Bone Tumors 101

Dr. N Colterjhon
Dr. W Husain
Keep It Simple and Basic

H History

P Physical Examination

P Plain X rays
History

Age & location
Circumstances of presentation
Symptoms (constitutional)
Associated Conditions
ROS
Natural History/prior investigations
Gait, inspection, body proportion & symmetry

Medical:
- Organs: Lungs, Breast, Thyroid, Prostate, Kidneys (PTBLK)
- Lymphatic
- Other masses
- Skin
Physical Examination
Site of Interest

Location
Superficial vs. deep
+/- tender
Soft vs. firm
Mobile vs. fixed
+/- vascular
Neural (Tinel)
Plain X rays

**SEVEN**

Location
Size
What’s it doing to bone
Host bone response
What type of matrix is being made
Cortical changes
Soft tissue mass
Location

Location will give you a hint

Type of bone
Long bone
centered - cortex or medulla;
epiphysis, metaphysis or diaphysis

Some tumors almost exclusively
occur at specific sites
In general

The larger the lesion the more likely it is to be aggressive or malignant.
The bigger the uglier
What’s it doing to bone?

Geographic

Key word in this question
Define the margin or interface between the host bone and the lesion

Narrow zone of transition “lesion is clear”

Wide zone of transition “difficult to be certain where the lesion starts and stops”
What is the Host bone response?

*Bone often responds to lesions by making new bone*

- Marginal sclerosis (o.o.)
- Periosteal new bone formation
- Orderly periosteal new bone formation, remodelling
- Periosteal newcortical response
- Poorly organized periosteal new bone (host vs. tumor)
AGGRESSIVE LYTIC LESION

INACTIVE RADIOLYTIC LESION

ACTIVE LYTIC LESION

AGGRESSIVE LYTIC LESION
Is the cortex eroded?

Cortical erosion (by the tumor) vs. remodeling (host response) is the hallmark of

Active
Aggressive
malignant
Is the lesion making matrix?

Matrix is the internal tissue of the tumor
Soft tissue matrix
  Most tumor have soft tissue matrix
Radiolucent (lytic) on X-ray
Cartilage matrix
  Calcified rings, arcs, dots
Ossific matrix
  Bone forming
BONE FORMING

OSTEOID OSTEOMA
OSTEOBLASTOMA
OSTEOSARCOMA
CARTILAGE FORMING

OSTEOCHONDROMA
CHONDROBLASTOMA
CHONDROMYXOID FIBROMA
CHONDROMA
CHONDROSARCOMA
NON MATRIX TUMORS

DESMOPLASTIC FIBROMA
GIANT CELL TUMORS OF BONE
EWING’S SARCOMA
LYMPHOMA
MYELOMA
CHORDOMA
ADAMANTINOMA
ANGIOSARCOMA
HEMANGIOENDOTHELIOMA
HEMANGIOMA
Soft tissue mass

How to discriminate between aggressive and malignant tumors?

Aggressive tumors develop there soft tissue components by destroying the cortex.

Malignant tumors extend by
1- Destroying the cortex
2- Directly through the haversian canal system leaving the cortex structurally intact
The Good the Bad the Ugly
Classify the Lesion

Benign: incidental
Benign: active
Benign: aggressive
The scary

Malignant

1- primary mesenchymal cell origin
   osteosarcoma, chondrosarcoma, fibrosarcoma

2- marrow element origin
   myeloma, lymphoma, leukemia

3- metastatic
   thyroid, breast, prostate, lung, renal, melanoma
Pearls of tumor rotation

Don’t ever look at

MRI or CT scan

Before plain X-rays
The differential diagnosis

Diaphyseal intramedullary lesions

- Ewing's sarcoma
- Lymphoma
- Myeloma
- Fibrous dysplasia
- Enchondroma
The differential diagnosis

Metaphyseal lesions centered in the cortex

- Non-ossifying fibroma (NOF)
- Osteoid osteoma
Diaphyseal lesions centered in the cortex

Adamantinoma

Osteoid osteoma
The differential diagnosis

Metaphyseal intramedullary lesions

- Osteosarcoma
- Chondrosarcoma
- Fibrosarcoma
- Osteoblastoma
- Enchondroma,
- Fibrous dysplasia
- Simple bone cyst
- Aneurysmal bone cyst
The differential diagnosis

- Metaphyseal exostosis
- Osteochondroma
The differential diagnosis

Epiphyseal lesions

Chondroblastoma (Ch)

Chondroblastoma is a rare tumor seen in children and adolescents with open growth plates
The differential diagnosis

Epiphyseal lesions:

Giant Cell Tumor (GCT)

GCT is the most common tumor of epiphyses in skeletally mature individuals with closed growth plates.

GCT often shows metaphyseal extension.
What Is Your Plan Next

Observation
Local staging
Systemic staging
Observation

- Reassurance
- Serial radiographs
- Clinical response (natural History)
- Review other sites
Local Staging

CT

MRI
### MRI Appearance of Tissues

<table>
<thead>
<tr>
<th>Tissue</th>
<th>T1 Appearance</th>
<th>T2 Appearance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Water</td>
<td>dark</td>
<td>bright</td>
</tr>
<tr>
<td>Air</td>
<td>dark</td>
<td>dark</td>
</tr>
<tr>
<td>Collagen</td>
<td>dark</td>
<td>dark</td>
</tr>
<tr>
<td>Fat</td>
<td>bright</td>
<td>grey</td>
</tr>
</tbody>
</table>

- TR < 0.5 = T1
- TR > 2.0 = T2
Systemic and local staging

Nuclear scan; Tc 99, indium, gallium systemic and local

PET scan
Systemic Staging

- Blood work
- Nuclear Scan
- U/S, Chest x-ray, CT
- Mammography
Enneking Staging

**Stage 0** benign (latent, active, aggressive); no risk of metastasis

**Stage I** low grade malignant; <15% risk of metastasis
- **IA** = intracompartmental
- **IB** = extracompartmental

**Stage II** high grade malignant; >15% risk of metastasis
- **IIA** = intracompartmental
- **IIB** = extracompartmental

**Stage III** established metastases
Most primary malignant bone tumors should be referred to orthopaedic tumor specialist for biopsy and definitive management. Perform local staging first to identify easiest site for biopsy.
BIOPSY PRINCIPLES

Closed

*Fine needle aspirate*
not good for sarcomas and primary bone tumors of mesenchymal cell origin
acceptable for cystic lesions (fluid sent for cytology) and hematologic tumors

*True-Cut/core needle biopsy*
good for soft tissue lesions
not useful for bone lesions lacking a soft tissue mass
BIOPSY PRINCIPLES

Open Biopsy

Discuss and plan with MSK oncologist
Ask pathologist in advance if special medium needed

Send intraoperative tissue samples for:
1 - frozen section – confirm adequate tissue
2 - gram stain, AFB
3 - C&S – aerobic/anaerobic, fungal, TB
4 - definitive pathology – histologic analysis, immuno-staining, DNA markers
BIOPSY PRINCIPLES

Use tourniquet but NOT Esmarch exsanguination – deflate tourniquet prior to closure to ensure hemostasis

Approach soft tissue mass through compartment that it already involves

Longitudinal incision

Direct dissection down to lesion – do NOT develop soft tissue planes

Avoid neurovascular structures

Meticulous hemostasis – electrocautery, gel-foam and cement

Use drain if necessary – exit through skin in line with and close to end of incision
Prevent pathologic fractures

Cement augmentation
Make oval hole in bone oriented to long axis
Postop activity restrictions
Pearls of tumor rotation

Biopsy should be done in consultation with a tumor surgeon.
Tissue compartments must not be contaminated.
Biopsy all cultures and culture all biopsies.
Tumor histology

What to ask to reach a diagnosis

1. Are the cells round or spindle shaped?
2. Is there a matrix?
3. Is the matrix osseous or cartilaginous?
Tumor histology

Cell Shape

Round Cells
Lymphoma
Ewings
Primitive Neuroectodermal

Spindle cells

NO Matrix Present
Fibrosarcoma
Malignant fibrous histiocytoma

Matrix Present
Cartilaginous Matrix
Chondrosarcoma
chondroma

Osseous Matrix
Osteosarcoma
Osteoma
Osteoid Osteoma

1. Where? Long bones
2. How large? <1 cm
3. What is tumor doing? Radiolucent nidus
4. What is bone doing? Sclerotic reaction around nidus
5. Matrix? No/ may calcified centrally
6. Cortex? Intact
7. Soft tissue? No

Benign active osteoblastic lesion
Osteoid Osteoma

Pathology
Small well circumscribed red nidus
Osteoid with mature trabeculae of bone in fibrovascular stroma
More mature bone toward periphery of lesion
Osteoblastoma

1. Where? Spine, femur, tibia and skull
2. How large? > 2cm
3. What is tumor doing? Sclerotic, lucent or mixed
4. What is bone doing? Sclerotic rim but less reactive than (o.o)
5. Matrix? Osteoid matrix
6. Cortex? Rim of neocortex
7. Soft tissue? No

Benign aggressive osteoblastic lesion
Osteoblastoma

Pathology
Interconnected trabeculae in fibrovascular stroma
Benign cells forming bone
Osteochondroma

1. Where? Metaphysis around knee, shoulder and hip flat bones pelvis and scapula
2. How large? Sessile or pedunculated
3. What is tumor doing? Bony base with cartilage cap
4. What is bone doing? Remodeling
5. Matrix? Cartilage matrix
6. Cortex? Surface lesion
7. Soft tissue? Can have a bursa

Benign latent lesion
Osteochondroma

Bone scan if remodeling hot

Pathology

Exophytic lesion with cartilage cap that matures into trabecular bone

Marrow is contiguous
Nonossifying Fibroma

1. Where? Metaphysis
2. How large? > 4 cm
3. What is tumor doing? Radiolucent
4. What is bone doing? Thin sclerotic margin
5. Matrix? Non
6. Cortex? Expanded and thinned
7. Soft tissue? no

Benign latent metaphyseal lesion

Pathology

Spindle cell
Giant cells
Foamy histiocytes
Aneurysmal Bone Cyst

1. Where? Metaphyseal can extend to epiphysis after maturity
2. How large? >5cm
3. What is tumor doing? Lytic, expansile, geographic and multilobulated
4. What is bone doing? Sclerotic rim
5. Matrix? No
6. Cortex? Erosions
7. Soft tissue? No

Benign aggressive lytic lesion
Aneurysmal Bone Cyst

Pathology

Gross hemorrhagic and cystic tissue
Blood filled spaces with **no** endothelial cell lining
Fibrous tissue and giant cells
Unicamiral Bone Cyst

1. Where? Central medullary metaphyseal migrates into diaphyseal 80% proximal humerus and proximal femur
2. How large? Can be very large
3. What is tumor doing? Lytic geographic fallen leaf sign
4. What is bone doing? Sclerotic rim
5. Matrix? No
6. Cortex? Thin
7. Soft tissue? No

Benign latent to active lytic lesion
Unicamiral Bone Cyst

Pathology
Fluid filled cyst
Aspiration yellow-green
Fibrous tissue & cyst wall
1. Where? Distal femur, proximal tibia, distal radias, proximal humerus, anterior column and sacrum, 75% epiphysis/metaphysic of long bones, eccentric and central in thin tubular bones.

2. Size? At least 0.5 of the diameter.

3. What is tumor doing? Lytic with geographic border.

4. What is bone doing? Expansile with poorly outlined periosteal reaction.

Giant cell tumor

6. Cortex? Cortical break through may be present
7. Soft tissue? May be present. 20% of lesions invade the cortex and break through and yield limited soft tissue

Benign aggressive lesion
Pathology
Sheets of multinucleated giant cells intermixed with oval mononuclear stromal cells
Spindle shaped but no atipia
Mitosis may occur but no clinical significance
Small amount of bone or osteoid may be seen up to 30%
**Osteosarcoma**

1. Where? Peri knee > proximal humerus > flat bones > epiphyseal-metaphyseal > diaphyseal
2. How large? 5cm generally large
3. What is tumor doing? Lysis with sclerosis but non geographic
4. What is bone doing? Onion skinning, sunburst
5. Matrix? Osteoid
6. Cortex? Cortical erosion
7. Soft tissue? Ossification

**Malignant primary bone lesion**
Osteosarcoma

Pathology
Malignant cells producing osteoid in lace or sheet like pattern
Chondrosarcoma

1. Where? Proximal & distal femur, proximal humerus, scapula, pelvis, ribs, sternum
2. How large? >5cm
3. What is tumor doing? Geographic with some permeative changes
4. What is bone doing? Endosteal expansion with thickened cortex
5. Matrix? Arcs and rings punctuated calcification
6. Cortex? Thinning and cortical destruction
7. Soft tissue? The matrix in soft tissue

Malignant cartilage producing lesion
Chondrosarcoma

Pathology
Myxoid or chondroid with increased cellularity and binucleate
The degree of cellularity, mitoses and cytologic atypia determines the grade of the tumor
1. Where? Pelvis and lower extremities
2. Size? Usually large >5 cm
3. What is tumor doing? Diffuse and permeative destruction of bone
4. What is bone doing? Poorly defined margins, moth-eaten bone destruction, onion skinning periosteal response
5. Matrix? No
Ewing’s sarcoma

6. Cortex ? Rapid growth of tumor through the haversian canals of the intact cortex which will give the sunburst or spiculated pattern

7. Soft tissue ? Large non-calcified, occasionally the bone lesion is imperceptible and the soft tissue mass can be the predominant radiographic finding

Malignant lesion

Pathology

Sheets or nests of small round cells separated by septa of fibrous tissue, cells may contain glycogen
Myeloma

1. Where? Vert bodies, ribs, skull, pelvis, femur > tibia > radius > humerus
2. Size? Variable
3. What is tumor doing? Permeative osteolytic destruction
4. What is bone doing? No scelerosis or periosteal reaction
5. Matrix? No
6. Cortex? Thinning
7. Soft tissue? May be present with cortical destruction

Malignant plasma cell tumor
Myeloma

Pathology
Infiltration by atypical plasma cells
Thank you very much

Wael M Husain