Chondromas

15

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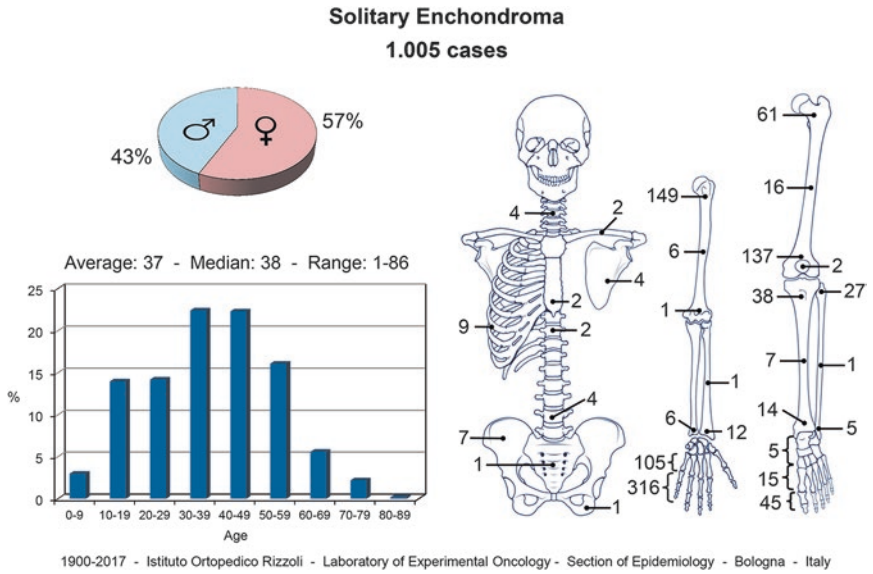
15.1 Solitary Chondroma (Enchondroma)

Definition: Intramedullary neoplasm made of well-differentiated hyaline cartilage.

Epidemiology: Relatively frequent. No predilection for either sex. May be diagnosed at any age.



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Localization: Chondromas occur only in bones that undergo secondary ossification of cartilage during skeletal development. They are very frequently seen in the tubular bones of the hand, where they represent the most common bone tumor. The other chondromas are mainly distributed in the long bones, with preference for the femur. The real incidence of chondroma is unknown, as most of these lesions do not cause any symptoms.

Clinical: Most chondromas are incidental findings on imaging studies. Pain can be due to small cracks or pathological fractures that often occur after minor trauma of the hands. A mild expansion of the cortex can be seen or palpated in superficial bones, such as tubular bones of the hand or foot, ribs, fibula.

Imaging: The lesion is frequently central, sometimes eccentric or intracortical. On radiographs it is usually an osteolysis, with rounded, lobulated, well-defined edges with a thin rind of reactive sclerosis. Often the lesion contains granular, popcorn, ring-like opacities that represent calcification and ossification at the periphery of the cartilage lobules. Chondroma can reach considerable extension in major long bones but rarely exceeds 10 cm. In the long tubular bones, the cortex is intact and does not show erosions. In small bones (hand, ribs), the cortex can be thinned and the bone expanded creating a palpable deformity. CT scan demonstrates the lobular or multi-island nature of the lesion, its sharp limits, and its radio-densities, as well as status of the cortex. MRI at best defines longitudinal extension; the lesion has low signal in T1, high signal in T2, and calcifications are seen as black signal-voids in both T1 and T2. Isotope scan is hot in most lesions and regularly reveals asymptomatic chondromas when performed as a staging examination in case of malignancies.

Histopathology: Lesion consists of lobules of cartilage. The aspect is typical of hyaline cartilage. Calcified areas appear as white opaque granules. Reactive or enchondral ossifications manifest as white-yellow hard rings and streaks around and between the lobules. The limits of the lesion are often irregular, as lobules of cartilage push toward the cancellous bone and excavate little niches in the cortex, but always well defined. The chondrocytes are sparse, with small, round, dense nuclei, of relatively uniform size. Occasionally, isogenous groups of cells can be seen. Double-nucleated cells are present, usually rare, but can be moderate. This does not really distinguish well benign enchondroma from chondrosarcoma. While diagnosis of cartilage tumor is usually easy even on clinico-imaging findings alone, the real problem is to differentiate chondroma from grade 1 chondrosarcoma. Histologically, this distinction is difficult:

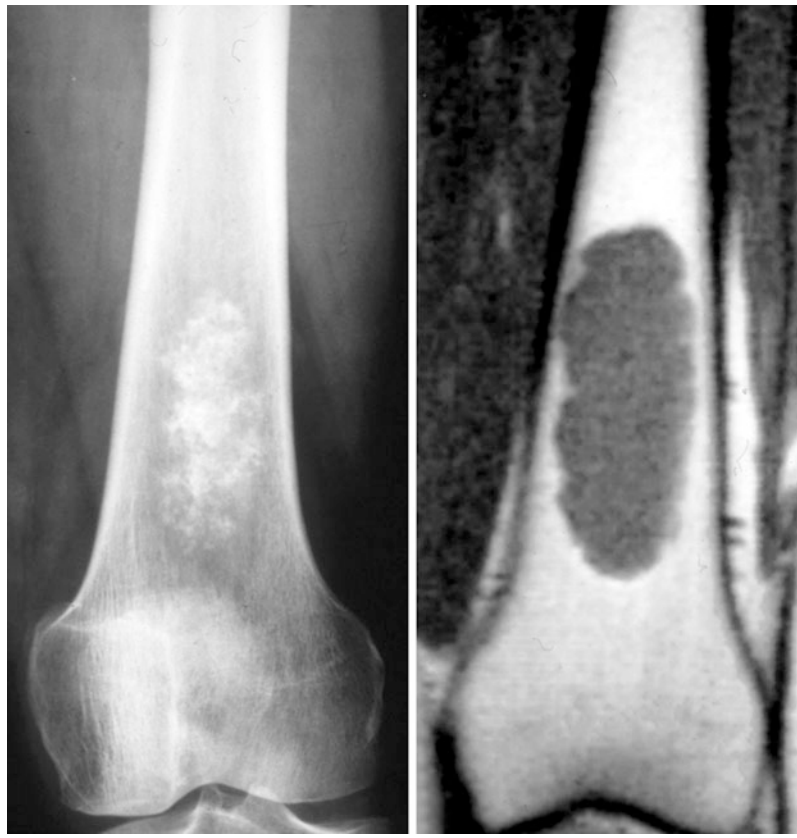
(a) Grade 1 chondrosarcoma has a higher cellularity, more plump nuclei, more than 4–5 double-nucleated cells per high-power field, but such features are subjective.
(b) Areas of chondroma and grade 1 chondrosarcoma may be found in the same tumor.
(c) The histological indicators of low-grade malignancy are meaningless if the lesion is in the hand, or in a child, or periosteal, or in chondromatosis. A useful differential element is represented by the relationship between tumor and host bone. Chondroma may present with cartilage islands scattered in the bone, usually encased by a shell of mature lamellar bone. Chondrosarcoma, on the contrary, permeates marrow spaces and haversian channels of the host in at least 90% of cases. The permeative pattern is 99% accurate, although cartilage permeation-like areas can be found in advanced osteoarthritis and in post fracture epiphyses.

Course and Staging: Until skeletal maturity, chondroma grows slowly, and then it tends to stop. Thus, chondroma is stage 2 in children and stage 1 in adults. The exact incidence of malignant transformation of a solitary chondroma is

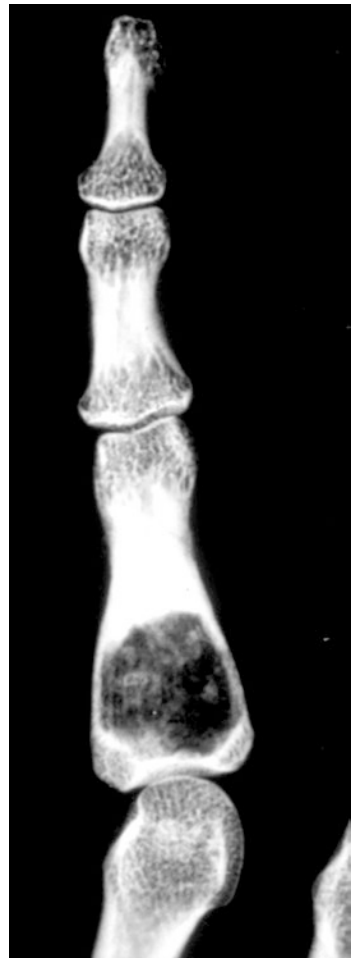
unknown and controversial. Transformation is very rare in the hands, probably less rare in the trunk and limb girdles.

Treatment and Prognosis: Diagnosis can usually be made on clinico-radiographic features. The majority of chondromas do not require biopsy or surgical treatment. Enchondromas of the hand are sometimes treated with curettage and bone grafting because of pain, pathologic fracture, or cosmesis. Rarely, biopsy may be indicated if the diagnosis of enchondroma is unclear and a chondrosarcoma is suspected. Follow-up with serial radiographs or MRI is helpful in the differential diagnosis vs. grade 1 chondrosarcoma. After the epiphyseal plate is closed, enchondromas show little to no growth, chondrosarcoma growth is slow but continuous, and is associated with increasing pain, also at rest.

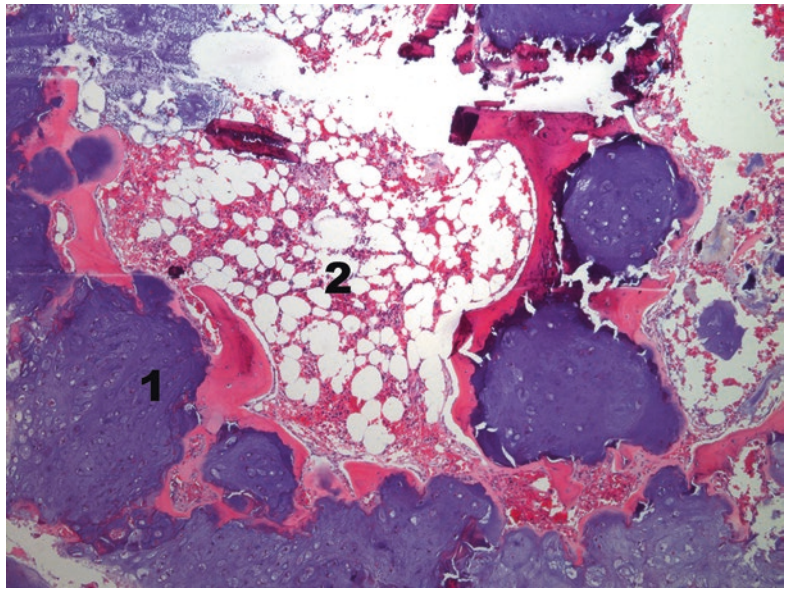
Key points	
• Clinical	Incidental findings
• Radiological	Central, lobulated, granular, and ring-like calcifications
• Histological	Lobules of benign cartilage ossified at the periphery without infiltration of the cancellous host bone
• Differential diagnosis	Low-grade central chondrosarcoma



AP radiograph and coronal T1 MR of the distal femur. Well-limited lesion, centered in the medullary cavity, with typical cartilaginous calcifications (round with a clear center). The cortex and soft tissues are not involved



Radiograph of a finger. Well-limited lesion, containing cartilaginous calcifications



(1) Cartilaginous lobules with mature matrix. (2) Normal fatty marrow in between the cartilaginous lobules

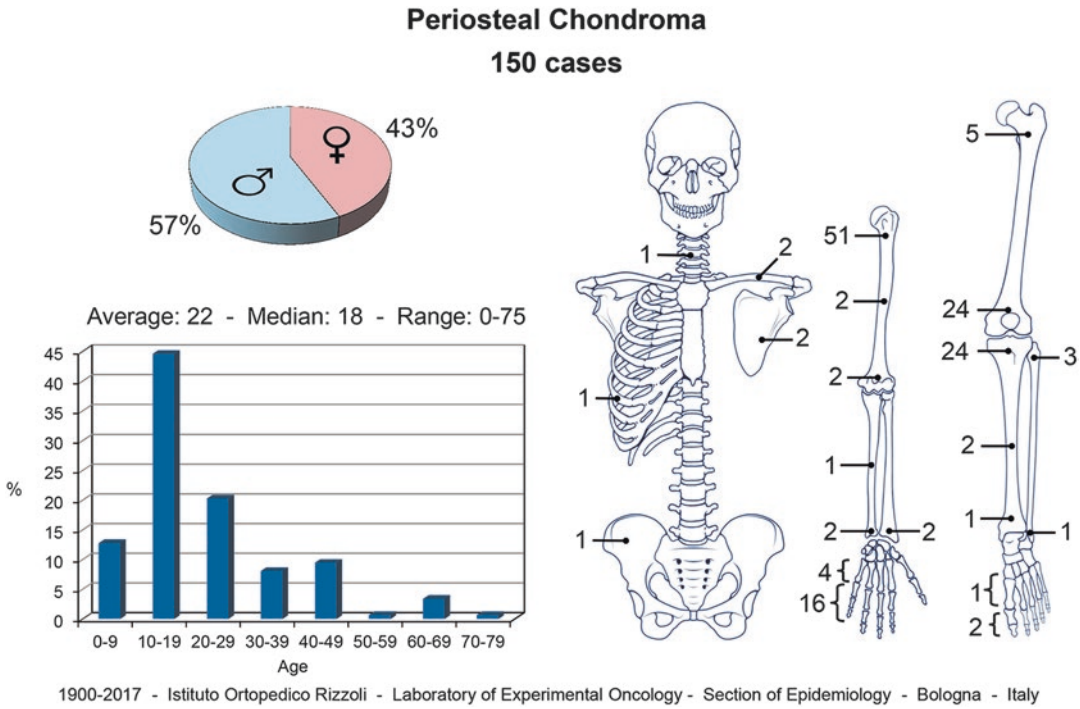
15.2 Periosteal Chondroma

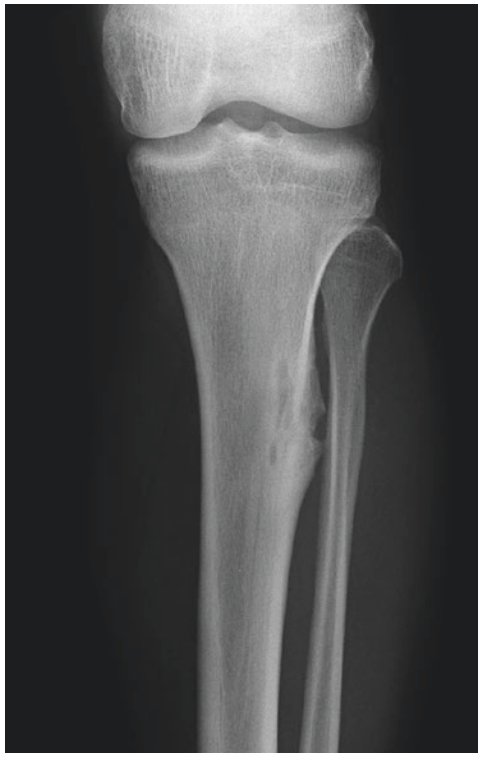
Definition: Benign cartilage neoplasm originating at the surface of the bone.

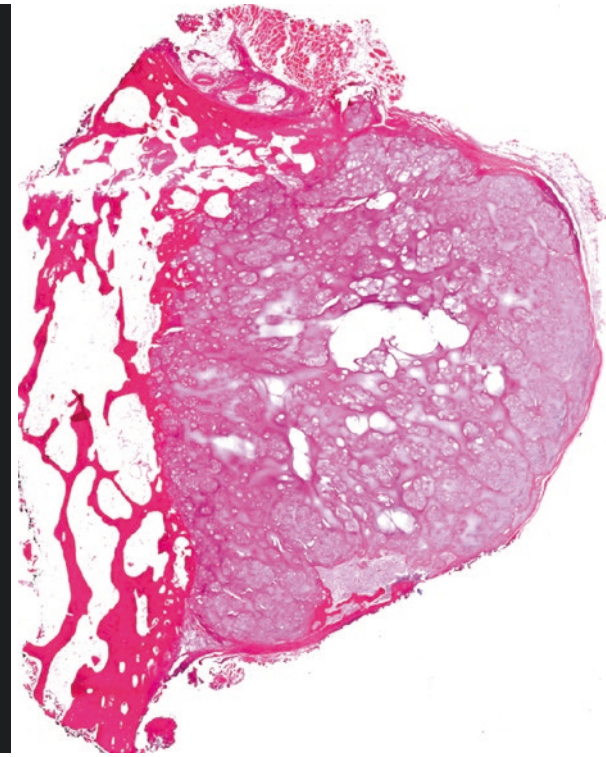
Rather rare, it is usually observed in children or young adults. It prefers the metaphysis of the long bones, particularly the proximal humerus. It is usually moderately painful because of nociception by the periosteum and typically presents as a hard, bony, swelling. Imaging shows a superficial erosion of the bone cortex, at times slightly scalloped, with regular borders. Such erosion is caused by a hemispherical periosteal cartilaginous mass, usually of small to moderate size (<3 cm). Granular or popcorn densities due to calcifications may be seen within the tumor.

Histologically the tumor is very similar to enchondroma, but it more frequently displays features of cell proliferation (high cellularity, nuclear plumpness, and frequent double nucleated cells). Being somewhat painful and causing some swelling in most instances, it usually requires surgical management consisting of either en bloc marginal excision or thorough curettage, equally effective.

Key points	
• Clinical	Some pain, young patients
• Radiological	Subperiosteal, metaphyseal, with erosion of the cortex, granular calcifications
• Histological	Lobules of benign cartilage. Possible hypercellularity
• Differential diagnosis	Periosteal chondrosarcoma



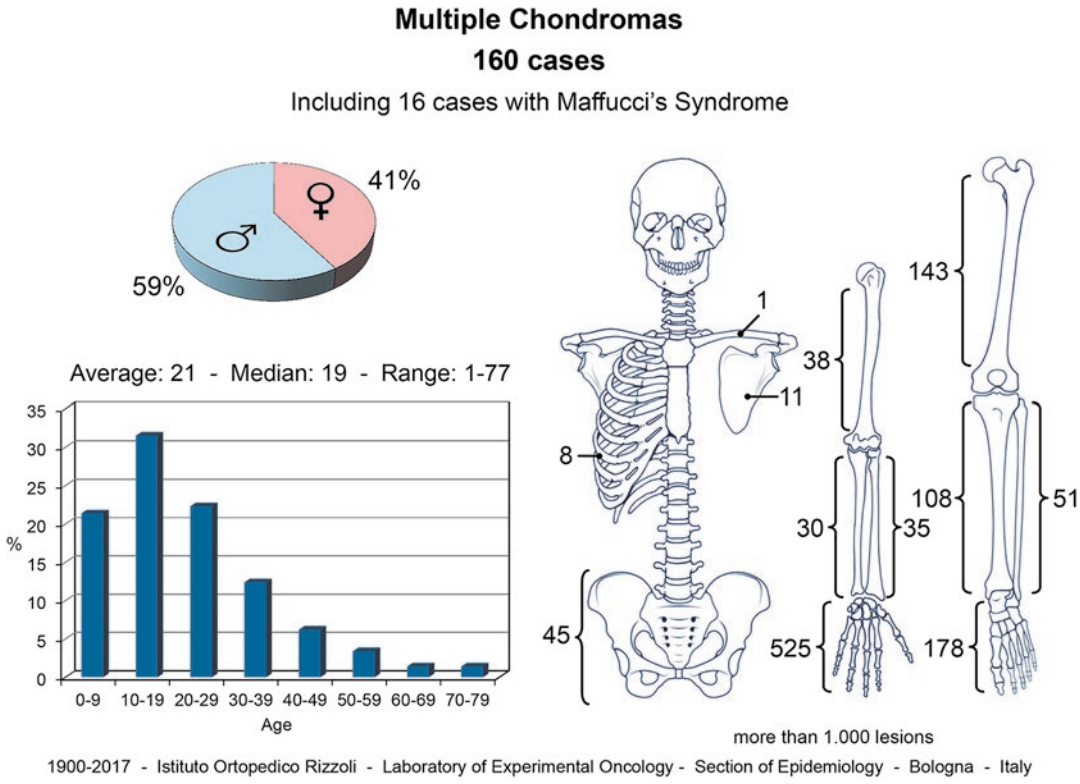




Male, 22 years old. Small (less than 3 cm) well-circumscribed lobulated lesion composed of hyaline cartilage. The lesion is beneath the periosteum with a sharp margin with the underlying cortex. The chondrocytes are frequently enlarged and hyperchromatic with increased cellularity and variability in nuclear size and shape. Chondrocytes are arranged in lobules. The lesion shows sharp borders, in particular without infiltration of the surrounding soft tissue (if present, this is consistent with periosteal chondrosarcoma)

15.3 Multiple Chondromas (Chondromatosis, Ollier's Disease) Associated Condition: Maffucci's Syndrome

Multiple chondromas are infrequent. The condition is not inherited and prevails in males**.** Their distribution and spectrum of presentation are extremely variable. It can present with only a few chondromas limited to the hands or one limb with minimal symptoms or show a hemisomic distribution. It can also be extended to the entire body with overall features of diffuse chondrodysplasia, known as Ollier's disease. The most affected bones are the small tubular bones of the hand and foot, but chondromas may present anywhere in the skeleton. Knobby swelling, bowing deformities, and lower limb length discrepancy (even >10 cm) are the dominant symptoms. Relationship between the polyostotic limited form and the fully expressed diffused chondrodysplasia (Ollier's disease) remains unknown.



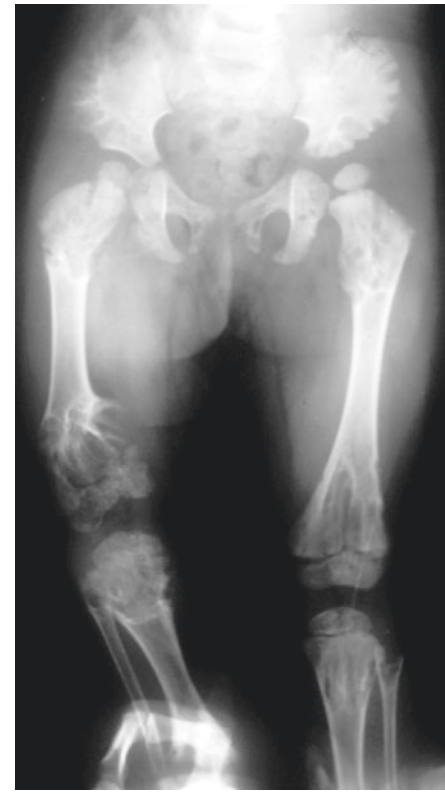
In Maffucci's syndrome (very rare), multiple, diffuse chondromas are associated with multiple hemangiomas. The hemangiomas can be either cutaneous, subcutaneous, or located in the deep soft tissues (not in the bone). Basic imaging is the same as described for solitary chondroma. In the metaphysis, longitudinal columns of radiolucency extend toward the diaphysis. They are divided by longitudinal bony septae. Chondromas can be very extensive, sometimes bubbly or trabeculated, with expansion of the bone, very thin cortex, or lack of any cortex. In Maffucci's syndrome, phleboliths reveal on X-rays the angiomas. MRI scans are useful to detect and follow up the lesions. Histologically, compared to solitary enchondroma, lesions in chondromatosis show features of more pronounced and persistent proliferative potential. Cartilage is more cellular, nuclei are sometimes hyperchromatic, and the histology overlaps with low-grade chondrosarcoma by cytology alone. The stage is 2 in children, more frequently 1 in adults. Transformation to secondary sarcoma, most commonly chondrosarcoma is frequent and probably ranges from 20% to 30% in Ollier and certainly higher in Maffucci (estimated >40%). Also in chondromatosis, malignant transformation to sarcoma is usually seen in adults but may occur even before age 20, especially in Maffucci.

Both Ollier and Maffucci are conditions at increased risk to develop extraskeletal malignancies, such as breast, liver, ovarian cancers and CNS tumors, suggesting an underlying genetic disorder predisposing to cancer in

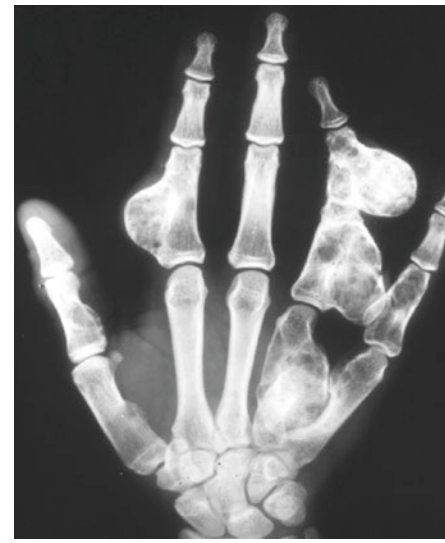
general. IDH1/IDH2 mutations are seen in approximately 90% of enchondromas in enchondromatosis (vs. 50% in solitary enchondromas). Surgical treatment is aimed to relieve symptoms, rather than excise chondromas. Skeletal deformities and limb length discrepancy are addressed by osteotomies and/or lengthening procedures. Prognosis is burdened by the incidence of malignant change.

15.4 Concept of "Active" Chondromas and Differential Diagnosis with Chondrosarcoma

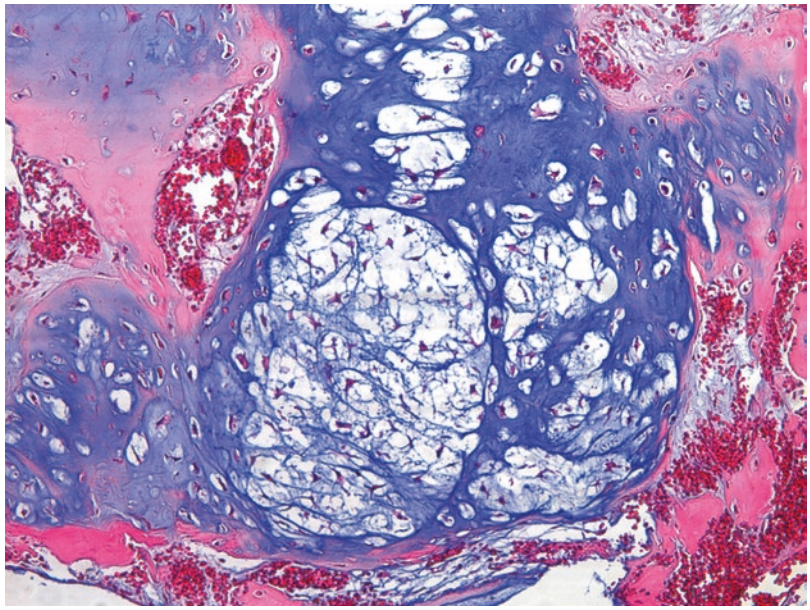
It is important to keep in mind that certain chondromas may show a histological pattern essentially similar to chondrosarcoma. In fact, multiple chondromas in Ollier and Maffucci, periosteal chondroma, enchondromas of the hands and feet, synovial chondromatosis, and soft tissue chondromas all show histologic features consistent with low-grade chondrosarcoma of bone. In other words, a grade 1 chondrosarcoma is cytologically indistinguishable from a benign lesion encountered in the abovementioned clinical settings. Therefore, diagnosis of malignant change is based on clinico-radiographic features and on the permeative growth pattern of tissue toward bone trabeculae. Secondarily, it is of paramount importance that the pathologist reviewing the slides has adequate clinical information, including site of biopsy and imaging studies, and discusses the case with the orthopedic surgeon before making the diagnosis of chondrosarcoma. Quite important to the recognition of malignant change is to have good radiologic baseline studies taken in early adulthood. Malignancy is characterized by a focal change in a bone of the baseline pattern often demonstrating a more "windblown" pattern often with a new soft tissue mass best seen on an MRI scan The patient also experiences chronic pain and may feel a mass.



Radiograph of the pelvis and proximal limbs. Multiple cartilaginous lesions, mainly metaphyseal, with bone deformations



Radiograph of the hand: multiple chondromas



Cartilaginous lobules show evident atypia with a pseudomalignant appearance of the lesion. This histology has to be considered benign if the lesion is in a child, or in Ollier/Maffucci disease, or in a periosteal site, in hands and feet, or in a joint (synovial chondromatosis)

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