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Case Report

Non-pigmented villonodular synovitis of knee joint in a 11-year-old boy on long term steroids: a case report

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ABSTRACT

Pigmented villonodular synovitis (PVNS) is a rare condition, commonly affecting the knee joint and hip joint. It is seldomly seen in pediatric age group. Aetiology is unknown. Magnetic resonance imaging (MRI) is the imaging modality of choice. Managed by surgical resection of the synovium performed with an open, arthroscopic or combined approach. Post-operative adjuvant external beam radiotherapy have reduced recurrence rate. In this case report we are going to present a rare case of eleven-year-old boy on long term steroid intake, who presented with a history of pain and swelling of right knee joint with restriction of range of movements. MRI suggested extensive villonodular synovitis of knee joint. Diagnostic arthroscopy showed extensive villonodular synovitis of suprapatellar pouch, medial gutter, lateral gutter, intercruciate space, posterior-medial and posterior-lateral compartments and villi had atypical yellow fatty appearance. Extensive arthroscopic synovectomy was done and histopathology showed chronic synovitis. Post-operatively pain and swelling reduced and range of movements improved. Arthroscopic synovectomy is the most effective line of management for villonodular synovitis.

Keywords: PVNS, Pediatric, Steroid, Yellow fatty, Arthroscopic synovectomy

INTRODUCTION

Pigmented villonodular synovitis (PVNS) is a benign lesion affecting the synovial joints, tendon sheaths and bursas.[1] It is relatively a rare condition, seldomly seen in pediatric age group. Knee is the most commonly affected joint followed by hip, ankle, shoulder and elbow.[2,3] Aetiology of this condition is still unknown. But there are studies suggesting it to be secondary to hemarthosis or chronic inflammation.[4] There are very few cases of PVNS reported in patients on anticoagulation therapy and with a bleeding disorder.[5,6] Presentation will be pain and swelling of the joint with restricted range of motion. MRI is the investigation of choice. Managed by surgical resection of the synovium performed in open, arthroscopic or combined manner.[7] Recurrence rates have been suggested to be decreased by radiation therapy and radiosynoviorthesis.[8]

CASE REPORT

Eleven-year boy came to our institute with complains of pain and swelling of right knee joint. Gives history of trivial trauma to knee three years back which was associated swelling. Aspiration of the knee joint was done elsewhere. Aspirate was straw coloured, biochemical and microbiological tests were normal. The treating consultant then started him on oral steroids (prednisolone).

He was on oral steroids for three years. Oral steroid was stopped and 20 days later he came with swelling and restricted range of motion of right knee joint. Supra, para and infra-patellar fullness was seen. Active and passive knee flexion was 0-ten degree with no distal neurovascular deficit. He was overweight for his age. MRI was done, which suggested extensive villonodular synovitis of knee joint.

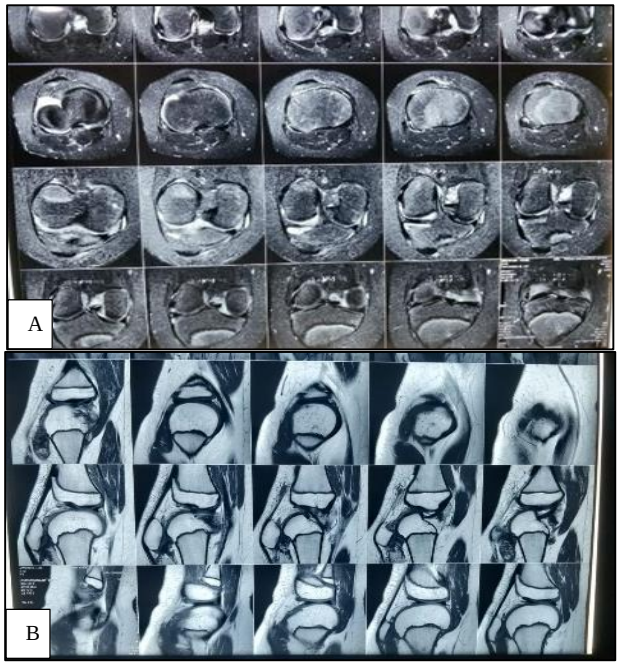

Figure 1 (A and B): MRI right knee (axial and sagittal sections) of villonodular inflammatory changes of synovium.

Diagnostic arthroscopy showed extensive villonodular synovitis of suprapatellar pouch, medial gutter, lateral gutter, intercruciate space, posterior-medial and posterior-lateral compartments. Surprisingly villi had yellow fatty appearance. Arthroscopic synovectomy done. Resected synovium sent for histo-pathological analysis. It revealed synovial cell hyperplasia with mild infiltration by mono-nuclear cells suggestive of chronic synovitis.

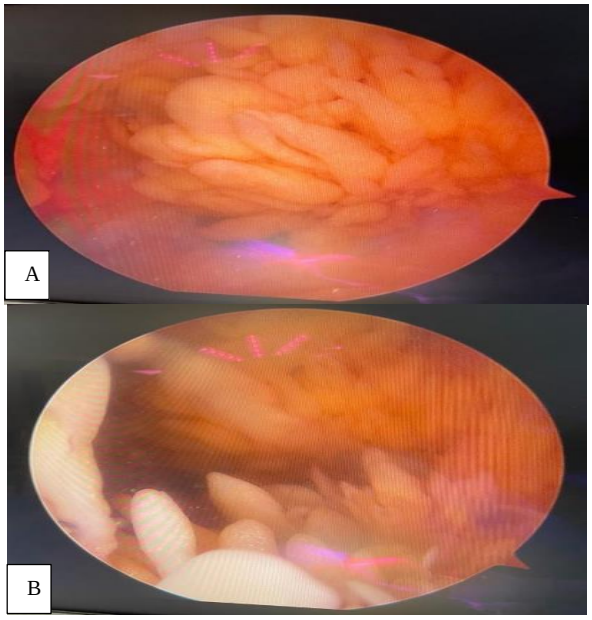

Figure 2 (A and B): Intra-operative arthroscopic images showing atypical yellow fatty appearing villonodular changes of the synovium.

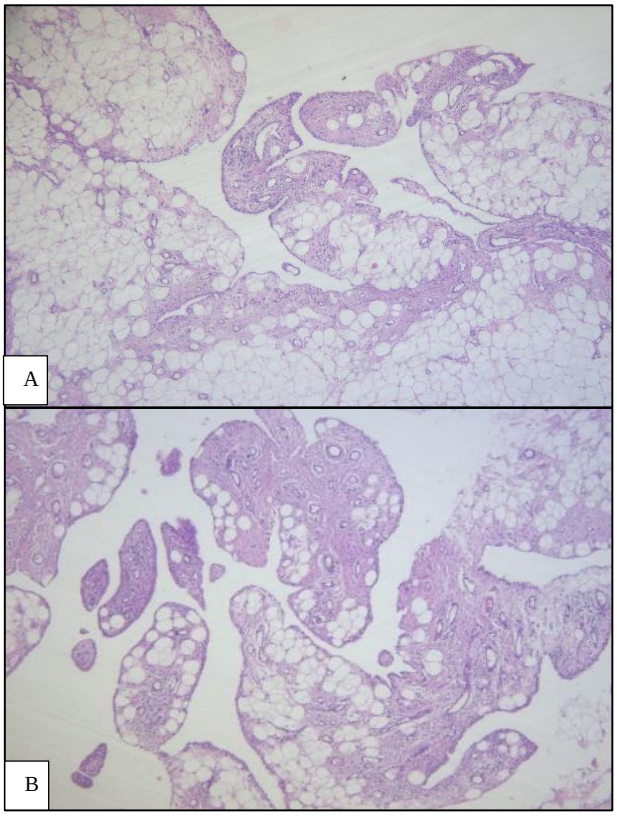

Figure 3 (A and B): Histopathology images showing synovial cell hyperplasia with mild infiltration by mononuclear cells suggestive of chronic synovitis.

Post-op period was uneventful. Knee ROM was started from post-op day one. Immediate pain relief and improvement of knee range to 90-degree flexion was noted. Patient was allowed full weight bearing mobilization from post-op day one. At one week follow up patient had active knee flexion of 0-100 degree and decrease in knee effusion. Sutures were removed on post-op day 15. At one month follow up patient had pain-free full knee range of movements and no effusion. Patient was followed up for a period of one year as well as had no relapse.

DISCUSSION

Adolescents acute knee effusions are most frequently associated with fractures, patellar dislocations, or internal derangements (ligamentous or meniscal injuries). Adolescent with a rarer chronic knee effusion, on the other hand, motivates us to widen our differential diagnosis. Juvenile rheumatoid arthritis (JRA), synovial osteochondromatosis, synovial hemangioma, synovial sarcoma, and PVNS are diagnoses to take into account in these patients.[9] PVNS is a benign proliferative disorder of the synovium. PVNS is a very uncommon disease with vague symptoms and a sneaky onset that frequently results in a delayed diagnosis. Most of the cause is unknown. It usually involves a large joint (most commonly the knee) and is diagnosed between the third and fifth decade of

life.[3,10] It frequently manifests as an insidious onset, painless joint swelling. Typically, a vague soreness or ache is noted along with some range of motion restrictions. To confirm the diagnosis, other imaging modalities are frequently required. Plain radiographs are typically negative in the early stages of the disease but may reveal articular erosions, effusions, or subchondral cysts.[10-12] An intraarticular nodular mass with low signal intensity on T1-weighted, T2-weighted, and matching proton density sequences due to hemosiderin deposition is the diagnostic marker of hypertrophic synovium on MRI.[10,11] The "blooming artefact" that appears on T2 weighted gradient-echo sequences is caused by the iron in hemosiderin's magnetic susceptibility.[13]

PVNS on plain film with multiple subchondral cysts has a range of differential diagnoses, including DJD, TB, haemophilia, amyloidosis, gout, and synovial chondromatosis.[14] Similar symptoms may be present in synovial osteochondromatosis (also known as synovial chondromatosis), but this is characterized by the proliferation and transformation of cartilaginous and synovial fragments into ossified loose bodies.[15] Plain radiographs show several calcifications, which aids in differentiating synovial osteochondromatosis from PVNS. On T2-weighted MRI scans, the signal intensity is typically quite strong.[15] Although synovial sarcoma, a slowly progressing cancer, should be taken into consideration, T2-weighted MRI images typically show increased signal intensity.[16] On the basis of the arthroscopic examination and biopsy, the final diagnosis is made. Synovial hemangioma a rare condition, may present with painful monoarticular hemarthrosis shows increased signal intensity on both the T1-weighted and T2-weighted MRI images.[17]

PVNS synovium is proliferative, with a villous appearance and a reddish-orange staining, according to an arthroscopic assessment. In order to alleviate pain and prevent recurrence, the goal of PVNS therapy is the excision of all aberrant synovial tissue. A synovectomy, including the posterior compartment if it is implicated, is the first course of treatment, done via arthroscopy or open arthrotomy.[18-20] Arthroscopic synovectomy is the preferred method, as it results in faster recovery and early return to normal activity.[21] However in our case, the villi were atypical with yellow fatty appearance. Long term steroid intake is present which might have triggered synovial hyperplasia and villonodular synovitis.

CONCLUSION

Atypical villonodular synovitis is a rare disease in pediatric age group with unknown aetiology, probably associated with chronic steroid intake. Usually presents with pain, swelling and restriction of range of movements. MRI is the investigation of choice. Arthroscopic synovectomy gives better functional outcome and early mobilization.

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