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Bone Tumors

A Practical Guide to Imaging

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To Ann, Alex, and Sonie, thanks for everything.

–JSW

With love and appreciation to my wonderful family and friends and with special thanks to Dr. R.E. Langevin for starting me on my radiology journey.

–MGH

Preface

The major goals of this book are to provide the reader with a practical way to analyze bone tumors and to highlight the most common tumors that one would expect to see in everyday clinical practice. We include high-yield facts and representative images for each tumor, focusing on their classic appearance and location, in order to provide a solid foundation for evaluating these tumors and to avoid the confusion that comes from including their uncommon presentations. In medicine, there are exceptions for almost everything; however, it is difficult to appreciate the extreme rarity of a chordoma in the tibia, if one does not first know that they occur almost exclusively in the axial skeleton.

The intended audience for this book is anyone interested in bone tumors, including radiologists, orthopedic surgeons, pathologists, and primary care physicians. However, we hope that the format of the book will make it particularly useful for the radiology resident who is preparing for rotations and for board exams. We imagine that this book is short enough to be read in a few days and we anticipate it being used as a quick reference during daily readouts.

We have divided the book into two sections: a didactic section and a section comprised of unknown cases. In the didactic section, we present key "bread and butter" facts and include a list of differential diagnoses with distinguishing features. We have tried to select images that illustrate the classic appearance and typical location for each tumor. Moreover, we also devote a chapter to lesions that can mimic bone tumors. In real life, physicians encounter many bone lesions that are not true tumors, but could be mistaken for a bone tumor. It would be impossible to adequately discuss the differential diagnosis for many bone tumors, without this section. In the unknown cases section, we have included a variety of cases, including: (1) tumors discussed in the didactic section but with a particular teaching point highlighted by the case; (2) examples of some unusual bone tumors; and (3) commonly-encountered "mimickers" that could be mistaken for true bone tumors. The cases are meant to be taken as if the reader were discussing them during an unknown case conference. We provide a short (usually relevant) clinical history, description of the imaging findings, a "best 3" differential diagnosis list, a short discussion that reveals the most likely diagnosis, and some relevant key facts.

It is our hope that after reading this book and working through the cases, you will find yourself with a solid foundation for evaluating bone tumors.

Jim S. Wu
Mary G. Hochman

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Contents

Abbreviations

ABC	Aneurysmal bone cyst
ADC	Apparent diffusion coefficient
AIDS	Acquired immune deficiency syndrome
AJCC	American Joint Committee on Cancer Staging
ALL	Acute lymphoblastic leukemia
AML	Acute myelogenous leukemia
AP	Anteroposterior
AS	Angiosarcoma
AVN	Avascular necrosis
BPOP	Bizarre parosteal osteochondromatous proliferation of bone
CBC	Complete blood count
CLL	Chronic lymphocytic leukemia
CML	Chronic myelogenous leukemia
CMF	Chondromyxoid fibroma
CT	Computed tomography
CXR	Chest X-ray
DF	Desmoplastic fibroma
DDx	Differential diagnosis
EIC	Epidermoid inclusion cyst
EG	Eosinophilic granuloma
ESR	Erythrocyte sedimentation rate
ES	Ewing sarcoma
FCD	Fibrous cortical dysplasia
FD	Fibrous dysplasia
FDG	Fluoro-deoxy-glucose
FLAIR	Fluid attenuated inversion recovery
FNA	Fine needle aspiration
FOV	Field of view
FS	Fat-saturated
GCT	Giant cell tumor
G-CSF	Granulocyte colony-stimulating factor
GI	Gastrointestinal
GY	Gray
HME	Hereditary multiple exostoses
HU	Hounsfield unit
IV	Intravenous
JC	Jaffe–Campanacci syndrome
LCH	Langerhan cell histiocytosis
LSMFT	Liposclerosing myxofibrous tumor
MCL	Medial collateral ligament
MFH	Malignant fibrous histiocytoma
MGUS	Monoclonal gammopathy of undetermined significance
MRA	MR angiography
MRI	Magnetic resonance imaging
NHL	Non-Hodgkin lymphoma
NF1	Neurofibromatosis type 1
NOF	Non-ossifying fibroma
NOS	Not otherwise specified
NSAIDS	Nonsteroidal anti-inflammatory drug
NSF	Nephrogenic systemic fibrosis
OFD	Osteofibrous dysplasia
PCNB	Percutaneous core needle biopsy
PET	Positron emission tomography
POEMS	Polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy
PNET	Primitive neuroectodermal tumor
PSA	Prostate specific antigen
PTH	Parathyroid hormone
PVNS	Pigmented villonodular synovitis
RCC	Renal cell carcinoma
RPMI	Roswell Park Memorial Institute (medium for flow cytometry of lymphoma)

SBC	Simple bone cyst
SBP	Solitary plasmacytoma of bone
SPECT	Single photon emission computed tomography
SPEP	Serum protein electrophoresis
STIR	Short tau inversion recovery
SUV	Standardized uptake value
T1W	T1-weighted
T2W	T2-weighted
TGFβ	Transforming growth factor beta
UPEP	Urine protein electrophoresis
US	Ultrasound
VEGF	Vascular endothelial growth factor
WBC	White blood cells
WHO	World Health Organization

1 Introduction to Bone Tumors

Focal lesions in bone are encountered frequently during everyday clinical practice. While some lesions are true neoplasms, many represent benign entities. Determining which lesions require evaluation and which should be left alone can be a daunting process. On occasion the imaging appearance is pathognomonic or highly suggestive of a specific entity; thus, imaging can play a determinative role in clinical management. Although the ultimate goal is always to arrive at a definitive correct diagnosis, this is often not possible based on the available clinical and imaging data. In practice, it is important to provide a short, reasonable list of relevant diagnoses, making sure that malignant tumors are not inappropriately omitted and that benign lesions are not overtreated. In order to do this effectively, it is important to understand some basic principles regarding the evaluation of bone tumors and their characteristic imaging features.

Classification of Bone Tumors

The World Health Organization (WHO) classification system of bone tumors provides uniformity for the reporting and treatment of bone tumors and is in common use (Table 1.1). The WHO classifies bone tumors based on their pattern of differentiation, meaning that they are grouped by the histologic tissue that they resemble. For instance, tumors that contain cartilaginous components are grouped under cartilage tumors and tumors that contain osseous matrix are grouped under osteogenic tumors. Many of these histologic subcategories contain both benign and malignant entities. Furthermore, there are several bone lesions that are not true neoplasms and therefore are not included in the WHO classification. These include bone islands, osteomas, and non-ossifying fibromas (NOF), among others. Bone islands and osteomas are hamartomas, in which normal cortical bone is seen in an atypical location. NOFs are considered developmental defects that resolve over time. Lastly, it is important to consider nonneoplastic lesions that can mimic a bone tumor on imaging, such as congenital anomalies, osteomyelitis, or posttraumatic changes. These are also not included in the WHO classification system.

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Incidence of Bone Tumors

The true incidence of each bone tumor is difficult to accurately assess and the reported numbers in the literature can be confusing, if not misleading. This is because many lesions are discovered incidentally on imaging and histologic diagnosis is not pursued in all cases. This is especially true for benign lesions, such as small asymptomatic enchondromas, osteochondromas, or intraosseous lipomas. Conversely, malignant lesions behave more aggressively and lead to symptoms if left untreated. It is therefore not surprising that three-fourths of all bone lesions that undergo biopsy are malignant. Moreover, there is more data on the epidemiology and features of malignant tumors than benign bone tumors, since malignant tumors are more likely to be evaluated.

Metastases are by far the most common tumor seen in bone. However, metastases are considered secondary tumors of bone because the primary tumor does not originate in bone. Primary tumors of bone, such as an osteosarcoma or chondrosarcoma, are actually quite rare. Prostate, breast, and lung cancers are the three most common primary sources of bone metastases. The ratio between primary and secondary (metastatic) bone lesions is believed to be around 20-to-1; however, this ratio is likely skewed in favor of metastases, since they are biopsied more commonly than benign lesions (Fig. 1.1).

Of the primary malignant tumors that occur in bone, plasma cell myeloma is most common; however, for practical purposes, it should only be considered in patients older than 40 years of age (Fig. 1.2). The various sarcomas of bone account for the next most common primary malignancies in bone. Of the primary bone sarcomas, the most common are: osteosarcoma—35%, chondrosarcoma—25%, Ewing sarcoma—16%, chordoma—8%, and malignant fibrous histiocytoma—5% (Fig. 1.3). Primary sarcomas of bone comprise only 0.2% of all neoplasms in the body and occur at one-tenth the rate of soft tissue sarcomas.

The incidence of primary benign tumors of bone is more difficult to determine since not every newly discovered lesion is biopsied or even completely evaluated. Biopsy or resection would be impractical and inappropriate in many instances if the lesion can be fully characterized as benign based on radiographic and clinical features. The most common benign neoplasms of bone are the following: osteochondroma—35%, enchondroma—20%, giant cell tumor of bone—15%, osteoid osteoma—10%, and fibrous dysplasia—5% (Fig. 1.4). Moreover, it is important to realize that even if a lesion is clearly benign based on radiographic and clinical data, the lesion nonetheless may require workup and treatment. For example, a giant cell tumor with extension to the tibial articular surface would be at risk for pathologic fracture and a fibular osteochondroma causing peroneal nerve compression would need to be resected to minimize nerve symptoms.

Table 1.1 World Health Organization (WHO) classification of bone tumors

Cartilage tumors
- Osteochondroma
- Chondroma
  - Enchondroma
  - Periosteal chondroma
  - Multiple chondromatosis
- Chondroblastoma
- Chondromyxoid fibroma
- Chondrosarcoma
  - Central, primary, secondary
  - Peripheral
  - Dedifferentiated
  - Mesenchymal
  - Clear cell

Osteogenic tumors
- Osteoid osteoma
- Osteoblastoma
- Osteosarcoma
  - Conventional
    - Chondroblastic
    - Fibroblastic
    - Osteoblastic
  - Telangiectatic
  - Small cell
  - Low grade central
  - Secondary
  - Parosteal
  - Periosteal
  - High grade surface

Fibrogenic tumors
- Desmoplastic fibroma
- Fibrosarcoma

Fibrohistiocytic tumors
- Benign fibrous histiocytoma
- Malignant fibrous histiocytoma

Ewing sarcoma/primitive neuroectodermal tumor
- Ewing sarcoma

Hematopoietic tumors
- Plasma cell myeloma
- Malignant lymphoma, NOS

Giant cell tumor
- Giant cell tumor
- Malignancy in giant cell tumor

Notochordal tumors
- Chordoma

Vascular tumors
- Hemangioma
- Angiosarcoma

Smooth muscle tumors
- Leiomyoma
- Leiomyosarcoma

Lipogenic tumors
- Lipoma
- Liposarcoma

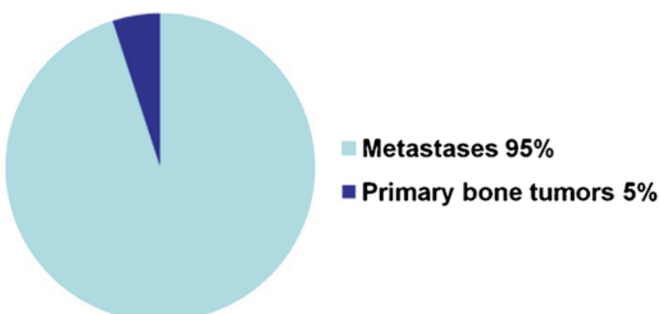
Neural tumors
- Neurilemmoma

Miscellaneous tumors
- Adamantinoma
- Metastatic malignancy

Miscellaneous lesions
- Aneurysmal bone cyst
- Simple cyst
- Fibrous dysplasia
- Osteofibrous dysplasia
- Langerhans cell histiocytosis
- Erdheim–Chester disease
- Chest wall hamartoma

Joint lesions
- Synovial chondromatosis

Reproduced with permission from: Fletcher CD, Unni KK, Mertens F, eds. WHO classification of tumours: pathology and genetics of tumours of soft tissue and bone. Lyon, France: IARC, 2002



(adapted with permission from: Murphey MD. Musculoskeletal Neoplasms: Fundamental Concepts. Radiologic Pathology. 2nd Ed. 2006)

Fig. 1.1 Bone tumors

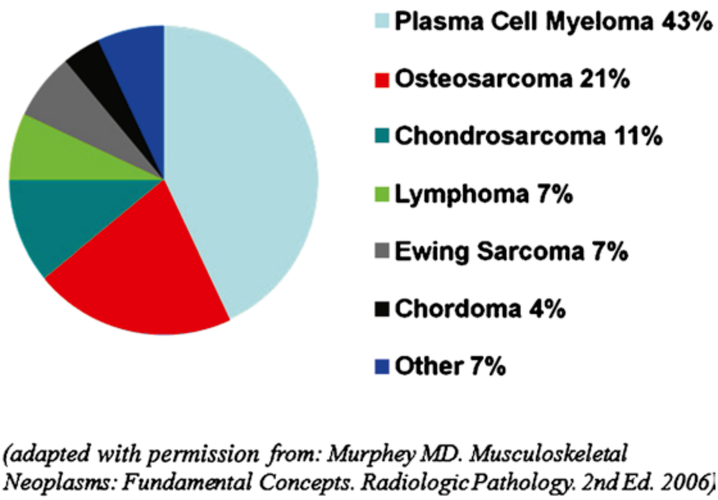


Fig. 1.2 Primary malignant bone tumors

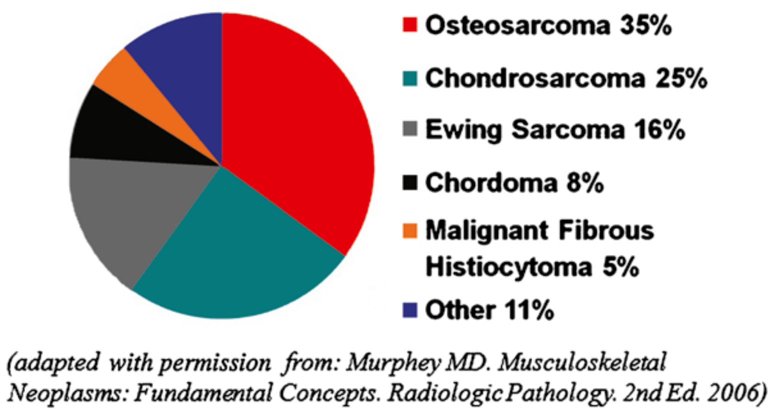


Fig. 1.3 Primary bone sarcomas

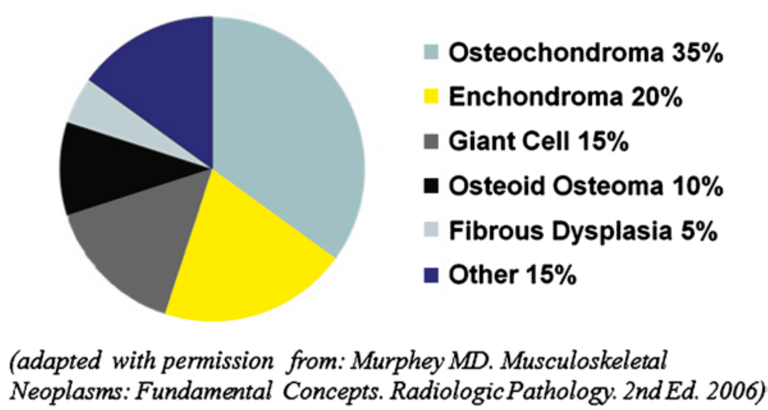


Fig. 1.4 Benign bone tumors

Value of Team Evaluation

Evaluation of a focal lesion in bone is best performed as a team effort by the clinician, radiologist, and pathologist (Fig. 1.5). All three team members have important roles that lead to the successful diagnosis and treatment of a patient with a bone lesion. The clinician provides data on the patient's history, physical examination, and laboratory values, and coordinates the patient's overall care. The radiologist performs the imaging studies and image-guided biopsy. Finally, the pathologist provides the tissue analysis which can include special stains, enzyme histochemistry, immunohistochemistry, electron microscopy, flow cytometry, and cytogenetics, leading to the identification of the lesion. Communication among all team members is vital in order to provide the best patient care.

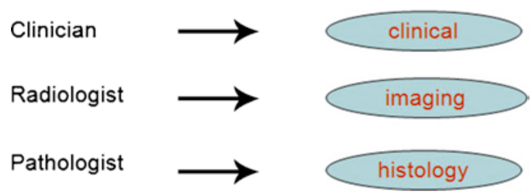


Fig. 1.5 Evaluation team

Clinical and Imaging Workup

The clinician is often the first to suspect a bone tumor. The patient may present with pain, swelling, limited mobility, weakness, and/or pathologic fracture, initiating the need for evaluation. Although pain can be nonspecific, it is invariably present with malignant tumors. Swelling occurs when the tumor enlarges and displaces adjacent tissue and often increases more rapidly with malignancies. However, both benign and malignant lesions can grow rapidly if there is bleeding or infection. Limited mobility can occur if the tumor develops around a joint. Lastly, any fracture may be the result of an underlying bone lesion. This is especially true if the degree of force is below the expected threshold to create a fracture or if the mechanism of injury is atypical. Children often present with pathologic fractures through benign lesions, such as non-ossifying fibromas (NOFs) and simple bone cysts. In some instances, the radiologist may be the first to discover the focal bone lesion, as an incidental finding. For instance, radiographs taken to exclude a foreign body in the soft tissues of the knee could reveal an enchondroma in the distal femur.

Once a focal bone lesion is discovered, it is important to decide if additional imaging is needed. Discussions between the clinician and the radiologist are important in order to determine the best imaging modality or modalities for arriving at the correct diagnosis. In cases where the clinician is not comfortable with the management of the bone lesion, she/he should consider referral of the patient to an orthopedic oncologist. The imaging workup typically begins with plain radiographs, as they are relatively inexpensive, easily to perform, and provide excellent assessment of the cortical features. CT can be useful to show that the lesion arises from the bone, to characterize internal matrix, and to determine the integrity of the cortex. MRI can be helpful in tumor characterization (showing the presence of fat or fluid–fluid levels); however, its main importance is in assessing the degree of tumor extension for staging and for identifying targets for biopsy. Bone scintigraphy and FDG-PET/CT can assess for multifocal disease and determine the osteoblastic and metabolic nature of the lesion, respectively. For some neoplasms such as multiple myeloma, a skeletal survey or whole-body MRI may be employed to assess for multifocal disease.

Management of the Focal Bone Lesion

After the imaging and clinical findings are gathered, the next step in clinical management is to determine whether the lesion requires tissue sampling to guide treatment (Fig. 1.6). There are three potential management recommendations: (1) do nothing; (2) perform follow-up imaging; or (3) perform biopsy/surgery. However, even lesions that are followed by imaging eventually either have to be left alone or biopsied. Practically speaking, only two real options really exist: (1) leave the lesion alone or (2) perform biopsy/surgery. Lesions can be categorized as "clearly benign," "indeterminate," or "clearly malignant." "Clearly benign" lesions such as a bone island or resolving fibroxanthomas are "don't touch" lesions that should be left alone. Lesions that are "clearly malignant" should undergo biopsy. This includes aggressive lesions with a high likelihood for malignancy, such as a large destructive tumor in a woman with invasive breast cancer. Lesions that are "indeterminate" can be further categorized into lesions that are "probably benign" or "suspicious for malignancy." "Probably benign" lesions, such as a large enchondroma with minimal endosteal scalloping, can be followed by imaging. Lesions that are "suspicious for malignancy" should go directly to biopsy; however, there can be biopsy decision factors that make imaging follow-up more practical. Recommendations for the interval and duration of imaging follow-up vary among lesions and between authors and are usually based on the degree of suspicion for malignancy generated by the imaging appearance of the lesion and the patient history. Typically, several years of stability is enough to suggest that the lesion is benign and argues against the need for additional imaging workup. "Indeterminate lesions" that show interval increase in lesion size and development of aggressive features such as cortical breakthrough, periosteal reaction, or soft tissue mass on follow-up imaging should raise the need for biopsy. Alternatively, on occasion, an indeterminate lesion may show changes on follow-up imaging that favor a benign entity, such as sclerosis seen in a resolving NOF.

Practically, the decision to go to biopsy is based not only on imaging features suspicious for malignancy (detailed in subsequent chapters) but also on other biopsy decision factors. Performing a biopsy may be the best management option for one patient, but not the appropriate course of action for another patient, even though the two lesions have identical imaging appearances. Factors such as known malignancy or severe pain at the site of the lesion can support the decision to biopsy. Conversely, factors such as medical comorbidities may make a biopsy risky to perform. In some instances, the lesion may be located adjacent to vital structures (nerves, vessels, lung, vital organs, and joint space), increasing the risk of procedure complications. Moreover, certain tumors and nonneoplastic conditions, such as infection, degenerative changes, ABC, SBC, and lymphoma, can have a relatively low diagnostic yield at core needle biopsy, suggesting that surgical biopsy would be a better option. In practice, patient anxiety can also have a major influence on the decision to biopsy. For some patients, a 10% risk of malignancy would be a compelling indication for biopsy, while, for others, a 10% risk of malignancy would be an indication for declining biopsy in favor of follow-up imaging. These decisions ultimately become a form of shared decision-making among the radiologist who evaluates the images and performs the percutaneous biopsy, the clinician caring for the patient, and the patient. However, it is important to understand that the radiologist's "recommendation" to perform or forego a biopsy should be based on the proper standard of medical care based on all the available information, while the final "decision" to actually perform or forego a biopsy must take into account patient preferences and other factors (Table 1.2).

Biopsy Considerations

Before a biopsy is undertaken, it is important for the radiologist and orthopedic oncologist to discuss the optimal approach to be used for percutaneous needle sampling and the specific biopsy technique. Percutaneous core needle biopsy (PCNB) with imaging guidance is performed by the radiologist and is the initial procedure to consider when tissue is required for pathologic diagnosis. PCNB can be performed on an outpatient basis utilizing CT, ultrasound, or fluoroscopy for image guidance and, when necessary, using conscious sedation for anesthesia. The procedure can be performed in less than an hour. Complications are rare, but can include hematomas, fractures, and injury to adjacent structures, such as pneumothoraces. Surgical biopsy is more invasive and is used when the percutaneous biopsy is nondiagnostic or when definitive resection is required. The benefits of surgical biopsy over PCNB are that the histologic samples are larger and small lesions can be completely resected (sometimes constituting definitive treatment). Fine needle aspiration (FNA) refers to a tissue sampling technique that uses a smaller size needle to sample a lesion than PCNB. FNA samples are sent for cytologic analysis (looking at cells), rather than histologic analysis (looking at tissue architecture). FNA can, at times, be helpful for lesions that are small and for lesions that carry a higher risk of complications if biopsied with a large core needle. However, the diagnostic yield with FNA is lower than with PCNB and most bone sarcomas cannot be adequately assessed with FNA alone.

Because of concerns related to possible seeding of tumor cells along the needle biopsy tract, the surgeon typically resects the needle tract at the time of definitive surgery. This is especially important for malignant primary tumors. Although technically appealing, biopsying along the shortest distance from the skin to the tumor may not always be the optimal trajectory. In general, one should avoid contamination of the neurovascular bundle, joint space, and certain muscles (e.g., partial resection of gluteus muscles and rectus femoris can lead to poor overall function). In addition, discussions with the pathologist prior to the biopsy are important for lesions that might require additional histologic and cytologic tests. In the majority of cases, samples can be submitted in formalin for histologic analysis. However, in cases where lymphoma is suspected, samples should be placed in Roswell Park Memorial Institute (RPMI) cell growth medium in order to perform flow cytometry.

Table 1.2 Non-imaging biopsy decision factors

Factor	Threshold to biopsy
Known malignancy	↓
Severe pain	↓
Comorbidities	↑
Risk of injury to adjacent structures	↑
Low diagnostic yield for certain lesions	↑
Patient anxiety	↑ or ↓

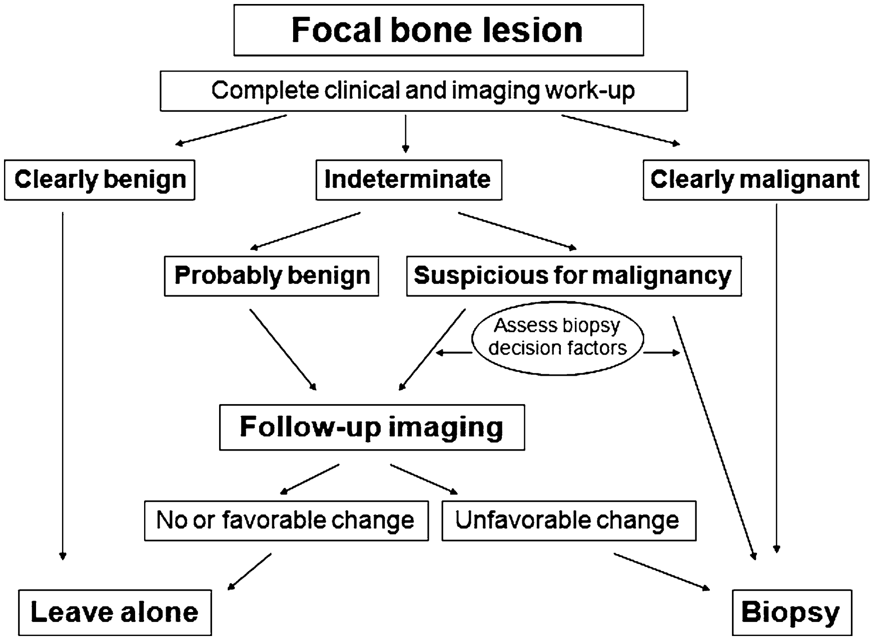


Fig. 1.6 Management of the focal bone lesion

Treatment

The treatment for each bone tumor depends on many factors, including whether the lesion is benign or malignant, the lesion location, the natural history of the disease entity, and the level of pain. Benign tumors that are asymptomatic often do not require treatment; however, benign lesions that are painful, injuring nearby anatomic structures, and/or at high risk for pathologic fracture, such as simple bone cysts, may require treatment. Curettage of the lesion followed by bone grafting or excisional biopsy can provide good results. However, these two treatment methods are inadequate for the treatment of malignant tumors, since residual tumors cells can remain at the margin of the lesion. Other nonsurgical treatment options exist for certain benign lesions. For instance, osteoid osteomas can be treated using radiofrequency ablation and Langerhans cell histiocytosis may resolve following steroid injections. Additionally, benign or malignant lesions that weaken the strength of the bone need to be stabilized with orthopedic hardware to prevent pathologic fracture.

Some malignant tumors, such as primary bone lymphoma, can be treated with chemotherapy or radiation without surgery. Other tumors will undergo treatment with radiation or chemotherapy in order to decrease the size of the tumor prior to definitive surgery, such as with a conventional osteosarcoma. Still other malignant processes can be treated with surgery alone. For instance, chondrosarcomas are typically low grade and do not respond well to chemotherapy, and in many instances, can be treated with surgery alone. Lastly, isolated metastatic lesions can be resected for potential cure. In general, surgery for bone metastases is reserved for preventing pathologic fracture, as the treatment is often limited to radiation and/or chemotherapy. Many malignant processes or aggressive benign processes can be treated with wide surgical resection, where a margin of normal tissue is resected along with the tumor. Others require radical resection, which involves removal of bone, muscle, or other tissues in the compartment along with the tumor.

Suggested Reading

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How to Evaluate a Bone Lesion

2

Using a systematic approach is key for evaluating a focal bone lesion. The ultimate goal should be to provide a definitive diagnosis; however, in many instances, this is not possible. Nonetheless, one should aim to provide a short, logical differential diagnosis and to comment on whether the lesion can be left alone as a "do not touch" lesion or whether additional workup is necessary. The analysis should begin with the patient's age and the location of the lesion, since these two factors play such a significant role in determining the differential diagnosis. Next, assessment of specific lesion characteristics can be used to help narrow the differential, i.e., the lesion's pattern of bone destruction and lesion margins and the presence of any matrix mineralization, periosteal reaction, or soft tissue component. If multiple lesions are present, that fact can help to refine the diagnosis even further. For example, the presence of multiple lytic lesions in an older individual would lead one to suspect metastatic disease or multiple myeloma. One should also search for relevant secondary findings, such as diffuse osteopenia in multiple myeloma or subperiosteal resorption and acro-osteolysis in hyperparathyroidism, and should consider relevant clinical information, such as a history of malignancy supporting a diagnosis of metastasis or fever and erythema supporting a diagnosis of osteomyelitis.

Factors to Consider

- Patient age
- Lesion location
- Lesion density
- Pattern of bone destruction and lesion margins
- Matrix and matrix mineralization
- Periosteal reaction
- Soft tissue component
- Single or multiple lesions

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Patient Age

Certain tumors tend to develop during specific age ranges (Tables 2.1 and 2.2). For example, most benign tumors occur before 30 years of age while lesions identified in persons over 40 years old are more likely to be metastases or multiple myeloma. Among benign lesions, non-ossifying fibroma, osteoid osteoma, simple (unicameral) bone cyst, Langerhans cell histiocytosis, and chondroblastoma occur in children or teenagers. However, giant cell tumor of bone almost always occurs in skeletally mature patients with closed physes and thus should not, for example, be mentioned in the differential diagnosis for a lytic lesion in a 10-year old. Among malignant bone lesions, metastases are by far the most common and should be considered high in the differential for patients over the age of 40 (especially if over 60 years of age). Among primary bone malignancies, osteosarcoma is the most common lesion biopsied in children and teenagers, while myeloma is the most frequent primary tumor of bone in adults (Fig. 2.1).

Table 2.1 Tumor and tumor-like lesions of bone: typical ages of patients

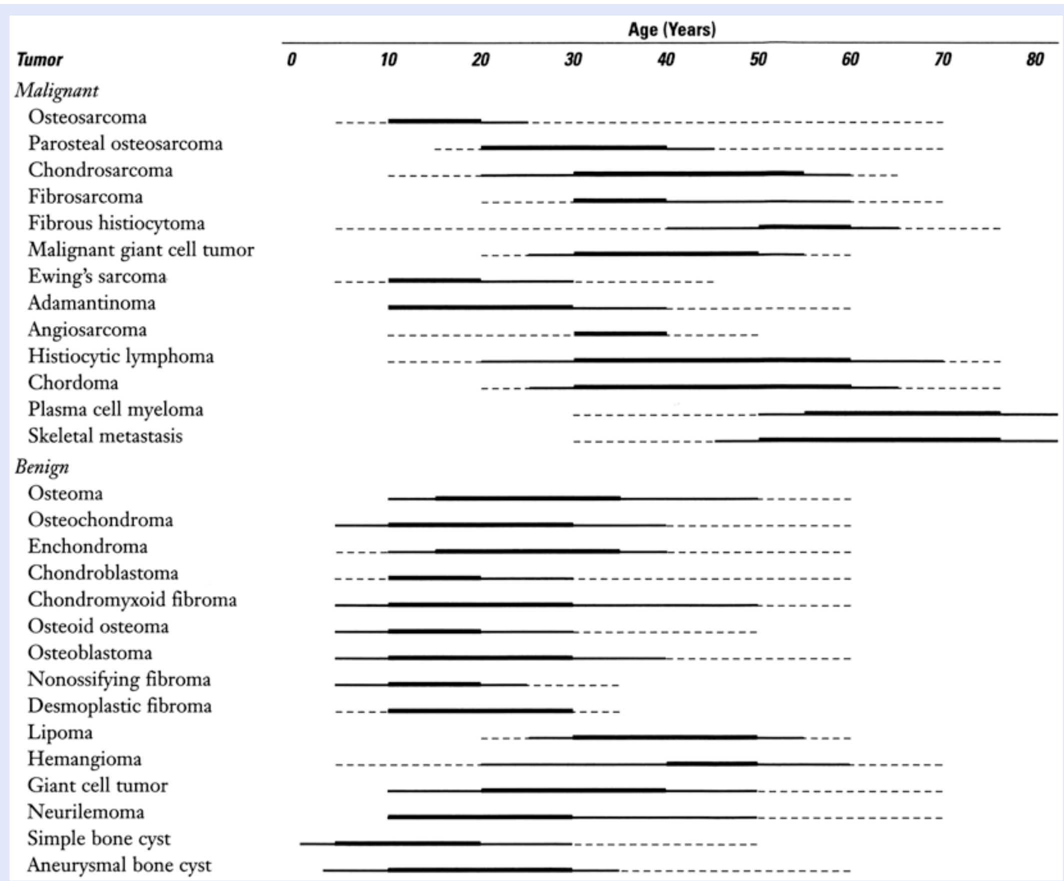

Table 2.2 Most likely tumors by age

Second decade	• Aneurysmal bone cyst (ABC) • Chondroblastoma • Ewing sarcoma • Langerhans cell histiocytosis • Osteosarcoma • Non-ossifying fibroma • Simple (unicameral) bone cyst
Third and fourth decades	• Giant cell tumor • Lymphoma • Parosteal osteosarcoma
Fifth to seventh decades	• Chondrosarcoma • Chordoma • Fibrosarcoma • Lymphoma • Metastases • Multiple myeloma

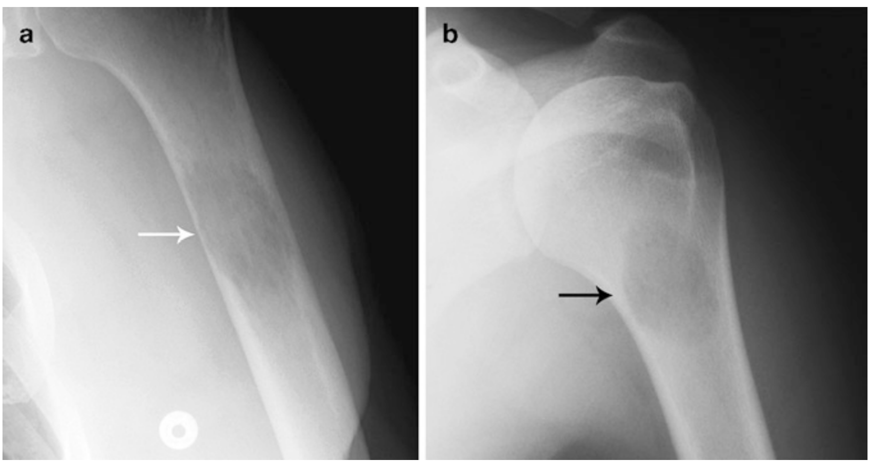

Fig. 2.1 Patient age plays a key role in shaping the differential diagnosis. Two different geographic, ovoid, lytic lesions in the humerus appear similar, but occur in patients of different ages and have very different diagnoses. (**a**) Sixty-eight-year-old man with a renal cell metastasis (*arrow*); (**b**) 17-year-old boy with Langerhans cell histiocytosis (LCH) (*arrow*)

Lesion Location

Some bone tumors and non-neoplastic bone lesions occur characteristically within specific bones or in specific locations within a bone. Bone tumors tend to develop near the ends of long bones, which are areas of rapid bone growth and remodeling. For example, osteosarcomas are commonly found about the knee in the distal femur or proximal tibia and in the proximal humerus, where rapid bone growth occurs. Non-ossifying fibromas tend to occur in these locations as well, probably for similar reasons. Enchondromas and osteochondromas, which are thought to arise from displaced cartilage that originates from the physis, tend to be associated with the metaphysis or metadiaphysis. The existence of a distinct relationship between a particular bone tumor and the anatomic site in which it typically arises has been described as the "field theory" of bone tumors: tumors of a particular cell type arise where the corresponding normal cells are most active, such that the composition of a tumor is related to the metabolic field in which it arises (Fig. 2.2). Additional factors contribute to the reproducibility of bone lesion locations. For example, seeding of either metastatic tumor or infection may occur in the metaphysis because of looping vessels and sinusoids which slow blood flow there. Round cell lesions tend to occur in areas of hematopoietic marrow and thus tend to be seen in the diaphysis or metadiaphysis. Because of differences in distribution of red marrow between children and adults, round cell tumors can occur in both the axial and appendicular skeleton in children, but are generally limited to the axial skeleton in adults. Chordomas, which arise from notochordal remnants, occur along the course of the spine, most often in the clivus and sacrum. Epidermoid inclusion cysts, which are thought to arise from implantation of epidermis into bone, tend to occur in the terminal phalanges and calvaria.

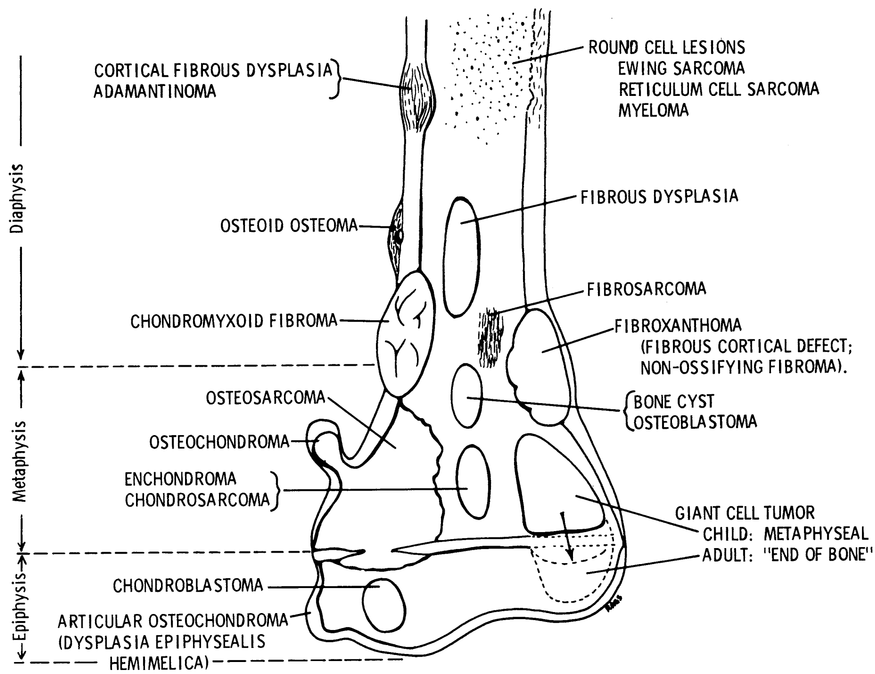

Fig. 2.2 Field theory of bone tumor locations. This diagram summarizes the most common location of various bone tumors within a long bone. For example, chondroblastoma tends to arise in the epiphysis, while osteochondromas and enchondromas tend to arise in the metaphysis and fibrous dysplasia, adamantinoma, osteofibrous dysplasia, and round cell lesions tend to arise in the diaphysis. In general, particular tumors tend to arise in the "field" where the corresponding cell type is normally most active. Adapted and reproduced with permission from Koeller KK, Levy AD, Woodward PJ, et al., editors, Radiologic-Pathology, Vol. 2: Musculoskeletal, Neuroradiologic, and Pediatric Radiologic Pathology Correlations, 3rd ed (2004) American Registry of Pathology, Armed Forces Institute of Pathology, Washington DC