Is that Tumor... A Cancer?

Jorge A. Fabregas, MD
Children’s Orthopaedics of Atlanta
Residency Program Director, Children’s Healthcare of Atlanta
Chief of Pediatric Orthopaedics at AHC

CONFLICT OF INTEREST
I hereby certify that, to the best of my knowledge, no aspect of my current personal or professional situation might reasonably be expected to affect significantly my views on the subject on which I am presenting.

I will not discuss off label use and/or investigational use in my presentation.

Never know how tumors show up!!

Dermatofibrosarcoma Protuberans
Objectives

- 1. Be able to identify radiographic findings of bone lesions
- 2. Be able to create a differential diagnosis
- 3. Familiarize yourself with common treatment options available for bone lesions.

What can go wrong? How can you prevent it?

- Missing a Malignancy
  - Diagnostic tips
- Treatment Options
  - Avoiding complications
- Overtreating Tumors
  - "no touch lesions"
- Common Malignancies

Diagnosis

- Stakes are high
  - Before the died regardless of treatment
  - Botched diagnosis didn’t affect outcome
- History
  - Night pain
  - Rapid symptoms
  - Won’t look sick
- Not all pain is due to injury
Work Up

- Infections and Ewings
- Xrays
- MRI
- ESR and CRPs
- Pathologic fractures
- Radiologist Communication
- Hold off on bone scan and CT chest

Clinical Evaluation

- Incidental Finding
- Pathologic Fracture
  - 50% of UBC
- Palpable mass
  - Duration
  - Malignancy ~ 6 months
  - Size variations

Clinical evaluation

- +/- Pain
  - Character
  - Length
  - Factors that make it better/ worse
  - Inflammatory signs
  - Neurological signs
  - Spine and proximal fibula lesions
Imaging

X-rays most helpful for bone lesions

- Have a routine for approaching unknown bone lesions
- 5 points in description of a lesion:
  1) Age
  2) Tumor location (Lists to remember)
  3) What lesion doing to bone?
  4) What bone doing to lesion?
  5) Periosteal response

Peak age of common pediatric musculoskeletal tumors

<table>
<thead>
<tr>
<th>Age (yrs)</th>
<th>Benign</th>
<th>Malignant</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-5</td>
<td>Langerhan cell histiocytosis (Hand-Schuller-Christian disease), polyostotic, mono-ostotic</td>
<td>Fibrosarcoma, Wilms' tumor (Mets)</td>
</tr>
<tr>
<td></td>
<td>Osteosarcoma</td>
<td>Langerhan cell histiocytosis (Letterer-Siwe disease)</td>
</tr>
<tr>
<td>5-10</td>
<td>Unicameral Cyst, Aneurysmal Bone Cyst, Nonossifying Fibroma, Fibrous Dysplasia, Giant Cell Tumor, Langerhan cell histiocytosis (polyostotic, mono-ostotic)</td>
<td>Osteosarcoma, Ewing's Sarcoma</td>
</tr>
<tr>
<td>10-20</td>
<td>Fibrous Dysplasia, Osteoid Osteoma, Osteosarcoma, Aneurysmal Bone Cyst, Chondrosarcoma, Giant Cell Tumor</td>
<td>Osteosarcoma, Ewing's Sarcoma, Chondroblastoma, Synovial Cell Sarcoma</td>
</tr>
</tbody>
</table>

Location, Location, Location !!!

- Certain tumors have a predilection for certain locations in the skeleton
**Epiphysis**

- Chondroblastoma
- Brodie abscess
- Giant cell
- Fibrous dysplasia

**Metaphysis: Anything**

**Diaphyseal (FAHEL)**

- Fibrous dysplasia
- Adamantinoma (adult)
  - Osteofibrous dysplasia
- Histiocytosis
- Ewings
- Leukemia, lymphoma

- Subacute osteomyelitis
  (occas. osteoid osteoma and UBC)
Posterior elements spine

- ABC
- Osteoblastoma
- Osteoid osteoma
- Osteochondroma
  - Multiple Hereditary Exostoses

When you hear hoof-beats...

X Rays

- Pathologic Fractures
- Check all fractures for possible pathology
  - Exam the whole film
- Difficult Films
  - Scapula
  - Spine
  - Pelvis
Clues for Diagnosis

- **Periosteal patterns** help determine the aggressiveness of a lesion.
  - Solid, smooth.
  - Lamellated ('onion skin') or spiculated ('sunburst') sarcoma and osteosarcoma.
  - Codman's.

- **Location of the lesion**
  - Chondroblastomas and giant cell
  - Osteofibrous dysplasia and adamantinoma
  - Nonossifying fibromas

- **Matrix mineralization**
  - Stippled or “arc and ring” calcifications
  - Amorphous, “cloudlike” mineralization
  - “ground-glass”.

Example

- AP/LAT of Knee
- 15 year old skeletally mature
- Closed Physys
- Well circumscribed radiolucent lesion
- Acrum
- Metaphysis into epiphysis involving the lateral femoral condyle

- Expansile
- Lateral cortical thinning
- No periosteal reaction
- Possible pathologic fx
- No soft tissue Mass

- Differential DX
  - Giant Cell
  - Chondroblastoma
  - Aneurysmal bone cyst

Oh...Oh!
CT Scan vs. MRI

CT Scan
- Assessing cortical destruction
- Superior at detecting mineralization and cortical abnormalities
- Localizing nidus of Osteoid Osteoma
- Differentiating ABC vs. UBC
  - Fluid-fluid levels

MRI
- Usually not indicated for evaluation of benign tumors
- IV contrast or Gadolinium
  - Not necessary
- Fluid-fluid levels of ABC
- MRI reveal soft tissue, bony edema

MRI Usually Advance Imaging Study need before biopsy

Do I need Blood work?
- CBC
- C-reactive Protein
- ESR
  - Infections
  - Ewing's Sarcoma
  - Leukemia
  - Lymphoma
  - Langerhan Cell
  - Histocytosis
  - Metastasis

Leukemia
- Most frequent childhood malignancy
- ALL ~ 80%; peak at 4 y.o. (> boys, whites)
- Symptoms: lethargy, pallor, purpura, hepatosplenomegaly, lymphadenopathy and bleeding
- Musculoskeletal pain in 20-50% (joint, bone pain, etc)
- Limp as a chief complaint 12%
- 75% of children with ALL have radiographic changes
- No pathognomonic osseous manifestation
Next Step

Do I want to mess with it?

Who is your Team?
Pathologist
Oncologist

Learn as much as you can prior to biopsy

Should I biopsy it?

Staging
– Biopsy
– MRI with/without Contrast
– Chest CT and Bone Scan can wait

When to worry

• Rapid growth
  – Exception Synovial Sarcoma
• Deep to Fascia
• Larger >5cm

How to do a Biopsy

• If not comfortable Refer
  – Use a tourniquet (gravity)
  – Longitudinal Incision
  – Stay away from joints
  – Perform Frozen Section
  – Cultures
  – Hemostasis
  – Drains in line with incision
  – Put tissue in formalin
  – Save tissue for genetic analysis
Create a Differential Dx

Your eyes see what your mind knows

General surgeon - hernia
Family - Is that poop?
Ortho - Osteopetrosis

“No Touch” Lesions

• Myositis ossificans
  – Consider MRI or CT
• Avulsion Fractures of Pelvis

“No Touch” Lesions

Stress Fractures
CoRTICAL dESMOID
Non Ossifying Fibroma

- Eccentric Cortically Based
- Metaphysis of Long Bones
- 30% of Population
- Observation
- Mayo > 50% treat
- Jaffe Campanaci Syndrome

Osteoid Osteoma

- benign solitary painful lesions
  – Nidus approximately <1.5 cm
  – Dull aching pain commonly at night present for several months
  – Relieved by NSAIDS and aspirin
  – Location- predominantly in long bones and posterior elements of the spine
  – If located in spine may be associated with painful scoliosis

- Natural History
  – No malignant transformation
  – Becomes asymptomatic and heals after ~3 to 5 years
  – Nidus will ossify, but surrounding sclerosis will persist
Treatment

- NSAIDS and aspirin
- CT guided radiofrequency thermoblature
- Surgical excision
  - En bloc
  - Burr down technique
- Cryo ablation

PROTECT LIMB

CLINICAL STUDY

Cryoblation of Osteoid Osteomas in the Pediatric and Adolescent Population

- 29 patients
- 3-18 years of age
- Complete resolution of pain at 19/21 at greater than a year
- 6 complications

Osteochondroma

- Cartilage capped bony projection on the external surface of a bone
- 1/3 of benign tumors
- 2nd decade
- Tx observation
- Excision pain, fracture, growth after skeletal maturity
**What is the most common talar lesion?**

- **Chondroblastoma**
  - Painful lytic lesion on epiphysis
  - Open growth plate
  - Edema on MRI
  - Metastasis <1%
  - Tx
    - Biopsy
    - Curettage and BG
    - +/- adjuvant

**Unicameral Bone Cysts**

- Common
- Usually found at time of pathologic fracture
- 90% proximal humerus and proximal femur
- Purely lytic, central
- Tx
  - Steroids
  - Bone graft
  - BMP Inj

**Percutaneous IM decompression and delivery of medical grade calcium sulfate**

- Radiopaque
- Placed percutaneously
- Biodegradable
  - Reabsorbed in 30-60 days
- Not intended to provide structural support
Recently

Aneurysmal Bone Cyst

- Solitary, expansile, radiolucent lesion
  - Wider adjacent physis
- Etiology unknown
- Affects patients between 5 to 20 years of age
- 75% percent in long bones
- Mainly in the metaphysis and posterior elements of the spine
- Bone graft
- Argon
- Tetracyline injection
  - Side effects.

Etiology

- Local circulatory disturbance leading to increased venous pressure and enlarged vascular bed within affected bone.
- Chromosome abnormalities
- USP6 hyper expression
- Dysregulation of BMP
- 16q22 and/or 17p13
Radiographic Findings

- Ballooned lesion with thin periosteal shell
- Located in the metaphysis, eccentric location with septations (soap bubble)
- MRI – spinal cord compression or rapidly expanding cyst
  - Fluid level
  - Septations
  - Low T1 and High T2

ABC 5 Step Approach:

- Biopsy
- Telangectatic Osteosarcoma
- Curettage
- High speed burr
- Cautery
- Phenol
- Bone Grafting

*Frozen section always first*

2006-2010
20 patients
6 to 14 injections
Increase cortical thickening
Recurrence rate of <5%
Langerhan’s Histiocytosis

• Prev known as histiocytosis X
• Spectrum of diseases in children
  – Eosinophilic granuloma
    • Osseous form
    • Self-limited
  – Hand-Schuller-Christian syndrome
    • Multifocal
    • Disseminated
    • Chronic
  – Letterer-Siwe syndrome
    • Disseminated form
    • Lethal

Histiocytosis

• Presentation
  – Localized, aching pain
  – Pain worse at night
  – Fever
  – Skin rash
  – Diabetes insipidus
  – Hepatosplenomegaly (L-S dz)
• Location
  – 50% skull, jaw
  – 25% spine, pelvis: can cause vertebra plana
  Usually no mass
• Needs Skeletal survey
• Wide variation “great imitator”

Histiocytosis

• Treatment
  ★ Refer to Hematologist
    – Self limited
    – Observation
      • Biopsy +/- steroids
    – No treatment needed for vertebral plana
Malignant Tumors

- Account for more 1/2 of amputations performed for disease processes in children
- Most common cause of death from disease in childhood and adolescence
- 8000 per year
- 12% musculoskeletal
- Multidisciplinary approach

That soft tissue mass: Rhabdomyosarcoma

- Most masses benign
  - Popliteal cyst
  - Ganglion
  - Deep Granuloma annulare

Do I Worry?

- Deep
- Larger than 2cm
- Midline… doesn't fit a baker's Cyst.
Soft Tissue Masses: Some are obvious

How about this one?

Rhabdomyosarcoma

- Arising in muscle
- MC ST sarcoma of childhood
- 40-50% of all Peds STS
  - 2 peaks 2-6 and 14 to 18 years of age
- 5% of childhood cancer
- Present anywhere
- Painless mass
**Osteosarcoma**

- Age distribution (2 peaks)
  - 10-20 adolescents (most rapid growth)
  - 6th decade (Pagets and Radiation)
- Any bone
- Metaphysis ends of long bones
- Get x rays if complaining of pain

**Osteosarcoma**

- If Pathologic fx
  - Tx conservatively
    - Splint or cast
- Please do not Rod
- Refer immediately to orthopaedic oncologist

**Ewing’s Sarcoma**

- The most lethal of the primary bone tumors
  - Survival >50% if no metastasis
- M > F (3:2)
- 5-25 y.o. (younger than O.S.)
- Any bone - femur M.C. (20%)
- May be multicentric
- 6-14% of all 1st tumors of bone
**Ewing’s Sarcoma**

- Commonly diaphyseal (75%)
- Diffuse permeative destruction of bone
- Extension of tumor through cortex into S.T.
  - MRI: large soft tissue mass
- Periosteal reaction
  - Codman’s Triangle
  - Onion skinning
  - Sunburst
- Wide zone of transition
- Large soft tissue mass on MRI

---

**Limb Sparing Surgery**

- LSS rather than amputation can be performed in up to 70 to 80%
- Reconstructed limb should have improved or equal function compared to amputation
- Does not appear to compromise survival

---

**Resection/Rotationplasty**
**Take Home**

- Recognizing Lesions
- Malignant vs. benign
  - Radiographic findings
- No touch Lesions
- Treatment Options
- What can go wrong? How can we prevent it?

---

**Warning**

- Don’t be fooled by pain presence for several weeks
- Pain in cancer and infections sometimes blamed on trauma
- Ewing and Infection look similar
  - Fever, ESR, CRP
- Look closely at x-rays
- If possible biopsy at center of definitive treatment