IDI: JCDR/2012/3882:1979

Case Report

Pathology Section

Bilateral Primary Synovial Chondromatosis: A Case Report

SHIVAKUMARSWAMY UDASIMATH, VIJAY P.M., PURUSHOTHAM R., KUMAR NAIK H., NAGESHA. K.R.

ABSTRACT

Synovial chondromatosis is a rare, generally benign condition which affects the synovial membranes and commonly involves the large joints such as the knee, hip, and the elbow. It is usually mono-articular and more common in males. Synovial chondromatosis is characterized by the presence of multiple cartilaginous nodules in the joint synovium or cavity. The definitive diagnosis is achieved after the histological examination of the synovial tissue. It can be very destructive and can cause severe osteoarthritis, pain and malignant transformation.

Herewith, we are presenting a rare case of bilateral, primary synovial chondromatosis of the knee joint in a female patient. An elderly aged woman presented with a history of pain and swelling of both the knees since six months. The imaging reports of the loose bodies suggested it to be synovial chondromatosis and histopathology studies confirmed this diagnosis. This case highlights the importance of careful clinical assessment, lateral thinking, the appropriate use of investigations, and careful pre-operative planning.

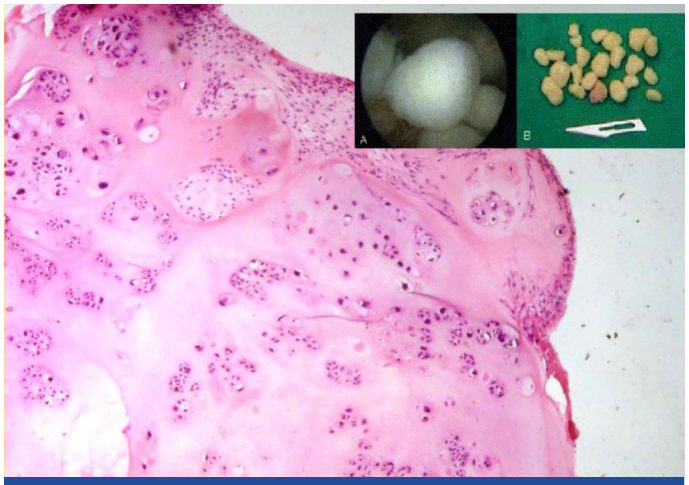
Key Words: Synovial chondromatosis, Loose bodies, Multiple cartilaginous nodules

INTRODUCTION

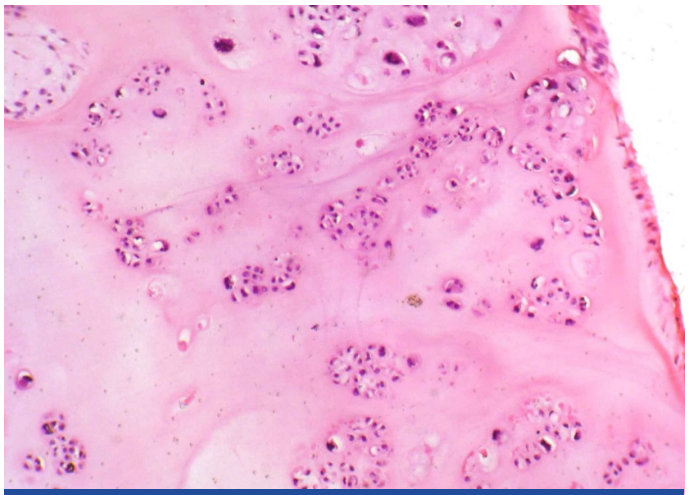
Synovial chondromatosis is an uncommon disorder of unknown aetiology. Mono-articular disease is the rule. It is characterized by the presence of cartilaginous nodules in the synovium of the joints, the tendon sheaths, and the bursae, which often occur without trauma or inflammation [1]. With the progression of this disease, these cartilaginous nodules may ossify and they can be identified only radiographically [2]. The diagnosis is commonly made on the basis of a thorough history, a physical examination, a radiographic examination and a histopathological examination of the cartilaginous nodules [2,3]. This condition tends to be progressive but self-limiting. The indications for surgery depend on the level of the symptomatic presentation in addition to the functional demands of the patient [1-3].

CASE REPORT

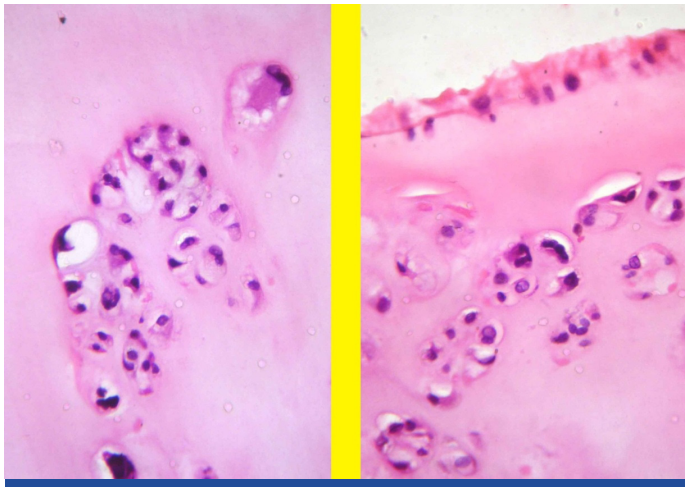
A 60-year old female presented with a history of pain and swelling and stiffness of both the knees since six months. The pain was aggravated by physical activity and it could be relieved by rest. She was otherwise healthy and her past medical history and systems review were unremarkable. There was no history of trauma or any other inflammatory or infectious disease. No neurological deficits of her lower limbs were found. The serologic tests for rheumatoid arthritis and for other seronegative arthritis or tuberculosis were negative. The radiographs of both the knees showed multiple calcified bodies which were diagnosed by histopathological studies as primary, bilateral synovial chondromatosis [Table/Fig-1]. CT imaging demonstrated the foci of increased attenuation (calcified areas). The conservative care was discontinued at this point and the loose bodies were removed. Histopathological examination of the loose bodies showed multiple, subsynovial cartilaginous nodules [Table/Fig-2] and mild to moderate cellular atypia in the shape and size of the chondrocytes of the subsynovial cartilaginous nodules [Table/Fig-3]. However, there



[Table/Fig-1]: Photomicrograph of loose bodies (H and E, 04 × magnifications), Inset: A: Radiography showing loose bodies, Inset B: Gross appearance of loose bodies.



[Table/Fig-2]: Photomicrograph of subsynovial cartilaginous nodule (H and E, 10 × magnifications).



[Table/Fig-3]: Photomicrograph of subsynovial cartilaginous nodule showing moderate atypia of chondrocytes (Left side) and synovial lining cells (Right side) (H and E, 10 × magnifications).

was no significant nuclear atypia, pleomorphism, and atypical mitosis, thus ruling out malignancy.

DISCUSSION

Synovial chondromatosis is also called as synoviochondrometaplasia, synovial chondrosis, synovial osteochondromatosis, and articular chondrosis [1]. In this condition, the cartilage cells are absent inside the synovial membrane. Milgram, in 1977, categorized the disease process into 3 distinct phases [4]. Phase I: (Active intrasynovial phase)- Cartilaginous metaplasia of the synovial intimal cells occurs with trauma, which is commonly thought of as an an inciting stimulus. Active synovitis and nodule formation is present, but no calcifications can be identified. Phase II: (Transitional lesions phase)- Nodular synovitis and loose bodies are present in the joint. The loose bodies are primarily still cartilaginous. Phase III: (Quiescent/Inactive intrasynovial phase)- The loose bodies remain but the synovitis has resolved. Synovial metaplasia occurs only in the first and second phase, while the free fragments are present in the second and third phases [5]. However, on the basis of the currently known molecular abnormalities, primary synovial chondromatosis is believed to be a benign neoplastic rather than metaplastic disease. These cytogenetic aberrations are absent in secondary synovial chondromatosis [6]. In addition, growth factors, such as the fibroblast growth factor –2 and –3 have been found in primary synovial chondromatosis. The fibroblast growth factor receptor 3 (FGFR3) is a specific marker of the mesenchymal precartilaginous stem cells. Histologically, the cells at the periphery of the cartilage nodules express FGFR3 [7]. Chromosome 6 abnormalities which are identified by the cytogenetic and molecular cytogenetic analyses, have been a recurrent finding in primary synovial chondromatosis [6].

Multiple (even hundreds), cartilaginous nodules, are commonly formed. The chondrocytes become pedunculated and encrusted inside the synovium and are eventually expelled into the joint as loose bodies. These loose bodies may continue to grow, being nourished by the synovial fluid and continue to calcify, in 2/3rds of the patients [4, 8].

This disease is commonly seen in the 3rd to 5th decades of life, with a male to female ratio of 2:1 [2]. The onset is described as insidious and it occurs over months to years. Monoarticular disease is the rule. The most frequent site of involvement is the knee, followed by the hip, shoulder, elbow and the ankle. It is also encountered in the tendon sheaths and in the periarticular bursa (extra-articular form). Occasionally, the process can be extended beyond the joint, into the adjacent soft tissue. A combination of intra- and extra-articular diseases can occur [1, 4, 8].

Synovial chondromatosis can be differentiated into primary and secondary forms. The primary form occurs in an otherwise normal joint and it is characterized by undifferentiated stem cell proliferation in the stratum synoviale [9]. The primary form is thought to be progressive and more likely to recur, and it may lead to severe degenerative arthritis with its long-term presence. Secondary synovial chondromatosis is thought to be caused by the irritation of the synovial tissue of the affected joint. This form is associated with degenerative joint disease, trauma, inflammatory and non-inflammatory arthropathies, avascular necrosis, and osteochondritis dissecans. This form is not likely to recur following its surgical removal [2, 9].

The diagnosis of synovial chondromatosis is made following a thorough history, a physical examination, and a radiographic examination [2, 3]. According to Milgram's classification, the plain film radiographs are only helpful in the third phase of the disease, once the calcification has occurred [10]. When mineralization occurs, the radiographs reveal radiopaque, round or oval, loose bodies within the joint [4, 10]. When imaging does not provide the specific diagnostic features, the definitive diagnosis is made histologically on the basis of a synovial tissue biopsy [11].

Recent interest in this diagnosis has increased due to its potential for malignant degeneration with a relative risk of 5% [12]. The clinical and radiographic features of the synovial chondromatosis and the chondrosarcoma conditions are similar. The literature reports only 33 cases of malignant transformation in the setting of histologically confirmed synovial chondromatosis. A key feature of all these cases is the recurrence of the benign disease prior to a diagnosis of the malignant disease [12].

As radiotherapy and chemotherapy have no effect on synovial chondromatosis, surgical excision is the preferred treatment for it [1, 2]. In asymptomatic patients, the nodules may resorb over time and invasive procedures should be avoided. In localized intra-articular disease, the complete excision of the abnormal synovium seems to provide a cure. In the phase III disease, the removal of the loose bodies alone is sufficient. When synovitis is present, resection of the loose bodies and synovectomy is done. The extra-articular disease treatment aims for the complete excision of the abnormal synovium. The recurrence rates for synovial chondromatosis after surgical treatment have been reported to vary from 7% to 23%. Complications like secondary degenerative osteoarthritis due to chronic mechanical irritation and bone destruction by the loose bodies is the rule. Surgery predisposes the patients to tissue scarring, subsequently compromising the joint function [2, 11].

The differential diagnoses of primary synovial osteochondromatosis include pigmented villonodular synovitis, secondary synovial osteochondromatosis, rheumatoid or other seronegative arthritis, septic arthritis which includes granulomatous infections, synovial hemangioma, synovial chondrosarcoma and osteochondromas with adjacent secondary bursal osteochondromatosis. Blood tests and the arthritis profiles can also help in ruling out the specific differential diagnoses [4, 11].

CONCLUSION

Synovial chondromatosis is a rare condition which can be highly aggressive and destructive. The lack of awareness of this condition may lead to incorrect diagnoses and unwarranted surgery. Because

of the concern of chondrosarcoma, if radiographic or MR imaging studies are inconclusive, a histological diagnosis is a prudent course for this condition.

REFERENCES

[1] Krebbs VE. The role of hip arthroscopy in the treatment of synovial disorders and loose bodies. *Clin Orthop Rel Rsch*. 2003;406:48–59.
[2] Valmassy R, Ferguson H. Synovial Osteochondromatosis. A brief review. *J Am Podiatr Med Assoc*. 1992;82:427–31.
[3] Forsythe B, Lou J, States L, Guttenberg M, Dormans JP. Painless ankle mass in a 12-year-old boy. *Clin Orthop Relat Res*. 2004;422:263–69.
[4] Milgram JWL. Synovial osteochondromatosis: A histopathological study of thirty cases. *J Bone Joint Surg*. 1977;59A:792–801.
[5] Milgram JW, Hadesman WM. Synovial osteochondromatosis in the subacromial bursa. *Clin Orthop Relat Res*. 1988;236:154–59.
[6] Murphey M E, Vidal JA, Fanburg-Smith JC, Gajewski DA. Imaging of synovial chondromatosis by using a radiologic-pathologic correlation. *Radio Graphics*, 2007;27:1465-88.
[7] Robinson D, Hasharoni A, Evron Z, Segal M, Nevo Z. Synovial chondromatosis: The possible role of FGF 9 and FGF receptor 3 in its pathology. *Int J Exp Pathol* 2000; 81:183-89.
[8] Kay PR, Freemont AJ, Davies DRA. The aetiology of multiple loose bodies. Snow storm knee. *J Bone Joint Surg Br*. 1989;71:501–04.
[9] Leu JZ, Matsubara T, Hirohata K. Ultrastructural morphology of the early cellular changes in the synovium of primary synovial chondromatosis. *Clin Orthop*. 1992;276:299–306.
[10] Zimmerman C, Sayegh V. Roentgen manifestations of synovial osteochondromatosis. *Am J Roentgenol Radium Threr Nucl Med*. 1960;83:680–86.
[11] Shearer H. A case report of bilateral synovial chondromatosis of the ankle. Available from: http://creativecommons.org/licenses/by/2.0 [Last accessed on 2007 November 24].
[12] Sah AP, Geller DS, Mankin HJ, Rosenberg AE, Delaney TF, Wright CD, et al. Malignant transformation of synovial chondromatosis of the shoulder to chondrosarcoma: a case report. *J Bone Joint Surg Am*. 2007;89:1321–28.

AUTHOR(S):

1. Dr. Shivakumarswamy Udasimath
2. Dr. Vijay P.M.
3. Dr. Purushotham R.
4. Dr. Kumar Naik H.
5. Dr. Nagesha K.R.

PARTICULARS OF CONTRIBUTORS:

1. Corresponding Author,
2. (Path) Assistant Professor,
3. (Path) Associate Professor,
4. (Path) Associate Professor,
5. (Path) Associate Professor.

NAME OF DEPARTMENT(S)/INSTITUTION(S) TO WHICH THE WORK IS ATTRIBUTED:

Department of Pathology, HIMS, HASSAN – 573201

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Shivakumarswamy Udasimath, MD, DNB, (Path)
Assistant Professor, Department of Pathology,
HIMS, HASSAN - 573201.
Phone: +91-9481730220
E-mail: udasimath@gmail.com

FINANCIAL OR OTHER COMPETING INTERESTS:

None.

Date of Submission: **Aug 02, 2011**
Date of Peer Review: **Oct 21, 2012**
Date of Acceptance: **Jan 09, 2012**
Date of Publishing: **May 01, 2012**