

The Creative Clinician

Synovial Chondromatosis

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LOOSE BODIES IN THE JOINT SPACE MAY BE DUE TO A NUMBER OF conditions. Loose bodies composed primarily of fibrin, "rice bodies", are due to bleeding into the joint or to synovitis as in rheumatoid arthritis or tuberculosis.¹ Cartilaginous or osteocartilaginous loose bodies are most commonly secondary to injury to the joint cartilage from trauma or fracture, or degeneration of the cartilage as in osteoarthritis or osteochondritis desiccans.² **Synovial chondromatosis (SC)**, sometimes referred to as primary synovial chondromatosis, is a rare disorder in which cartilaginous or osteocartilaginous loose bodies, usually in large numbers, form in the joint space without an apparent underlying injury to the cartilage or synovium.² Although morphologic overlap with the secondary types may occur, the histopathologic features of loose bodies in SC are usually distinctive, allowing the pathologist to identify them as such, whereas the clinical findings may be ambiguous. Herein we describe a case of SC with typical morphologic and clinical features.

CASE REPORT

A 45 year-old male plumber presented with a two-month history of knee pain, swelling and stiffness. He recalled no injury to the knee, only worsening symptoms over time, and there was no history of preexisting arthritis or joint disease. On physical examination, he had a large joint effusion, which was aspirated, yielding 20-30 cc of serous fluid and resulting in immediate improvement in his symptoms and range of motion. There was no instability of the cruciate, collateral or posterolateral corner ligaments, but the examination elicited symptoms of a probable medial meniscus tear and mild patellofemoral pain. The distal nerve and vascular examination was normal. **Magnetic resonance imaging (MRI)** of the knee confirmed the presence of an extensive tear of his medial meniscus as well as mild degenerative change in his medial and patellofemoral compartments. The patient elected to have arthroscopic repair of the meniscus tear, during which numerous loose bodies, possibly 250 or more, each measuring between two and ten millimeters in greatest dimension, were encountered in the joint space (Figure 1. Arthroscopic image). These were removed by aspiration and lavage, the medial meniscus tear as well as an unsuspected tear of the lateral meniscus were repaired, and the entire synovium, which appeared to be inflamed or reactive, was removed. No injury of the cartilage or patellofemoral joint was identified. A sample of the loose bodies and synovium was submitted for pathologic examination. The patient recovered uneventfully from his surgery, but he eventually developed recurrent pain and effusions in the knee and underwent a second arthroscopic procedure 14 months after his initial operation.

PATHOLOGIC FINDINGS

The specimen obtained from the first operation consisted of about twenty-five ivory-colored cartilaginous nodules, each four to five millimeters in diameter with a smooth, slightly

lobular surface. Microscopically, the nodules were composed of hyaline cartilage matrix containing lobular clusters of chondrocytes (Figure 2). Each of the nodules was surrounded by a shaggy rim of fibrin or a thin mantle of synovium, which occasionally enclosed several coalescing nodules. The chondrocytes displayed mild-to-moderate variation in nuclear size and configuration (Figure 3). A rare nodule demonstrated focal peripheral endochondral bone formation. The specimen from the second procedure contained similar cartilaginous nodules with more extensive ossification as well as a few fragments of hyalinized synovium.

DISCUSSION

In contrast to those that develop secondary to cartilaginous injury, which typically have a concentric lamellar matrix surrounding a nidus of ossified cartilage, the loose bodies of SC have a distinctive lobular configuration with clusters of mildly-to-moderately atypical chondrocytes in a chondroid matrix as

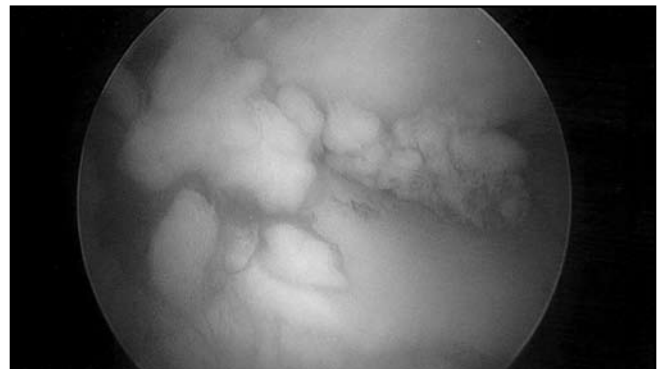


Figure 1. Arthroscopic image of intraarticular loose bodies.

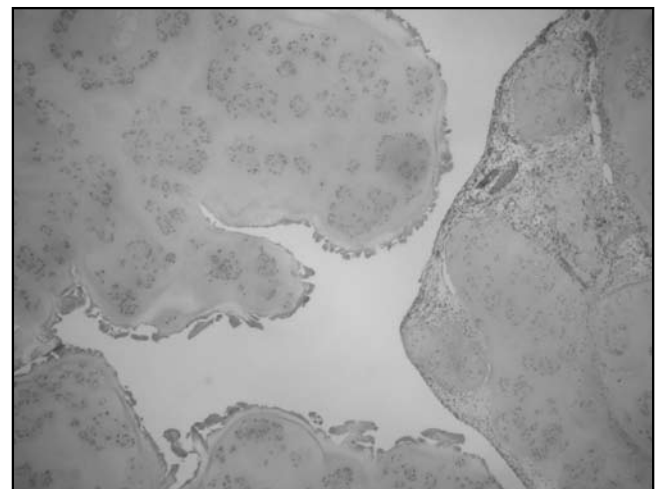


Figure 2. Cartilaginous nodules with lobular arrangement of chondrocytes in hyaline cartilage matrix and surface covered with layer of fibrin or thin mantle of synovium (right).

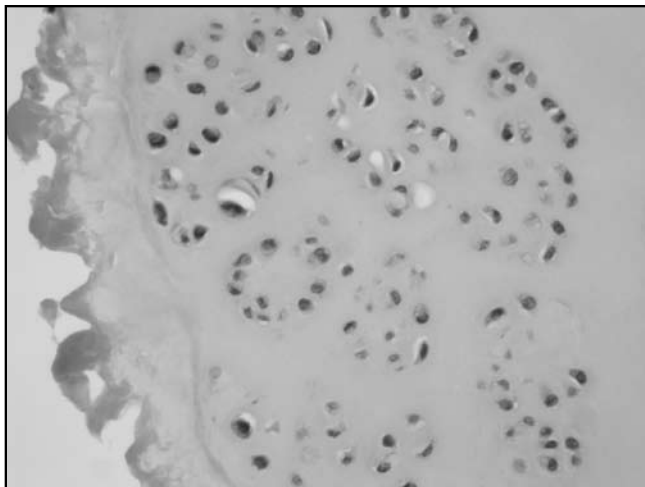


Figure 3. Chondrocytes showing lobular arrangement and mild-to-moderate nuclear atypia.

seen in this case. As the disease progresses, the cartilaginous bodies break free of the synovium eventually becoming ossified. What initiates the disease is not known, although the cartilaginous nodules are believed to develop within the synovium from chondrocytic metaplasia of fibroblasts, perhaps with the influence of **transforming growth factor-beta (TGF- β)** and **bone morphogenic proteins (BMP)**, both of which have been identified in lesional tissue.^{3,4,5} Attempts to identify a genetic event associated with the development of SC so far have yielded inconsistent results,^{6,7} leaving open the question of whether the lesion is neoplastic rather than simply metaplastic in nature; however, malignant transformation of SC, usually to a form of chondrosarcoma with aggressive behavior, is seen in about 5% of cases.⁸

SC usually affects a single large joint, most often the knee or hip, but any joint may be involved, including the joints of the hand and foot or the temporomandibular joint. Roughly twice as many men as women develop SC, and it is rare in children, the average age at presentation being 41 years.⁸ SC is often difficult to detect by x-ray or MRI in the early stages before the cartilaginous nodules become ossified.⁹ Although the symptoms of SC may mimic those of a torn meniscus,¹⁰ the significance of the meniscal tear in this case is uncertain. It has not been previously identified as an associated or complicating factor in SC and may be coincidental. About 5% of patients eventually develop osteoarthritis in the affected joint.⁸

Arthroscopic or open removal of the loose bodies, with or without complete synovectomy, is usually successful in treating SC. Five (11.1%) of 45 knees in 42 patients so treated in two studies required a second arthroscopic procedure.^{11,12} Twenty-three (20.7%) of 111 patients with SC of the hip, 69 of whom had arthroscopy only, required further arthroscopy for recurrence; twenty-two (19.8%) of the 111 patients eventually went on to have total hip replacement.¹³ Arthroplasty may provide significant symptomatic relief, but SC recurred in 1 of 4 (25%) knee and 1 of 7 (14%) hip replacement patients followed by Ackerman et al.¹⁴ Interestingly, radiation therapy has been reported to be successful in the treatment of one case of refractory SC of the knee,¹⁵ but caution regarding this would seem to be advisable.

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Disclosure of Financial Interest

The authors and/or their significant others have no financial interests to disclose.

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