Primary Bone Malignancy: Common Neoplasms, Imaging Features, & Clinical Implications

Suzanne Shepherd

3 March 2011
Goals:

• Review criteria utilized in diagnosis of bone tumors
• Discuss role of multimodality imaging
  – ACR appropriateness criteria
  – Staging
• Discuss some of the most common malignant primary bone tumors and distinguishing features in deriving useful ddx
Primary Bone Malignancy

IMAGING CRITERIA
Characteristics in Diagnosis of Bone Tumors

- Patient age (#1 factor to consider) & gender
- Tumor location
- Lesion margin
- Matrix formation
- Periosteal reaction
## Generalizations by patient age

(adapted from Table 1, Miller TT. Rad 2008; 246:662-674)

<table>
<thead>
<tr>
<th>Age (yrs)</th>
<th>Benign</th>
<th>Malignant</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 20</td>
<td>Fibrous cortical defect/NOF</td>
<td>Leukemia</td>
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<tr>
<td></td>
<td>Simple bone cyst</td>
<td>Ewings sarcoma</td>
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<td></td>
<td>ABC</td>
<td>Osteosarcoma</td>
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<tr>
<td></td>
<td>Chondroblastoma</td>
<td>(conv, periost, telang)</td>
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<td></td>
<td>LCH</td>
<td>Mets (rare)</td>
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<td></td>
<td>Osteoblastoma</td>
<td>NBoma</td>
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<tr>
<td></td>
<td>Osteoid osteoma</td>
<td>RBoma</td>
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<tr>
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<td>Osteofibrous dysplasia</td>
<td>Rhabdomyosarcoma</td>
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<tr>
<td></td>
<td>Chondromyxoid fibroma</td>
<td>Hodgkin lymphoma</td>
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<td></td>
<td>FD</td>
<td></td>
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<tr>
<td></td>
<td>Enchondroma</td>
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<tr>
<td>20-40</td>
<td>Enchondroma</td>
<td>Osteosarcoma (parosteal)</td>
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<tr>
<td></td>
<td>GCT</td>
<td>Adamantinoma</td>
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<td></td>
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<td></td>
<td>Non-Hodgkin Lymphoma</td>
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<td>Chondrosarcoma</td>
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<tr>
<td></td>
<td>Malig fibrous histiocytoma</td>
<td></td>
</tr>
<tr>
<td></td>
<td>O-sarc (d/t Pagets or rad’n)</td>
<td></td>
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</table>
Generalizations by patient age & location

- **Location**
  - Flat vs. tubular bone
  - Longitudinal location:
    - Epiphyseal vs. metaphyseal vs. diaphyseal
  - Axial location:
    - Central vs. eccentric
    - Cortical vs. juxtacortical vs. soft tissue

(www.radiologyassistant.nl/en/494e15cbf0d8d)
Generalizations about Location

• Epiphyseal:
  – Subchondral cyst (sk mature, OA)
  – Chondroblastoma (sk immature)

• Metaphyseal: active area of bone formation
  – NOF
  – Osteochondroma
  – Sarcomas

• Diaphyseal:
  – Ewing sarcoma

Courtesy, George Nomikos
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Courtesy, George Nomikos
Pattern of Bone Destruction: Lesion Margin

• Type I: Geographic
  – A: Well-defined, with surrounding sclerosis
  – B: Well-defined, without surrounding sclerosis
  – C: Ill-defined

• Type II: Motheaten

• Type III: Permeative

Adapted from Murphey, AFIP notes 2009 p.812
Geographic Lesions, Type I

A  
NOF

B  
UBC

C  
GCT

Courtesy, George Nomikos
Margins reflect biological activity

• Nonaggressive
  – Geographic, well-defined lesion
  – Narrow zone of transition
  – Sclerotic lesion margins

• Aggressive
  – Permeative, moth-eaten
  – Wide zone of transition
  – Nonsclerotic lesion margins

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Tumor Matrix

• Osteoid
  – Ivory
  – Fluffy
  – Cloud-like

• Chondroid
  – Arcs/rings
  – Punctate
  – Stippled

• Fibrous
  – Ground glass
  – Hazy

Osteosarcoma

Courtesy, George Nomikos
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Courtesy, George Nomikos

Enchondroma
Tumor Matrix

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Fibrous dysplasia

Dr. Sartoris Teaching File
Periosteal Reaction

(www.radiologyassistant.nl/en/494e15cbf0d8d)
Periosteal Reaction

- Non-aggressive:
  - Solid
  - Buttressing
  - Expansion
  - Septation

- Aggressive:
  - Codman triangle
  - Sun-burst
  - Hair-on-end
  - Laminated

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Periosteal OGS

Courtesy, George Nomikos
Lesion Number

- Monostotic
- Polyostotic
  - Benign
    - LCH
    - Enchondromatosis
    - FD
  - Malignant
    - Mets/Myeloma

Solitary enchondroma

UCSD Thornton, Evelyn Fliszar
Lesion Number

- Monostotic
- Polyostotic
  - Benign
    - LCH
    - Enchondromatosis
    - FD
  - Malignant
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Multiple enchondromas

Georgetown University Hospital
Primary Bone Malignancy

ROLE OF MULTIMODALITY IMAGING
ACR Appropriateness Criteria for Evaluation of Bone Tumors

• **Radiographs** for initial evaluation of bone lesion

• Additional imaging dept on 1 of 4 conditions:
  
  – 1. nl XR, but pt has persistent sxs
    • Lytic lesions not seen on XR till 30-50% loss of mineralization
    • If pt can localize sxs, go to MR; if not, go to scintigraphy

  – 2. abnl XR, clinician suspects mets or MM on basis of hx, lab values or both
    • Next step, bone scan

  – 3. abnl XR, non-aggressive-appearing tumor

  – 4. abnl XR, aggressive-appearing primary bone tumor
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  – 1. nl XR, but pt has persistent sxs
    • Lytic lesions not seen on XR till 30-50% loss of mineralization
    • If pt can localize sxs, go to MR (r/o occult frx, infection, etc)
    • If not, go to scintigraphy
  – 2. abnl XR, clinician suspects mets or MM on basis of hx, lab values or both
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Modalities

- Conventional radiographs
  - Ddx is best derived from XR
- CT/MRI
  - Pre-operative assessment, biopsy, & staging
  - Further matrix characterization
  - ST component
- Bone scintigraphy
  - Degree of lesion radiotracer uptake
  - Lesion multiplicity

Courtesy, Evelyn Fliszlar
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Courtesy, Evelyn Fliszar
Staging

• 2 systems
  – Enneking 1\textsuperscript{st} in 1980 (3 criteria)
  – American Joint Committee on Cancer (AJCC) in 1983 and revised in 2003 (4 criteria)
    • Does NOT apply to lymphoma or myeloma
Enneking Staging System for Primary Malignant Bone Tumors

1. Tumor extent
   - T1: intracompartmental
   - T2: extracompartmental

2. Mets
   - M0: no mets
   - M1: + mets

3. Histologic grade
   - G1: low grade (<25% risk of mets)
   - G2: high grade (>25% risk of mets)

- INTRAcompartmental = entirely intraosseous or parosseous (ex: parosteal o-sarc)

Enneking Staging System for Primary Malignant Bone Tumors

- Based on 3 criteria

1. Tumor extent
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3. Histologic grade
   - G1: low grade (<25% risk of mets)
   - G2: high grade (>25% risk of mets)

- EXTRACompartmental = intraoss. w/ST-extension or parosseous w/ intraoss. or extrafascial extension (ex: parosteal o-sarc)

Enneking Staging System for Primary Malignant Bone Tumors

- Based on 3 criteria
  1. Tumor extent
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### TABLE 2: Enneking Staging System [14] for Primary Malignant Tumors of Bone

<table>
<thead>
<tr>
<th>Stage</th>
<th>Tumor</th>
<th>Metastases</th>
<th>Grade</th>
</tr>
</thead>
<tbody>
<tr>
<td>IA</td>
<td>T1</td>
<td>M0</td>
<td>G1</td>
</tr>
<tr>
<td>IB</td>
<td>T2</td>
<td>M0</td>
<td>G1</td>
</tr>
<tr>
<td>IIA</td>
<td>T1</td>
<td>M0</td>
<td>G2</td>
</tr>
<tr>
<td>IIB</td>
<td>T2</td>
<td>M0</td>
<td>G2</td>
</tr>
<tr>
<td>III</td>
<td>T1 or T2</td>
<td>M1</td>
<td>G1 or G2</td>
</tr>
</tbody>
</table>

Note—T1 = tumor is intracompartmental, T2 = tumor is extracompartmental, M0 = no regional or distant metastasis, M1 = regional or distant metastasis, G1 = low grade, G2 = high grade.

AJCC Staging System for Primary Malignant Bone Tumors (after 1/1/2003)

• Based on 4 criteria
  (bold = worse px):
  1. Tumor size:
     – T1 = < 8cm
     – T2 if > 8cm
     – **T3** if skip mets
  2. Regional LN mets:
     – N0 = absent
     – N1 = present
  3. Mets:
     – M1a = lung mets only
     – **M1b** = mets other sites/LNs
  4. Grade:
     – G1 = well-, G2 = moderately-, **G3** = poorly-, **G4** = un-differentiated

<table>
<thead>
<tr>
<th>Stage</th>
<th>Tumour</th>
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<tr>
<td>IVA</td>
<td>Any T</td>
<td>N0</td>
<td>M1a</td>
<td>Any G</td>
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<tr>
<td>IVB</td>
<td>Any T</td>
<td>Any N</td>
<td>M1b</td>
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</tr>
</tbody>
</table>

AJCC Staging System for Primary Malignant Bone Tumors (after 1/1/2003)

- Rather than intra- or extra-osseous tumor extent, **tumor size** found to be better px indicator
- Stage III if **skip** mets
- Stage IV if **distant** mets:
  - Lung mets are IVA
  - Elsewhere is IVB

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MRI vs CT

• MRI superior to CT in detecting intraosseous extent

• Best sequence is debatable:
  – For osarc, must include a T1-wted spin echo
  – STIR may overestimate intraosseous tumor when compared to histopath specimens
Epiphyseal involvement

- Physis was thought to be barrier to tumor extension
- MRI = extension of intermed T1 SI across GP w/ physeal destruction
  - Hi sensitivity
  - Low specificity due to FP cases from low SI red marrow at the physis
- T1 more specific and STIR more sensitive to determine epiphyseal involvement
Sample MR protocol for assessing primary tumor

- Large field-of-view coronal or sagittal sequence covering the entire bone
- Small field of view sequences to cover the primary tumor in its entirety:
  - T1- and fat-suppressed T2-weighted sequences in the axial plane
  - T1- and fat-suppressed T2-weighted sequences performed in at least one orthogonal plane
Skip mets

• Intramedullary osteosarcoma (\{\}) with skip mets (arrows) on coronal T1 MR image
• Must include entire length of bone

Courtesy, George Nomikos, AFIP
Determining joint invasion:

• Important for surgical planning:
  – Limb sparing surgery
  – Vs. joint amputation

• Joint effusion does not = Joint involvement

Courtesy, George Nomikos, AFIP
Joint invasion: osteosarcoma (*) along ACL (^)

Courtesy, George Nomikos, AFIP
Role of IV Gad

• Limited value b/c of inherent contrast b/t the tumor and normal marrow signal
  – Intermed/low T1 and nl/high SI marrow fat

• Helpful to differentiate solid f/ hemorrhage & necrosis
  – Aids in biopsy planning
  – Arguably, T2 can be of similar utility

• Dynamic CE-MRI \(\rightarrow\) not useful in initial staging
  – May help discern tumor f/ reactive edema post chemotx or to see residual tumor postop
Bone Scan (Scintigraphy)

- Can overestimate extent of intraoss tumor d/t falsely extended uptake
- May show apparent joint involvement
- Correlates poorly with path specimens, underestimates and overestimates

Role of advanced imaging

• Traditionally, relied on bone scan during initial w/u
• More recent advances include WB MRI
  – Improved techniques have reduced imaging time to < 1hr
  – (rolling platform, parallel imaging, phased-array coils)
• WB MRI better than PET for brain & liver mets
• Chest CT preferred for evaluating lung mets, lymph nodes
  – PET limited for detecting sub-cm nodules
• Currently, PET best to confirm suspicious or equivocal findings & assess areas not in FOV on other modalities
• MRI best for skip lesion detection
  – PET shows promise in peds as red marrow can limit assessment for skip mets on MRI & bone scan

Sample images Whole Body MRI – LCH (arrowhead)

Daldrup-Link, et al. AJR 2001:177
Role of whole body MRI and FDG-PET

• Some evidence that WB-MRI + PET have higher sensitivity for primary bone mets detection than skeletal scintigraphy
Primary Bone Malignancy

OSTEOSARCOMA
Osteosarcoma (Osteogenic Sarcoma (OGS))

- Malignant mesenchymal neoplasm which makes **osteoid** (arrows) or immature bone

- **Histo:**
  - Osteoblastic (predominates in 50-80%)
  - Chondroblastic
  - Fibroblastic

Murphey MD, RG 1997;17:1205-1231
Osteosarcoma Types

• Intramedullary (75%)
  – High-grade*
  – Telangiectatic*
  – Low-grade
  – Small cell
  – Osteosarcomatosis*
  – Gnathic tumors

• Surface (10%)
  – Intracortical
  – Parosteal*
  – Periosteal*
  – High-grade surface tumors

• Extraskeletal

• Secondary (malignant transformation)*

Murphey MD, RG 1997;17:1205-1231
Osteosarcoma – Epidemiology

- Most common primary bone malignancy in pts < 20 yo
  - <6 or >60 yo unusual
- 2\textsuperscript{nd} most common in all ages following myeloma
- More common in white pts
- M:F, 1.5:1

Osteosarcoma Treatment

- Chemotx
- Wide surgical resection
- Limb salvage if possible (or amputation)
Osarc - Metastatic disease

- Lungs
  - Spontaneous ptx
- Regional/distant LN's
- Bones

Courtesy, George Nomikos
Osarc - Metastatic disease

- Lungs
- Regional/distant LNs
- Bones:
  - Skip mets (MR of entire length of bone)
    - Intramedullary osteosarcoma (\{\}) with skip mets (arrows) on coronal T1 MR image

Courtesy, George Nomikos
Osteosarcoma: high-grade intramedullary

- Also called central or conventional
- 75% of all osarcs
- 15-25 yo
- 5 yr survival of 60-80%
- About the knee (50-55%)
  - Femur > tibia > humerus

Osteosarcoma: high-grade intramedullary

- 90% metaphyseal
  - Majority cross to epiphysis
- 5-10% diaphyseal
- < 1% epiphyseal

(Murphey, AFIP notes)

Courtesy, George Nomikos
Osteosarcoma: high-grade intramedullary - XR

- Mixed sclerosis/lysis
- Aggressive periostitis & ST mass
- Violates cortex w/o expanding it
- Large at dx, > 6cm
- Rapid growth (doubles in 20-30 days)

Courtesy, George Nomikos
Osteosarcoma: high-grade intramedullary - MR

- Essential for staging and preoperative planning
- Tumor is intermediate SI on T1

Courtesy, George Nomikos
Osteosarcoma: high-grade intramedullary - MR

- Tumor is high SI on T2
- Mineralized matrix = areas of low SI on both T1 and T2
- Areas of hemorrhage = high SI on both T1 and T2
- Areas of necrosis (ST or bone) = low T1, high T2

Courtesy, George Nomikos
Telangiectatic osteosarcoma

- 4.5-11% of all osarcs
- Sim distribution to intramedullary:
  - Most around knee
  - 90% metaphyseal, 10% diaphyseal
  - May have better px t/others (68% 5-yr survival)
- Can be secondary (FD, Paget, & after XRT)
- Rarely extraskeletal

Courtesy, George Nomikos
Telangiectatic osteosarcoma

- Must have hemorrhagic, cystic, or necrotic spaces occupying > 90% lesion
- Cystic cavities = cavernous vessels, blood filled spaces
  - Fluid/fluid levels

Courtesy, George Nomikos
Telangiectatic osteosarcoma

- Largely osteolytic & expansile
  - Geographic bone destruction
  - Wide zone of transition
- Osteoid formation in periphery
  - “Donut sign” on bone scan
  - RN accumulation in periphery, central photopenia

Courtesy, George Nomikos
Telangiectatic osteosarcoma

- Aggressive periosteal rxn
- Cortical destruction
- ST mass
- Pathologic frx

Courtesy, George Nomikos
... Not to be confused with Aneurysmal Bone Cyst (ABC)

- Term aneurysmal is based on its radiographic appearance
- Interval of 4 mos b/t images
- Rapid lesion expansion has been reported

http://www.bonetumor.org/tumors-bone/aneurysmal-bone-cyst

Courtesy, George Nomikos
Not to be confused with Aneurysmal Bone Cyst (ABC)

- Expansile osteolytic lesion with a thin wall, containing blood-filled cystic cavities
- True cause unknown (possibly post-traumatic)

Courtesy, George Nomikos
Ddx of Secondary ABC includes . . .

- Giant cell tumor of bone (distal radius)
- Osteoblastoma
- Chondroblastoma

Courtesy, Brady Huang
Ddx of Secondary ABC includes . . .

- Giant cell tumor of bone
- Osteoblastoma
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Courtesy, George Nomikos
Ddx of Secondary ABC includes . . .

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Courtesy, George Nomikos, AFIP
Osteosarcomatosis

- Also known as multifocal osteosarcoma, or multiple sclerotic osteosarcoma
- Multiple intraosseous foci of osarc at time of presentation
- Uncommon, 3-4% of osarc cases
Osteosarcomatosis

- Possibly represents rapidly progressive metastatic disease
  - Murphey strongly believes this
- Rapidly appearing, usually symmetric, sclerotic lesions
Osteosarcomatosis

- Typically a large, dominant lesion can be identified
- Dominant lesion:
  - Ill-defined margins
  - Aggressive periosteal rxn
  - Cortical disruption
  - Adjacent ST extension
  - Can be sclerotic or lytic
Osteosarcomatosis

• Secondary foci:
  – Smaller
  – More sclerotic
  – Better defined
  – Lack periosteal rxn or cortical destruction

Georgetown University Hospital
Osteosarcoma
tosis

- Horrible prognosis
- Despite chemotherapy, aggressive surgery, reported mean survival of 12 months

Georgetown University Hospital
Osteosarcomatosis

Georgetown University Hospital
Surface osteosarcoma: Parosteal OGS

- 65% of all juxtacortical osteosarcomas
- 3\textsuperscript{rd}-4\textsuperscript{th} decades of life
- Slight female predominance
- 80-90% long-term survival
- Presents as palpable mass

Courtesy, George Nomikos
Parosteal OGS

- Metaphyseal (80-90%)
- Post distal femur (50-65%)
- Arise from the outer layer of periosteum
- Exophytic sclerotic mass

Courtesy, George Nomikos
Parosteal OGS

- Large, lobulated, ossific, juxtacortical mass
- "Cauliflower-like"
- Excellent px
  - Surgical resection
  - No neoadjuvant chemotx or XRT

Courtesy, George Nomikos
Parosteal OGS

- CT & MR
- Important for planning surgical resection
  - Show ST-extent
  - Determine if medullary involvement
  - Look for lucent cleavage plane

Courtesy, George Nomikos
Parosteal OGS

- Determine medullary involvement (*)
  - Deep medullary invasion may require limb salvage
  - Joint replacement if involved
Parosteal OGS Mimic = Myositis Ossificans

- More dense peripherally
- Usually not attached to cortex

Courtesy, George Nomikos
Parosteal OGS Mimic = Myositis Ossificans

Courtesy, George Nomikos
Periosteal OGS

• 25% of all juxtacortical osteosarcomas
• 2\textsuperscript{nd} – 3\textsuperscript{rd} decades, M>F
• Diaphyseal or metadiaphyseal
• Femur & tibia >> ulna & humerus
• Arise f/ deep layer of periosteum
• Cortical thickening, scalloping, w/o intramedullary invasion
• 55-83% long-term survival

Children’s National Medical Center
Periosteal OGS

- Broad-based surface ST mass
- Causes scalloping of thickened underlying cortex, w/o intramedullary invasion
- Periosteal rxn – perpendicular to bone length axis

Children’s National Medical Center
Periosteal OGS

- Perpendicular periosteal rxn into a broad-based ST mass
- Codman’s triangles at sup & inf margins of the lesion
Periosteal OGS

- Perpendicular periosteal rxn into a broad-based ST mass
- Codman’s triangles at sup & inf margins of the lesion

Children’s National Medical Center
Periosteal OGS

- Perpendicular periosteal rxn into a broad-based ST mass
- Codman’s triangles at sup & inf margins of the lesion
Periosteal OGS

- ST mass of intermed T1 SI
- Erodes thick cortex extrinsically

Children’s National Medical Center
Periosteal OGS

- ST mass of intermed T1 SI
- Erodes thick cortex extrinsically
Periosteal OGS

- Fluid sensitive sequence better shows the ST mass
- *Important to note high water content of ST mass reflects cartilage component
  - Biopsy of ST mass may lead to mis-dx of a *chondrosarcoma*
  - Substantial tx implications (no chemotx for chondrosarc, but would for osarc)
  - (Murphey Rad 2004)
Periosteal OGS

- Enhancing ST mass
Periosteal OGS

- Axial fluid sensitive images show well-defined ST mass of approximately 75% cortical circumference
- Rays of low SI = periosteal rxn
- Small foci in marrow of high SI are not continuous with ST mass (reactive marrow)
Periosteal OGS

- Bone scan
- Marked, eccentric radionuclide uptake
- No other lesions, good px

Children’s National Medical Center
Other lesions that involve the cortex....

- Osteoid osteoma
- LCH
- FCD/NOF
- Infn

Case from CVI, 2/18/2011 UCSD Teleradiology
Ddx of Cortical Lesions

- Osteiod osteoma
- LCH
- FCD/NOF
- Infn

Courtesy, George Nomikos
Ddx of Cortical Lesions

- Osteoid osteoma
- LCH
- FCD/NOF
- Infn

Courtesy, George Nomikos
Ddx of Cortical Lesions

- Osteiod osteoma
- LCH
- FCD/NOF
- Infn

Courtesy, George Nomikos
Osteoid Osteoma: look closely for the nidus

- Benign
- Nidus consists of bone in various stages of maturity
- Has highly vascular stroma of connective tissue w/ many dilated capillaries

Case from CVI, 2/18/2011 UCSD Teleradiology
Osteoid Osteoma: nidus

- Osteoid w/in nidus will calcify
  - Associated with irregular trabeculae of woven bone
- Nidus is surrounded by compact lamellar bone made by the periosteum

Case from CVI, 2/18/2011 UCSD Teleradiology
Reviewed 38 path-proven cases of OO

- Femur (13), tibia (15), humerus (4)
- Determined location of nidus center of OO
- Subperiosteal (18), Intracortical (18), endosteal (0), intramedullary (2)

Kayser F. AJR:170, March 1998
Evidence of the Subperiosteal Origin of Osteoid Osteomas in Tubular Bones: Analysis by CT and MR Imaging

- Proposed that osteoid osteoma arises in a surface location (subperiosteal)
- Inward migration of the nidus (intracortical, endosteal, intramedullary)

Kayser F. AJR:170, March 1998
Continuous remodeling of bone with subperiosteal deposition, endosteal erosion, & cortical drift

Cortical drift phenomena of immature bone
  - Bone develops in response to mechanical load, to fracture healing, etc.

Case from CVI, 2/18/2011 UCSD Teleradiology
Osteoid Osteoma: extensive marrow edema

- CVI case was an arthrogram
- Wider FOV coronal fluid sensitive seq showed marrow edema
- Patient brought back for CT with suspicion of OO
Ddx of lesions causing extensive marrow edema

- Osteoid osteoma
- Chondroblastoma
- Osteoblastoma
- LCH
- Brodies abscess

Case from CVI, 2/14/2011 UCSD Teleradiology
Ddx of lesions causing extensive marrow edema

- Osteoid osteoma
- Chondroblastoma
- Osteoblastoma
- LCH
- Brodies abscess
Ddx of lesions causing extensive marrow edema

- Osteoid osteoma
- Chondroblastoma
- Osteoblastoma
- LCH
- Brodies abscess

Courtesy, George Nomikos
Ddx of lesions causing extensive marrow edema

- Osteoid osteoma
- Chondroblastoma
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Courtesy, George Nomikos
Ddx of lesions causing extensive marrow edema

- Osteoid osteoma
- Chondroblastoma
- Osteoblastoma
- LCH
- Brodies abscess
Primary Bone Malignancy

CHONDROSARCOMA
Primary Chondrosarcoma - Epidemiology

- 3\textsuperscript{rd} most common primary bone malignancy
  - follows MM & osarc
- 8-17\% of all biopsied primary bone tumors
  - Vs. 15\% of biopsied primary bone tumors are osteosarcoma
  - (AFIP notes)

Murphey M D et al. Radiographics 2003;23:1245-1278
Chondrosarcoma

- Malignancy of cartilage cells, often with myxoid changes
- “Rings & arcs”
- Deep endosteal scalloping
- Cortical break-through, ST mass
- Pathologic fracture common

Specimen XR; Courtesy, George Nomikos
Chondrosarcoma

- Hallmark of dx:
  - Entrapment & destruction of osseous trabeculae (T) by cartilage lobules (C)
  - “Islands of normal bone within the neoplasm”
- Higher the grade, more cellular the tumor = less chondroid matrix

Murphey M D et al. Radiographics 2003;23:1245-1278
Chondrosarcoma Types

- Primary
  - (central or surface)
    - Conventional intramedullary*
    - Clear cell
    - Juxtacortical
    - Myxoid
    - Mesenchymal
    - Extraskeletal
    - Dedifferentiated*
- Secondary
  - Enchondroma*
  - Osteochondroma*
  - Paget dis
  - XRT-induced

Murphey M D et al. Radiographics 2003;23:1245-1278
Chondrosarcoma – Grading System

• Grade 1 – low
  – Clear cell (*high glycogen)
• Grade 2 – intermediate
• Grade 3 – high

Murphey M D et al. Radiographics 2003;23:1245-1278
Chondrosarcoma – Grading System

- Grade 1 – low
- Grade 2 – intermediate
  - Myxoid
- Grade 3 – high

Murphey M D et al. Radiographics 2003;23:1245-1278
Chondrosarcoma – Grading System

- Grade 1 – low
- Grade 2 – intermediate
- Grade 3 – high
  - Dedifferentiated
    (“collision tumor” of low grade chondrosarc & high grade fibrosarc)

Murphey M D et al. Radiographics 2003;23:1245-1278
Chondrosarcoma – Grading System

- Grade 1 – low
- Grade 2 – intermediate
- Grade 3 – high
  - Dedifferentiated
  ("collision tumor" of low grade chondrosarcoma & high grade fibrosarcoma)

Murphey M D et al. Radiographics 2003;23:1245-1278
Biopsy & risk of underestimating lesion grade

- Acquire & carefully review images
- Direct biopsy toward aggressive endosteal scalloping, ST component, & diffusely enhancing regions
- Avoid areas of matrix mineralization
- Biopsy tract resected w/ surgical excision

Murphey M D et al. Radiographics 2003;23:1245-1278
Grade 1 conventional chondrosarcoma vs. enchondroma of long bone

• Look for signs of higher grade chondrosarc:
  – Cortical destruction
  – ST mass

• If sx-ic intramedullary cartilage tumor of long bone with endosteal scalloping of < 2/3 cortical thickness . . .
  – Follow 4-6 mo intervals x2 yrs
  – Then annually up to 5 yrs
Chondrosarcoma Imaging: XR

- Mixed lytic and sclerotic
  - Sclerotic = chondroid matrix = rings & arcs, or flocculent calcs
  - Lucent = geographic lysis, multilobulated, corresponds to lobular growth of hyaline cartilage
- Higher grade = more moth-eaten, permeative
  - (mesenchymal, myxoid, & dedifferentiated types)
  - *Clear cell can appear radiolucent*
- Lobulated endosteal scalloping → cortical breakthrough → ST mass

Courtesy, George Nomikos
Chondrosarcoma Imaging: Scintigraphy

- Marked increased radionuclide uptake >> anterior iliac crest
- Heterogenous pattern if conventional intramedullary chondrosarc

Courtesy, George Nomikos
Chondrosarcoma Imaging:

CT

• Deep endosteal scalloping (>2/3 cortical thickness)
• Cortical remodeling, thickening & periosteal rxn, common
• ST component

Courtesy, George Nomikos
Chondrosarcoma Imaging: CT

• Best for detection of matrix mineralization
  – Matrix mineralization not helpful in distinguishing chondrosarcoma/enchondroma
  – Less calcification seen with higher grade

• Mild periph rim & septal enhancement
  – Higher grade lesions more diffuse & nodular enh d/t increased cellularity & decr’d water content

Courtesy George Nomikos
Chondrosarcoma Imaging: MR

- Best for extent of marrow involvement
- Matrix mineralization: low to intermediate on T1
  - Speckled high T1 = entrapped yellow marrow
- Non-mineralized compts are high on T2
  - High water content of hyaline cart
  - Peritumoral edema
- Cart lobules may be surrounded by low SI septa
- PD: best to depict endosteal scalloping

Courtesy George Nomikos
Chondrosarcoma - Treatment

- **Low grade:**
  - Intrallesional curettage, chemical/thermal ablation, cement or bone graft
- **Intermediate/high grade:**
  - Wide surgical excision
- **Chemotx and XRT have limited role**
  - Conventional chondrosarcoma not particularly sensitive
  - XRT for higher-grade conventional chondrosarcoma (gr 2-3) if incompletely excised
  - More aggressive chondrosarcoma (mesenchymal, dedifferentiated) – use both
  - Clear cell chondrosarcoma – use chemotx
Chondrosarc – Metastatic disease

• Low grade:
  – Relatively non-existent

• Intermediate grade:
  – Lungs
  – Lymph nodes
  – Bones

• High grade:
  – Above + viscera (liver)
Chondrosarcoma – Conventional Intramedullary

• Most common primary chondrosarcoma
• 4th-5th decades of life
• Pain & ST mass
• 5 yr survival:
  – Gr1: 90-94%
  – Gr2: 61-81%
  – Gr3: 43-44%
• Long bones & pelvis (65%)
  – Femur > tibia > humerus

Murphey M D et al. Radiographics 2003;23:1245-1278
Intramedullary chondrosarcoma

- Lobular growth
- Multilobulated lesion replacing the marrow space (C)
- Deep endosteal scalloping (>2/3 cortical thickness) with expansile remodeling of bone (arrows)
  - Reflects slow tumor growth

Murphey M D et al. Radiographics 2003;23:1245-1278
Intramedullary chondrosarcoma: Sites of involvement

• Large at dx (>4cm length)

• Long tubular bones
  – Metaphysis > diaphysis
    >> Epiphysis
    • Epiphyseal enchondroma is exceedingly rare
  – Proximal femur > tibia > fibula

Courtesy, George Nomikos, AFIP
Intramedullary chondrosarcoma: T1

- Signal of the ST mass follows that of high water content of hyaline cartilage
  - (sim to periosteal osarc, w/o the perpendicular periosteal rxn)

Courtesy, George Nomikos, AFIP
Intramedullary chondrosarcoma: T1 & T2

Courtesy, George Nomikos, AFIP
Intramedullary chondrosarcoma: post Gad & gross

Courtesy, George Nomikos, AFIP
Dedifferentiated

- 10% of chondrosarc
- Consists of conventional low-grade chondrosarcoma with foci of higher-grade non-cartilaginous malignancy
  - such as MFH, osteosarcoma, or fibrosarcoma
- “Collision of two tumors”

Courtesy, George Nomikos, AFIP
Dedifferentiated
Dedifferentiated
Dedifferentiated
Clear Cell Chondrosarcoma

- Rare, low grade malignant cartilaginous tumor
  - (Glycogen-heavy chondrocytes that appear clear/vacuolated)
- Epiphyseal/apophyseal
- May histologically resemble osteoblastoma
  - Large areas of hemorrhage and cyst formation, unlike conventional chondrosarcoma
- About ½ of clear cell chondrosarcoma contain areas of conventional chondrosarcoma
Clear cell chondrosarcoma in the left proximal femur of a 30-year-old man with hip pain.

Murphey M D et al. Radiographics 2003;23:1245-1278

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Secondary Chondrosarcomas

• When enchondromas and osteochondromas go bad
Enchondroma

- Geographic
- Narrow zone of transition
- Chondroid matrix (except hand, may not see matrix)

Complications:
- Fracture
- Malignant transformation to chondrosarcoma

Multiple:
- Maffucci (w/ ST hemangiomas) & Olliers
- Increased risk of malignant transformation

Courtesy, George Nomikos
Enchondroma
Chondrosarcoma features when ddx incl enchondroma:

• Deep endosc scalloping (>2/3 cortical depth)
• Uptake on bone scan > ASIS
• Cortical destruction or ST mass
• Periostitis, cortical thickening
• Size > 5cm
• Epiphyseal location (unlikely for enchondroma)
• Pain directly attributable to lesion
Enchondroma Gone Bad

• Brady’s case
• 59 yo male with palpable, painful 4\textsuperscript{th} digit mass

Courtesy, Brady Huang
Secondary Chondrosarcoma from Enchondroma

- **1st** – clinically sx-ic
- **2nd** – imaging
  - Expansile
  - Marrow replacing, endosteal thinning
  - Enhancement

Courtesy, Brady Huang
Secondary Chondrosarcoma from Enchondroma

T1

T2 FS

T1 FS +

Courtesy, Brady Huang
Enchondroma Gone Bad

Courtesy, Brady Huang
Osteochondroma

- Most common benign bone neoplasm
- 10-15% of all primary bone tumors
- Malig transformation:
  - 1% for solitary osteochondromas
  - 2-5% for hereditary multiple exostoses (HME)
Osteochondroma: Cartilage cap
Osteochondroma: Malignant Transformation

- Growing osteochondroma in a skeletally mature patient
- Irregular or indistinct lesion surface
- Focal radiolucent regions w/in the lesion
- Erosion or destruction of adj bone
- Significant ST mass w/ scattered calc’n
- Hyaline cart cap thickness > 1.5cm sk mature suspicious for harboring malignant transformation
- Bone scan does not help differentiate benign f/malignant
Osteochondroma Gone Bad - MHE
Improved Differentiation of Benign Osteochondromas from Secondary Chondrosarcomas with Standardized Measurement of Cartilage Cap at CT and MR Imaging.


- Murphey and colleagues set out to verify cartilage cap thickness and concern for malign
- Reviewed 67 benign enchondromas, 34 exostotic chondrosarcomas
- Greatest percentage of malignancy was in lesions derived from pelvis
Determining cartilage cap thickness

- Identify tidemark (arrows) of mature mineralization at the cartilage interface with the osteochondroma stalk
- Exclude crevases of cartilage b/t undulations in the tidemark (dotted line)

Bernard S A et al. Radiology 2010;255:857-865
Determining cartilage cap thickness

- Measure cartilage thickness perpendicular to the tidemark
- Include full thickness of relatively high-fluid-content cartilage
  - Fluid attenuation on CT
  - Low-intermed T1
  - Intermed PD
  - High T2

Bernard S A et al. Radiology 2010;255:857-865
Murphey 2010 article results

- Cartilage cap thickness results:
  - Benign: 79% < 1cm; 7% > 1.5cm, 18% > 1cm
  - Malignant: 2-14cm at CT, 2-17cm at MR; 0 < 2cm, 79% > 3cm

- 2cm cutoff:
  - Sens/spec = 100%/98% MR (100%/95% CT)
  - PPV/NPV = 96%/100% MR (93%/100% CT)

- If cap > 2cm at CT, confirm with MR or US to exclude bursal fluid

- Consider close surveillance of central lesions, esp pelvic osteochondromas
Correlation of cartilage cap thickness and pathologic findings.

- Abrupt transition b/t malig (green) & benign (red) at 2cm
- Arrow = 1 exception, benign lesion of 2.2cm

Bernard S A et al. Radiology 2010;255:857-865

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In conclusion . . .

- Reviewed imaging criteria
- Discussed imaging modalities and role in staging
- Discussed some of the most common primary malignancies and their importance in deriving useful ddx.

Washington Monument - National Cherry Blossom Festival 2010
Photo by Brent Shepherd
Thank you!

- Special thanks to George Nomikos and his excellent teaching file which he hoped I would share with all of you.

Tidal Basin, Jefferson Memorial - National Cherry Blossom Festival 2007
Photo by Brent Shepherd
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