Bone Tumors

- A tumor is a lump or mass of tissue that divide uncontrollably.

- <u>Classification:</u>

1. primary bone tumor .. Arise originally from the bone
   - benign or Malignant
   - in first 3 decades of life.
   - Benign tumors > malignant.
   - commonest sites around the knee , distal femur and proximal tibia
2. secondary bone tumor ..
   - metastasize to the bone form (breast , prostate etc ..)
   - malignant transformation of benign lesions.
   - Most commonly noticed above the fifth decade of life

- **Histological classification based on dominant tissue:**

Cell type	Benign	Malignant
Chondrogenic	Osteochondroma Enchondroma Chondroblastoma Chondromyxoid fibroma	Chondrosarcoma
Osteogenic	Osteoid osteoma Osteoblastoma	Osteosarcoma
Histocytic	Fibrous histiocytoma	Malignantf fibrous histiocytoma
Fibrogenic	fibrous cortical defect(non-ossifying fibroma), fibrous dysplasia, fibroma	Fibrosarcoma
Vascular	Hemangioma	Angiosarcoma
Others	Giant cell tumor, aneurismal bone cyst, simple bone cyst	Malignant Giant cell tumor

Clinical presentation:

History:

1. **asymptomatic** accidentally discovered on x-ray, more likely with benign lesions.
2. **Pain:** it may be caused by:
    1. rapid expansion
    2. central hemorrhage.
    3. pathological fracture.
3. **Swelling or a lump.**
4. **Neurological symptoms .. Compression by mass**
5. **Pathological fracture**

Examination :

- **Possible mass**
- **Joint effusion and \or limitation of movemet** in tumor around joint
- **muscle spasm and back stiffness, or painful scoliosis** in case of .Spinal lesions
- **Lymphadenopathy** should be checked
- **neurovascular check** for tumors in the limbs

Imaging:

- **X-ray** : check for

1. Which bone is involved
2. Where is the lesion in the bone? (epiphysis , metaphysis or diaphysis.)
3. lesion solitary or multiple?
4. centric or eccentric.
5. osteolytic or osteoblastic / is center calcified?
6. margins of the lesion well- or ill-defined?
7. Any cortical destruction?
8. Any periosteal reaction?
9. Any soft tissue extension

Benign bone lesion

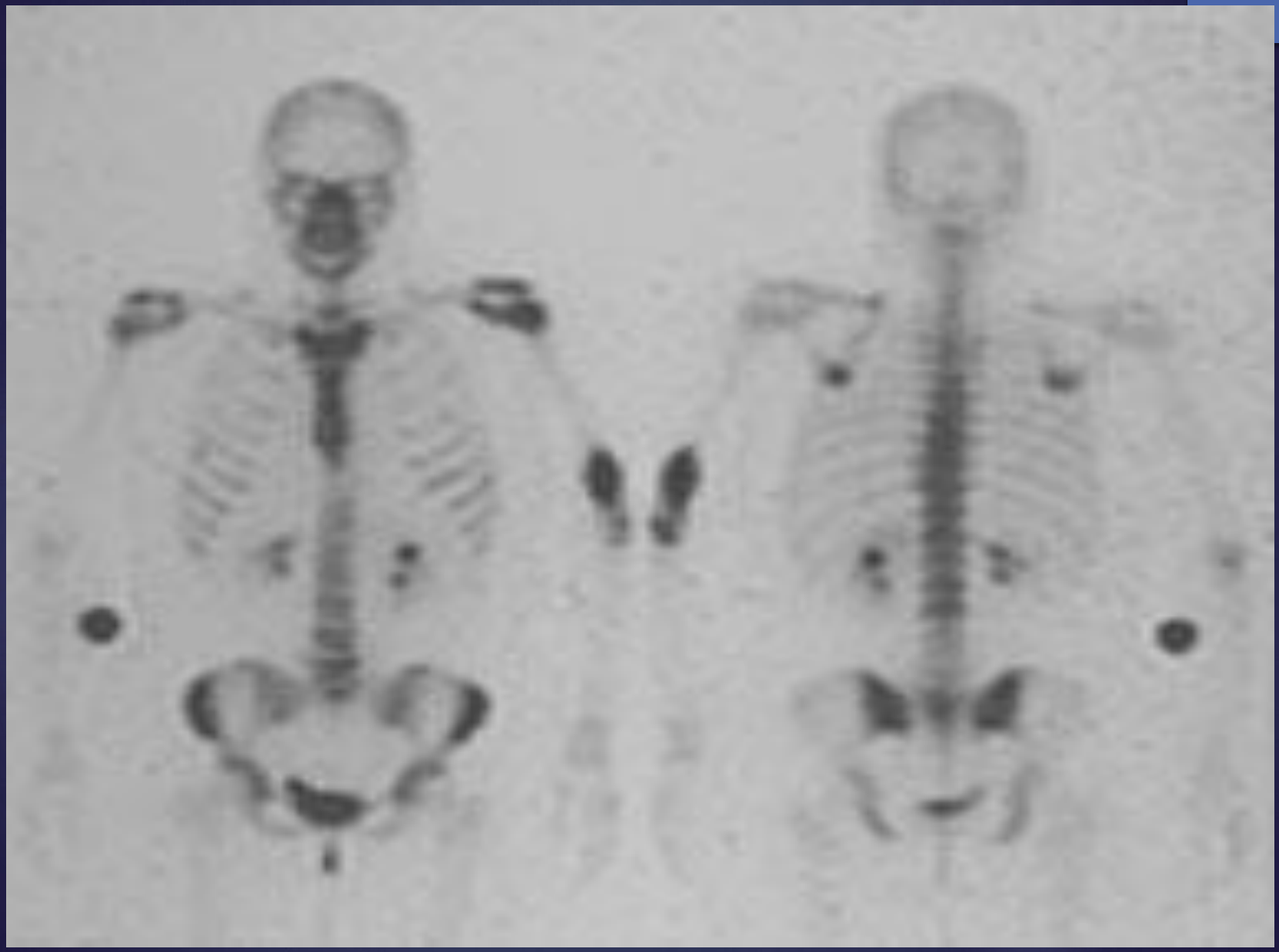
malignant bone lesion

Other imaging:

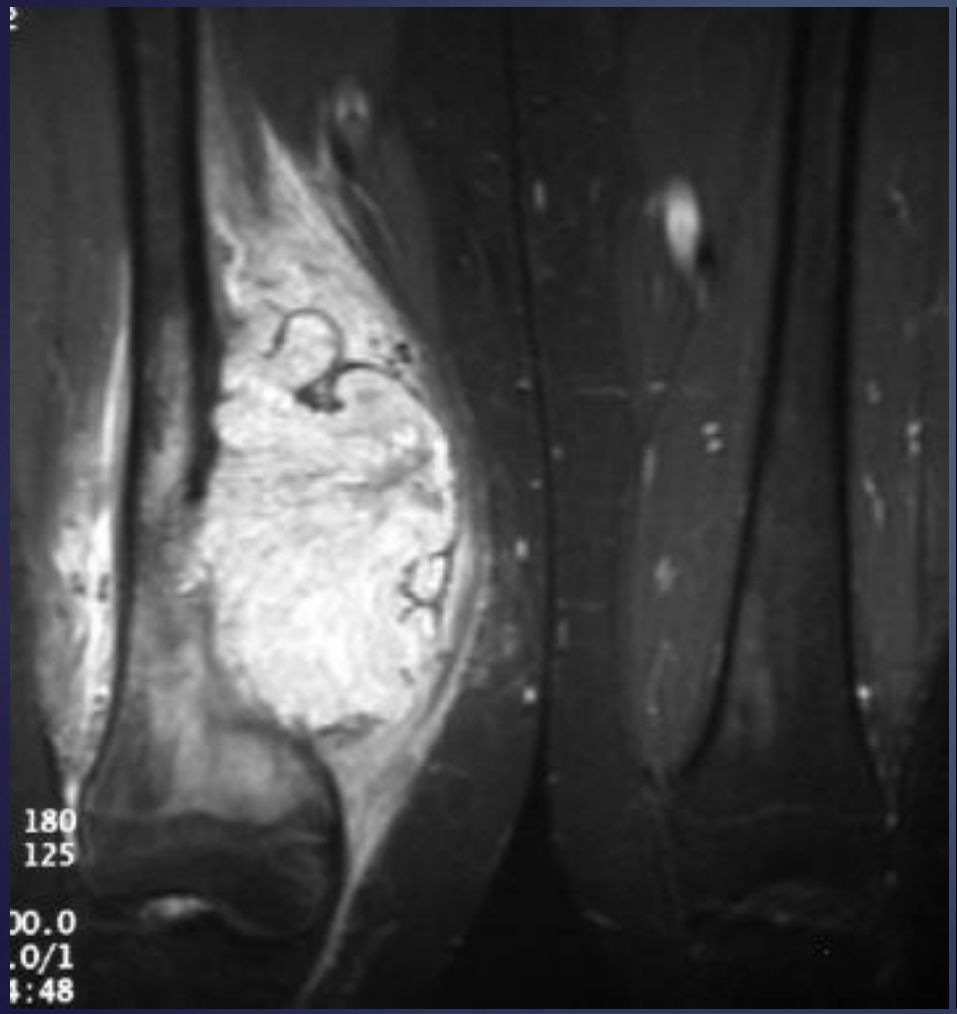
- **Radioisotope scanning** : Helpful in metastatic and skip lesions.

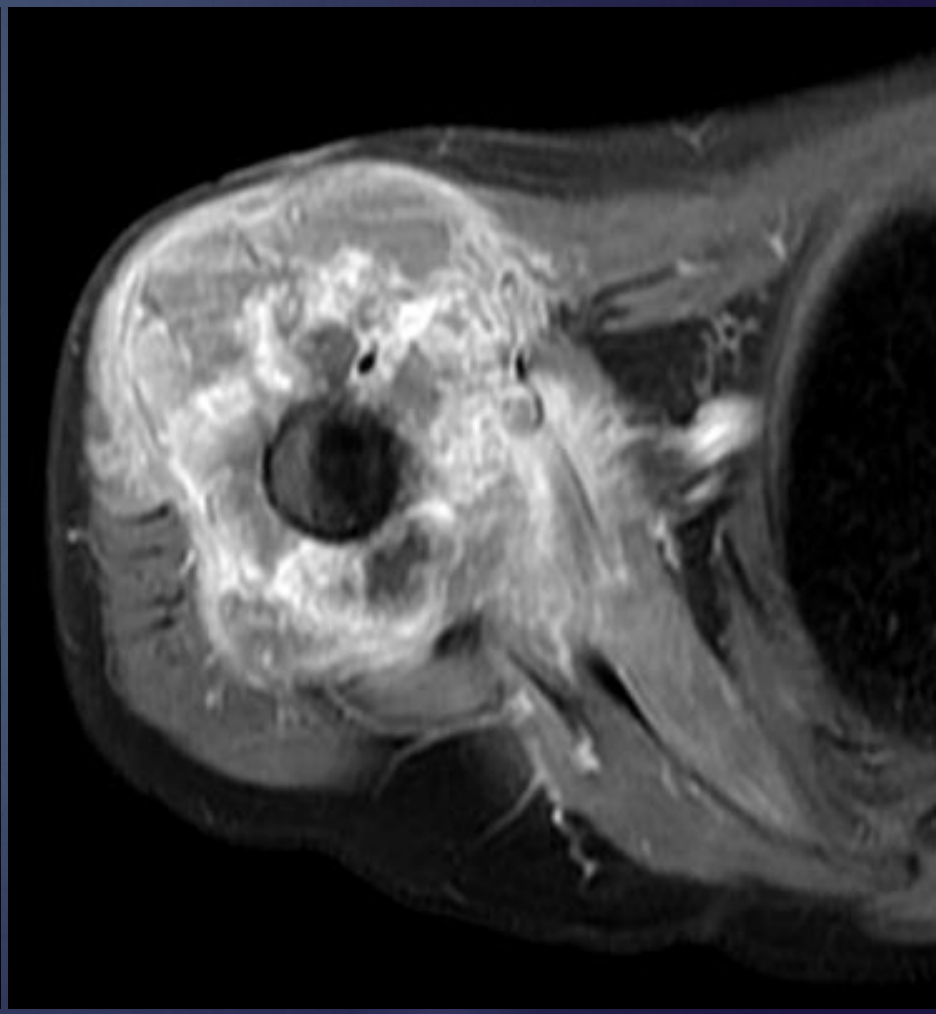
- **CT** & **MRI** : can determine:
  - ✓ The intra osseous and extra osseous extension of the tumor.
  - ✓ Skip lesion in the same bone
  - ✓ Lesions in inaccessible sites ,like the spine or pelvis.
  - ✓ Pulmonary metastasis.

Bone Isotope Scan



MRI





Lab. investigations

- **Blood tests to exclude other conditions e.g.**
  - infection
  - metabolic bone disorders
  - "brown tumor" in hyperparathyroidism.
- **Serum and urine protein electrophoresis**
  - for abnormal globulin and Bence-Jones protein in myeloma.
- **serum acid phosphatase**
  - for prostatic carcinoma.

Biopsy

- for accurate diagnosis

two basic methods of doing a biopsy:

1. **Needle biopsy**
   - under local anesthesia or GA using an X-ray or CT guidance
   - value is in sampling inaccessible tumors.
2. **Open biopsy:**
   - done through a small incision under general anesthesia in an operating room.
   - Several samples should be taken

Needle biopsy

Open biopsy

Differential Diagnosis
tumor mimicker

1. myositis ossificans.
2. stress fracture.
3. bone infection.
4. brown tumor of hyperparathyroidism.

Staging the lesion:

- ***Enneking's staging of benign lesion:***
  - *latent*
  - *active*
  - *aggressive*.
- ***Enneking's staging of malignant tumor:***

Stage1: low grade sarcomas

- *1A: intracompartmental*
- *1B: extracompartmental*

Stage 2: high grade lesions.

- *2A: intracompartmental*
- *2B: extracompartmental*

Stage3: sarcomas which have metastasized. e.g. to lung.

Management of Primary Benign tumors:

- Observation only / might disappear over time(e.g. fibrous cortical defect, simple bone cyst)
- Excision to reduce the risk of pathological fracture
- Excision because its symptomatic / or have a risk of malignant potentials like Giant cell tumor

Management of Primary Malignant tumors:

- If the lesion is suspected to be a malignant tumor ,the patient is admitted for
  - detailed examination
  - blood tests
  - CXR
  - pulmonary CT
  - biopsy.

- **Treatment goals include**
  - Removing the tumor
  - preserving the function of the body .

Methods of treatment of malignant tumor

- **Tumor excision with wide excision or radical excision.**
  - **Limb-sparing surgery:** removes cancerous section of bone but keeps nearby muscles, tendons, nerves and blood vessels . The excised bone is replaced with a metallic implant (prosthesis) or bone transplant.
  - **Amputation** : removes all or part of an arm or leg when the tumor is large and/or nerves and blood vessels are involved.

- **Radiotherapy** : uses high-dose X-rays.
  - Shrinks the tumors
  - suitable for inaccessible sites

- **Multi-agent chemotherapy** : neoadjuvant for malignant bone tumors

Benign Bone Tumors

Osteochondroma

the most common benign bone tumor

- common locations include
  - knee (proximal tibia, distal femur)
  - proximal femur
  - proximal humerus
- Can be either
  1. solitary ostoechondroma
  2. Multiple Hereditary Exostosis (MHE)
- Clinical presentation
  - Asymptomatic / painless mass
  - mechanical symptoms
  - symptoms of neurovascular compression

Osteochondroma

- **Radiograph**
  - sessile (broad base) or

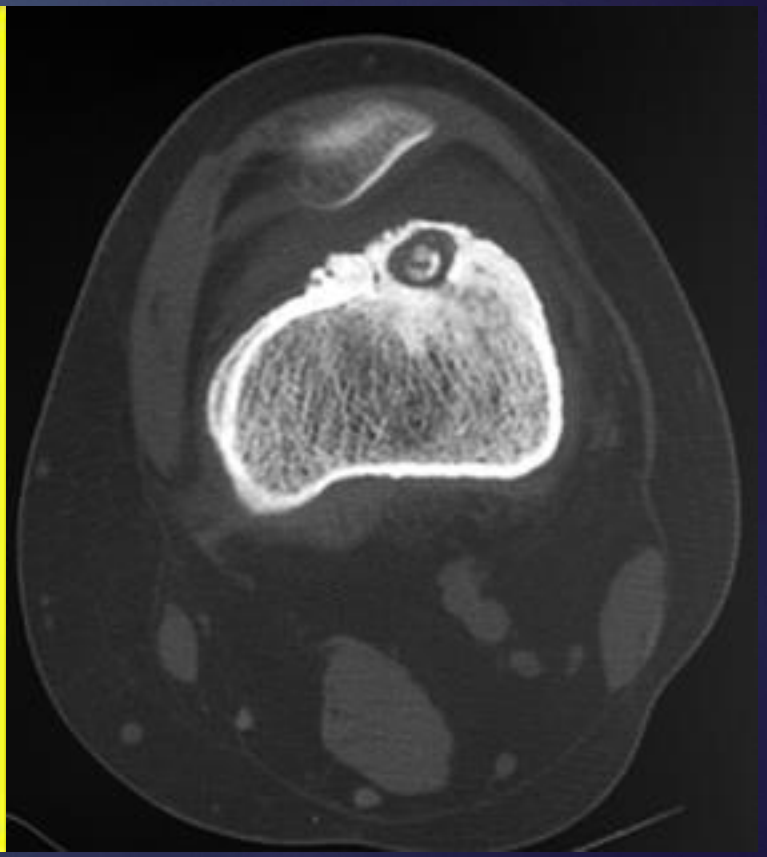
  - pedunculated

- **Treatment**
  - **Observation alone .. If Asymptomatic**
  - **Operative .. If symptomatic or growing fast**

Osteoid osteoma

- A small, discrete, painful, benign bone lesion
- **Commonest** location
  - proximal femur > tibia diaphysis
    - usually within bone cortex
  - Spine .. Produce scoliosis
- Characterized by central nidus with surrounding sclerotic rim
- Pain is constant at night and relived with NSAIDS
- Radiographs
  - reactive bone around radiolucent nidus
- CT imaging is the study of choice

Osteoid osteoma



Osteoid osteoma

Management

- **clinical observation and NSAID administration**
- **percutaneous radiofrequency ablation**
- **surgical resection/curettage**
  - complete marginal resection of nidus (sclerotic bone is normal and can be left behind)

Non-ossifying Fibroma

- fibrogenic lesion /dysfunctional ossification
- Locations … metaphysis of long bones

- **Symptoms**
  - asymptomatic found incidentally
  - Or pathologic fracture

- **Radiographs is diagnostic**
  - metaphyseal cortical eccentric "bubbly" lytic sclerotic rim

- **Treatment**
  - **observation ..** most resolve spontaneously
  - **curettage and bone grafting ..** If symptomatic or at risk of fracture

Unicameral bone cyst
simple bone cyst

- A non-neoplastic, serous fluid-filled bone lesion / failure of bone formation
- usually found in the metaphysis of long bones in young patients <20 years
- found in the
  - proximal humerus
  - Proximal femur
  - Distal tibia and radius
- Symptoms
  - most asymptomatic unless fracture occurs (usually with minor trauma)
  - pathologic fracture in ~50%

Unicameral bone cyst (simple bone cyst)

- Radiographs
  - central, lytic, well-demarcated metaphyseal lesion
  - thinning of cortices

- **Treatment**
  - **Observation if at low risk of fracture**
  - **aspiration/methylprednisolone injection**
  - **curettage and bone grafting +/- internal fixatio location**

Giant cell tumor

- A **benign** aggressive tumor found in the metaphysis of long bones in **mature** adults
- distal femur > proximal tibia > distal radius
- **Clinical features**
  - pain in the involved joint
  - palpable mass
- **Radiograph**

eccentric lytic epiphyseal/metaphyseal lesion extends subchondral bone

- Chest radiograph or chest CT .. 5% pulmonary metastsasis
- Bone scan is very hot
- MRI ,… signal change

Giant cell tumor

Giant cell tumor

Treatment

- **medical management ??? New modality ..**
  - bisphosphonates
  - denosumab

- **Operative**
  - **extensive curettage and reconstruction (with adjuvant treatment)**
  - 10-30% recurrence with curettage alone verses 3% with adjuvant treatment (phenol , hydrogen peroxide , argon llaser ..)

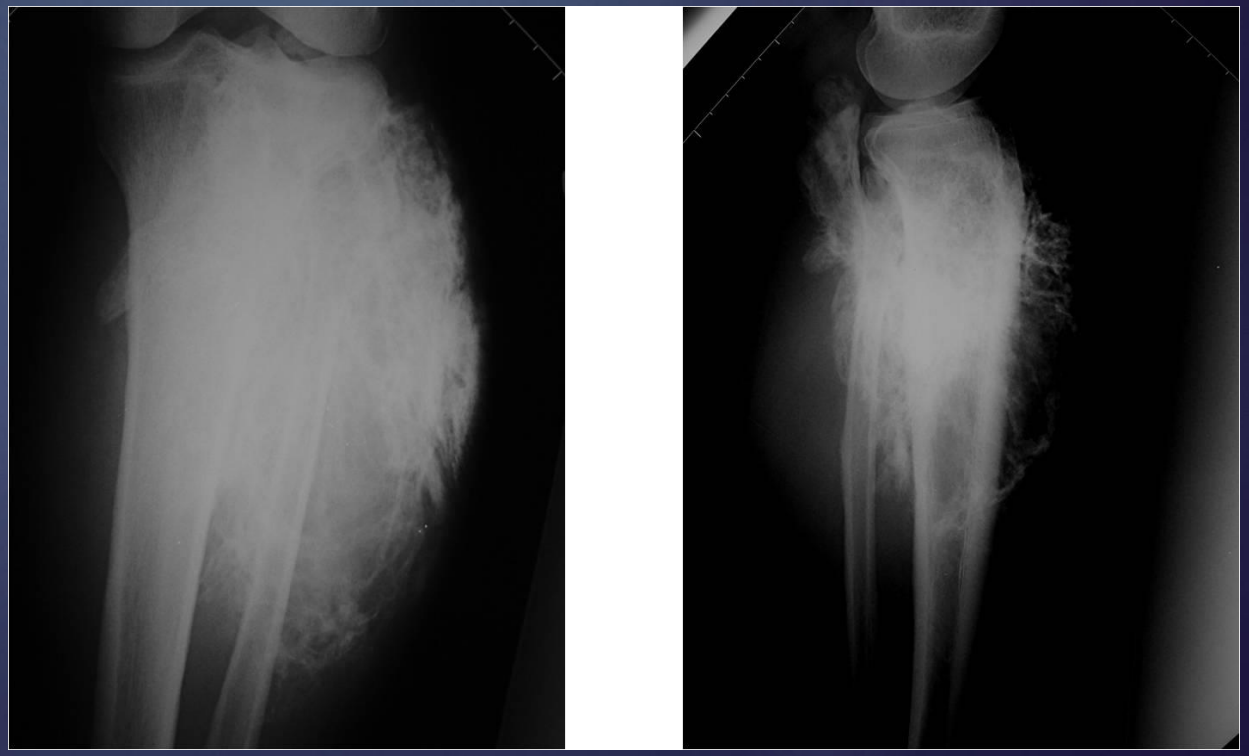
Malignant Bone Tumors

Osteosarcoma

- the most common primary sarcoma of bone
- in children and young adults <25 years
- common site / distal femur & proximal tibia
- commonly diagnosed at Stage IIB (high grade, extra-compartmental, no metastases)
- 10-20% of patients has pulmonary metastases
- **Presentation**
  - rapidly progressive pain, fever, and swelling
  - may feel mass on examination
- Radiographs
  - mixed blastic and destructive lesion
  - sun-burst or hair on end pattern
  - periosteal reaction (Codman's triangle)

Osteosarcoma

- MRI must include entire involved bone to determine
  - soft tissue
  - neurovascular involvement
  - skip metastases in same bone
- Bone scan
- chest Ct for metastasis



Osteosarcoma

Treatment

- **multi-agent chemotherapy and limb salvage resection**
  - preoperative chemotherapy given for 8-12 weeks followed by …
  - resection then …
  - maintenance chemotherapy for 6-12 months after surgical resection

- Prognosis 76% long-term survival with modern treatment.

Ewing's Sarcoma

- typically from 5-25 years of age
- second common malignant bone tumor in children
- ~50% are found in the diaphysis of long bones
- Genetics t(11:22) translocation in all cases
- **Presentation**
  - pain with fever
  - mimics an infection !!!!!
  - swelling and local tenderness
- **Radiographs**
  - destructive lesion in the diaphysis or metaphysis with a moth-eaten appearance
  - periosteal reaction give "onion skin" or "sunburst" appearance
- **MRI** .. soft-tissue extension and marrow involvement
- CT chest and bone scan for metastasis

Ewing's Sarcoma

Ewing's Sarcoma

Treatment

- **Neoadjuvant chemotherapy with limb salvage resection followed by postoperative chemotherapy**
- **the standard of therapy in most patients**
  - Neoadjuvant chemotherapy given for 8-12 weeks followed by surgical resection then maintenance chemotherapy for 6-12 months

- Prognosis
  - 60-70% long term survival with **isolated** extremity disease
  - 15% long term survival if patient presents with **metastatic** disease

Bone Metastasis / Secondary Bone Tumor

- most common malignancy of bone is metastatic disease
- metastatic lesions are usually found in older patients (> 40 years)
- carcinomas commonly spread to bone include (Breast, lung, thyroid, renal, prostate)
- common sites of metastatic lesions include spine> proximal femur> humerus
- Symptoms
  - pain
  - pathologic fracture
  - metastatic hypercalcemia

Evaluation of bone metastasis

Workup for older patient with bone lesion and unknown primary includes

- **Imagining**:
  - plain radiographs in two planes of affected limb
  - CT of chest / abdomen / pelvis
  - bone scan to detect extent of disease
- **Labs**
  - CBC , ESR
  - LFTs, Ca, Phos, alkaline phosphatase
  - serum and urine immuno-electrophoresis
- **biopsy** .. where a primary carcinoma is not identified, obtaining a biopsy is necessary to rule out a primary bone lesion.

Treatment of metastatic bone disease

- **Nonoperative** …
  - bisphosphonate therapy
  - chemotherapy, radiotherapy, and hormone therapy

- **Operative .. aim is not cure but to improve the quality of life !!!**
  - **stabilization of complete fracture with postoperative radiotherapy**
  - **prophylactic stabilization of impending fracture, postoperative radiation**