Bone Tumors

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Agenda

• Look at different types of lesions
  – Benign
  – Malignant
• Characteristics
• What you need to know
Which would you rather?

Benign lesions are slower growing, small, have narrow zone of transition

Malignant lesions tend to be larger, wide transition zone, cortical erosion (sunburst, Codman’s triangle, onion skinning)

WHAT MAKES IT BAD???

- P. Ferguson, OKU F&A 3
Generally

- There are over 50 types of osseous tumors
- About one half have been documented to effect the F&A
- Calcaneus and metatarsals are the most common sites
- M>F 2:1 and benign more common
- Metastatic more common than primary
- Malignant lesions more common in younger pts due to high bone turnover

Where do you start?
Patterns of destruction

- Geographic
  - Sharp borders
  - Narrow transition
  - Implies less aggressive
- Moth-eaten
  - More ragged
  - More rapid growth
  - Implies malignancy
- Permeative
  - Ill-defined
  - Wide zone of transition
  - Malignant

Periosteal reactions
Location of lesion - Axial

- Central
  - Enchondroma
- Eccentric
  - GCT
  - Osteosarcoma
- Cortical
  - NOF
  - Osteoid osteoma
- Parosteal
  - Osteochondroma
Location of lesion - Longitudinal

- Epiphyseal
  - Chondroblastoma
  - GCT (cross over)
- Metaphyseal
  - Osteosarcoma
  - Chondrosarcoma
- Diaphyseal
  - ABC, solitary cysts, NOF
  - Enchondroma

TUMOR GRADING/STAGING - AJCC STAGING PROTOCOL FOR SARCOMAS OF BONE

<table>
<thead>
<tr>
<th>STAGE</th>
<th>GRADE</th>
<th>PRIMARY TUMOR</th>
<th>REGIONAL NODES</th>
<th>DISTANT METS</th>
</tr>
</thead>
<tbody>
<tr>
<td>IA</td>
<td>Low</td>
<td>Intra-compartmental</td>
<td>N0</td>
<td>None</td>
</tr>
<tr>
<td>IB</td>
<td>Low</td>
<td>Extra-compartmental</td>
<td>N0</td>
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</tr>
<tr>
<td>IIA</td>
<td>High</td>
<td>Intra-compartmental</td>
<td>N0</td>
<td>None</td>
</tr>
<tr>
<td>IIB</td>
<td>High</td>
<td>Extra-compartmental</td>
<td>N0</td>
<td>None</td>
</tr>
<tr>
<td>III</td>
<td>High</td>
<td>Skip Mets</td>
<td>N0</td>
<td>None</td>
</tr>
<tr>
<td>IVA</td>
<td>Any</td>
<td>Any</td>
<td>N1</td>
<td>None</td>
</tr>
<tr>
<td>IVB</td>
<td>Any</td>
<td>Any</td>
<td>N0-1</td>
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</tbody>
</table>

American Joint Committee on Cancer,
### TUMOR GRADING/STAGING—ENNEKING PROTOCOL FOR STAGING OF BENIGN BONE LESIONS

<table>
<thead>
<tr>
<th>STAGE</th>
<th>DEFINITION</th>
<th>BEHAVIOR</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>Latent</td>
<td>Remains static or heals spontaneously</td>
</tr>
<tr>
<td>2</td>
<td>Active</td>
<td>Progressive growth, but limited by natural barriers</td>
</tr>
<tr>
<td>3</td>
<td>Benign Aggressive</td>
<td>Progressive growth NOT limited by natural barriers</td>
</tr>
</tbody>
</table>

**BENIGN BONE LESIONS**

- Non-Ossifying Fibroma (NOF)
- Osteochondroma
- Osteoid Osteoma
- Osteoblastoma
- Enchondroma
- Chondroblastoma
- Subungal Pathology
- Unicameral Bone Cyst (UBC)
- Aneurysmal Bone Cyst (ABC)
- Giant Cell Tumor Of Bone (GCT)
Non-Ossifying Fibroma (NOF)

- AKA metaphyseal fibrous defect, fibrous cortical defect, benign fibrous histiocytoma.
- May occur in ~1/3 of pts with open physes
  - Caffey et al, Advances in Pediatrics, 1955
- Location = distal femur > prox tibia > dist tibia > prox humerus > dist radius.

Non-Ossifying Fibroma (NOF)

- Cortical, becomes eccentric with increased size
- Lytic lesion
- Scalloped, sclerotic border
- Overlying cortex can be thinned but intact
- Lesion migrates toward diaphysis with age, becomes sclerotic
Non-Ossifying Fibroma (NOF)

• Histo=Fibroblastic stromal tissue
• Scattered giant cells

Non-Ossifying Fibroma (NOF)

• Tx = usually observation, may require curettage and grafting when big & painful
• Criteria for repair = over 43mm long, over 33mm wide, involves over 50% of cortex.
  – Frassica, Ortho Boards Review, 2007
Osteochondroma

- Surface lesion which is continuous with medullary cavity
- Bone produced by progressive ossification of a cartilage cap
- Most are solitary lesions which parallel Pt's growth, stop when physes close
- Grow away from physes
- Overall most common benign tumor

Osteochondroma

Medullary Blending
Osteochondroma

- Histo of cartilage cap looks like a physis with columns of chondrocytes

- Tx = exostectomy if lesion becomes prominent, interferes with joint function
- Multiple hereditary osteochondromatosis also known as multiple exostoses, diaphyseal acalasia
- Osteochondromas tend to be big, sessile, have big (over 10mm) cartilage caps
- Malignant transformation with solitary osteochondroma less than 1%, for Mult Exostoses 5-25% (10%). Transform into low grade chondrosarcomas
- Malignant transformation heralded by new pain, sudden increase in size, heterogeneity on X-ray, calcified ST masses, bony destruction
Osteoid Osteoma

- Benign bone-forming lesion
- **Size less than 2 cm**
- Has limited growth potential
- Most common location=prox femur
- May occur in any bone of the foot but is most common in the tarsal bones
- Most cases before the 3rd decade
- Presents with night pain unrelated to activity, reliably relieved with NSAID’s

Osteoid Osteoma

- **Nidus** typically intracortical, 5-10mm, surrounded by sclerotic bone
- CT better than MR for evaluation
Osteoid osteoma

- Nidus is interlacing network of fibrovascular stroma and haphazard osteoid trabeculae mineralized in the center
- Tx = RFA, NSAIDs for up to 2y (50% effective), surgical removal with “burr down” technique
Osteoblastoma

- Benign bone forming tumor with marked growth potential
- 2nd decade (avg age=22.5)
- Presents with pain, but not nocturnal, NOT relieved by NSAIDs
- Rare tumor but foot is 3rd most common site, usually talar neck

Osteoblastoma

- OB is arbitrarily designated by its larger size (over 2cm) compared with OO
- Radiographic appearance is variable, non-specific, expansile
- 25% resemble malignancy
- Lytic with some surrounding sclerosis but lacks robust sclerotic bone of OO
Osteoblastoma

- Histologically similar to OO
- Irregular osteoid arranged haphazardly in loose, vascular connective tissue
- Tend to be more aggressive than OO
- Tx = curettage, graft or cement

Enchondroma

- Benign, inactive hyaline cartilage tumor
- Most common in small bones of the hand and foot
- Most common benign tumor of the foot
- Usually asymptomatic
- Incidental finding
**Enchondroma**

- Cortex is intact
- Radiographs show lucent areas with variable mineralization, rings, arcs, stipples
- May cause cortical expansion or FX

**Enchondroma**

- Hypocellular, abundant hyaline cartilage, absence of myxoid change, cellular atypia
- Tx=observation & serial X-rays (3m, 6m, 1y, yearly)
- Surgery when (1)-path FX or (2)-suspicious for low-grade chondrosarcc
- Tx=curettage, graft
**Enchondroma-Multiple Lesions**

- Multiple enchondromas have sporadic inheritance
- Ollier’s DZ has multiple enchondromas with deformity
- Maffucci’s Syn has multiple enchondromas, dysplasia, hemangiomas of skin and ST viscera
- Malignant transformation to secondary chondrosarcoma=1% per year in solitary DZ, 25% in Ollier’s DZ, 100% in Maffucci’s syndrome

**Subungual exostosis and osteochondroma**

- Bony growth protruding from dorsal tip of distal phalanx
- Histologically, subungal exostosis has a base or stalk of normal-appearing trabecular bone with a fibrocartilagenous cap
- Cap on osteochondroma is composed of hyaline cartilage
- Subungal osteochondroma shows classic medullary continuity
- Tx=complete surgical resection
Chondroblastoma

- Pain in 86%, also effusion, limp, joint stiffness
- **EPIPHYSEAL** lesion in skeletally immature Pts, most common in males in 2nd decade
- Mostly long bones, but talus & calcaneus also common locations

![Graph showing distribution of chondroblastomas according to age and sex of the patient and site of the lesion](image)
Chondroblastoma

- Radiographically lytic, 50% have sclerotic border, 60% in both epiphysis and metaphysis
- Little or no matrix, calcifications present in 1/3, can be very destructive
- Intense edema adjacent to lesion on MR

Chondroblastoma

- Proliferating cell is the chondroblast, ovoid cell with ovoid nucleus
- Histologically very cellular, field of mononuclear cells with fine “chicken wire” calcifications
Chondroblastoma

- Tx = curettage, BG or cement as necessary
- Local recurrence rate is ~10%
- Can develop occasional (1-2%) benign pulm mets.

Unicameral Bone Cyst (UBC)

- Common cystic lesion of skeletally immature Pts (Avg age=15)
- 2:1 M:F
- Prox humerus and prox femur most common (2/3)
- In series of 429 UBCs, only 18 (4%) in calcaneus
  - Hanna et al, F&A Int, 2004
- Most foot UBC’s are incidental
- FX is rare (~2%)
- In contrast to UBCs in long bones which tend to spontaneously involute, UBCs of the calcaneus tend to persist thru adulthood, usually without consequence
Unicameral Bone Cyst (UBC)

• Purely lucent, thin but intact cortex, metaphyseal expansion
• “Fallen fragment sign”

Unicameral Bone Cyst (UBC)

• Histo=thin fibrous membrane, giant cells, hemosiderin, chronic inflammatory cells
Unicameral Bone Cyst (UBC)

- Tx=aspiration & injection of steroid, injection of orthobiologic, curettage and bone graft
- If path FX, allow it to heal then graft
- Generally F&A lesions are asymptomatic and require no Tx

Aneurysmal Bone Cyst (ABC)

- Common benign aggressive cystic lesion of the young
- Often found with other neoplasia such as CGT, chondroblastoma, FD, CMF, osteosarcoma
- Avg age of ABC is 15y
  - Casadei, F&A Int, 1996
- Locations: distal femur/prox tibia, pelvis, spine, distal tibia, hands, feet.
- Most common site of ABC in F&A are metatarsals
Aneurysmal Bone Cyst (ABC)

- Metaphyseal, eccentric, lytic, can expand/destroy cortex, extends into ST’s with thin shell of reactive bone
- Typically has sharp, trabeculated margin and intralesional septae
- "Soap bubble"
- Fluid/fluid levels on MRI

Aneurysmal Bone Cyst (ABC)

- Histology=cavernous blood-filled spaces
- Cyst lining has giant cells, spindle cells, lacy new bone formation
- Tx=curettage, bone grafting
- Very high local recurrence rate in children (up to 50%)
Giant Cell Tumor Of Bone (GCT)

- Benign aggressive
- Occurs in Pts with closed physes
- F>M 20-40 yrs
- Pain, swelling, pathologic FX
- Most common about the knee, fairly common in distal tib, less common in the foot
- 19% of GCT’s are in tarsal bones

Giant Cell Tumor Of Bone (GCT)

- Eccentric, expansile, lucent metaphyseal lesion
- Adjacent to epiphyseal scar
- Matrix without mineralization
- Permeative, poorly marginated, purely lytic
- Typically, rim of periosteal new bone
- “Moth eaten”
Giant Cell Tumor Of Bone (GCT)

- Histologically, NUMEROUS giant cells are scattered uniformly throughout
- Stroma of mononuclear cells whose nuclei should be identical to giant cell nuclei

Tx=extended curettage if the joint can be spared, resection/recon if it cannot
- Extended curettage-intralesional curettage + adjuvant (cryotherapy, burring, argon laser, cement, phenol) and bone graft or cement
- Previous Tx c XRT is the factor most commonly associated with malignant transformation
- GCTs of the phalanges should be resected rather than curetted
Giant Cell Tumor Of Bone (GCT)

- Very locally aggressive, high (10-40%) local recurrence rate
- “Benign” lung mets in 2-5%.
- Mortality in benign metastasizing GCT is 10-20%, no standard Tx
- Can develop secondary sarcoma if multiple local recurrences or radiation Tx

MALIGNANT BONE LESIONS

- Chondrosarcoma
- Osteosarcoma
- Ewing’s Sarcoma
- Metastatic Carcinoma
Chondrosarcoma

- Most common bone sarcoma >20 years old, usually in Pts 50y +
- Primary complaint is a painful mass
- Most common locations in order=pelvis, proximal femur, shoulder girdle
- Rarely observed in F&A(1-5%)
- Most common malignant bone tumor in the foot
- In F&A, often secondary chondrosarcs (found in Pts with MHE or Ollier’s DZ)

- Bone destruction with calcification
- 75% mineralized with rings, arcs, stipples
- 84% cortical abnormalities, large erosions involving over 50% of cortex, expansile
- Tumors tend to be very large, very destructive, poorly marginated
- High on T2 MRI due to high H2O content in cartilage
Chondrosarcoma

- Solid chondroid matrix with myxoid change, broad spectrum of atypia
- Tx=wide excision
- Can use chemo in high-grade lesions but no benefit has been shown
- 10 year survival ~70% with adequate margins

Chondrosarcoma vs. Enchondroma

- Differentiating Clinical and Radiographic Features of Enchondroma and Secondary Chondrosarc of the Foot
  - Gajewski et al, F&A Int, 2006
- Reviewed AFIP records and found 14/340 foot secondary chondrosarcs (4.1%). 34 of 755 enchondromas were of the foot
- Poor (51%) intraobserver reliability
- Traditionally, chondrosarcomas have pain, deep endosteal scalloping, cortical destruction, ST mass, periosteal rxn, uptake on bone scan. No sig difference in these parameters in this study.
- Lesions of the midfoot and hindfoot tended to be malignant, and forefoot lesions tended to be enchondromas.
- Concern is greatest when size is over 5cm
Osteosarcoma

- AKA osteogenic sarcoma
- Most common primary bone tumor of the young
- 70-75% of Pts 10-25y/o
- CC=Painful mass
- Distal femur>proximal tibia>proximal humerus
- Most common location is knee (50-65%), followed by humerus and pelvis
- ~4% F&A
- Distal tib most common site in F&A, but MT and tarsal lesions have been reported

Osteosarcoma

- Metaphyseal lesion
- Poorly-defined, destructive bone-forming lesion on X-rays
- Periosteal rxn, ST mass common
Osteosarcoma

- Histology marked cellular atypia
- Immature woven bone (osteoid) by malignant cells

- Metastasis to lung common and reason for most deaths
- If caught early (2yrs), treated with excision and chemo, can be 60% survival
Ewing’s Sarcoma

• 1st described by Sir James Ewing in 1921
• 2nd most common bone malignancy of childhood
• Uncertain cell of origin
• Thought to be primitive neuroectodermal tumor
• Most common in pelvis, long bone diaphyses, ribs, spine
• 1.2-5% of Ewing’s sarcoma occur in the foot
• 10-15% path FX
• M>F, avg age ~12y (range=4-20)
• Painful mass, constitutional sx’s, night pain

Ewing’s Sarcoma

• Diaphyseal/meta-diaphyseal region
• Permeative pattern of destruction
• Thinned cortex, onion skinning, Codman’s triangle, ST mass
Ewing’s Sarcoma

- No osteoid formation
- Must be differentiated from round cell sarcomas ie leukemia, lymphoma, metastatic lesions with special stains

Almost done but remember...
Metastatic Carcinoma

- Metastatic lesions (PBKTL) are more common in older Pts and should be specifically investigated, esp with Hx of CA and a lesion
- Mets to the foot are rare, with lung CA being most common primary
- Lung CA is most common tumor to spread distal to knee
- Tx is to prevent FX or help with palliation

Metastasis

<table>
<thead>
<tr>
<th>Primary site</th>
<th>Metastases</th>
</tr>
</thead>
<tbody>
<tr>
<td>(Where the cancer starts)</td>
<td>(Where it often spreads to)</td>
</tr>
<tr>
<td>Breast</td>
<td>Lymph nodes (underarm), lung, bone, liver, brain</td>
</tr>
<tr>
<td>Colon and rectum</td>
<td>Lymph nodes (next to bowels), liver, lung, bone</td>
</tr>
<tr>
<td>Lung</td>
<td>Lymph nodes (next to lungs), other lung, adrenals,</td>
</tr>
<tr>
<td></td>
<td>liver, bone, brain</td>
</tr>
<tr>
<td>Ovary</td>
<td>Lymph nodes (in pelvis), liver, lung</td>
</tr>
<tr>
<td>Pancreas</td>
<td>Lymph nodes (in abdomen), liver, lung, bone, brain</td>
</tr>
<tr>
<td>Prostate</td>
<td>Lymph nodes (in pelvis), bone, lung, liver</td>
</tr>
<tr>
<td>Soft tissues (sarcomas)</td>
<td>Lungs, bone, lymph nodes, brain</td>
</tr>
<tr>
<td>Stomach</td>
<td>Lymph nodes (in abdomen), liver, lungs, brain</td>
</tr>
<tr>
<td>Thyroid</td>
<td>Lymph nodes (in neck), lungs, liver, bone</td>
</tr>
</tbody>
</table>
End thoughts

• Can seem overwhelming
• Remember the easy things
  – Location
  – Appearance
  – Unique clues
• Categorize and characterize – this will give you a good start
• Although not common, be on guard

Thank You