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Title Benign Bone Tumors

Permalink https://escholarship.org/uc/item/7h86k14t

Journal Radiologic Clinics of North America, 49(6)

ISSN 0033-8389

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Publication Date 2011-11-01

DOI

10.1016/j.rcl.2011.07.002

Peer reviewed

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Benign Bone Tumors

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KEYWORDS

- Benign bone tumor Radiography Computed tomography
- Magnetic resonance imaging

The imaging characteristics of benign bone tumors reflect their histopathologic appearance. A solid knowledge of this underlying histopathology aids in differential diagnoses of these tumors. Important factors in diagnosis of a bone tumor include patient age and gender; the bone involved; the location of the tumor along, within, or on the bone; lesion margin; matrix proliferation; and periosteal reaction. This article provides a review of the origin of the tumor matrix and its influence on the imaging properties of these tumors.

TUMORS WITH AN OSTEOID MATRIX

These tumors contain a matrix derived from the bone and cartilage progenitor cells of the embryonal mesenchyme. They encompass only 3.6% of biopsied bone tumors.

Enostosis

These osteoblastic lesions, commonly known as "bone islands," are usually asymptomatic and considered incidental findings. There is no gender or age preference, yet bone ensotoses are unusual in children. They are usually less than 2 cm in diameter, although larger variants have been described as giant bone islands. Histologically, ensotoses consist of dense intramedullary lamellar bone with normal haversian canals, which blend in with the normal surrounding trabecular bone. They are most commonly found in the axial skeleton (spine, pelvis, and ribs). In the long bones they are usually located in the epiphysis or metaphysis (Table 1).¹

An enostosis characteristically appears on radiography and computed tomography (CT) as an oval, densely sclerotic intramedullary focus with a spiculated or thorny margin.² This translates into a low signal focus on MR imaging on all pulse sequences with a normal surrounding marrow signal (**Fig. 1**). Enostosis is usually inactive on bone scintigraphy. Further work-up is rarely necessary and biopsy is generally not indicated. A bone island may slightly fluctuate in size because of intrinsic osteoblastic or osteoclastic activity, but a growth of more than 25% over 6 months is unusual and may warrant a biopsy. The differential diagnosis includes sclerotic metastasis, but a metastatic lesion often demonstrates increased activity on bone scintigraphy and displays a halo of surrounding bone marrow edema on MR imaging on fluid-sensitive sequences.³

Multiple bone islands may be associated with osteopoikilosis (osteopathia disseminata); striped osteopathy (Voorhoeve disease); or melorheostosis. Osteopoikilosis consists of numerous circular or ovoid bone islands, usually on both sides of joints, whereas striped osteopathy is characterized by bandlike sclerotic foci in the bones. Melorheostosis is characterized by long segments of thickened cortical bone.²

Osteoma

This benign slow-growing osteoid tumor is usually an incidental finding. At times it becomes symptomatic because of its location. There is no age or gender predilection for this tumor. Histologically, an osteoma is composed of both woven and dense bone and arises strictly from the cortex of the bone. It is most commonly found in the skull and occasionally in the long bones (see **Table 1**). In the sinuses it can cause headache, sinusitis, and other symptoms related to obstruction of the paranasal airspace.

On all imaging modalities an osteoma is a sharply defined bony surface lesion arising from the cortex.

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The authors have nothing to disclose.

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Radiol Clin N Am 49 (2011) 1115–1134 doi:10.1016/j.rcl.2011.07.002 0033-8389/11/\$ – see front matter © 2011 Elsevier Inc. All rights reserved.

Table 1 Location	of benign bone tumors				
Matrix	Bone Tumor	Location in Skeleton		Location Within the Bone	
		Most common	Other locations	Long axis	Short axis
Osteoid		Most common			
oscolu	Fnostosis	Axial skeleton	Anywhere	Epiphysis and metaphysis	Medullary
	Osteoma	Sinuses, skull	Rarely in long bones	Anywhere	Cortical
	Ostoid osteoma	Femur and tibia	Other long bones > spine > hand and foot	Diaphysis	Cortex or adjacent
	Osteoblastoma	Spine, femur, and tibia	Skull > hands and feet > pelvis	Diaphysis or metaphysis	Central
Chondro	id				
	Osteochondroma	Long bones	llium and scapula	Metaphysis	Surface
	Enchondroma	Long bones	Hand and feet	Metaphysis	Central
	Juxtacortical chondroma	Long bones	Humerus > femur > tibia > hands and feet	Metaphysis	Surface
	Chondroblastoma	Long bones	Femur > tibia > proximal humerus, hands and feet	Epiphysis	Eccentric
	Chondromyxoid fibroma	Tibia	Other long bones	Metaphysis	Eccentric in long bones, central in short bones
Fibrous					
	Fibroxanthoma	Long bones	Around the knee	Metaphysis	Eccentric
	Fibrous dysplasia	Long bones, skull	Femur > tibia > skull and facial bones > ribs	Diaphysis	Central
	Osteofibrous dysplasia	Tibia and fibula	Tibia > fibula	Diaphysis	Cortex
	Desmoplastic fibroma	Long bones	Femur > tibia > humerus > radius	Metadiaphysis	Central
Fat					
	Intraosseus lipoma	Calcaneus	Calcaneus > femur > tibia	Metaphysis	Central
Vascular					
	Osseous hemangioma	Vertebrae	Calvarium > calncaneus > long bones	Metaphysis	Central
Unknow	n origin				
	Giant cell tumor	Long bones	Femur > tibia > radius > humerus	Epiphysis and metaphysis	Eccentric
	Simple bone cyst	Long bones	Fumerus > femur > calcaneus	Metaphysis	Central
	Aneurysmal bone cyst	Long bones > spine	About knee > spine	80% metaphsys, 20% diaphysis	Eccentric



Fig. 1. A 58-year-old man with a bone island in the distal tibia (*arrow*). (A) Frontal radiograph of the ankle shows the elongated sclerotic focus in distal tibia. (B) Sagittal T1-weighted MR image shows the low signal bone island with surrounding normal marrow.

It demonstrates a density identical to cortex on radiography and CT (Fig. 2), and follows the low signal intensity characteristics of cortex on all MR imaging pulse sequences.² Multiple osteomas are associated with Gardner syndrome and tuberous sclerosis.

Osteoid Osteoma

Osteoid osteoma (OO) is a painful benign bony lesion of unknown cause. Although OO most commonly occurs in the long bones, it is the most common cause of painful scoliosis in a skeletally immature patient; the lesion is usually located along the concavity of the scoliosis. Pain, which can be worse at night, is usually relieved with nonsteroidal anti-inflammatory drugs through inhibition of prostaglandin 2. OO is more common in males (2–3:1) and mostly affects patients between 5 and 25 years of age. The "nidus," representing the actual lesion, is usually less than 1 cm in diameter, although the lesion diameter can vary between 0.1 and 2 cm. Histologically, the nidus is composed of osteoid and woven bone on a background of highly vascularized fibrous connective tissue. This nidus may be surrounded by a variable degree of sclerosis depending on the location within or along the bone. The cortical variant is the most common and is usually located in the shaft of the long bones. Less common locations are cancellous (subcortical), intra-articular, and subperiosteal.² The most common locations in the skeleton and within the bone are listed in **Table 1**.

The radiographic and CT imaging characteristics of OO depend on its location. The nidus appears as an osteolucent focus, which may or may not have



Fig. 2. A 45-year-old man with an osteoma in the left frontal sinus (*arrow*). (*A*) Axial head CT. (*B*) Head CT, coronal reformation. (*Courtesy of* Ali Sepahdari, MD, University of California, Los Angeles, CA.)

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a dense central focus of mineralization. In long bones, surrounding extensive fusiform sclerosis is the hallmark of this lesion. Cancellous and intraarticular lesions may have no or only limited surrounding sclerosis. The rare subperiosteal lesion may have no adjacent sclerosis and may present as only subtle erosion.² On MR imaging the intense bone marrow edema adjacent to the nidus is paramount for detection and localization of the lesion (**Fig. 3**). The nidus itself may not be readily visible on MR imaging depending on its size and the degree of mineralization.⁴ Adjacent soft tissue edema and periosteal reaction are often noted. An intra-articular lesion may be masked by reactive changes in the surrounding tissue; in a child with joint pain, and synovitis, joint effusion, and adjacent marrow edema on MR imaging, OO should be strongly considered. A CT examination, preferably with triplanar reformatted images, demonstrates the nidus.² The differential diagnosis for OO in the long bones includes stress fractures and osteomyelitis (Brodie abscess). Toxic and inflammatory synovitis can be considered in the differential diagnosis for the intra-articular lesions.



Fig. 3. An 8-year-old girl with an osteoid osteoma of mid-tibial shaft. (A) Lateral radiograph with anterior tibial cortical thickening (arrow). (B) Axial CT shows the nidus (black arrow). White arrows mark tracks of a previously attempted biopsy. (C) Axial fat-saturated T2-weighted MR image with marrow edema and cortical thickening (arrow).

Osteoblastoma

Osteoblastoma (OB) is a rare osteoid-producing tumor that on routine histologic analysis is essentially indistinguishable from OO. A tumor size of 2 cm or larger is the main histopathologic criterion to distinguish this lesion from an OO. OB exhibits different clinical behavior: it presents with a dull ache and possible swelling that may not be worse at night and usually does not respond to nonsteroidal antiinflammatory drug intake. Scoliosis is common with spinal OB along with neurologic symptoms in up to 40% of cases in the spine. The patients are usually younger than 30 years of age and the lesion is more common in males (by \approx 2:1). The common locations in the skeleton are the spine (particularly the posterior elements) and long bones. In most cases in long bones an eccentric location in the diaphysis or metaphysis is noted (see Table 1).^{2,5}

OB may have three different presentations on radiographs. The first presentation is similar to OO, only larger; this is termed a "giant osteoid osteoma." Second, it may have an expansile appearance with a mineralized matrix and a narrow zone of transition. This is typical of the spinal OB. Finally, it may appear more aggressive with prominent bone expansion, partial cortical destruction, and soft tissue infiltration. This is more common in long bones. CT examination may better demonstrate the mineralization and the lesion margin. In complex bony structures, such as in the spine or pelvis, CT demonstrates the extent of the lesion and helps differentiate between bone and soft tissue components.^{2,5} MR imaging may show reactive marrow edema of the surrounding bone, usually less than with OO, with a variable degree of increased T2 signal depending on matrix mineralization and cellularity of the lesion (Fig. 4). MR imaging helps to determine the extension of the spinal lesions into the neural foramina and spinal canal.⁶

Approximately 16% of OBs have components of aneurysmal bone cyst (ABC). Aggressive-appearing inflammatory variants have been described.⁷ A malignant transformation into an osteosarcoma is rare.

TUMORS WITH A CHONDROID MATRIX

Cartilage-forming tumors have imaging characteristics that aid with their diagnosis. In particular, the chondroid "ring and arc" mineralization pattern is often a prominent imaging feature. Chondroid lesions encompass approximately 14% of biopsied primary bone tumors.

Osteochondroma

Osteochondroma (OC) is a relatively common surface lesion of the bone composed of lamellar

bone covered by a cartilage cap.8 Clinically, it presents most commonly as painless swelling and cosmetic deformity. Further clinical presentations may include neurovascular impingement, fracture, overlying bursal or pseudoaneurysm development, and malignant transformation. There is a male predominance, and 75% of patients are younger than 20 years of age. Its growth comes to a halt by the time the physis closes, and the cartilage cap usually involutes after skeletal maturity. This lesion is usually classified as a neoplasm, but it is generally believed that it occurs as a result of physeal cartilage displacement onto the bone surface. This displacement may be secondary to trauma or radiation. OC can arise from any bone undergoing enchondral maturation, but it is most common in tubular bones and in particular around the knee. The ilium and scapula are the two most commonly affected flat bones. In long bones it is located at the metaphysis and grows away from the joint (see Table 1).8

On imaging, OC is a surface lesion that demonstrates cortical and medullary continuity. It may be on a stalk (pedunculated) or broad based (sessile). Cross-sectional imaging with CT and MR imaging may demonstrate a cartilage cap (Fig. 5). After skeletal maturity the cartilage cap should not be thicker than 2 cm; if it is, then transformation to chondrosarcoma should be considered. Ultrasound or contrast-enhanced CT and MR imaging aid in distinguishing bursa formation over the cap from the actual cap. Cartilage cap growth after skeletal maturity and pain are worrisome for malignant transformation.^{9,10} Associated entities include hereditary multiple exostoses and Trevor disease (dysplasia epiphysealis hemimelica, or epiphyseal OC).

Enchondroma

Enchondroma (EC) is a common incidental finding in long bones. EC comprises 3% to 5% of biopsied primary bone lesions. EC is encountered equally in men and women and has a peak incidence in the third decade. Histologically it is characterized by rests of hyaline cartilage within the medullary bone intermixed at times with a myxoid matrix. EC may have a variable degree of amorphous matrix mineralization. It is common in bones of the hand and feet and in long bones. EC occurs mostly as a central lesion in the metaphysis (see **Table 1**).¹¹

On radiography EC presents as an expansile lucent lesion with a narrow zone of transition; varying degree of chondroid mineralization (rings and arcs); and often cortical thinning (endosteal scalloping). EC in long bones of the hands and



Fig. 4. A 13-year-old boy with an osteoblastoma of distal tibia. (*A*) Lateral radiograph demonstrates the expansile lesion of the distal tibia (*arrow*). (*B*) Axial CT shows mineralization within the lesion (*arrow*). (*C*) Axial fat-saturated T2-weighted MR image shows the lesion (*arrow*) and surrounding bone marrow edema (*asterisk*).

feet often lacks any degree of matrix mineralization. In other locations CT readily detects chondroid mineralization, even in those cases with only subtle or no radiographic evidence of matrix mineralization. MR imaging properties reflect the high water content of the hyaline cartilage (ie, high signal on fluid-sensitive sequences with scattered low signal foci of chondroid mineralization). A lobular margin can usually be identified in larger central ECs on all imaging modalities (Fig. 6). These lesions may undergo a chondrosarcomatous transformation, presenting with a more destructive lytic lesion with a soft tissue mass. However, the most sensitive indicator of sarcomatous transformation is pain. Ollier disease (multiple enchondromatosis) and Maffucci syndrome (enchondromatosis and soft tissue hemangiomas) are associated entities. Both of these entities are associated with an increased rate of malignant transformation of ECs.¹¹

Juxtacortical Chondroma

Juxtacortical or periosteal chondroma (JC) compromises less than 2% of primary bone lesions and presents as focal swelling. It is most common in men younger than the age of 30. Histologically, JC demonstrates a lobular cartilaginous growth limited to the cortex and beneath the periosteum without an extension into the medullary cavity. It



Fig. 5. A 35-year-old woman with a large osteochondroma arising from the posterior cortex of the proximal tibia. (*A*) Lateral radiograph. (*B*) Axial CT clearly demonstrates the continuity of cortex with underlying bone (*arrow*). (*C*) Fat-saturated T2-weighted MR image with a thin cartilage cap (*arrow*).

is most common in the proximal humerus followed by the femur, tibia, hands, and feet (see **Table 1**). Pelvis and ribs are uncommon sites.⁹

On radiography JC presents as a focus of cortical saucerization with a soft tissue mass that may contain a chondroid matrix. A well-defined sclerotic margin separates the lesion from the medullary cavity. Overhanging cortical edges may be present and may entirely cover the lesion. CT aids in demonstration of the matrix mineralization and MR imaging shows the high water content of the cartilage tissue. Both modalities confirm the lack of communication of this lesion with the medullary canal (Fig. 7). The main differential

diagnosis is a surface chondrosarcoma, which usually presents later in life; is larger (>3 cm); and on imaging demonstrates invasion of the medullary cavity. Other lesions that may arise from the surface of bone, such as a surface osteosarcoma, ABC, or chondromyxoid fibroma (CMF), also need to be excluded.¹²

Chondroblastoma

Chondroblastoma (CB) is an uncommon lesion compromising less than 2% of benign primary bone neoplasms. It occurs close to joints and may present with some loss of joint function. CB



Fig. 6. A 57-year-old woman with an incidental enchondroma of the distal femur. (A) Anteroposterior radiograph of the knee shows a central mineralized lesion (arrows). (B) Axial CT shows the chondroid matrix with characteristic "ring-and-arc" mineralization (arrow). (C) Coronal fat-saturated proton density MR image demonstrates the high signal lesion (arrow) with lobulated superior margins.

is twice as common in males as females, and most cases occur between the ages of 5 and 25 years. Histologically, it presents as a sharply marginated and lobulated lesion containing chondroblasts, occasional giant cells, and abundant chondroid matrix. ABC components may be present in up to 15% of cases and matrix mineralization occurs in a lacelike pattern with a histologic appearance similar to chicken wire. CB occurs predominantly in the epiphyses or epiphysis equivalents (apophyses and sesamoids). It is most commonly seen in the proximal femur followed by the distal femur, proximal tibia, proximal humerus, and hands and feet (see Table 1).⁹

On radiographs CB presents as a lucent lesion of the epiphysis with a thin sclerotic rim. It may have a subtle chondroid matrix, which is better demonstrated on CT. CT also may characterize the adjacent periosteal reaction, which is usually an inflammatory response. CB may appear more expansile depending on the size of the ABC component. A more aggressive appearance may be encountered featuring cortex destruction and invasion of the surrounding soft tissues or the joint space. MR imaging readily demonstrates the surrounding marrow edema and the typical low-tointermediate signal of the tumor on fluid-sensitive sequences (Fig. 8). This is a distinguishing feature of this cartilage lesion, which has been attributed to the high hemosiderin content or highly cellular matrix. The differential diagnosis for epiphyseal lesions includes giant cell tumor (GCT), subchondral cyst, infection, Langerhans cell histiocytosis, OB, and clear cell chondrosarcoma.^{9,13}

Chondromyxoid Fibroma

CMF is rare (<1% of primary bone tumors). It is composed of varying degrees of cartilaginous, fibrous, and myxoid components. It presents with swelling and may occasionally be painful. CMF has a slight male predilection and affects a wide range of age between 3 and 70 years; however, most cases occur younger than age 30. Histologically, CMF presents as a sharply demarcated lesion containing the aforementioned components with small foci of calcification and common multinucleated giant cells, usually at the periphery of





Fig. 7. A 21-year-old man with a JC arising from the proximal humerus (*arrow*). (*A*) Frontal shoulder radiograph in internal rotation shows a surface lesion of the humeral head and neck. (*B*) Axial CT shows mineralization of the lesion. (*C*) Axial T1-weighted MR image shows a low-to-intermediate signal surface lesion of the proximal humerus.

the cartilaginous lobules. It arises most often around the knee with the proximal tibia as the single most common location. The foot and pelvic bones also are common sites. CMF is usually located in the metaphysis of the long bones as an eccentric lesion, and it is often cortical (see **Table 1**).⁹

On radiography CMF presents as an eccentric, expansile metaphyseal lesion of the long bones with an inner narrow zone of transition with the adjacent normal bone and a thinned or absent outer margin simulating a more aggressive lesion. In the foot and flat bones it presents as a central expansile lesion. CT may reveal trabeculation and a mineralized matrix. CT and MR imaging may demonstrate a lobulated margin. MR imaging also demonstrates the characteristic high signal water-laden cartilage matrix (Fig. 9).⁹

TUMORS FORMING OR ARISING FROM FIBROUS TISSUE

The common feature of these often eccentric benign tumors is a fibrous matrix with adjacent bone sclerosis.

Fibroxanthoma

The histopathologic term "fibroxanthoma" (FX) covers both the larger nonossifying fibroma and the smaller fibrous cortical defect. FX encompasses



Fig. 8. An 11-year-old boy with a chondroblastoma of the proximal humerus. (*A*) Frontal shoulder radiograph in internal rotation shows a lucent lesion (*arrows*) involving the humeral head and neck. (*B*) Sagittal reformatted CT images demonstrates epiphyseal location of the lesion with extension into the metaphysis (*arrows*) and matrix mineralization suggesting a chondroid lesion. (*C*) Axial fat-saturated T2-weighted MR image shows the chondroblastoma (*arrow*) with surrounding soft tissue edema (*arrowhead*).

about 2% of biopsied primary bone tumors. This lesion is usually asymptomatic and demonstrates a tendency for spontaneous healing. The larger nonossifying fibroma may present with a painful pathologic fracture. FX is more common in males with a peak incidence in the first two decades of life. Histologically, it presents with bundles of fibrous tissue with a few multicentric giant cells along with foam or xanthoma histiocytes. Occasional foci of necrosis and hemorrhage (hemosiderin) may be visible.¹⁴ FX is a metaphyseal lesion that can migrate into the diaphysis with growth. It affects in up to 90% of cases the long tubular bones. FX is eccentric ("cortically based") and up to 55% of cases arise about the knee (femur, tibia, and fibula). It is uncommon in the upper extremity (see **Table 1).**¹⁵

Radiographically, the main differentiation between nonossifying fibroma and fibrous cortical defect is the size of the lesion. An exact size criterion may be unnecessary because histologically they have a similar pattern. The fibrous cortical defect is smaller, cortically based, and well marginated. The nonossifying fibroma tends to be lobulated and expansile with trabeculation and a longitudinal growth pattern. A rim of sclerosis and thinned or absent cortices may be encountered on radiographs, CT, and MR imaging. Because of the high fibrous content FX is often of predominantly low signal on all MR pulse sequences (Fig. 10). The involuted lesion may appear as normal bone with mild expansion or contain a sclerotic matrix. This lesion has a typical appearance in long bones and a differential diagnosis is often unnecessary. The most well-known associated entity is the Jaffe-Campanacci syndrome with multiple FXs, skin changes, and mental retardation.14,15



Fig. 9. A 33-year-old man with a distal femoral chondromyxoid fibroma (*arrow*). (*A*) Frontal radiograph of the knee shows a lucent metaphyseal lesion with small foci of mineralization. (*B*) Axial CT scan shows the thin anterior margin of lesion with foci of calcification. (*C*) Axial fat-saturated T2-weighted MR image demonstrates the partially lobulated margins of the intermediate-to-high signal lesion.

Fibrous Dysplasia

Fibrous dysplasia (FD) is usually a noninherited developmental anomaly of bone formation in which the normal bone marrow and cancellous bone is replaced by immature bone and fibrous tissue. It compromises up to 7% of benign bone tumors and less than 1% of biopsied bone tumors. FD is usually an incidental finding unless complicated by a pathologic fracture or rare malignant transformation. Males and females are equally affected, and although the lesion can be detected at any age, there is a peak incidence between 2 and 30 years. Histologically, it presents as a fibroosseous metaplasia with islands of pure woven bone creating a typical "alphabet soup" pattern. ABC components also may be present. Most cases are monostotic (70%-80%), but up to 30% may be polyostotic. FD is most commonly found in the femur followed by tibia, skull and facial bones, and ribs. It is uncommon in the hand and foot, spine, and clavicle. In the long bones it presents as a medullary diaphyseal lesion (see **Table 1**).¹⁶

The radiographic features include a usually welldefined lucent expansile lesion with a sclerotic rim. FD may be multiloculated and the woven bone creates a "ground glass" appearance. Because of growth disturbance or underlying fracture, it may create a bowing deformity of the bone. CT can aid in detailing these radiographic features. On MR imaging FD is of low signal on T1-weighted images with a variable appearance on T2-weighted images ranging from intermediate to high signal intensity (**Fig. 11**). The differential diagnosis may include a simple bone cyst (SBC), Paget disease, and in the skull meningioma. The risk of a pathologic fracture depends on the location of the lesion and may



Fig. 10. A 17-year-old boy with a distal femur nonossifying fibroma (*arrow*). (A) Frontal radiograph shows an eccentric lesion with sclerotic lobulated margins. (B) Axial CT scan confirming the eccentricity of the lesion and lack of a mineralized matrix.



Fig. 11. Fibrous dysplasia of the proximal femur (*arrow*). (*A*, *B*) A 46-year-old man. Frontal radiograph (*A*) shows a lesion with a "ground glass" appearance and sclerotic margins in the femoral neck. Fat-saturated proton density weighted MR image (*B*) shows a hyperintense lesion with low signal intensity margins. (*C*) A 37-year-old man. CT (coronal reformation) shows the sclerotic rim (*arrow*) and foci of ground glass density along the lesion margin.

be related to the size of the ABC component. Malignant transformation into a sarcoma occurs in only 1% of cases and presents with pain and swelling; radiologically, cortical destruction and a soft tissue mass are prominent features. Syndromes associated with the polyostotic variant include McCune-Albright (precocious puberty and skin pigmentation in girls) and Mazabraud (rare combination with soft tissue myxomas).¹⁶

Osteofibrous Dysplasia

Osteofibrous dysplasia (OFD) is an unusual lesion compromising less than 0.2% of biopsied benign bone tumors. It presents with focal swelling and is painless unless complicated by a fracture. The patients are usually younger than 10 years and rarely older than 16. There is no gender predilection. Histologically, OFD presents with a vascularized fibrous stroma similar to FD without the alphabet soup woven bone pattern. Up to 80% of cases are found in the tibia, the remainder occurring in the fibula only or in combination with a fibular lesion. Radius and ulna lesions are extremely rare. OFD is usually based in the anterior cortex of the tibia (see Table 1).¹⁷

On radiographs and CT, OFD presents as an expansile lytic lesion of the anterior tibial cortex with a mixed lytic and sclerotic matrix. On MR imaging, OFD is usually of intermediate signal on T1- and high on T2-weighted images (Fig. 12). In many cases, patients with OFD can simply be observed closely after biopsy confirms the diagnosis; however, the most crucial entity in the differential diagnosis is the adamantinoma, a low-grade malignant tumor. OFD and adamantinoma components may both be present in a lesion and needle biopsy may not be conclusive. Imaging features suggesting adamantinoma, such as a more locally aggressive appearance with multiple lytic lesions, invasion of the medullary canal, and a soft tissue mass, may aid in the differential diagnosis; however, in these cases the entire lesion should undergo curettage and histologic investigation.¹⁷

Desmoplastic Fibroma

Desmoplastic fibroma (DF) is a rare lesion accounting for only 0.1% of benign bone tumors. It is considered the bony counterpart of the more common soft tissue desmoid. The clinical presentation is nonspecific and may include pain and swelling. There are controversial reports about gender distribution, but one review mentions a 1.5:1 male to female ratio.¹⁸ DF is mostly found in the third and fourth decades of life. On histologic analysis it reveals a similar appearance to its soft tissue counterpart with a white-to-gray mass containing fibroblasts producing well-formed collagen. DF is mostly located centrally in the metadiaphysis of tubular bones around the knee (femur and tibia) followed by the humerus and radius (see **Table 1**).¹⁹



В

Fig. 12. A 15 year old with anterior tibial osteofibrous dysplasia. (A) CT, sagittal reformation shows readily the location of the lesion in the anterior cortex of the bone (*arrow*). (B) CT, axial image shows the location of the lesion within the anterior cortex and an additional lucent focus in the lateral cortex of the tibia (*arrow*).

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On radiography DF frequently has an aggressive appearance presenting as a lytic expansile lesion. The lesion may have internal trabeculation and a sclerotic margin. CT confirms lack of a mineralized matrix and may demonstrate cortical breaching and a soft tissue component. The hallmark of DF on MR imaging is intermediate-to-low signal intensity on fluid-sensitive sequences because of the high collagen content of its fibrous component (**Fig. 13**).¹⁸ The differential diagnosis includes a low-grade central osteosarcoma and FD. Rare association with FD has been described.¹⁹

HISTIOCYTIC OR FIBROHISTIOCYTIC TUMORS

The main benign representative of this group is the Langerhans cell histiocytosis, which is discussed elsewhere in this issue.

TUMORS WITH A FATTY MATRIX Intraosseous Lipoma

Intraosseous lipoma (IL) is the main benign variant of this group. It is usually an incidental finding and only presents with pain if there is an associated



Fig. 13. A 28-year-old woman with a desmoplastic fibroma of the distal femur (*arrow*). (*A*) Anteroposterior radiograph of the knee shows a large lucent lesion of the metaphysis with expansile remodeling and trabeculation. (*B*) CT, coronal reformation shows a sclerotic margin and lack of matrix mineralization. (*C*) Coronal proton density weighted MR image confirms presence of low-to-intermediate signal components within the lesion.

fracture. IL has been considered to be a very rare bone tumor, although a general increase in use of cross-sectional imaging has resulted in more frequent reporting. The absolute etiology of this tumor is not well known and speculation includes its possible association with focal stress, osteoporosis, or involution of an underlying primary bone tumor. There is an equal gender distribution with a possible slight male predominance. There is a wide age range (from 4-85 years) with a mean of 40 years. Histologically, IL demonstrates a mature fatty matrix devoid of medullary trabeculation. Areas of fat necrosis with associated foamy histiocytes or cyst formation are encountered. It is most commonly found in the calcaneus followed by the metaphysis of the long bones, such as femur, tibia, and fibula. It is usually centrally located within the bone (see Table 1).²⁰

On radiography IL presents as a well-defined lucent lesion with a narrow zone of transition without remodeling of the bone. A central calcification or ossification may be present. The mature fatty matrix can be confirmed by Hounsfield measurement on CT and fat-saturation on MR imaging (Fig. 14). On CT and MR imaging a central ossification is readily visible and cystic components are encountered commonly. The differential diagnosis includes a metastatic or myeloma lesion on radiography; however, demonstration of a fatty matrix on cross-sectional imaging secures the correct diagnosis, with no further work-up necessary.²⁰

TUMORS WITH A VASCULAR MATRIX

The main representatives of this tumor type in the bone are osseous hemangioma (OH) and the glomus tumor (GT). Solitary or multiple osseous lymphangiomas may have an appearance similar to bony hemangiomas, but are not as common.

Osseous Hemangioma

The predominant matrix of an OH is vascular. The capillary subtype is most common; however, mixed tumors with other vascular subtypes may be present. OH is usually an incidental finding and asymptomatic. It is twice as common in males as females, and affects individuals in the fourth or fifth decade of life. Histologically, OH demonstrates vascular replacement of the marrow with possible calcified or cystic foci and a reactive sclerotic margin. One article reports that up to 75% of OH occur in the vertebrae, but they also occur in the calvarium, calcaneus, and the long bones.²¹ Hemangiomas in the vertebrae may partially or completely replace the vertebral body. OH commonly presents in the long bones as a central metaphyseal lesion (see Table 1).22

Vertebral hemangiomas demonstrate a typical striated appearance on radiography similar to corduroy. This translates into a polka dot appearance on axial CT.²³ In other bones OH presents as a lytic lesion with a honeycomb pattern and cortical erosion. Advanced imaging with CT and MR imaging may show a soft tissue component with



Fig. 14. A 78-year-old man with an incidental intraosseous lipoma of the proximal tibia (*white arrow*) with a central ossification (*black arrow*). (*A*) Anteroposterior radiograph of the knee. (*B*) CT, axial image. Note attenuation of the lesion similar to that of subcutaneous fat.

phleboliths. On MR imaging the fatty component interspersed between the vascular channels may exhibit high T1 signal; the predominant T2 signal is usually high (Fig. 15).6,22 The differential diagnosis of extraspinal OH on plain radiography may include infection and metastatic disease, although cross-sectional imaging usually confirms the presence of a hemangioma. OH may be encountered as the smaller component of extensive diffuse soft tissue hemangiomatosis.

Glomus Tumor

GT is a rare benign lesion that almost exclusively occurs in the subungual region of the terminal phalanx. It is technically a soft tissue tumor that arises from the specialized arteriovenous anastomosis responsible for thermoregulation of the fingertips. GT commonly erodes into the tuft of the distal phalanx, thus simulating a bony tumor.²⁴ GT is described in further detail elsewhere in this issue.

TUMORS OF MISCELLANEOUS OR **UNKNOWN ORIGIN**

This group consists of bony lesions of unknown etiology. The more common representatives are discussed next. Other entities commonly grouped in this category but not discussed here include epidermoid inclusion cyst, subchondral cyst, intraosseous ganglion, and posttraumatic cyst.



В

spine shows the "corduroy" appearance of the hemangioma. (B) CT, axial image demonstrates the characteristic polka dot appearance. (C) Axial T1-weighted MR image shows the replacement of the vertebral body with a soft tissue mass with low signal striation and extraosseous soft tissue component (black arrow).

Giant Cell Tumor

GCT of the bone is a relatively common bone tumor comprising up to 10% of primary bone tumors. It may present with pain, swelling, and at times a pathologic fracture. GCT may grow with pregnancy or intake of oral contraceptives. It is slightly more common in females and affects individuals after skeletal maturity with a peak incidence in the third decade of life. Histologically, GCT presents with an abundance of multinucleated giant cells on a background of mononuclear cells. The stroma is usually vascular with numerous capillaries, thus microscopic foci of internal hemorrhage and ABC-like components are possible. GCT is usually located at the ends of the tubular bones extending to the subchondral bone and typically is eccentric in location (see **Table 1).**25

Radiography reveals a characteristic expansile lytic lesion at the end of the bone with a narrow zone of transition. CT may reveal a partially sclerotic rim and confirms the lack of matrix mineralization. CT and MR imaging may demonstrate a soft tissue component. On MR imaging a lowto-intermediate signal intensity prevails on T1weighted and the fluid-sensitive sequences (**Fig. 16**). The lower T2 signal compared with other subarticular lesions may be caused by hemosiderin deposition or increased cellularity. GCT has a high recurrence rate (up to 25%) after curettage or resection. It is the most common lesion associated with secondary ABC. The term "benign metastasizing GCT" is reserved for a histologically benign tumor with aggressive growth and distant metastatic deposits.²⁵

Simple Bone Cyst

A SBC is a true cystic lesion of the bone. Other terms used, such as a "unicameral," "solitary," or "juvenile" bone cyst are misleading because the lesion may be multiloculated, rarely multiple, and occasionally seen in adults. SBC comprises about 3% of all biopsied primary bone tumors. SBC frequently presents with pain after a pathologic fracture; however, incidental asymptomatic occurrence is not uncommon. There is a predilection for males with a male to female ratio of 2.5:1. SBC mostly occurs in the first two decades of life with



Fig. 16. A 27-year-old man with a fibular head giant cell tumor (*white arrow*). (*A*) Frontal radiograph of the knee shows the extension of the metaphyseal lesion into the epiphysis and subcortical bone. (*B*) CT, axial image, confirms lack of a mineralized matrix and thinning of the cortex. (C) Axial fat-saturated T2 image shows the predominantly intermediate signal lesion with a cystic focus (*black arrow*).

less than 15% occurring after 20 years of age. Histologically, the uncomplicated SBC demonstrates a thin cyst wall composed of a single layer of mesothelial cells. The serous fluid content is usually under pressure. SBC complicated by a fracture contains serosanguinous fluid and fibro-osseous repair tissue. In the pediatric population up to 60% of cases occur in the proximal humerus followed by 30% in the proximal femur. In the adult population there is a preference of location in the calcaneus and iliac bones adjacent to the sacroiliac joints. SBC is usually located centrally in the metaphysis of the bone (see **Table 1**).^{26,27}

In a child SBC presents radiographically as a mildly expansile lucent lesion of the metaphysis invariably abutting the open growth plate. With skeletal growth the lesion migrates into the diaphvsis, which has been termed the "latent" form. The appearance may be more complex with an underlying fracture. A "fallen fragment" sign has been described as a pathognomic finding with the fracture fragment in the dependent portion of the lesion confirming the "simple" fluid content of the lesion. This has been reported in up to 5% of lesions. The "rising bubbles" sign is based on similar priniciples.²⁸ MR imaging, and in particular CT, may better demonstrate the details of an underlying pathologic fracture. In an uncomplicated SBC, CT and MR imaging exhibit characteristic features of a lesion with simple fluid content (ie, a Hounsfield unit equal to simple fluid on CT and high T2 signal on MR imaging) (Fig. 17).²⁶

Aneurysmal Bone Cyst

ABC is a benign bone lesion that is neither a cyst nor a neoplasm.²⁹ Several theories have been postulated regarding its origin. The most entertained is a reactive vascular process of native bone caused by antecedent trauma or a precursor tumor. ABC is rare (<2% of biopsied benign bone lesions) and usually presents with pain and swelling. A juxta-articular ABC may be the source of limited range of motion and the spinal variants may cause nerve and cord impingement. Pathologic fractures occur in up to 20% of patients. ABC is slightly more common in females and is seen in patients younger than age 20. The gross pathologic appearance has been described as that of a blood-filled sponge. On histology, cavernous blood-filled spaces lined by fibrous walls and osteoclast-rich giant cells prevail. A primary ABC has no associated underling lesion. A secondary ABC refers to those associated with an underlying bone tumor, most commonly GCT, followed by CB, OB, and FD. Over half of ABC cases occur in long bones and about 20% are encountered in the spine (predominantly the posterior elements). Pelvic bones are the most common flat bones affected. In long bones an eccentric metaphyseal location is seen in 80% of cases. A diaphyseal location, usually cortical or subperiosteal, is seen in approximately 20% of cases (see Table 1).^{26,27}

ABC is the only bone lesion named for its radiographic appearance. Three phases have been



Fig. 17. A 10-year-old boy with a simple bone cyst of the proximal humerus (*white arrow*). (*A*) Lateral radiograph of the humerus with a "fallen fragment" (*black arrow*). (*B*) CT, coronal reformation image with a cortical fracture (*black arrow*).



Fig. 18. A 15-year-old boy with a subperiosteal distal tibia aneurysmal bone cyst (*white arrow*). (*A*) Lateral radiograph of the ankle. *Black arrow* points to the extraosseous component. (*B*) CT, axial image, demonstrates a thin sclerotic rim. (*C*) Axial fat-saturated T2-weighted MR image shows the fluid–fluid levels in the lesion.

described. Initially, a focal area of osteolysis may be seen mimicking other benign bone tumors, such as FD or SBC. This is followed by an active or growth phase characterized by locally aggressive enlargement, bone destruction, and marked expansile remodeling. The outer border demonstrates extreme thinning, whereas the inner margin appears usually less aggressive. Periosteal reaction may occur suggesting a malignant tumor, although bone expansion with an intact rim of cortex usually supports the presence of a benign lesion. The stabilization phase is characterized by progressive calcification and ossification of the rim. CT imaging aids in excluding a mineralized matrix and reveals in up to 35% of cases one or multiple fluid-fluid levels. MR imaging is more sensitive in revealing the fluid-fluid levels, which may have a variable T1 and T2 appearance depending on the stage of blood products (Fig. 18). With secondary ABC, MR imaging also more readily demonstrates the solid underlying primary tumor. The differential diagnosis includes teleangiectatic osteosarcoma, GCT, OB, and CB, all of which may also contain fluid–fluid levels in the absence of a secondary ABC.^{26,27}

SUMMARY

At first glance the benign bone tumors may appear confusing. However, thorough knowledge of radiologic characteristics of these benign lesions aids in a succinct differential diagnosis. The radiologic appearance of this group of bone tumors plays a major role in appropriate clinical management, preventing unnecessary patient anxiety, medical intervention, and ultimately added healthcare cost.

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