Case series of synovial chondrosarcoma of the knee joint: A very rare but challenging differential diagnosis in benign synovial lesions

Saam Haji Aliloo Saami, Ashkan Abrari, Milad Haji Agha Bozorgi

From MD, ShafaYahyaeian educational and clinical center, Iran University of Medical Science, Tehran, Iran

Correspondence to: Ashkan Abrari, ShafaYahyaeian hospital, Shohada square, Tehran, Iran, E-mail: ashkan_abrari@yahoo.com

Received - 03 November 2019 Initial Review - 20 November 2019 Accepted - 14 April 2020

ABSTRACT

Synovial chondrosarcoma (SCH) is a very rare malignancy and its differentiated from benign synovial lesion such as pigmented villonodular synovitis (PVNS) and synovial chondromatosis (SC) is challenging but crucial. Here, we present a case series of two different cases of 44 and 47 years old men with unilateral knee pain and swelling for the past 3 and 2 years, respectively. The knee gradually swells more in both cases. The medical history and constitutional symptoms were negative. There was a mass with a size of nearly 3 × 4 cm in the left knee of the first case and nearly 5 × 5 cm on the same side of the second case. Based on the magnetic resonance imaging and X-Ray, a diagnosis of synovial chondromatosis or PVNS was made. After marginal resection in both cases, the histologic analysis was consistent with the diagnosis of synovial chondrosarcoma. These patients were monitored carefully and both of them are asymptomatic. Although SCH is very rare, its misdiagnosis with PVNS and SC might lead to devastating consequences.

Key words: *Synovial chondrosarcoma, Pigmented villonodular synovitis, Synovial chondromatosis*

Benign neoplasms of synovium, including pigmented villonodular synovitis (PVNS) and synovial chondromatosis (SC), could affect any joint of the body such as the knee, hip, and elbow. Both of the lesions generally occur at the age of <50 years. Differentiation of these entities might be challenging both clinically and radiographically [1]. Localized PVNS and SC, if symptomatic, can be removed with marginal resection and carry a negligible risk of recurrence [1].

Intra-articular or synovial chondrosarcoma (SCH) is a very rare type of chondrosarcoma that most frequently presents in the knee joint [2]. It usually occurs at the age of >50 years and can be either primary or secondary to an SC. Primary SCH is extremely rare [3,4]. The optimal SCH treatment is extra-articular resection with wide surgical margins [2]. However, the diagnosis of SCH is not well characterized, and misdiagnosis with PVNS or SC is most likely [3,5]. Since SCH is a malignant neoplasm, its delayed diagnosis may result in the involvement of the neurovascular bundle and limb amputation. Therefore, timely diagnosis of SCH and avoiding its misdiagnosis with benign neoplasms such as PVNS and SC is of critical value.

In this study, we report two cases of SCH of the knee joint that was initially misdiagnosed as benign neoplasms of synovium and was resected marginally. Later histopathologic analysis of the resected tumors confirmed the diagnosis of SCH.

CASE SERIES

Case 1

A 44-year old male was referred to our orthopedic oncology center with complaints of pain and swelling at the left knee for 3 years. He was consulted to take nonsteroidal anti-inflammatory drugs (celecoxib) which proved not to be helpful. No history of trauma and constitutional symptoms was recalled by the patients. Vital signs were stable and there were no significant findings at the systemic examination. There exists a firm and mobile mass in infra patellar pouch with a size of nearly 3*4 cm and the knee range of motion (ROM) was 140 degree and painful. The laboratory testing, including white blood cell (WBC) count, C-reactive protein (CRP = 8), and erythrocyte sedimentation rate (ESR = 15), was all within the normal range.

Pre-operative radiographs of the knee revealed a clump of calcification in infra patellar pouch with no osteoarthritic changes (Fig. 1a). The magnetic resonance imaging (MRI) of the knee revealed evidence of intra-articular soft-tissue mass (Fig. 1b). Based on the radiologic observations, a benign synovial lesion (PVNS or SC) was suspected, and the patient underwent marginal resection due to the uncontrollable pain.

The resected tumor was sent to the pathology department for histologic evaluations. The histologic examinations revealed a proliferation of chondroid tissue with myxoid changes, slight atypia, and hyperchromatic nuclei that were consistent with

the diagnosis of chondrosarcoma Grade I (Fig. 2). The patient was monitored carefully on a regular basis at the outpatient clinic. Two years of follow-up of the patient was event-free and asymptomatic.

Case 2

A 47-year old male was referred to the oncology clinic of our hospital with complaints of pain and swelling of the left knee from 2 years ago. No history of trauma was remembered by the patient. Constitutional symptoms were also absent. Vital signs and systemic examination were normal. There exists a firm and mobile mass in infra patellar pouch with a size of nearly 5*5 cm and the knee range of motion was slightly restricted due to the pain and knee flexion was equaled nearly to 130 degree. Laboratory data including ESR = 19 and CRP = 6 were detected as well.

Pre-operative radiographs of the knee revealed no calcifications and osteoarthritic changes (Fig. 3a). MR imaging of the knee revealed a heterogeneous mass at the anterior knee joint (Fig. 3b). Accordingly, a benign synovial lesion (PVNS or SC) was suspected, and the lesion was extracted with marginal resection. Similar to case 1, the histologic evaluation was consistent with the diagnosis of chondrosarcoma Grade I. The patient was monitored carefully, and 3 years follow-up of the patient was event-free and asymptomatic.

DISCUSSION

SCH is a very rare entity and its differential diagnosis from the benign synovial tumors, particularly PVNS and SC, is crucial to avoid its devastating consequences. In this study, we reported two cases of SCH which occurred in the knee joint of two males with the age of fewer than 50 years. The lesions were not clinically and radiologically distinguishable with benign synovial lesions. For this reason, the mass was extracted with marginal resection. Even so, histologic evaluation of the extracted specimens was consistent with the diagnosis of SCH Grade I.

Bertoni *et al*. reported the long-term outcome of ten cases of SCH, which is by far the largest series of such entity. Only two cases of this series were considered primary. The remaining cases were secondary to SC or other pre-existing diseases. Five lesions were in the knee joint. Five patients had persistent pain. Three of these patients also had a palpable mass. Painless swelling was observed in two patients. Clumps of calcification were seen around the knee joint of four cases. Three cases (30%) of this series were misdiagnosed as chronic synovitis or PVNS.[3] These results reveal that misdiagnosis of SCH with benign synovial lesions is more prevalent than generally thought. Biazzo *et al*. performed a systematic review of the SCH cases reported in the literature, either primary or secondary. In total, 67 cases of SCH were included in their study. The average age of the patients was 56.9 years, with male dominancy. The knee joint was the most affected joint (47.7%), followed by hip and ankle (34.3% and 5.9%, respectively). The surgical intervention led to limb amputation in 59.7% of cases. The rate of local recurrence was 28.3%. They concluded that the SCH prognosis is worse than conventional chondrosarcoma, and timely diagnosis is essential [6]. Both cases of the present series were below the introduced age of higher incidence for SCH (44 and 47 years). These findings reveal that the diagnosis

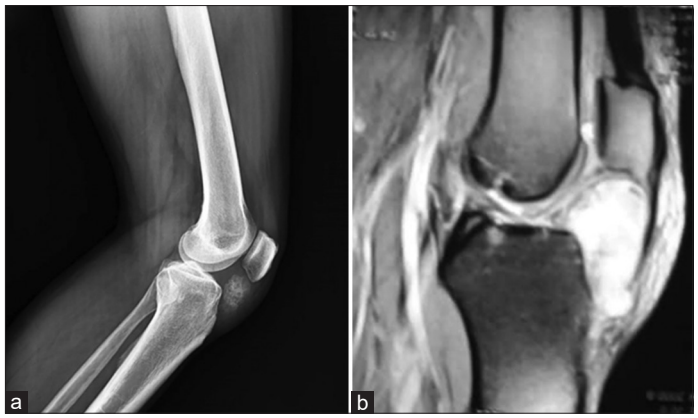

Figure 1: (a) Lateral radiograph of the knee showing intra-articular calcification; (b) sagittal T2-fat suppress magnetic resonance imaging showing a high-intensity intra-articular mass in infra patellar pouch (Case 1)

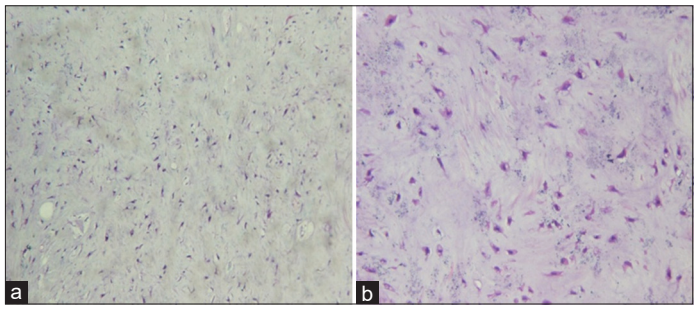

Figure 2: Histologic slides of the lesion showing chondroid tissue proliferation with myxoid changes, slight atypia, and hyperchromatic nuclei (a: ×10 and b: ×40) (Case 1)

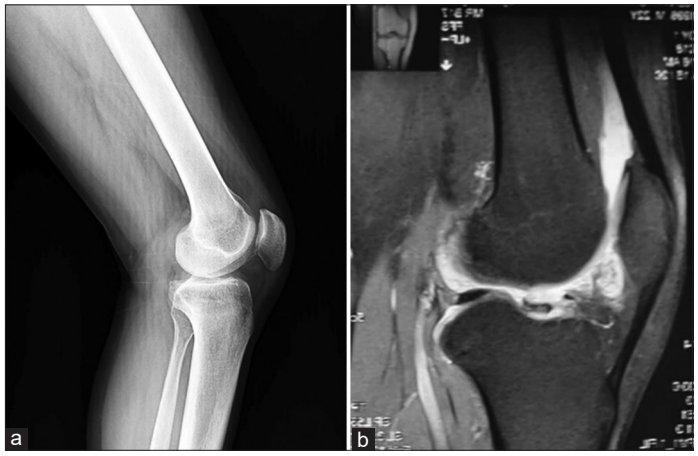

Figure 3: (a) Lateral radiograph of the knee showing no intra-articular calcification; (b) sagittal T2-fat suppress magnetic resonance imaging showing an intra-articular mass with mixed signal changes at the anterior joint (Case 2)

of SCH should be suspected even if the age of the patients favors the diagnosis of a benign synovial lesion. Furthermore, the report of the Biazzo *et al*. reveals that more awareness is required regarding the differential diagnosis of PVNS and SC from SCH.

PVNS is generally presented with a size of ≤3 cm and the age of <50 years. Similar to the SCH, the knee is the most commonly involved lesion [1]. The male dominancy was also a characteristic of PVNS. Although calcification is not seen in PVNS [1,7], effusion could be observed in a subset of patients so that eight out of 21 (38%) cases of the study of Huang *et al*. had effusion [1]. Therefore, the lack of calcification and the presence of effusion could be in favor of PVNS diagnosis.

SC usually affects the patients at their third to fifth decades of life [8]. The clinical symptoms of PVNS and SC are similar and typically include pain, swelling, and restricted joint movement. Similar to the SCH and PVNS, the knee is the most commonly involved lesion [1]. By contrast to PVNS, SC could be presented with radiographic calcifications in a considerable number of patients [9,10]. Therefore, the differentiation of SC from SCH could be even more challenging.

Muramatsu *et al*. reported a case of SCH in the right elbow joint of a 64-year old male who was inadequately treated with marginal resection, as there was no suggestion of a malignant tumor before the surgery. The surgical intervention led to the malignant transformation of the lesion over time. They suggested including the SCH in the differential diagnosis of the intra-articular tumor, as well as preoperative histologic evaluation to prevent iatrogenic contamination of tumor cells [11].

Altogether, although SCH of the knee is a very rare disease, it can be very devastating if not appropriately diagnosed. In this respect, its differentiation from the benign synovial lesion is of considerable importance as inadequate treatment of SCH could result in unfavorable consequences. Therefore, SCH should be included in the differential diagnosis of benign intra-articular lesions. If suspected, a pre-operative biopsy might be a reasonable choice to prevent tumor spread.

CONCLUSION

Although SCH is very rare, its misdiagnosis with PVNS and SC might lead to inadequate treatment and devastating consequences such as limb amputation. Therefore, it should be included in the differential diagnosis of these lesions.

REFERENCES

1. Startzman A, Collins D, Carreira D. A systematic literature review of synovial chondromatosis and pigmented villonodular synovitis of the hip. Phys Sportsmed 2016;44:425-31.
2. Campanacci DA, Matera D, Franchi A, Capanna R. Synovial chondrosarcoma of the hip: Report of two cases and literature review. Chir Organi Mov 2008;92:139-44.
3. Bertoni F, Unni KK, Beabout JW, Sim FH. Chondrosarcomas of the synovium. Cancer 1991;67:155-62.
4. Wenger D, Sundaram M, Unni KK, Janney C, Merkel K. Acral synovial chondrosarcoma. Skeletal Radiol 2002;31:125-9.
5. Sperling BL, Angel S, Stoneham G, Chow V, McFadden A, Chibbar R. Synovial chondromatosis and chondrosarcoma: A diagnostic dilemma. Sarcoma 2003;7:69-73.
6. Biazzo A, Confalonieri N. Synovial chondrosarcoma. Ann Transl Med 2016;4:280.
7. Jabalameli M, Jamshidi K, Radi M, Hadi H, Bagherifard A. Surgical outcomes of 26 patients with pigmented villonodular synovitis (PVNS) of the knee at a mean follow-up of 4 years: Introducing a novel technique. Med J Islam Repub Iran 2014;28:123.
8. Mankin HJ, Rosenthal DI, Xavier R. Gaucher disease. New approaches to an ancient disease. J Bone Joint Surg Am 2001;83:748-63.
9. Abolghasemian M, Gharanizadeh K, Kuzyk P, Masdari Z, Fakharian M, Safir O. Hips with synovial chondromatosis may display the features of femoroacetabular impingement. J Bone Joint Surg Am 2014;96:e11.
10. Boyer T, Dorfmann H. Arthroscopy in primary synovial chondromatosis of the hip: Description and outcome of treatment. J Bone Joint Surg Br 2008;90:314-8.
11. Muramatsu K, Miyoshi T, Moriya A, Onaka H, Shigetomi M, Nakashima D, *et al*. Extremely rare synovial chondrosarcoma arising from the elbow joint: Case report and review of the literature. J Shoulder Elbow Surg 2012;21:e7-11.

Funding: None; Conflicts of Interest: None Stated.

How to cite this article: Saami SHA, Abrari A, Bozorgi MHA. Case series of synovial chondrosarcoma of the knee joint: A very rare but challenging differential diagnosis in benign synovial lesions. Indian J Case Reports. 2020;6(4):157-159.

Doi: 10.32677/IJCR.2020.v06.i04.003