MSK TUMORS

Lecture objectives:

1. To be able to specify the symptoms and signs; outline the assessment and appropriate investigations; propose a limited differential diagnoses and; outline the principles of management of a patient with:
   - Metastatic bone disease
   - Primary bone lesions
   - Benign bone tumor:
     A. Osteoid osteoma
     B. Bone Cyst:
        -Unicameral bone cyst (UBC)
        -Aneurysmal bone cyst (ABC)
     C. Osteochondroma
2. Malignant tumors: Osteosarcoma and Ewing's Sarcoma
3. Discuss presenting history and physical examination features of bone tumours
4. Discuss imaging characteristics of bone tumours
5. Discuss biopsy principles and techniques for biopsy.

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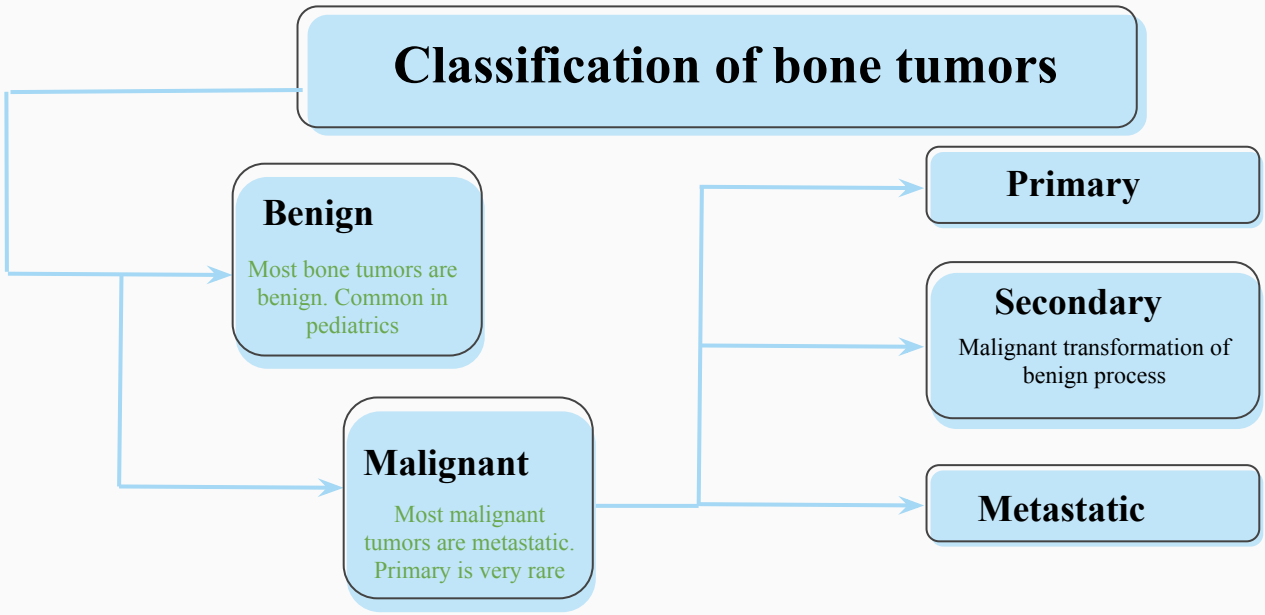
Revised by: Sondos AlHawamdeh
References: 436 team, dr's slides and notes.



Classification of tumors and tumorlike lesions by tissue of origin:

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

Focus on Red and Golden Notes!!!

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

★ History:

Age, sex. may present with: pain especially at night.

Key history Questions:

- Onset of pain - traumatic vs. atraumatic
- Progressive pain
- **Night pain, rest pain**
- Relieving factors (NSAIDS)
- Family history
- History of radiation, 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 presentation:

1- Pain. 2 - Mass. 3 - Pathologic Fracture. 4 - Incidental finding on x-ray.

★ Physical examination:

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

- Mass: fixed vs. mobile; deep to fascia or superficial (contract muscle group underneath – if deep to fascia then it becomes more fixed)
- Estimate size of mass.
- Lymphadenopathy.
- Neurovascular examination.

★ Investigations:

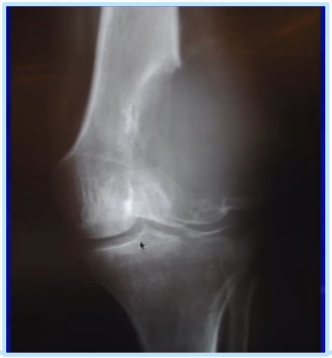
- Lab: CBC (infection or anemia), ESR, Hematological investigations.
- Imaging:
  1. Local: X-ray العلامة الفارقة هي, MRI, CT.
  2. Systemic: chest CT, Bone scan.
- Biopsy: biopsy should be done by who will do the definitive surgery, ideally orthopedic oncologist or interventional radiologist who works with the orthopedic oncologist.

★ Investigation/Staging:

➔ **9 Questions:**

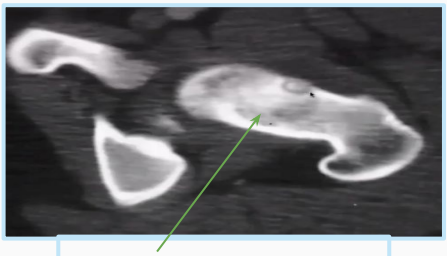
1. Where is the lesion?

- Epiphyseal, metaphyseal, diaphyseal.
- Surface.
- Peri-articular.
- Central or eccentric.

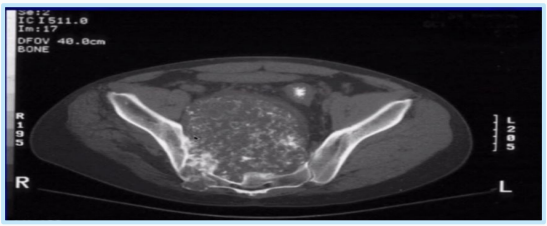


A tumor in the epiphyseal, metaphyseal area - eccentric - periarticular- with cortical erosion - no periosteal reaction= معناته الورم قاعد يكبر بشكل سريع

2. How big is it?

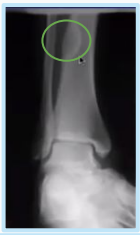


Neck lesion 1x0.5cm

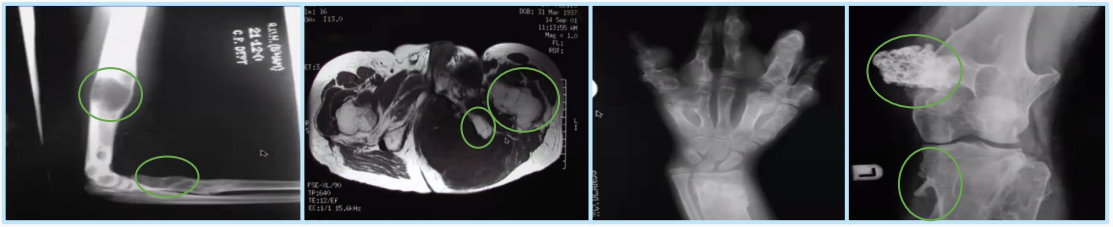


15x8 cm

3. Is it solitary or multifocal?

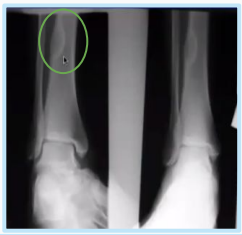


Solitary

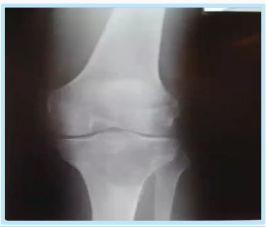


Multifocal

4. **What is the interface between the bone and the lesion?** Sclerotic or Lytic or Mixed.



Good demarcated sclerotic lesion



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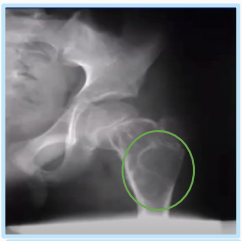
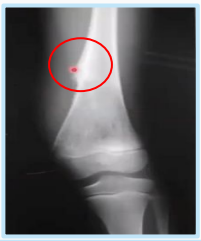
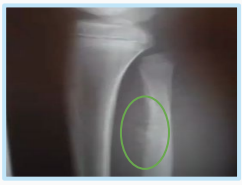


Well defined

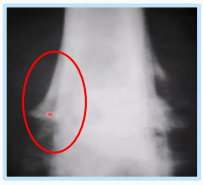


Ill defined Periosteal reaction (codman triangle)= bone try to form periosteal but مايلحق

5. Is there periosteal reaction?

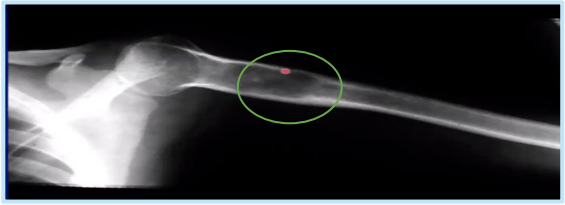


Sunburst usually in Ewing Sarcoma



Codman triangle
(Commonly with osteosarcoma)

6. Is the cortex eroded?



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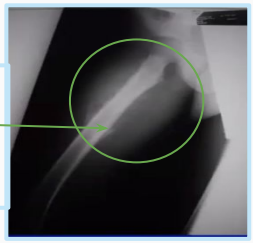
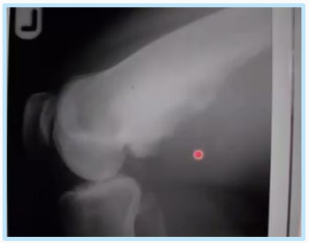
6. Is

7. Is there bony remodeling?

8. Is there a soft tissue mass?

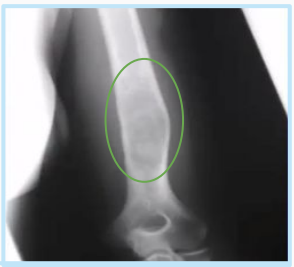
It is like Codman triangle



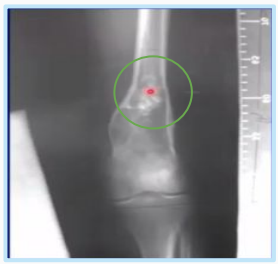


9. Is there any matrix?

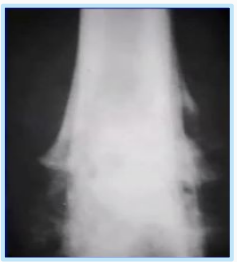
3 types: 1- Fibrous, 2- Cartilaginous, 3- osseous bone.



Fibrous: Ground-glass



Cartilage: Popcorn-like



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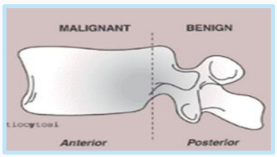
★ X-Ray reading describing a lesion should include the following:

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

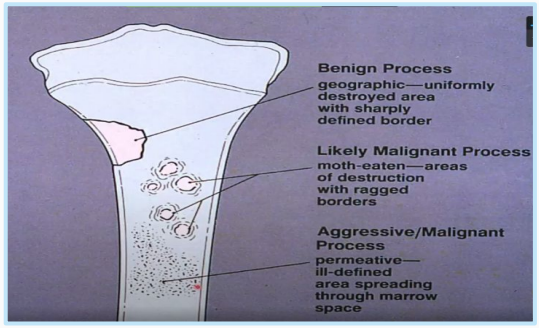
➔ **2- Site:** What bone is involved & where is the lesion in the bone?

- 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**:
  - 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.



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

- 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.



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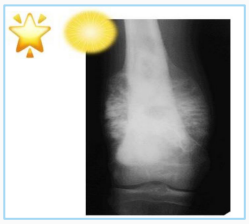
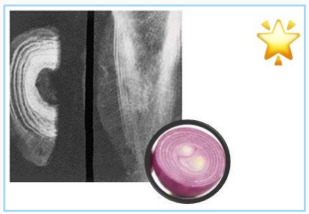
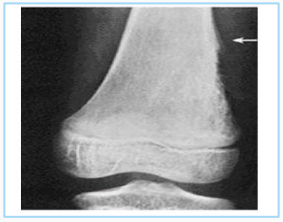
[1] DDx for Multiplicity: metastases, myeloma, lymphoma, fibrous dysplasia, enchondromatosis

➔ **4- Border of the tumor or zone of transition:**

- Well defined & sharp borders (Iill defined لو لا معناه sharp معناه تشفونه وقدرتوا شفاف ورق تجيبون لو)(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:**

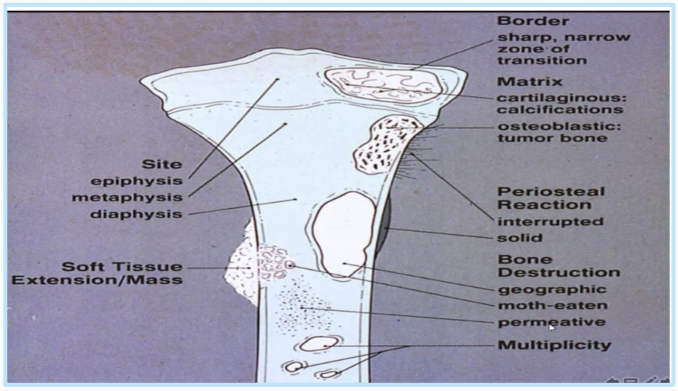
- Uninterrupted periosteal reaction usually indicates a benign process (solid clear buttress).
- Interrupted / large (teeth like) reaction indicates a malignant or aggressive nonmalignant process:

Sunburst pattern[2] in osteosarcoma and in Ewing Sarcoma	Lamellated or onion-skin type in Ewing Sarcoma	Codman triangle (arrow) in Ewing Sarcoma and usually appears in 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).

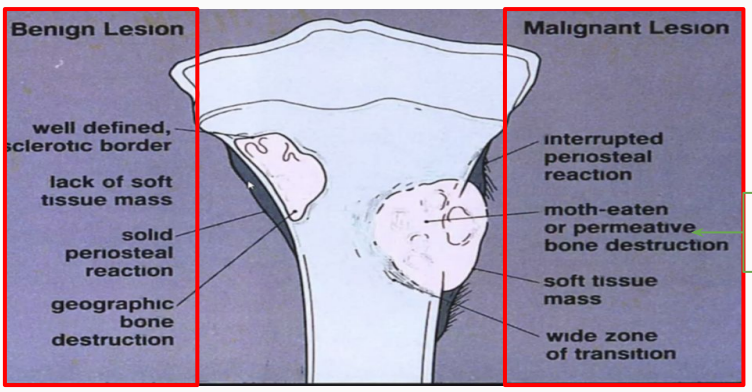
➔ **7- Soft tissue extension (swelling/shadow):** Mostly with malignant tumors (MRI is ideally used) .



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

★ **The spectrum:**

1. Benign Latent.
2. Benign Active.
3. Aggressive.
4. Lo-grade Malignant.
5. Hi-grade Malignant.



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[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

Benign latent:

- Asymptomatic.
- Well defined.
- Narrow zone of transition (geographic).
- No soft tissue mass.
- No periosteal reaction.
- No fracture.
- May or may not have matrix.
- eg's- enchondroma, non-ossifying fibroma.

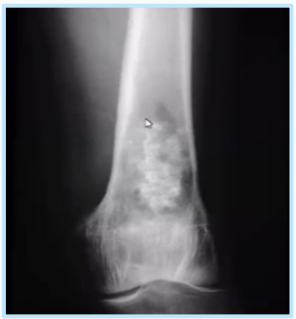
Benign Active:

- Symptomatic.
- Geographic.
- Well-ordered periosteal reaction.
- No soft tissue mass.
- May or may not have matrix.
- eg's - osteoid osteoma, UBC, eosinophilic granuloma, fibrous dysplasia, osteochondroma.

Benign Aggressive:

- Symptomatic.
- Geographic or permeative.
- Usually lytic, cortical erosion.
- May have soft tissue mass.
- Periosteal neocorticalization.
- eg's-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, Adamantinoma, Parosteal OSA, Chordoma.

High Grade Malignant:

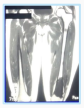
- Permeative.
- Usually has soft tissue mass.
- Cortex usually intact.
- Malignant periosteal reaction (onion skinning, sunburst, codman' s triangle)
- May or may not have matrix.
- Osteosarcoma, Ewing' s sarcoma, high grade chondrosarcoma, non-osteogenic spindle cell sarcoma (eg. MFH).

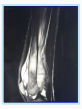
★ What next?

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

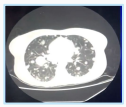
★ Local and systemic staging:

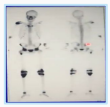
- Bloodwork – CBC, ESR, CRP, serum calcium, Alkaline phosphatase, bone profile , LDH (latter 2 are prognostic in sarcoma if increase meaning a poor prognosis).





- Local x-ray = whole bone (done), chest x-ray.
- MRI of local site: must cover entire bone (full length MRI) may find lesions in proximal which called skip metastatic(occasionally CT = if there is soft tissue)





- CT chest to rule out metastasis.
- Total body bone scan for other lesions.
- For Ewing's sarcoma – gallium scan, **bone marrow aspirate (**for lymphoma and Ewing's sarcoma)

★ What if you think it's metastatic disease?

above age of 40 think about metastasis, < 40; primary.

- Bloodwork – same bloodwork plus PSA prostate surface antigen, serum immunoelectrophoresis (In case of lymphoma and multiple myeloma).
- CT chest and abdomen to look for the source.
- Mammogram.
- Bone scan other area are involved.

★ How to stage bone tumors?

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

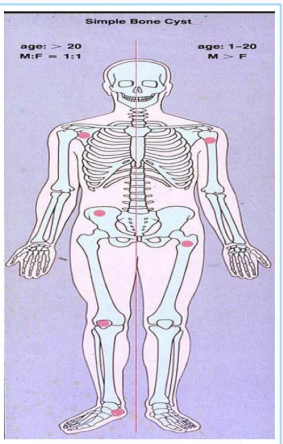
OSMOSIS

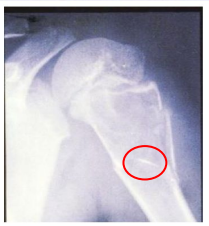
PRIMARY BENIGN BONE TUMORS:

In general, benign tumors are treated by curettage and bone graft.

BENIGN BONE TUMOR

1) Simple bone cyst (unicameral cyst): كيسة عظمية بسيطة/وحيدة المسكن(UBC)

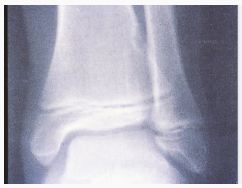


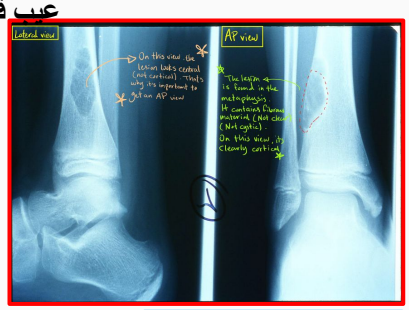


fallen -leaf sign
في كسر بس داخلها سوائل فتجي عائمة

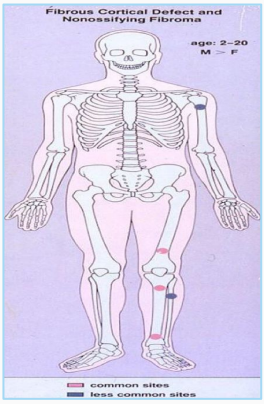
- A common benign tumor, it is benign latent except if it ruptures.
- Usually seen in young patients. Less than 20 year olds.
- **Sites:**
  - Pelvis, Calcaneus, Scapula, around the knee (lower limb more).
  - Proximal part of long bones (e.g. proximal humerus, femur or tibia).
- **Presentation:**
  Most commonly as incidental **finding** (asymptomatic) or pathological fracture.
- **Radiological features:**
  On x-ray, you will most likely find it in the metaphysis or in growth plate in case of children. If you see it in diaphysis, it's unlikely to be a cyst.
  1. Metaphysic lytic lesion. It contains fluid like **suntop juice.**
  2. Well defined sharp border.
  3. No periosteal reaction.
- **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. Never resection.
  - **Fracture**: correction of fracture will heal the cyst[3]

2) Fibrous Cortical Defect and Non-Ossifying Fibroma: عيب قشري ليفي و ورم ليفي غير متعظم



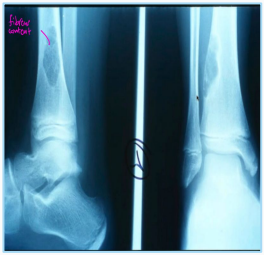


- A benign incidental finding, it is benign latent.
- Found in children. Never transfers to malignant.
- **Sites:**
  - Around knee: (lower femur, upper tibia).
  - Lower tibia.
  - May in proximal humerus
- **Presentation:**
  Asymptomatic
- **Radiological features:**
  - On x-ray, always found on the cortex (eccentric). Contains fibers and not clear material.
  - **FIBROUS COMPONENT!!!**
  - Metaphyseal Mixed lesion (cystic + sclerotic components)[4]
  - 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.
- **Treatment:**
  - Self-limited. Reassurance.
  - If large or fractured → curettage + bone graft.



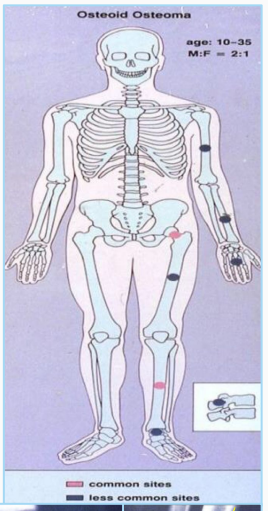
Describe this x-ray? A defect in the metaphyseal area. Geographic shape and has fibrous material. This x-ray shouldn't be worrisome and could be observed with no tx.

Is this x-ray of an adult or a child? Child, growth plate is seen.



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

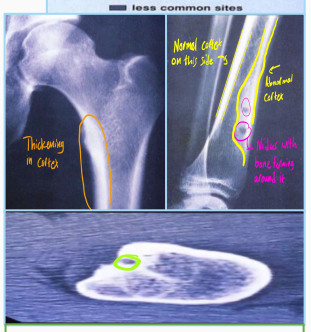
3) Osteoid osteoma: الورم العظمي العظماني



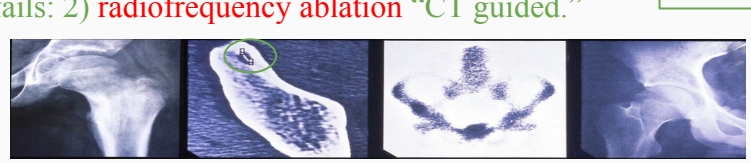
- Bone forming bone.
- it is benign active.
- **Site**:
  - May arise in the cortex of long bones, or occasionally in the cancellous bone of the spine and less commonly talus.
  - found in diaphysis and neck of femur, tibia, humerus, posterior element of spine.
- **Presentations:**
  - Main presentation 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.
- **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).
  - Cortical hyperplasia (thickening) on x-ray.
- **Treatment:**

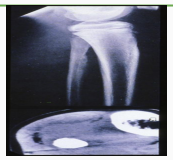
Without treatment, the lesion will slowly increase but over time will regress and usually burns out over a variable number of years.

- 1) NSAIDs challenge "aspirin challenge." If it fails: 2) radiofrequency ablation "CT guided."
3) Excision if all others fail.



The **nidus** is forming the bone and making the cortex thicker.

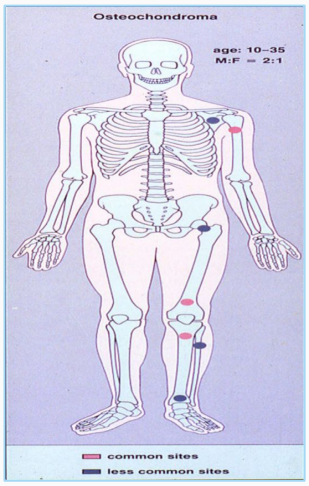




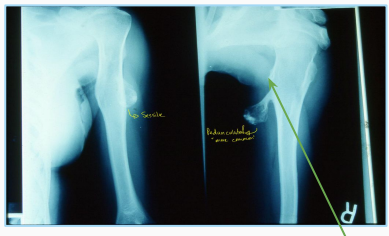
4) Osteochondroma: ورم عظمي غضروفي

- 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.
- **Sites:**
  - 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.
  - - 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.



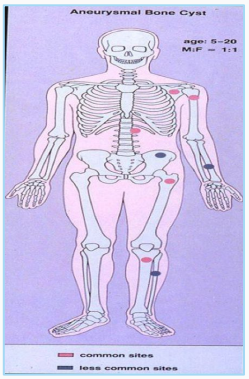
- **Radiological features:**
  1. Exostosis[5] (fungated). Mushroom-like stalk of the bony tumor (connected to the bone).
  2. Benign features.
  3. Metaphyseal lesion.
  4. MRI: cartilaginous cap.
  5. According to the shape of the neck of the tumor, we divide it into:
     A. Pedunculated type (more common): long & thin neck. Directed away from the bone.
     B. Sessile type نايمة على الكورتكس : has short & thick neck.
     - Its feature: medullary content of bone is continuity with tumor.



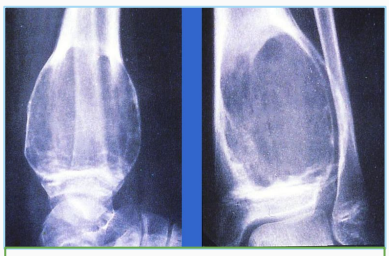
- **Treatment:**
  - Usually nothing is needed, reassure the patient.
  - Surgery is NEVER DONE for cosmetic reasons. Only done in (**he mentioned this three times**):
    1. If the lesion is obstructing a nerve, vessel or tendon.
    2. If the lesion limits movement.
    3. If the lesion is quickly increasing in size.

AGGRESSIVE BENIGN BONE TUMORS:

1) Aneurysmal bone cyst: كيسة عظمية شبيهة بأم الدم (ABC)



- They can become recurrent and may transfer to osteosarcoma.
- Blood-filled cystic spaces,[6] bigger than simple cysts,Simple cyst is not wider than growth plate and has fallen leaf sign , aneurysm bone cyst is wider than growth plate.
- It being active.
- **Sites:**
  - Around joints: Upper humerus (proximal humerus), Upper femur, Proximal tibia.
  - Spine (neural arch: lamina & pedicle) may give pressure effect.
  - Flat bones: Scapula and pelvis.
- **Presentation:**
  - Asymptomatic
- **Radiological features:**
  a. Large "balloon-like" lesion
  b. Metaphyseal lytic lesion.
  c. Well- defined & sclerotic margin.
  d. Expansile →thin cortex (like egg shell).
  e. No periosteal reaction, No Soft tissue swelling



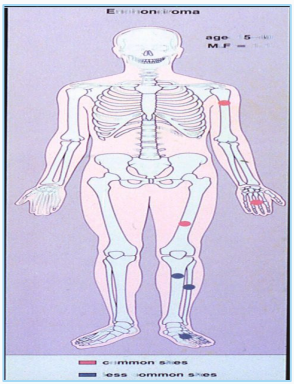
Bone منتفخ and thin cortex.
Expansile
أعرض من growth plate

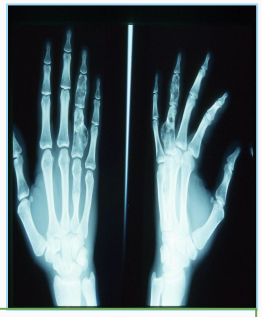
Why not malignant here?
No fracture, ground glass appearance of contents and found in metaphysis.

How to know aneurysmal cyst and not simple?
Here size is much bigger (balloon like bone) and aspiration of content will give **BLOOD**

[5] is the formation of new bone on the surface of a bone.

2) Enchondroma: ورم غضروفي داخلي





- Middle aged patients (15-50 age group).
- It composed of translucent hyaline cartilage (chondroid).
- Mostly found in the **fingers.**
- **Sites:** 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.
- **Radiological features:**
  - Popcorn matrix is characteristic for cartilaginous content.
  - Benign features.
  - Can be seen on CT.
- **Treatment:**

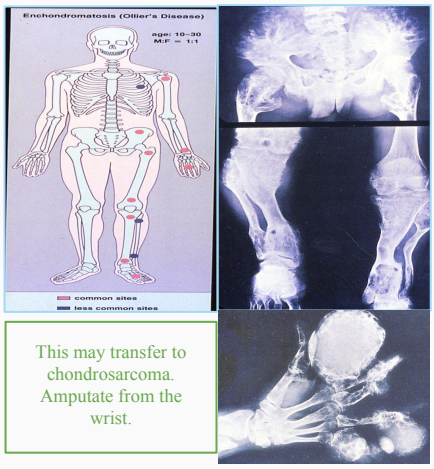
  If symptomatic (pain, bulky, fracture) > curettage + bone graft+\-fixation.

Why is this Enchondroma and not ABC?
1. LOCATION (found in fingers).
2. The ground substance here is made of chondrocytes and not blood.

3) Multiple Enchondromatosis (Ollier's disease): أورام غضروفية باطنة متعددة

- Multiple sites of enchondroma in the body most common sites are proximal humerus/tibia , which are premalignant.
- Autosomal recessive[6]
- 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. Can't remove all you will slaughter him.

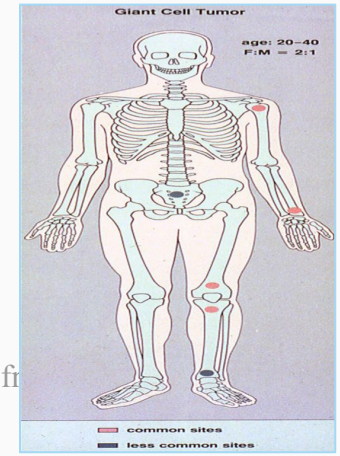


This may transfer to chondrosarcoma. Amputate from the wrist.

4) Giant cell tumor: أورام الخلايا العملاقة

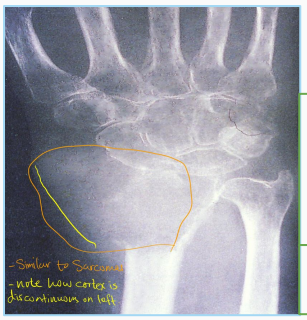
- Benign but very aggressive tumor (most aggressive benign one)
- The only benign that can metastasize (to the lung. So, it's important to get chest x-ray)
- GCTs can occasionally be seen in conjunction with Paget's disease of bone and can arise in association with focal dermal hypoplasia (Goltz syndrome).
- In middle age group, In 20-40 years.
- **Sites:**
  - Proximal humerus, distal femur proximal tibia.
  - - Most common: distal radius.
  - - Less common are distal tibia & sacrum.
  - - Very bad in the sacrum because it will affect nerve roots.
- **Presentation:**

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



[6]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.

- **Radiological features:**
  1. Aggressive features:
     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).



AP x-ray with a lesion on metaphysis or epiphysis with involvement of soft tissue but NO cortical involvement.

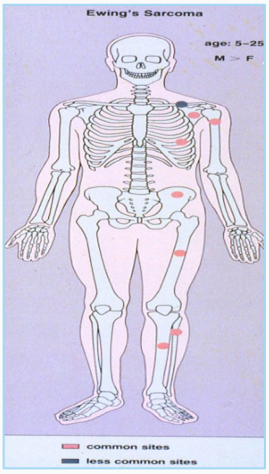
Either aggressive benign or malignant.

كأني مسحته بمساحة

- **Investigations:**
  - Soft tissue extension: by MRI.
  - Joint extension: the articular cartilage is 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.

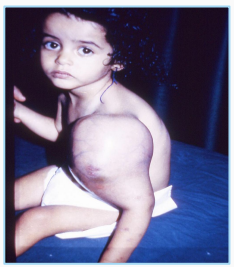
MALIGNANT BONE TUMORS:

MALIGNANT BONE TUMOR



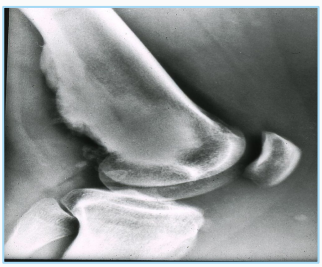
1) Ewing's Sarcoma:

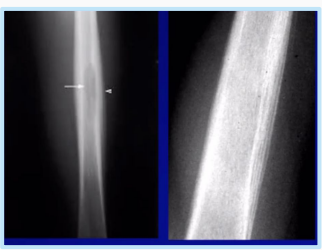
- **one** of the most common tumors in **children (3-5y)**
- **Sites:**
  - Around the growth plate → femur (most common), tibia, and humerus.
  - It is the only bone tumor which takes origin from **diaphysis,** Iliac flat bone, and ribs.
- **Presentations:**
  - Pain.
  - Huge swelling.
  - Lump with constitutional symptoms.



Most Ewing's sarcomas are 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. Even with biopsy some **pus** comes out

- **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 or little pus and you will find tumor tissue >You do biopsy > Ewing's sarcoma.
  - N.B. you can't differentiate from osteosarcoma unless in biopsy. But hints are
  - age group, and radiological location (diaphysis in Ewing and metaphysis in osteosarcoma).

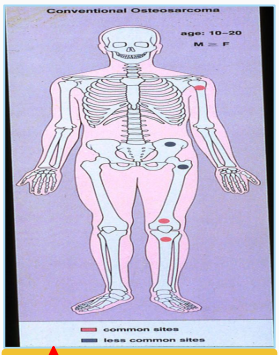




- **Treatment:**
  - (for both Ewing and osteosarcoma) neoadjuvant chemotherapy followed by tumor excision and reconstruction.
  - Why do we give neoadjuvant chemotherapy prior to surgical resection?

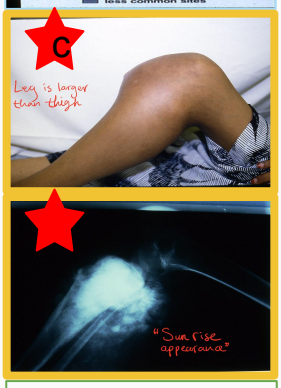
To shrink the tumor for easier removal. 2. To kill micrometastasis. 3. Gives me a hint on what chemotherapeutic agent I might use post surgery.

2) Osteosarcoma: الساركوما العظمية أو الورم الغرني العظمي

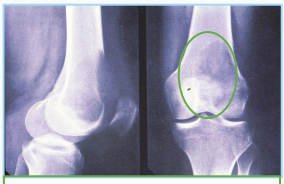


- More common than Ewing's sarcoma. **Age 10-20.**
- History of radiation and paget's disease.
- **Types:**
  1. Primary (conventional, low-grade central, telangiectatic, multicentric (multifocal), juxtacortical; no need to know types of primary sarcoma)
  2. Secondary (malignant transformation of benign process):
     a. Paget's disease > Paget's sarcoma
     b. post radiation sarcoma
  3. Metastatic from breast, lungs or bones.
- **Sites:**
  - 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 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:**
  - Codman triangle. Irregular medullary and cortical destruction of the metaphysis, unlike Ewing which is seen in the diaphysis. Cortex is intact
- **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:**
  - similar to Ewing's Sarcoma.

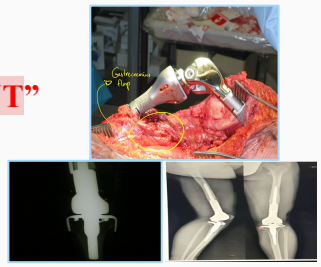
Neoadjuvant Chemotherapy then Surgery then Chemotherapy " VERY IMPORTANT"



Bone producing, cortex is intact.

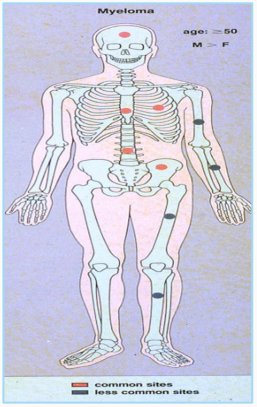


juxtacortical metaphyseal-epiphyseal

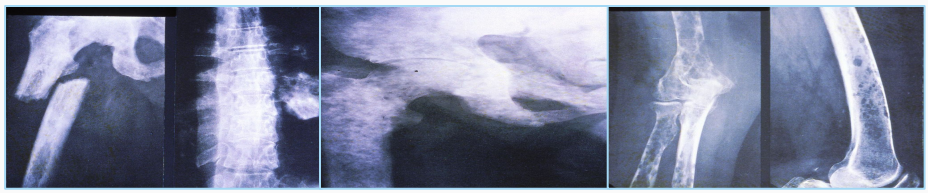


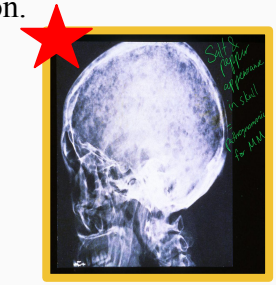
3) Multiple Myeloma: الورم النقوي المتعدد أو الورم النخاعي المتعدد

- 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
- **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. " Moth-eaten 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) (urine & serum electrophoresis) UPEP and SPEP.



- Only definitive diagnosis is by bone marrow aspiration.
- **Treatment:** (mainly medical)
  - Radiotherapy, Chemotherapy.
  - Bone marrow transplant: Success rate is 30% and it costs millions.
  - The role of orthopedic surgeon is only fix pathological fractures, do internal fixation.



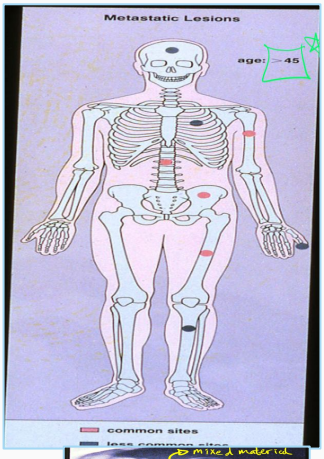


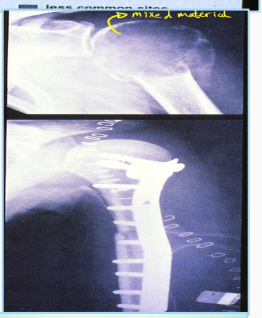
4) Metastasis:

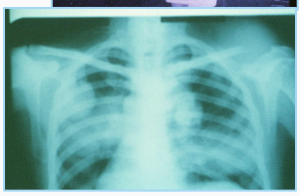
- More common than all primary tumors. Common in adults (>45 y)
- The most common tumors are: thyroid, lung, breast, colorectal, prostate and kidney.
- Usually metastasis occurs in the **highly vascular bones** e.g. vertebral body, hip, ribs, pelvis, upper end of femur, and humerus.

الاورام بعد اربعين سنة تروح لأماكن ثانية خصوصا العظم

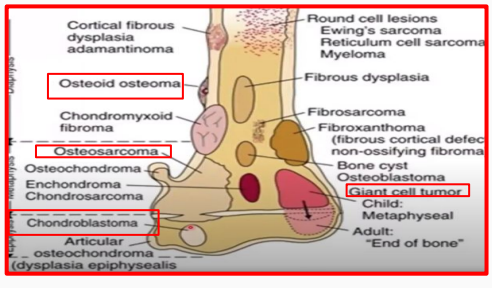
- **Presentation:**
  - Patient may present with known primary tumor.
  - May present with secondary metastasis. So, we must identify the primary site and treat it.
- **Investigations:**
  - X-ray shows lytic lesions (bone eaten away) with thinning of the cortex. Resembles bone cysts but the **age group** directs the diagnosis.
  - CT scan of the chest.
  - Take biopsy to know where is the primary.
  - You can do bone scan to check metastasis in other areas.
- **Treatment:**
  1- Palliative chemo (not surgical). The new trend is to downgrade the tumor then allograft it.
  2- 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)

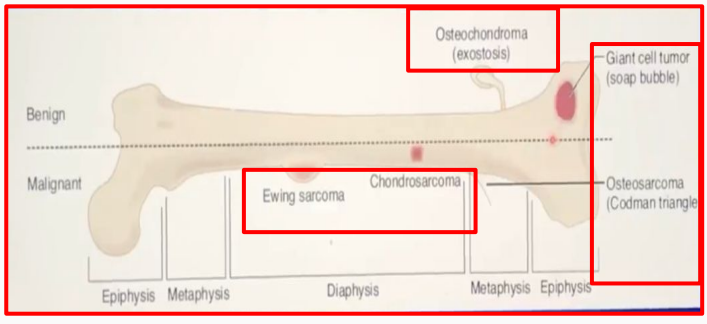






- Chondroblastoma: is in epiphysis
- Giant cell tumor: Cross metaphysis and epiphysis and
- Osteosarcoma : Cross metaphysis and epiphysis
- Osteoid osteoma: cortex





What's next ? **If you don't know what this lesion before biopsy you will not know after.**

The biopsy

- Is not a substitute for thorough history, physical examination and investigation.
- 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.

Every lesion doesn't need 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.

1-Indications for biopsy:

- Aggressive or malignant appearing bone or soft tissue lesions.
- For soft tissue lesions > 5 cm, deep to fascia or overlying bone or neurovascular structures.
- Unclear diagnosis in symptomatic patient
- Special situation - solitary bone lesion in a patient with a history of carcinoma

2-Prerequisites for biopsy: very very very important

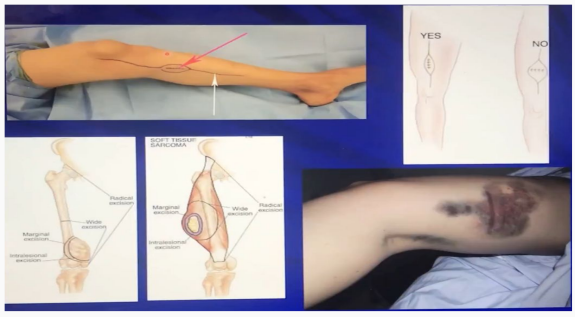
- CBC, platelets, coagulation screen if you do biopsy and pt has bleeding disorders may die
- Cross sectional imaging- depicts local anatomy , solid areas of tumor
- Experienced musculoskeletal pathologist available

3-Technique of biopsy

- Fine needle aspirate - gives cytologic specimen (adequate for some pathologists experienced with this technique
- Core biopsy (tru-cut) allows for ultrastructural examination
- Incisional biopsy
- Excisional biopsy (remove whole mass) it is biopsy and surgery pain same time - selected indications (small < 5c ,superficial soft tissue masses)

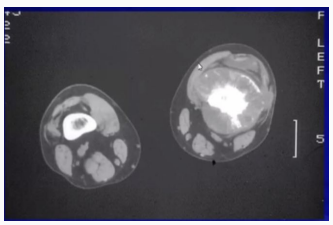
Principles of Open Biopsy:

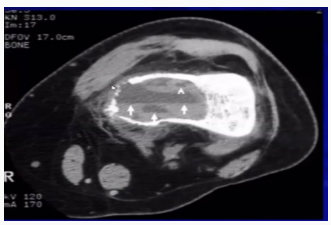
1. Extensile incision – longitudinal in extremities.
2. Avoid developing planes.
3. Use involved compartment اروح لمكان واحد .
4. Do not expose neurovascular structures.
5. Meticulous hemostasis
6. Release tourniquet prior to wound closure.
7. If using drain, bring out in line with incision.



General Recommendations:

- ❑ 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.





Summary						
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:[14]		5-20			Sclerotic. **sunburst** periosteal reaction	
11) Multiple myeloma		> 50	spine or femur	pain, osteoporosis, pathological #	lytic or sclerotic	chemo
12) Metastasis:		> 35				

Bone Tumours

- primary bone tumours are rare after 3rd decade
- metastases to bone are relatively common after 3rd decade

Clinical Features

- malignant (primary or metastasis): local pain and swelling (weeks to months), worse on exertion and at night, ± soft tissue mass
- benign: usually asymptomatic
- minor trauma often initiating event that calls attention to lesion

Red Flags

- Persistent skeletal pain
- Localized tenderness
- Spontaneous fracture
- Enlarging mass/soft tissue swelling

X-ray Findings

- Lytic, lucent, sclerotic bone
- Involvement of cortex, medulla, soft tissue
- Radiolucent, radiopaque, or calcified matrix
- Periosteal reaction
- Permeative margins
- Pathological fracture
- Soft tissue swelling

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

Benign	Malignant
No periosteal reaction	Acute periosteal reaction • Codman's triangle • "Onion skin" • "Sunburst"
Thick endosteal reaction Well developed bone formation Intraosseous and even calcification	Broad border between lesion and normal bone Varied bone formation Extraosseous and irregular calcification

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

Diagnosis

- malignancy is suggested by rapid growth, warmth, tenderness, lack of sharp definition
- staging should include:
  - blood work including liver enzymes
  - CT chest
  - bone scan
  - bone biopsy
    - should be referred to specialized centre prior to biopsy
    - classified into benign, benign aggressive, and malignant
  - MRI of affected bone

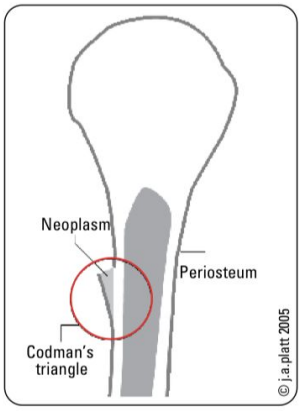


Figure 56. Codman's triangle – a radiographic finding in malignancy, where the partially ossified periosteum is lifted off the cortex by neoplastic tissue

Benign Active Bone Tumours

BONE-FORMING TUMOURS

Osteoid Osteoma

- bone tumour arising from osteoblasts
- peak incidence in 2nd and 3rd decades, M:F = 2:1
- proximal femur and tibia diaphysis most common locations
- not known to metastasize
- radiographic findings: small, round radiolucent nidus (<1.5 cm) surrounded by dense sclerotic bone ("bull's-eye")
- symptoms: produces severe intermittent pain from prostaglandin secretion and COX1/2 expression, mostly at night (diurnal prostaglandin production), thus is characteristically relieved by NSAIDs
- treatment: NSAIDs for night pain; surgical resection of nidus

FIBROUS LESIONS

Fibrous Cortical Defect

- i.e. non-ossifying fibroma, fibrous bone lesion
- most common benign bone tumour in children, typically asymptomatic and an incidental finding
- occur in as many as 35% of children, peak incidence between 2-25 yr old, higher prevalence in males
- femur and proximal tibia most common locations, 50% of patients have multiple defects that are usually bilateral, symmetrical
- radiographic findings: diagnostic, metaphyseal eccentric 'bubbly' lytic lesion near physis; thin, smooth/lobulated, well-defined sclerotic margin
- treatment: most lesions resolve spontaneously

Osteochondroma

- cartilage capped bony tumour
- 2nd and 3rd decades, M:F = 1.8:1
- most common of all benign bone tumours – 45%
- 2 types: sessile (broad based and increased risk of malignant degeneration) vs. pedunculated (narrow stalk)
- metaphysis of long bone near tendon attachment sites (usually distal femur, proximal tibia, or proximal humerus)
- radiographic findings: cartilage-capped bony spur on surface of bone ("mushroom" on x-ray)
- may be multiple (hereditary, autosomal dominant form) – higher risk of malignant change
- generally very slow growing and asymptomatic unless impinging on neurovascular structure ('painless mass')
  - growth usually ceases when skeletal maturity is reached
- malignant degeneration occurs in 1-2% (becomes painful or rapidly grows)
- treatment: typically observation; surgical excision if symptomatic

Enchondroma

- hyaline cartilage tumour; majority asymptomatic, presenting as incidental finding or pathological fracture
- 2nd and 3rd decades
- 60% occur in the small tubular bones of the hand and foot; others in femur (20% - Figure 57), humerus, ribs
- benign cartilaginous growth, an abnormality of chondroblasts, develops in medullary cavity
  - single/multiple enlarged rarefied areas in tubular bones
  - lytic lesion with sharp margination and irregular central calcification (stippled/punctate/popcorn appearance)
- malignant degeneration to chondrosarcoma occurs in 1-2% (pain in absence of pathologic fracture is an important clue)
- not known to metastasize
- treatment: observation with serial x-rays; surgical curettage if symptomatic or lesion grows

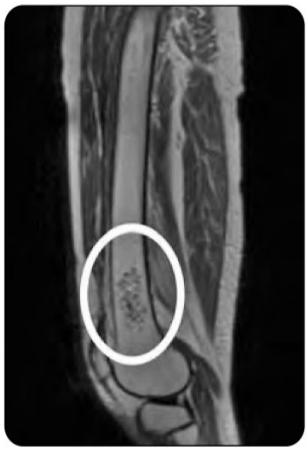


Figure 57. T1 MRI of femoral enchondroma

CYSTIC LESIONS

Unicameral/Solitary Bone Cyst

- most common cystic lesion; serous fluid-filled lesion
- children and young adults, peak incidence during first 2 decades, M:F = 2:1
- proximal humerus and femur most common
- symptoms: asymptomatic, or local pain; complete pathological fracture (50% of presentations) or incidental detection
- radiographic findings: lytic translucent area on metaphyseal side of growth plate, cortex thinned/expanded; well-defined lesion
- treatment: aspiration followed by steroid injection; curettage ± bone graft indicated if re-fracture likely

Benign Aggressive Bone Tumours

Giant Cell Tumours/Aneurysmal Bone Cyst/Osteoblastoma

- affects patients of skeletal maturity, peak 3rd decade
- osteoblastoma: found in the distal femur, proximal tibia, distal radius, sacrum, tarsal bones, spine
- giant cell tumour: pulmonary metastases in 3%
- aneurysmal bone cysts: either solid with fibrous/granular tissue, or blood-filled
- radiographic findings
  - giant cell tumour: eccentric lytic lesions in epiphyses adjacent to subchondral bone; may break through cortex; T2 MRI enhances fluid within lesion (hyper-intense signal)
  - aneurysmal bone cyst: expanded with honeycomb shape
  - osteoblastoma: often nonspecific; calcified central nidus (>2 cm) with radiolucent halo and sclerosis
- symptoms: local tenderness and swelling, pain may be progressive (giant cell tumours), ± symptoms of nerve root compression (osteoblastoma)
- 15% recur within 2 yr of surgery

Treatment

- intralesional curettage + bone graft or cement
- wide local excision of expendable bones

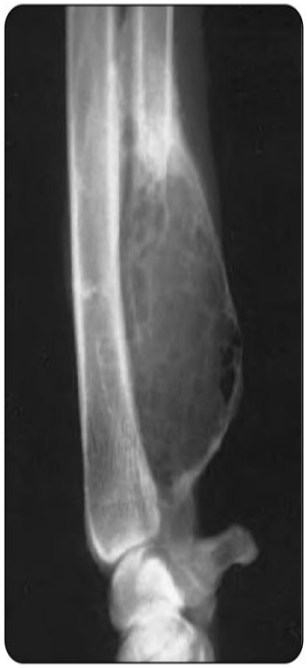


Figure 58. X-ray of aneurysmal bone cyst
Note the aggressive destruction of bone

Malignant Bone Tumours

Table 26. Most Common Malignant Tumour Types for Age

Age	Tumour
<1	Neuroblastoma
1-10	Ewing's of tubular bones
10-30	Osteosarcoma, Ewing's of flat bones
30-40	Reticulum cell sarcoma, fibrosarcoma, periosteal osteosarcoma, malignant giant cell tumour, lymphoma
>40	Metastatic carcinoma, multiple myeloma, chondrosarcoma

Osteosarcoma

- malignant bone tumour
- most frequently diagnosed in 2nd decade of life (60%), 2nd most common primary malignancy in adults
- history of Paget's disease (elderly patients), previous radiation treatment
- predilection for sites of rapid growth: distal femur (45% - Figure 59), proximal tibia (20%), and proximal humerus (15%)
  - invasive, variable histology; frequent metastases without treatment (lung most common)
- painful symptoms: progressive pain, night pain, poorly defined swelling, decreased ROM

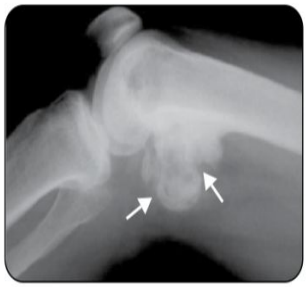


Figure 59. X-ray of osteosarcoma of distal femur

- radiographic findings
  - characteristic periosteal reaction: Codman's triangle (Figure 56) or "sunburst" spicule formation (tumour extension into periosteum)
  - destructive lesion in metaphysis may cross epiphyseal plate
- management: complete resection (limb salvage, rarely amputation), neo-adjuvant chemo; bone scan – rule out skeletal metastases, CT chest – rule out pulmonary metastases
- prognosis: 90% survival for low-grade; 70% survival for high-grade

Chondrosarcoma

- malignant chondrogenic tumour
- primary (2/3 cases)
  - previous normal bone, patient >40 yr; expands into cortex to cause pain, pathological fracture
- secondary (1/3 cases)
  - malignant degeneration of pre-existing cartilage tumour such as enchondroma or osteochondroma
  - age range 25-45 yr, better prognosis than primary chondrosarcoma
- symptoms: progressive pain, uncommonly palpable mass
- radiographic findings: in medullary cavity, irregular "popcorn" calcification
- treatment: unresponsive to chemotherapy, treat with aggressive surgical resection + reconstruction; regular follow-up X-rays of resection site and chest
- prognosis: 90% survival for low-grade (10yr survival); 20-40% survival for high-grade

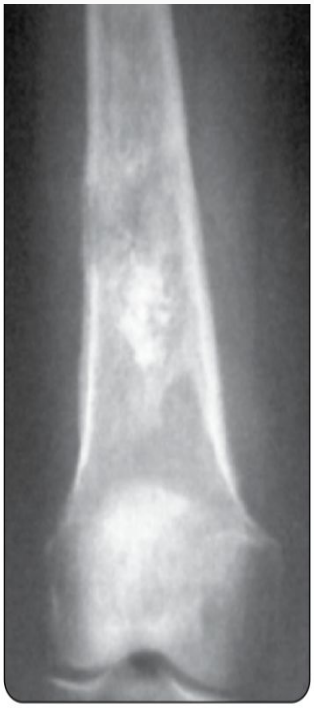


Figure 60. X-ray of femur chondrosarcoma

Ewing's Sarcoma

- malignant, small round cell sarcoma
- most occur between 5-25 yr old
- florid periosteal reaction in metaphyses of long bone with diaphyseal extension
- metastases frequent without treatment
- signs/symptoms: presents with pain, mild fever, erythema, and swelling; anemia, increased WBC, ESR, LDH (mimics an infection)
- radiographic findings: moth-eaten appearance with periosteal lamellated pattern ("onion-skinning")
- treatment: resection, chemotherapy, radiation
- prognosis: 70% survival, worst prognostic factor is distant metastases

Multiple Myeloma

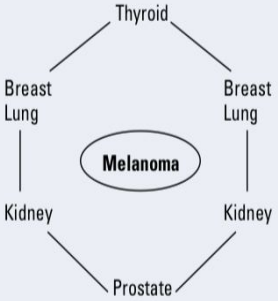
- proliferation of neoplastic plasma cells
- most common primary malignant tumour of bone in adults (~43%)
- 90% occur in people >40 yr old, M:F = 2:1; twice as common in African-Americans
- signs/symptoms: localized bone pain (cardinal early symptom), compression/pathological fractures, renal failure, nephritis, high incidence of infections (e.g. pyelonephritis/pneumonia), systemic (weakness, weight loss, anorexia)
- labs: anemia, thrombocytopenia, increased ESR, hypercalcemia, increased Cr
- radiographic findings: multiple, "punched-out" well-demarcated lesions, no surrounding sclerosis, marked bone expansion
- diagnosis
  - serum/urine immunoelectrophoresis (monoclonal gammopathy)
  - CT-guided biopsy of lytic lesions at multiple bony sites
- treatment: chemotherapy, bisphosphonates, radiation, surgery for symptomatic lesions or impending fractures – debulking, internal fixation
- prognosis: 5 yr survival 30%; 10 yr survival 11%
- see Hematology, H49

Signs of Hypercalcemia
"Bones, Stones, Moans, Groans, Psychiatric overtones"
CNS: headache, confusion, irritability, blurred vision
GI: N/V, abdominal pain, constipation, weight loss
MSK: fatigue, weakness, unsteady gait, bone and joint pain
GU: nocturia, polydipsia, polyuria, UTIs

Bone Metastases

- most common cause of bone lesions in adults; typically age >40
- 2/3 from breast or prostate; also consider thyroid, lung, kidney
- usually osteolytic; prostate occasionally osteoblastic
- may present with mechanical pain and/or night pain, pathological fracture, hypercalcemia
- bone scan for MSK involvement, MRI for spinal involvement may be helpful
- treatment: pain control, bisphosphonates, stabilization of impending fractures if Mirel's Critera >8 (ORIF, IM rod, bone cement)

Most Common Tumours Metastatic to Bone



BLT with a Kosher Pickle
Breast
Lung
Thyroid
Kidney
Prostate

Table 27. Mirel's Criteria for Impending Fracture Risk and Prophylactic Internal Fixation

Variable	Number Assigned		
	1	2	3
Site	Upper arm	Lower extremity	Peritrochanteric
Pain	Mild	Moderate	Severe
Lesion	Blastic	Mixed	Lytic
Size	<1/3 bone diameter	1/3-2/3 diameter	>2/3 diameter

QUESTIONS

1). 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 (98.8°F), 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?

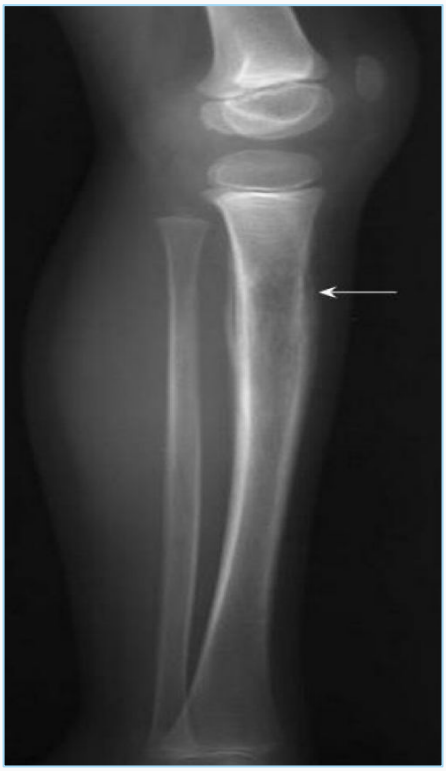
A. Osteosarcoma
B. Ewing's sarcoma
C. Giant cell tumor
D. Metastasis

2). A 15-year-old man 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?

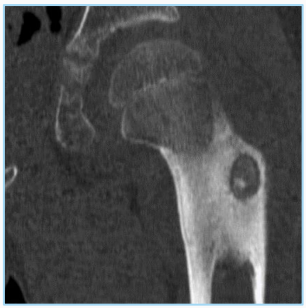
A. Onion-skin appearance
B. Punched-out lytic lesions
C. Necrosis surrounded by sclerosis of bone

3). 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 20mm/hr. An X-ray of this patient's left leg is shown . What is the most likely diagnosis?

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

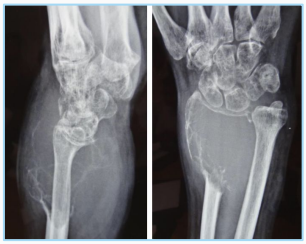


4). A 12-year-old boy comes to the clinic with his mother because of severe pain in his right leg for the past month. He says that the pain is mostly located in his thigh and that it was initially mild but now the pain is so bad that he cannot walk on his leg. His mother says that aspirin relieves his 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 1cm 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. Osteochondroma
D. Osteoid osteoma

5). 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

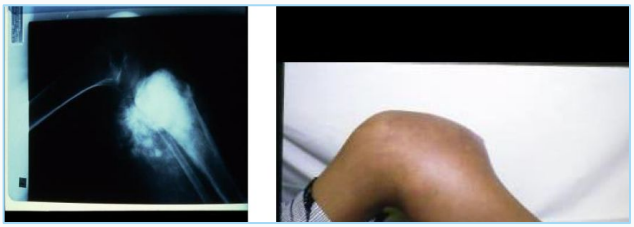
6). Which one of the following is the most common benign bone tumor?

A. Osteoid osteoma
B. Enchondroma
C. Osteochondroma
D. Unicameral cyst

7). What is the most aggressive benign tumor?

A. Osteoid osteoma
B. Enchondroma
C. Giant cell tumor
D. Simple bone cyst

8). 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 and is shown below. What is the confirmatory diagnosis test?



A- MRI of knee
B- CT of knee
C- Bone scan with gallium
D- Biopsy

Answers: **1-A. 2-A. 3-D. 4-D. 5-D. 6-C. 7-C. 8-D.**