

# **Tumor Information**

# **Bone Tumors Soft Tissue Tumors**

#### **Bone Tumors**

Bone tumors are a rare cause of musculoskeletal pain but should always be considered in the patient with otherwise unexplained pain. Most bone tumors present with pain and/or a mass. Care must be taken to ensure the correct diagnosis is made, and early consultation with an orthopaedic oncologist is advised to avoid potential complications. In general, these tumors are best treated at a referral practice that specializes in bone tumors.

# Benign

# **Presenting symptoms**

Benign bone tumors can have a wide variety of presenting symptoms; in general, benign bone tumors present with pain. Tumors can occur in any bone, and can occur in all age groups. In general, these tumors are a rare cause of musculoskeletal pain, but should be considered when the diagnosis is in question. Often, benign tumors are found incidentally when patients are imaged for other reasons (i.e., a football player hurts his knee and gets an X-ray to rule out fracture; a suspicious tumor is seen). These are usually benign tumors, but need to be carefully evaluated by an orthopaedic tumor specialist.

# **Diagnostic Imaging**

Imaging is necessary to diagnose a bone tumor. Often, multiple tests are ordered, but must be evaluated carefully by an orthopaedic tumor specialist to make sure that the most accurate diagnosis is rendered.

X-Rav

Usually done in the office, this is the most basic imaging test. Plain X-rays can provide essential diagnostic information, and must be of high quality. It is not uncommon to have to repeat these in order to make sure a high quality digital image is obtained. Often, this is the only test that is needed to diagnose and follow a benign bone tumor.

CT Scan

A 3-demensional study that allows orthopaedic oncologists to see the bone and soft tissue better than a plain X-ray. Bone destruction, matrix production, and degree of bone involvement are assessed. In addition, soft tissues are visualized better than an X-ray, but not as well as with an MRI.

#### MRI

MRI provides excellent bone and soft tissue detail, and is often necessary to diagnose and treat these tumors. Images are usually obtained with IV contrast. In the pediatric population, sedation, and occasionally general anesthesia is required to make sure a quality image is obtained. Most adults can tolerate the test quite well. Occasionally claustrophobic patients require oral sedation to complete the MRI. These tests may need to be repeated by the orthopaedic oncologist if the quality of the image is poor, or if the entire area is not seen on the scan.

#### Bone Scan

A nuclear medicine test that shows areas of bone turnover. Often ordered when the diagnosis is difficult or for staging purposes (to make sure there are not other areas of bone involvement). MRI has made this test less necessary for benign bone tumors.

#### **PET Scan**

A new imaging modality that allows doctors to see metabolic activity in a tumor. It is usually used for imaging in patients with cancer, but will occasionally be used for patients with benign bone diseases.

#### Staging

For benign bone tumors, most surgeons use the Enneking staging system:

Grade 1- the tumor is latent or inactive.

The patient has no symptoms, the tumor is not growing, and the x-rays are consistent with a benign process. Usually these tumors do not require treatment, but should be observed annually.

Grade 2-The tumor is benign appearing, but is growing and causing symptoms. There is not overt bone destruction, and treatment usually consists of minimally invasive surgery to remove the tumor and place bone graft in its place.

Grade 3- The tumor has destroyed bone and must be removed by excision of the area. Often, these tumors are locally aggressive, and have the potential to recur even after aggressive local treatment.

### **Biopsy**

Biopsy for benign bone tumors is often necessary to confirm the diagnosis. The biopsy is usually done in the setting of an outpatient minor surgery. Often tissue is sent as a frozen section so that the diagnosis is known the same day.

Minimally invasive core needle biopsy or minimally invasive open biopsy is the current standard. Biopsy of musculoskeletal tumors should be performed by specialists who do this procedure regularly. Inappropriate biopsy can lead to significant preventable morbidity and complications. In general, biopsy is best performed by a musculoskeletal surgical oncologist.

#### **Treatment**

Often, asymptomatic tumors are treated by observation. Serial X-rays are usually ordered to make sure that the tumor is not growing. In the case of tumors that are symptomatic or enlarging, minimally invasive surgery is performed where possible. Usually, the tumor is removed carefully, and then a bone graft substitute is paced in the defect to restore the patient's skeletal structure.

# **Malignant**

# **Presenting symptoms**

Malignant bone tumors can have a wide variety of presenting symptoms. In general, malignant bone tumors present with pain and a mass in the effected extremity. Tumors can occur in any bone, and can occur in all age groups. In general, these tumors are a rare cause of musculoskeletal pain, but should be considered when the diagnosis is in question. For certain tumors, patients may have fever, chills, and weight loss. Early diagnosis is the key to success in these patients, and early referral to a musculoskeletal tumor surgeon is necessary to ensure the correct diagnosis and treatment plan is instituted.

#### **Diagnostic Imaging**

Imaging is necessary to diagnose a bone tumor. Often, multiple tests are ordered, but must be evaluated carefully by an orthopaedic tumor specialist to make sure that the most accurate diagnosis is rendered.

X-Ray

Usually done in the office, this is the most basic imaging test. Plain X-rays can provide essential diagnostic information, and must be of high quality. It is not uncommon to have to repeat these in order to make sure a high quality image is obtained.

CT Scan

A 3-demensional study that allows orthopaedic oncologists to see the bone and soft tissue better than a plain X-ray. Cortical destruction, matrix production, and degree of bone involvement are assessed. In addition, soft tissues are visualized better than a X-ray, but not as well as an MRI. In patients with a bone malignancy, CT of the chest is often ordered to evaluate for tumor metastasis.

MRI

MRI provides excellent bone and soft tissue detail, and is often necessary to diagnose and treat these tumors. Images are usually

obtained with IV contrast. In the pediatric population, sedation, and occasionally general anesthesia is required to make sure a quality image is obtained. Most adults can tolerate the test with the occasional patient needing oral sedation. These tests may need to be repeated by the orthopaedic oncologist if the quality of the image is poor, or if the entire area is not seen on the scan. Surgical planning for removal of the tumor is often based on the MRI findings. Therefore, it may need to be repeated after chemotherapy and prior to surgery.

#### Bone Scan

A nuclear medicine test that shows areas of bone turnover. For malignant bone tumors, it is ordered to rule out metastatic disease.

#### **PET Scan**

A new imaging modality that allows doctors to see metabolic activity in a tumor. It is used often for sarcoma patients to determine if they have cancer at other sites in their body.

### Staging

For malignant bone tumors, most surgeons use the MSTS (musculoskeletal tumor society) staging system. Low-grade tumors are less aggressive and in general have less ability to metastasize when compared with high-grade ones.

Stage	Grade/Compartment	Metastasis
IA	Low, confined to bone	n
IB	Low, bone tumor with soft	N
	tissue mass	
IIA	High, confined to bone	n
IIB	High, bone tumor with soft	n
	tissue mass	
III	Any	у

### Biopsy

Biopsy for malignant bone tumors is always necessary to confirm the diagnosis. The biopsy is usually done in the setting of an outpatient minor surgery. Often tissue is sent as a frozen section so that the diagnosis is known the same day. Minimally invasive core needle biopsy or minimally invasive open biopsy is the current standard. Biopsy of musculoskeletal tumors should only be performed by surgeons who do this procedure regularly. Inappropriate biopsy can lead to significant preventable morbidity and complications

#### **Treatment**

Treatment for individual bone malignancies is complex and should only be undertaken by specialists who treat these routinely. A team approach (medical oncologists, radiation oncologists, among others) is often required to ensure the best possible outcome.

# Benign

Bone forming tumors

Osteoid Osteoma

Osteoblastoma

Bone Island

Osteopoikilosis

Osteoma

Melorheostosis

Cartilage Tumors

Enchondroma

Maf

Osteochondroma

Chondroblastoma

Chondromyxoid Fibroma

**BPOP** 

Periosteal chondroma

Fibrous Tumors

Benign Fibrous Histiocytoma

Solitary Fibrous tumor

Non-ossifying fibroma/fibroxanthoma

Desmoplastic Fibroma

Fibrous Dysplasia

Liposclerosing Myxofibrous Tumor of Bone

Osteofibrous dysplasia

Vascular Tumors of Bone

Hemangioma

**Arteriovenous Malformation** 

Glomus Tumor

Other

Giant Cell Tumor of Bone

Aneurysmal Bone Cyts

Simple/Unicameral Bone Cyst

**Brown Tumor** 

Intraosseus ganglion/Geode

Eosiniphilic Granuloma

Lipoma of Bone

# **Malignant Bone Tumors**

# **Bone Forming Tumors**

Osteosarcoma

Conventional

Periosteal

Parosteal

High Grade Surface Osteosarcoma

Secondary Osteosarcoma

Small cell osteosarcoma

Telangiectatic

# **Cartilage Tumors**

Chondrosarcoma

Primary

Secondary

Clear Cell

Mesenchymal

Ewing's Sarcoma/PNET

Lymphoma of Bone

Multiple Myeloma

Fibrous Tumors

Fibrosarcoma

Malignant Fibrous Histiocytoma of Bone

#### Vascular Tumors

Angiosarcoma

Hemangioendotheliama

# Other

Leiomyosarcoma of bone

Adamantinoma

Chordoma

Radiation induced sarcoma

#### Metastatic Bone Disease

Renal

Thyroid

Breast

Prostate

Lung

Melanoma

Other

#### **Soft Tissue Tumors**

The most common complaint of patients with a soft tissue tumor is a mass. Usually, there is no pain, but occasionally pain is the presenting complaint. The vast majority of soft tissue tumors are benign; however, diagnosis and treatment should only be undertaken by surgeons who routinely treat soft tissue tumors.

# **Benign**

Benign soft tissue tumors are outlined below. The most common presenting symptom is a mass, or "growth". Depending on the type of tumor, they may grow slowly or remain at a stable size. Treatment is observation for asymptomatic tumors. Tumors that are symptomatic may be excised. Imaging by MRI may be required prior to excision for larger tumors.

# Malignant

Malignant soft tissue tumors (soft tissue sarcoma) represent a rare family of malignant tumors. Multiple imaging tests are routinely ordered for a suspected soft tissue sarcoma, and tissue diagnosis by biopsy is necessary to appropriately diagnose and treat sarcoma. Treatment of these rare tumors is complex, and may consist of chemotherapy, limb sparing surgery, and radiation.

# Benign

**Fibrous Tumors** 

Nodular fasciitis

Aggressive Fibromatosis/Desmoid Tumor

Proliferative myositis

**Fibroma** 

Elastofibroma

Fibrous histiocytoma

**Lipomatous Tumors** 

Lipoma

**Muscle Tumors** 

Leiomyoma

Blood/lymphatic vessel tumors

Hemangioma

Lymphangioma

**Glomus Tumor** 

Synovial Tumors/Joint tumors

Pigmented Villonodualr Synovitis

Giant Cell Tumor of tendon sheath

**Synovial Chonromatosis** 

Lipoma Arborenses

Nerve Tumors

Neuroma

Schwannoma Neurofibroma

Other

Myxoma Ganglion Cysts

# **Malignant Soft Tissue Tumors**

Fibrous Tumors

Fibrosarcoma

Infantile fibrosarcoma

Myxofibrosarcoma

Malignant Fibrous Histiocytoma

Dermatofibrosarcoma Protuberans

Malignant Fibrous Histiocytoma

**Unclassified Sarcoma** 

Low Grade

High Grade

**Lipomatous Tumors** 

Atypical Lipoma

Liposarcoma

Myxoid Liposarcoma

Muscle tunors

Leiomyosarcoma

Rhabdomyosarcoma

Vascular Tumors

Angiosarcoma

Kaposi's Sarcoma

Hemangiopericytoma/Malignant Solitary Fibrous Tumor

Nerve Tumors

Malignant Peripheral Nerve Sheath Tumors

Primative neuroectodermal tumors

Ewing's Sarcoma

Cartilage and Bone

Extraskeletal Chondrosarcoma

Extraskeletal Osteosarcoma

# Other

Clear Cell Sarcoma (malignant melanoma of soft parts)

Synovial Sarcoma

Epithelioid Sarcoma

Alveolar Soft Parts Sarcoma

Advanced Skin Cancer

Subungal Tumors Bone invasion Melanoma, Squamous Cell Carcinoma

# **Treatment of Bone/Soft Tissue Tumors**

# **Benign Bone**

Observation for asymptomatic tumors Serial x-rays, CT, or MRI may be used

# **Curretage/grafting**

The tumor is scraped out of the bone. The resultant defect is replaced with bone graft substitute, or bone cement. This is usually done as an outpatient, but occasionally an overnight stay is required for larger tumors or if hardware is put in. Biopsy is often performed prior to this procedure to confirm the diagnosis.

# **Prophylactic Fixation**

If the tumor is large enough to significantly alter the structural properties of the bone, then plates, screws, and rods are often used to stabilize the bone so it does not fracture.

# **Radiofrequency Ablation**

For certain tumors (osteoid osteoma) a probe is inserted into the tumor and used to heat the tumor with minimal damage to surrounding tissues. This is usually done as a day surgery

#### **Malignant Bone tumors**

Depending on the type of tumor that is diagnosed, treatment may consist of a combination of chemotherapy and surgery. Some tumors, such as chondrosarcoma, are usually only treated with surgery.

#### Wide Resection

The tumor is removed with a normal cuff of tissue around it. More than 95% of these procedures are limb-sparing.

#### Reconstruction

Endoprosthesis- a metallic joint replacement is used to reconstruct the part of the bone/joint removed.

Allograft- cadaveric bone used to replace the defect Free Fibula grafting- the patients fibula is used to reconstruct the defect. Microvascular anastamosis is used to provide a blood supply to the bone.

#### Treatment of Metastatic Bone Disease

Several issues are key in treating patients with metastatic bone disease that has spread from other sites to the skeleton. Experience with these patients is necessary to formulate the best reatment plan. Breast, Prostate, Lung, Thyroid, and renal cell carcinoma are the most common tumors to spread to the skeleton. Treatment of these tumors is generally palliative and includes the following:

# **Prophylactic Fixation**

A metastatic tumor has destroyed enough of the bone that it is structurally unsound. A metal rod is placed in a minimally invasive procedure to stabilize the bone so that fracture does not occur.

#### **Biopsy**

Often a biopsy is needed to determine if the assumed diagnosis of metastatic cancer is correct.

# Radiofrequency ablation

Thermal energy is used to kill tumor cells. This is minimally invasive and is used in tumors where radiation has been unsuccessful, or in painful tumors in high risk areas. Often, this procedure is combined with cementoplasty (see below)

# **Cementoplasty**

Bone cement is injected into tumors to provide support. Commonly used around the pelvis, we commonly combine this treatment with radiofrequency ablation and radiation to provide optimal results.

#### Resection

Survival benefit has been shown to occur with resection of certain types of cancer. In cases of renal cell carcinoma, thyroid carcinoma, and melanoma, removal of metastasis can provide survival benefit. The decision to resect a maetastasis is made by the medical and orthtopaedic oncologist.

### Benign soft tissue

In general, asymptomatic tumors are observed. Occasionally MRI is required to evaluate the tumor completely. Removal is reserved for large and/or symptomatic tumors. Most of this can be done as outpatient surgery.

#### Malignant soft tissue

A multidisciplinary team consisting of the musculoskeletal oncologic surgeon, medical oncologist, and radiation oncologist usually determines optimal treatment of soft tissue sarcomas. Depending on the type of tumor, all modalities may be used. Low-grade tumors usually do not require chemotherapy and radiation. All malignant tumors are removed unless there is a contraindication to surgery.

#### Wide resection

The tumor is removed with a wide cuff of normal tissue around it. Limb sparing surgery is possible in 95% of cases. Usually, several days in the hospital to recover are necessary.

# Treatment of tumor bed

Occasionally, sarcomas are inadvertently excised when surgeons believe that they are treating a benign tumor. When this happens, in general, an MRI is performed to evaluate the tumor bed. Repeat excision of the tumor bed is performed for high-grade tumors followed by radiation. Low-grade tumors are often observed, provided there is no gross tumor present in the bed by MRI.

# **Information on Complex Reconstruction**

Primary Joint Replacement

Hip/Knee-http://www.stryker.com/en-us/education/PatientEducation/028940

shoulder, elbow http://www.zimmer.com/

#### **Revision Joint replacement**

Unfortunately, joint replacement procedures can fail for a variety of reasons, and often revision surgery is required to restore function. Dislocation, infection, and loosening are the main problems that often require revision surgery.

Hip, knee, shoulder, elbow
Use of hinged, megaprosthesis
Treatment of recurrent dislocation/instability
Use of Achilles tendon allograft to reconstruct abductor

# Treatment of infected total joint replacement

Any joint replacement can become infected. Treatment of these difficult problems requires expertise to achieve optimal outcome

2 stage revision

The joint is removed and a temporary spacer is inserted. Often, an articulating spacer is perfered to allow weight bearing and range of motion. Antibiotics are administered for at least 6 weeks. After two weeks off antibiotics, the joint is aspirated to make sure the infection is gone. If the aspirate looks fine, then a revision total joint replacement is performed

#### Treatment of complex and basic fractures

Periprosthetic fracture Complex articular fractures, especially needing replacement Hip fracture

### **Treatment of tendon ruptures**

Primary rupture of the Achilles, patellar, and quadriceps tendon Secondary reconstruction for neglected ruptures Reconstruction for failed repair of tendon rupture Periprosthetic tendon rupture Extensor mechanism rupture after TKR Extensor Mechanism Allograft

# Treatment of avascular necrosis

Core decompression, Bisphosphonates JOINT REPLACEMENT' FREE FIBULA GRAFTING