Benign Bone Tumors of Childhood

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Abstract

The diagnosis of a bone tumor in a child can be a source of great anxiety for the patient, the parents, and the treating physician. Fortunately, most bone tumors in children are benign. Although there are a variety of benign bone tumors that affect skeletally immature patients, most have such characteristic clinical and radiographic presentations that the diagnosis can be made with reasonable accuracy without a biopsy. However, some benign bone tumors can simulate a malignant process and may be best handled by referral to a person trained in orthopaedic oncology for additional evaluation. Treatment alternatives are in part related to the Musculoskeletal Tumor Society stage of the lesion. Recurrences of certain lesions, such as aneurysmal bone cysts and osteoblastomas, can be problematic. By becoming familiar with the presentation of the more common benign bone tumors in children, physicians will be able to alleviate fears, establish a diagnosis, and make treatment recommendations in the most effective manner.

The true incidence of benign bone tumors in children is unknown. Much of the data regarding the incidence of benign bone tumors is based on material from series of biopsied or treated lesions. Furthermore, many benign bone tumors in children are diagnosed radiographically and never require further treatment. It is estimated that the incidence of fibrous cortical defects in children is between 30% and 40%. Because of their characteristic radiographic appearance and relatively predictable clinical course, nonossifying fibromas represent only 2% of biopsy-analyzed benign tumors.[1] Codman,[2] who was the first to recognize the rarity of bone tumors, established the registry concept of tumor studies; similar series that followed formed the basis on which tumor incidence has been estimated.[3]

As a group, benign bone tumors in children represent a heterogeneous mix of lesions. However, most benign lesions have a specific and characteristic clinical and radiographic presentation. Fibrous cortical defects and enchondromas are usually asymptomatic and are discovered only as an incidental finding. In contrast, osteoid osteomas and aneurysmal bone cysts are usually associated with symptoms of pain, which prompt the patient to seek medical attention.[4,5] Other tumors may present as a mass (e.g., osteochondroma) or as a pathologic fracture (e.g., unicameral bone cyst).

The natural history of these tumors of childhood and the requirements for treatment vary widely as well. Some lesions, such as fibrous cortical defects, usually require no treatment and resolve spontaneously. Other lesions, such as aneurysmal bone cysts, chondroblastomas, and osteoblas-

tomas, usually require surgical treatment and can be prone to local recurrence. Still others (e.g., Langerhans-cell histiocytosis and osteoid osteoma) have a more unpredictable course and may either resolve spontaneously or require treatment. Despite the apparent differences of the various benign bone tumors affecting children, the diagnosis of any given tumor can frequently be established on the basis of the characteristic clinical and radiographic presentation. It is important that the orthopaedist be able to identify the more common benign bone tumors in children so that unnecessary biopsy can be avoided, fears can be alleviated, and appropriate treatment recommendations can be made.

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Musculoskeletal Tumor Society Classification

Benign bone tumors are staged according to their radiographic appearance and apparent clinical behavior. The Musculoskeletal Tumor Society staging system for benign bone tumors has three stages. Stage 1 lesions are static, latent lesions, which are typically self-healing. Stage 2 lesions are active but remain within the confines of the bone and are associated with bone destruction or remodeling. Stage 3 lesions are active and locally aggressive and tend to extend beyond the cortex into surrounding soft tissue.

Assessing the stage of a benign tumor is useful not only in establishing the diagnosis but also in appropriately planning treatment. Stage 1 lesions usually require no surgical intervention and can be followed periodically to confirm that the lesion is static. Stage 2 lesions may require intervention if they cause structural weakness or are markedly symptomatic. The nature of the intervention depends on several factors, including the specific tumor type, its location, and the patient's age. Stage 3 lesions usually require surgical treatment. In most cases, intralesional procedures are recommended but may need to be augmented with adjuvant modes of therapy. Incomplete or inadequate treatment may make such lesions prone to local recurrence.

Clinical Presentation

The presentation of a child with a benign bone tumor to an orthopaedist is usually precipitated by discovery of a bone lesion as an incidental finding on a radiograph taken after an injury or because of the onset of signs or symptoms, such as pain, a palpable mass, or a pathologic fracture. There is no single characteristic presentation for all benign bone tumors, but there may be a highly characteristic presentation for a given type of tumor, usually a specific constellation of signs, symptoms, and radiographic findings (Table 1). In rare instances, a benign bone tumor may be the initial presentation of a systemic process, as in Albright syndrome and Langerhans-cell histiocytosis.[6] In other instances, the presentation may simulate a malignant process, which can lead to unnecessary anxiety and diagnostic studies.

Pain

Whenever a child complains of musculoskeletal pain, the physician

Table 1
Clinical Characteristics of Benign Bone Tumors

Tumor type	Location			Presentation				Stage		
	Epiphyseal	Metaphyseal	Diaphyseal	Pain	Palpable mass	Pathologic Fracture	Incidental	1	2	3
Benign osseous tumors										
Osteoid osteoma		x	x	x			x	x		
Osteoblastoma		x		x					x	x
Benign cartilage tumors										
Osteochondroma		x			x		x	x		
Chondroblastoma	x			x					x	x
Enchondroma		x	x				x	x	x	
Chondromyxoid fibroma		x		x				x		
Fibrous lesions										
Fibrous dysplasia		x	x			x	x	x	x	
Nonossifying fibroma		x				x	x	x	x	
Fibrous cortical defect		x					x	x		
Cystic lesions										
Unicameral bone cyst		x	x	x		x		x	x	
Aneurysmal bone cyst		x		x		x			x	x
Histiocytic tumorlike lesions										
Langerhans-cell histiocytosis		x	x	x	x			x		

should enquire into the nature and location of the pain, the duration of symptoms, and any aggravating or alleviating factors. This may be difficult in young children who are very fearful or are unable to provide an accurate history. The parents may be able to provide additional information, although the onset of crying or wanting to be carried may be the only symptom. The child's symptoms are often initially attributed to "growing pains" or unwitnessed trauma. This is especially true when radiographs are not obtained, are of poor quality, or are misinterpreted. The possibility of a benign bone tumor should always be included in the differential diagnosis of unexplained musculoskeletal pain in a child.

Benign bone tumors may be a source of pain without any underlying fracture, depending on the tumor type, size, and location. Tumors that commonly present with localized pain include aneurysmal bone cyst, Langerhans-cell histiocytosis (previously known as eosinophilic granuloma), osteoblastoma, and osteoid osteoma. Symptoms may be related to a structural weakness in the affected bone or, in the case of osteoid osteoma, to high local concentrations of prostaglandins within the tumor.[3] Even metaphyseal fibrous defects may become painful if they grow large enough to impair bone structure. Other typically asymptomatic tumors, such as osteochondromas, may be symptomatic due to secondary causes, such as fracture, repeated trauma, and local irritation of surrounding structures (tendon, muscle, artery, or nerve) (Fig. 1). When pain associated with an osteochondroma is the result of mechanical irritation, symptoms are localized to the site of the tumor and are typically aggravated by specific activities.

Benign bone tumors are capable of producing referred pain when they irritate an adjacent nerve, which can make the diagnosis even more elusive. An osteochondroma involving the proximal fibula may compress the common peroneal nerve and present as foot pain. Similarly, osteoblastoma of the spine may present as leg pain, mimicking a disk lesion, or as painful scoliosis; in either case, the condition may remain undiagnosed until appropriate radiographic examination of the spine reveals the tumor.

The size and location of the tumor may also be a factor in whether or not that tumor becomes symptomatic. Fibrous dysplasia, nonossifying fibroma, and fibrous cortical defect are usually asymptomatic unless they are large enough to weaken the bone and create microfractures that cause symptoms.

The character of pain can help in establishing the diagnosis of a benign bone tumor. The history of a dull aching pain for weeks to months, which is worse at night and is relieved by aspirin or nonsteroidal anti-inflammatory drugs, is so common with osteoid osteoma as to be nearly diagnostic.[7] When the physician elicits such a history, the suspicion of an osteoid osteoma must be strongly considered even if initial radiographs of the site fail to reveal an abnormality. A child with an osteoid osteoma involving the hip may complain of a dull aching pain in the knee, which is relieved by aspirin or nonsteroidal anti-inflammatory medication. The combination of a high index of suspicion for a small tumor and a knowledge of patterns of referred pain should lead the physician to obtain a bone scan and/or radiographs of the hip when radiographs of the knee fail to reveal a cause for the patient's symptoms.

Palpable Mass

The most common benign bone tumor that presents as a palpable mass in a growing child is osteo-chondroma. The mass associated with an osteochondroma is firm and immobile and may be tender. The physical examination may help determine whether the mass is of osseous origin. In the case of a pedunculated osteochondroma, it may be relatively easy to appreciate that it arises from bone. When the osteochondroma has a broad base and is sessile, it may be more difficult to discern that the mass is arising from the underlying bone. This type of lesion may appear to be expanding the bone, as in the case of a unicameral bone cyst, fibrous dysplasia, or aneurysmal bone cyst.

The parents and child are often unable to provide a history of growth of the lesion if it has only recently been noticed; however, they may be able to give a history of recent skeletal growth or a familial

Fig. 1 Radiograph of a previously asymptomatic 13-year-old boy who was kicked in the distal aspect of the thigh during a soccer game. A bony pedunculated surface lesion can be seen arising from the metaphysis of the lateral distal femur and pointing away from the epiphysis, which is contiguous with the adjacent cortex. A fracture of the osteochondroma accounted for the patient's symptoms.

inheritance pattern. The physician should examine the patient carefully, looking for osseous masses or angular deformities of other extremities, in order to assess whether the patient has multiple osteochondromas. Solitary exostoses outnumber multiple hereditary exostoses by at least 10:1. An autosomal dominant mode of transmission is evident in 70% of patients with multiple osteochondromas.[8-10] After the history and physical examination, plain radiographs of the affected area should be obtained.

Pathologic Fracture

Some benign bone tumors go unrecognized until they structurally compromise the bone to the point that it breaks. Tumors that may present with a pathologic fracture usually grow slowly and weaken bone (stage 2 lesions). The most common include unicameral bone cyst, fibrous dysplasia, and nonossifying fibroma.[3,11,12] Approximately 50% of unicameral bone cysts are first diagnosed after a pathologic fracture (Fig. 2). Less likely to initially present with a pathologic fracture are the more aggressive benign tumors, such as aneurysmal bone cysts, which tend to produce symptoms of pain before fracture. Therefore, when a child presents with a pathologic fracture, a careful history of prior symptoms should be elicited. Benign latent lesions, such as fibrous cortical defect, may produce symptoms when the bone is weakened to the point of creating microfractures. In such cases, the fracture may not be visible on plain radiographs.

Incidental Finding

Many benign bone tumors in children are discovered as an incidental finding on radiographs or bone scans obtained for unrelated reasons. In such instances, radiographs may have been obtained in the emergency department or the pediatrician's office, and the parents may have been informed that their child has a bone tumor. Physicians who are not familiar with the clinical and radiographic presentation of benign bone tumors are unable to reassure the patient and family that the lesion is benign, which leads to unnecessary anxiety, imaging studies, and even biopsy. It is incumbent on the orthopaedic surgeon to be able to recognize the benign nature of the lesion and to provide reassurance that the tumor is not life-threatening.

The benign bone tumors that tend to be discovered as an incidental finding are usually stage 1 or stage 2 lesions. Stage 3 lesions often present with pain before being discovered radiologically. A benign tumor may also be detected incidentally on a bone scan performed for unrelated reasons. Benign bone tumors that are generally asymptomatic but demonstrate increased activity on bone scan include fibrous dysplasia, fibrous cortical defect, and enchondroma. In such cases, plain radiographs of the affected area and the knowledge that benign lesions (especially stage 2 lesions) may be active on bone scan are all that is needed for the physician to provide reassurance that the lesion is most probably benign. Other benign bone tumors that are active on bone scan include aneurysmal bone cyst and osteoid osteoma. However, as these two types of tumors are frequently associated with pain, most patients seek medical attention before the lesion is detected radiologically. Because benign tumors are often discovered as an incidental finding, it follows that many others go undiagnosed.

Radiologic Findings

The most helpful imaging study for the evaluation of a bone tumor is plain radiography. At least two or-

Fig. 2 Typical radiographic appearance of a unicameral bone cyst with an associated pathologic fracture and "fallen leaf sign" (arrow).

thogonal views centered over the lesion should be obtained. In most cases, the plain radiographs, combined with the clinical history, are all that is required to establish the correct diagnosis.

In some instances, computed tomography (CT) and, less commonly, magnetic resonance (MR) imaging may be helpful in evaluating specific lesions. Technetium bone scanning is used to assess other sites of possible bone involvement in conditions that may be polyostotic, such as fibrous dysplasia, multiple enchondroma, and Langerhans-cell histiocytosis. In the latter case, the bone scan is unreliable because of poor radioisotope uptake by the lesions; a skele-

tal survey should be performed to evaluate the possibility of other osseous sites.[13,14]

Plain Radiography

Plain radiography is the most helpful imaging study in establishing a diagnosis when a bone tumor is suspected. In most cases, the specific diagnosis can be made without additional imaging studies. The critical factors that can usually be gleaned from review of the radiographs and that are useful in narrowing the differential diagnosis are tumor location (flat vs tubular bone), the segment of bone involved (epiphysis, metaphysis, or diaphysis), the growth characteristics of the lesion (as judged by the tumor margins and the presence or absence of periosteal reaction), and the presence or absence of calcified tumor matrix.[15]

Unicameral (simple) bone cyst, enchondroma, osteoblastoma, and nonossifying fibroma have a predilection for specific bones. Unicameral bone cysts are most likely to occur in the proximal humerus and proximal femur; these sites account for approximately 80% to 90% of cases.[16,17] Osteoblastomas have a predilection for the posterior elements of the spine. Nonossifying fibromas are most commonly found in the distal femur and proximal tibia.

Most benign bone tumors in children affect the metaphyses of long bones. This is true of osteoid osteoma, osteochondroma, enchondroma, chrondromyxoid fibroma, fibrous dysplasia, nonossifying fibroma, fibrous cortical defect, unicameral bone cyst, and aneurysmal bone cyst. Langerhans-cell histiocytosis has a predilection for involving flat bones, with nearly 70% of such lesions occurring in the skull, jaw, spine, pelvis, and ribs[1]; it rarely affects the tubular bones of the hands and feet. In contrast, osteochondroma and chondroblastoma involve flat bones in only about 10% of cases. Fibrous dysplasia may affect any long bone as well as the ribs and skull. Involvement of the spine, pelvis, scapula, hands, or feet is rare.

The segment of bone involved is also a factor. Most bone tumors are located within the metaphysis or diaphysis. One significant exception is chondroblastoma; more than 95% of all chondroblastomas are epiphyseal.[18] Thus, when a lesion is located within the epiphysis of a long bone, particularly the proximal humerus, the diagnosis of chondroblastoma should be strongly considered (Fig. 3), along with giant cell tumor, atypical osteomyelitis, and enchondroma. Lesions that tend to arise within the metaphysis include osteochondroma, enchondroma, osteoblastoma, osteoid osteoma, nonossifying fibroma, unicameral bone cyst, and aneurysmal bone cyst. Few benign tumors typically occur within the diaphysis; osteoid osteoma and Langerhans-cell histiocytosis are much more common at that site.

There is considerable overlap for some tumors with respect to the segment of bone they involve. Fibrous dysplasia may involve the metaphysis and/or the diaphysis of a given bone. Similarly, unicameral bone cysts are thought to begin in the metaphysis but migrate away from the epiphysis as the bone undergoes longitudinal growth, so that they may be located within the diaphysis when ultimately discovered.

The location of the tumor should be defined as being cortical or intramedullary. Intramedullary lesions should be further characterized as being central or eccentric. Cortically based tumors include osteochondroma, fibrous cortical defect, and osteoid osteoma. Tumors that tend to be centrally located within the medullary portion of bone include enchondroma, fibrous dysplasia, and unicameral bone cyst. Eccentric medullary lesions include nonossifying fibroma, chondromyxoid fibroma, and aneurysmal bone cyst.

Primary benign tumors of bone may involve the spine, although uncommonly. Children with vertebral involvement may present with torticollis or scoliosis with or without back pain. The benign osteoblastoma in particular has a distinct predilection for the spine, with 40% of all osteoblastomas and 10% of all osteoid osteomas occurring in the
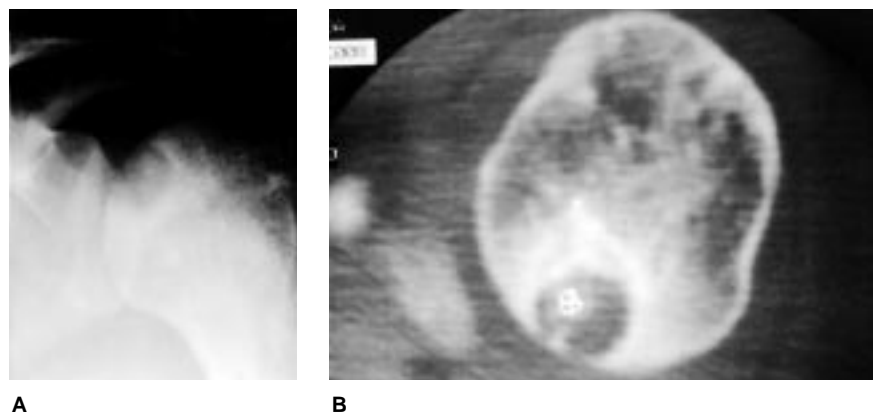

Fig. 3   A, Plain radiograph of the proximal humerus demonstrates a well-circumscribed osteolytic lesion in the epiphysis. Open biopsy confirmed the diagnosis of chondroblastoma. **B,** CT scan through the involved area shows internal calcification within the lesion.

spine and sacrum.[19] Other lesions that may involve the spine include Langerhans-cell histiocytosis, osteochondroma, and aneurysmal bone cyst.

Plain radiographs can provide clues about the stage, and therefore the biologic behavior, of a lesion. Three radiographic patterns have been described by Madewell et al[15]: geographic, moth-eaten, and permeative. These patterns of destruction represent increasing growth rates, from slow for the geographic pattern to rapid for the permeative pattern. The most common pattern seen in benign lesions is geographic, with a typically slow growth rate. In contrast, most primary malignant bone tumors in children have a permeative pattern radiographically. There is, however, some overlap; not all benign tumors are geographic in appearance, nor are all malignant tumors permeative. Langerhans-cell histiocytosis may simulate an aggressive malignant lesion radiographically (Fig. 4).

Geographic lesions may be characterized by sclerosis at the margins. Sclerotic margins are associated with indolent lesions, such as fibrous cortical defect (Fig. 5), fibrous dysplasia, chondromyxoid fibroma (Fig. 6), and, occasionally, chondroblastoma. The absence of a sclerotic border is indicative of an increasing growth rate. Tumors that may present with that pattern include fibrous dysplasia, aneurysmal bone cyst, chondroblastoma (Fig. 7), and, occasionally, chon-
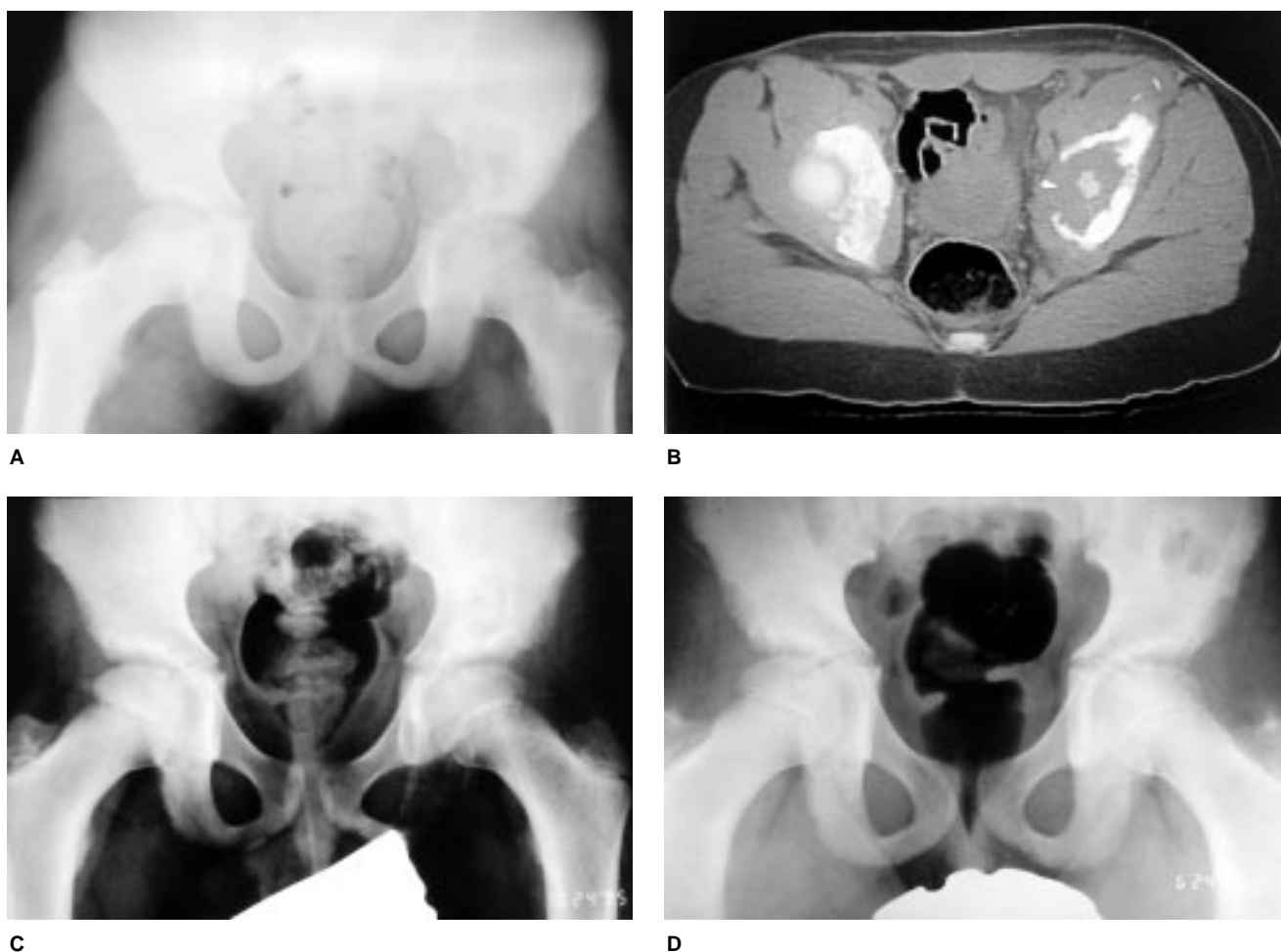
A

B

C

D

Fig. 4 Images of an 8-year-old child who complained of left hip pain. **A,** Plain radiograph of the pelvis demonstrates an ill-defined osteolytic lesion in the supra-acetabular portion of the left pelvis. **B,** CT scan demonstrates the aggressive osteolytic nature of the lesion. **C,** Image obtained 6 months after open-biopsy confirmation of the diagnosis of Langerhans-cell histiocytosis and intralesional injection of corticosteroids shows partial resolution of the lesion in the supra-acetabular area; however, there is evidence of new involvement in the ischium and inferior pubic ramus. **D,** Radiograph obtained 2 years and 2 months after the initial diagnosis shows no evidence of tumor. The patient remained asymptomatic.
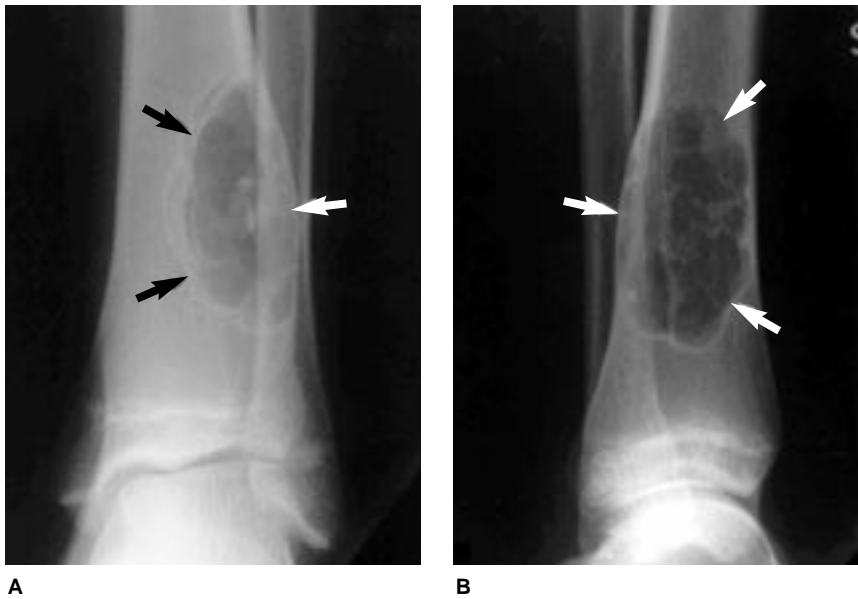
Fig. 5 Radiographic appearance of a fibrous cortical defect (arrows) involving the left distal tibia in a 10-year-old child. **A,** Anteroposterior radiograph of the distal tibia demonstrates an ovoid radiolucent lesion with expansion and thinning of the adjacent cortex but with sclerotic borders. **B,** Lateral radiograph shows that the margins are scalloped but well defined.

dromyxoid fibroma. However, there is considerable overlap in the radiographic appearance and biologic activity of some tumors. Chondroblastoma and fibrous dysplasia, for example, may be either indolent or active lesions. In addition to providing clues to the diagnosis, the pattern of bone offers information about whether intervention is likely to be necessary. In the case of fibrous dysplasia, a sclerotic border suggests that the lesion is indolent and is less likely to progress than a similar lesion without a sclerotic border.

Finally, the matrix calcification on plain radiographs may provide a hint as to the tissue type of the tumor. This is particularly true of cartilage-producing tumors, such as chondroblastoma, enchondroma (Fig. 8), and chondromyxoid fibroma. Stippled, or punctate, calcifications within the lesion should alert the physician to the probability of a tumor of cartilaginous origin. Although CT and MR imaging are more sensitive for identifying cartilage within a bone tumor than plain radiography, they are not usually required for establishing the diagnosis.[20]

Bone Scintigraphy

The role of technetium bone scanning in the evaluation of children with benign tumors is either to help define the precise location of a small pain-producing lesion in an area of complex anatomy (e.g., osteoid osteoma) or to assess the child for other sites of disease in conditions that may involve multiple sites, such as fibrous dysplasia and multiple enchondroma. Localization of small lesions within the spine, pelvis, or ribs is often accomplished with technetium bone scanning.

Patients with Langerhans-cell histiocytosis should be evaluated for multiple osseous sites, but bone scintigraphy is not reliable, as some lesions may show increased activity while others do not. Therefore, a skeletal survey is recommended to assess other sites for disease. Because most malignant bone tumors are active on bone scan, the fact that Langerhans-cell histiocytosis may not be active can be helpful when plain radiographs are insufficient to distinguish Langerhans-cell histiocytosis from a malignant tumor. If the lesion does not show increased
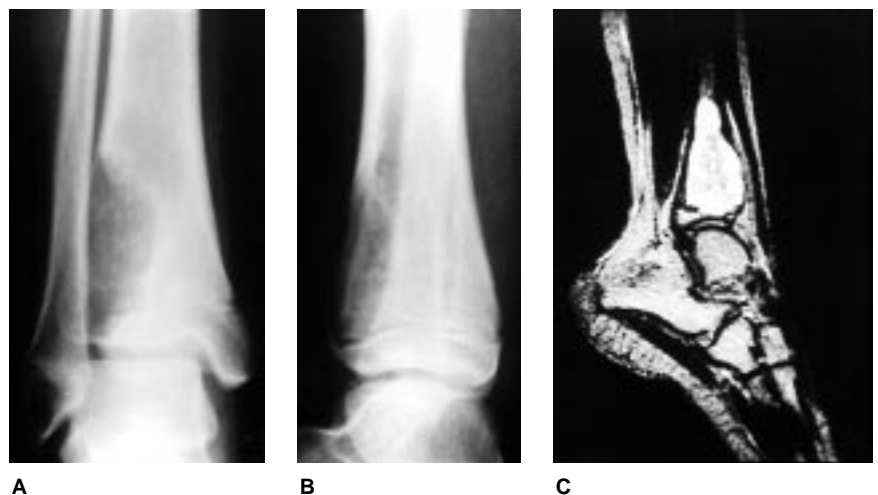

Fig. 6 Radiologic appearance of chondromyxoid fibroma in a child. Anteroposterior **(A)** and lateral **(B)** radiographs demonstrate an ovoid eccentric metaphyseal lesion with a geographic margin. **C,** T2-weighted MR image demonstrates hyperintense lobular cartilage matrix within the lesion.

Fig. 7 Plain radiograph of the proximal tibia of a child with knee pain demonstrates a well-defined radiolucent lesion (arrows) without sclerotic borders arising from the epiphysis. Open biopsy confirmed the diagnosis of chondroblastoma.

activity on bone scan, the diagnosis of Langerhans-cell histiocytosis should be strongly considered. Biopsy or aspiration is frequently necessary to establish the diagnosis of Langerhans-cell histiocytosis.[21]

Although bone scans are not recommended for the evaluation of unicameral bone cysts, they may demonstrate a central area of photopenia. This may be helpful in the uncommon situation in which one must differentiate a unicameral bone cyst from fibrous dysplasia (which tends to demonstrate increased activity on bone scintigraphy).

Computed Tomography

Computed tomography is best used as an adjunct to plain radiography for the purposes of staging and preoperative planning rather than as a diagnostic tool. It should be used in preference to MR imaging because definition of bone architecture is of prime importance. Computed tomography is especially useful for assessing the extent of cortical destruction due to active or aggressive tumors, such as aneurysmal bone cyst, fibrous dysplasia, enchondroma, and fibrous cortical defect. Although no imaging study can predict the risk of pathologic fracture, CT can more accurately assess cortical integrity than plain radiography. In cases in which there is concern about impending fracture or there is already a microfracture, cortical integrity is best assessed with CT. Computed tomography may be indicated when a tumor involving the ribs, spine, or pelvis cannot be adequately imaged with plain radiography because of anatomic considerations.

Computed tomography is especially helpful in localizing the nidus of an osteoid osteoma. It is particularly useful for preoperative planning when the nidus is in a subperiosteal location.[22] Because the central nidus is typically 2 to 4 mm in diameter, thin-section CT may be required to visualize the lesion (Fig. 9). On rare occasions, an osteochondroma may appear to arise directly from the bone cortex, rather than involving blending between medullary host bone and the tumor. When this is the case, the lesion must be distinguished from a parosteal osteosarcoma. A CT scan of an osteochondroma will demonstrate continuity between the medullary host bone and the tumor. With a parosteal osteosarcoma, there is no such continuity.

Computed tomography may also be helpful in distinguishing a unicameral bone cyst from an aneurysmal bone cyst.[23] When multiple fluid-fluid levels are seen, the diagnosis of aneurysmal bone cyst should be suspected. However, not all lesions with multiple fluid-fluid levels are aneurysmal bone cysts. For example, CT may demonstrate multiple fluid-fluid levels in cases of osteolytic osteosarcoma. Fluid-fluid levels may be seen more accurately on MR imaging.

Magnetic Resonance Imaging

Generally, MR imaging is not indicated for the diagnosis or evaluation of benign bone tumors in children. However, patients frequently present to the orthopaedist after an MR study has already been obtained. Therefore, a brief discussion of the MR imaging appearance of some benign bone lesions may be useful.

The cartilaginous cap of an osteochondroma has signal characteristics similar to those of articular cartilage (i.e., increased signal intensity on T1- and T2-weighted sequences). In addition, MR imaging may be useful for evaluating the size of the cartilaginous cap when considering the risk of secondary chondrosarcoma. Chondroid-containing lesions, such as chondroblastoma, enchondroma, and chondromyxoid fibroma, are hyperintense on T2-weighted images
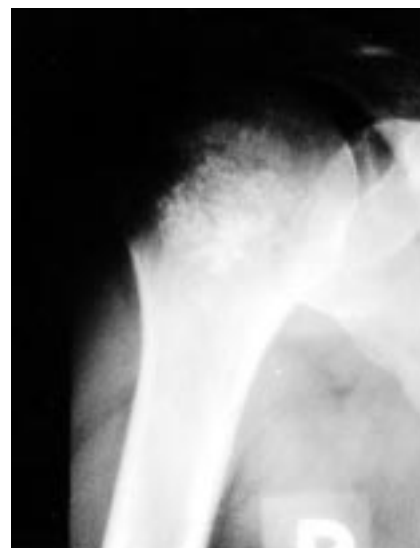

Fig. 8 Radiographic appearance of a benign enchondroma with stippled calcifications in a 16-year-old boy. The tumor was discovered as an incidental finding after trauma.
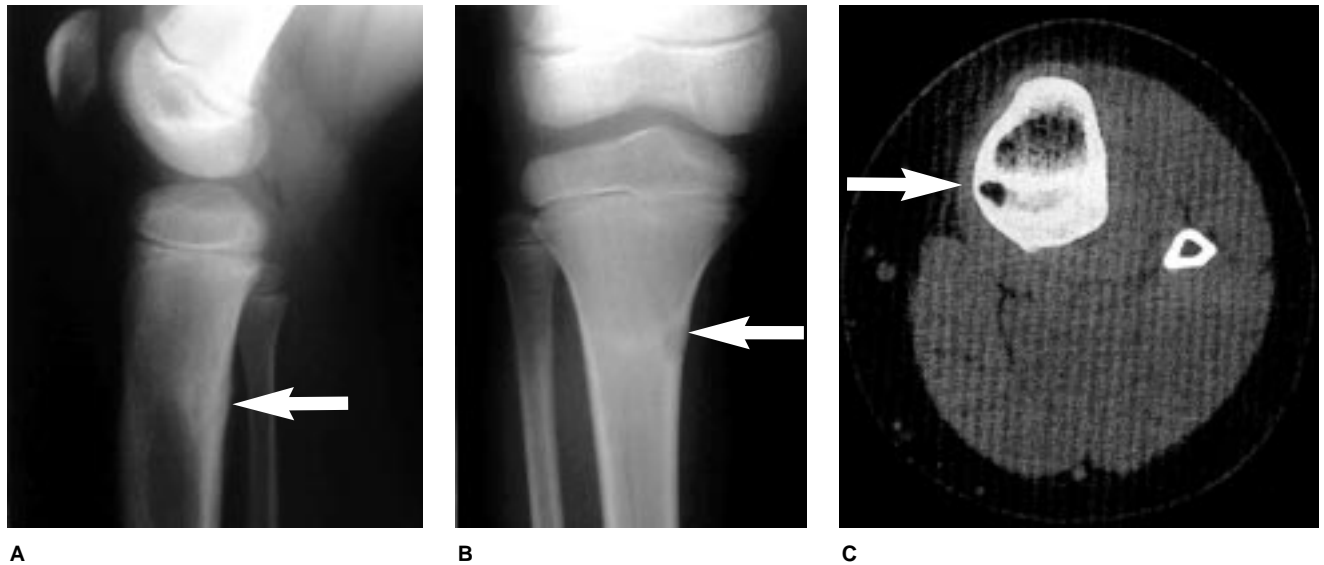
A **B** **C**

Fig. 9 Images of the tibia of a child who complained of a dull aching pain in the leg unrelated to activity, which was worse at night and was relieved with nonsteroidal anti-inflammatory agents. Lateral **(A)** and anteroposterior **(B)** radiographs demonstrate an area of cortical thickening along the posteromedial tibia as well as a subtle radiolucency (arrows). **C,** CT scan demonstrates the subcortical location of the nidus (arrow) of the osteoid osteoma.

and may be lobular in appearance. Areas of dense calcification seen on plain films may appear as focal areas of low signal intensity on T2-weighted images. Chondroblastomas are known to incite prominent peritumoral edema. When examined with MR imaging, a large area of edema (very bright on T2-weighted sequences) surrounding the tumor may lead to the erroneous conclusion that the underlying disorder is infectious, traumatic, or malignant.

The MR appearance of fibrous dysplasia is markedly variable. In some cases, fibrous dysplasia has predominantly dark signal intensity on both T1-weighted and T2-weighted images. In other patients, however, the signal intensity on the T2-weighted images may be increased, with a speckled pattern. The MR appearance does not add to the workup; the lesion is best imaged by plain radiography.

Hemorrhagic fluid-fluid levels on MR images of aneurysmal bone cysts are typically visualized as low signal on T1-weighted images and hyperintense signal on T2-weighted images. However, this finding is not pathognomonic for aneurysmal bone cysts; fluid-fluid levels may be seen on MR imaging in other conditions.

Treatment

Once the diagnosis of a benign bone tumor in a child has been established, the clinician should consider a number of factors before deciding on treatment. These include the natural history of the tumor, the risk of pathologic fracture, and the risks and benefits associated with operative and nonoperative treatment. The Musculoskeletal Tumor Society staging system for benign tumors is useful for determining which tumors are most likely to require treatment and which can be safely observed. Stage 1 tumors are usually self-limiting or stable and in most cases require no surgical treatment. This is certainly true of some fibrous

cortical defects, enchondromas, osteochondromas, unicameral bone cysts, and small nonossifying fibromas. However, large unicameral bone cysts and nonossifying fibromas may pose a risk for pathologic fracture.

There is no reliable method for predicting which large fibrous cortical defects will go on to pathologic fracture. Some authors have used lesion size (e.g., whether the lesion is more than 5 cm in diameter or occupies more than 50% of the transverse diameter of the bone) or persistent pain with or without repeated pathologic fracture as relative indications for curettage and bone grafting. Lesions located in close proximity to an active physis may be best managed nonoperatively until the lesion is no longer adjacent to the physis in order to minimize the risk of surgical injury to the growth plate.[24] Local recurrence is exceptionally rare, and the use of adjuvants is not necessary. Pathologic fractures will heal with operative or nonoperative treatment.

Osteoid osteomas are also stage 2 lesions. It has been shown that the natural history of this tumor is one of spontaneous resolution over the course of several years. Symptoms can sometimes be managed medically with salicylates or nonsteroidal anti-inflammatory drugs.[25] Patients who cannot tolerate or do not want medical management may elect tumor excision. En bloc excision of the tumor reduces the risk of local recurrence compared with less aggressive methods, but it poses a risk of subsequent fracture. In weight-bearing bones, en bloc excision may need to be augmented with bone grafting and/or internal fixation. To minimize the amount of resected bone and the risk of subsequent fracture, Ward et al[26] advocate the use of the burr-down technique for excision of osteoid osteomas. This technique involves using a high-speed burr to remove cortical bone until the nidus has been identified. The nidus is then excised with use of a curette, and the tissue is sent for histologic confirmation and culture.

Other minimally invasive techniques described for excision of osteoid osteoma include CT localization followed by percutaneous drilling and radiofrequency ablation.[27] The advantages of radiofrequency ablation include the fact that it can be performed in an outpatient setting, is associated with fewer complications than open procedures, and is nearly equivalent to operative excision with respect to local tumor control.[28]

The natural history of osteochondromas is growth of the cartilaginous cap by enchondral ossification during periods of skeletal growth, which ceases at skeletal maturity. Indications for surgical excision in the growing child include neurovascular compromise, pain, and interference with function. In the absence of specific noncosmetic indications, removal of an osteochondroma should be avoided in a growing child. When surgical excision is indicated, the surgeon should take care to remove the entire tumor along with its base and perichondrium and the surrounding periosteum in order to minimize the risk of local recurrence. Patients and their parents should be informed about the signs and symptoms associated with malignant degeneration, such as pain or growth of the tumor after skeletal maturity.

Tumors such as enchondromas, fibrous dysplasia, and unicameral bone cysts, may present as stage 1 or stage 2 lesions. Most enchondromas in children are stage 1 and can be managed nonoperatively. In cases of repeated fracture, curettage and bone grafting may be indicated. Similarly, fibrous dysplasia that presents as a stage 1 lesion can also usually be followed nonoperatively. Stage 2 lesions, depending on their location, the patient's age and symptoms, and the fracture risk, may require surgical treatment. In skeletally immature patients, the indications for surgery include repeated fracture and progressive deformity. Unfortunately, simple curettage and bone grafting in children and adults is associated with local recurrence. Surgical treatment is directed toward complete excision of lesional tissue, followed by use of cortical strut grafts with internal fixation when necessary.[29]

The natural history of unicameral bone cysts is a tendency to heal with skeletal maturity, and many can simply be observed without specific treatment. Prior to skeletal maturity, unicameral bone cysts may cause repeated fractures and disability. This is especially true of lesions associated with significant cortical thinning; those in weight-bearing areas, such as the proximal femur; and those in areas subjected to torsional forces, such as the humerus. Initially, most pathologic fractures should be treated nonoperatively and be allowed to heal before surgical treatment is considered. Displaced fractures involving the proximal femur may require open treatment and stabilization.

Prior to 1979, treatment of unicameral bone cysts consisted primarily of open curettage and bone grafting. In that year, Scaglietti et al[30] published their initial favorable experience with percutaneous aspiration and injection of methylprednisolone acetate. More recently, autogenous bone marrow and bone-graft substitutes have been injected percutaneously in lieu of corticosteroids, with improved results. Percutaneous aspiration and injection is associated with less morbidity than open procedures; however, patients and their parents should be advised that more than one injection will likely be necessary, and an open procedure may be indicated if closed procedures fail. Bone-graft substitutes, including demineralized bone powder, bone morphogenetic protein, freeze-dried allograft, and inorganic ceramic composites, in conjunction with open curettage are currently under study.[31]

Langerhans-cell histiocytosis may occasionally resolve spontaneously over time. In practice, many patients with Langerhans-cell histiocytosis undergo biopsy or aspiration to confirm the diagnosis when it is impossible to rule out infection or Ewing sarcoma on the basis of clinical and radiographic criteria alone. Biopsy may be performed percutaneously or as an open procedure. If the diagnosis is confirmed during open biopsy, the lesion can be treated by simple curettage. Capanna et al[32] reported variable results with the use of intralesional corticosteroid injections. Low-dose radiation (500 to 600 cGy) is highly effective for large or inaccessible lesions, such as

those in the spine or pelvis, but may be used for any lesion. Few long-term complications of radiation therapy have been reported.

Osteoblastoma, chondroblastoma, chondromyxoid fibroma, and aneurysmal bone cyst usually present as stage 2 or stage 3 lesions. Chondromyxoid fibroma can be treated with thorough curettage with bone grafting. Gherlinzoni et al[33] have suggested that bone grafting may reduce the incidence of local recurrence by virtue of "the added zeal with which curettage was performed." In children with open epiphyseal plates, surgery should be delayed as long as possible to avoid injury to the physis.

Chondroblastomas present as stage 2 or stage 3 lesions but, in contrast to chondromyxoid fibroma, are prone to more locally aggressive behavior and local recurrence. For this reason, treatment by thorough curettage is recommended and should not be delayed. Lesions that involve the growth plate require complete excision even at the risk of premature closure. The use of physical adjuvants, such as liquid nitrogen, has been advocated, but carries with it the risk of secondary fracture and premature closure of the growth plate.[34] Large defects may necessitate bone grafting.

Stage 2 and stage 3 osteoblastomas differ in their responses to treatment. For lesions contained within the bone (stage 2), curettage and grafting is usually sufficient. With stage 3 lesions, however, the same treatment has resulted in a recurrence rate of approximately 20%. Therefore, lesions in "expendable bones," such as the fibula, may be treated by wide excision. The surgeon should attempt to remove the entire lesion; however, this may not always be possible without significant morbidity, especially in the spine. Some patients will be cured following incomplete excision but must be followed up aggressively for local recurrence. The role of radiation therapy remains controversial. Radiation may be indicated in cases of spinal lesions with an extensive soft-tissue component and epidural spread or following local recurrence.[19]

Aneurysmal bone cysts also present as stage 2 or stage 3 lesions. Their natural history is one of continued growth if left untreated. Simple curettage is associated with unacceptably high rates (>50%) of local recurrence. In an effort to decrease the incidence of local recurrence, extended curettage should be performed with the use of a high-speed mechanical burr. The technique of extended curettage involves creating a large cortical window so as to visualize the entire cavity, followed by removal of all involved bone with a high-speed burr. The use of physical adjuvants such as liquid nitrogen and phenol is generally recommended. The defect should be filled with bone graft or polymethylmethacrylate; in some cases, this may need to be supplemented with internal fixation. Inoperable lesions, particularly those involving the spine, may be treated preoperatively with embolization and postoperatively with radiation.

Summary

While the true incidence of benign bone tumors in children is unknown, it remains a clinical situation that is often encountered by the general orthopaedist. By becoming familiar with the clinical and radiologic characteristics of the most common benign bone tumors in children, physicians will be able to accurately establish the correct diagnosis without the need for biopsy in most cases. With an understanding of the natural history of the tumor and the stage at presentation, appropriate treatment recommendations can be made. This will allow the parents and child to be reassured earlier in the evaluation process and will also prevent both overtreatment and undertreatment of these tumors.

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