14- MSK Tumors

Objectives:

- To be able to specify the symptoms and signs; outline the assessment and appropriate investigation; propose a limited differential diagnosis and; outline the principles of management of a patient with:
  - **Metastatic bone disease.**
  - **Primary bone lesions.**
  - **Benign tumors:**
    - Osteoid osteoma.
    - Bone Cyst:
      - Unicameral bone cyst (UBC).
      - Aneurysmal bone cyst (ABC).
      - Giant-cell tumor (GCT).
    - Osteochondroma.
  - **Malignant tumors:**
    - Osteosarcoma.
    - Ewing's sarcoma.

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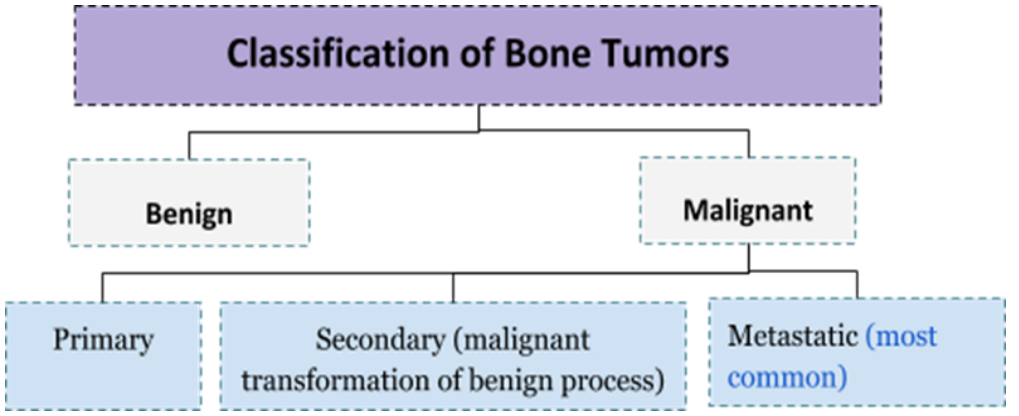
References: 435 team, Doctors' notes, 436 slides, Tornto notes



Kaplan notes high yield osmosis notes Toronto notes

Most are classified according to the **normal cell of origin** and apparent pattern of differentiation لا تتعبون نفسكم فيها كلها بالنهاية ترجع لأساسها اللي هو بيناين ومماليقنانت.

Histologic Type	Benign	Malignant
Hematopoietic (40%)		**Myeloma** **Malignant lymphoma**
Chondrogenic (22%) "cell of origin: cartilage"	**Osteochondroma** **Chondroma** Chondroblastoma Chondromyxoid fibroma	**Chondrosarcoma** **Dedifferentiated chondrosarcoma** **Mesenchymal chondrosarcoma**
Osteogenic (19%) "cell of origin: bone"	**Osteoid osteoma** **Osteoblastoma**	**Osteosarcoma**
Unknown origin (10%) classified as benign or malignant based on cell pathology, but cell of origin is unknown	**Giant cell tumor**	**Ewing tumor** **Giant cell tumor** **Adamantinoma**
Histiocytic origin	Fibrous histiocytoma	Malignant fibrous histiocytoma
Fibrogenic	Metaphyseal fibrous defect (fibroma)	Desmoplastic fibroma Fibrosarcoma
Notochordal		Chordoma
Vascular	Hemangioma	Hemangioendothelioma Hemangiopericytoma
Lipogenic	Lipoma	Liposarcoma
Neurogenic	Neurilemmoma	

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

★ History:

- Age, sex.
- Present with:
- Pain, especially night pain
- **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.

★ Physical Examination:

If there is a lump: 2Ts = tenderness, temperature, **3Ss** = Site, size & shape

★ Investigations:

- Lab: **CBC** (infection or anemia), **ESR**, Hematological investigations.
- Imaging: X-ray, MRI, CT, Bone scan قبل نستخدمها الحين استبدلناها ب **PET 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.

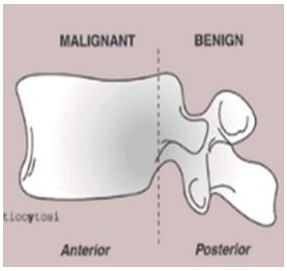
التيومرز شعوبًا وقبائل كل واحد يسكن المكان المعروف له لكن عادي نحصّل جنوبي ساكن بالشمال!

X-Ray Reading describing a lesion should include the following (VERY IMPORTANT):

1- Number: is the lesion solitary or are there multiple lesions?[1]

2- Site: What bone is involved & where is the lesion in the bone?

- Epiphysis, Metaphysis (most common) or Diaphysis
- Centric (in the middle of the bone) or eccentric (in the bone border).



Distribution of various lesions in a **vertebra**:

- Malignant lesions are seen predominantly in its anterior part (body).

"Most likely" exceptions: hemangioma, Langerhans cells, fibrous dysplasia.

- Benign lesions predominate in its posterior elements.

3- Types of bone destruction (morphology: most important):

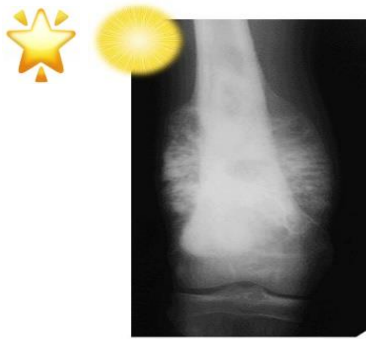
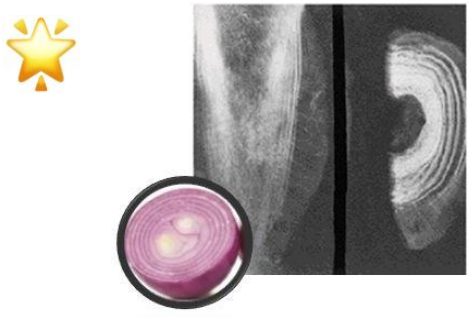
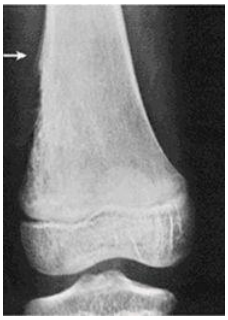
- Geographic (Organized uniformly destroyed area with sharply defined border): benign.
- Mouth eaten (areas of destruction with ragged/disorganized border): likely malignant.
- Permeative **"همجية"** (ill-defined areas spreading through bone marrow): aggressive/malignant process.

4- Border of the tumor 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).

5- 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:

Sunburst pattern[2] in osteosarcoma	lamellated or onion-skin type in Ewing sarcoma	Codman triangle (arrow) in Ewing sarcoma & osteosarcoma
		

6- Matrix of the tumor:

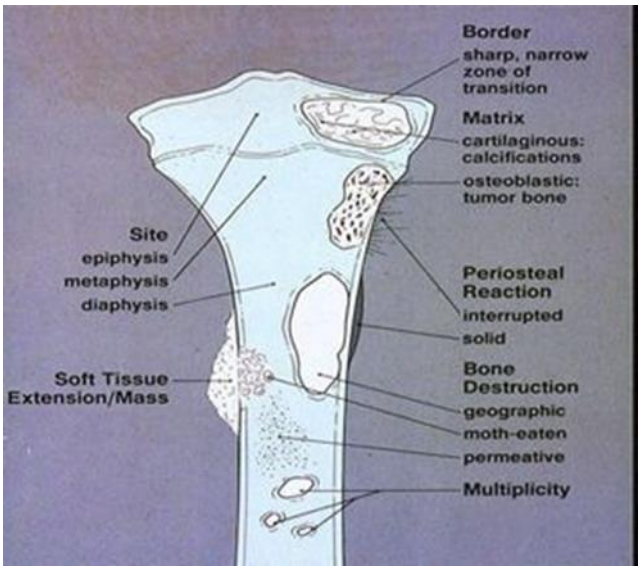
- Sclerotic: it means bone forming (osseous). So, on x-ray →opaque/white.
- Lytic: it means forming tissue other than bone "fluid" (it may be cartilage, fibrous tissue, or cystic), on X-ray →translucent/ black (e.g. popcorn calcification = chondroid)

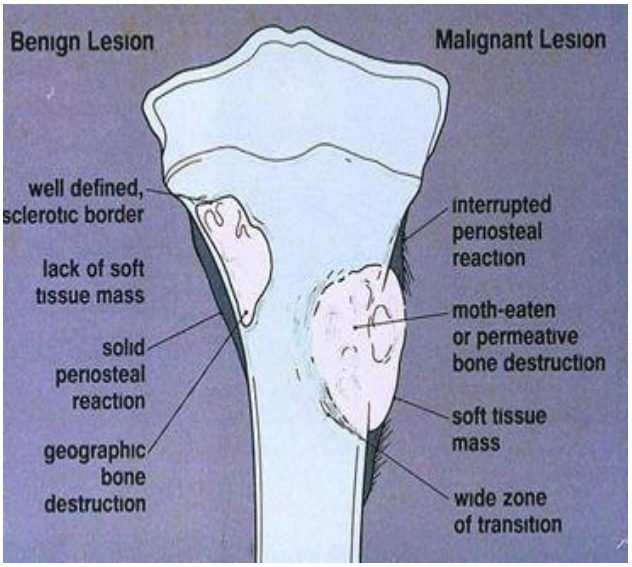
[1] DDx for Multiplicity: metastases, myeloma, lymphoma, fibrous dysplasia, enchondromatosis

[2] 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

7- Soft tissue extension (swelling/shadow):
Mostly with malignant tumors (MRI is ideally used)

All you need to know is to recognize bone lesions in x-ray and classify them as benign or malignant.

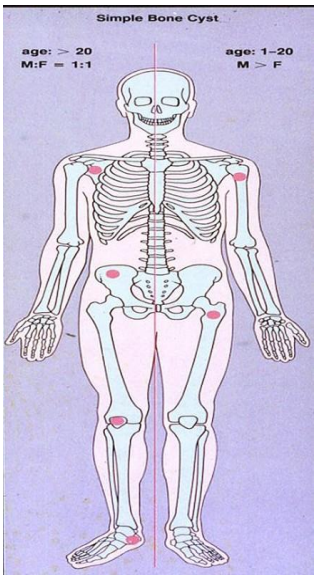




1- Primary Benign bone tumors

Sketchy benign bone tumor

1) Simple bone cyst (unicameral cyst): (UBC)



Usually seen in young patients
#Presentation:
Most commonly as **incidental finding** (asymptomatic) or **pathological fracture**.
#Sites:

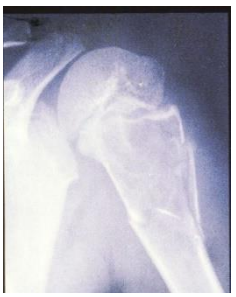
- Pelvis, Calcaneus, Scapula, around the knee (lower limp more)
- Proximal part of long bones (e.g. proximal humerus, femur or tibia).

#Radiological features:

1. **Metaphysic** lytic lesion.
2. Well defined sharp border.
3. No periosteal reaction.

#Treatment:

- Treatment is often **supportive**. lesions will regress following skeletal maturity.
- **Curettage** and bone **grafting** may be required in areas at risk of fracture. Never resection
- **Fracture:** correction of fracture will heal the cyst[3]

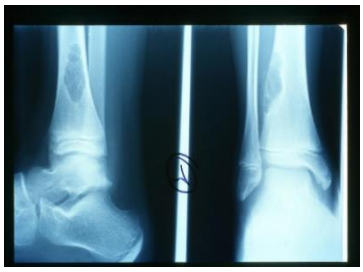


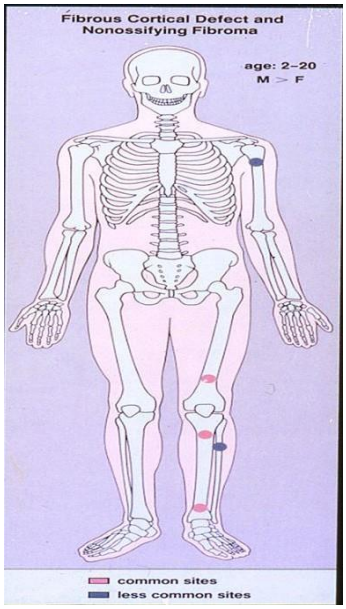
2) Fibrous Cortical Defect and Non-Ossifying Fibroma:

The commonest benign lesion of bone. Never transfer to malignant.
- In children.

[3] Pathological fracture in young age group: cast the fracture & patient education regarding recurrence of fracture in this area. The same fracture in older age with displacement & angulation: ORIF + Bone graft.

Eccentric جاية على الكورتكس نفسها





#Sites:

- Around knee: (lower femur, upper tibia).
- Lower tibia.

#Presentation: Asymptomatic

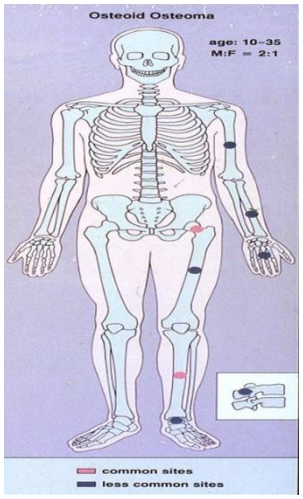
#Radiological features:

- Metaphyseal **Mixed** lesion (cystic + sclerotic components)[4]
- Lesion that appears to be 'central' is actually **adjacent to or within the cortex**, hence the alternative name 'fibrous cortical defect'.
- Benign features: No periosteal reaction, No Soft tissue swelling, Well-defined, sclerotic margin.

#Treatment:

- Self-limited. Reassurance.
- If large or fractured →curettage + bone graft.

3) Osteoid osteoma: الورم العظمي العظماني , Nothing on physical exam



#Site:

May arise in the cortex of **long bones**, or occasionally in the cancellous bone of the **spine** and less commonly talus.

#Presentation:

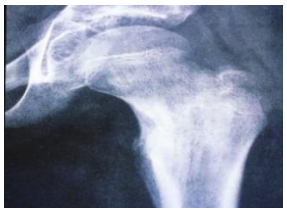
Main presentation is **pain**.

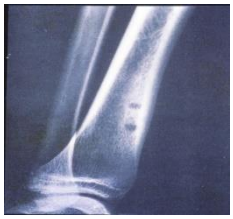
- Well localized pain (in the back or groin area)
- Worse at night and prevents patient from sleep.
- Responds well to NSAIDs, leading to complete resolution.

Painful scoliosis if affecting the spine. While idiopathic scoliosis is painless

#Radiological features:

- Lytic lesion (central nidus), surrounded by a reactive zone of dense sclerotic new bone formation.
- **CT scan** →nidus (modality of choice in the diagnosis)



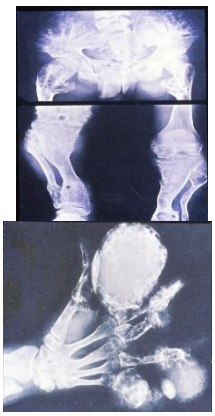


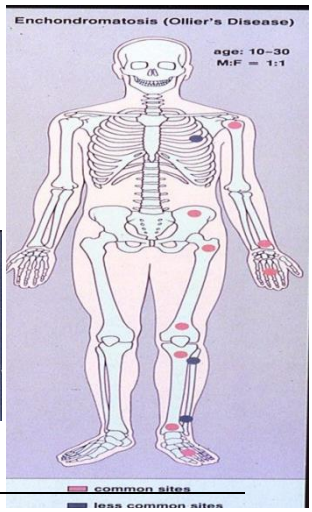
#Treatment:

Without treatment, the lesion will slowly increase but **over time will regress.**and usually burns out over a variable number of years.

- NSAIDs challange "aspirin challenge"

4) Multiple Enchondromatosis (ollier's disease):





Autosomal recessive[5].

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**).

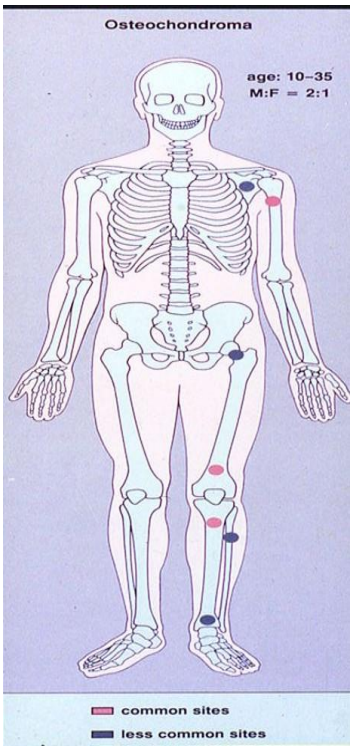
#Presentation:

- Not painful
- Very disabling deformity →restricts movement.

[4] Although it looks cystic on X-rays, it is a solid lesion consisting of unremarkable fibrous tissue with a few scattered giant cells.

[5] Enchondromatosis encompasses several different subtypes of which Ollier disease and Maffucci syndrome are most common. Most subtypes are non-hereditary, while some are autosomal dominant or recessive

5) Osteochondroma (exostosis) زيادة عظمية



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.

#Sites:

- The commonest are around the knee (distal femur & proximal tibia)
- proximal humerus, Scapula and neck of femur.

#Presentation:

- Painless progressive Swelling. Patients usually present for cosmetic purposes.
- Painful in children due to growth plate compression and pressure effects on adjacent nerve or vascular structures.
- Symptoms of complications:
  1. Pressure symptom:
     - Pseudoaneurysm →artery.
     - Hypoesthesia or paresthesia →nerve.
     - Rendering the movement →tendon.
     - Restrict the movement of the nearby joint
     - formation of an overlying bursa due to friction.
  2. Fracture especially with pedunculated type If it transfers to malignant.

#Radiological features:

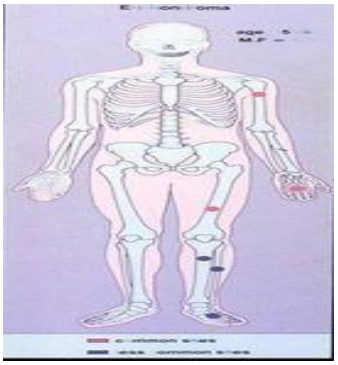
1. Exostosis[6] (fungated). Mushroom-like stalk of the bony tumor (connected to the bone).
2. Benign features.
3. Metaphyseal lesion.
4. MRI: cartilaginous cap.
5. According to the shape of the neck of the tumor, we divide it into:
   a. **Pedunculated type**: long & thin neck. Directed away from the bone
   b. **Sessile type:** has short & thick neck.

#Treatment:

- Usually nothing is needed, reassure the patient.
- Surgery (just excision, no need for bone graft b/c it is a surface tumor) The surgery is indicated if there is: مهم never cosmetic

1. Presence of any complication (compression on nerves, vessels, or tendons).
2. Transformation to malignancy "chondrosarcoma" (1%). Identified by:
   - Sudden increase in **pain**.
   - Sudden increase in **size**.
   - If cartilaginous cap exceeds 2 cm on X-Ray.

2- Aggressive Benign bone tumors



1) Enchondroma:

Middle aged patients (15-50 age group).

It composed of translucent hyaline cartilage (chondroid).

#Sites: Mainly small bone e.g. phalanges in hand & foot ...etc.

#Presentation:

[6] is the formation of new bone on the surface of a bone

- Asymptomatic: Usually found **incidentally**.
- Rings may become tight due to the swelling.
- **Pathological fracture** which may cause pain, affect one side and prolonged healing.

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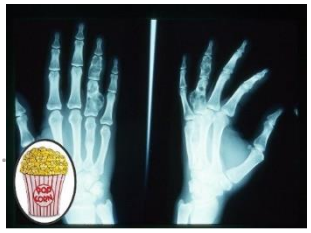


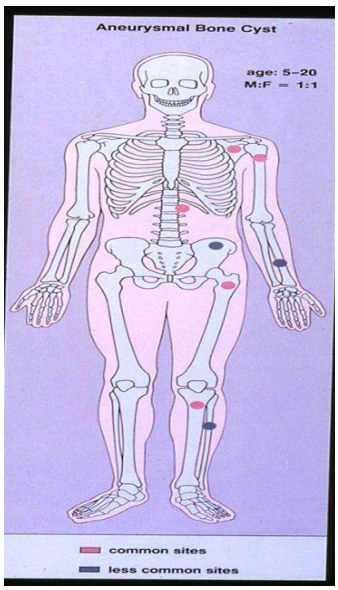
#Radiological features:

- Popcorn matrix is characteristic for cartilaginous content.
- Benign features.

#Treatment: If symptomatic (pain, bulky, fracture) >curettage + bone graft +\- fixation.

2) Aneurysmal bone cyst (ABC): تتحول **osteosarcoma**

Blood-filled cystic spaces.[7] Although called "aneurysmal" it has nothing to do with blood vessels.



#Sites:

- Around joints: Upper humerus, Upper femur, Proximal tibia.
- Spine (neural arch: lamina & pedicle) may give pressure effect.
- Flat bones: Scapula and pelvis.

#Presentation: Asymptomatic.

#Radiological feature:

1. Large "balloon-like" lesion
2. Metaphyseal lytic lesion.
3. Well- defined & sclerotic margin.
4. Expansile →thin cortex (like egg shell).
5. No periosteal reaction, No Soft tissue swelling

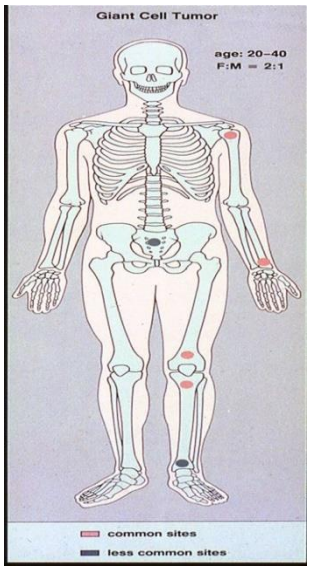
نفرّق بالاكس راي والماتيريال اللي جوّاها

Why aggressive?

1- High recurrence rate

2- Can become malignant

3) Giant cell tumor (GCT):



Benign but very aggressive tumor (most aggressive one)

- The only benign that can metastasize (to the lung. So, it's important to get chest x-ray)
- GCTs can occasionally be seen in conjunction with Paget's disease of bone and can arise in association with focal dermal hypoplasia (Goltz syndrome).
- In middle age group

#Sites:

- Most common: distal radius.
- Less common are distal tibia & sacrum.
- Very bad in the sacrum because it will affect nerve roots.

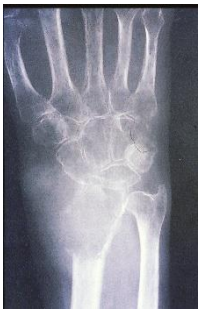
#Presentations:

Mostly patients present first with: pain, then swelling & later with pathological fracture.

#Radiological features:

1. Aggressive features:

[7] Fusiform cyst and Arteriovenous formation. Aneurysmal cysts have bloody content, while simple cysts have Orange colored fluid (Looks like Straw/sun top juice).



a. **Permeative** destruction.
b. Ill-defined borders
c. Huge soft tissue component.

2. No new bone formation b/c the osteoclast will eat this new bone (little or no periosteal reaction).

#Investigations:

- Soft tissue extension: by MRI.
- Joint extension: the articular cartilage is a very resistant against progression of the tumor from going to the joint.
- Bone extension: by CT scan.
- Bone scan: for metastasis.
- Biopsy: to confirm that it does not transform into malignant tumor.

#Treatment:

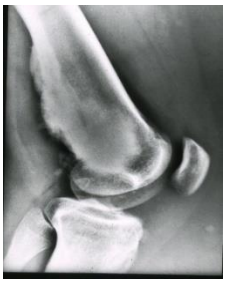
- No place for conservative treatment.
- Excision (Curettage) followed by either bone graft or prosthesis depending on the site of the tumor

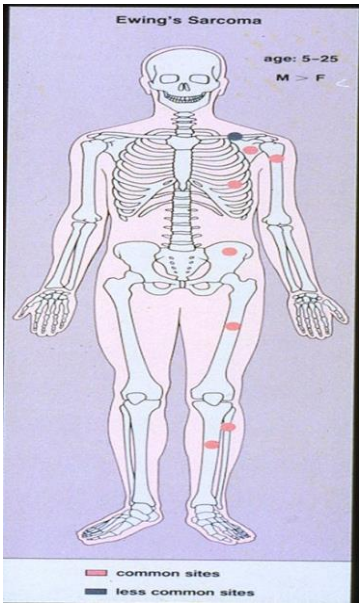
#Management of benign bone tumors: الزبدة

- Latent benign lesions may not require any treatment unless symptomatic.
- Active benign lesions usually require intervention to halt the active process and allow healing. This can range from **limited biopsy and curettage** to **detailed curettage and bone grafting**.

3- Malignant bone tumors

sketch malignant tumo





1) Ewing's sarcoma: One of the **most common** tumors in children

Origin: Unknown

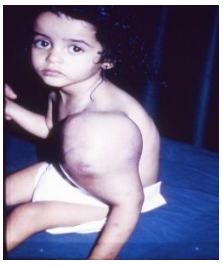
#Sites:

- Around the growth plate →femur (most common), tibia, and humerus.
- It is the only bone tumor which takes it origin from diaphysis

#Presentations:

- Pain
- Huge swelling

Most of the Ewing's sarcoma is misdiagnosed as acute osteomyelitis. Because of systemic symptoms and elevated ESR and elevated CRP: Febrile patient, High WBC's, Local Pain & redness, Ulceration of skin, Swelling. pusحتى البايوبسي يطلع معك



#Radiological features:

- Onion-skin periosteal reaction.
- **Very significant soft tissue component** *characteristic of Ewing sarcoma*

#Investigations:

- MRI and biopsy make definitive diagnosis
- You think it might be osteomyelitis: You do aspiration for drainage of pus > there will be no pus and you will find tumor tissue >You do biopsy > Ewing's sarcoma.
- N.B. you can't differentiate it from osteosarcoma unless in biopsy.

#Treatment: excision and chemotherapy

الساركوما بشكل عام ريسيكشن وكيمو لا تدخلون بالتفاصيل

2) Osteosarcoma: More common than Ewing's sarcoma. 15-35y until 60y

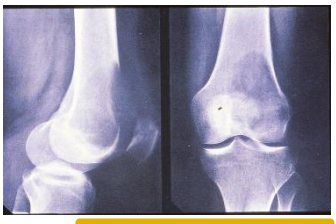
#Types:

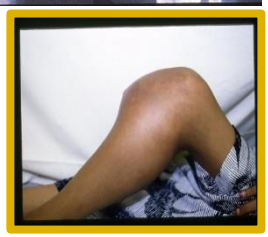
1. Primary (no need to know types of primary sarcoma)
2. Secondary (malignant transformation of benign process):
    a. Paget's disease > Paget's sarcoma
    b. post radiation sarcoma
3. Metastatic from breast, lungs or other bones.

#Sites: Around the growth plate > Around the shoulder, knee. ميتافيسيال ودايما هيوج

#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 could 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).





#Radiological features:[8]

#Investigations:

- 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:

Excision and chemotherapy

3) Multiple myeloma

- 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

#Sites: 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)

#Diagnosis:

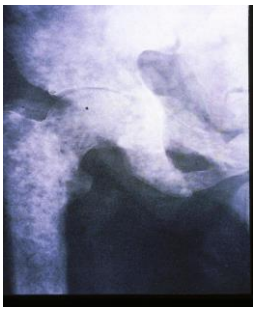
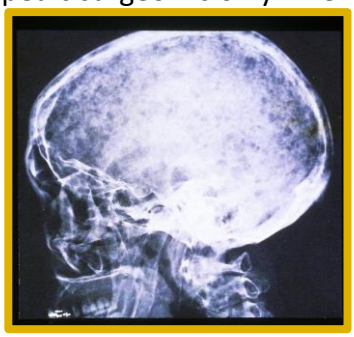
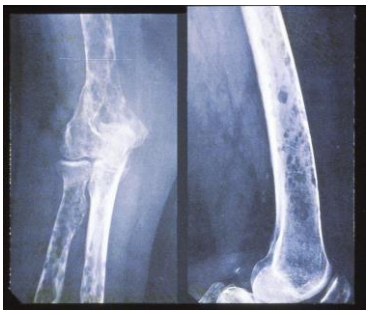
- Radiological features: Multiple Lytic or sclerotic lesions. "Salt-pepper appearance"
- In skull, there will be pepper (lytic) & salt (sclerotic) appearance (pepper-pot). you have to do skull x-ray
- Bence jones proteins test found in 24h urine collection (highly suggestive)(protein electrophoresis) Only definitive diagnosis is bone marrow aspiration.

#Treatment: (mainly medical)

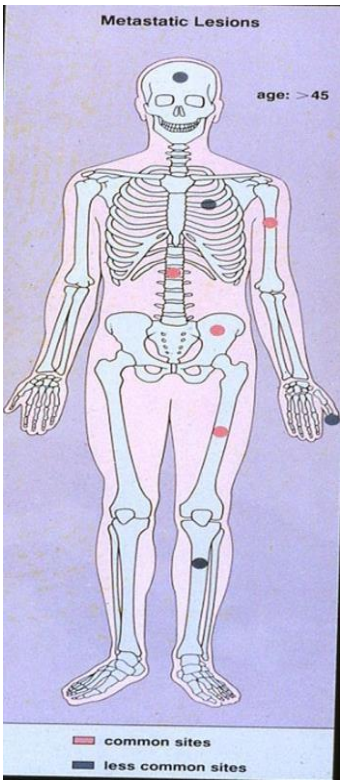
- Radiotherapy, Chemotherapy.
- Bone marrow transplant: Success rate is 30% and it Costs millions

[8] Irregular medullary and cortical destruction of the metaphysis, unlike Ewing which is seen in the diaphysis.

- The rule of orthopedic surgeon is only when you have pathological fracture, do internal fixation.

4) Metastasis



- More **common** than primary tumors in later adult life (age > 35).
- The most common tumors are: thyroid, lung, breast, colorectal, and prostate kidney.
- 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 site and treat it.

#Radiological features:

#Investigations:

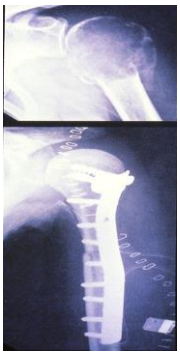
- X-ray shows lytic lesions (bone eaten away) with thinning of the cortex. Resembles bone cysts but the **age group** directs the diagnosis.
- CT- scan of the chest.
- Take biopsy to know where is the primary

Treatment:

Palliative chemo (not surgical)

Surgery indications:

- severe pain
- mechanical instability: pain in the spine when they stand
- neurological symptoms (spinal cord compression)
- prophylactic (preventive fixation): in weight bearing bones with involvement of >60% of cortex
- Fixate the fracture. (after curettage)



Bone tumor	nature	age	site	symptoms	X-ray	treatment
1) Simple bone cyst (unicameral cyst):	**benign**	young < 20	pelvis, scapula, etc.	Asymptomatic & pathological fracture.	lytic	No need for treatment, **If symptomatic**> curettage + bone graft
2) Aneurysmal bone cyst:			around joints		lytic "ballon like"	
3) Fibrous Cortical Defect (Non-Ossifying Fibroma):			around knee		Mixed (cystic + sclerotic)	
4) Osteoid osteoma:		10 - 35	cortex of long bones	**PAIN** (at night, painful scoliosis)	Lytic nidus within sclerotic bone.	NSAIDs + burn the of nidus
5) Enchondroma:		Around 40	phalanges	Asymptomatic & pathological fracture.	**Popcorn matrix** (cartilaginous).	No need for treatment, **If symptomatic**> curettage + bone graft / prosthesis
6) Multiple Enchondromatosis (AR):	**benign**, with High risk of malignant **transformation**	10-30		disabling deformity	multiple aggressive lesions	
7) Osteochondroma:	**benign**	10-35	around the knee	swelling	**Exostosis.** Benign features.	
8) Giant cell tumor (GCT):	**benign** but aggressive	20-40		Pain, swelling, & pathological fracture	Aggressive features lytic, Permeative destruction	
9) Ewing's sarcoma:	**malignant**	5-25			Sclerotic. **onion-skin** periosteal reaction.	chemo & surgery (limb salvage or amputation)
10) Osteosarcoma:[9]		5-20			Sclerotic. sunburst periosteal reaction	
11) Multiple myeloma		> 50	spine or femur	pain, osteoporosis, pathological #	lytic or sclerotic	chemo
12) Metastasis:		> 35				

[9] primary, secondary (Paget's sarcoma, post radiation sarcoma), or metastatic from breast or **other bones**.