Current Management of Bone Sarcomas: An Orthopaedist’s Perspective

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LEARNING OBJECTIVES

At the end of this presentation the participant will be able to:

1. Understand current pediatric bone sarcoma management
2. Understand the role of an orthopaedic surgeon in the treatment of bone sarcomas
3. Appreciate the multi-disciplinary approach to management of pediatric bone sarcomas

WHAT IS AN ORTHOPAEDIC ONCOLOGIST/MUSCULOSKELETAL ONCOLOGIST?

• Somebody who has gone to school for >90% of their life
• Somebody who is more rare than a billionaire in the US
• Somebody who will never join that group

• Surgical management of benign and malignant soft tissue and bone diseases of the skeleton

MUSCULOSKELETAL ONCOLOGIST

• Requires an understanding of pathology, hematology/oncology, psychiatry, radiation oncology, prosthetics, radiology, and vascular surgery.

• I guess that explains the bags under my eyes

• Multi-disciplinary field with a team approach to treatment.

BONE SARCOMAS

Primarily: Osteosarcoma and Ewing’s sarcoma

(rarely chondrosarcoma)
PRIMARY BONE SARCOMAS

- Rare
- Most pediatricians will see one or two in their life of practice
  - Benign diseases are far more common (don’t be fooled to assuming this)
  - Although benign diseases sound just that, don’t forget that many require treatment

BONE SARCOMAS

- How rare?
  - Large centers see < 20/year
  - Require multi-disciplinary approach
  - Hem-onc does the vast majority of the work
  - I’d like to believe that I’m important too (but I know I’m not)

WHAT IS A SARCOMA

- Malignant tumor of mesenchymal tissue origin

Modeling sarcomagenesis using multipotent mesenchymal stem cells. Review Cell Research 2012

SARCOMAS

- Osteosarcoma = bone
- Chondrosarcoma = cartilage
- Rhabdomyosarcoma = skeletal muscle
- Ewing’s = neuroectodermal tumor (kind of)

MANAGEMENT

- Varies dramatically for each type of sarcoma

OSTEOSARCOMA

- Bone forming sarcoma
  - Total of about 800 new cases of osteosarcoma in the US last year
  - JUST 800! (thankfully)
  - Just over 95% of these occur in pediatric patients (400-500/year)
  - Most common in adolescents around the knee
    - Distal femur, proximal tibia, proximal fibula
    - Proximal humerus, pelvis
  - Antecdotally, occurring in younger patients
    - Likely due to environmental factors
OSTEOSARCOMA MANAGEMENT
- History, physical examination
- Most complain of pain that started insidiously as an occult sprain
- Pain persists or recurs after rest, most common around the knee
- Tenderness
- X-rays
- May be subtle; poor clinic x-rays can disguise tumor
- Referral
- Early referral and let the advanced imaging be decided by the specialist
- Specialist
- MRI of the entire involved bone
- Bone scan
- CT chest

OSTEOSARCOMA MANAGEMENT
- Biopsy
- Neoadjuvant chemotherapy
- Doesn't shrink tumor but typically turns to bone and resolves surrounding edema
- Surgical resection
- Resection = complete removal of all tumor tissue
- Not scoop or scrape out but as a whole piece
- Adjuvant chemotherapy
- Determined by percent necrosis of tumor on histologic evaluation

WHERE DOES THE SURGEON FIT IN?
- Simply: Get rid of ALL tumor (don't leave any around)
- Although > 90% necrosis from chemotherapy is "good", what about the other 10%?

General concept: Limb salvage vs. Amputation

<table>
<thead>
<tr>
<th>Necrosis (%)</th>
<th>Limb Salvage</th>
<th>Amputation</th>
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<tbody>
<tr>
<td>90% - 20%</td>
<td>Their own leg</td>
<td>No local recurrence</td>
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<tr>
<td>20% - 80%</td>
<td>Quick recovery</td>
<td>Requires wound healing and long surgery</td>
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<tr>
<td>95% - 5%</td>
<td>Minimal risk of long term pain</td>
<td>Metal or allograft bone has own set of complications</td>
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LIMB SALVAGE VS. AMPUTATION

Ewing's sarcoma
- Combination of ectoderm and mesoderm
- Young children (<15)
- Diaphyseal part of femur, tibia, or fibula
- Pelvis
- Can present as soft tissue mass and swelling alone
- Categorized as a "small round blue cell tumor"
- Cells have a EWS-FLI1 translocation t(11,22)

Ewing's sarcoma treatment
- Staging
- CT Chest/Abdomen/Pelvis
- MRI of extremity
- Bone Scan
- Bone marrow biopsy to rule out marrow involvement
- Biopsy
- Neoadjuvant Chemotherapy
- Surgery or radiation therapy
- Adjuvant chemotherapy
SURGERY VS. RADIATION

- If a resectable site of disease, surgery
  - Femur, tibia, fibula, humerus, forearm, hand, or foot

- If a non-resectable or profound morbidity, radiate
  - Pelvis or spine
  - Equal outcomes

Ewing’s sarcoma radiation vs. surgery

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<tr>
<th></th>
<th>Radiation</th>
<th>Surgery</th>
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<tr>
<td><strong>Advantages</strong></td>
<td>Equal outcomes in trials to surgery</td>
<td>Removal of ALL tumor</td>
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<tr>
<td><strong>Disadvantages</strong></td>
<td>No certainty of removing disease</td>
<td>Morbidity of recovering and reconstruction</td>
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<td>Long-term sequelae of radiation; secondary sarcoma; fibrosis, scarring, pain</td>
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CHONDROSARCOMA

- Cartilage based tumor
- Very rare in pediatric population
  - < 10% of all chondrosarcomas
  - Occurs in pelvis most commonly
- Can arise “de novo” or “secondary”
  - If arises in the setting of a low grade chondroid lesion (enchondroma, osteochondroma) then secondary

CHONDROSARCOMA

- X-rays
- Staging
  - MRI or CT scan of lesion
  - Bone scan
  - CT chest
  - Biopsy

- Surgery alone
  - Chemo and radiation insensitive

CASE EXAMPLES WITH DISCUSSION

CASE 1

- 13 yo female with left hip pain
- TTP: fullness on left pelvis
- RH: Trisomy 21
AP x-ray shows a left pelvis lesion that has eroded the bone.  
- Appears to have calcification (small white flecks)  
- Consistent with a chondroid lesion = chondrosarcoma

CT AND MRI
- Erosion of the cortex  
- Needle biopsy verified chondrosarcoma

PATHOLOGY SECTION

16 MONTHS LATER

DISCUSSION
- No chemotherapy, no radiation therapy
- Pulmonary surveillance clear
- Reconstruction unnecessary and healed uneventfully
- Chondrosarcoma only requires surgery with a clear margin

CASE 2
- 5 yo male with left knee pain
- No injury
- TTP distal femur
- PMH negative
**Xrays**

- Xrays alone are pathognomic
- High-grade osteosarcoma
  - Bone formation
  - Extra-osseous mass
  - Periosteal reaction of the bone
- Staging negative, neo-adjuvant chemotherapy

**Pediatric Specific Issues**

- Children grow
- Removing the tumor would involve removing the growth plate
  - Having one leg much shorter than the other is not very useful
  - At 5 years old, he is going to grow a ton!
- If the survival, reconstruction options can potentially fail and require more and more surgery

**Reconstruction Options**

- **Allograft** - somebody else’s bone
  - Advantage: fills the space and is structurally strong
  - Disadvantage: higher infection rate and complication rate, does not grow
- **Arthrodesis** - fuse the knee together
  - Advantage: should be one surgery
  - Disadvantage: knee does not move
- **Amputation** - cut the whole leg off above the tumor
  - Advantage: should be one surgery and done
  - Disadvantage: above the knee prosthesis isn’t fantastic
- **Expandable prosthesis and rotationplasty**

**Expandable Prosthesis**

- Sounds like a panacea
- Invasive or non-invasive implants that can be expanded as the child grows
- Replaces the section of bone that is removed

**Rotationplasty**

- Remove the tumor, but leave the nerves and blood vessels intact to the foot
- Rotate the leg 180 degrees so that the ankle functions as the knee
CASE 3
- 13 yo female with right thigh mass
- Moderately painful, growing mass
- No injury
- Otherwise healthy

X-RAYS
- Scalloped erosive tumor
- Note you can even tell the tissues are enlarged medially
- MRI ordered

MRI
- Bright signal is tumor
- The femur is being eroded through

BONE SCAN
- Non-specific but uptake in the distal femur

BIOPSY
- CD99 positive and lots of small round blue cells consistent with Ewing’s sarcoma
PLAN
- Neoadjuvant chemotherapy
- Tumor shrinks! (Lymphoma and Ewing’s sarcoma shrink)

SURGERY
- Since the femur is "expendable," let’s take it out, replaced with an allograft in the center

IMPLANTS
- For adolescents, we use implants to replace the bone that is removed
- All implants need soft tissue coverage that is not just skin or it will break down

GROWTH PLATE PRESERVATION 8 YO MALE
OSTEOSARCOMA

PRESERVATION GROWTH PLATE

GROWTH PLATE PRESERVATION - ALLOGRAFT