Introduction to Musculoskeletal Tumors

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Definitions

• Primary Bone / Soft tissue tumors
  • Mesenchymally derived tumors (Mesodermal)
  • Benign or Malignant (Sarcoma)
  • Sarcoma=fleshy (Greek), fish flesh
  • Sarcoma—ability to metastasize systemically and invade locally
Classification

• Derived from primitive pluripotential mesenchymal cell
• Pluripotential mesenchymal cell can form
  • Bone
  • Cartilage
  • Fibrous Tissue
  • Lipogenic
  • Blood Vessels
  • Nervous tissue
  • Small Round Blue Cells
Classification

- Bone and soft tissue tumors are classified according to the predominant type of tissue (Pattern of Differentiation).
- Important to think in terms of these categories when evaluating.
- There are benign and malignant types and matrix and non-matrix producing tumors.
## Classification

### Bone Forming Tumors

<table>
<thead>
<tr>
<th>Benign</th>
<th>Malignant</th>
</tr>
</thead>
<tbody>
<tr>
<td>Enostosis (Bone Island)</td>
<td>Osteosarcoma</td>
</tr>
<tr>
<td>Osteoma</td>
<td></td>
</tr>
<tr>
<td>Osteoid Osteoma</td>
<td>Intramedullary</td>
</tr>
<tr>
<td>Osteoblastoma</td>
<td>Conventional</td>
</tr>
<tr>
<td></td>
<td>Telangiectatic</td>
</tr>
<tr>
<td></td>
<td>Low Grade Intraosseous</td>
</tr>
<tr>
<td></td>
<td>Surface/Juxtacortical</td>
</tr>
<tr>
<td></td>
<td>Parosteal</td>
</tr>
<tr>
<td></td>
<td>Periosteal</td>
</tr>
<tr>
<td></td>
<td>High Grade Surface</td>
</tr>
<tr>
<td></td>
<td>Intracortical</td>
</tr>
<tr>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Classification
Cartilage Forming Tumors

Benign
- Enchondroma
- Osteochondroma
- Chondroblastoma
- Chondromyxofibroma
- Periosteal Chondroma

Malignant
- Chondrosarcoma
  - Conventional
    - Grade I
    - Grade II
    - Grade III
  - Dedifferentiated
  - Clear Cell
  - Mesenchymal
  - Periosteal Chondrosarcoma
Classification

Fibrous/Fibroosseous Tumors

Benign

• Nonossifying fibroma
• Fibrous cortical defect
• Benign Fibrous Histiocytoma
• Fibrous Dysplasia
• Osteofibrous dysplasia
• Desmoplastic Fibroma

Malignant

• Fibrosarcoma
• Malignant Fibrous Histiocytoma (MFH)
Classification
Small Round Blue Cell Tumors
(Non Matrix Producing)

**Benign**
- Eosinophilic Granuloma

**Malignant**
- Ewing Sarcoma
- Primary Lymphoma of Bone
- Plasmacytoma/Myeloma
Classification
Giant Cell Rich Lesions

Benign
• Giant Cell Tumor
• Giant Cell Reparative Granuloma
• Aneurysmal Bone Cyst (ABC)

Malignant
• Malignant Giant Cell Tumor of Bone
Classification
Vascular Tumors

**Benign**
- Hemangioma
- Lymphangioma
- Glomus Tumor

**Malignant**
- Epithelioid Hemangioendothelioma
- Hemangiopericytoma
- Angiosarcoma
Classification

Benign Cysts:
- Unicameral Bone Cyst
- Intraosseous Ganglion

Miscellaneous:
- Intraosseous Lipoma
- Intraosseous Liposarcoma
- Parosteal Lipoma
- Leiomyosarcoma of Bone
- Adamantinoma
- Rhabdomyosarcoma
- Chordoma

Synovial Lesions:
- Synovial Hemangioma
- Synovial Chondromatosis
- Pigmented Villonodular Synovitis (PVNS)
- Synovial Lipoma
- Synovial Chondrosarcoma
Natural History

Benign
- Latent
- Active
- Aggressive

Malignant
- Low Grade
- Intermediate
- High Grade
Staging

• Purpose
  • Determine tumor type
  • Determine prognosis
  • Guide treatment
  • Compare results between study groups
  • Delineate extent of local and distant disease
Staging Studies

- Plain Radiograph
- MRI
- CT scan
- Chest CT
- Bone Scan
Staging for Benign Bone Tumors

Benign Staging System (Enneking)

**Stage 1: Latent**
- Grow slowly with growth of individual and then stop; tendency to heal spontaneously
- (ex. NOF; UBC)

**Stage 2: Active**
- Progressive growth; Minimal surrounding sclerosis

**Stage 3:**
- Aggressive; Break through bone
# Staging for Malignant Bone Tumors

## Enneking Staging System

<table>
<thead>
<tr>
<th>Stage</th>
<th>Grade</th>
<th>Site</th>
</tr>
</thead>
<tbody>
<tr>
<td>IA</td>
<td>G1</td>
<td>T1</td>
</tr>
<tr>
<td>IB</td>
<td>G1</td>
<td>T2</td>
</tr>
<tr>
<td>IIA</td>
<td>G2</td>
<td>T1</td>
</tr>
<tr>
<td>IIB</td>
<td>G2</td>
<td>T2</td>
</tr>
<tr>
<td>III</td>
<td>Mets</td>
<td>Mets</td>
</tr>
</tbody>
</table>

*(based on biological behavior)*
# Staging for Malignant Bone Tumors

## Malignant Bone Tumors

### TNM Staging System (AJC)

<table>
<thead>
<tr>
<th>Stage</th>
<th>Grade</th>
<th>Tumor</th>
<th>Node</th>
<th>Mets</th>
</tr>
</thead>
<tbody>
<tr>
<td>IA</td>
<td>G1,2</td>
<td>T1</td>
<td>N 0</td>
<td>M 0</td>
</tr>
<tr>
<td>IB</td>
<td>G1,2</td>
<td>T2</td>
<td>N 0</td>
<td>M 0</td>
</tr>
<tr>
<td>IIA</td>
<td>G3,4</td>
<td>T1</td>
<td>N 0</td>
<td>M 0</td>
</tr>
<tr>
<td>IIB</td>
<td>G3,4</td>
<td>T2</td>
<td>N 0</td>
<td>M 0</td>
</tr>
<tr>
<td>III</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>IVA</td>
<td>Any G</td>
<td>Any T</td>
<td>N1</td>
<td>M 0</td>
</tr>
<tr>
<td>IVB</td>
<td>Any G</td>
<td>Any T</td>
<td>Any N</td>
<td>M 1</td>
</tr>
</tbody>
</table>

III  Undefined for bone tumors
### Grading

**Biological Behavior / Natural History**

<table>
<thead>
<tr>
<th>G1</th>
<th>G2</th>
</tr>
</thead>
<tbody>
<tr>
<td>LG Chondrosarcoma</td>
<td>High Grade Chondrosarcoma</td>
</tr>
<tr>
<td>Secondary Chondrosarcoma</td>
<td>Conventional Osteosarcoma</td>
</tr>
<tr>
<td>Parosteal Osteosarcoma</td>
<td>Ewing’s Sarcoma/PNET</td>
</tr>
<tr>
<td>Adamantinoma</td>
<td>Malignant Fibrous Histiocytoma</td>
</tr>
<tr>
<td></td>
<td>Angiosarcoma</td>
</tr>
</tbody>
</table>
Soft Tissue Sarcoma Staging

• Important Prognostic Characteristics
  • Tumor Size (>5cm, worse prognosis)
  • Tumor Depth (Deep, worse prognosis)
  • Grade (High grade, worse prognosis)
  • Presence of Mets
### Staging for Soft Tissue Sarcomas

#### Malignant Tumors

**TNM Staging System (AJC)**

<table>
<thead>
<tr>
<th>Stage</th>
<th>Grade</th>
<th>Tumor</th>
<th>Node</th>
<th>Mets</th>
</tr>
</thead>
<tbody>
<tr>
<td>IA</td>
<td>G1,2</td>
<td>T1a-b</td>
<td>N 0</td>
<td>M 0</td>
</tr>
<tr>
<td>IB</td>
<td>G1,2</td>
<td>T2a</td>
<td>N 0</td>
<td>M 0</td>
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<tr>
<td>IIA</td>
<td>G1,2</td>
<td>T2b</td>
<td>N 0</td>
<td>M 0</td>
</tr>
<tr>
<td>IIB</td>
<td>G3,4</td>
<td>T1a-b</td>
<td>N 0</td>
<td>M 0</td>
</tr>
<tr>
<td>IIC</td>
<td>G3-4</td>
<td>T2a</td>
<td>N 0</td>
<td>M 0</td>
</tr>
<tr>
<td>III</td>
<td>G3,4</td>
<td>T2b</td>
<td>N 0</td>
<td>M 0</td>
</tr>
<tr>
<td>IVA</td>
<td>Any G</td>
<td>Any T</td>
<td>N1</td>
<td>M 0</td>
</tr>
<tr>
<td>IVB</td>
<td>Any G</td>
<td>Any T</td>
<td>Any N</td>
<td>M 1</td>
</tr>
</tbody>
</table>
Grading of Soft Tissue Sarcomas

- **Soft Tissue Sarcomas (Biological Behavior)**
  - Tumors that are definitionally high grade
    - Ewing’s Sarcoma
    - PNET
    - Rhabdomyosarcoma
    - Angiosarcoma
    - Pleomorphic Liposarcoma
    - Soft Tissue Osteosarcoma
    - Mesenchymal Chondrosarcoma
Grading of Soft Tissue Sarcomas

- **Soft Tissue Sarcomas (Biological Behavior)**
  - Tumors that are definitionally low grade
    - Well Differentiated Liposarcoma
    - Dermatofibrosarcoma Protuberans
    - Infantile Fibrosarcoma
    - Angiomatoid MFH
Grading of Soft Tissue Sarcomas

- **Soft Tissue Sarcomas**
  - Tumors not gradable but which metastasize often
    - Alveolar soft part sarcoma
    - Clear cell sarcoma
    - Epitheloid sarcoma
    - Synovial sarcoma
    - Low grade fibromyxoid sarcoma
Grading of Soft Tissue Sarcomas

- Soft Tissue Sarcomas
  - Tumors of varying behavior—grading may be useful
    - Myxoid liposarcoma
    - Leiomyosarcoma
    - MPNST
    - Fibrosarcoma
    - Myxoid MFH
Grading of Soft Tissue Sarcoma

- **Soft Tissue Sarcomas**
  - Tumors of varying behavior—grading parameters not yet established
    - Hemangiopericytoma
    - Myxoid chondrosarcoma
    - Malignant granular cell tumor
    - Malignant mesenchymoma
Thank You!