BENIGN BONE TUMORS

Dr Srinivas C H
M S Ortho, Fellowship Ortho Oncology,
Asst. Prof., Orthopaedic Oncosurgeon,
BGS Global Hospitals

Bone School @ Bangalore
Overview of today’s talk

- **Classification**
- Clinical Features
- Radiology: salient features
- Evaluation
- Management
What is expected of you in your exam

- GCT – Radius, Femur/Tibia
- Bone Cyst – Unicameral Bone cyst, ABC
- Chondroblastoma
- Osteochondroma / Exostosis
- Osteoid Osteoma
- Enchondroma

Bone School @ Bangalore
Case: Lytic lesion proximal Tibia
After 6 months of Rx

OSTEOSARCOMA

Now what ????

Bone School @ Bangalore
# W.H.O. Classification of Bone Tumours (2002- Revised)

## I. Osteogenic Tumours

### Benign
- **Osteoma**
- **Osteoid Osteoma**
- **Osteoblastoma**

### Malignant
- **Osteosarcoma**

## II. Cartilage Forming Tumours

### Benign
- **Chondroma**
- **Osteochondroma**
- **Chondromyxoid fibroma**
- **Chondroblastoma**

### Malignant
- **Chondrosarcoma**

---

Bone School @ Bangalore
III. FIBROGENIC

IV. FIBROHISTIOCYTIC
- Benign fibrous histiocytoma
- Malignant fibrous histiocytoma

V. NEUROECTODERMAL
- Ewing’s sarcoma

VI. HEMATOPOETIC
- Lymphoma
- Multiple Myeloma

VII. GCT

VIII. NOTOCHORDAL TUMOURS

IX. VASCULAR
- Benign
  - Hemangioma
  - Lymphangioma
  - Glomangioma
- Malignant
  - Angiosarcoma

XI. MISCELLANEOUS
- Bone cysts-simple or aneurysmal
- Fibrous dysplasia-mono or polystotic
- Reparative giant-cell granuloma (e.g. Epulis)
- Fibrous cortical defect
- Eosinophilic granuloma
- Non ossifying fibroma
- Osteitis fibrosa cystica (brown tumour)

Bone School @ Bangalore
ANATOMICAL LOCATION IN BONE
**Surgical Staging System for Musculoskeletal Tumors**  
(Enneking and MSTS)

**Benign:**
1. Latent  
2. Active  
3. Aggressive

<table>
<thead>
<tr>
<th>Stage</th>
<th>Grade</th>
<th>Tumor</th>
<th>Metastasis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Latent</td>
<td>G0</td>
<td>T0</td>
<td>M0</td>
</tr>
<tr>
<td>Active</td>
<td>G0</td>
<td>T0</td>
<td>M0</td>
</tr>
<tr>
<td>Aggressive</td>
<td>G0</td>
<td>T1-2</td>
<td>M0-1</td>
</tr>
</tbody>
</table>

**Site:**
- **T1:** Intracompartamental (Confined within limits of periosteum)
- **T2:** Extracompartamental (Breach in an adjacent joint cartilage, bone cortex or periosteum, fascia lata, quadriceps, and joint capsule)

**Metastasis:**
- **M0:** No identifiable skip lesions or distant metastases.
- **M1:** Any skip lesions, regional lymph nodes, or distant metastases.

**Discussion:**
- **Benign tumor staging uses Arabic numbers (1,2,3)**
- Malignant tumors identified with Roman numerals and a letter (Ia, Ib, IIa, IIb, IIIa, IIIb)
STAGING- Enneking

Stage-1 (latent stage)

- remains **static or heals spontaneously**  Eg: NOF
- intra capsular
- no growth
- well defined margins
- thick reactive bone
- not expanding cortex
Stage-2 (active)

- progressive growth but limited by natural barriers Eg: Simple bone cyst
- intra capsular
- actively growing
- well defined margins
- thin rim of reactive bone
- cortical expansion with thinning
Stage 3 (aggressive)

- Progressive growth not limited by natural barriers
  eg: GCT
- Extracapsular
- Break through reactive bone/cortex
GEOGRAPHIC PATTERN

- Least aggressive
- Narrow zone of transition
- Benign
MOTH EATEN APPEARANCE

- Multiple holes 2-5mm
- Wide zone of transition
- More aggressive

Bone School @ Bangalore
PERMEATIVE

- Multiple tiny holes <1mm
- Wide zone of transition
- Aggressive type

Bone School @ Bangalore
OSTEOMA

- Benign bony outgrowth of membranous bones.
- Multiple osteomas are associated with Gardner's syndrome

Highest incidence in the sixth decade
Male: female is 3:1
Asymptomatic
Excision if symptomatic

Bone School @ Bangalore
OSTEOID OSTEOMA

• Commonest benign osseous tumour
• Common in 1\textsuperscript{st} & 2\textsuperscript{nd} decade of life
• 10% of all benign bone tumours
• M:F – 2:1
• SITE: diaphysis, metaphysis of long bones
OSTEOID OSTEOMA

CLINICAL FEATURES
- Dull pain, worse at night (night cries) & responds to salicylates (aspirin)
- Swelling uncommon
- Tenderness

RADIOLOGICAL FEATURES
- A sharp round or oval lesion.
- Less than 2 cm in diameter.
- Radiolucent nidus surrounded by reactive sclerosis
- Nidus- osteolytic/partially/entirely calcified
Investigations

**CT SCAN:**
Nidus is best localized with CT (1 mm cuts)
Bull’s eye lesion

**BONE SCAN: Tc99**
- Due to intense radioisotope uptake by nidus and decreased uptake by surrounding sclerotic bone, a **double density image** is created that is typical of osteoid osteoma.

Headlight in fog appearance
Management

Course: Self limiting
On maturation, ossify and merge with surrounding bone
No reports of malignant transformation till date

Treatment:
- Conservative - not recommended because of severity of pain
- Surgical: En Bloc resection, Burr down
- Percutaneous radiofrequency ablation (PRA)
OSTEOBLASTOMA

- Benign osseous tumour similar to osteoid osteoma
- Progressive growth, absence of reactive perifocal bone formation
- 2\textsuperscript{nd}, 3\textsuperscript{rd} decade.
- M > F
- Sites: vertebrae- posterior elements
- Pain, long duration
- CT scan- ‘cotton wool’ if calcified
ENCHONDROMA

AGE: Most common between 2\textsuperscript{nd} & 4\textsuperscript{th} decades

SITES: Short tubular bones of hand (phalanges and metacarpals), followed by femur, humerus and ribs

RADIOGRAPHS: well circumscribed distinct area of rarefaction, expands the cortex

Calcification in older lesions - spotty/punctate
Management

- Asymptomatic lesions - follow-up with serial radiographs
- Symptomatic – PET Scan or biopsy to r/o any malignancy
- Curettage and bone grafting
- Wide excision to avoid recurrence
- Pathologic fractures are allowed to heal with closed treatment, curettage and bone grafting is then required after fracture healing.
OLLIERS DISEASE

• Multiple enchondromatosis
• Non-hereditary disorder common in children
• Affects metaphysis of long bones
• Presentation – bony swellings leads to thickening and shortening and deformities

MAFFUCCI SYNDROME

• hereditary familial disease
• multiple enchondroma and cavernous haemangioma
Osteochondroma

Also known as: Osteocartilaginous Exostosis

- Cartilage capped bony projection on external surface of bone.
- Commonest benign tumour of bone.
- Lesion has its own growth plate, usually stops growing at skeletal maturity.

AGE GROUPS: first two decades

SEX PREDILECTION: M:F-1.5:1
SITES OF PREDILECTION: Around the knee(40%) and proximal humerus.

LOCATION: Metaphysis/diaphysis of long bones.

- Asymptomatic
- Pain: Mechanical, Ischemic necrosis, Perilesional bursitis, Fracture of stalk, Malignancy <1%
Osteochondroma
Osteochondroma

- Growth disturbance of the extremity
- Block to joint motion
- No growth after skeletal maturity

**X RAY**
- Pedunculated / sessile – exophytic
- Metaphysis / diaphysis
- Marrow and cortices of lesion continuous with bone
- Directed away from growing end
- Cartilage cap not seen on x ray
PATHOGENESIS:
• Herniation of a fragment of growth plate through periosteal bone cuff
• Misdirected growth of that portion of physis

HISTOLOGY:
• Cartilage cap resembles layers of the normal growth plate
• The cartilage is more disorganized than normal
• Binucleate chondrocytes in lacunae
• Covered with a thin layer of periosteum.
• Cartilage cap 1-3 mm thick, thicker in children

**SIGNS OF MALIGNANCY:**
• >2 cm after skeletal maturity indicates possible malignant transformation
• Growth spurt of lesion beyond skeletal maturity
• Development of soft tissue with calcifications
• Dispersed calcifications within the cartilaginous cap
Osteochondroma with Chondrosarcoma

Thickened Cartilage Cap
Treatment

- Observe
- Delay excision until later adolescence
  - Pain, deformity, nerve palsy, movement restriction
HEREDITARY MULTIPLE EXOSTOSES (H.M.E)

- Also known as: Multiple Exostoses, Diaphyseal aclasis

- **Autosomal dominant** hereditary disorder, 10% no family history. EXT1,2,3 genes

- Knees, ankles and shoulders are most frequently affected.
- Knobby appearance, Short stature
- Forearm deformity, Tibio-fibular synostosis, Genu valgum, Coxa valga

**Rx** - Excision of symptomatic exostosis
Correction of deformity and limb length discrepancy
CHONDROBLASTOMA

ALSO KNOWN AS: Codman’s Tumour
Arises from immature cells of epiphyseal cartilage

AGE GROUPS: 10 to 20 yrs.

SEX PREDILECTION: Males more affected than females.

SITES OF PREDILECTION: Proximal part of the tibia, proximal part of the humerus and femur.

LOCATION: Epiphysis/ apophysis

SYMPTOMS: Pain and local swelling of joint without h/o trauma
RADIOGRAPHS:

- Well defined oval lytic lesion
- Sclerotic margin
- Epiphysis
- Eccentric
- Cottonwool calcification
- HPE: Chicken-wire calcification
CHONDROMYXOID FIBROMA

- Rare benign bone tumour (<1%)
- Chondroid tumour with myxoid and fibrous elements

AGE GROUPS: Adolescents and young adults.

SEX PREDILECTION: Males more affected than females (2:1).

SITES OF PREDILECTION: Lower extremities usually proximal tibia.

LOCATION: Metaphysis.

SYMPTOMS: Peripherally located mass with local pain and swelling.
**SIMPLE BONE CYST**

**Etiology:** Unknown

**C/F:** 3-14 yrs, 2:1 male predominance. Silent until pathological fracture occurs.

**Location:** 75% - humerus and proximal femur.

**Xray** - *Fallen-leaf or Fallen-fragment sign*

**Histology:** cyst filled with clear yellowish fluid, wall lined with fibrous tissue & hemosiderin.

**Rx:** Path # - Immobilize Drainage, Steroid injection
SBC of humerus (truncated cone appearance)

SBC with pathological fracture
X ray of 6 yr old boy showing SBC with path #

Bone School @ Bangalore
Steroid injection in the cyst
ANEURYSMAL BONE CYST:
expansile, blood filled cystic cavity.

Etiology:

- Primary ABC (65-99%): Unknown
- Secondary ABC (1-35%): preexisting bone
C/F:

- 5-20 yrs, 60% in females.
- Presents with pain at the site.
- 80% of lesions – long bone like femur and tibia.
- Spinal lesions affect the neural arch, spinous process, transverse process, and lamina. The thoracic and lumbar spine are the common regions.

**Histology:** ABC consists of multiple blood filled sinusoid spaces. The solid, numerous multinucleated giant cells.
**CT scan features:** “blood filled sponge”, fluid levels due to sedimentation of blood.

**MRI:** Multiple cysts: Fluid – fluid levels

**Nuclear study:** “donut sign” i.e. peripheral increased uptake.

**Angiography:** hypervascularity in the periphery of the lesion.

**Rx:** Surgical curettage with bone grafting.

Recurrence rate is high
NONOSSIFYING FIBROMA

• Solitary
• Eccentric
• Radiolucent, ovoid, bubbly
• Metaphysis
FIBROUS DYSPLASIA

- Developmental anomaly of bone formation.

**C/F** : 3-15 yrs, M:F 1:1, Bowing deformities and pathologic fractures, Café-au-lait spots are present in 30% of patients.

**Monostotic fibrous dysplasia** (solitary lesion): 70-80%

**Polyostotic fibrous dysplasia** (multiple bones): 20-30%

**McCune Albright syndrome:**

Polyostotic fibrous dysplasia, Endocrine dysfunction: precocious puberty, hyperthyroidism, Café-au-lait spots (coast of Maine)
Fibrous dysplasia with Shepherd’s crook deformity and pathological fracture
GIANT CELL TUMOUR (OSTEOCLASTOMA)

INCIDENCE: 5% of biopsied primary bone tumors.

CLINICAL FEATURES: 5Es- Elderly

- Epiphysis
- Eccentric
- Expansive
- Egg shell crackling
Campanacci Grading:
Grade I: Tumour associated with well defined margins and thin rim of mature bone

Grade II: Tumour is well defined but has no radiopaque rim

Grade III: Tumour has fuzzy borders
Campanacci Grading:

Grade I

Grade II

Grade III

Bone School @ Bangalore
Radiological Signs

- Lytic lesion
- Epiphysis
- Narrow zone of transition
- Thinning of cortex
- Honey comb appearance
- Soap bubble appearance
**DD:** GCT has to be differentiated from **giant cell variants.**

- Unicameral bone cyst
- Aneurysmal bone cyst
- Non-ossifying fibroma
- Chondroblastoma
- GCT of hyperparathyroidism (Brown tumor)
Treatment:

- **Curettage** and bone grating
- **Curettage** and placement of bone cement
- **Curettage** and cryosurgery
- Enbloc resection: Endoprosthesis, Arthrodesis
- Angioembolization
- Zolindronic acid

Follow-up:

Follow-up examination is essential for at least 5 years.
Biopsy

- Needle True Cut Biopsy
- Image Guided
- Open Biopsy
When to do what?

Bone School @ Bangalore
When to do what?
Curettage

High speed Burr

Adjuvant Therapy

Bone School @ Bangalore
T - Construct

Auto fibula
Iliac crest

Bone School @ Bangalore
< 2 mm

Autograft Iliac crest

Morcellised graft

Fibula