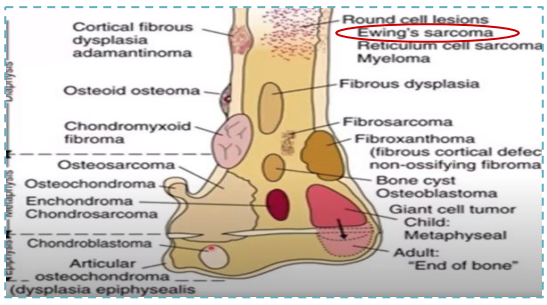
Important locations

Tumors usually have a common site (not necessary)

Common sites of tumor in bones:

- **Chondroblastoma**: epiphysis
- **Giant cell tumo**r: cross metaphysis and epiphysis
- **Osteosarcoma**: cross **metaphysis** and epiphysis
- **Ewing sarcoma**: **diaphysis**
- **Osteoid osteoma**: cortex





What's Next? Biopsy.

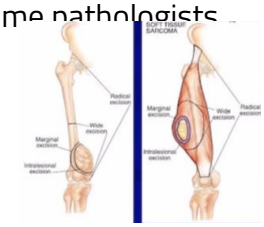
Biopsies

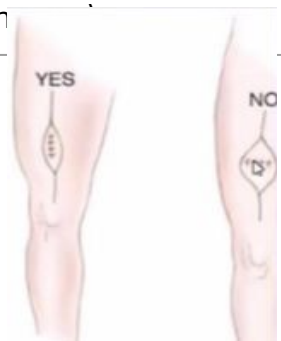
- Is not a substitute for thorough history, physical examination and investigations.
- Serves to confirm diagnosis suspected from above
- If you don't know what it is before the biopsy you won't know what it is after.

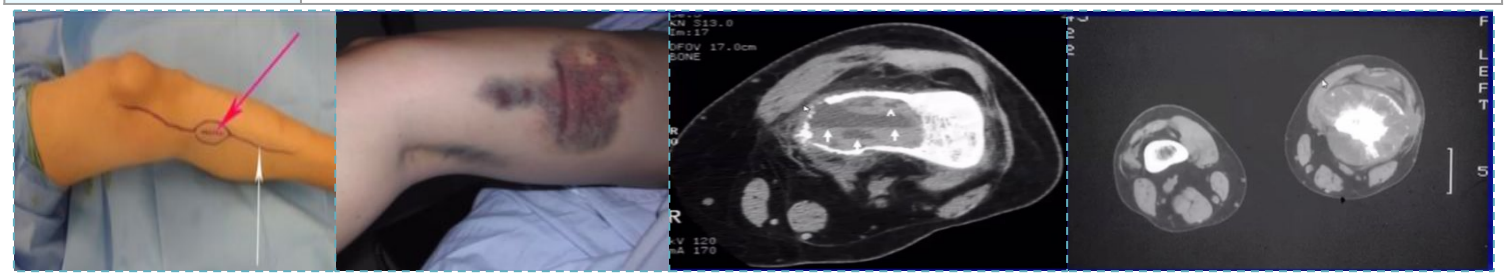
Not every lesion needs a biopsy!

- An asymptomatic (latent) or symptomatic bone lesion (active) that appears entirely benign on imaging doesn't need a biopsy
- A soft tissue lesion that appears entirely benign on MRI (lipoma, hemangioma) does not need a biopsy
- When in Doubt, it is safer to do a biopsy

Indications for Biopsy **V. Important**	• Aggressive or malignant appearing bone or soft tissue lesions. • For soft tissue lesions > 5 cm, deep to fascia, bone or neurovascular structures. • Unclear diagnosis in symptomatic patient • Special situation - solitary bone lesion in a patient with a history of carcinoma
Prerequisites for Biopsy	• CBC, platelets, coagulation screen • Cross sectional imaging- depicts local anatomy , solid areas of tumor • Experienced musculoskeletal pathologist is available.
Biopsy Technique	• Fine needle aspirate - gives cytologic specimen (adequate for some pathologists experienced with this technique) • Core biopsy (tru-cut) allows for ultrastructural examination • Incisional biopsy= open biopsy (go in take small piece and go) • Excisional biopsy (remove whole mass) when? if superficial to fascia and less than 5 cm. • Selected indications (small < 5c ,superficial soft tissue m
Principles of Open Biopsy	• Extensile incision – longitudinal in extremities. • Avoid developing planes (cut only one time) • Use involved compartment • Do not expose neurovascular structures. • Meticulous hemostasis • Release tourniquet prior to wound closure. • If using drain, bring out in line with incision.
General Recommendations Not important for you	• For benign aggressive tumours without soft tissue mass, plan biopsy through area of maximal cortical weakening based on CT or MRI. • For malignant tumours or benign aggressive with soft tissue mass, biopsy soft tissue rather than creating hole in bone.







Quiz

Q1: A 13-year old boy comes to the clinic because of a 6-week history of pain in his right shoulder. His pain is aggravated by the recent start of football practice and associated increased activity, but it persists after practice as well. He has no prior history of trauma. He denies fevers, weight loss and night sweats. His temperature is 37.1°C, pulse is 70/min, respirations are 16/min, and blood pressure is 120/82 mm Hg. Physical examination shows a hard immobile mass on the right proximal humerus which is slightly tender. His range of motion in the shoulder is minimally restricted. A radiograph obtained of his right upper extremity shows a sunburst pattern. Which of the following is the most likely diagnosis?

- **A** Ewing Sarcoma
- **B** Osteosarcoma
- **C** Giant cell tumor
- **D** Metastasis

Q2: A 15-year-old comes to his pediatrician's office because of pain in his right humerus for the past 3 months. He also reports a recent low grade fever. Further examination reveals midshaft swelling on his right humerus. The patient denies any recent history of trauma. Genetic analysis shows an 11;22 translocation. An x-ray of his right arm is taken. Which of the following answer choices will most likely show up on imaging for this patient?

- **A** Onion-skinned appearance
- **B** Punched-out lytic lesion
- **C** Necrosis surrounded by sclerotic bone
- **D** Central nidus, hyperactive cortex

Q3: A 10-year-old boy is brought to his pediatrician's office by his parents because of worsening pain over his left lower leg for the past 2 months. He denies any recent trauma to his leg within the past year and has been generally healthy. Physical examination shows a discrete, hard, and exquisitely tender mass over the anterior aspect of the right tibia about 3 inches below the knee. ESR today is 20 mm/hr. An X-ray of this patient's left leg is shown . What is the most likely diagnosis?

- **A** Osteosarcoma
- **B** Ewing Sarcoma
- **C** Giant cell tumor
- **D** Osteoid osteoma

Q4: Q4: A 17 year old with swelling and pain at the right knee for the last 6 months, lost 7 kg, no history of trauma or infection, blood work done and was normal besides high alkaline phosphatase. X-ray was done which showed highly suspicious tumor most likely to be an osteosarcoma. What is the confirmatory diagnosis test?

- **A** Knee MRI
- **B** Knee CT scan
- **C** Bone scan
- **D** Biopsy

1:B 2:C 3:A 4:D 5:C

Quiz

Q5: A 12-year-old boy comes to the clinic with his mother because of severe pain in his right leg for the past month. He says that the pain is mostly located in his thigh and that it was initially mild but now the pain is so bad that he cannot walk on his leg. His mother says that aspirin relieves his pain temporarily. Physical examination shows some localized pain over the area of the proximal femur. There is no palpable joint deformity, redness or swelling. He also walks with a visible limp. Neither increasing nor decreasing activity alleviates the pain, which tends to be worse at night. He has not had any recent trauma to the leg. An MRI of the leg shows a 1 cm lesion within the femur (shown below). Which of the following is the most likely explanation for this patient's symptoms?

- A: Aneurysmal bone cyst
- B: Non-ossifying fibroma
- C: Osteochondr-oma
- D: Osteoid osteoma

Q6: A 23-year-old man comes to the emergency department because of a 10-month history of gradual painful swelling of his wrist. His temperature is 37.9°C (100.2°F), pulse is 82/min, respirations are 16/min, and blood pressure is 120/77 mm Hg. An X-ray is obtained and is shown below. Which of the following is the most likely diagnosis?

- A: Osteoid osteoma
- B: Osteosarcoma
- C: Ewing sarcoma
- D: Giant cell tumor

Q7: Which of the following is the most aggressively benign tumor?

- A: Osteoid osteoma
- B: Endochondr-oma
- C: Giant cell tumor
- D: Simple bone cyst

SAQs

List three bone tumors that require bone marrow aspiration

1- Multiple Myeloma
2- Ewing Sarcoma
3- Lymphoma

What are 3 prerequisites to biopsy a highly suspicious bone tumor?

1- CBC, platelets, coagulation screen
2- Cross sectional imaging to depict local anatomy and solid areas of the tumor
3- Experienced musculoskeletal pathologist available

MCQ	
Q1	A
Q2	A
Q3	B
Q4	D
Q5	D
Q6	D
Q7	C

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