Tumors for the general orthopedist: how to save your patients and practice

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It is likely that most orthopedic surgeons will see a patient with a benign or a malignant musculoskeletal tumor sometime during their career. However, because of the rarity of these entities, many surgeons may benefit from a review of how to evaluate a patient with a bone lesion or a soft-tissue mass. A logical approach is necessary for the evaluation of imaging studies as well as in the workup of children and adults with a possible tumor. It is important to have a good working relationship with a musculoskeletal radiologist to assist in interpreting the images. If the treatment algorithms lead to a conclusive diagnosis of a benign bone tumor, benign soft-tissue mass, or metastatic bone disease, the orthopedic surgeon may choose to definitively treat the patient. If the workup indicates an indeterminate lesion, it may be prudent to discuss the situation with an orthopedic oncologist or transfer the care of the patient to a physician with more specialized knowledge. A careful, logical workup is needed before surgery to limit risks to the patient and optimize the chances for a favorable outcome.

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Introduction

Practicing orthopedic surgeons are likely to periodically encounter a patient with a benign or a malignant musculoskeletal neoplasm during their careers. Approximately 12 000 new malignant musculoskeletal neoplasms are diagnosed each year in the USA, and it is important to categorize them correctly. Benign lesions are encountered much more often than malignant lesions.

Interpreting imaging studies of musculoskeletal tumors

Recognizing the imaging characteristics and anatomic details of musculoskeletal tumors from different imaging modalities provides the orthopedic surgeon with considerable information and generally leads to an appropriate differential diagnosis.

Conventional radiography

Good-quality biplanar radiographs remain the gold standard for evaluating bone lesions and providing useful information on soft-tissue lesions, such as the presence of mineralization, bone involvement, or reactive changes. Plain radiographs should be obtained first [1]. Attention should be paid to the zone of transition between the lesion and the bone. The Lodwick classification system helps determine the biologic activity of the lesion and response of the bone [2]. Type I lesions are geographic and have a sclerotic (type IA), well-defined (type IB), or poorly defined (type 1C) margin. Type II lesions appear moth eaten (confluence of small lytic areas in bone) and suggest more aggressive processes (Fig. 1). Type III lesions are permeative with diffuse destruction. Generally, an increasing number in the Lodwick classification system corresponds to the increasing aggressiveness of the lesion.

The characteristics of the lesion help to differentiate aggressive from benign lesions. Larger lesions tend to be more aggressive. Cortical destruction, poor margination, and extension into the soft tissues are generally characteristics of a malignant tumor. Evaluation of periosteal patterns, if present, can also be helpful. Benign or low-grade lesions generally show thick, wavy, and uniform changes on the bone surface, reflecting a slow response to the underlying condition. Lamellated (onion skin), spiculated (sunburst), or Codman triangle periosteal changes reflect aggressive lesions that are 'blowing out' the bone.

The location of the lesion provides clues to an appropriate differential diagnosis (e.g. chondroblastomas or giant cell tumors in the epiphyseal area, unicameral bone cysts in the proximal humerus or proximal femur). Patient age is similarly helpful in the diagnosis because many tumors manifest in specific age groups (e.g. Ewing sarcoma in teenagers and multiple myeloma in older adults) [3].

Matrix mineralization is another feature that provides useful diagnostic clues. For example, the presence of stippled calcifications (rings and arcs) indicates a chondroid tumor. Amorphous ossification is common in osteosarcoma or

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osteoblastic metastasis. Fibrous dysplasia shows a 'ground-glass' appearance; a central, swirling 'smoke up the chimney' mineral pattern is consistent with a bone infarct [4].

Bone scintigraphy

Bone scintigraphy (bone scanning) is useful in determining whether a bone lesion is active (increased uptake) or indolent. The scan actually shows bone formation (in response to a stimulus) and blood flow; slow-growing or inactive lesions typically show minimal radiopharmaceutical uptake, whereas active bone-forming lesions, irrespective of their biologic potential, typically show intense uptake. This test is generally sensitive in screening for distant metastatic bone disease, regional skip lesions within the bone, and radiographically occult disease. The transition from mild to intense radiopharmaceutical uptake in a known lesion is generally a poor prognostic sign. Multiple myeloma, renal cell carcinoma, thyroid cancer, and rapid infection with necrosis may show little uptake, yielding a false-negative result [5,6].

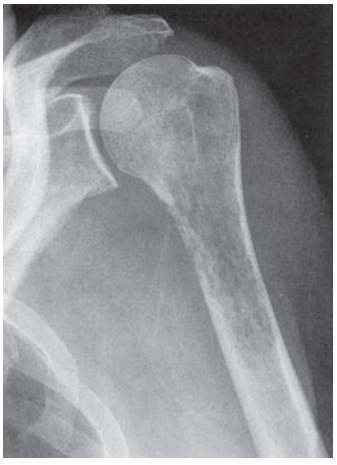
Computed tomography

Computed tomography (CT) is particularly useful for identifying mineral densities in bone and soft-tissue lesions. This modality helps differentiate chondrogenic (calcification), osteogenic (cloud-like bone formation), and soft-tissue lesions that may have calcifications (hemangiomas, liposarcomas, and synovial sarcomas) in addition to myositis ossificans. CT also readily shows cortical thinning or breakthrough, the zone of transition (margination), and ostcolytic and sclerotic changes in fiat bones [7]. CT is helpful in the evaluation of axial skeletal (pelvis, spine) lesions where the anatomy is complex and in planning and performing needle biopsies and/or thermal ablation of lesions (Fig. 2) [8]. A contrast-enhanced CT scan of the chest, abdomen, and pelvis is one of the principal imaging tools used to detect the primary source of metastatic disease to bone.

Magnetic resonance imaging

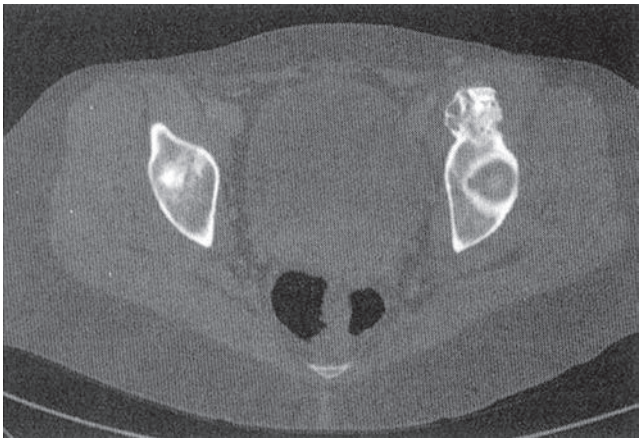
MRI provides superior soft-tissue contrast and fine anatomic detail. This modality improves the physician's ability to appropriately stage musculoskeletal tumors and adequately plan limb-salvage surgery. Various sequences, which are influenced by relaxation and echo times between pulses, can be manipulated to maximize the visualization of different tissue types on the basis of their signal characteristics. T1-weighted images (short echo and relaxation times) produce the greatest anatomic detail and tend to be the best method for evaluation of the bone marrow, which is largely composed of fat in an adult. Marrow replacement from a tumor or an infection tends to be at least as dark as adjacent muscle. Fat, methemoglobin (a blood product), gadolinium, proteinaceous fluid, melanin, and particulate calcium appear bright (white) on T1-weighted MRI scans [9]. Reduced venous flow, surgical packing, and hemostatic agents may also be bright on a postoperative T1-weighted MRI scan [10]. T2-weighted images (longer pulse sequences) are best for contrasting the difference between normal and abnormal tissues. Edema, cystic lesions, joint or synovial fluid, most tumors, and free water appear bright (white) on T2-weighted images (Fig. 3). Fluid–fluid levels, the layering of different density materials (e.g. blood and transudate or exudate), are common in lesions including an aneurysmal bone cyst, telangiectatic osteosarcoma,

Figure 1



Radiograph showing a lymphoma in the humerus with the moth-eaten appearance typical of round cell tumors.

Figure 2



Computed tomography scan showing a benign osteochondroma arising from the anterior acetabular area. These lesions are poorly visualized with plain radiographs.

and synovial sarcoma. Other lesions, such as complex unicameral bone cysts, chondroblastoma, giant cell tumor, and fibrous dysplasia, may also show fluid–fluid levels.

Gadolinium-contrasted T1-weighted images are used to further define tumor involvement and help distinguish between a tumor and postoperative scar tissue [11]. It should be noted that tumoral hyperemia associated with neoplasia shown on gadolinium T1-weighted images can occasionally lead to an overestimation of the amount of tissue involvement by tumor. Gradient echo sequences (short relaxation, low angle) are useful in the evaluation of subtle calcifications or chronic blood products because of their sensitivity to signal loss from these tissues (bloom artifact). Certain conditions, such as pigmented villonodular synovitis, hemophilic hemorrhagic synovitis, and synovial chondromatosis, may be best shown with this sequence. Although MRI sensitivity is impaired by the presence of large metal implants, a prescanning consultation with a musculoskeletal radiologist can often produce a scan with useful information. Modified fast spin echo and inversion recovery techniques can reduce the intravoxel dephasing and frequency misregistration adjacent to these implants [12].

Despite the limitations associated with fine bony detail, subtle calcification, and implanted hardware, MRI is the modality of choice for the evaluation of many musculoskeletal tumors.

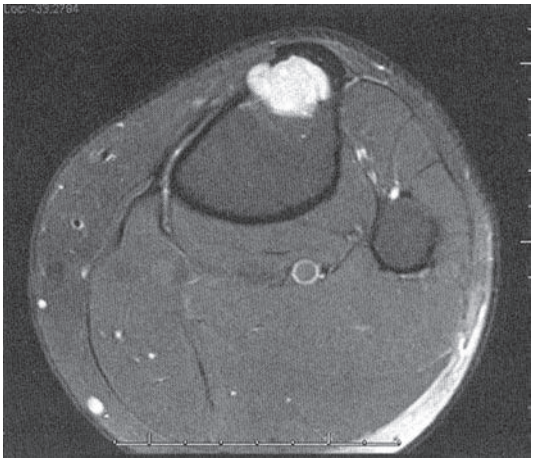
Positron emission tomography

PET is a functional imaging modality that is based on the presumption that neoplastic tissue has an increased metabolism compared with surrounding baseline tissue(s) and can be identified using radioactive glucose [13]. To increase its localization sensitivity, PET is often fused to a CT scan (PET-CT). To assess lesions, 18F-fluorodeoxyglucose (18F-FDG) is typically used and is quite sensitive in identifying small and/or skip lesions. Increased 18F-FDG uptake is usually correlated with an increasing degree of tumor aggressiveness [13]. Typically, a standard uptake value greater than 2.0 or a tumor-to-background ratio greater than 3.0 is a cause for concern (Fig. 4) [14]. The wide overlap between normal and abnormal tissue leads to a relatively low specificity with a PET scan. PET is less useful in the diagnosis of inflammatory diseases such as osteomyelitis, Paget disease, fibrous dysplasia, and osteoblastic bone metastases. Sarcomas with extensive extracellular matrix can dilute the 18F-FDG signal, leading to an underestimation of the biologic aggressiveness of the lesion. In the future, PET may offer benefits in estimating tumor necrosis after neoadjuvant chemotherapy. A decreasing standard uptake value may be correlated with necrosis and, theoretically, will allow earlier or changing intervention for tumors that are responding poorly to treatment [14,15].

Approach for the older patient with a destructive bone lesion

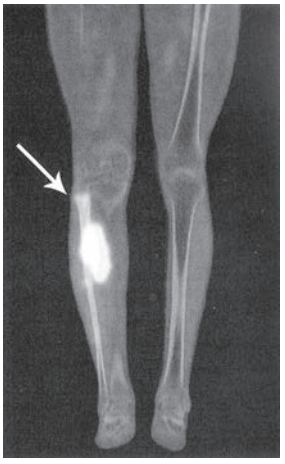
In patients older than 40 years of age, destructive bone lesions will usually be diagnosed as metastatic carcinoma. Primary malignant bone tumors occur less commonly in this age group, and the surgical treatment is quite different for this subset of patients. A careful, stepwise approach to the evaluation of an older patient with a destructive bone lesion will minimize the chance of a misdiagnosis that

Figure 3



T2-weighted axial MRI scan of the tibia showing plasmacytoma with early cortical breakthrough of the soft-tissue mass.

Figure 4



PET-computed tomography fusion image of fibular Ewing sarcoma showing intense uptake.

could compromise the optimal outcome for the patient [16,17].

Evaluation patient history

An appropriate patient evaluation begins with a thorough assessment of history [16,17]. Because destructive bone lesions are almost always painful, the nature of the pain, such as timing, duration, location, relation to weight bearing, exacerbating factors, and medications needed for pain relief, must be determined. Progressive pain that occurs at rest and at night is more suggestive of a malignant process. Constitutional symptoms, such as weight loss and lethargy, are important. Determining a personal or a family history of cancer is critical. It is important to keep in mind that cancers can metastasize more than 10 years after the primary site is treated. A history of radiation treatment or Paget disease may suggest the possibility of a secondary malignancy. Dates of screening mammograms, prostate examinations, and colonoscopies should be recorded. Cancer in any bodily site can metastasize to bone, but the most common primary sites of metastatic bone cancer include the breast, the lung, the prostate, the kidney, and the thyroid. Symptoms related to abnormal function of these organs should be documented.

Physical examination

The physical examination should be complete and should focus on the site of the painful lesion as well as the surrounding areas (e.g. the entire affected extremity) [16,17]. Potential primary sites of cancer (such as the breast, prostate, lung, or thyroid) should be evaluated. Any decreased joint range of motion, swelling, tenderness, neurologic deficits, gait abnormalities, and lymphadenopathy should be documented.

Laboratory studies

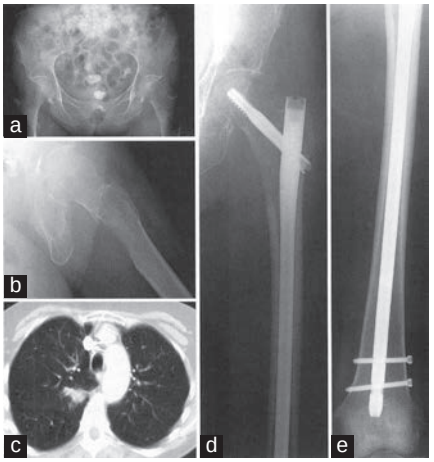
A complete blood count with a differential, basic metabolic panel, and urinalysis should be performed. Normal laboratory studies do not eliminate the possibility of malignancy or metastasis, but abnormal results are sometimes helpful in decision making. Anemia may suggest multiple myeloma, hypercalcemia occurs with hyperparathyroidism or widespread bone metastasis, and microscopic hematuria is associated with renal cell carcinoma. Alkaline phosphatase, phosphorus, and vitamin D levels may be helpful in identifying a particular metabolic bone disease. Other studies such as serum and urine electrophoresis, thyroid function tests, the prostate-specific antigen test, and other tumor markers can be ordered as necessary depending on radiographic findings and clinical suspicions.

Imaging

Plain radiographs of the destructive bone lesion will often provide clues to the eventual diagnosis [16,17]. Most bone metastases are osteolytic, with the exception of prostate and some breast cancers, which create osteoblastic metastases. Multiple myeloma bone lesions are described as punched-out holes without a visible matrix within the lesion. Lymphoma within the bone may not be visible on plain radiographs. Some bone tumors or metabolic abnormalities have unique characteristics (ossification in osteosarcoma, calcifications in chondrosarcoma, and coarsened trabeculae in Paget disease). The location of the bone lesion can be helpful. Most bone metastases occur in the thoracic spine. A lesser trochanteric avulsion implies an underlying pathologic process and impending hip fracture (Fig. 5). Chordoma typically affects the sacrum. It is important to obtain orthogonal radiographic views and to image the entire bone.

Additional imaging is usually necessary for a definitive diagnosis. A total-body ^{99m}Tc bone scan is a screening study for additional bone lesions and is often positive in metastatic disease. Because a bone scan documents osteoblastic response, it is usually falsely negative in patients with multiple myeloma.

Figure 5



(a) Anteroposterior pelvic radiograph of a 67-year-old woman with left hip pain. Note the absence of the left lesser trochanter, indicating a destructive lesion. (b) A lateral left hip radiograph better defines the osteolytic lesion, which places this patient at high risk for a pathologic fracture. A thorough assessment of history and physical examination indicated no history of cancer, a 30-pack per year history of cigarette use, pain on range of motion of the left hip, and an antalgic gait. (c) A full radiographic staging workup was performed. This chest computed tomography (CT) scan shows a probable primary lung cancer in the right upper lobe. Of note, the patient's abdominal CT scan showed multiple liver lesions and vertebral metastasis. A needle biopsy of the liver lesions showed metastatic lung cancer. Anteroposterior radiographs of the proximal (d) and distal (e) left femur after the placement of an intramedullary femoral reconstruction nail. A bone sample after reaming was sent to pathology to confirm the metastatic disease. This method of stabilization prevents a future femoral neck or diaphyseal fracture.

A CT scan of the chest, abdomen, and pelvis will usually identify a primary site of cancer in metastatic bone lesions [16]. A skeletal survey is suggested if the appearance of the bone lesion suggests multiple myeloma. Additional imaging, such as an 18F-FDG-PET scan, is not used routinely in the workup of a destructive bone lesion nor is it used for a staging workup for a high-grade sarcoma; however, it is frequently used to stage and measure treatment response in patients with lymphoma. MRI is not necessary to define a destructive bone lesion unless it occurs in the spine, where it often cannot be clearly defined on plain radiographs. An MRI of the spine can also help differentiate between a vertebral compression fracture caused by osteoporosis and a fracture caused by metastatic disease.

Biopsy

Sometimes, the previously described workup will lead to a definitive diagnosis (Fig. 5); however, if there is any uncertainty, a biopsy must be performed. Bone lesions can be diagnosed by a needle biopsy under image guidance or by an incisional biopsy. If experienced radiologists and pathologists are available, a needle biopsy can be performed expediently with minimal stress to the patient and a high rate of accuracy. With sophisticated immunohistochemical techniques, the primary site of a bone metastasis can often be identified, even if it was not apparent from the imaging workup.

If the workup and biopsy lead to a diagnosis of a primary malignant bone tumor, such as osteosarcoma, chondrosarcoma, chordoma, or malignant fibrous histiocytoma, referral to an orthopedic surgeon with expertise in treating these conditions is warranted. If the lesion is a lymphoma, referral to a medical oncology team is suggested. Most destructive bone lesions caused by lymphoma are secondary to the systemic disease and will often heal with chemotherapy, thereby precluding surgical treatment. If the lesion is metastatic bone disease or multiple myeloma, surgical stabilization may be warranted. A proper understanding of the indications for fixation and the oncologic principles for the treatment of metastatic bone disease is essential; much of this information can be found in the literature.

Associated medical disorders

Patients with metastatic bone disease should be treated by a multidisciplinary approach because their medical condition often requires expertise from physicians specialized in radiation oncology, medical oncology, interventional radiology, neurosurgery, and orthopedic surgery. Depending on the extent of disease, patients may have physiologic disruptions related to the cancer [17]. Hypercalcemia is present in 10–15% of patients with bone metastasis, most commonly from lung and breast cancer. Early symptoms include polyuria and/or polydipsia, fatigue, weakness, and anorexia. Late symptoms include irritability, depression, nausea and/or vomiting, vision abnormalities, and coma. Hypercalcemia is a potentially life-threatening condition and requires hydration and possible intravenous bisphosphonate therapy. Other physiologic disruptions relate to the hematopoietic system (anemia), thromboembolic disease (hypercoagulability), and constipation (often related to the extensive medication required for adequate pain control).

Biomechanical considerations and indications for stabilization

Patients with metastatic bone disease often present with a pathologic fracture. After stabilization with a brace or traction, the patient should be evaluated as described previously. If a patient presents with a destructive lesion, it is necessary to determine whether the lesion places the patient at risk for an impending fracture. A stress riser in the bone occurs whenever there is cortical destruction. When the length of an open section longitudinal bone defect exceeds 75% of the bone diameter, there is a 90% reduction in torsional strength. Cortical defects also reduce the bending strength of the bone. Indications for fixation of an impending fracture have been studied, but remain somewhat imprecise because fractures are not entirely predictable and depend on other factors, including patient comorbidities and balance, histology of the tumor, and underlying general bone density [18,19]. Lesions are more likely to cause a fracture when they are osteolytic, located in the peritrochanteric region, affect more than two-thirds of the cortex, and are painful. The decision to surgically treat extremity lesions is usually made on the basis of plain radiographs; further local imaging is not necessarily required, although CT can better define the cortical integrity.

Noninvasive or minimally invasive alternative treatments that can be used alone or in combination with surgical treatment of metastatic bone disease include bisphosphonates, external beam radiation, Cyber Knife (Accuray, Sunnyvale, California, USA) radiation, radiofrequency ablation, cryoablation, embolization, vertebroplasty, and kyphoplasty.

The complete technical details related to the stabilization of actual or impending fractures caused by metastatic bone disease or multiple myeloma are beyond the scope of this paper [20]; however, a few technical pearls should be considered: (a) methylmethacrylate can be used to supplement fixation when necessary;

(b) prosthetic replacement should be used for pathologic femoral neck fractures rather than in-situ pinning or hip screw/side plate constructs; (c) the entire femur should be stabilized with a reconstruction-type nail (Fig. 5) irrespective of where the femoral lesion or fracture is located; (d) highly vascular lesions (such as renal cell carcinoma or thyroid cancer metastasis) should be embolized if a tourniquet cannot be used; (e) postoperative external beam radiation can be used to supplement fixation after the skin heals to minimize the risk of disease progression and continued pain.

Approach for the young patient with a destructive bone lesion

Three goals are necessary in the treatment of a destructive bone lesion in a child. First, a diagnostic strategy is needed that leads to an effective and confident treatment plan. Second, an error that will result in the loss of the patient's life or limb must be avoided. Third, the treatment strategy should minimize unnecessary tests and interventions. It is important that the surgeon makes sound recommendations to the child and the parents. These recommendations should provide a sense of competence and reassurance and should advise them of the need for the necessary tests and treatments that will achieve the best possible outcome.

Children differ from adults in many ways. They frequently have unrecognized infections and incidental trauma. As a result, most bone lesions are post-traumatic or inflammatory. Both children and adults may have metabolic bone disease but, with the exceptions of Paget disease, osteoporosis, and renal ostcodystrophy, most metabolic bone diseases are diagnosed in patients younger than 20 years of age. Children frequently have developmental abnormalities or reactive, non-neoplastic lesions visible in bone. These non-neoplastic lesions often heal in children near the time of skeletal maturity. In contrast to adults, skeletal metastases are relatively rare in children, and primary malignant bone tumors in children are often high grade and associated with significant mortality.

Because there is a low incidence of primary malignant bone tumors in children (fewer than 2000 cases/year in the USA), most surgeons have only a vague familiarity with the evaluation and treatment of these conditions. Similarly, radiologists involved in the interpretation of imaging studies may lack education and experience in reviewing primary malignant bone tumors in children and may provide opinions that are too generic to be helpful, are inaccurate, or lead to unnecessary tests and procedures.

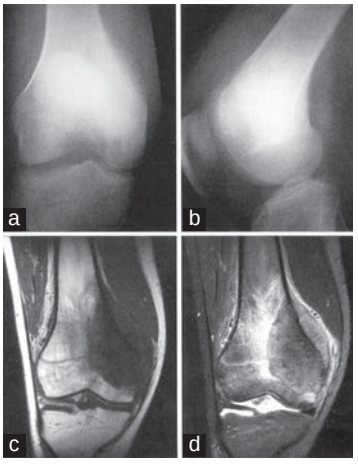
Children and parents may not be able to provide an accurate patient history. Children often have vague symptoms that may not be reported to the parents. These may be interpreted, often correctly, as growing pains, muscle pulls, strains, and sprains. Once a bone abnormality is diagnosed, anxiety, guilt, and a need for a clear answer will be expressed by the parents. It is important to use a diagnostic rationale that will achieve the treatment goals and lessen anxiety for everyone who is involved. This scheme will involve a series of decision points with proposed action steps.

Findings suggestive of a primary malignant bone tumor

If the child has a history of a focal lesion, increasing pain, physical findings of a soft-tissue mass and/or localized tenderness, and imaging findings consistent with a malignant tumor, a decision is needed on whether the orthopedic surgeon will provide care or whether the child will be referred to a trained orthopedic oncologist (Fig. 6). If the lesion is destructive, the child's ability to bear weight should be protected with an assistive device, and the family should be advised on the risk of a pathologic fracture. If the lesion is located in the upper extremity, a sling or a splint is recommended.

Additional workup and a possible biopsy are indicated. If the surgeon is not familiar with the management of malignant bone tumors, including limb-salvage

Figure 6



Anteroposterior (a) and lateral (b) radiographs of a 17-year-old adolescent boy with right knee pain and swelling for 3 months. The radiographs show increased density in the medial femoral condyle with expansion of a mineralizing process into the adjacent soft tissues. The zone of transition is wide. These findings are most consistent with an osteosarcoma. (c) The T1-weighted MRI scan shows decreased signal within the medial femoral condyle and a soft-tissue mass extending into the vastus medialis. (d) The contrast-enhanced MRI scan shows extensive surrounding bone edema and a small effusion. The loss of the normal low signal cortex is particularly worrisome for a malignancy. A subsequent open biopsy confirmed a diagnosis of osteosarcoma.

techniques, consultation with or referral to a colleague with such experience is necessary. Studies show improved outcomes for patients who are referred to physicians familiar with the treatment of bone sarcomas [21].

The incidental bone lesion

Incidental bone findings on plain radiographs are rarely worrisome, but merit observation. Occasionally, additional testing may be necessary, but treatment is usually not indicated. The absence of symptoms is most often associated with a latent bone lesion. In contrast, most active benign and malignant bone tumors are painful.

The presence of a fracture may add uncertainty to the diagnosis. The physician should be guided by imaging characteristics as described previously. If open treatment of the fracture is necessary, a biopsy to confirm the presence of a benign tumor is usually recommended.

In attempting to determine whether a bone lesion is symptomatic, the physician should look for physical evidence of atrophy, deformity, tenderness, or a palpable mass. If the lesion is symptomatic, an active process may be involved. If there are no signs or symptoms associated with the radiographic findings, it is safe to observe most incidentally noted lesions with latent imaging characteristics. In general, follow-up examinations and radiographs at 3, 6, and 12 months are adequate to determine whether there are any worrisome changes in the appearance of the lesion.

The indeterminate lesion

Indeterminate lesions require a diagnostic strategy involving additional imaging studies and possible biopsy [22]. After a careful assessment of history and physical examination, the physician should obtain laboratory tests including a complete blood count with differential, erythrocyte sedimentation rate, and C-reactive protein level to look for infection. Rarely, the differential will indicate a hematologic malignancy. A chemistry panel including calcium, phosphate, and alkaline phosphatase is helpful if metabolic bone disease is suspected.

After the lesion is evaluated with plain radiographs, additional imaging may be required. A ^{99m}Tc bone scan can detect multifocal disease. CT can accurately assess cortical destruction or identify a nidus in an osteoid osteoma, but MRI with contrast is most helpful in determining the extent and nature of the lesion [23]. Marrow involvement may extend further than shown on plain radiographs. Soft-tissue involvement is worrisome, and MRI can define the relationship of the tumor with the surrounding neurovascular structures. Fluid-filled lesions with only peripheral enhancement arc often unicameral bone cysts. Fluid–fluid levels suggest an aneurysmal bone cyst (Fig. 7).

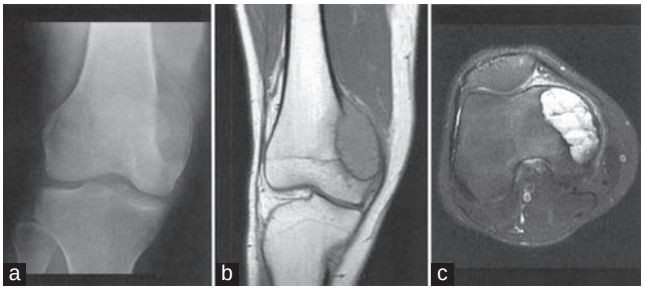
The most common primary malignant bone tumors in children are osteosarcoma and Ewing sarcoma. Both diagnoses have radiographic findings that often include cortical destruction and a soft-tissue mass. If the eventual diagnosis proves to be eosinophilic granuloma, a skeletal survey is important in staging, but it is not as important in the initial workup.

Biopsy

When the diagnosis is not clear and the imaging findings suggest an active process, a biopsy is necessary. Biopsy is a simple technical procedure but requires significant planning [24]. The surgeon should be aware of nonconventional approaches and limb salvage options that allow the biopsy tract to be resected later with the tumor if it proves to be malignant. Complications may alter the ability to perform limb salvage surgery.

Biopsies can be open (incisional) or closed (core-needle or fine-needle aspiration). If a musculoskeletal radiologist performs the needle biopsy, it is recommended that the surgeon discuss the preferred site and route of needle entry. Many bone lesions that are suggestive of a particular lesion (such as chondroblastoma or an aneurysmal bone cyst) can be

Figure 7



(a) Anteroposterior radiograph of a 15-year-old boy with 3 weeks of right knee pain. A lucency can be seen in the medial femoral condyle with expansion of the cortex and periosteum. A fine line of peripheral ossification is present. Proximal periosteal elevation is apparent. There is no internal mineralization or calcification. The zone of transition is narrow. These are characteristics of an active lesion. (b) The T1-weighted MRI scan shows a narrow zone of transition and a small rim of low signal bone about the lesion medially and is most consistent with a benign tumor. (c) The T2-weighted axial MRI scan shows fluid–fluid levels consistent with an aneurysmal bone cyst. An open biopsy with frozen section confirmed the diagnosis, and definitive surgery with curettage and bone grafting was performed at the same setting.

approached with an open biopsy because an accurate frozen section diagnosis may allow definitive treatment at the same surgical setting. It is recommended that prophylactic antibiotics be withheld until tissue cultures are obtained for bacteria, acid-fast bacilli, and fungus. Osteomyelitis can often be confused with an active bone tumor in a child.

Proper oncologic techniques involve selecting a direct approach to the tumor without exposing neurovascular structures. No flaps should be raised. The smallest necessary longitudinal incision should be used, and joint and tendon insertions should be avoided. A biopsy of the soft-tissue mass is preferable to creating a hole in the bone. If a bone biopsy is necessary, rounded edges should be made with a circular hole saw to minimize the chance of fracture. Meticulous hemostasis with polymethylmethacrylate or bone wax is important. A frozen section is important to ensure sufficient tissue for diagnosis and should be done before definitive surgery. Drains may be used but should be close to and in line with the incision.

Immediate definitive surgical treatment

Definitive treatment should be performed in patients only when the preoperative evaluation suggests a benign tumor, which is confirmed by analyzing the frozen section. If findings are inconsistent, the surgeon should achieve hemostasis, close the incision, and await findings on the basis of permanent sections. This approach requires the availability of an experienced musculoskeletal pathologist and is especially important when performing fracture fixation. Splinting or remote traction should be considered if there is any possibility that the underlying cause of the fracture is malignant. Plates and screws through a lateral approach will result in less contamination than intramedullary fixation, but neither technique is ideal.

Intralesional curettage is the definitive treatment for most benign bone lesions in children. A high-speed burr may be used to make a sufficiently large window in the bone to allow visualization and extend the curettage. Adjuvants such as polymethylmethacrylate, cryotherapy, phenol, or argon beams are often used. The approach, the extent of the resection, and the use of adjuvants and internal fixation must all be considered in the context of open growth plates in pediatric patients. Most surgeons use allograft or bone graft substitutes to reconstruct the defects. Children are followed postoperatively with serial radiographs at 3–4-month intervals for ~2 years to monitor for local relapse. A longer period of follow-up may be necessary to evaluate the development of a growth deformity in the child.

If the biopsy suggests a malignant process, such as an osteosarcoma or Ewing sarcoma, the patient should be treated by an orthopedic surgeon experienced in limb-salvage techniques who has access to a multidisciplinary team. Malignant bone tumors in children generally require neoadjuvant and adjuvant chemotherapy.

A consistent and rational approach to treating a child with a destructive bone lesion will allow a confident evaluation of the significance of the lesion, preserve life and limb, and avoid unnecessary interventions and testing. Treatment may include observation, further testing, biopsy, or definitive surgery.

Soft-tissue masses

Soft-tissue masses are common. Primary care physicians and surgeons often must evaluate patients and decide whether a soft-tissue mass should be observed, biopsied, or removed [25]. Incorrect decisions may lead to a delay in the correct diagnosis or inappropriate excisional biopsy. In a 1982 study, Mankin *et al*. [26] reported a significant rate of biopsy errors that resulted in either a change in the management plan or even unnecessary amputation. In a second study by Mankin *et al*. [21] carried out in 1992, there was no improvement in biopsy results for musculoskeletal malignancies in the 10-year interval between the two studies. Simply classifying the lesion as determinate or indeterminate can guide clinicians in making the correct decisions following the imaging evaluation of a soft-tissue tumor [27].

Evaluation

If a patient presents with a soft-tissue mass on the extremities or the trunk, a thorough assessment of history and physical examination are necessary; however, these steps may provide only a few clues to the nature of the mass. Radiography and MRI are the two essential imaging studies that are necessary for making proper treatment decisions.

Radiographic imaging

The first step in evaluating a soft-tissue mass is to obtain two orthogonal plain radiographs of the region. The radiographs are evaluated to ensure that the underlying osseous structures are normal. A fixed soft-tissue mass (one that does not move with palpation) may be the result of soft-tissue extension from an intramedullary or a surface bone tumor (one arising from the cortex).

An MRI is the most definitive imaging technique for decision making and can be performed with or without contrast. A contrast-enhanced study should be

performed if the clinician needs to differentiate a fluid-filled structure (cyst) from a solid tumor. The clinician should evaluate the MRI scan with an experienced musculoskeletal radiologist to classify the soft-tissue mass as determinate or indeterminate [28].

Evaluation and treatment of a determinate soft-tissue mass

Determinate soft-tissue masses are defined as masses for which the clinician and radiologist can establish a definitive diagnosis on the basis of imaging studies [27]. If a definitive diagnosis can be established, the clinician can plan the treatment without a biopsy. Common determinate masses include lipomas, synovial cysts, and soft-tissue hemangiomas [29].

Lipomas

Lipomas show high signal intensity on T1-weighted images and low signal intensity on T2-weightcd images (Fig. 8). Fat suppression turns the entire lesion dark (low signal). To establish the diagnosis conclusively, the entire mass must be fat suppressed in a homogeneous manner with no areas of high signal intensity. If there are areas of intermediate or high signal intensity within the lipomatous lesion, the mass may represent an atypical lipoma or dedifferentiated liposarcoma.

Synovial cysts

Cysts are common, and patients may present with a soft-tissue mass as a manifestation of an articular disorder. Cysts are fluid-filled structures and have a uniform MRI appearance. On T1-weighted images, the cyst shows very low signal intensity; on T2-weighted images, the cyst shows high signal intensity. When a cyst arises from a joint in a characteristic location, contrast enhancement is not necessary to establish a diagnosis. For example, if the stalk (area of joint communication) of a popliteal (Baker) cyst can be identified arising between the semimembranosus and medial gastrocnemius muscles, contrast is not necessary to establish the diagnosis. However, if a soft-tissue mass is identified in an atypical location, contrast enhancement is necessary to establish the diagnosis.

Soft-tissue hemangiomas

Radiographs may show characteristic phleboliths, which allow a definitive diagnosis. On an MRI scan, the features of a soft-tissue hemangioma are round structures with low signal intensity interspersed within high signal areas on T1-weighted images. The low signal, round structures represent vascular channels, whereas the areas of high signal intensity represent adipose tissue. T2-weighted images with fat suppression show round structures with very high signal intensity, with complete suppression for the previous high signal areas.

There are other determinate masses; however, the clinician and the radiologist must be able to identify the nature of the mass (Fig. 9). Some teams of clinicians and radiologists may be able to identify only a few soft-tissue masses as determinate, whereas others may be capable of identifying many masses as determinate.

Treatment options for determinate masses include observation, excisional biopsy, or percutaneous treatment. The clinician and the radiologist must establish the exact diagnosis of a mass before recommending observation to the patient. Patients may choose observation as the method of treatment for an intramuscular lipoma or hemangioma if they are asymptomatic or only mildly symptomatic.

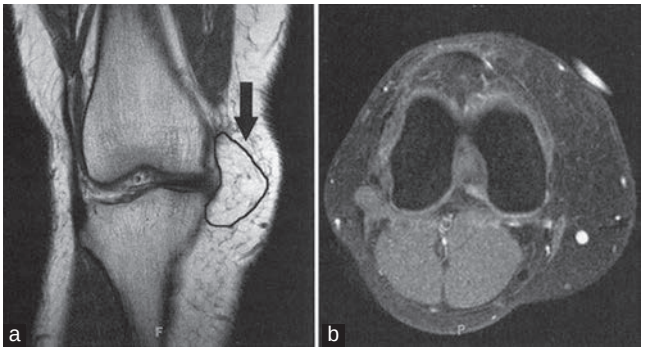
If the clinician is sure of the diagnosis, the patient may be advised to have the mass excised. If the mass is determinate, the clinician knows the diagnosis and can design the surgical procedure (or percutaneous treatment such as sclerotherapy for an intramuscular hemangioma) on the basis of the biologic behavior of the process.

Workup and treatment of indeterminate soft-tissue masses

Soft-tissue masses that cannot be identified definitively by the clinician–radiologist team are classified as indeterminate. Examples include the entire spectrum of low-grade and high-grade soft-tissue sarcomas.

There are no characteristic features of these sarcomas that enable accurate diagnosis on the basis of the

Figure 8



(a) A T1-weighted coronal MRI scan shows a soft-tissue mass along the medial aspect of the knee (arrow, outlined area). It abuts the medial capsule and has high signal intensity (same signal as the subcutaneous fat). (b) MRI scan with all of the medial soft tissues fat suppressed. The patterns shown in these images are diagnostic of a lipoma. This is a determinate mass, and treatment can be planned without a biopsy.

MRI scan (Fig. 10). Many benign entities are also indeterminate [30].

A needle or an incisional biopsy must be performed to establish a diagnosis if the soft-tissue mass has been classified as indeterminate following an MRI evaluation. An indeterminate soft-tissue mass may be a malignant (low-grade, intermediate-grade, or high-grade sarcoma or other type of cancer) or a benign process. The surgeon cannot plan treatment without establishing a diagnosis through either needle or incisional biopsy [31].

In two previously mentioned studies, Mankin *et al.* [21,26] documented a high error rate in diagnostic and technical factors following biopsy of bone and soft-tissue masses, with no reduction in management errors in the 10-year period between the studies. A possible explanation for a lack of improvement in management errors is that a method of decision making that would reduce errors was not developed. Removal (excisional biopsy) of sarcomas before a diagnosis had been established was one of the most frequently reported errors. Soft-tissue sarcomas may have an appearance similar to that of benign lesions, and both clinicians and radiologists often do not recognize that a patient has a malignant soft-tissue tumor.

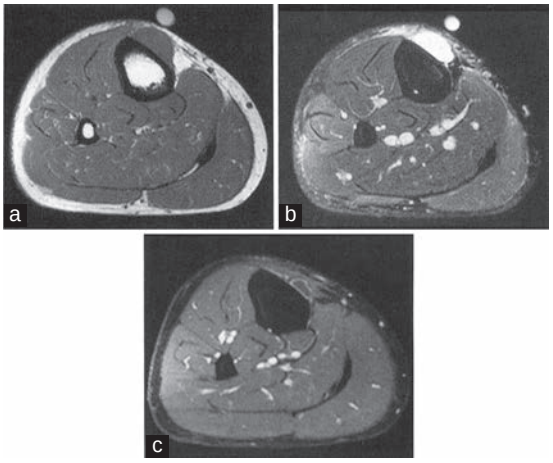
Unplanned removal of soft-tissue sarcomas has several deleterious effects on the patient's outcome. Subsequent re-excision often requires a more extensive surgical procedure with removal of all areas possibly contaminated by the unplanned excision. Of even more concern is the potential for a higher local failure rate following re-excision [32]. Several studies have documented a compromised ability to obtain wide margins following an unplanned excision [32,33].

A simple classification system that all clinicians and radiologists can use may help to reduce the number of errors in the decision-making process. The system of classifying lesions as determinate or indeterminate is simple and can be modified for all teams of clinicians and radiologists. These teams may have a small or a large number of determinate masses in their armamentarium depending on their level of experience.

Summary

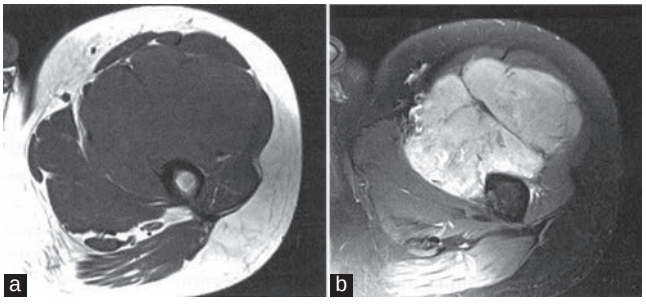
A logical evaluation of imaging studies and a stepwise, careful workup of a patient with a bone or soft-tissue lesion can help prevent mistakes in diagnosis and treatment. Patients frequently present to an orthopedic oncologist after a soft-tissue sarcoma has been inadequately excised on the basis of the false assumption that the mass was benign. Another scenario is the older patient with hip pain who is treated with a total hip arthroplasty for presumed osteoarthritis when the pain is actually emanating from an acetabular chondrosarcoma. Although benign bone lesions, benign soft-tissue lesions, and metastatic bone disease are much more common than primary malignant bone and soft-tissue tumors, the orthopedic surgeon must remain vigilant and not make assumptions that are not substantiated by the patient's history or findings from the examination and imaging studies. A biopsy is required if a conclusive diagnosis cannot be made. If the general orthopedic surgeon lacks the training, experience, or team members needed to provide definitive care, the patient should be referred to an orthopedic oncologist for treatment.

Figure 9



(a) A T1-weighted axial MRI scan of a soft-tissue mass over the medial aspect of the tibia. On this scan, the mass has uniform low signal intensity characteristics. (b) A T2-weighted axial MRI scan shows uniform high signal intensity within the lesion. (c) A contrast-enhanced axial scan shows the rim enhancement (high signal intensity at the periphery of the mass). This lesion is a cyst. Because the cyst is over the tibial periosteum, this is a periosteal ganglion cyst. This patient is asymptomatic and can be managed with observation.

Figure 10



(a) T1-weighted MRI scan showing a low signal intensity mass in the anterior compartment of the thigh. (b) T2-weighted MRI scan showing a high signal mass in the anterior compartment that is an indeterminate lesion. A needle or an incisional biopsy is necessary to establish the diagnosis.

Acknowledgements

Conflicts of interest

Dr Weber or an immediate family member serves as a board member, owner, officer, or committee member of the American Orthopaedic Association, the Ruth Jackson Orthopaedic Society, and the American Academy of Orthopaedic Surgeons Research Council. Dr Peabody or an immediate family member serves as a board member, owner, officer, or committee member of the American Orthopaedic Association and the Musculoskeletal Tumor Society, and has received research or institutional support from Biomet. Dr Frassica or an immediate family member is a member of a speakers' bureau or has made paid presentations on behalf of Stryker; serves as a paid consultant to or is an employee of Stryker; has received research or institutional support from Zimmer; has stock or stock options held in Zimmer and Stryker; and has received nonincome support (such as equipment or services), commercially derived honoraria, or other non-research-related funding (such as paid travel) from SLACK Orthopedics, Orthopedics Today, and Stryker. Dr Mott or an immediate family member serves as a board member, owner, officer, or committee member of the Mid-America Orthopaedic Society and has stock or stock options held in Johnson & Johnson. Dr Parsons or an immediate family member is a member of a speakers' bureau or has made paid presentations on behalf of Biomet and has received research or institutional support from Smith & Nephew, Tornier, Synthes, Zimmer, Medtronic, DePuy, and Biomet.

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