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Chondroid Tumors: Review of Salient Imaging Features and Update on the WHO Classification

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A B S T R A C T

Chondrogenic tumors are typically well recognized on radiographs, but differentiation between benign and malignant cartilaginous lesions can be difficult both for the radiologist and for the pathologist. Diagnosis is based on a combination of clinical, radiological and histological findings. While treatment of benign lesions does not require surgery, the only curative treatment for chondrosarcoma is resection. This article (1) emphasizes the update of the WHO classification and its diagnostic and clinical effects; (2) describes the imaging features of the various types of cartilaginous tumors, highlighting findings that can help differentiate benign from malignant lesions; (3) presents differential diagnoses; and (4) provides pathologic correlation. We attempt to offer valuable clues in the approach to this vast entity.

Introduction

Chondroid tumors represent the largest group of bone tumors. Chondrosarcoma is the third most common primary bone tumor following multiple myeloma and osteosarcoma.[1] Cartilage tumors are a heterogeneous group of neoplasms that all share the presence of tumor cells producing cartilaginous matrix. This matrix is characterized by a lobulated growth pattern and popcorn-like or ring- and arc-like calcifications, which can be visualized on conventional radiography and computed tomography (CT).[2] However, the chondroid matrix is visible on conventional radiography only in 65% of chondrosarcomas.[3]

The World Health Organization (WHO) updated in 2020 the chondroid tumor classification. Differences compared to the 2013 classification are: (1) Chondroblastomas and chondromyxoid fibromas were moved to the benign group; (2) synovial chondromatosis was moved to the intermediate group; (3) atypical cartilaginous tumors (ACT) of the appendicular skeleton were assigned to the intermediate group and those of the axial skeleton (including scapula and pelvis) were designated as chondrosarcomas grade 1 (Table).[4] The differentiation of enchondromas from atypical cartilaginous tumor/low-grade chondrosarcomas is one of the most difficult distinctions for radiologists, pathologists and clinicians. Therefore, we discuss radiological and clinical characteristics that can help in this differentiation.

Throughout this pictorial review, our goal is to provide an overview of the imaging and histological features of benign and malignant chondrogenic tumors in order to aid in their diagnosis. We explain in detail the recent changes in the WHO classification and how they affect imaging and management strategies.

Benign Chondrogenic Tumors

Osteochondroma

Osteochondromas (or exostoses) are the most common benign skeletal neoplasms and represent 20%-50% of all benign bone tumors and 10%-15% of all bone tumors. Most of the cases are incidental and asymptomatic. They most commonly arise from the appendicular skeleton, especially around the knee. Osteochondromas are composed of cortical and medullary bone with an overlying hyaline cartilage cap. They may be solitary or multiple (referred to as hereditary multiple exostoses). The lesion is composed of cortical and medullary bone protruding from and continuous with the underlying bone. The areas of osseous continuity between parent bone and osteochondroma may be broad (sessile) or narrow (pedunculated) and can be detected on all imaging techniques.[5]

The authors declare that there is no conflict of interest.

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TABLE
WHO Chondrogenic tumor classification

Benign
Osteochondroma Enchondroma Periosteal chondroma Osteochondromyxoma Subungual exostosis Bizarre paraosteal osteochondromatous proliferation Chondroblastoma Chondromyxoid fibroma
Intermediate (Locally aggressive)
Synovial chondromatosis Atypical cartilaginous tumor
Malignant
Chondrosarcoma grade 1/2/3 Dedifferentiated chondrosarcoma Clear cell chondrosarcoma Mesenchymal chondrosarcoma

CT is the optimal technique to demonstrate lesion continuity and also clearly identifies the typical chondral calcifications of the cartilage cap. The thickness of the cartilage cap can be difficult to assess with CT and therefore magnetic resonance imaging (MRI) is the best modality to evaluate the cartilage cap. Nonmineralized portions of the cartilage cap have intermediate-low signal intensity (SI) on T1 weighted images (T1WI) and very high SI on T2 weighted images (T2WI). A cartilage cap thicker than 1.5-2 cm is suspicious for malignant degeneration.[6] After intravenous gadolinium administration, enhancement of benign lesions is normally seen in the fibrovascular tissue covering the cartilaginous cap; however, the cartilaginous cap should not appear enhanced (Fig. 1, and 2).[7]

Osteochondroma is the most common precursor lesion of secondary chondrosarcoma. About 0.4%-2% of solitary osteochondromas (1%-5% in patients with multiple osteochondromas) undergo malignant transformation (Fig 3).[8]

Enchondroma

Enchondroma is probably the second most common primary bone tumor in adults representing 12%-14% of all benign bone tumors and 3%-10% of all bone tumors.[9] These tumors are the most common benign lesions in the phalanges and they present as an incidental finding in 2.9% of knee MRI.[10] The true prevalence of enchondroma is unknown since many lesions are asymptomatic. They can appear in any bone formed from cartilage and they are typically located in the center of the medullary cavity of tubular bones. They appear mostly (40%-65%) in the small bones of the hand.[11]

Enchondromas show endosteal scalloping and contain calcified chondroid matrix except for the phalanges, where they appear as small lytic lesions with sharply defined margins (Fig 4). The characteristic rings and arcs pattern seen on radiography and CT reflects the pattern of chondral calcification around lobules of mature hyaline cartilage, with a narrow transition zone (Fig 5). On MRI, enchondromas show well-defined lobulated margins, with intermediate T1WI and high T2WI SI consistent with hyaline cartilage and central linear areas of low signal intensity (rings and arcs) corresponding to areas of calcification (Fig 6).[9] It is often difficult to differentiate between an enchondroma and a bone infarction. A key feature is that the areas of mineralization are more central in chondroid lesions and peripheral in bone infarct.[12]

Multiple enchondromas are rare occurrences, presenting either as enchondromatosis, seen in Ollier disease and together with hemangiomas in Maffucci syndrome, or as a hereditary syndrome. Ollier disease is a nonhereditary syndrome, often unilateral and it may regress or undergo malignant degeneration (Fig 7).[13]

Differentiation of enchondroma from atypical cartilaginous tumor/chondrosarcoma grade 1 is challenging for radiologists and pathologists. The presence of permeation on histopathology is diagnostic for an ACT/chondrosarcoma. In the absence of clear-cut diagnostic criteria on histopathology, it is frequently stated that the differentiation between the two disease entities is based on a consensus decision between radiology, histopathology and clinical

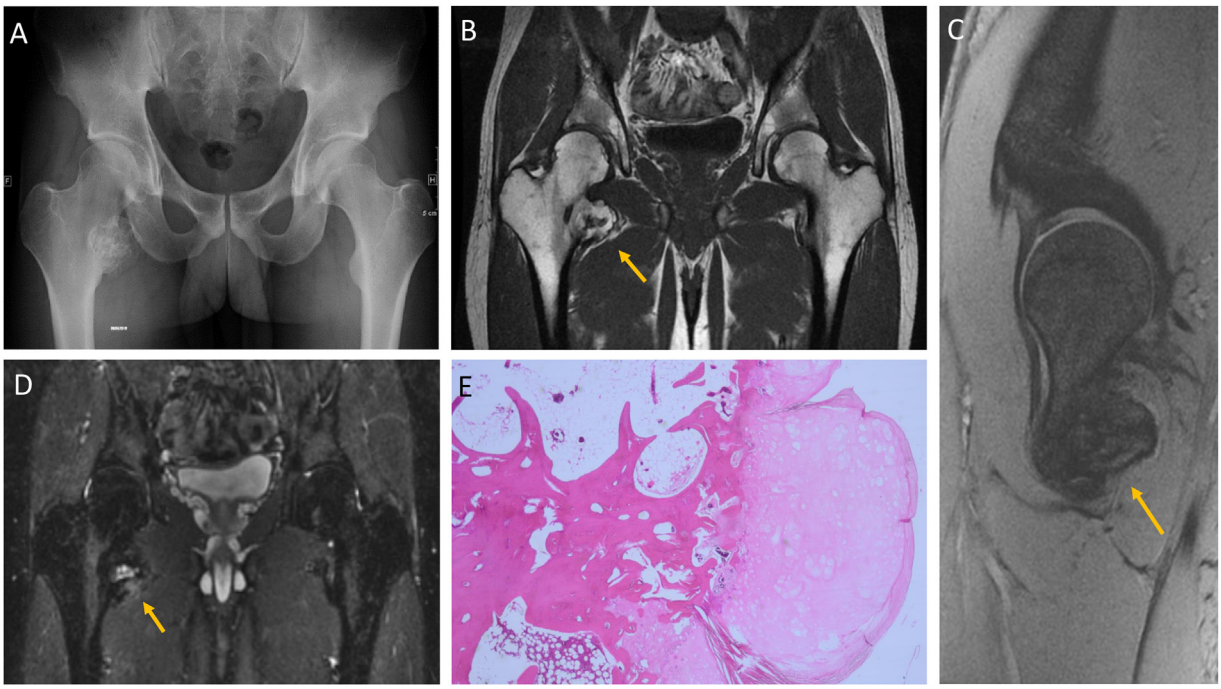

FIG 1. A. 26-year-old male with a 3-month history of right groin pain. Plain radiograph (A) shows an exophytic lesion in the proximal right femur, greater than 3 cm with chondral calcifications suggestive of osteochondroma. Coronal T1 (B), sagittal GRE T2 (C) and coronal STIR (C) confirm the continuity of the femoral neck's cortical and medullary (arrow in B and C) with those of the lesion. There is central intralesional high signal (C) in relation to chondral matrix with a 2 mm cap (arrow). Low power view of an osteochondroma (E) showing a cartilage cap lined by perichondrium, contiguous with mature bone (hematoxylin-eosin [H-E] stain). (Color version of figure is available online.)

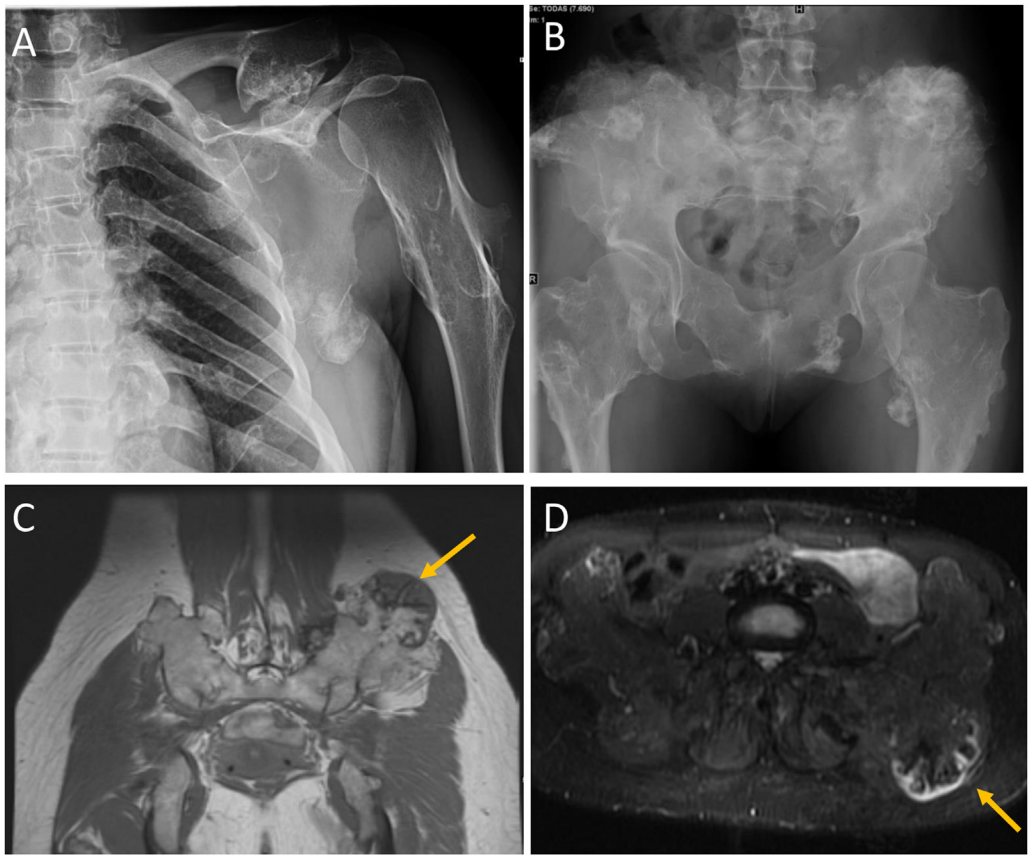

FIG 2. A. 24-year-old female with multiple osteochondromatosis diagnosed in childhood. Plain radiograph (A, B) show multiple pedunculated and sessile exostoses with chondral calcifications in both axial and peripheral skeleton with no signs of complication. The largest one emanates from the left iliac bone with low signal in T1WI (arrow in C) and has a 7 mm width cartlage cap visible in STIR sequence (arrow in D) with smooth edges. (Color version of figure is available online.)

findings.[14] Core needle biopsy might be prone to sampling error due to the heterogeneity of these tumors, which present areas of benign hyaline cartilage even in high-grade chondrosarcoma. For this reason, needle biopsy is not recommended to differentiate between an enchondroma and a low-grade chondrosarcoma or an ACT as this procedure has a considerable risk of error, and may result in a non-representative sample.[15]

Douis et al[14] found that clinical assessment and conventional MRI aid in this differentiation. They found that features such as tumor length (enchondromas are typically <5 cm), depth of endosteal scalloping (more than two-thirds of the depth of the cortex), cortical destruction, bone expansion, soft tissue mass, and pain favor ACT/chondrosarcomas grade 1. Ferrer-Santacreu et al[16] proved a statistical relationship between 3 features and chondrosarcoma grade 1 in long bones: pain on palpation, cortical scalloping in CT/MRI, and Tc99 uptake similar to or higher than the anterosuperior iliac crest. Nevertheless Crim et al[17] proved that scalloping should be better considered a correlate of large size and/or subcortical location rather than as an independent imaging sign of chondrosarcoma. No or minimal enhancement in MRI after more than 10 seconds is suggested in the literature as sufficient evidence to confidently diagnose enchondroma.[9] On the other hand, when enhancement of a chondral lesion is present, dynamic contrast-enhanced MRI is not useful in differentiating enchondromas from ACT/grade 1 chondrosarcomas.[14] Currently the use of diffusion weighted images (DWI) has not shown value to differentiate between enchondromas, ACT/chondrosarcomas grade 1 and high-grade chondrosarcomas.[18]

Positron emission tomography/computed tomography (PET/CT) is a useful technique together with conventional images in the characterization of chondroid tumors since the maximum standardized uptake value (SUVmax) correlates with the histological grade. Very low SUVmax (<2.0) supports a diagnosis of enchondroma or ACT, while elevated SUVmax (>4.4) is suggestive of a chondrosarcoma grade 2 or 3; however, 46% of the tumors are in the indeterminate range.[19]

Asymptomatic enchondromas can be treated nonsurgically, with radiological follow up. Indications for curettage are continuous symptoms; enlargement or radiographic changes during follow up to rule out a low-grade malignant variant; or an actual fracture of the host bone.[20]

Periosteal Chondroma

They arise from the periosteum of long bones, especially proximal humerus, distal femur, and phalanges. Radiological features include cortical scalloping with sclerosis, chondroid matrix mineralization and rarely intramedullary invasion. On MRI they are typically seen as soft tissue lesions abutting the cortex, with lobulated margins showing typical signal intensity characteristics of chondral tissue. They are difficult to distinguish from juxtacortical chondrosarcomas, being the size of the lesion the most important distinctive factor. Mean size for these tumors is 2.4 cm vs 5.5 cm of periosteal chondrosarcoma.[21]

Osteochondromyxoma

It is a benign but locally aggressive tumor with both osteoid and chondroid production. This rare tumor arises in approximately 1% of patients with Carney complex and sites of involvement include the

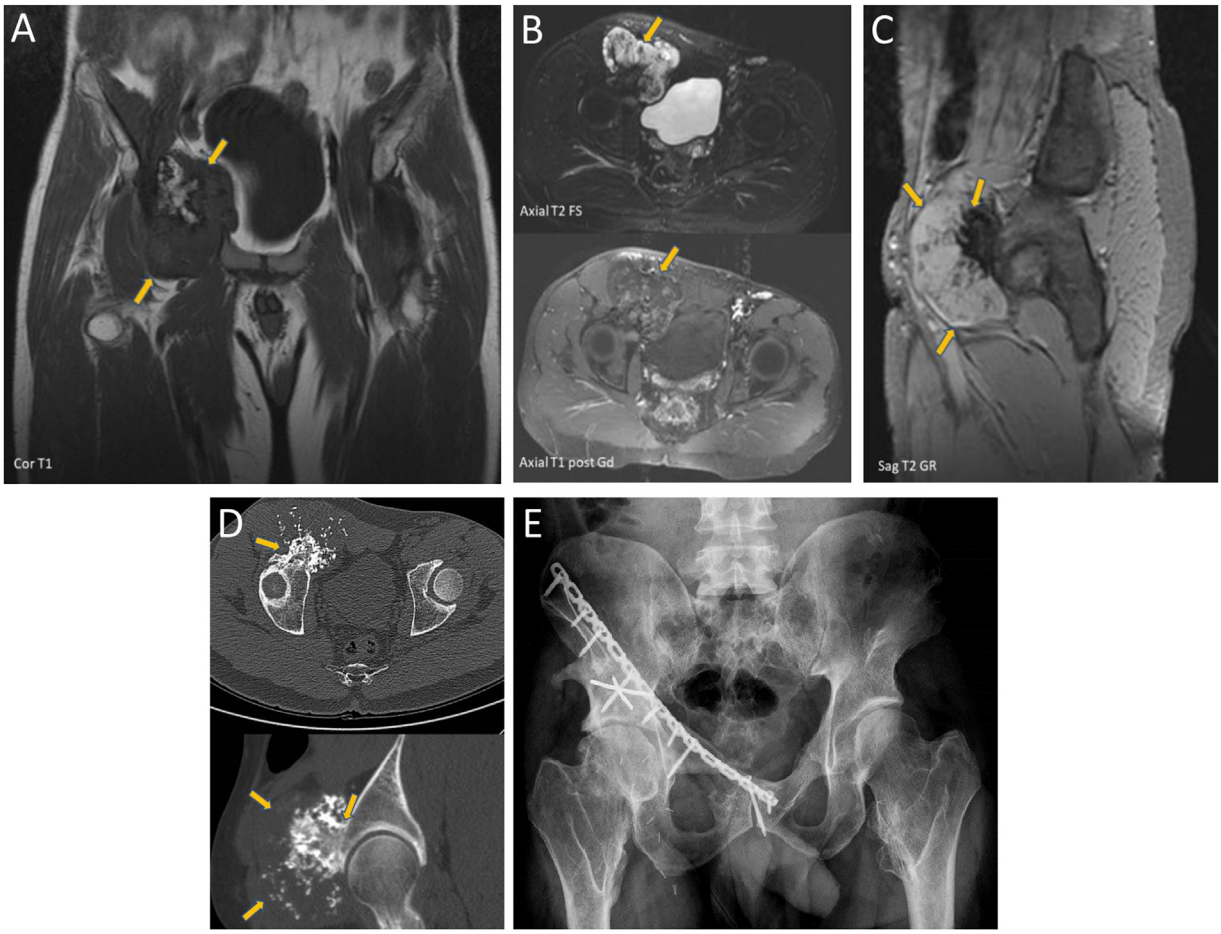

FIG 3. T1WI (A), FS T2WI and contrast enhanced FS T1WI (B), GRE T2WI (C), and CT (D) images show multiple osteochondromas affecting both iliac bones, sacrum, ilio-pubic ramus and both femoral necks and diaphysis. There is an expansive lytic lesion in the right puboacetabular junction with thick cartilaginous cap (>1.5cm) that suggests neoformative transformation (arrows in A, B, C, D). Radiograph after surgery where chondrosarcoma was confirmed (E). (Color version of figure is available online.)

tibia and sinonasal bones. Destructive growth with extension into soft tissue may occur but metastases have not been reported.[22]

Subungual Exostosis

These lesions are bony projections, which arise from the dorsal surface of the distal phalanx, most commonly of the first toe. Although they usually appear as well-circumscribed bone lesions, they lack a clear contiguity of both the medullary cavity and cortex, which helps distinguish them from osteochondromas.[23]

Bizarre Parosteal Osteochondromatous Proliferation (BPOP)/Nora Lesion

These lesions are benign wide based bony growth lesions similar to an osteochondroma and are typically located in hands and feet. However, they often lack the characteristic orientation away from the physis, the cartilage cap as well as the medullary involvement seen in osteochondromas. BPOPs may show aggressive radiographic features and have aggressive behavior locally, but there is no reported risk of malignant degeneration.[9]

Chondroblastoma

Chondroblastomas are rare tumors that arise in the epiphysis of long bones, patella or tarsal bones, usually in skeletal immature patients with pain and joint swelling. On plain radiograph and CT, chondroblastomas are seen as well-defined lucent lesions, lobulated with a thin sclerotic margin. A total of 40%-60% of these tumors present internal calcifications and regular benign periosteal reaction. On MRI, these lesions show typical cartilage features. MRI demonstrates prominent bone marrow and soft tissue associated edema, which is almost always present (Fig 8).[24,25]

Most cases are successfully treated by curettage. Recurrence rates range from 5% to 14% and the reported rate of metastases is <1%.[26] Consequently, chondroblastoma is better classified as a benign tumor instead of locally aggressive (intermediate) lesion in the updated WHO classification.[4]

Chondromyxoid Fibroma

It is a rare neoplasm that comprises a combination of chondroid, myxoid and fibrous tissue components and typically occurs in young adults. Most chondromyxoid fibromas involve the medullary cavity of the metaphyseal region of long bones. However, they can also appear in hand and foot and they rarely show malignant degeneration. When they appear in elders, paraosteal chondrosarcoma should be excluded.[27]

Radiographically they present as eccentric lytic lesions, with well-defined sclerotic margin, often expansive, without periosteal reaction. They also present geographic bone destruction and septations (pseudotrabeculation). Internal matrix calcification is usually absent. On MRI, they show low signal in T1WI and heterogeneous intermediate signal in T2WI due to fibrous, chondroid and myxoid components.

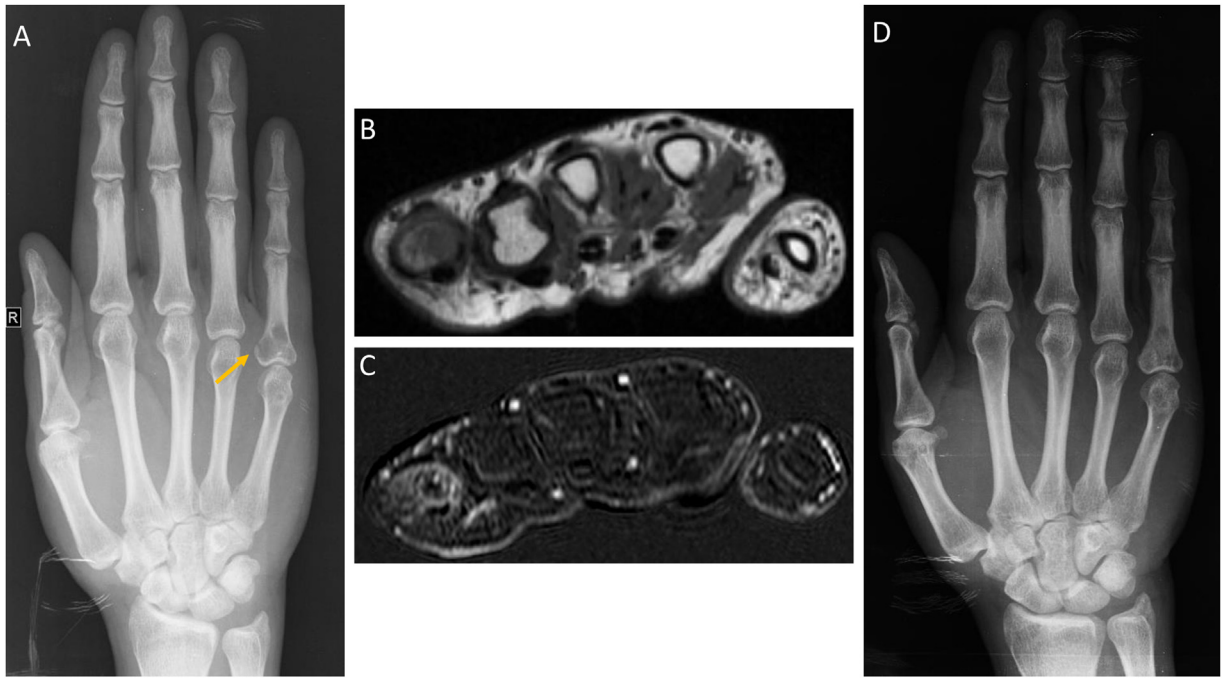

FIG 4. Plain radiograph (A) of the left hand shows a lytic lesion with well-defined margins in the base of the proximal phalanx of the fifth finger, with cortical rupture (arrow). The lesion is hypointense on T1WI (B) and enhances after contrast administration as seen in FS T1WI subtraction imaging (C), compatible with enchondroma. The enchondroma fracture was treated with curettage and autologous bone graft obtained from iliac crest (D). (Color version of figure is available online.)

Most of them show peripheral nodular enhancement after gadolinium administration (Fig 9).[28]

Juxtacortical or surface-type chondromyxoid fibromas are even rarer (Fig 10). Baker et al[29] reviewed all previously reported juxtacortical or surface-type chondromyxoid fibromas, publishing the largest case series and describing 20 cases collected over a 47-year period. They concluded that the presence of calcification was found more frequently within surface-type chondromyxoid fibroma than in the conventional intramedullary chondromyxoid fibromas. Malignant transformation is exceptional and prognosis is excellent; therefore, they are now classified as benign instead of locally aggressive.[4]

Intermediate (Locally Aggressive) Chondrogenic Tumors

Synovial Chondromatosis

Primary synovial chondromatosis is a synovial metaplasia and proliferation characterized by multiple intra-articular cartilaginous loose bodies of relatively similar size. It most commonly occurs in the third through fifth decades of life, presents with joint pain, swelling, and stiffness and it most commonly affects the knee, hip, and elbow. These cartilaginous bodies frequently calcify with a typical “ring and arc” pattern and are easily seen on radiography. CT confirms that the

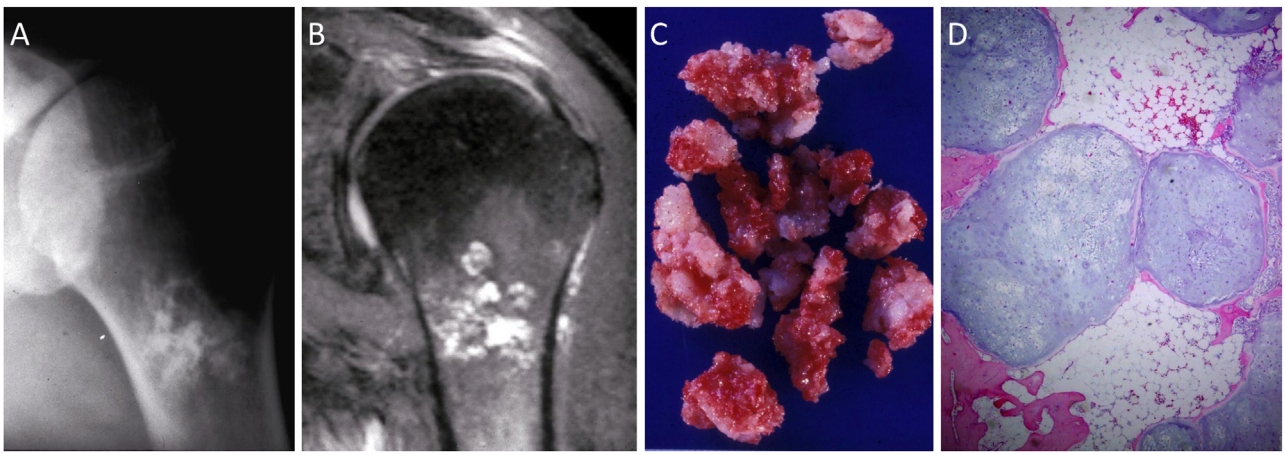

FIG 5. A. 52-year-old male with 2 months shoulder discomfort. Plain radiograph (A) and FS T2WI (B) show an intraosseous lytic lesion in the proximal diaphysis of the humerus, with nodular calcifications; it does not deform or thicken the contours of the cortical bone. Gross examination of the lesion after curettage demonstrates a proliferation of hyaline cartilage. Photomicrograph (D) shows an enchondroma characterized by lobules of hyaline cartilage separated one another by bone marrow (H-E stain). (Color version of figure is available online.)

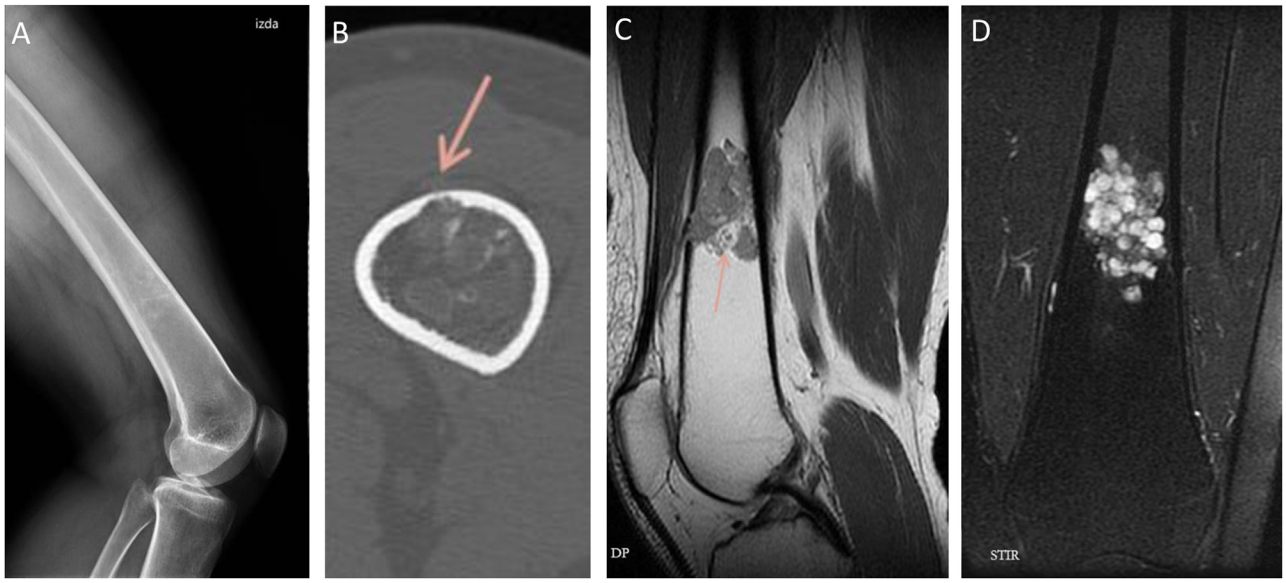

FIG 6. A. 42-year-old woman in study due to a painless lesion in the distal femur. Plain radiograph (A) and CT (B) show a chondral matrix lesion in distal femur with cortical scalloping (arrow in B). These findings are also present on MR, which shows a lesion with popcorn calcifications, intermediate signal on PD (C) and high signal on STIR (D). The result of the biopsy was an enchondroma (not shown). (Color version of figure is available online.)

loose bodies are intra-articular. In 30% of the cases they do not calcify, and plain radiographs may show joint effusion or less frequently bone erosions secondary to pressure.[30] On MRI, loose cartilaginous bodies have typical chondroid signal characteristics: intermediate to low signal in T1WI and high signal in T2WI. Ossified loose bodies have cortical edges of low signal with a central fat intensity. Some ossified bodies are diffusely dense and sclerotic (low signal in all sequences).

In secondary synovial chondromatosis the intra-articular loose bodies are a result of trauma, osteoarthrosis, or neuropathic arthropathy. It typically associates with degenerative changes of the joint. The intra-articular bodies tend to be larger, less numerous and more varied in size and shape than in primary synovial chondromatosis (Fig 11).

Although controversy remains regarding the optimal treatment, it generally includes loose body removal combined with an open or

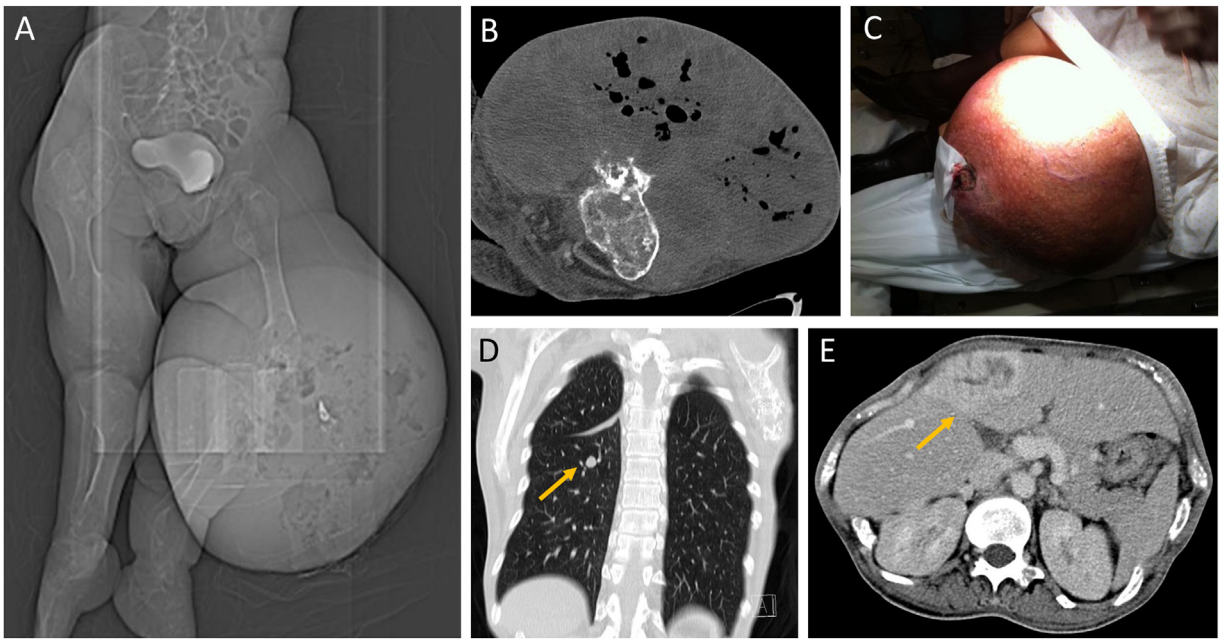

FIG 7. Woman diagnosed with Ollier disease without follow-up. On CT topogram (A) a voluminous mass in the left lower extremity is observed. CT image (B) shows a bone lesion with chondroid calcifications and destruction of the bone cortex due to degeneration of the disease to chondrosarcoma. The lesion is associated with a soft tissue mass with areas of heterogeneous attenuation and gas, compatible with abscess. Intraoperative photograph of the mass (C). Pulmonary (D) and hepatic (E) metastasis were reported on CT (arrows). (Color version of figure is available online.)

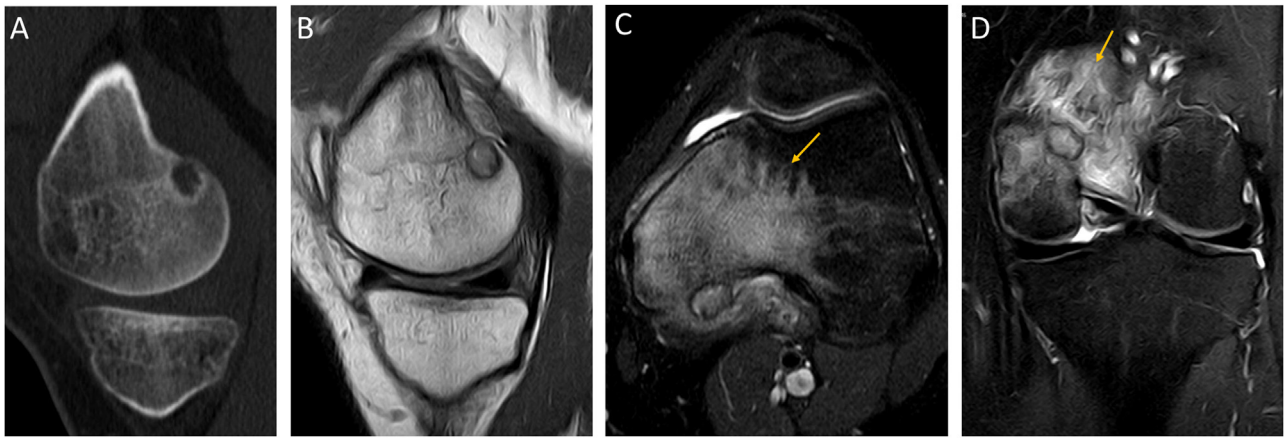

FIG 8. A. 20-year-old-male with pain and swelling of the knee. CT (A) of the left knee shows a well-defined lytic lesion with a thin sclerotic margin in the posterior area of the medial femoral condyle. On T1WI (B) and axial and coronal FS T2WI (C and D) the lesion shows typical chondroid signal as well as intense bone marrow and soft tissue edema (arrows). The pathological anatomy (not shown) confirmed the diagnosis of chondroblastoma. (Color version of figure is available online.)

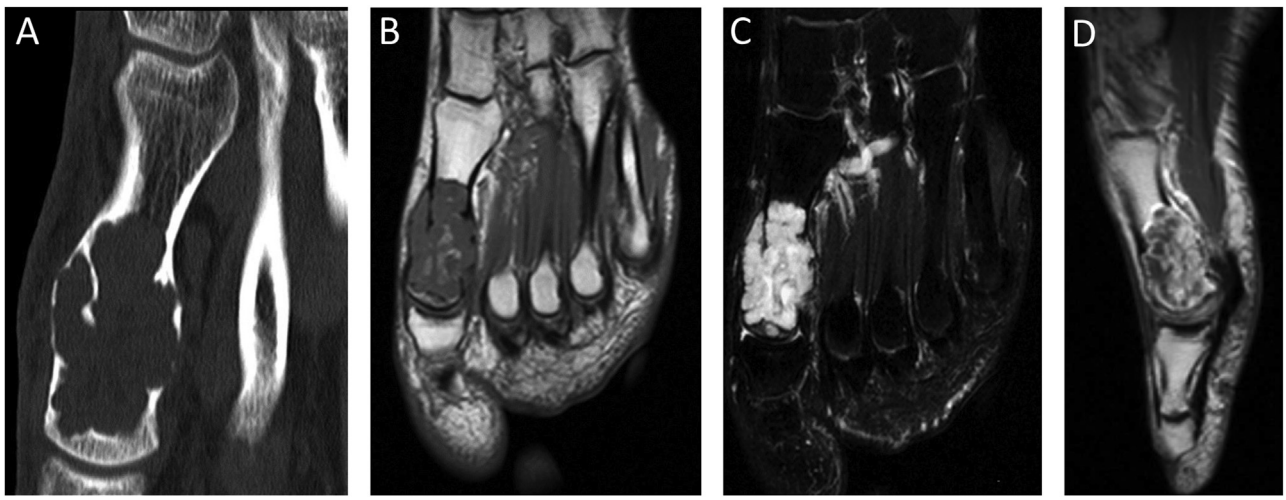

FIG 9. CT (A) shows a lytic lesion in the distal diaphysis and head of the first metatarsal that causes bone cortical expansion. It has intermediate signal on T1WI (B), hyperintense signal on STIR (C), with heterogeneous enhancement on T1WI (D) after administration of intravenous contrast. Anatomopathology (not shown) was compatible with chondromyxoid fibroma.

arthroscopic synovectomy. The addition of synovectomy has been recommended because it reduces the recurrence rate of loose body removal alone.[31]

In the WHO 2020 classification of synovial chondromatosis was moved from the benign to the intermediate group to reflect the locally aggressive growth pattern and the high risk for local recurrence.[4] This change highlights the importance of an early radiological diagnosis and treatment to decrease the risk of recurrence and degenerative changes.[31] Synovial chondrosarcoma can rarely arise from synovial chondromatosis and may be considered if there are multiple recurrences, rapid lesion enlargement, extra-articular extension, or clear bone marrow invasion.[32]

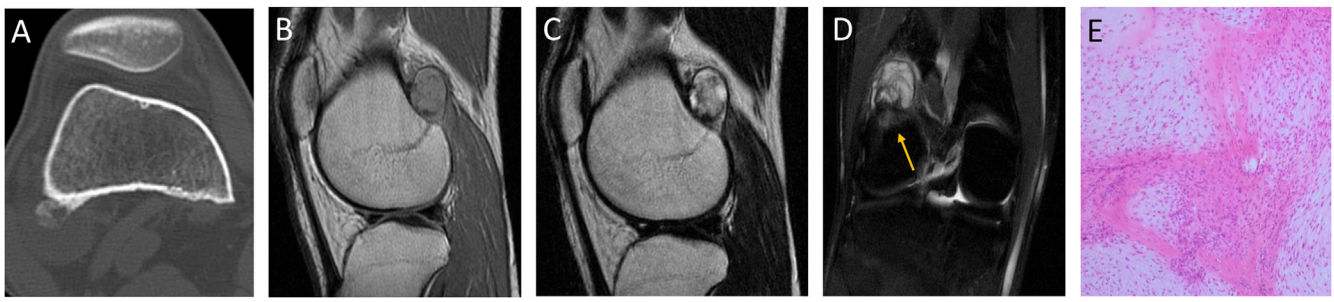

FIG 10. A. 22-year-old male with a 2-year history of pain in the right knee. CT of the right knee (A) shows an exophytic lesion, with irregular but well-defined sclerotic margin and periosteal reaction in continuity with the cortical of the posterolateral face of the lateral femoral condyle. A linear fracture was also present (not shown). The previous findings are confirmed on the MR images, which demonstrate an irregular but well-defined bony outgrowth of low intensity on PD (B) and heterogeneous high-predominant signal on T2WI (C). FS PD shows condyle bone marrow edema (arrow in D). En bloc excision was performed, showing a juxtacortical chondromyxoid fibroma (E) characterized by hypocellular lobules of poorly formed hyaline cartilage with fibrous septae (H-E stain). (Color version of figure is available online.)

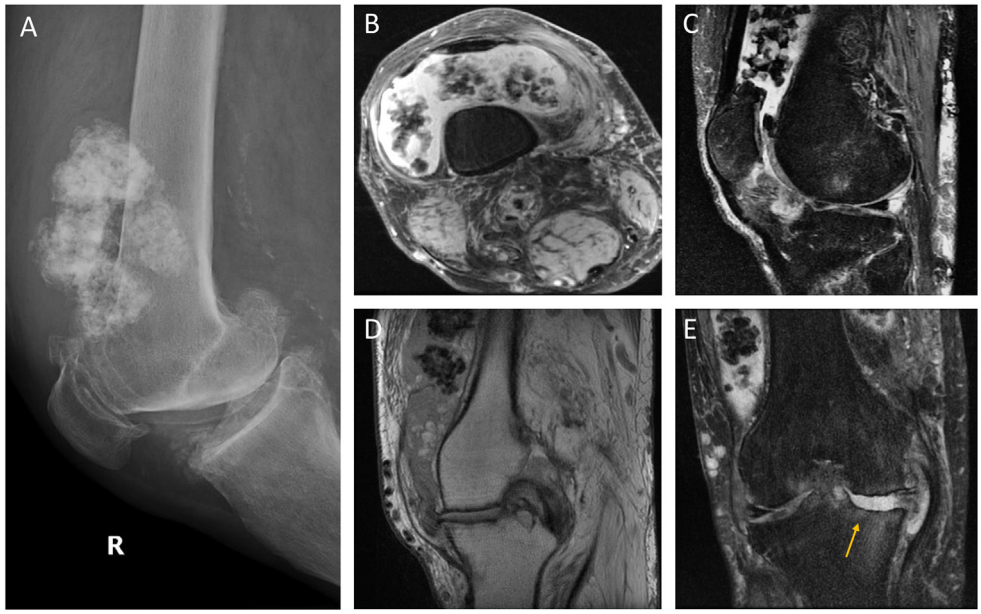

FIG 11. Plain radiograph of the right knee (A) demonstrates coarse ossified loose bodies and severe degenerative disease. Moreover, FS PD (B, C, E) and PD (D) show medial meniscus tear with grade 4 chondropathy (arrow in E). Findings are compatible with secondary synovial chondromatosis. (Color version of figure is available online.)

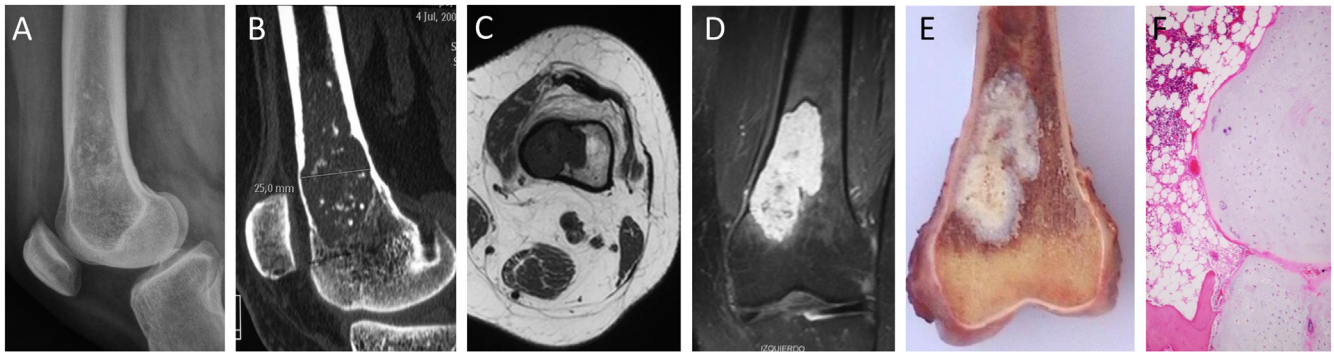

FIG 12. Plain radiograph (A) shows an intraosseous lytic lesion with punctiform calcifications, in the lower metadiaphysis of the femur. CT (B) shows a slight bone deformation and endosteal scalloping. On MR, the lesion is hypointense on T1WI (C) and hyperintense on T2WI (D). Gross image (E) of atypical cartilaginous tumor showing a lobulated endomedullary lesion permeating through the marrow with erosion of the surrounding cortex. Low power view (F) of atypical cartilaginous tumor showing a lobular growth pattern, and abundance of predominantly hyaline cartilaginous matrix (H-E stain). (Color version of figure is available online.)

Atypical Cartilaginous Tumor

Chondrosarcomas are divided into grades ranging from 1 to 3 according to their increasing aggressiveness. Since grade 1 chondrosarcomas of the long bones rarely metastasize and show no signs of local malignant behavior, in the WHO classification of 2013 the term "chondrosarcoma, grade 1″ was replaced with the term "atypical cartilaginous tumor" and these tumors were considered as locally aggressive instead of malignant.[33] In the new edition of the WHO classification of 2020, a clear distinction was made between chondrosarcoma grade 1 in the axial skeleton and appendicular skeleton. Chondrogenic tumors in the appendicular skeleton (long and short tubular bones) should be termed ACTs due to their more favorable prognosis (Fig 12). The term chondrosarcoma grade 1 (with identical histomorphology as ACT) is reserved for tumors in the axial skeleton (including pelvis, scapula and skull base) reflecting the poorer clinical outcome at these sites (Fig 13). Secondary conventional central ACT/chondrosarcoma grade 1 are tumors arising centrally in bone in association with a pre-existing enchondroma. Secondary peripheral ACT / chondrosarcoma grade 1 are tumors arising within the cartilaginous cap of a pre-existing osteochondroma.[4]

Due to the increase in patients undergoing MRI examinations for joint-related complaints, the incidental detection of ACT has increased substantially.[34] Radiographically and histologically ACTs/chondrosarcomas grade 1 are difficult to distinguish from enchondromas as we have previously discussed. A systematic review realized by Deckers et al[34] found that compared with ACT, high-grade chondrosarcoma (ie, grades 2 and 3) may present more often with the following MRI characteristics: loss of entrapped fatty marrow, cortical breakthrough, and extraosseous soft tissue expansion.

Changes in the WHO classification together with the recent insight from low transformation risk of ACTs (<1%), has resulted in a more conservative treatment approach of ACTs in literature. Presumed diagnosis of ACT based on imaging and clinical examination without a diagnostic biopsy is considered safe and appropriate. The recommended management of long bone ACTs has assumed less aggressive surgical management over the last 20 years, although surgeons have been performing diagnostic biopsies and wide resections at similar to historical rates. Actual recommendations include extended intralesional excision with curettage.[15] However, many authors have questioned if the negative side effects of surgical treatment of ACTs outweigh the potential benefits. Active surveillance

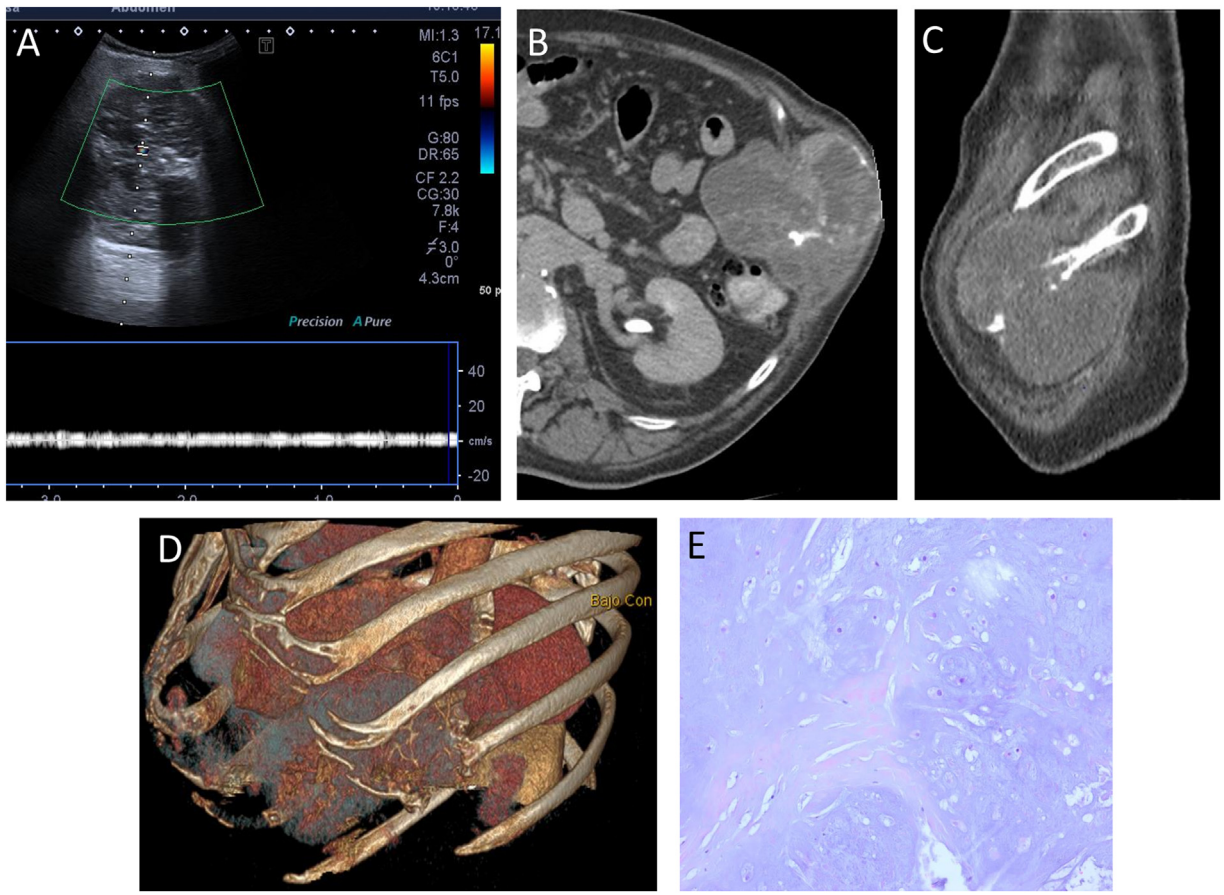

FIG 13. A. 81-year-old woman with a slow growing (10 years) hard lump in the left hypochondrium. No skin changes. US (A) demonstrates a well-defined 10 cm solid mass, heterogeneous, with some echogenic foci that suggest calcifications, and vascularization. Axial contrast enhanced CT (B), sagittal (C), and 3D (D) reconstructions confirm chondroid calcifications and reveal that the mass depends on the 11th costal arch with intraabdominal extension. Grade 1 chondrosarcoma (E) with myxoid matrix changes (H-E stain). (Color version of figure is available online.)

seems effective for asymptomatic ACTs in the long bones with nonaggressive imaging characteristics (ie, cortical destruction, presence of soft tissue mass, moth-eaten, permeative or extensive osteolysis, multilamellar or aggressive periosteal reaction). Nevertheless, follow-up schemes should be tailored according to biological behavior to prevent overutilization of costly advanced imaging.[35] It is extremely important to establish multidisciplinary teams early on in the diagnosis and evaluation steps of ACT, as this may lead to less aggressive surgical treatments with overall better functional outcomes.[15]

Malignant

Chondrosarcoma is the third most common primary bone tumor following multiple myeloma and osteosarcoma. Chondrosarcomas are almost always symptomatic and they frequently metastasize, primarily to the lungs. Chondrosarcomas may be classified as primary (*de novo*) or secondary. Secondary chondrosarcomas arise in association with a pre-existing cartilaginous lesion, such as enchondroma (Fig 7), osteochondroma (Fig 3) or Paget disease (Fig 14).[2] There are various histological subtypes of chondrosarcomas, of which conventional chondrosarcoma is by far the most common. Rarer subtypes include clear cell chondrosarcoma, mesenchymal chondrosarcoma, and dedifferentiated chondrosarcoma.

Chondrosarcoma (Grade 1, Grade 2, Grade 3)

Conventional chondrosarcomas are classified according to the lesion site into intramedullary (most common) (Fig 15), peripheral (its precursor is always an osteochondroma) and periosteal (Fig 16). The localization of grade 2 and 3 is similar to that of central ACT/chondrosarcoma grade 1, and therefore all parts of the skeleton can be affected.[4] A histological differentiation between grade 1, 2, and 3 is relevant for prognosis. It is well acknowledged among those in the orthopedic community that grading from biopsies and the possible seeding of tumor material in the biopsy tract can be inaccurate. Indeed, some centers avoid preoperative biopsy in obvious chondrosarcoma cases.[36]

On plain radiographs and CT, chondrosarcomas appear as lytic lesions with intralesional calcifications in rings and arcs. The degree of calcification is variable, but higher-grade tumors tend to show less mineralization. As the lesion grows it causes cortical thickening and breach, periosteal reaction, deep endosteal scalloping (involving more than 2/3 of cortical thickness) and soft tissue mass. There are typical imaging-based morphological criteria of malignancy such as moth-eaten pattern (Lodwick type II) or permeative growth (Lodwick type III).[2] On MRI, chondrosarcomas show low to intermediate signal in T1WI and very high intensity of noncalcified portions in T2WI (because of their high-water content) with hypointense internal septa. Perilesional marrow edema is unusual, a feature that helps in the distinction from chondroblastoma. After gadolinium administration, most demonstrate moderate to intense heterogeneous contrast enhancement, which can be septal and peripheral rim-like, corresponding to fibrovascular septations between lobules of hyaline cartilage (Fig 17).[1]

On PET/CT, chondrosarcomas typically demonstrate increased uptake of fluorodeoxyglucose as previously explained (Fig 18).[19] Chondrosarcomas (all grades) are managed by excision and negative margins.[4]

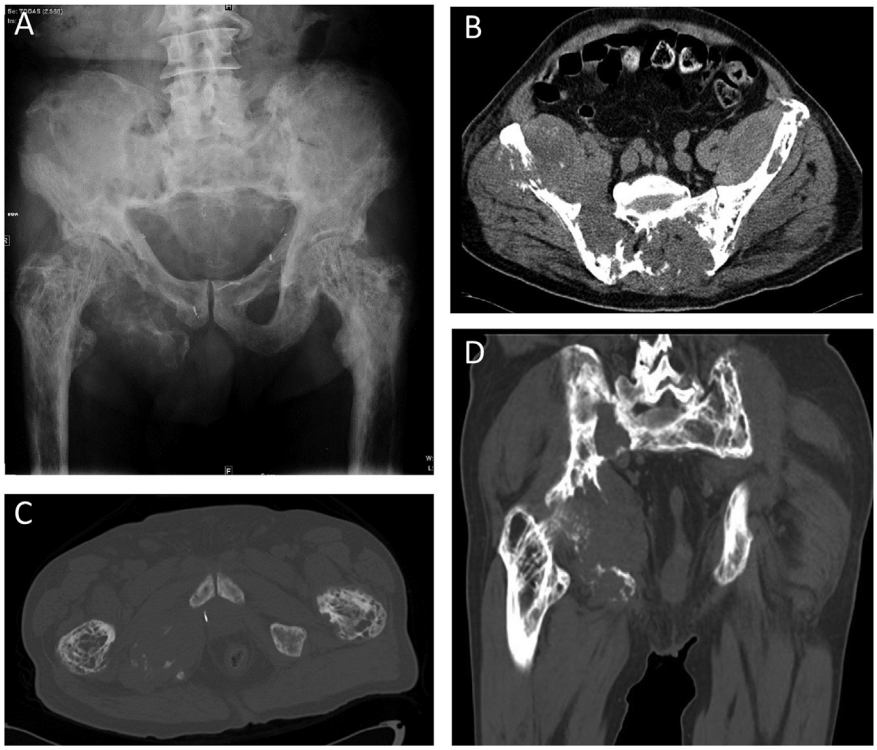

FIG 14. Plain radiograph (A) and CT (B-D) images show extensive lytic and blastic lesions in the pelvis in a patient with Paget's disease. Note an expansive lytic lesion that breaks the right ischiopubic ramus with a mass in the adductor musculature and gluteus, suggestive of neoformative transformation. The soft tissue mass in the sacrum invades the medullary cone. Anatomopathology confirmed grade 3 chondrosarcoma transformation in Paget's disease. (Color version of figure is available online.)

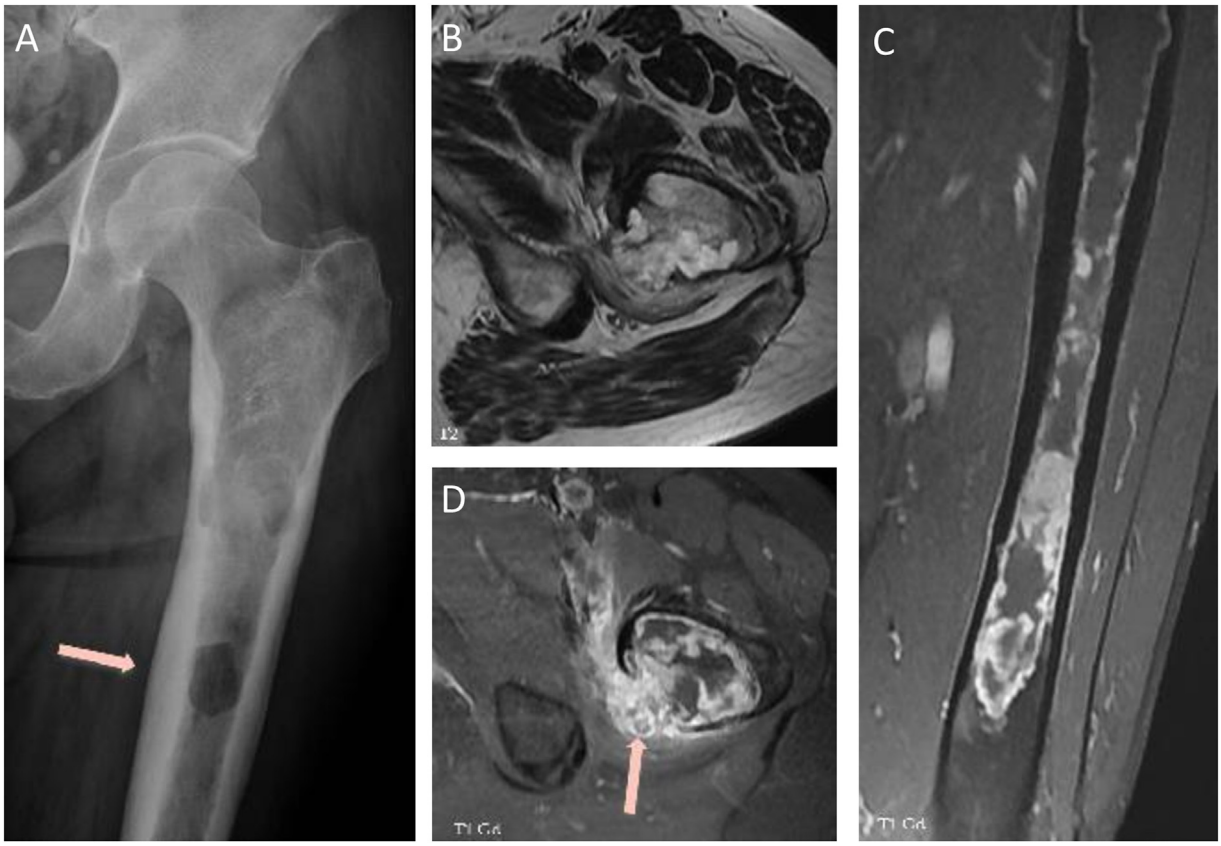

FIG 15. AP plain radiograph of the left femur (A) shows a well-defined lytic lesion (arrow) within an infiltrating intramedullary lesion in the left femoral diaphysis, with deep endosteal scalloping. MR demonstrates a high heterogeneous T2 signal (B) with cortical breakthrough and soft-tissue mass. FS T1WI with contrast shows intense peripheral enhancement (C) and cortical rupture (arrow in D). Anatomopathology confirmed a grade 3 chondrosarcoma. (Color version of figure is available online.)

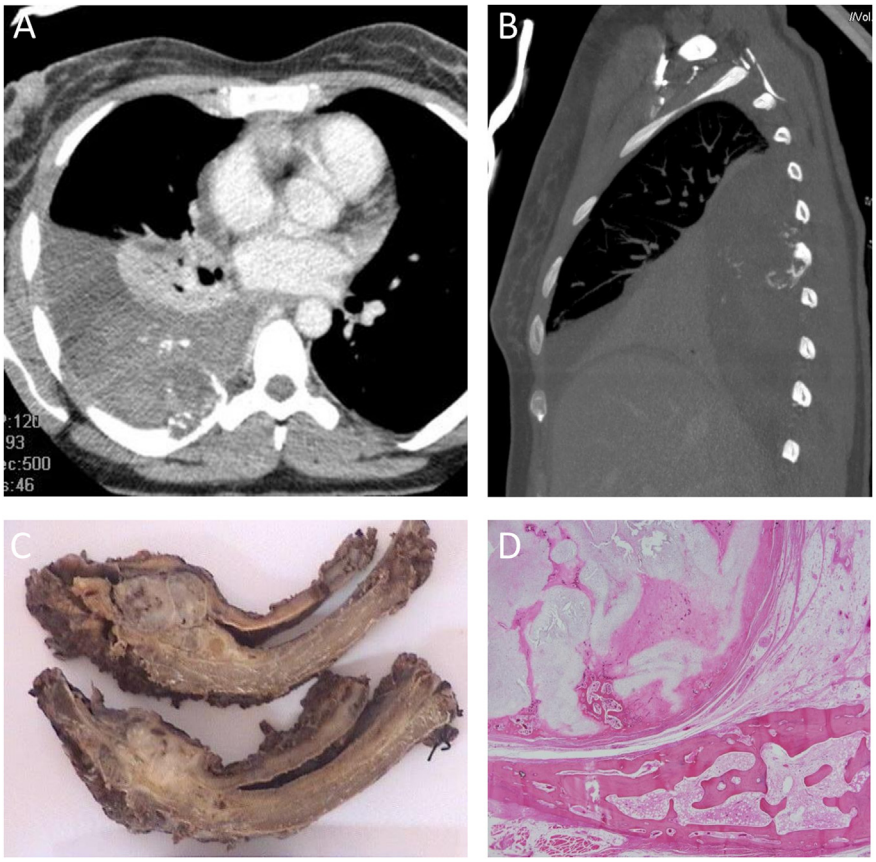

FIG 16. A. 29-year-old woman with acute pleuritic chest pain that is interpreted as metapneumonic. Two days later she developed a massive right hemothorax that needed embolization of intercostal arteries and drainage. CT images (A, B) show an exophytic periosteal tumor in the posterior arch of the seventh right rib, with chondral calcifications projected into the thoracic cavity. Gross image (C) and photomicrograph (D) (H-E stain) showing a lobulated cartilaginous tumor at the surface of bone (periosteal chondrosarcoma grade 3). (Color version of figure is available online.)

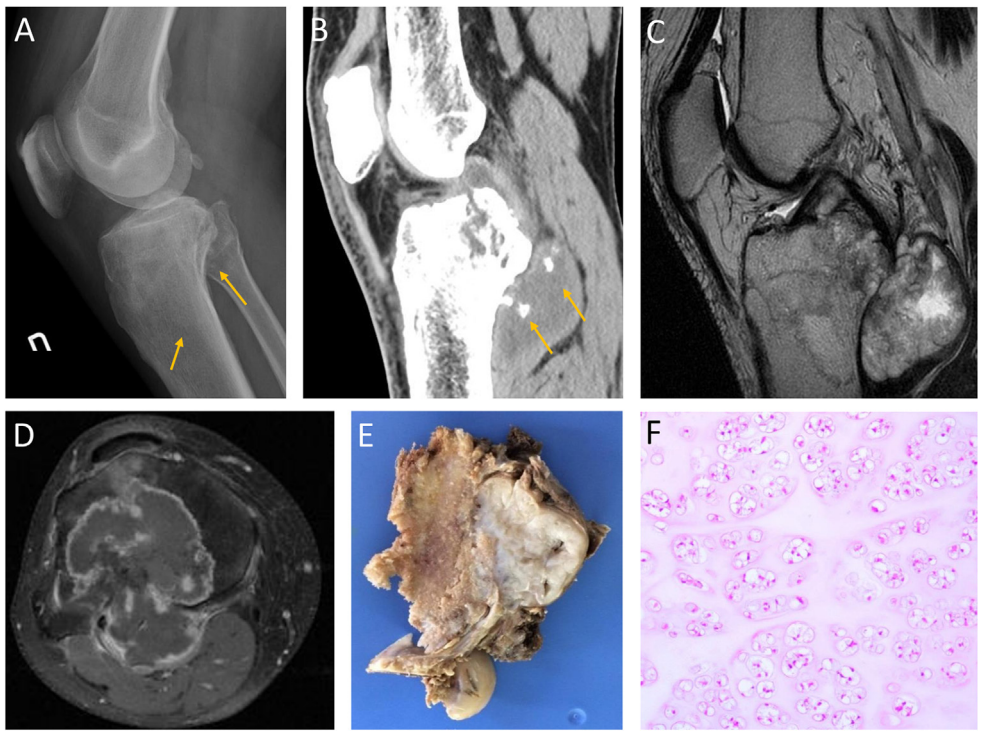

FIG 17. A. 51-year-old male with knee pain. At the diagnosis, knee radiograph (A) shows a metaphyseal bone lesion with cortical destruction and periosteal reaction (arrows). CT (B) shows cortical rupture, intra and extraosseous chondroid matrix and soft tissue mass with calcifications (arrows). PD (C) shows a heterogeneous soft-tissue mass with heterogeneous contrast enhancement on FS T1WI (D). Postsurgical gross image (E) shows a lobular cartilaginous tumor with erosion and destruction of the cortex with soft tissue extension. Grade 2 chondrosarcoma (F) with increased cellularity and myxoid matrix changes (H-E stain). (Color version of figure is available online.)

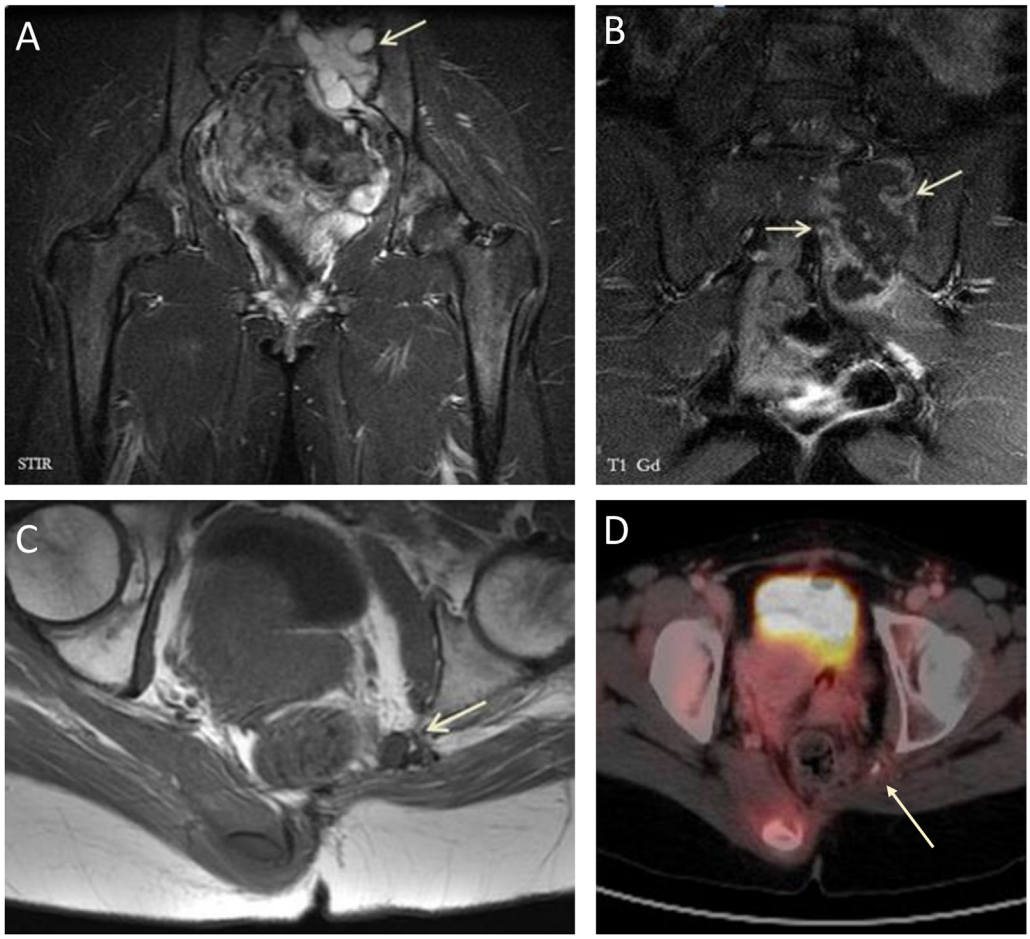

FIG 18. A. 30 years old female with pain in the left hip. STIR (A) MR images show an expansive lesion in the left sacrum with high signal with chondroid appearance (arrow) and poor peripheral enhancement on FS T1WI (arrows in B). The surgically removed piece confirmed a grade 2 chondrosarcoma. Two years after the surgery a recurrent lesion appeared in the remaining sacrum on T1WI (arrow in C) with high uptake on PET-CT (arrow in D). (Color version of figure is available online.)

Dedifferentiated Chondrosarcoma

Dedifferentiated chondrosarcomas represent approximately 11% of all chondrosarcomas and the mean age at diagnosis is 50-60 years.[37] The most common bones involved are the femur, pelvis and humerus, and the tumor arises centrally in most cases, although dedifferentiated peripheral chondrosarcomas are also described.[38] These highly malignant tumors are formed of 2 components, which are often sharply demarcated from one another: well differentiated cartilage (grade 1 chondrosarcoma) juxtaposed to a high-grade non-cartilaginous sarcoma (Fig 19).[39]

Both MRI and CT improve the detection of extraosseous tumor extension. Evidence of a large unmineralized soft-tissue mass associated with a lesion with radiologic features of a chondrosarcoma is suggestive of a bimorphic tumor pattern and should raise the suspicion of dedifferentiation.

Mesenchymal Chondrosarcoma

It is an aggressive tumor with a strong tendency to metastasize. It represents 2%-13% of all chondrosarcomas. Mesenchymal chondrosarcoma is diagnosed in the second to fourth decade equally in men and women. Unlike conventional chondrosarcoma, it most commonly involves the axial skeleton and craniofacial region. Although it normally arises as a primary lesion, it may also appear secondarily in fibrous dysplasia.[39]

Radiographically, they are aggressive moth-eaten to permeative processes with ill-defined periosteal reaction and osseous destruction with a large associated soft tissue mass. In up to 67% of the cases, chondroid matrix calcification is present. On MRI, mesenchymal chondrosarcomas show heterogeneous intermediate SI in T2WI rather than the high SI of hyaline cartilage seen in conventional chondrosarcomas. The enhancement pattern is diffuse and lacks the typical septal and peripheral enhancement seen in typical chondrosarcomas.[1] This pattern is the imaging manifestation of their bimorphic histology: highly undifferentiated noncartilaginous small round cells, similar to those observed in Ewing sarcoma, with hemangiopericytomatous pattern and islands of well differentiated hyaline cartilage.[40]

Clear Cell Chondrosarcoma

It is a rare subtype of chondrosarcoma, which accounts for approximately 2% of all chondrosarcomas. It appears more predominantly in males in their fifth decade. The typical radiographic manifestation of this tumor is a slow growing epiphyseal osteolytic lesion in the proximal femur.[41] These tumors adopt their name from the presence of clear cell chondrocytes which have abundant clear, vacuolated cytoplasm containing large amounts of glycogen (Fig 20).[39]

The typical radiological characteristics of clear cell chondrosarcoma are well-delineated osteolytic lesions, often with typical chondroid matrix mineralization surrounded by a sclerotic rim.[41] Typical

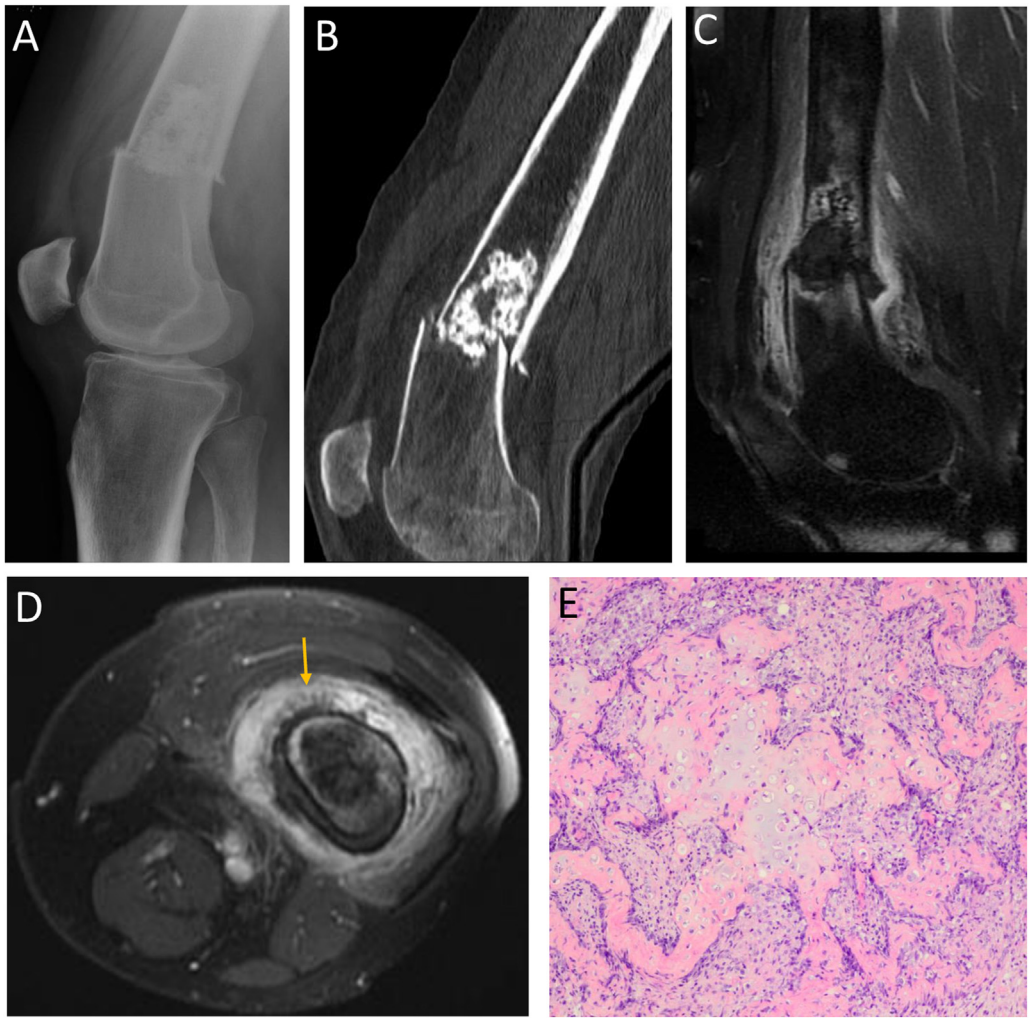

FIG 19. Knee radiograph (A) and CT (B) show a bone lesion with flocculent calcifications in the metaphyseal region of the femur with pathological fracture. FS PD (C) shows a bone lesion with perilesional edema and fracture line. FS T1WI with contrast (D) confirm the same findings and periosteal reaction (arrow). Histology of the lesion (E) showed chondrosarcoma juxtaposed to a high-grade noncartilaginous sarcoma, findings compatible with dedifferentiated chondrosarcoma (H-E stain). (Color version of figure is available online.)

signal on MRI includes heterogeneous low-intermediate SI in T1WI, heterogeneous high SI in T2WI and heterogeneous enhancement after gadolinium administration (Fig 21). Heterogeneous signal is due to hemorrhage or to chondroid mineralization. It is not uncommon for these tumors to show cystic changes with fluid levels that lead to their misdiagnosis as cystic bone lesions. Perilesional bone marrow edema is unusual and mild, which allows distinguishing them from chondroblastomas. Clear cell chondrosarcomas are relatively slow growing, low-grade malignant tumors that have a much better prognosis than conventional chondrosarcomas. However, metastases have been reported.[42]

Conclusion

Detecting cartilage tumor malignancies is essential to allow early adequate treatment. Distinguishing between benign and malignant chondroid lesions can be challenging, both radiographically and pathologically. Diagnostic biopsy is unreliable in assessing the genuine histological grade and malignant potential of chondrosarcomas. Therefore, physicians need to rely on imaging and clinical findings.

The most recent update of the WHO classification reflects the importance of a correct and early radiological diagnosis of newly benign (chondroblastoma and chondromyxoid fibroma) and locally aggressive lesions (synovial chondromatosis). This change highlights the relevance of early surgical treatment of synovial chondromatosis in order to reduce the risk of recurrence and degenerative changes. According to the updated WHO classification, the location of chondral tumors is crucial, and lesions in the axial skeleton are more likely to be malignant.

Due to the increase in patients undergoing MRI examinations the incidental detection of enchondromas and ACTs has increased substantially. Treatment of ACTs has assumed less aggressive surgical management over the last 20 years and new trends propose active surveillance. These insights make the imaging differentiation between enchondroma/ACT and high-grade chondrosarcoma clinically relevant.

Correct diagnosis of chondrosarcoma grade is crucial for determining both treatment and prognosis. It remains essential that decisions about treatment are made by a multidisciplinary team. Understanding and recognizing the spectrum of appearances of the

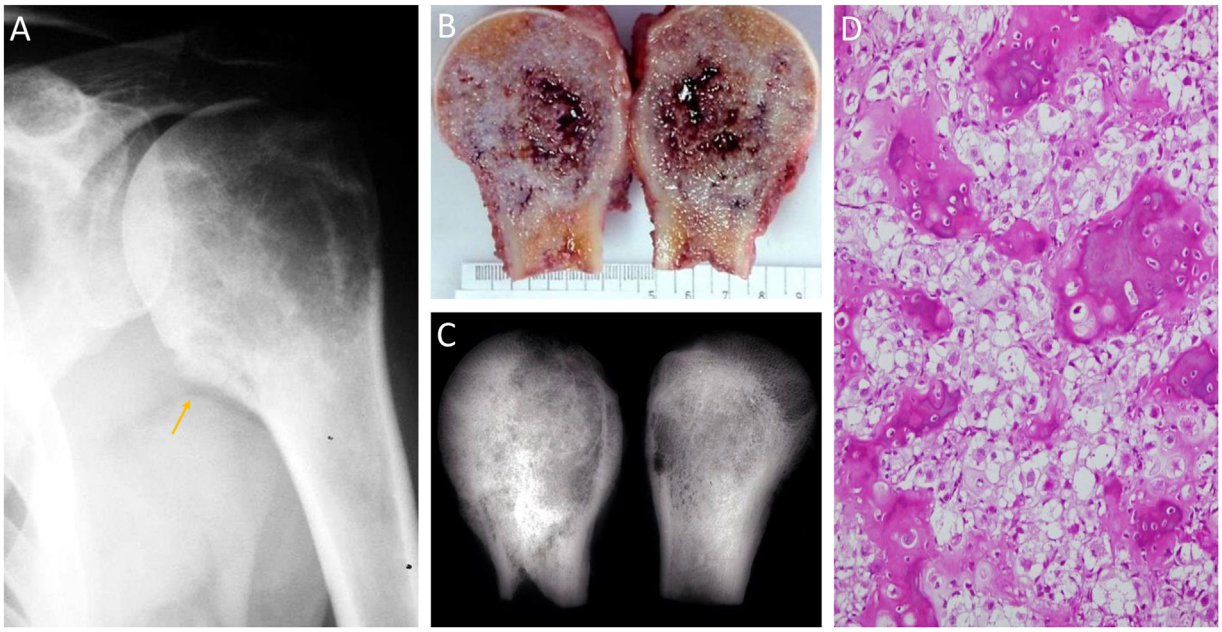

FIG 20. A. 43-year-old male with pain in the left shoulder. The radiograph (A) shows an intraosseous lesion in the metaphysis of the humerus, with periosteal reaction in the inner aspect (arrow). After the diagnostic biopsy, a wide resection of the lesion was performed (B) with gross specimen radiograph (C). Anatomopathology (D) revealed clear cells with abundant cytoplasm admixed with trabeculae of woven bone, consistent with clear cell chondrosarcoma (H-E stain). (Color version of figure is available online.)

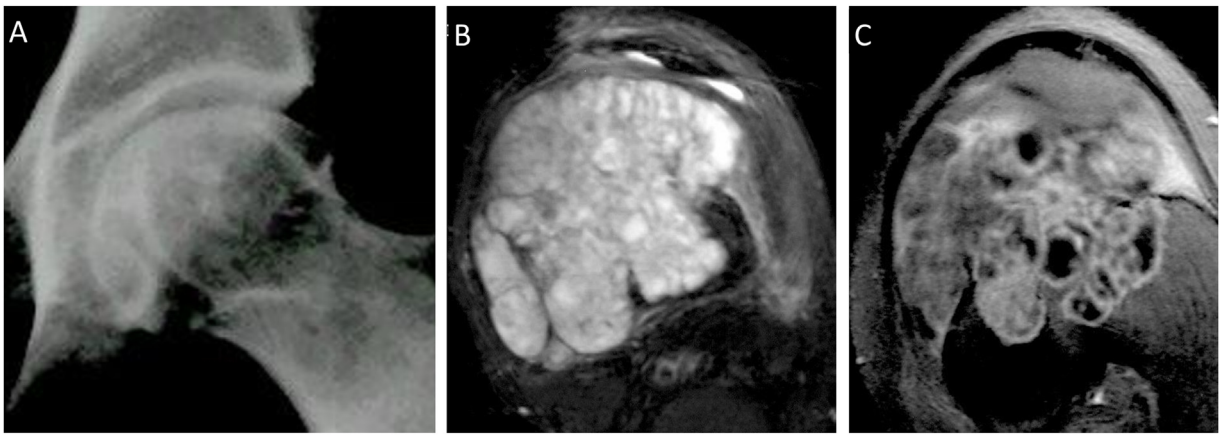

FIG 21. Plain radiograph (A) demonstrates a lytic lesion with partial sclerotic margins and chondral calcifications located in the central region of the left femoral head. MR shows a lobulated lesion with high signal on T2WI (B) and small foci of signal absence due to the calcifications. Intense peripheral enhancement (C) is shown on FS T1WI with contrast. Anatomopathology (not shown) revealed a clear cell chondrosarcoma.

various types of benign and malignant chondroid tumors allow improved patient assessment and are vital for optimal clinical management.

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