Benign Tumors and Tumor-like Lesions II

Lesions of Cartilaginous Origin

Benign Chondroblastic Lesions

Diagnosis of a bone lesion as originating from cartilage is usually a simple task for the radiologist. The lesion’s radiolucent matrix, scalloped margins, and anicular comma-shaped or punctate calcifications usually suffice to establish its chondrogenic nature. However, whether a cartilage tumor is benign or malignant is sometimes extremely difficult for the radiologist to determine.

Enchondroma (Chondroma)

Enchondroma is the second most common benign tumor of bone, constituting approximately 10% of all benign bone tumors and representing the most common tumor of the short tubular bones of the hand. When the lesion is located centrally in the bone, it is termed an enchondroma (Fig. 18.1); if it is extracortical (periosteal) in location, it is called a chondroma (periosteal or juxtacortical) (see Figs. 18.10 and 18.11). Regardless of location, this benign lesion is characterized by the formation of mature hyaline cartilage. It has been widely postulated that enchondroma is formed as a consequence of displacement of embryonic rests of cartilage from the growth plate into the metaphysis. This contention, however, was recently challenged by some investigators whose work failed to confirm this theory. Furthermore, the study by Amary and colleagues identified somatic mutations in isocitrate dehydrogenase 1 and 2 (IDH1 and IDH2) in many central low-grade cartilaginous tumors, thus supporting the neoplastic origin of enchondromas. Moreover, most enchondromas contain clonal chromosomal abnormalities involving chromosomes or chromosomal regions 4q, 5, 7, 11, 14q, 16q22-q24, 20, and particularly rearrangement of chromosome 6 and 12q12-q15. Although occurring throughout life, enchondromas are usually seen in patients in their second through fourth decades. There is no sex predilection.

The short tubular bones of the hand (phalanges and metacarpals) are the most common sites of occurrence (Fig. 18.2), although the lesions are also encountered in the long tubular bones (Fig. 18.3). Sporadic cases have been reported in the rib, clavicle, cuboid, and carpal bones. They are often asymptomatic; a pathologic fracture through the tumor (Figs. 18.4 and 18.5) often calls attention to the lesion.

Enchondroma protuberans is a rare variant. It is a lesion that arises in the intramedullary cavity of a long bone and forms a prominent exophytic mass on the cortical surface. This lesion must be distinguished from an osteochondroma or central chondrosarcoma that penetrates the cortex and forms a juxtacortical mass.

In most instances, radiography suffices to demonstrate the lesion. In the short bones, the lesion is often entirely radiolucent (Fig. 18.6), whereas in the long bones, it may display visible calcifications. If the calcifications are extensive, enchondromas are called calcifying (Fig. 18.7). The lesions can also be recognized by shallow scalloping of the inner (endosteal) cortical margins because the cartilage in general grows in a lobular pattern (see Fig. 18.1).

Computed tomography (CT) and magnetic resonance imaging (MRI) may further delineate the tumor and more precisely localize it in the bone. On spin echo T1-weighted MR images, enchondromas demonstrate intermediate to low signal intensity, whereas on T2-weighted images, they exhibit high signal intensity. The calcifications within the tumor will image as low-signal intensity structures (Figs. 18.8 and 18.9). It must be stressed, however, that most of the time neither CT nor MRI is suitable for establishing the precise nature of a cartilaginous lesion, nor can CT or MRI distinguish benign from malignant lesions. Despite the use of various criteria, the application of MRI to the tissue diagnosis of cartilaginous lesions has not brought satisfactory results, although preliminary results of recent trials with fast contrast-enhanced MR imaging showed that this technique might assist in differentiation between the benign and malignant cartilaginous tumors.

Skeletal scintigraphy usually reveals mild to moderate increased uptake of the tracer in uncomplicated enchondromas, whereas the presence of a pathologic fracture or malignant transformation is revealed by marked scintigraphic activity.

Intracortical chondroma is a very rare variant of conventional enchondroma. The lesion is located in cortical bone and is surrounded by sclerosis of the medullary bone and periosteal reaction. Some of these lesions may actually represent periosteal chondroma with an atypical radiographic appearance, as reported by Abdelwahab and associates. Intracortical chondroma can occasionally simulate an osteoid osteoma.

Periosteal chondroma is a slow-growing, benign cartilaginous lesion that arises on the surface of a bone in or beneath the periosteum. It occurs in children as well as adults, with no sex predilection. There is usually a history of pain and tenderness, often accompanied by swelling at the site of the lesion, which is most commonly located in the proximal humerus. As the tumor enlarges, it is seen radiographically eroding the cortex in a saucer-like fashion, producing a solid buttress of periosteal new bone (Fig. 18.10). The lesion has a sharp sclerotic inner margin demarcating it from the buttress of periosteal new bone. Scattered calcifications are often seen within the lesion (Fig. 18.11).

CT may show to better advantage the scalloped cortex and matrix calcification (Fig. 18.12). It also may demonstrate the separation of a lesion...
FIGURE 18.1 Enchondroma. A radiolucent lesion in the medullary portion of the proximal femur of a 22-year-old man is seen eroding the inner aspect of the lateral cortex. Note scalloped borders and matrix calcification.

FIGURE 18.2 Enchondroma. (A) A radiolucent lesion in the proximal phalanx of the middle finger of a 40-year-old woman and (B) a similar lesion with central calcification in the proximal phalanx of the ring finger of a 42-year-old man are typical examples of enchondroma in the short tubular bones.

FIGURE 18.3 Skeletal sites of predilection, peak age range, and male-to-female ratio in enchondroma.
FIGURE 18.4  Enchondroma. Radiograph of a 31-year-old man who had injured his left thumb reveals a pathologic fracture through an otherwise asymptomatic lesion.

FIGURE 18.5  Enchondroma. Pathologic fracture through a large enchondroma is present in the proximal phalanx of the middle finger.

FIGURE 18.6  Enchondroma. A typical, purely radiolucent lesion at the base of the proximal phalanx of the ring finger of a 37-year-old woman represents an enchondroma. Note the marked attenuation of the ulnar side of the cortex.

FIGURE 18.7  Calcifying enchondroma. In this heavily calcified enchondroma of the proximal humerus of a 58-year-old woman, note the lobular appearance of the lesion and the minimal degree of scalloping of the lateral endocortex.
**FIGURE 18.8 MRI of enchondroma.**
Anteroposterior (A) and lateral (B) radiographs of the left knee of a 61-year-old man demonstrate only a few calcifications in the distal femur (arrows). The extent of the lesion cannot be determined. Coronal (C) and sagittal (D) T1-weighted MR images show a well-circumscribed, lobulated lesion displaying intermediate signal intensity. The darker area in the center represents calcifications. Coronal T2-weighted image (E) shows the lesion displaying a mixed-intensity signal: The brighter areas represent cartilaginous tumor and the darker areas calcifications.

**FIGURE 18.9 MRI of enchondroma.**
(A) Lateral radiograph of the knee shows chondroid calcifications in the distal femur (arrows). Coronal (B) and sagittal (C) spin echo T1-weighted MR images show the lesion being predominantly of low signal intensity. Coronal (D) inversion recovery T2-weighted with fat saturation and sagittal (E) fast spin echo T2-weighted images demonstrate the full extent of enchondroma. Calcifications exhibit low signal intensity.
**FIGURE 18.10** Periosteal chondroma. A radiolucent lesion (arrow) is eroding the external surface of the cortex of the proximal humerus of a 24-year-old man.

**FIGURE 18.11** Periosteal chondroma. A periosteal chondroma at the medial aspect of the neck of the left femur eroded the cortex in a saucer-like fashion. The characteristic buttress of a periosteal reaction is seen at the inferior border of the lesion (arrow). Note also cluster of calcification in the soft tissue (curved arrow).

**FIGURE 18.12** CT of periosteal chondroma. (A) An oblique radiograph of the right ankle shows a lesion containing calcifications eroding the medial cortex of the distal fibula. CT using a bone window (B) and a soft-tissue window (C) better demonstrates the extent of the lesion and the distribution of the calcifications.
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A large periosteal chondroma eroded the cortex of the proximal fibula and extended into the medullary cavity. Coronal (B) proton density (spin echo [SE]; repetition time [TR] 2000/echo time [TE] 19 msec) and sagittal (C) T2-weighted (SE; TR 2000/TE 70 msec) MRI show the lesion’s extension into the bone marrow.

MRI is marrow edema mimicking tumor invasion or vice versa. Unlike enchondroma and osteochondroma, periosteal chondroma may continue to grow after skeletal maturation. Some lesions may attain a large size (up to 6 cm) and may resemble osteochondromas (Figs. 18.14 and 18.15). Some lesions may mimic an aneurysmal bone cyst. Very rarely, the lesion may encase itself intracortically, thus mimicking other intracortical...
lesions (such as intracortical angioma, intracortical fibrous dysplasia, or intracortical bone abscess).

Histologically, enchondroma consists of lobules of hyaline cartilage of varyingcellularity and is recognized by the features of its intracellular matrix, which has a uniformly translucent appearance and contains relatively little collagen. The tissue is sparsely cellular, and the cells contain small and darkly staining nuclei. The tumor cells are located in rounded spaces known as lacunae. On histologic examination of periosteal chondroma, the findings are identical to those of enchondroma, although the lesion sometimes exhibits higher cellularity, occasionally with atypical cells.

Differential Diagnosis
The main differential diagnosis of enchondroma, particularly in lesions of the long bones, is a medullary bone infarct (Fig. 18.16). At times, the two lesions may be difficult to distinguish from one another, particularly if the enchondroma is small, because both lesions present with similar calcifications. The radiographic features helpful in the differential diagnosis are the lobulation of the inner cortical margins in enchondroma, the annular, punctate, and comma-shaped calcifications in the matrix, and the lack of sclerotic rim that is usually seen in bone infarcts (Fig. 18.17).

The most difficult task for the radiologist is to distinguish a large solitary enchondroma from a slowly growing low-grade chondrosarcoma. One of the most significant findings pointing to a chondrosarcoma in the early stage of development is localized thickening of the cortex and deep endosteal scalloping (Fig. 18.18). The size of the lesion should also be taken into consideration. Lesions longer than 4 cm (or, according to some investigators, longer than 7 cm) are suggestive of malignancy. In more advanced tumors, destruction of the cortex and the presence of a soft-tissue mass are the hallmarks of malignancy.

Complications
The single most important complication of enchondroma, aside from pathologic fracture (see Fig. 18.4), is its malignant transformation to chondrosarcoma. With solitary enchondromas, this occurs almost exclusively in a long or flat bone and almost never in a short tubular bone. The radiographic signs of the transformation are thickening of the cortex, destruction of the cortex, and a soft-tissue mass. The development of pain in the absence of fracture at the site of the lesion is an important clinical sign.

**Treatment**
Curettage of the lesion with the application of bone graft is the most common course of treatment.

**Enchondromatosis, Ollier Disease, and Maffucci Syndrome**

Enchondromatosis is a condition marked by multiple enchondromas, generally in the region of the metaphysis and diaphysis (Fig. 18.19). If the skeleton is extensively affected, with predominantly unilateral distribution, the term *Ollier disease* is applied. The clinical manifestations of multiple enchondromas, such as knobby swellings of the digits or gross disparity in the length of the forearms or legs, are frequently recognized in childhood and adolescence; the disease has a strong preference for one side of the body. The disorder has no hereditary or familial tendency. Some investigators claim that it is not a neoplastic lesion but rather a developmental bone dysplasia. Maffucci syndrome is a congenital, nonhereditary disorder, characterized by enchondromatosis and soft-tissue angiomatosis (hemangiomatosis). The hemangiomas may occur anywhere in the skin and subcutaneous tissue. They are usually cavernous in type and may form unilaterally or bilaterally. The enchondromas in Maffucci syndrome have predilection for the tubular bones and have the same distribution as in Ollier disease, with a strong predisposition for one side of the body, the metacarpals and the phalanges being the most common sites. The pathogenesis of Ollier disease and Maffucci syndrome is unknown. The recent investigations, however, suggested that these disorders represent two entities within continuum of enchondromatosis and that both conditions bear the risk of mesodermal and nonmesodermal malignancy, caused by somatic mosaic mutations of *IDH1* and *IDH2* genes.

Conventional radiography is usually sufficient to demonstrate the typical features of enchondromatosis/Ollier disease. Characteristically, interference of the lesion with the growth plate causes foreshortening of the limbs. Deformity of the bones is marked by radiolucent masses
Figure 18.17 Bone infarct. (A) Conventional radiograph of the proximal tibia shows the typical coarse calcifications of medullary bone infarct. Note the sharply defined peripheral margin separating necrotic from viable bone and the lack of characteristics for chondroid tumor annular and comma-shaped calcifications. (B) In another patient with a bone infarct in the distal femur, a CT section reveals central coarse calcifications and no endosteal scalloping of the cortex.
FIGURE 18.18 Low-grade chondrosarcoma. (A) A 48-year-old woman presented with pain in the upper leg. A radiograph shows a radiolucent lesion in the proximal tibia with a wide zone of transition and central calcifications. Note focal thickening of the cortex (arrows), an important feature that distinguishes chondrosarcoma from similarly appearing enchondroma. In another patient, a 57-year-old woman, the radiograph of the distal femur (B) and coronal T1-weighted fat-suppressed MR image obtained after intravenous injection of gadolinium (C) show deep endosteal scalloping (arrows). Excision biopsy revealed a low-grade chondrosarcoma. (D) Classic enchondroma is shown for comparison. Note that cortex is not thickened, and despite the size of the lesion that abuts the endocortex, there is lack of endosteal scalloping.
of cartilage, often in the hand and foot, containing foci of calcification (Fig. 18.20). Enchondromas in this location may be intracortical and periosteal. They sometimes protrude from the shaft of the short or long tubular bone, thus resembling osteochondromas (Fig. 18.21). Linear columns of cartilage in the form of radiolucent streaks extend from the growth plate to the diaphysis, and a fan-like pattern is common in the iliac bones (Fig. 18.22). MRI demonstrates lobulated in contour masses exhibiting low-to-intermediate signal intensity on T1-weighted images and high signal on T2 weighting. After injection of gadolinium, there is various degree of enhancement (Fig. 18.23). The Maffucci syndrome, in addition to the typical osseous alterations of enchondromatosis, is recognized radiographically by the presence of multiple calcified phleboliths (Fig. 18.24).

Histologically, the lesions of enchondromatosis are essentially indistinguishable from those of solitary enchondromas, although on occasion they tend to be more cellular.

**Complications**

The most frequent and severe complication of Ollier disease is malignant transformation to chondrosarcoma. In contrast to solitary enchondromas,
even lesions in the short tubular bones may undergo sarcomatous change (Fig. 18.25). This is also true in patients with Maffucci syndrome (Fig. 18.26).

**Osteochondroma**

Also known as *osteoarticular cartilage*, this lesion is characterized by a cartilage-capped bony projection on the external surface of a bone. It is the most common benign bone lesion, constituting approximately 20% to 50% of all benign bone tumors, and is usually diagnosed in patients before their third decade. It has been postulated that sporadic osteochondromas represent developmental abnormality; however, recent cytogenetic studies revealed mutations in the EXT gene encoding exostosin 1, suggesting their neoplastic nature. Apparently, these genetic mutations lead to abnormal processing and accumulation of heparan sulphate proteoglycans (HSPG) in the cytoplasm of the chondrocytes. This leads to a loss of polar organization of the growth plate allowing chondrocytes to grow in the wrong direction. Continued growth of these chondrocytes coupled with endochondral ossification result in the formation of outpouching of both medullary and cortical bone covered by the cartilaginous cap, thus forming an exostosis. Osteochondroma, which has its own growth plate, usually stops growing at skeletal maturity. The most common sites of involvement are the metaphyses of the long bones, particularly in the region around the knee and the proximal humerus (Fig. 18.27). Variants of osteochondroma include subungual exostosis, turrett exostosis, traction exostosis, bizarre parosteal osteochondromatous proliferation (BPOP), florid reactive periostitis, and dysplasia epiphysealis hemimelica (also called intraarticular osteochondroma or Trevor-Fairbank disease).

The radiographic presentation of osteochondroma is characteristic according to whether the lesion is pedunculated, with a slender pedicle usually directed away from the neighboring growth plate (Fig. 18.28A), or sessile, with a broad base attached to the cortex (Figs. 18.28B,C). The most important characteristic feature of either type of lesion is uninterrupted merging of the cortex of the host bone with the cortex of the osteochondroma; additionally, the medullary portion of the lesion and the medullary cavity of the adjacent bone communicate. CT scanning can establish unequivocally the lack of cortical interruption and the continuity of cancellous portions of the lesion and the host bone (Fig. 18.29). These are important features that distinguish this lesion from the occasionally similar looking bone masses of osteoma, periosteal chondroma, BPOP, juxtacortical osteosarcoma, soft-tissue osteosarcoma, and juxtacortical myositis ossificans (Fig. 18.30). The other characteristic feature of osteochondroma involves calcifications in the chondro-osseous portion of the stalk of the lesion (see Fig. 18.28) and cartilaginous cap. The thickness of the cartilaginous cap ranges from 1 to 3 mm and rarely exceeds 1 cm. On MRI, the cartilaginous cap shows high signal intensity on T2-weighted and gradient-echo sequences. A narrow band of low signal intensity surrounding the cap represents the overlying periosteum (Fig. 18.31).

Histologically, the osteochondroma cap is composed of hyaline cartilage arranged similarly to that of a growth plate. A zone of calcification in the chondro-osseous portion of the stalk corresponds to the zone of provisional calcification in the physis. Beneath this zone, there is vascular invasion and replacement of the calcified cartilage by new bone formation, which undergoes maturation and merges with the cancellous bone of the host bone’s medullary cavity.

**Complications**

Osteochondroma may be complicated by a number of secondary abnormalities, including pressure on nerves or blood vessels (Fig. 18.32), pressure on the adjacent bone (Fig.18.33; see also Fig. 16.66), with occasional fracture (Fig. 18.34), fracture through the lesion itself, and inflammatory changes of the bursa exostotica (“exostosis bursata”) covering the cartilaginous cap (Fig. 18.35).

The least common complication of osteochondroma, seen in solitary lesions in less than 1% of cases, is malignant transformation to chondro-
FIGURE 18.22  Ollier disease. The classic features of Ollier disease in a 17-year-old boy are exhibited in extensive involvement of multiple bones. (A) Anteroposterior radiograph of the pelvis demonstrates crescent-shaped and ring-like calcifications in tongues of cartilage extending from the iliac crests and proximal femora. (B) A radiograph of both legs shows growth stunting and deformities of the tibia and fibula. (C) In another patient, a 6-year-old boy, note extensive involvement of the tibia and distal femur.
FIGURE 18.23 MRI of Ollier disease. (A) Anteroposterior radiograph of the right humerus of a 23-year-old woman shows numerous enchondromas affecting proximal half of the bone. Observe also the lesions within the scapula. (B) Coronal T1-weighted MRI shows heterogeneous signal intensity of the lesions. (C) Coronal T1-weighted fat-suppressed MR image obtained after intravenous administration of gadolinium demonstrates strong peripheral enhancement of the lesions. (D) Coronal T2-weighted fat-saturated MRI of the distal femur of another patient demonstrates linear columns of cartilage in the distal metaphysis of the femur (arrowheads) and more globular cartilage tumors (arrows). Note the involvement of the epiphysis.
Figure 18.24  Maffucci syndrome. Radiograph of the hand reveals typical changes of enchondromatosis, accompanied by calcified phleboliths in soft-tissue hemangiomas. (From Bullough PG. Atlas of orthopedic pathology, 2nd ed. New York: Gower; 1992:14:9.)

Figure 18.25  Chondrosarcoma in Ollier disease. In this case of sarcomatous transformation of enchondroma in the hand in a patient with Ollier disease, note the large, lobulated masses of cartilage in all fingers. The lesion of the middle phalanx of the ring finger shows destruction of the cortex and extension into the soft tissues.

Figure 18.26  Chondrosarcoma in Maffucci syndrome. A 26-year-old woman, known to have Maffucci syndrome for several years, presented with slowly enlarging mass in the ring finger of her right hand (arrows). Excision biopsy revealed a chondrosarcoma.
Osteochondroma

**FIGURE 18.27** Skeletal sites of predilection, peak age range, and male-to-female ratio in osteochondroma (osteocartilaginous exostosis).

Sarcoma. Nevertheless, it is important to recognize this complication at an early stage. The chief clinical features suggesting malignant transformation are pain (in the absence of a fracture, bursitis, or pressure on nearby nerves) and a growth spurt or continued growth of the lesion beyond the age of skeletal maturity. Certain imaging features have also been identified that may help in the determination of malignancy (Table 18.1).

The most reliable imaging modalities for evaluating the possible malignant transformation of an osteochondroma are conventional radiography, CT, and MRI; the results of a radionuclide bone scan, which may show increased uptake of radiopharmaceutical at the site of the lesion, may not be reliable. The radiography usually demonstrates whether the calcifications in an osteochondroma are contained within the stalk of the lesion—a clear indication of benignity (see Fig. 18.28). Similarly, CT can demonstrate both dispersed calcifications in the cartilaginous cap and increased thickness of the cap, cardinal signs of malignant transformation of the lesion, as Norman and Sissons have pointed out (Fig. 18.36).

The unreliability of radionuclide imaging is related to the fact that even benign exostoses exhibit an increased uptake of radiopharmaceutical caused by endochondral ossification. Exostotic chondrosarcoma is also marked by isotope uptake, which is related to active ossification, osteoblastic activity, and hyperemia within the cartilage and bony stalk of the tumor. Thus, although the uptake is more intense in exostotic chondrosarcomas than in benign exostoses, various investigations show that this is not always a reliable feature distinguishing these lesions.

**Treatment**

Solitary lesions of osteochondroma usually can simply be monitored if they do not cause clinical problems. Surgical resection is indicated if the lesion becomes painful, if there is suspected encroachment on adjacent nerves or blood vessels, if pathologic fracture occurs, or if there is concern about the diagnosis.

**Multiple Osteocartilaginous Exostoses**

This condition, also known as multiple hereditary osteochondromata, familial osteochondromatosis, or diaphyseal aclasis, is classified by some authorities in the category of bone dysplasias. It is a hereditary, autosomal dominant disorder with incomplete penetrance in females. Approximately two thirds
**FIGURE 18.28** Osteochondroma. (A) The typical pedunculated type of osteochondroma is seen arising near the proximal growth plate of the right humerus in a 13-year-old boy. (B) In the typical sessile or broad-based variant, seen here arising from the medial cortex of the proximal diaphysis of the right humerus in a 14-year-old boy, the cortex of the host bone merges without interruption with the cortex of the lesion. The cartilaginous cap is not visible on the conventional radiographs, but dense calcifications in the stalk can be seen. (C) In another patient, a 28-year-old man, a sessile osteochondroma of the distal femur exhibits no visible calcifications.

**FIGURE 18.29** CT of osteochondroma. (A) Lateral radiograph of the knee shows a calcified lesion at the posterior aspect of the proximal tibia (arrows). The exact nature of this lesion cannot be ascertained. (B) CT clearly establishes the continuity of the cortex, which extends without interruption from the osteochondroma into the tibia. Note also that the medullary portion of the lesion and the tibia communicate.
FIGURE 18.30  Differential diagnosis of osteochondroma. Radiographic features characterizing lesions similar in appearance to osteochondroma.

FIGURE 18.31  MRI of osteochondroma. (A) Radiograph of the right proximal humerus shows a sessile osteochondroma at the medial aspect of metadiaphysis. (B) T1-weighted coronal MRI reveals that the lesion exhibits low signal intensity because of extensive mineralization. (C) T2-weighted image shows the thin cartilaginous cap as a band of high signal intensity (arrows), covered by a linear area of low signal representing perichondrium (open arrow).
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FIGURE 18.32  Complication of osteochondroma. A 14-year-old boy with a known osteochondroma of the right humerus complained of pain and numbness of the hand and fingers. (A) Radiograph of the right shoulder demonstrates a sessile-type osteochondroma arising from the medial aspect of the proximal diaphysis of the humerus. (B) Arteriography reveals compression and displacement of the brachial artery.

FIGURE 18.33  Complication of osteochondroma. (A) Sessile lesion of the distal tibia has caused erosion of the medial aspect of the fibula. (B) Continued growth of the sessile osteochondroma of the proximal ulna resulted in pressure erosion of the head and neck of the radius. (C) Pedunculated osteochondroma of the distal ulna has eroded medial aspect of the shaft of the radius.
**FIGURE 18.35 Bursa exostotica.** (A) A 25-year-old man with a known solitary osteochondroma of the distal right femur reported gradually increasing pain. The capillary phase of an arteriogram reveals a huge bursa exostotica. Inflammation of the bursa, with the accumulation of a large amount of fluid (bursitis), was the cause of the patient's symptoms. (B) Another patient, a 12-year-old girl, presented with pain in the popliteal fossa. Coronal T1-weighted MR image (SE; TR 650/TE 25 msec) demonstrates a large osteochondroma arising from the posterolateral aspect of distal femur (arrows). (C) Axial T2-weighted image (SE; TR 2200/TE 70 msec) demonstrates a bursa exostotica distended with fluid.

**FIGURE 18.34 Complication of osteochondroma.** A 9-year-old boy had a sessile osteochondroma of the distal tibia. The lesion produced pressure erosion, and later bowing and attenuation of the fibula, with subsequent fracture of the bone.
of affected individuals have a positive family history. The specific genetic defect has been recently identified, a novel mutation in genes \( \text{EXT1} \) that maps to chromosome 8q24.1, \( \text{EXT2} \) that maps to chromosome 11p13, and \( \text{EXT3} \) that maps to the short arm of chromosome 19. There is a decided 2:1 male predilection. The knees, ankles, and shoulders are the sites most frequently affected by the development of multiple osteochondromas (Fig. 18.37). The radiographic features are similar to those of single osteochondromas (see Fig. 18.28), but the lesions are more frequently of the sessile type (Figs. 18.38 to 18.40). CT and 3D CT show spatial distribution of the lesions (Fig. 18.41). The histopathologic features of multiple osteochondromas are the same as those of solitary lesions.

Two syndromes associated with multiple osteochondromas have been identified: \textit{Langer-Giedion syndrome} and \textit{Potocki-Shaffer syndrome}. The first one, also known as \textit{trichorhinophalangeal syndrome type II (TRPS2)} or \textit{Langer-Giedion chromosome region (LGCR)} is an autosomal dominant genetic disorder caused by deletion of gene \( \text{EXT2} \) and probably \( \text{ALX4} \). Recent investigations points to loss of functional copies of the trichorhinophalangeal syndrome type I (\( \text{TRPS1} \)) gene encoding a zinc-finger protein, and \( \text{EXT7} \) gene at 8q23.2-q24.1 chromosome. Clinically, it characterizes by short stature, joint laxity, short fingers, microcephaly, craniofacial dysmorphism, mental retardation, and multiple osteochondromas. Potocki-Shaffer syndrome is caused by deletion of 11p11.2-p12 chromosome, and clinically, it manifests by enlarged parietal foramina, multiple osteochondromas, and sometimes craniofacial dysostosis and mental retardation.

**Complications**

There is a greater incidence of growth disturbance in multiple osteocartilaginous exostoses than in solitary osteochondroma. Growth abnormalities are primarily seen in the forearms (Fig. 18.42; see also Fig. 16.64) and legs. Malignant transformation to chondrosarcoma is also more common, seen in 5% to 15% of cases, with lesions at the shoulder girdle and around the pelvis at greater risk of undergoing transformation. The clinical and imaging signs of this complication are identical to those in the malignant transformation of a solitary osteochondroma (Fig. 18.43; see also Fig. 18.36 and Table 18.1).

**Treatment**

Multiple osteochondromas are treated individually. Like solitary lesions, they are likely to recur in younger children, and surgery may be deferred to a later date.

**Bizarre Parosteal Osteochondromatous Proliferation**

Also known as \textit{Nora lesion}, named for a pathologist F. E. Nora from Mayo Clinic, who first described this benign surface lesion in 1983, BPOP

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### TABLE 18.1

**Clinical and Imaging Features Suggesting Malignant Transformation of Osteochondroma**

<table>
<thead>
<tr>
<th>Clinical Features</th>
<th>Radiologic Findings</th>
<th>Imaging Modality</th>
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<tbody>
<tr>
<td>Pain (in the absence of fracture, bursitis, or pressure</td>
<td>Enlargement of the lesion</td>
<td>Conventional radiography (comparison</td>
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<td>on nearby nerves)</td>
<td>Development of a bulky cartilaginous cap usually</td>
<td>with earlier radiographs)</td>
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<td>Growth spurt (after skeletal maturity)</td>
<td>2–3 cm thick</td>
<td>CT, MRI</td>
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<td>Dispersed calcifications in the cartilaginous</td>
<td>Radiography, CT, MRI</td>
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<td></td>
<td>cap</td>
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<td></td>
<td>Development of a soft-tissue mass with or without</td>
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<tr>
<td></td>
<td>calcifications</td>
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<tr>
<td></td>
<td>Increased uptake of isotope after closure of growth</td>
<td>Scintigraphy</td>
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<td>plate (not always reliable)</td>
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CT, computed tomography; MRI, magnetic resonance imaging.

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**FIGURE 18.36** Transformation of osteochondroma to chondrosarcoma. A 28-year-old man had pain in the popliteal region and also noticed an increase in a mass he had been aware of for 15 years—important clinical information that warranted further investigation to rule out the malignant transformation of an osteochondroma. (A) Lateral radiograph of the knee demonstrates a sessile-type osteochondroma arising from the posterior cortex of the distal femur. Note that calcifications are present not only in the stalk of the lesion but also are dispersed in the cartilaginous cap (arrows). (B) An arteriogram demonstrates displacement of the small vessels, which are draped over the invisible cartilaginous cap. (C) CT section confirms the increased thickness of the cartilaginous cap (2.5 cm) and dispersed calcifications within the cap (arrows). These imaging features are consistent with a diagnosis of malignant transformation to chondrosarcoma, which was confirmed by histopathologic examination.
**FIGURE 18.37** Skeletal sites of predilection, peak age range, and male-to-female ratio in multiple osteocartilaginous exostoses (multiple osteochondromata, diaphyseal aclasis).

**FIGURE 18.38** Hereditary multiple exostoses. (A) Anteroposterior radiograph of the shoulder of a 22-year-old man demonstrates multiple sessile lesions involving the proximal humerus, scapula, and ribs. (B) Involvement of the distal femur and proximal tibia is characteristic of this disorder.
**FIGURE 18.39**  
Hereditary multiple exostoses. An anteroposterior radiograph of both knees of a 17-year-old boy shows numerous sessile and pedunculated osteochondromas.

**FIGURE 18.40**  
MRI of hereditary multiple exostoses. (A) Anteroposterior radiograph of the hips shows multiple sessile osteochondromas mainly affecting proximal femora. Some lesions are also present at the pubic bones. Coronal (B) and axial (C) T1-weighted (SE; TR 600/TE 20 msec) MR images demonstrate continuity of the lesions with the medullary portion of the femora. Note also dysplastic changes expressed by abnormal tubulation of the bones.
commonly affects the metacarpals and phalanges of the hand. The long bones are involved in about 25% of reported cases. The lesion is seen in the third and fourth decades, with equal frequency in men and women. Patients typically present with a firm, slow-growing, nontender mass. The cause is unknown, but it may be related to trauma, although recently reported by Zambrano and associates, the cytogenetic changes put in question the lesion’s nonneoplastic nature. Imaging studies commonly show a mushroom-like–shaped osseous or cartilaginous mass attached to the cortex (Fig. 18.44). The contour of the mass is usually smooth but may be slightly lobulated. The absence of continuity between the lesion and medullary cavity of the adjacent bone differentiates this lesion from osteochondroma. The other similarly appearing lesions to consider in the differential diagnosis are juxtacortical myositis ossificans, periosteal chondroma, turret exostosis, florid reactive periostitis, and parosteal or periosteal osteosarcoma. The characteristic histologic feature of BPOP is the presence of irregular calcified matrix stained blue on hematoxylin-eosin staining referred to as blue bone. There is lack of cellular atypia of osteoblasts or fibrous tissue, and the bone is lamellar and well organized, features that distinguish this lesion from osteosarcoma. The treatment of BPOP is surgical excision; however, recurrence rate is high.

Chondroblastoma
Also known as a Codman tumor, chondroblastoma, representing fewer than 1% of all primary bone tumors, is a benign lesion occurring before skeletal maturity, characteristically presenting in the epiphyses of long bones such as the humerus, tibia, and femur (Fig. 18.45). Although secondary involvement of the metaphysis after skeletal maturity is recognized, a predominantly metaphyseal or diaphyseal location is exceedingly rare. Equally unusual is involvement of the vertebra or intracortical location in the long bones. Occasionally, the patella, which is considered equivalent to an epiphysis, is affected. Ten percent of chondroblastomas involve the small bones of the hands and feet, with the talus and calcaneus representing the most common sites. Although the lesion is usually seen in growing bones, some cases have been reported after obliteration of the growth plate. Chondroblastoma is usually located eccentrically, shows a sclerotic border, and often demonstrates scattered calcifications of the matrix (25% of cases) (Fig. 18.46). Brower and colleagues noticed a distinctively thick, solid periosteal reaction distal to the lesion in 57% of chondroblastomas in long bones (Fig. 18.47). This most likely represents an inflammatory reaction to the tumor. In most cases, radiography suffices to demonstrate the lesion (Fig. 18.48), but CT scan can help demonstrate the calcifications if they are not visible on the standard radiographs (Fig. 18.49). MRI usually reveals a larger area of involvement than can be seen on radiographs, including regional bone marrow and soft-tissue edema (Fig. 18.50).

Histologically, chondroblastoma is composed of nodules of fairly mature cartilage matrix surrounded by a highly cellular tissue containing uniformly large round cells with ovoid nuclei and clear cytoplasm. Multinucleated osteoclast-like giant cells are a common finding. The matrix shows characteristic lattice-like fine calcifications surrounding apposing
chondroblasts, having a spatial arrangement resembling the hexagonal configuration of chicken wire.

Clonal abnormalities in chondroblastoma have been reported, including recurrent structural alterations in chromosomes 5 and 8 with rearrangements of band 8q21, and recurrent breakpoints at 2q35, 3q21-q23, and 18q21.

**Treatment and Complications**

Chondroblastomas are usually treated by curettage and bone grafting. Only few reported cases have been treated with percutaneous radiofrequency ablation.

In rare cases, pulmonary metastases develop in the absence of any histologic evidence of malignancy in either the primary bone tumor or the pulmonary lesions. Only in exceptional circumstances, pulmonary or widespread metastases led to patient death.

**Chondromyxoid Fibroma**

Chondromyxoid fibroma is a rare tumor of cartilaginous derivation, characterized by the production of chondroid, fibrous, and myxoid tissues in variable proportions, and accounts for 0.5% of all primary bone tumors and 2% of all benign bone tumors. It occurs predominantly in adolescents and young adults (males more than females), most commonly in the patient’s second or third decade. It has a predilection for the bones of the lower extremities, with preferred sites in the proximal tibia (32%) and distal femur (17%) (Fig. 18.51). Exceedingly rare, the lesion may be located in the vertebra. Few cases of chondromyxoid fibroma have been reported in juxtacortical location. Its clinical symptoms include local swelling and pain, which are occasionally caused by pressure on adjacent neurovascular structures by a peripherally located mass.

Its characteristic radiographic picture is that of an eccentrically located radiolucent lesion in the bone, with a sclerotic scalloped margin often eroding or ballooning out the cortex (Figs. 18.52 and 18.53). The lesion may range from 1 to 10 cm in size, with an average of 3 to 4 cm. Calcifications are not apparent radiographically, but focal microscopic calcifications have been reported in as many as 27% of cases. Frequently, a buttress of periosteal new bone can be observed. MRI reveals characteristics of most cartilaginous tumors: intermediate-to-low signal intensity on T1-weighted and high signal on T2-weighted sequences (Fig. 18.54).

Pathologically, the most important feature of the lesion is its lobular or pseudolobular arrangement into zones of varying cellularity. The center of the lobule is hypocellular. Within the matrix, loosely arranged spindle-shaped and stellate cells with elongated processes are present. The periphery of the lobule is densely cellular, containing a mixture of mononuclear spindle-shaped and polyhedral stromal cells with a variable number of multinucleated giant cells.

Recently, a pericentric inversion of chromosome 6 [inv(6)(p25q13)] has been proposed as a specific genetic marker for chondromyxoid fibroma, and some other studies disclosed a breakpoint on a long arm (q25) of this chromosome. In addition, the clonal translocation t(1;5)(p13;p13) was suggested as a novel sole clone abnormality in this tumor.
FIGURE 18.43  Malignant transformation. (A) Oblique radiograph of the right hand of a 22-year-old man shows multiple osteochondromas. A large soft-tissue mass situated between the index finger and thumb and containing chondroid calcifications indicates malignant transformation to chondrosarcoma. (B) Sagittal T1-weighted (SE; TR 600/TE 16 msec) MRI reveals volar extension of a large soft-tissue tumor. (C) Coronal inversion recovery (fast multiplanar inversion recovery [FMPIR]/90; TR 4000/TE 64 msec/EF) MR image shows malignant lobules of the cartilage invading the bones and soft tissues of the hand. (D) Axial T2-weighted fat-saturated MRI in another patient with a large osteochondroma of the pelvis that underwent malignant transformation. Observe the thin, hyperintense cartilage cap of the anterior aspect of the lesion (arrowhead), in comparison to the thick cartilage cap of the posterior aspect undergoing malignant transformation to chondrosarcoma (arrows). Biopsy of the posterior cartilage cap demonstrated malignant chondrocytes.

FIGURE 18.44  Bizarre parosteal osteochondromatous proliferation. Anteroposterior (A) and lateral (B) radiographs of the small finger of an 8-year-old boy show an osseous mass adjacent to the posteromedial cortex of the proximal phalanx. The lesion was excised and histopathologic examination showed typical changes of Nora lesion. (From Greenspan A, Jundt G, Remagen W. Differential diagnosis in orthopaedic pathology, 2nd ed. Philadelphia: Lippincott Williams & Wilkins; 2007, Fig. 2-131A,B, p. 140.)
FIGURE 18.45  Skeletal sites of predilection, peak age incidence, and male-to-female ratio in chondroblastoma.

FIGURE 18.46  Chondroblastoma. A lesion located in the proximal tibia (arrows) of a 17-year-old boy exhibits faint sclerotic border and central calcifications.
**FIGURE 18.47** Chondroblastoma. A lesion in the proximal humerus (arrows) elicited periosteal reaction along the lateral cortex (open arrow).

**FIGURE 18.48** Chondroblastoma. (A) Lateral radiograph and (B) anteroposterior tomogram of the knee show the typical appearance of a chondroblastoma in the proximal epiphysis of the tibia. Note the radiolucent, eccentrically located lesion with a thin, sclerotic margin (arrows). There are small, scattered calcifications in the center of the lesion, which are better seen on the tomogram.
FIGURE 18.49 CT and MRI of chondroblastoma. (A) Anteroposterior radiograph of the right shoulder of a 16-year-old boy shows a lesion in the proximal humeral epiphysis, but calcifications are not well demonstrated. Note the well-organized layer of periosteal reaction at the lateral cortex (arrow). (B) CT section shows the calcifications clearly. The tumor was removed by curettage, and a histopathologic examination confirmed the radiographic diagnosis of chondroblastoma. (C) Anteroposterior radiograph of the shoulder in another patient demonstrates a well-demarcated lesion in the epiphysis of the humerus with a sclerotic rim (arrow) and internal calcifications. (D) Axial CT image shows the sclerotic rim (arrow) and chondral calcifications inside the lesion. (E) Axial T2-weighted MRI demonstrates the tumor (arrow) containing low-signal intensity calcified chondral matrix.
Chondromyxoid Fibroma

**FIGURE 18.50** MRI of chondroblastoma. (A) Axial T2-weighted (SE; TR 2000/TE 80 msec) MRI of the shoulder in an 18-year-old man shows sharply marginated lesion with sclerotic border and central calcifications within the left humeral head. Note the small amount of joint effusion and peritumoral edema. In another patient, (B) sagittal proton density-weighted (SE; TR 2000/TE 28 msec) and (C) axial T2-weighted (SE; TR 2000/TE 80 msec) MR images of the knee show extension of chondroblastoma located in the posterior aspect of proximal tibia into the soft tissues.

**FIGURE 18.51** Skeletal sites of predilection, peak age range, and male-to-female ratio in chondromyxoid fibroma.
**FIGURE 18.52** Chondromyxoid fibroma. Anteroposterior (A) and lateral (B) radiographs of the left leg of an 8-year-old girl demonstrate a radiolucent lesion extending from the metaphysis into the diaphysis of the tibia, with a geographic type of bone destruction and a sclerotic scalloped border.

**FIGURE 18.53** Chondromyxoid fibroma. Anteroposterior (A) and lateral (B) radiographs of the left knee of a 12-year-old girl show a radiolucent, slightly lobulated lesion with a thin sclerotic margin in the proximal tibial diaphysis. Note the lack of visible calcifications.
FIGURE 18.54 MRI of chondromyxoid fibroma. (A) Sagittal T1-weighted (SE; TR 600/TE 19 msec) MRI in a 10-year-old girl shows a well-demarcated lesion in the plan- tar aspect of the calcaneus, displaying low signal intensity. (B) An axial T1-weighted (SE; TR 600/TE 17 msec) image shows significant amount of peritumoral edema. (C) Sagittal T2-weighted (SE; TR 2000/TE 80 msec) MRI shows the lesion displaying high signal intensity. A sclerotic border is imaged as a rim of low signal intensity.

FIGURE 18.55 Chondromyxoid fibroma resembling an aneurysmal bone cyst. (A) Anteroposterior radiograph of the knee of an 18-year-old woman shows a lesion in the lateral aspect of the proximal tibia. The tumor balloons out from the cortex and is supported by a solid periosteal buttress resembling that seen in an aneurysmal bone cyst. The periosteal buttress (arrows) is better appreciated on a tomographic cut (B).
Differential Diagnosis

Commonly, one can observe a characteristic buttress of periosteal new bone formation (Fig. 18.55), in which case a chondromyxoid fibroma may be radiographically indistinguishable from an aneurysmal bone cyst. In unusual locations such as in short tubular or flat bones, it may mimic a giant cell tumor or desmoplastic fibroma.

Treatment

The treatment of this lesion usually consists of curettage and a bone graft. Recurrences are frequent, with the reported rate between 20% and 80% (see Fig. 16.59).

PRACTICAL POINTS TO REMEMBER

[1] Enchondroma is characterized by the formation of mature hyaline cartilage and is seen:

• most commonly in the short tubular bones of the hand, where the lesion is usually radiolucent
• in the long bones, where scattered calcifications may be seen, resembling a medullary bone infarct.

[2] The characteristic radiographic features of enchondroma include:

• popcorn-like, annular, or punctate calcifications
• a lobulated growth pattern with frequent shallow scalloping of the endosteal cortex.

[3] Important clinical and radiographic features of the malignant transformation of an enchondroma include:

• the development of pain, in the absence of a fracture, in a previously asymptomatic lesion
• thickening or destruction of the cortex
• development of a soft-tissue mass.

[4] Enchondromatosis is a condition marked by multiple enchondromas, commonly in metaphysis and diaphysis. If skeleton is extensively affected and the lesions are distributed unilaterally, the term Ollier disease is applied.

[5] Ollier disease and Maffucci syndrome (an association of Ollier disease with soft-tissue hemangiomas) both carry an increased risk for malignant transformation to chondrosarcoma.

[6] In the radiographic evaluation of osteochondroma, the most common benign bone lesion, note that:

• it can be seen as enchondral or sessile (broad-based) variants
• its two important radiographic features are uninterrupted merging of the lesion's cortex with the host bone cortex and continuity of the cancellous portion of the lesion with the medullary cavity of the host bone.

[7] The most important differential diagnoses in suspected osteochondroma include:

• juxtacortical osteoma
• juxtacortical osteosarcoma
• soft-tissue osteosarcoma
• juxtacortical myositis ossificans.

[8] Osteochondroma may be complicated by:

• pressure on adjacent nerves or blood vessels
• pressure on the adjacent bone, frequently leading to fracture
• bursitis exostosis
• malignant transformation to chondrosarcoma.

[9] In the malignant transformation of osteochondroma, imaging signs include:

• enlargement of the lesion
• marked thickening of the cartilaginous cap of the lesion
• dispersion of calcifications into the cartilaginous cap
• development of a soft-tissue mass
• increased isotope uptake by the lesion after skeletal maturity.

[10] Variants of osteochondroma include subungual exostosis, turrett exostosis, traction exostosis, BPP0, florid reactive periostitis, and dysplasia epiphysealis hemimelica (Trevor-Fairbank disease).

[11] Multiple osteocartilaginous exostoses, a familial hereditary condition, carry the increased risk of malignant transformation of an osteochondroma to a chondrosarcoma, particularly in the shoulder girdle and pelvis.

[12] Chondroblastoma is characterized radiographically by:

• its eccentric epiphyseal location
• sclerotic margin
• scattered calcifications
• periosteal reaction (>50% cases).

[13] Chondromyxoid fibroma is characterized radiographically by:

• its location close to the growth plate
• its scalloped, sclerotic border
• a buttress of periosteal new bone
• lack of visible calcifications.

It may mimic an aneurysmal bone cyst.

CHAPTER 18 Benign Tumors and Tumor-like Lesions II

SUGGESTED READINGS


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