Bone Tumors

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

- **Classification:**

  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
<table>
<thead>
<tr>
<th>Cell type</th>
<th>Benign</th>
<th>Malignant</th>
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<tbody>
<tr>
<td>Chondrogenic</td>
<td>Osteochondroma</td>
<td>Chondrosarcoma</td>
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<td>Enchondroma</td>
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<td>Chondroblastoma</td>
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<td>Chondromyxoid fibroma</td>
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<td>Osteogenic</td>
<td>Osteoid osteoma</td>
<td>Osteosarcoma</td>
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<td>Osteoblastoma</td>
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<td>Histocytic</td>
<td>Fibrous histiocytoma</td>
<td>Malignant fibrous histiocytoma</td>
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<tr>
<td>Fibrogenic</td>
<td>fibrous cortical defect (non-ossifying fibroma), fibrous dysplasia, fibroma</td>
<td>Fibrosarcoma</td>
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<td>Vascular</td>
<td>Hemangioma</td>
<td>Angiosarcoma</td>
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<tr>
<td>Others</td>
<td>Giant cell tumor, aneurismal bone cyst, simple bone cyst</td>
<td>Malignant Giant cell tumor</td>
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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 movement 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
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
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:**

  **Stage 1:** low grade sarcomas
  - 1A: intracompartmental
  - 1B: extracompartmental

  **Stage 2:** high grade lesions.
  - 2A: intracompartmental
  - 2B: extracompartmental

  **Stage 3:** 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 it is 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 fixation based on tumor 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 metastasis
- 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