Title

Primary Synovial Chondromatosis of the Knee in a 12-Year-Old Boy: A Case Report

Journal

UCLA Radiological Sciences Proceedings, 4(1)

Authors

McNulty, Catherine A

Narasimhalu, Tara

Ghahremani Koureh, Shahnaz

Publication Date

2024

DOI

10.5070/RS44153838

Radiological Sciences Proceedings

UCLA Health

Primary Synovial Chondromatosis of the Knee in a 12-Year-Old Boy: A Case Report

McNulty C, MD[1] | Narasimhalu T, MD[2] | Ghahremani Koureh S, MD[1]

Author Affiliation: [1] Department of Radiological Sciences, David Geffen School of Medicine at UCLA
[2] Department of Pathology and Laboratory Medicine, David Geffen School of Medicine at UCLA

Corresponding Authors: C.M. (CAMcnulty@mednet.ucla.edu)

UCLA Radiol Sci Proc. 2024;4(1):1-5

Abstract: Primary synovial chondromatosis is an uncommon, benign synovial proliferative disorder rarely seen in children. The most commonly affected joint is the knee, followed by the hip. The symptoms of this condition are progressive and insidious and may include pain, joint stiffness, and swelling. Radiography is an appropriate first imaging modality that typically shows multiple calcified loose bodies within the affected joint space. Magnetic resonance imaging should then be performed to evaluate for the presence of noncalcified cartilaginous loose bodies and soft tissue involvement prior to surgical treatment that is necessary to prevent permanent joint damage. The recurrence of primary synovial chondromatosis is common, especially in cases of incomplete surgical excision. We report a case of primary synovial chondromatosis in a child. The diagnostic and the treatment considerations of the case are briefly discussed.

Keywords: *synovial chondromatosis, loose bodies, chondrosarcoma*

Key Points

- Primary synovial chondromatosis is an uncommon, benign synovial proliferative disorder that most commonly affects the knee joint and is exceptionally rare in children.
- Radiologically, primary synovial chondromatosis appears as multiple, mostly calcified/ossified loose bodies within the joint space.
- Synovial chondromatosis is treated by surgical excision of loose bodies and often by complete synovectomy to prevent permanent joint damage.

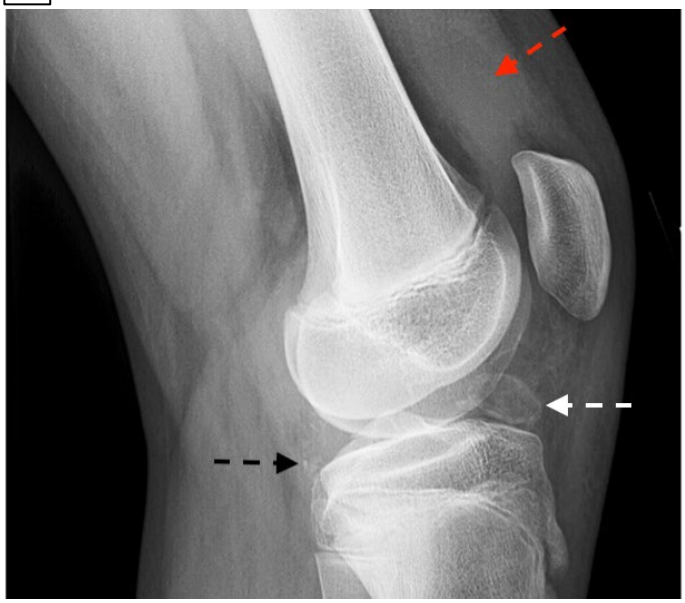
Case Presentation

A 12-year-old boy presented to their primary care physician with a 6-month-long experience of symptoms of intermittent pain in the left knee as well as stiffness and a locking sensation when ambulating. The patient denied prior trauma or episodes of joint pain. Physical examination revealed no obvious abnormalities, and the patient was treated conservatively with rest, ice, compression, elevation, and physical therapy. Despite adherence to the recommended therapy, the patient returned to his physician six months later with recurrent knee swelling in addition to the previous symptoms. At this time, physical examination revealed left knee effusion and a palpable, firm mass behind the patellar tendon. Radiography of the left knee revealed a round, 2.5-cm calcified, dense formation in the anterior joint space, slightly hyperdense spots in the posterior joint space, and a small effusion in the suprapatellar region (Figure 1A). These findings were suggestive of synovial chondromatosis.

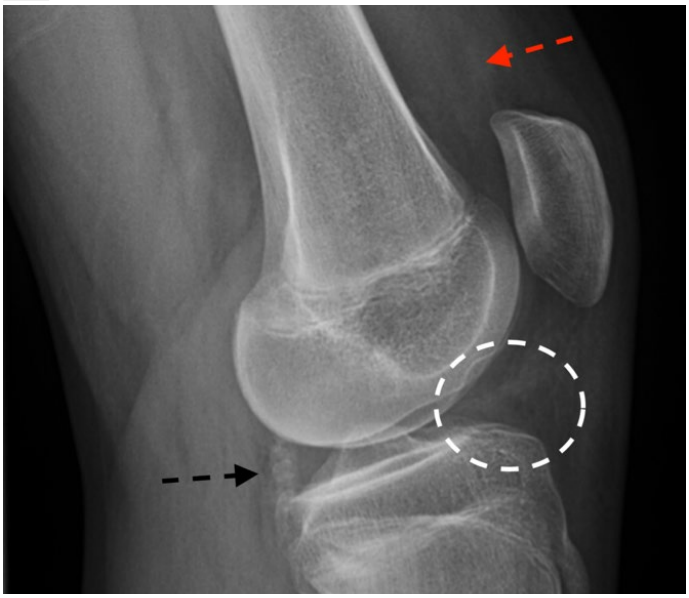
The patient was referred to an orthopedic surgeon and underwent magnetic resonance imaging (MRI) for preoperative planning. The imaging findings (Figures 2A and 2B) were consistent with synovial chondromatosis, and the patient underwent

Figure 1. Radiography of the Left Knee of a 12-Year-Old Boy with Primary Synovial Chondromatosis

A Initial radiograph, lateral view



B Postoperative radiograph, lateral view



(A) The initial radiograph, lateral view, reveals a round 2.5-cm calcified loose body in the anterior tibiofemoral aspect of the knee joint (A, white arrow), slightly hyperdense spots in the posterior aspect of the knee joint (A, black arrow), and an effusion within the suprapatellar recess (A, red arrow).
(B) The postoperative radiograph shows the anterior tibiofemoral aspect of the knee (B, white circle) cleared of the calcified loose body. A decrease in the size of the effusion (B, red arrow) is seen in the suprapatellar region. There are spots of increased radiographic density within the posterior aspect of the knee joint (B, black arrow), indicative of recurrence of synovial chondromatosis.

exploratory and therapeutic arthroscopy with removal of several loose bodies from the medial and the lateral gutters as well as extraction of a 2.5 x 1.5 x 1.5-cm osteochondral body that was scarring the infrapatellar fat pad. Histopathologic examination of removed specimens revealed mature cartilage with zones of ossification (Figure 3). As the patient did not have evidence of degenerative changes of the knee joint, the diagnosis was primary synovial chondromatosis.
The patient recovered from surgery without complications and no longer reported left knee discomfort, swelling, or locking. Multiple postoperative follow-up radiographic examinations of the left knee have revealed dense formations that were widening the posterior space of the tibiofemoral joint (Figure 1B). Given the patient's lack of symptoms, the decision has been made to monitor the patient's condition with annual radiographic examinations and to repeat surgical intervention if knee discomfort recurs.

Discussion

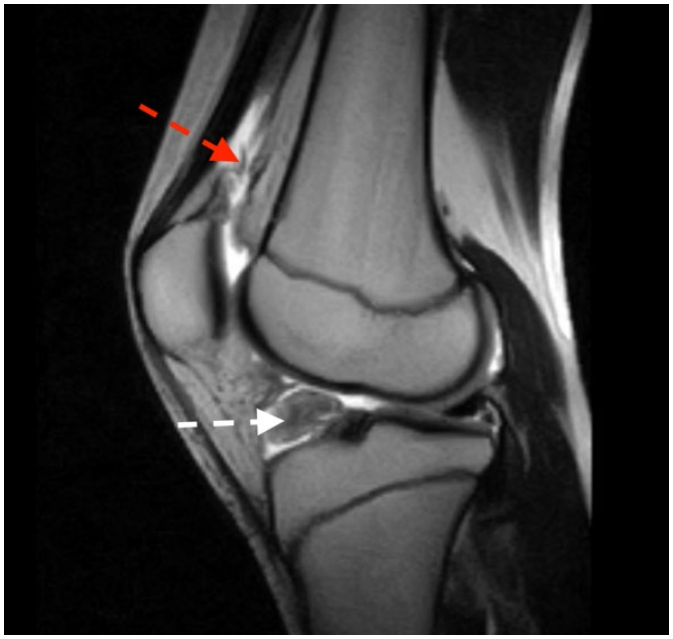
Synovial chondromatosis is a benign neoplasm characterized by the formation of cartilage nodules within the joint space.[1,2] This disease most commonly affects the knee,[1-3] followed by the hip.[1,3] Other typically involved joints include the elbow, the shoulder, and the ankle.[1,3] Primary synovial chondromatosis is exceedingly rare, and its causes are unknown.[1,2] Secondary synovial chondromatosis is caused by underlying joint abnormalities, including osteoarthritis, trauma, and infections.[1,4] Both subtypes rarely occur in children and more often affect males aged 20 to 50 years.[1,4] The symptoms of synovial chondromatosis are progressive and insidious and may include pain, joint stiffness, and swelling,[4] such as were observed in our patient. The results of physical examination are nonspecific and commonly reveal a palpable mass, crepitus, locking, muscle atrophy, and/or other evidence of joint destruction.[1,4]
As described in this case report, in addition to the patient's medical history and physical examination, imaging is an important part of the initial evaluation of patients with synovial

chondromatosis. Although radiography may be the first imaging modality to diagnose synovial chondromatosis, MRI is often used to detect noncalcified nodules, extracapsular spread of the disease, and the extent of involvement of the muscle, the bursa, or the fascia.[1,3] On imaging, innumerable, similarly sized calcified bodies are more specific for primary synovial chondromatosis,[1,3,5] whereas a small number of variably sized calcified bodies is characteristic of secondary synovial chondromatosis.[1,3] The appearance of synovial chondromatosis on MRI varies depending on the degree of calcification of intra-articular bodies; it is most commonly characterized by a high-intensity signal on T2-weighted images and a low-to-medium-intensity signal on T1-weighted images.[1,3,5] The imaging findings observed in our patient—multiple calcified loose bodies within the knee joint and a small knee effusion—are consistent with synovial chondromatosis. A lesion with focal ossification was identified on both radiographic images (Figure 1) of our patient and photomicrographs (Figure 3) of surgically removed specimens from the patient's left knee. Except for this lesion, the other loose bodies, consisting mainly of cartilage, were radiographically occult and therefore better assessed with MRI.

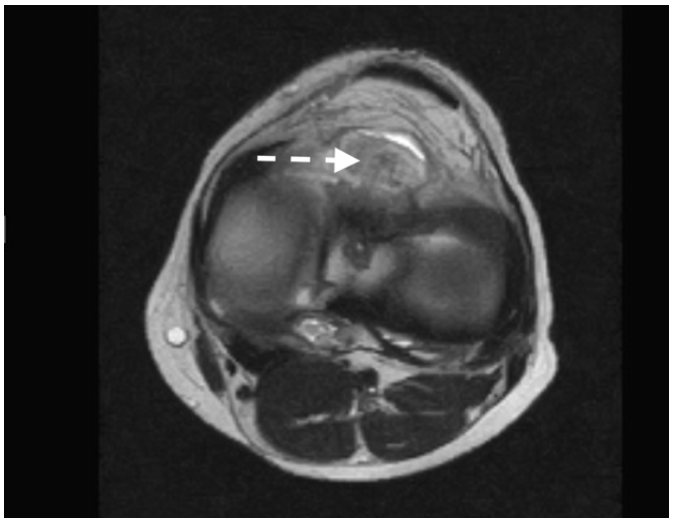
In children and adolescents with persistent pain, stiffness, and swelling in a monoarticular joint (the symptoms suggestive of primary synovial chondromatosis), differential diagnosis should include monoarticular juvenile idiopathic arthritis (JIA),[4] chondrosarcoma,[1] diffuse and localized types of tenosynovial giant cell tumor (tGCT), and synovial hemangioma.[6] Junaid et al[7] provided comprehensive analysis of imaging appearances of intra-articular tumors and tumor-like lesions in children and adolescents. According to this analysis, a large effusion of the joint is pathognomonic for both diffuse tGCT and JIA. In cases of diffuse tGCT affecting the knee joint, this effusion, the result of widespread villous and/or nodular synovitis that appears hypointense on T2-weighted images, extends into a popliteal cyst, whereas in cases of JIA, the effusion is accompanied with synovial thickening that is diffusely enhanced on MRI. In cases of localized tGCT, the lesion appears within the joint as a well-defined, solitary mass with intermediate-to-low-intensity signal. According to the authors, synovial hemangioma, the second most common neoplastic lesion after tGCT that affects children and adolescents, is characterized by a poorly defined,

Figure 2. Magnetic Resonance Imaging (MRI) of the Left Knee of a 12-Year-Old Boy with Primary Synovial Chondromatosis.

A T2-weighted image, sagittal view



B T2-weighted fat-saturated image, axial view



(A) T2-weighted, sagittal view, and (B) T2-weighted fat-saturated, axial view, MR images of the left knee reveal an intra-articular loose body (A and B, white arrow) in the anterior compartment of the knee joint, with signal intensity similar to that of the bone. (A) A small effusion (A, red arrow) is seen within the suprapatellar recess.

lobulated, capillary or cavernous mass with fluid signal intensity and a serpentine or septate appearance on MRI.[7]

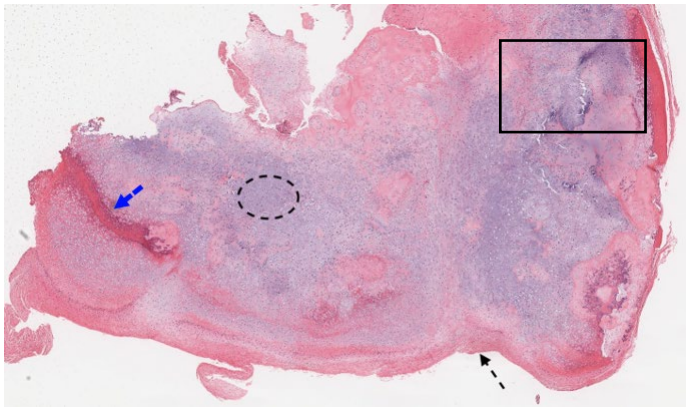
Differentiating primary synovial chondromatosis from chondrosarcoma is challenging because radiologically both lesions might have a remarkably similar appearance.[1] In addition, it is difficult to diagnose chondrosarcoma on histopathologic examination alone.[1,8] However, in cases of a rapidly growing lesion, the histopathologic signs of necrosis and chondrocytes arranged in sheets as well as the signs of destruction of the bone cortex and invasion of the bone marrow seen on both histopathologic examination and MRI are suggestive of chondrosarcoma.[1]

Although nonsteroidal anti-inflammatory drugs may help reduce the symptoms of inflammation and pain in the early stages of synovial chondromatosis, the typical treatment of this condition is the surgical removal of the cartilaginous bodies and, in some cases, a complete synovectomy.[4,9] Recurrence is common, particularly in primary synovial chondromatosis, and seems to be directly correlated with incomplete surgical resection.[4,6] If left untreated, synovial chondromatosis may result in permanent joint damage.[1,4]

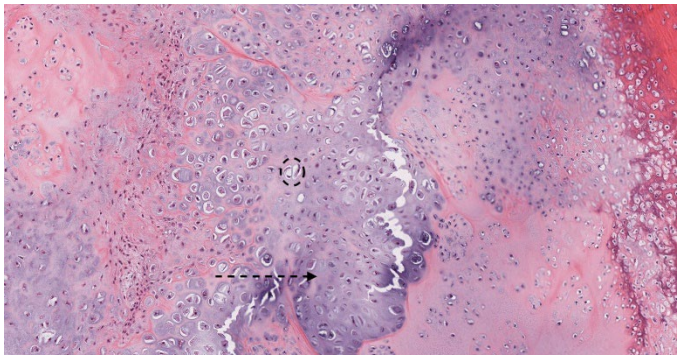
Synovial chondromatosis in children and adolescents is exceedingly rare. It can be misdiagnosed and requires the radiologist to have the detailed knowledge of imaging appearances of the lesions as well as the ability to correlate radiologic, clinical, and histopathologic data to expedite the diagnosis and to facilitate the beginning of appropriate treatment to prevent degenerative changes in the affected joints.

Figure 3. Histopathologic Examination of a Specimen Surgically Removed from the Left Knee of a 12-Year-Old Boy with Synovial Chondromatosis.

A Hematoxylin-eosin staining



B Hematoxylin-eosin staining, detail from the inset of photomicrograph (A)



(A) Photomicrograph of the specimen, with inset (A, rectangle) drawn, shows a lobulated osteochondral loose body within the synovium (A, black arrow), consisting of cartilage (A, black circle) with focal endochondral ossification (A, blue arrow) (hematoxylin-eosin, original magnification x20). (B) Detail from the inset of photomicrograph (A), at 100x magnification, shows the typical nodular architecture of synovial chondromatosis, a matrix of pale blue hyaline cartilage (B, black arrow) with inclusion of the cartilage-producing chondrocytes (B, black circle).

Author Contributions

Conceptualization, S.G.K.; Acquisition, analysis, and interpretation of data, C.M. and T.N.; Writing – original draft preparation, C.M.; Review and editing, S.G.K. and C.M.; Supervision, S.G.K. All authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All authors had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis.

Disclosures

None to report.

References

1. Murphey MD, Vidal JA, Fanburg-Smith JC, Gajewski DA. Imaging of synovial chondromatosis with radiologic-pathologic correlation. *Radiographics*. 2007;27(5):1465-1488. doi:10.1148/rg.275075116

2. Garner HW, Bestic JM. Benign synovial tumors and proliferative processes. *Semin Musculoskelet Radiol*. 2013;17(2):177-178. doi: 10.1055/s-0033-1343095
3. Ballehr LO. Primary Synovial Chondromatosis. Radsource LLC; MRI Web Clinic. July 2015. Accessed March 7, 2021. https://radsource.us/primary-synovial-chondromatosis/
4. Giancane G, Tanturri de Horatio L, Buonuomo PS, Barbuti D, Lais G, Cortis E. Swollen knee due to primary synovial chondromatosis in pediatrics: a rare and possibly misdiagnosed condition. *Rheumatol Int.* 2013;33(8):2183-2185. doi: 10.1007/s00296-012-2382-8
5. Cantarelli Rodrigues T, Serfaty A. MRI Assessment of Benign Tumor And Tumor-Like Synovial Diseases. *Semin Roentgenol*. 2022;57(3):191-200. doi: 10.1053/j.ro.2022.02.005
6. Al-Mohrej OA, Al-Jarallah SA, Al-Dakhil Allah HH, Pant R, Al-Zayed ZS. Synovial sarcoma presenting as an intra-articular mass in a pediatric patient: a case report. *BMC Musculoskelet Disord*. 2020;21(1):283. Published 2020 May 7. doi: 10.1186/s12891-020-03312-3
7. Junaid S, Gnanananthan V, Malhotra K, Saifuddin A. Tumours and tumour-like lesions of joints: Differential diagnoses in a paediatric population compared to adults. *Br J Radiol*. 2021;94(1120):20201389. doi: 10.1259/bjr.20201389
8. O'Connell JX. Pathology of the synovium. *Am J Clin Pathol*. 2000;114(5):773-784. doi: 10.1309/LWW3-5XK0-FKG9-HDRK
9. Zacharia B, Inassi J, Paulose S. Retrospective analysis of the clinical and radiological profile of few cases of synovial osteochondromatosis with a literature review. *Eur J Rheumatol*. 2021;8(1):40-45. doi: 10.5152/eurjrheum.2020.20132