Primary synovial chondromatosis in children: a case report and literature review

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Abstract

Primary synovial chondromatosis is an uncommon and benign metaplastic condition that usually affects large joints. Though shoulder involvement is scarce, and there are only a few cases in the pediatric population.

INTRODUCTION:

Primary synovial Chondromatosis (PSC), also called Reichel syndrome, is a benign tumor with cartilaginous nodules in the synovium joints. It usually affects diarthrodial joints and occurs between the third and fifth decade of life [5,7,17]. The glenohumeral joint is an unusual location in PSC, particularly in pediatric patients [4,7,16]. To our knowledge, there are only 5 cases of children reported in the literature. Herein, we report a rare case of PSC in a 14-year-old boy with an uncommon localization in the shoulder revealed by the chronic pain and the limited motion in the right arm. We also discuss clinical presentation, imaging findings and treatment of this rare condition in children based on current literature.

CASE PRESENTATION:

A 14-year-old, right-handed boy presented to the Pediatric Orthopedics department with right shoulder pain. He complained of 14 months history of pain and discomfort in his right shoulder. He had no trauma or family history of osteochondroma. He reported no symptoms of weight loss, fatigue, systemic signs, or any other arthralgia.

On physical examination, there was no obvious deformity or atrophy involving the affected shoulder. We noted a decreased range of motion, in comparison to the uninvolved side, with respectively: flexion to 160°, extension to 40°, abduction to 140°, adduction to 40°, internal rotation to L4, and external rotation to 50°. Subacromial impingement signs, as well as rotator cuff tear tests, were negative. The joint was stable. A neurovascular exam revealed no deficits in the upper limb.

The laboratory results were slightly normal, particularly the calcium and the phosphorus rate. Plain radiographs showed multiple radio-opaque bodies distributed throughout the glenohumeral joint, without bone defection or joint narrowing (Figure 1). Subsequent Magnetic resonance imaging (MRI) revealed a high number of calcified intra-articular loose bodies around the joint and the biceps tendon (Figure 2). PSC was strongly suspected.

We choose a shoulder arthroscopy using a deltopectoral approach to remove the tumors nodules. More than 50 shiny and solid bodies, with an average size of 10-15 millimeters, were retrieved (Figure 3). There were also several bodies within the coracoid process and the conjoint tendon. The synovial tissue, bursas, and cartilages appeared intact. Upon removing the particles, we complete a partial synovectomy to avoid the relapse. Histology of loose bodies and synovium confirmed the diagnosis of PSC by revealing multiple

cartilaginous nodules, composed of clustered chondrocytes and embedded in synovium (Figure 4). No sign of malignant transformation was noted.

Postoperative shoulder X-rays did not show any densities. The patient was discharged after the surgery using the arm sling. At six months of follow-up**,** the patient remains free of symptoms, and shoulder radiographs showed no recurrence of calcification.

DISCUSSION:

First mentioned by Jaffe et al., PSC is a rare benign tumor affecting the synovial cavity [5]. It is a proliferation of multiple cartilaginous nodules in the synovium of joints, tendon sheaths, and bursae.

These nodules may break free from the synovial membrane to become cartilaginous loose bodies in the joint space [4,10,15,17-20].

According to the literature, this disorder usually occurs in men between the ages of 30 and 50 old-years. It has been reported that knees, hip, elbow, and wrists are the main affected joints in descending order of frequency [10,12,15]. The involvement of the shoulder is unusual in adults, and more exceptional during childhood. To the best of our knowledge, only five cases have been reported in the literature [4,7,12,14,16].

Based on the underlying pathogenesis, synovial chondromatosis may be primary or secondary. The PSC, called idiopathic synovial osteochondromatosis or Reichel syndrome, usually occurs in a previously healthy joint. In contrast, secondary osteochondromatosis is a sequela of intra-articular pathology as osteochondral fracture, osteochondritis dissecans, and osteoarthritis [4,9,7,11,15,20]. In our case, the young-onset, the absence of a history of trauma, and the unremarkable results of blood tests strengthen the diagnosis of PSC.

It is noteworthy that histopathologic analysis is mandatory to distinguish between these conditions. [4,6,10,15,19]. In a series of 136 presumed synovial osteochondromatosis, Villain et al showed that the histopathological patterns are different. In fact, in primary lesions, the foci of chondrometaplasia in both synovium cells and loose bodies are noticeably disorganized. The chondrocytes are frequently binucleate, and the pattern of calcification is patchy and diffuse. However, in secondary lesions, calcification is zonal and uniformly distributed, and fragments of articular cartilage or subchondral bone are usually in the loose bodies [19]. In the present case, histologic evaluation revealed multiple cartilaginous nodules arranged in clusters and embedded in synovium.

Clinical presentation is often nonspecific [7,15,17]. As a result, patients may experience long symptoms delays before the final diagnosis. Like the current case, most children with PSC of the shoulder were diagnosed between 10 to 14 years old with a diagnosis delay ranging from 1 to 18 months from symptoms onset [4,7,11,12,16].

In a recent literature review of cases with osteochondromatosis occurring in the shoulder, the most common reported symptoms were mainly shoulder pain, uncomfortable feeling during exercises, and locked joint movement [18]. Our report is following data in the literature. Interestingly, a palpable bony mass may occur, as described in two children with PSC of the shoulder [3,20].

Radiographic features vary according to the degree of ossification. In the later stages of the disease, the plain radiographs showed a characteristic image with multiple intraarticular radio-opacities. These calcifications are frequently very similar and uniform in size with a typical nest-like arrangement. Thus, plain radiographs may be normal in the earlier stages (30% of cases) [3,20]. Sometimes, the diagnosis overlaps between differential diagnostics such as osteosarcoma and chondrosarcoma.

Hence, MRI plays a pivotal role in confirming the diagnosis by revealing intra-synovial hypo-intense nodules on T1 and T2-weighted images. MRI also aids in the management of surgical approaches. [2,3,5,17,18].

The optimal therapeutic management of the disease requires surgical removal of any loose bodies [7,8,13,16,17]. Partial synovectomy, optional but often recommended, may decrease the recurrence rate

[15,16,20]. Histopathological analysis of the loose bodies and synovial tissue is mandatory as a malign transformation may occur in up to 5% [1,7,17]. The choice of surgical procedure is still a matter of debate [15]. Open surgery remains the mainstay of treatment and is highly recommended in cases of osteochondromatosis with soft-tissue involvement and limited anatomic space access [7]. Moreover, this approach was often preferable in pediatric patients with shoulder involvement [7,12,14,16].On the other hand, the arthroscopic surgery approach may offer less morbidity and earlier postoperative recovery [20]. In line with Hamada et al, we opted for shoulder arthroscopy using the deltopectoral approach [4]. To elucidate further the clinical presentation and the therapeutic management of PSC of the shoulder in children, we analyzed findings reported in the literature (Table 1).

According to the literature, recurrence is common and ranges between 15% and 30% [1,2,15,16]. It's noteworthy to mention that no recurrence of calcification was reported among pediatric patients with PSC of the shoulder [6,7,16]. However, close follow-up including imaging in the first two years is warranted [7,16]. In our case, the short duration of follow-up was not sufficient to make a definitive conclusion.

In sum, the present case illustrates a rare entity of PSC that combined the intra and extraarticular involvement of the shoulder. MRI is a powerful key for early diagnosis. The management of this affection, like in adult patients, is based on the chondromyxoid bodies remove through open or arthroscope-assisted surgery. Histological analysis is mandatory since that a malign transformation might occur. Prognosis is excellent, associated with a low-moderate risk of recurrence.

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Table 1. Clinical Presentation, Treatment, and Outcome in patients with Synovial Chondromatosis in the literature and the present case

	Age	Sex	Duration of symptom (months)	Trigger factor	Clinical presentation	Surgical option	Follow-up Period (months)	Recurr
Nashi et al. 1998 [14].	14	male	6	Sporting activities	-Shoulder pain	- (under observation)	24	-
Miranda et al. 2004 [12].	10	female	1	Sporting activities	-Shoulder pain -Discomfort	Arthrotomy and synovectomy	12	No
Hamada et al. 2005 [4].	14	female	18	Sporting activities	-Shoulder pain -Discomfort	Arthroscopy	36	No
Kirchoff et al. 2008 [7].	14	male	12	no	-Shoulder pain -Palpable mass	Arthrotomy and synovectomy	9	No
Sinikumpu et al. 2020 [16].	14	male	12	Sporting activities	-Shoulder pain -Stiffness -Palpable mass	Arthrotomy and synovectomy	12	No

	Age	Sex	Duration of symptom (months)	Trigger factor	Clinical presentation	Surgical option	Follow-up Period (months)	Recurr
The present case 2021	14	male	14	no	-Shoulder pain -Discomfort	Arthroscopy and synovectomy	5	No

FIGURE LEGENDS

Figure 1:

Radiograph of the right shoulder demonstrates the presence of numerous radiopaque loose bodies in the glenohumeral joint.

Figure 2:

STIR magnetic resonance image shows excess of synovial joint fluid and multiple centers of nodular calcification around the joint and within the biceps tendon sheath.

Figure 3:

Numerous synovial Chondromatosis nodules removed from the shoulder joint (size between 10 and 15 mm).

Figure 4:

Histological image of the lesion showing multiple cartilaginous nodules composed of clustered chondrocytes, embedded in synovium (hematoxylin-eosin, original magnification × 400).