S
ynovial chondromatosis is an uncommon, benign condition characterized by multiple nodules of hyaline cartilage in the subsynovial layer of joints, tendons, or bursal cavities. Three times more common in men than in women,1 it is most commonly diagnosed between the third and fifth decades of life.2

The knee is involved in two thirds of reported adult cases, followed in frequency by the hip, elbow, shoulder, and wrist.2 Extra-articular involvement of synovial chondromatosis is less common, with involvement of the biceps tendon reported only three times in the literature.3-5 Extra-articular involvement has not been reported in children.

This article presents synovial chondromatosis of the left shoulder and biceps tendon sheath occurring in a pediatric patient.

CASE REPORT

A 10-year-old girl presented with left shoulder pain of 1 month’s duration. Symptoms included mild to moderate pain and clicking within the joint occurring with movement, especially with exercise. Pain also occurred at night with rest. No constitutional symptoms or history of trauma to the left shoulder were reported.

Physical examination was remarkable for mildly decreased internal rotation of the left shoulder with tenderness over the biceps tendon and posterior glenohumeral joint, as well as a palpable effusion of the glenohumeral joint. Shoulder abduction was normal. Atrophy was reported, and no masses or lymphatic nodules were palpable. Strength and sensation were normal and symmetric in both shoulders.

Radiographs of the left shoulder demonstrated focal areas of ossification around the glenohumeral joint. Computed tomography (CT) revealed multiple centers of ossification around the glenohumeral joint, as well as similar centers within the biceps tendon sheath (Figure 1).

Based on physical examination and radiographs, a diagnosis of synovial chondromatosis was made. Intraoperatively, multiple cartilaginous loose bodies and diffuse hypertrophic synovitis were evident in the joint, with foci of new loose bodies attached to the synovium (Figure 2). Notably, the articular surface of the humeral head and glenoid showed no evidence of premature arthritis.

Open exploration of the left biceps tendon using a deltopectoral approach revealed multiple loose bodies and synovitis of the biceps tendon sheath (Figure 3). The loose bodies were subsequently removed, and a synovectomy of the biceps tendon was performed. In total, >30 loose bodies of ranging sizes were removed.
from the shoulder and biceps tendon sheath (Figure 4). Histology of intra- and extra-articular loose bodies and synovium confirmed the diagnosis of synovial chondromatosis.

Postoperatively, the shoulder was immobilized in an adduction sling. Physical examination performed 12 months postoperatively revealed only a subtle loss of strength and slight decrease in internal rotation and adduction compared to the nonoperative side. The patient has returned to full activity with no reports of pain or limitation.

DISCUSSION

Although the exact pathogenesis of synovial chondromatosis is poorly understood, it is generally accepted that the process involves a benign reactive metaplasia of the subintimal connective tissue of the synovial membrane. As primitive mesenchymal cells differentiate into mature chondrocytes, nodules of hyaline cartilage form and break off to become loose bodies. These loose bodies, nourished by synovial fluid, grow inside the joint. The central cores of hyaline cartilage may calcify, allowing for visualization on plain radiographs. Histologic evaluation reveals multinucleated chondrocytes arranged in clusters within a hyalinized matrix. Although trauma has been associated with development of this condition, the cause of this reactive metaplasia is unknown.

Characteristic symptoms of synovial chondromatosis include pain and limited range of motion of the affected joint. Swelling, muscle atrophy, palpable nodules, and locking of joints are also common presenting signs and symptoms. Synovial chondromatosis can be complicated by secondary degenerative osteoarthritis caused by loose bodies eroding the surrounding bone.

Imaging techniques for synovial chondromatosis include plain radiographs, CT, and magnetic resonance image (MRI). Murphy et al. found that calcifications were not appreciated in 5%-30% of synovial chondromatosis cases, and Christensen and Poulsen revealed that 12 of 22 patients diagnosed with the disease had normal plain radiographs preoperatively.

Computed tomography and MRI allow visualization of uncalcified cartilaginous bodies and erosions not seen with plain radiographs. In addition, they allow the observer to better confirm the intra-articular position of nodules. Although noncalcified bodies are well demonstrated on arthrography of T2-weighted MRI, in most cases it is believed that a combination of radiographs and CT is superior to MRI alone. The diagnosis must be confirmed using histology by identifying the synovial origin of the cartilaginous proliferation.

Treatment of synovial chondromatosis involves surgical exploration and loose body removal. No long-term intervention studies have assessed outcomes of open versus arthroscopic treatment. The benefits of arthroscopic treatment of synovial chondromatosis have been well outlined in the literature. However, arthroscopy may not always be technically feasible and open surgical treatment may be preferred, especially in cases of extra-articular
involvement where anatomical space may be limited.

Although many authors suggest loose body removal followed by excision of the synovial membrane, studies have yielded conflicting results. After reviewing 26 patients with synovial chondromatosis, Shpitzer et al\(^9\) concluded that no difference existed between those treated with excision of the synovium compared with those with only loose body removal. Ogilvie-Harris and Saleh\(^9\) compared 13 patients who underwent loose body removal or loose body removal with synovectomy and found a recurrence of disease in 3 patients who underwent loose body removal alone, but no recurrence in those treated with loose body removal and arthroscopic synovectomy. Jeffrey\(s,\)\(^{11}\) however, found no recurrence of disease in 5 patients treated with partial synovectomy and in 12 patients managed only with loose body removal, and therefore concluded that both treatments produced the same outcome.

Extra-articular manifestations of synovial chondromatosis are rare, and the only reported cases to date have occurred in adult bursae, tendon sheaths of the hands, feet, temporomandibular joint, and knees.\(^3,12-14\) A thorough literature review revealed several theories as to how these loose bodies come to exist at extra-articular locations. The first theory proposes that synovial chondromatosis is a reactive process of cells secondary to injury or irritation. In true primary bursal osteochondromatosis, Milgram and Keagy\(^4\) argued that an exostosis of the distal femur in apposition to the bursal cavity was the likely culprit in inducing proliferation of bone and cartilage nodules from the bursal lining with subsequent production of loose bodies. This idea is supplanted by the knowledge that the bursae and tendon sheaths are closely related to joints embryologically, and are thus subject to the same pathological conditions, which can lead to loose body formation in extra-articular locations.\(^15\)

A second theory states that normal communication between two compartments, such as the shoulder and biceps tendon, may facilitate the introduction of new cartilaginous foci from a more prominent synovial-producing area to one in which less synovium is present. This was based on a case reported by Covall and Fowble\(^8\) in which a 22-year-old man previously treated arthroscopically for synovial chondromatosis limited to the shoulder presented 18 months postoperatively with disease limited only to the biceps tendon. A final hypothesis suggests that smaller loose bodies originally formed within a joint can enter a bursa or cystic cavity that communicates with the joint, and continue to grow into larger lesions.\(^14,17\) This communication may occur not only naturally, but also through a tear secondary to injury. In either case, histological demonstration of articular surface fragments within extra-articular loose bodies supports an intra-articular origin.\(^14,18\)

In suspected cases of extra-articular involvement, plain radiographs, CT, and MRI are recommended. Depending on the site of involvement, arthroscopy may be indicated. Synovectomy usually is not performed for bursal lesions; total bursectomy is indicated. Synovectomy generally is used for symptomatic cases.\(^5,9\) Although extra-articular synovial chondromatosis generally is assumed to have a benign clinical course, extension of the growing loose bodies into surrounding bone may complicate the diagnosis with malignant transformation in extra-articular synovial chondromatosis to chondrosarcoma.\(^20\) Sim et al\(^3\) noted the importance of identifying the synovial origin of extra-articular lesions to avoid the erroneous diagnosis of chondrosarcoma. Additionally, cytologic atypia found in synovial chondromatosis may be mistaken for chondrosarcoma.\(^12\) Therefore, careful attention must be paid to both rule-out and rule-in suspected cases of extra-articular synovial chondromatosis.\(\Box\)

**REFERENCES**