Clinical Presentation & Evaluation and Staging

Pediatric Orthopedic Department
University of Basel, Switzerland
• 8-year old boy
• for 10 days pain in his right knee and thigh
• questions?
• measures to take?
• pain persists now for 4 weeks
• they still are about the same
• questions?
• measures to take?
What to do?
Evaluation and Staging

- history
- clinical findings
- laboratory findings
- imaging
- biopsy
- staging
History

- pain?
- functional limitations?
- weight?
Pain history

when did it start?

trauma?

activity?

night pain?

duration?

how often?
Night pain

- high-grade malignant bone tumors (osteosarcomas, Ewing-sarcomas etc.)
- bone producing benign bone tumors (osteoid-osteoma, osteoblastoma)
- tumors with instability because of (micro-)fractures (mainly metastases)
- infections (bone and soft-tissue)
- growing pain (usually bilateral)
No night pain in:

- All soft tissue sarcomas (incl. high-grade sarcomas)
- All benign soft tissue tumors (except for mechanical pain)
- Low grade malignant bone tumors
- Benign bone tumors (except bone producing tumors)
Clinical findings

- consistency
- displaceability of the skin
- displaceability on the bone
- tenderness
- reddening
- warming up
Functional limitations

- limping
- reduced ROM
- reduced force
Laboratory findings

- serum alkaline phosphatase (osteosarcoma?)
- lactate dehydrogenase (LDH) (Ewing’s sarcoma?)
- sedimentation rate (Ewing’s sarcoma)
- catecholamines (neuroblastoma?)
- monoclonal proteins (plasmocytoma?)
Making the diagnosis (I)

- XR
- CT-scan
- MRI
- Scintigram
Making the diagnosis (II)
Making the diagnosis (III)
~0.5 % of all malignant tumors: Bone tumors
(without plasmocytoma: ~0.5%)
Metastases (CUP) ~ 6-8x more frequent
Age at diagnosis of the 3 most common malignant bone tumors

* Bone Tumor Registry of Basle 2006
Bone tumors in children and adolescents

Benign Tumors
- osteochondroma 19.8%
- non-ossifying bone fibroma 15.8%
- unicameral bone cyst 8.9%
- aneurysmal bone cyst 4.6%
- fibrous dysplasia 2.6%
- other tumorlike lesions 4.7%
- enchondroma 5.8%
- chondroblastoma 1.8%
- osteoid-osteoma 8.3%
- giant cell tumor 1.3%

Malignant Tumors
- Ewing-sarcoma 4%
- other malignant 4.0%

Tumorlike Lesions
- other benign tumors 9.3%
- osteosarcoma 9.1%

Basle Bone Tumour Registry, tumors at age < 20 y., n=4408
Bone tumors in adults

Basle Bone Tumour Registry, tumors at age > 20 y., n=7534
Age related occurrence of soft-tissue tumors

- Malignant fibrous histiocytoma / pleomorphic sarcoma
- Malignant peripheral nerve sheath tumour
- Liposarcoma
- Fibrosarcoma
- Synovial sarcoma
- Rhabdomyosarcoma
## Topography within the bone

### benign vs malignant

<table>
<thead>
<tr>
<th>Region</th>
<th>Tumors</th>
</tr>
</thead>
<tbody>
<tr>
<td>epiphysis</td>
<td>chondroblastoma, giant cell tumors, clear cell chondrosarcoma,</td>
</tr>
<tr>
<td>metaphysis</td>
<td>non-ossifying fibroma, solitary bone cyst, aneurysmal bone cyst, giant cell tumors, osteoblastoma, chondroma, osteosarcoma, (chondrosarcoma)</td>
</tr>
<tr>
<td>diaphysis</td>
<td>fibrous dysplasia, osteofibrous dysplasia (Campanacci), Ewing-sarcoma, primitive neuroectodermal tumor (PNET), Non-Hodgkin-lymphoma (NHL) of bone, adamantinoma, chondrosarcoma</td>
</tr>
</tbody>
</table>
Age distribution and topography of bone tumors
Localization of bone tumors in children and adults
Screening for metastases

- x-ray of thorax, when suspicious CT-scan
- ultrasound abdomen, when suspicious CT-scan
- PET (or PET-CT)
- occasionally a whole body MRI is more efficient
Classification of Bone Tumors

Benign Tumors

Malignant Tumors

Tumor-like Lesions
Histological Classification (WHO): according to the type of tissue imitated by the tumor cells

- Bone > Osteosarcoma
- Cartilage > Chondrosarcoma
Classification of Bone Tumors

- Tumors that produce bone
- Tumors that produce cartilage
- Tumors containing giant cells
- Tumors of the bone marrow
- Tumors that produce vessels
- Tumors that produce connective tissue
- Other tumors
- Tumor-like lesions
## Classification of Bone Tumors

### benign vs malignant

<table>
<thead>
<tr>
<th>Category</th>
<th>Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>Osteogenic tumors</td>
<td>osteoma, osteoid-osteoma, osteoblastoma, osteosarcoma, parosteal osteosarcoma, low-grade central osteosarcoma</td>
</tr>
<tr>
<td>Cartilage producing tumors</td>
<td>enchondroma, osteochondroma, periosteal chondroma, chondroblastoma, chondrosarcoma, clear cell chondrosarcoma, dedifferentiated chondrosarcoma</td>
</tr>
<tr>
<td>Fibrous tissue producing tumors</td>
<td>benign fibrous histiocytes, desmoplastic fibroma, malignant-fibrous histiocytes, fibrosarcoma</td>
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## Classification of Bone Tumors

### benign vs malignant

<table>
<thead>
<tr>
<th>Tumors Containing Giant Cells</th>
<th>Giant Cell Tumors</th>
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<tr>
<td>Tumor Producing Vessels</td>
<td>Hemangioma, Hemangioendothelioma, Angiosarcoma</td>
</tr>
<tr>
<td>Tumors of the Bone Marrow</td>
<td>Ewing-Sarcoma, Primitive Neuroectodermal Tumor (PNET), Non-Hodgkin-Lymphoma (NHL) of Bone</td>
</tr>
<tr>
<td>Other Tumors</td>
<td>Chordoma, Adamantinoma</td>
</tr>
<tr>
<td>Tumor-Like Lesions</td>
<td>Solitary Bone Cyst, Aneurysmal Bone Cyst, Langheans‘Cell Histiocytosis, Fibrous Dysplasia, Non-Ossifying Fibroma</td>
</tr>
</tbody>
</table>
Diagnostic procedure in bone tumors

MORPHOLOGY
- Border
- Bone Destruction
- Periosteal Reaction
- Matrix
- Soft Tissue Mass

LOCATION
IN PARTICULAR BONE:
What Part of Bone?
(Epipysis, Metaphysis,
Diaphysis, Central,
Eccentric)

AGE, SEX

SINGLE
MULTIPLE

LOCATION
IN THE
SKELETON:
What Bone?
Exam-ales of histo-logical grading

Grade 1
liposarcoma, well differentiated

Grade 2
malignant peripheral nerve sheath tumor (MPNST)

Grade 3
pleomorphic rhabdomyosarcoma
Dedifferentiated chondrosarcoma

enchondroma

chondrosarcoma

osteosarcoma
### Staging-system according to Enneking

<table>
<thead>
<tr>
<th>G-Grade</th>
<th>histological differentiation</th>
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<tr>
<td>T=Site</td>
<td>anatomical localisation, compartments</td>
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<tr>
<td>M=Metastases</td>
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**UNIKLINIK BALGRIST**

«Musculoskeletal Oncology»

October 6, 2011
## Staging-system according to Enneking

<table>
<thead>
<tr>
<th>Stage:</th>
<th>Grade:</th>
<th>Site:</th>
<th>Metastases</th>
</tr>
</thead>
<tbody>
<tr>
<td>IA</td>
<td>G1 (diff.)</td>
<td>T1 (intracomp.)</td>
<td>M0 (none)</td>
</tr>
<tr>
<td>IB</td>
<td>G1 (diff.)</td>
<td>T2 (extracomp.)</td>
<td>M0 (none)</td>
</tr>
<tr>
<td>IIA</td>
<td>G2 (dediff.)</td>
<td>T1 (intracomp.)</td>
<td>M0 (none)</td>
</tr>
<tr>
<td>IIB</td>
<td>G2 (dediff.)</td>
<td>T2 (extracomp.)</td>
<td>M0 (none)</td>
</tr>
<tr>
<td>IIIA</td>
<td>G2 (dediff.)</td>
<td>T1 (intracomp.)</td>
<td>M1 (present)</td>
</tr>
<tr>
<td>IIIB</td>
<td>G2 (dediff.)</td>
<td>T2 (extracomp.)</td>
<td>M1 (present)</td>
</tr>
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American Joint Committee (AJC) staging system of soft tissue tumors

<table>
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<tr>
<th>G-Grade</th>
<th>histological differentiation</th>
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<tr>
<td>T=Site</td>
<td>anatomical localisation, compartments</td>
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<tr>
<td>N=regional lymph nodes</td>
<td></td>
</tr>
<tr>
<td>M=distant metastases</td>
<td></td>
</tr>
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</table>
# The AJC Staging-system of soft-tissue tumors

<table>
<thead>
<tr>
<th>Stage:</th>
<th>Grade:</th>
<th>Site:</th>
<th>Lymph-nodes</th>
<th>Metastases</th>
</tr>
</thead>
<tbody>
<tr>
<td>IA</td>
<td>G1 (diff.)</td>
<td>T1 (intracomp.)</td>
<td>NO (none)</td>
<td>MO (none)</td>
</tr>
<tr>
<td>IB</td>
<td>G1 (diff.)</td>
<td>T2 (extracomp.)</td>
<td>NO (none)</td>
<td>MO (none)</td>
</tr>
<tr>
<td>IIA</td>
<td>G2 (moder.)</td>
<td>T1 (intracomp.)</td>
<td>NO (none)</td>
<td>MO (none)</td>
</tr>
<tr>
<td>IIB</td>
<td>G2 (moder.)</td>
<td>T2 (extracomp.)</td>
<td>NO (none)</td>
<td>MO (none)</td>
</tr>
<tr>
<td>IIIA</td>
<td>G3,4 (dediff.)</td>
<td>T1 (intracomp.)</td>
<td>NO (none)</td>
<td>MO (none)</td>
</tr>
<tr>
<td>IIIB</td>
<td>G3,4 (dediff.)</td>
<td>T2 (extracomp.)</td>
<td>NO (none)</td>
<td>MO (none)</td>
</tr>
<tr>
<td>IVA</td>
<td>G1-4</td>
<td>T1-2</td>
<td>N1 (present)</td>
<td>MO (none)</td>
</tr>
<tr>
<td>IVB</td>
<td>G1-4</td>
<td>T1-2</td>
<td>NO (+/-)</td>
<td>M1 (present)</td>
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Survival rates of soft tissue tumors depending on stage

Survival rates for stages 1, 2, 3, and 4 are illustrated. The graph shows the percentage of patients surviving over years after diagnosis, with each stage having a different line color and stage designation.

- Stage 1 (177 cases)
- Stage 2 (86 cases)
- Stage 3 (329 cases)
- Stage 4 (110 cases)
Assessment of bone tumors

Heterogenous structure of bone tumors

Histological-radiological-clinical correlation is always necessary

Bone tumors are interdisciplinary problems
Multidisciplinary Tumor Center in Basel

Universitäts-Kinderspital beider Basel (UKBB)

Universitätsspital Basel (USB)

Interdisziplinäres Knochen- und Weichteiltumorzentrum der Universität Basel (KWUB)

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