Pediatric Bone and Soft Tissue Tumors Practice Tool

Evaluation, Diagnosis and Referral

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Background

Bone and soft tissue tumors arise in the body's connective and soft tissues, including bone, cartilage, muscle, fat, blood vessels, lymph vessels, nerves, tendons, ligaments, synovial tissue, and other fibrous tissue. Benign tumors are more common than malignant tumors (sarcomas).

Osteosarcomas are the most common malignant bone tumor and develop most frequently in children and young adults 10 to 20 years of age; they most often occur in the distal femur. Osteochondroma is the most common type of benign bone tumor in children, accounting for approximately 35% of benign tumors in children.

Bone:
~80% benign
~20% malignant

Soft Tissue:
~99% benign
~1% malignant

Clinical Presentation of Bone and Soft Tissue Tumors

The clinical presentation of bone and soft tissue tumors is highly variable; however, some general trends are observed:

- Patients often present with a mass; typically one that is increasing in size.
- Constitutional symptoms are rare, but fever, malaise and weight loss may be observed, especially in patients with Ewing sarcoma.
- Pain is the most important symptom to consider. Bone tumors tend to be painful, while soft tissue tumors usually are not; however, there are exceptions to this general rule.

Delayed presentation and diagnosis are common, especially when a mass is painless.

General Presentation	
Bone tumors	• Pain or swelling, usually in the arms, legs, chest, back or hips • Pain may be worse at night • A lump that might feel soft and warm • A bone that breaks for no reason • Fever, tiredness or weight loss
Soft tissue tumors	• A painless lump or swelling under the skin • As the tumor grows, it may cause pain, muscle weakness or limping

Workup

Choosing the right imaging modality is critical to the diagnosis and management of patients with suspected bone or soft tissue tumors.

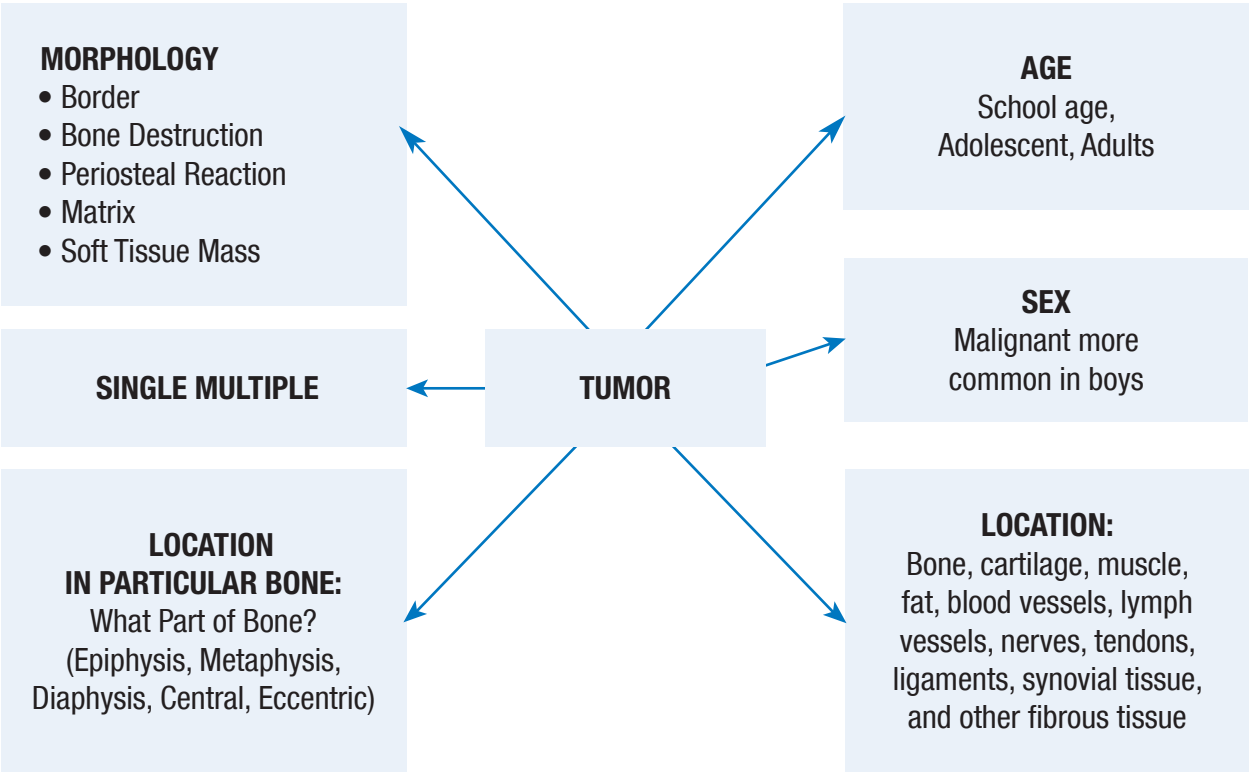

Figure 1: Basic Bone Lesion Work-up

Bone - Bone tumors often have defining radiographic hallmarks (Table 1). Radiographs are valuable for characterizing osseous lesions and enable accurate differential diagnosis of benign and malignant tumors.

Soft Tissue - Radiographs have limited to no value in the evaluation of soft tissue tumors. These almost always require MRI assessment.

During the initial stages of patient evaluation, a comprehensive medical history, physical examination and laboratory workup should be conducted. Chest radiographs may be obtained and are helpful for observing large nodules or masses that may result from metastatic disease.

Biopsy is gold standard for diagnosis and will typically be conducted by a specialist after referral.

When to Refer

Every patient with a mass with indeterminate imaging findings should be referred to or reviewed by a pediatric orthopedic or musculoskeletal oncologist.

If you have a concern about a patient and would like to consult, please contact the Nationwide Children's Orthopedic Oncology team at (614) 722-5175 or visit NationwideChildrens.org/Orthopedic-Oncology

About the Orthopedic Oncology Program

Led by orthopedic oncologist Dr. Tom Scharschmidt, our program is designed to investigate, treat and provide lifelong support for patients with all types of bone and soft tissue tumors. Whether a patient needs comprehensive medical and surgical support for a malignant tumor or surgery for a benign tumor causing concern, our team is ready to help you and your patient families.

Our treatment options include everything from embolization and bone grafting to limb salvage and joint replacement surgery. We will ensure your patient and their family has the information they need to make the best decision and move forward with confidence.

Referrals Information

To discuss a patient with our team, please call (614)-722-5175
or visit **NationwideChildrens.org/Orthopedic-Oncology**

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W1859001
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Bone and Soft Tissue Tumors in Pediatric and Young Adult Patients: An Overview

Tumor Type	Age (decade of life)	Sex (M:F)	Location	Presentation	Radiographic Hallmarks	Treatment	Prognosis
Malignant							
Osteosarcoma	1st, 7th+	1.3:1	• Ends of long bones in the arms and legs ∘ Femur (42%) ∘ Tibia (19%) ∘ Humerus (10%)	• Pain (especially at night/rest) • Swelling • Mass • Pathologic fracture	• Sun-burst appearance • Periosteal lifting/formation of Codman's triangle • New bone formation in soft tissues with permeative pattern of destruction of bone	• Neoadjuvant chemotherapy (MAP) • Surgery • Adjuvant chemotherapy	• 5-year survival localized disease: up to 75% • 14-23% of patients with metastatic disease at presentation ∘ 5-year OS: 45% • Local recurrence with limb-salvage: ~5-10%
Ewing Sarcoma	1st-3rd	1.5:1	• Lower extremity (41%, femur) • Pelvis (26%) • Chest wall (16%) • Upper extremity (9%) • Spine (6%) • Metadiaphyseal/ diaphyseal	• May have systemic symptoms • Often with large soft tissue mass • 25% with metastatic disease	• Bone destruction with moth-eaten to permeative changes (76% to 82%) • Aggressive onionskin or spiculated periosteal reaction (58% to 84%) • Wide transitional zone (96%)	• Neoadjuvant chemotherapy (dose compressed VDC/IE + surgery) • Support for trimodality therapy in pelvic Ewing	• Overall 60-80% ∘ <40% for pelvic location ∘ <20% when presenting with metastases • Worse prognosis with: ∘ Larger tumors (>200 mL), older age (>15 years) ∘ Male sex, poor histologic response, axial location • Local recurrence rate: 20% with radiation vs 6% with surgery
Rhabdomyosarcoma	1st, 2nd	1.3:1	• Head and neck area • Genital or urinary organs	• Persistent (or growing) swelling or mass • May be painful • Crossed-eyes or bulging eye • Headache • Trouble with urinating or bowel movements • Blood in urine • Bleeding in nose, throat, vagina, or rectum		• Surgery • Radiation therapy • Chemotherapy	• 5-year survival for low-risk disease: 70-90% • 5-year survival for intermediate-risk disease: 50-70% • 5-year survival for high-risk disease: 20-30% • Risk group depends on patient age, tumor location and histology, and metastasis
Benign							
Osteoid Osteoma	1st-3rd	3:1	• 80% in long bones at or near cortex ∘ Proximal femur ∘ Tibia ∘ Fibula ∘ Humerus • Posterior spine	• Pain ∘ Night pain ∘ Worse with alcohol ∘ Better with NSAIDs	• Small (<1 cm) lytic nidus with surrounding sclerosis • Very hot on bone scan CT for localization	• Suppression with NSAIDs • Curettage/burring • Radiofrequency ablation	• Rare recurrence and excellent prognosis
Osteoblastoma	1st-3rd	3:1	• Posterior spine • Sacrum • Femur	• Pain ∘ Not responsive to NSAIDs	• Variable • Lucent, slightly expansile • No sclerotic rim • >2 cm	• Not self-limiting • Cutterage and bone grafting • Resection	• Local recurrence: up to 20% • Excellent prognosis
Osteochondroma	2nd		• Ends of long bones (femur, tibia, humerus) or hip	• Hard, painless (usually) mass near joint • Can be painful if near nerve or rubbing muscle	• Classic stalk-like appearance/bony spur • Continuity with the underlying bone	• Monitored until child finished growing • Surgery if large or bothersome	• Extremely low risk of recurrence • Excellent prognosis

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Tumor Type	Age (decade of life)	Sex (M:F)	Location	Presentation	Radiographic Hallmarks	Treatment	Prognosis
Benign (continued)							
Unicameral bone cyst (true bone cyst)	1st-2nd	1:1	• 90% in proximal humerus and proximal femur • Proximal tibia • Fibula	• Asymptomatic • Pathologic fracture	• Metaphyseal, cystic lesion • May be multiloculated • Minimal expansion • 20% "fallen leaf" sign	• Controversial • Aspiration and injections • Autogenous bone marrow injections • DBM and bone graft substitutes • Curettage and bone grafting (allow to move away from physis first) • Allow pathologic fracture to heal before initiating treatment	• Local recurrence: 10-20% • Prognosis ◦ Excellent ◦ Eventual resolution of cyst Problematic local recurrence • DDX - ABC
Aneurysmal bone cyst (Cystic bone lesion with central cavity of blood)	2nd	1:1.3	• Posterior spine • Femur • Tibia • Any bone	• Pain • Swelling	• Expanded ("aneurysmal") bone • Lytic appearance • No matrix • Can have aggressive appearance • Fluid-fluid levels on MRI	• Doxycycline injections • Curettage, +/- adjuvant treatment, bone graft or PMMA	• Local recurrence - high (17-59%) • Overall prognosis ◦ Good ◦ Problematic local recurrence
Tenosynovial Giant Cell Tumor (TGCT)	4th-5th	1:1.5	• Knees • Fingers • Wrists • Elbows	• All related to the joint ◦ Pain ◦ Swelling ◦ Locking/stiffness ◦ Instability/popping • Possible pathologic fracture		• Surgery	• Good prognosis • High rate of recurrence
Intraosseous ganglion cyst	3rd-5th	1:1	• Medial malleolus • Proximal tibia • Carpal bones • Acetabulum	• Often asymptomatic • Pain may occur with weight bearing/increased activity	• Well-defined lytic lesion in the epiphysis beneath the subchondral plate	• Observation vs curettage and bone grafting	• Local recurrence: uncommon • Overall prognosis: excellent, may develop DJD • DDX - GCT, chondroblastoma
Non-ossifying fibroma (Metaphyseal Fibrous Defect)	1st-2nd	1:1	•Distal femur • Distal tibia • Proximal tibia	• Usually asymptomatic • Occasional pathologic fracture	• Eccentric • Longer than wide • Sclerotic margin • Lobulated or scalloped border	• Observation • Curettage, bone graft, internal fixation if concerned for pathologic fracture	• Local recurrence: rare • Overall prognosis: excellent, eventual involution of lesion • DDX - enchondroma, fibrous dysplasia
Fibrous dysplasia	2nd-3rd	1:1.3	• Skull • Ribs • Proximal femur • Any bone • Note: ◦ Monostotic = common ◦ Polyostotic = rare	• Typically asymptomatic • Occasional deformity and pathologic fracture	• Well-defined with sclerotic margin • Hazy, "ground glass" matrix • Expansion of bone • Bowing/deformity • "Shepherd's crook"	• Observation • Curettage, bone graft, internal fixation if symptomatic or concern for pathologic fracture • Will frequently heal with dysplastic bone	• Local recurrence: common • Overall prognosis: good ◦ Local recurrence and deformity can be problematic ◦ Malignant transformation has been reported ◦ McCune-Albright syndrome - polyostotic fibrous dysplasia, endocrine abnormalities, "coast of Maine" café au lait spots • DDX - NOF, well-differentiated fibroblastic osteosarcoma