12-Year-Old Boy With Left Knee Pain

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This case is presented to illustrate the imaging and clinical findings of a condition of interest to orthopedic surgeons. The initial findings are noted on the first 2 pages, along with diagnostic considerations and differential diagnoses as additional information is obtained as the clinical investigation proceeds. The correct diagnosis is discussed beginning on the third page.

Case Presentation

A 12-year-old boy was referred for evaluation of left knee pain and inability to bear weight because of the pain. He reported experiencing the pain for a month, and he rated it 3 to 6 on a subjective pain scale (10 points maximum). He had injured the knee jumping off a diving board 4 weeks before presentation, and he had not received any treatment for the condition or taken any pain medication. He denied any history of fever, chills, or prior knee pain. His previous medical history was otherwise normal.

On examination, the patient ambulated with an antalgic gait. The left knee showed some soft-tissue swelling in the prepatellar region but no effusion. The superolateral pole of the patella was tender to deep palpation. There was some warmth over the area, compared with the right knee, but no erythema. The examination did not show any signs of ligamentous instability.

Left knee flexion was limited to 90° with full knee extension. Pain was elicited with flexion. There were no signs of patellar instability. The patient was able to straight-raise the left leg. Standard blood tests (including complete blood cell count and erythrocyte sedimentation rate) were normal.

Plain films (Figures 1A, 1B), magnetic resonance imaging (MRI) scans (Figures 2A, 2B), and computed tomography (CT) scans (Figures 3A, 3B) of the left knee were obtained.

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☐ **Given the history, physical examination, and imaging studies, the differential diagnoses at this point are aneurysmal bone cyst (ABC), chondroblastoma, enchondroma, giant cell tumor (GCT), solitary bone cyst, and subacute osteomyelitis.**

Radiographic Interpretation

Plain films (Figures 1A, 1B) showed a radiolucent lesion in the superolateral aspect of the patella with minimal reactive sclerotic rim. An associated bipartite patella was also noted. T_2 MRI scans (Figures 2A, 2B) showed a

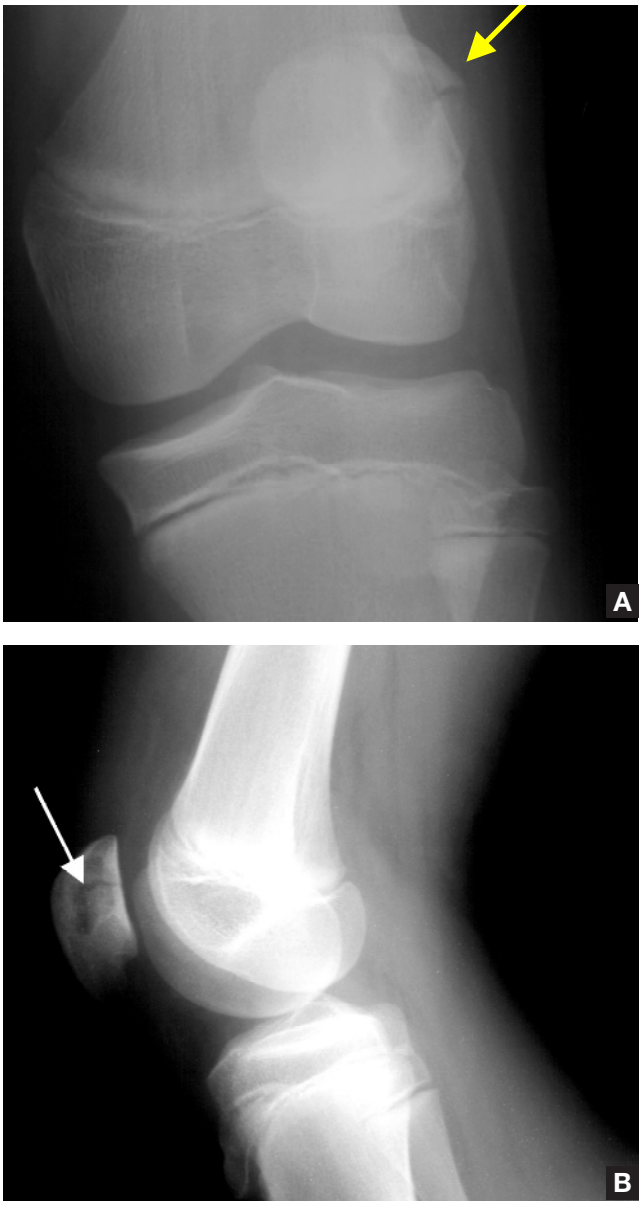

Figure 1. Anterior plain film **(A)** and lateral roentgenogram **(B)** of the left knee.

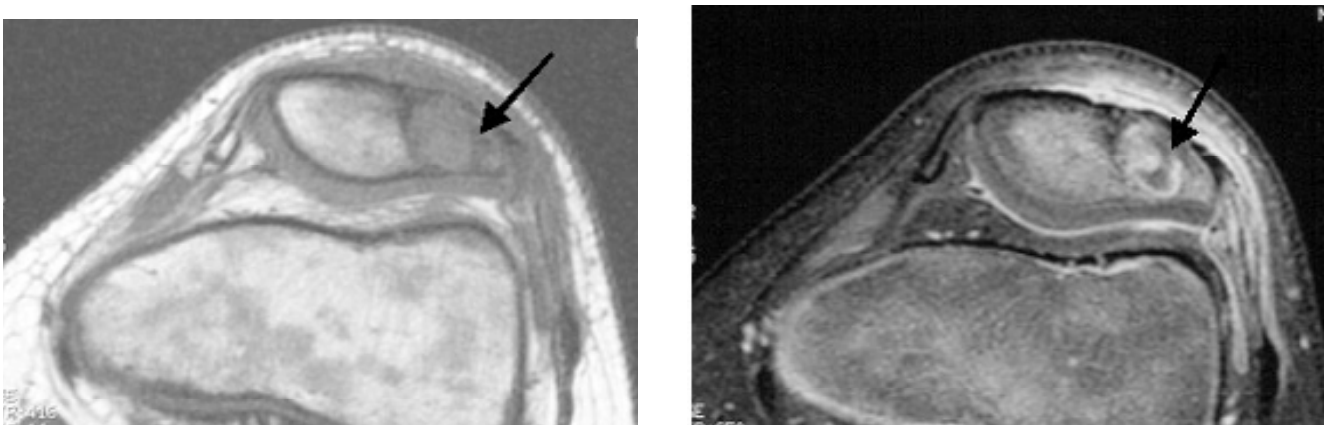

Figure 2. Magnetic resonance imaging scans of the left knee: (A) axial T_1-weighted and (B) T_2-weighted.

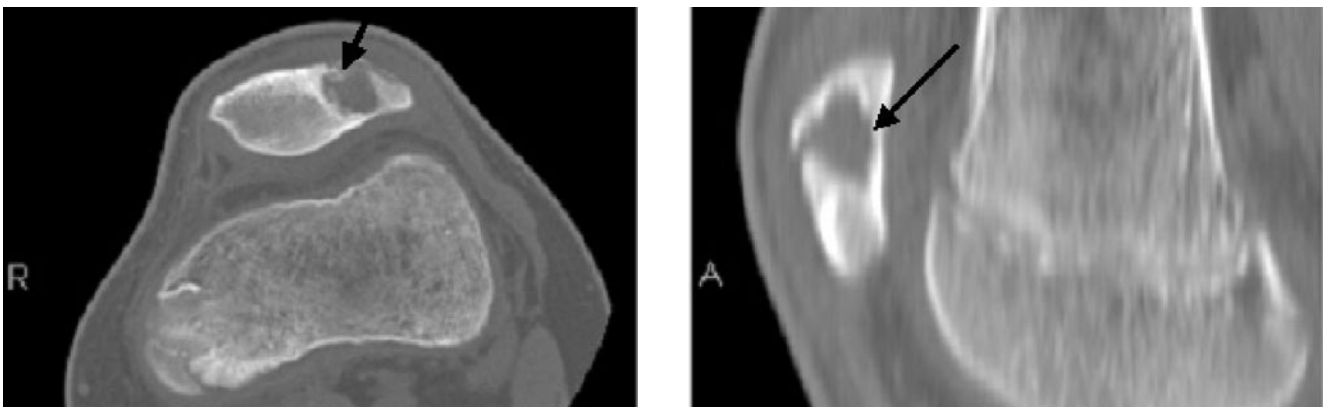

Figure 3. Computed tomography scans of the left knee: **(A)** axial and **(B)** sagittal.

hypointense lesion with some edema extending into the soft tissues and no associated soft-tissue mass. CT scans (Figures 3A, 3B) showed a well-defined lytic lesion in the left lateral patella with anterior cortical disruption measuring 1.2x1.2x1.4 cm in maximum diameter. Nonaggressive periosteal reaction indicated an old pathologic fracture. There was minimal soft-tissue swelling surrounding the patella.

Given the history, physical examination, imaging studies, and laboratory studies, the differential diagnoses are aneurysmal bone cyst, giant cell tumor, enchondroma, chondroblastoma, and simple bone cyst.

Histologic Interpretation

An open biopsy was performed, and an intraoperative frozen section was obtained. Grossly, the biopsy specimen consisted of multiple irregular fragments of pink-tan to red soft and bony tissue. Histologically, the tumor was composed of relatively small, round to polygonal cells with eosinophilic cytoplasm and angular or grooved nuclei set in a fairly abundant predominantly pink chondroid matrix. In some areas, calcific deposits lined lacunar spaces around tumor cells. Focal osteoid formation and infarction were also noted (Figure 4).

Given the history, physical findings, radiographic studies, and histologic picture, what is the diagnosis, and how should this lesion be treated? The correct diagnosis is given on the next page, with a discussion.

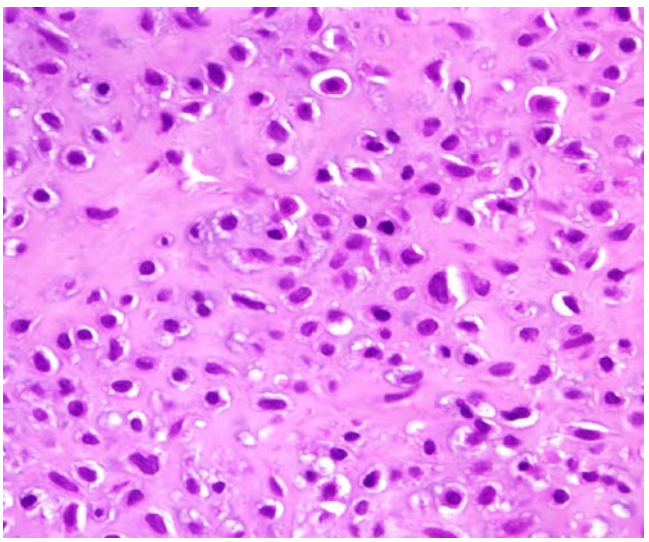

Figure 4. High-power photomicrograph of biopsy specimen (hematoxylin-eosin, original magnification x 200).

Correct Diagnosis, Treatment and Discussion

Chondroblastoma is the correct diagnosis.

Chondroblastoma, a rare, benign bone tumor first described by Ewing in 1923,[1] represents 1% to 3% of all primary tumors. Although most chondroblastomas occur between the ages of 10 and 30 years, age of onset ranges from 3 to 73 years. There is a male predominance, 2:1 to 3:1. The most common sites of involvement are the epiphyseal and epimetaphyseal regions of the distal and proximal femur, proximal tibia, and proximal humerus; other sites (eg, acetabulum, ilium, talus, calcaneus, skull) have also been documented.[1,2]

In 1963, benign chondroblastoma in the knee was first described by Cohen and Cahen.[3] Chondroblastoma accounts for 1% of all tumors in the patella but in some series has been the tumor most commonly found at that site.[4] Mercuri and colleagues[5] collected 279 cases of benign tumors of the patella including 126 GCTs, 61 chondroblastomas, 19 chondromas, 15 primary ABCs, 14 bone cysts, 10 osteoid osteomas, 9 osteoblastomas and osteochondromas, 8 hemangiomas, 1 osteoma, and 1 nonossifying fibroma. In the literature, we found only 6 case reports addressing the association of chondroblastoma of the patella and microfractures.[4-10]

The clinical presentation of chondroblastoma in this area is highly variable.[1] Most patients initially present with localized pain. Less frequent findings include soft-tissue swelling, difficulty walking, and movement limitation.[1,3,11] Joint effusion, especially around the knee, is not uncommon. There is no known relationship to trauma.[1] Our patient's history of trauma to the left knee before symptom onset was probably a coincidence.

On plain films, typically a patient with chondroblastoma presents with a small central or eccentric osteolytic lesion, with or without a thin sclerotic rim in the epiphysis of the long bones.[1,11-13] Lesions as large as 19 cm have also been reported.[2] Metaphyseal involvement is recognized in 25% to 50% of cases, but this is by extension rather than primary origin.[1] Punctuate calcifications within the lesion may be seen on plain film and CT. The calcific foci and opacity caused by septa are best differentiated on CT. CT scans can be useful in identifying extent and proximity to articular cartilage and the physeal plate.[11,13-15]

MRI shows a low signal on T_1-weighted images and a variable, usually hypointense signal on T_2 images. The intensity of the imaging signal correlates with the histologic characteristics of the lesion, including abundance of immature chondroid matrix, chondroblastic hypercellularity, calcification, and hemosiderin deposition.[1] No soft-tissue mass is present unless cortical breakthrough has occurred.

Pathologic assessment of chondroblastoma is essential.[1] The typical picture is represented by a well-circumscribed, blue-gray to gray-white tumor with focal areas of yellow calcification, necrosis, or both. The tumor may be soft, firm, rubbery, or friable, and it averages 3 to 6 cm in size. Chondroblastomas are slow-growing tumors; differences in evolutionary phase account for their wide morphologic spectrum.[1,9,16,17] Hypercellularity occurs early, followed by necrosis. In time, the necrosis is replaced by fibrous or chondroid areas with occasional spindle cells. Some lesions have prominent osseous metaplasia and rich vascularity and thus are easily confused with osteoblastomas. The typical chondroblastoma is characterized by sheets of round to polygonal cells with distinct cytoplasmic borders; eosinophilic cytoplasm; and oval, round, or uniform nuclei, which may be indented or cleaved. Infrequently, larger cells with hyperchromatic nuclei may be identified. However, significant cellular atypia is absent. Some lesions may have a spindle cell component that represents spindle-shaped mononuclear cells or reparative cells of fibroblastic origin. Multinucleated osteoclast-type giant cells are interspersed within the polygonal cells or may be concentrated in areas of hemorrhage or necrosis. These giant cells are different from the larger, more abundant kind that occur in GCTs. Calcification, seen in one third of cases, occurs linearly around the individual neoplastic cells, creating a characteristic "chicken-wire" pattern. Cystic changes are also commonly seen in chondroblastoma. A lesion with a unilocular cyst or multilocular cysts is termed a *cystic chondroblastoma*. Secondary ABC formation is found in 20% to 25% of cases. Conversely, when ABC formation is diagnosed, particularly in a young patient, presence of an underlying chondroblastoma should be ruled out.[18] Presence of chondroid-like, osteoid-like, or fibrous matrix—an important diagnostic feature—was the subject of a recent study, by Aigner and colleagues,[18] sputing the histogenesis and current concept of chondroblastoma. Those investigators indicated that chondroblastomas are bone-forming rather than cartilage-forming tumors. Other less frequent histologic findings include hemosiderin deposition, cortical and soft-tissue invasion, vascular invasion, and presence of myxoid areas.[1,7]

Other Conditions in the Differential Diagnosis

The differential diagnosis of a solitary cystic lesion of the patella should include (in addition to chondroblastoma) GCT, solitary bone cyst, ABC, and enchondroma.[10,17,19-21]

Giant cell tumor can be mistaken for a chondroblastoma.[22,23] However, GCTs occur almost exclusively in skeletally mature patients. Most GCTs are located within the epiphyses of long bones, but often they extend into the metaphysis.[1] Histologically, giant cells in GCT are generally more clustered, larger, and more numerous than those found in chondroblastoma.[1] Radiographically, GCT appears more destructive.[10,22,23]

Solitary bone cyst arises in the metaphyseal region of long bones. Histologically, the walls or even small masses of fibrous tissue that are characteristic of solitary bone cyst are not seen in chondroblastoma.[5,10]

Aneurysmal bone cyst has been found in patellar bone.[1] Abellar and colleagues[1] reported the case of an unusual association of chondroblastoma and secondary ABC formation. Histologically, multiple small vascular lakes surrounded by loose spindle cells with scattered multinucleated giant cells and hemosiderin deposition are typically observed.[10] MRI helps to differentiate several fluid/fluid levels and solid tumor components. Mercuri and colleagues[5] reported 9 cases of chondromas in a group of 279 patients with benign tumors of the patella. Chondroma is characteristically composed of confluent masses of bluish, semitranslucent hyaline cartilage in a lobular arrangement. Histologically, the chondrocytes are small cells within lacunar spaces and have a round, regular nucleus similar to the nucleus of the chondrocyte seen in nonneoplastic hyaline cartilage.[10,21]

Enchondroma, a relatively common bone tumor, is usually located in the small bones of the hands and feet.[10]

Our patient's knee pain and physical findings raised the suspicion of a possible bony lesion. The plain films, CT scans, and MRI scans of the left patellar lesion showed a marginated lytic lesion with sclerotic rim. Moreover, there was a bipartite patella on the same side.

Treatments and Outcomes for Chondroblastoma

The standard surgical treatment for chondroblastoma is extended intralesional excision with or without adjuvants such as phenol and grafting.[12,16,21] Chemotherapy and radiotherapy currently have no role in the treatment of chondroblastomas. Radiotherapy was previously used for unresectable lesions. Because of the 14% recurrence rate (15%-25% with intralesional resection), care is taken not to penetrate the adjacent joint, except when better visualization or exposure is required.[14-19] Even though the incidence of pulmonary metastasis is less than 1%, serial chest plain films are recommended every year for 5 years.[6] Mean age at pulmonary metastasis is 18.5 years.[5,20] Recurrence is attributed to risk factors such as open epiphyseal plates and an ABC component.[9] There is a rare report of sarcomatous transformation secondary to radiotherapy.[14]

In our patient's case, surgery was the chosen treatment. Curettage, cauterization, high-speed burring, and phenolization with 5% phenol were performed. The lesion was grafted with commercially available demineralized bone matrix allograft cancellous cubes and demineralized bone allograft or paste. Clinical evaluation of the left knee 7 months after surgery revealed resolution of pain, full range of motion in the knee, and a well-healing surgical wound with no signs of recurrence. Plain films obtained at that visit showed a healed patella and no signs of recurrence. The patient had returned to his preinjury level of activity.

Chondroblastomas are rare, benign tumors of bone with distinctive radiologic and pathologic characteristics. In children, the highly variable and sometimes nonspecific clinical presentation and radiographic findings of chondroblastomas require a multidisciplinary evaluation-and-management approach.

Author's Disclosure Statement and Acknowledgement

The authors report no actual or potential conflict of interest in relation to this article.

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