Caring for Pediatric Patients with Malignant Bone Tumors: The Advanced Practice Nurse’s Perspective

Elizabeth Cummings MSN RN CPNP-AC CPHON
Kristen Dalton MSN RN CPNP-AC CPHON
Amy Rapino MSN RN CPNP-PC

APHON National Conference - September 2020
Disclosures

• No relevant conflicts of interest or disclosures to discuss!
Objectives

• To describe the presentation, diagnosis, and management of pediatric malignant bone tumors
• To explore conference identified learning gaps
• To address how to support patients through the acute and late effects of these tumors and their therapy
• To discuss the APP role and the importance of interprofessional practice in the care of these patients
Case Study #1

• 18 year old male
• 2 month hx of left calf stiffness and weakness
• Symptoms progressed to sharp shooting pain in the back of his left thigh, buttocks, and radiating to the calf
• No urinary or bowel incontinence, saddle anesthesia, fever, decrease in appetite
• 5 pound weight loss over the past 1-2 months
Case Study #1

- Seen in outpatient ortho clinic
- Physical examination
  - 4/5 strength in left leg
  - Hyporeflexia on left leg
  - Decreased sensation over Left thigh
  - Normal rectal tone
  - Limp
  - Unable to plantar flex left foot or do left leg calf raises
- MRI lumbar spine revealed lesion in S2
- Referral to pediatric oncology
Diagnostic Work Up

- MRI lumbar spine
- Biopsy
- CT chest
- PET scan
Diagnostic Work Up: Biopsy

- Needle
- Open incisional
- Lab testing

https://www.chop.edu/pediatric-fellowships/interventional-radiology/fellowship
Ewing Sarcoma

- Epidemiology
- Biology
- Genetics
- Risk factors
- Clinical presentation
- Staging
- Prognostic factors

http://europepmc.org/article/PMC/5808087
Supportive Care at Diagnosis

- Pain control
- Physical therapy
- Education
- Interprofessional support

Plan

- Diagnostic and staging workup
- Baseline evaluations
- Central line placement
- Chemotherapy
- Re-staging scans
- Local control
- Monitoring after therapy
Chemotherapy

• Neoadjuvant and adjuvant therapy
• Importance of interval compression
• Standard of care
  • VDC
  • IE
• Notable toxicities r/t chemotherapy
• Supportive care during chemotherapy
Surgery

• S2 vertebral tumor - challenging location
• Risks
  • Loss of bladder or bowel function
  • Loss of sensation to foot or ankle
  • Loss of motor function to foot or ankle
  • Tumor recurrence or positive margins
IN DEPTH: Radiation for Ewing Sarcoma

- Radiosensitive tumor
- Efficacy
  - Tumor size and location may influence outcome
  - No randomized controlled trials comparing radiation and surgery
- Generally used when surgery is not an option or if poor functional outcome is expected with surgery
Radiation: Dose

- Measured in Gy
  - Absorption of one joule of radiation per kilogram of matter
  - 1 Gy = 100 cGy

- Definitive Dose for Ewing Sarcoma: 55.8 Gy (5580 cGy)
  - Pre-chemo volume to 45 Gy (4500 cGy)
  - Boost to residual tumor (determined by re-staging scans)

- Dose is divided into daily fractions
Radiation: Planning Process

- CT-Simulation
  - Non-contrast scan
  - Create immobilization devices and place tattoos to replicate position
  - Anesthesia may be required for young children
- SIM CT is fused with diagnostic imaging
- Radiation Oncologist contours the target volumes
- Dosimetrists and Physicists design the beam angles
- Radiation Oncologist reviews and approves the plan (or sends back for revisions)
- Rigorous quality assurance process
Initial Radiation Plan

- Pre-chemo volume to 4500 cGy

Proton Plan
Boost Volume

- Residual tumor volume to 5580 cGy

Proton Plan
Complete Radiation Plan

- Plan Sum

Proton Plan
Radiation Treatment

- Goals
  - #1: Treat the tumor
  - #2: Minimize toxicity to normal cells
- Consider the real estate
  - Bladder
  - Bowel (large and small)
  - Cauda
  - Rectum
  - Femoral heads
- Determine the optimal modality
Radiation Modalities

- **Photons** (x-rays)
  - AP/PA (2D)
    - Most basic, used in the emergent setting
  - 3D
    - Basic - extremities, whole abdomen, whole brain
  - Intensity Modulated Radiation Therapy (IMRT)
    - Superconformal

- **Protons**
  - Double scatter
    - Less conformal, increased dose to skin
  - Pencil Beam Scanning
    - Dose paint, conformal, less dose to normal tissue
Plan Comparison of Radiation Modalities

- Photons vs. Protons

IMRT Plan

Proton Plan
Dose Volume Histogram

- Dose to bladder, bowel, and rectum very low
- Cauda- mean dose is 46 Gy
- Highest dose to tumor
Acute Side Effects of Pelvic Radiation

- Fatigue
- Skin erythema (radiation dermatitis)
- Nausea and vomiting
- Diarrhea
- Anemia, thrombocytopenia
- Delayed count recovery
Late Side Effects of Pelvic Radiation

- Decreased velocity of bone growth
- Arthritis in hip joint (if in the field, usually avoided)
- Slightly increased risk of bowel obstruction, altered bowel habits
- Increased risk of fracture
- Risk of second malignancy
Case Study #2

- 15 year old female
- Presented to orthopedic clinic with 4 weeks hx of right shoulder pain
- Pain when doing push ups during color guard practice
- Pain worsened over several months
- PCP ordered x-rays → referred to local orthopedist
- MRI right shoulder → concern for tumor
Case Study #2

• On exam
  • Firm and fixed mass palpable at right proximal humerus
  • Pain with shoulder abduction + internal/external rotation
  • Tenderness to palpation
  • Neurovascular exam intact
  • No deficits in strength
Diagnostic Work Up

- MRI right shoulder
- IR guided biopsy
- CT chest
- PET scan
Diagnostic Work Up: Imaging
Osteosarcoma

- Epidemiology
- Biology and genetics
- Risk factors
- Predisposition syndromes
- Clinical presentation
- Staging
- Prognostic factors

http://medcell.med.yale.edu/histology/bone_lab/osteosarcoma.php
Supportive Care at Diagnosis

- Pain control
- Occupational vs physical therapy
- Weight bearing precautions
- Education
- Interprofessional support
Plan

- Diagnostic + metastatic workup
- Baseline evaluations
- Central line placement
- Chemotherapy
- Surgery
- Monitoring after therapy
Chemotherapy

- Neoadjuvant and adjuvant therapy
- Standard of care = MAP
  - Methotrexate
  - Doxorubicin
  - Cisplatin
- Local control
- Notable toxicities
- Supportive care

https://link.springer.com/chapter/10.1007/978-1-4419-0284-9_18
Role of Radiation in Osteosarcoma

- NOT a radiosensitive tumor
  - Requires high doses (7000-8000 cGy)
  - Limited by the normal structures in the field
- Considered if no surgical option
- Palliation
IN DEPTH: Surgery

• Local control surgery = wide resection

• Goals
  • Negative margins
  • Maintain function

• Pre-op MRI
Surgery: Limb Salvage

Radical Resection
- Physeal-sparing
- Joint-sparing
- Limb-sparing

<table>
<thead>
<tr>
<th>Huvos Classification</th>
<th>Necrosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Little or no effect, &lt;60% tumor necrosis</td>
</tr>
<tr>
<td>II</td>
<td>Areas of viable tumor, 60-89% necrosis</td>
</tr>
<tr>
<td>III</td>
<td>Scattered foci, 90-99% tumor necrosis</td>
</tr>
<tr>
<td>IV</td>
<td>No histological evidence of viable tumor – 100%</td>
</tr>
</tbody>
</table>
Local Control Surgery: Amputation
Limb Salvage: Vascularized Fibular Reconstruction

- Fibula = appendix
- Microsurgery
- Plate & screws
- Long term considerations
Limb Salvage: Endoprosthetic Reconstruction

- Joint replacement
- Static vs Growing
- Recovery to walk
- Metal and plastic < bone
- Activity level
Local Control Surgery: Van Nes Rotationplasty

- “Almost” Limb Salvage, “Functional Amputation”
- Full activities
- Often will be one and done
- For distal femur/proximal tibial tumors
Case Study #2: Surgery

• Limb salvage
  • Endoprosthetic reconstruction
  • 100% tumor necrosis, negative margins
• OT for shoulder rehabilitation
• Is now >3 years post-surgery
• Doing well!
Local Control Surgery: Postoperative Management

• Wound healing- 2-3 weeks post-op before resuming chemo
• Monitor @ 2 weeks, 6 weeks, 3 months; then every 3-4 months x 3 years, every 6 months year 4 & 5
• Possible complications
  • Infection
  • Non-union
  • Fracture
  • Limb length discrepancy
Local Control Surgery: Take Home Points

• Not “one size fits all” approach
• Primary goal = wide resection
• Chemo is a double edged sword
• Kids are resilient!
Role of the Nurse Practitioner

- Education
  - Patient and family
  - Nursing
  - Medical team
- Advocacy
- Clinical management, monitoring, and supportive care
- Research
Future Directions

- Standard of care
- Phase 2 clinical trials
- Future breakthroughs
  - Chemotherapy
  - Immunotherapy
  - Personalized medicine
  - Imaging
  - Radiation
  - Surgical techniques

https://nfer.org/blog/sarcoma-awareness-month
Questions?
References


References


