A Case Report

Extra –articular pigmented villonodular synovitis of knee

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ABSTRACT

This study is a case report of a 30 year old male with one month old history of rapid growing swelling over left lower thigh and mass which was associated with mild pain. Imaging was inconclusive and biopsy (open) showed, inflammatory process with synovial cell and gross specimen showed Pigmented villonodular synovitis. This case study highlights the clinical presentation of this rare disorder with rare presentation and recommends its consideration as a differential diagnosis when dealing with non-traumatic persistent peri or extra-articular knee pain and swelling.

Key Words: Pigmented villonodular synovitis, Extraarticular, Hemosiderin

INTRODUCTION

Pigmented villonodular synovitis is a locally destructive fibroelastic proliferation, characterised
by many villous and nodular synovial protrusions, which affects joints, bursa, and tendon sheaths. PVNS was first described by Jaffe, Lichtenstein and Sutro in 1941, who used this name to identify the lesion because of the yellow brown, villous and nodular appearance. The yellow-brown pigmentation is due to excessive deposits of lipid and hemosiderin. The condition can be diffuse or localized. When entire synovium is affected and there is major villous component, the condition is called as diffuse. When a discrete intrarticular mass is presented, the condition is called as localized PVNS. When it affects the tendon sheath it is called lozalized giant cell tumour of tendon sheath or nodular tenosynovitis. The diffuse form usually occurs in knee, hip, elbow and wrist. Localised / Nodular form most often seen in fingers. In the new revised classification of soft tissue tumour, the WHO classified localized lesion as Giant cell tumour of sheath and diffuse as, diffuse type GCT.

One of the most characteristic finding in PVNS is the ability of the hyperplastic synovium to invade the subchondral bone. Aetiology of PVNS is unknown. Some investigations has suggested autoimmune pathogenesis¹ and some as a genetic², Trauma is also a suspected cause. Some suggested a disturbance in lipid metabolism as an etiologic factor. Pigmented villonodular synovitis is usually monoarticular, affecting adults in the third or fourth decade of life; the knee is the most commonly involved anatomic location, followed by the hip, ankle,
shoulder, and elbow. Pigmented villonodular synovitis is usually intra-articular and only rarely extra-articular. In cases of extra-articular pigmented villonodular synovitis, there is usually evidence of extension from an intra-articular focus. As a result, the finding of pigmented villonodular synovitis in the suprapatellar area, which does not communicate with the knee joint is rare. In extraarticular form PVS can also be either diffuse with multifocal lesions of synovial or nodular type with localized nodular changes that are frequently called gigantocellular tumor of tendon sheaths. It is widely accepted that the process is of reactive inflammatory nature. However, because of its aggressive growth with bone destruction or recidivism, low malignant local aggressive neoplasm is frequently suspected.
CASE REPORT

A 30 year old male presented in our unit with a diffuse soft tissue mass over anterolateral aspect of distal femur (left) proximal to left knee since 4 months. The mass has been increasing in size for one month. 15 days prior to admission mass became tender and was associated with popping sensations. He denied any history of trauma, weight loss, anorexia or fever. Review of the respiratory, cardiovascular, gastrointestinal, and genitourinary and nervous system was essentially normal. His past medical and family history were not significant.

The general physical examination was within normal limits, except for the presence of a left sided antalgic gait. Local examination of the left knee revealed ill defined swelling of dimensions 10x6 cm, soft-elastic consistency, without any wide movements and increased local temperature. The terminal range of movements were normal. Neurovascular status was normal.

Radiographic findings of the left knee were normal except increased soft tissue density along anterolateral aspect of left femur, without a visible soft tissue mass, bone erosion, acute fracture, dislocation, or effusion.

MRI showed large irregularly marginated ill defined lesion is noted in suprapatellar recess of left knee joint. It was hyperintense in both T1W and T2W images and showed flow voids in it. The lesion was associated with a minimal loculated hemarthrosis with septations. Possibilities were synovial neoplastic lesion; synovial hemangioma, synovial lipoma, PVNS, synovial sarcoma. (Fig. 1, 2)
Based on these findings the conclusion was established: intraperiarticular tumefaction inside the suprapatellar recessus predominantly cystic with soft tissue proliferation. The lesion had aggressive characteristics. Differential diagnosis included lipoma arborescens, liposarcoma,
synovial sarcoma, synovial hemangioma, inflammatory arthropathy, synovial chondromatosis, bursitis with synovial tissue proliferation, hemosiderotic synovitis.

The patient was taken to the operating room, under general endotracheal anesthesia, by means of lateral suprapatellar incision, operative exploration showed an irregular, easily palpable, pink to purple, soft tissue mass of size 10 x 4 cms, in the distal femoral (left) space, after meticulous dissection partial synovectomy with complete extirpation of mass was done and sent for routine histopthologic examination. (Fig. 3) In addition, culture swab was also taken for microbiologic examination.

![Figure 3 Gross pathological specimen](image)

Histopathological examination revealed-marked hyperplastic synovium thrown into villous pattern, dense inflammatory cell infiltrate in the synovial tissue composed of lymphocytes, plasma cell, histiocytes and large clusters of hemosiderin laden macrophages. There is no any granuloma. (Fig. 4)
Postoperative period was normal, applied physical therapy gave satisfactory result and was found to be pain free on day 14 of surgery, on follow up after one month range of motion was achieved fully. 3 months after the surgery patient showed normal physical status with full physical activity and full range of motion, without pain, swelling or reactivism.

CONCLUSION-

Pigmented villonodular synovitis (PVNS) is a rare benign proliferative disorder of uncertain etiology that affects synovial lined joints, bursae, and tendon sheaths. The disorder results in various degrees of villous and/or nodular changes in the affected structures. Patients usually present with painless joint swelling of insidious onset that mimics joint effusion. Although recurrent hemorrhage is a common presentation but our patient did not have any such episode. Joint pain subsequently supervenes, but the swelling is disproportionate to the degree of pain. The pain is mild and of insidious onset, and it progressively worsens and frequently is accompanied by decreased range of motion and locking of the joint. Recurrent mild to moderate effusion creates the impression of recurrent joint swelling.
Blood fluid aspiration and MRI are valuable tools for early diagnosis\(^7\). In our patient radiographs failed to demonstrate a soft tissue mass, which is common in pigmented villonodular synovitis. The diagnosis of pigmented villonodular synovitis was established histologically. It is very important to analyze the form and place of appearance of villonodular synovitis. The treatment of this disease is challenging\(^{11}\). Nonsurgical treatment includes use of triamcinolone hexacetonid, use of 90 Yttrium or radiotherapy\(^{12,13}\). Surgical treatment by synovectomy has appeared to be the most effective in symptomatic diffuse type PVNS\(^{14}\). Sometimes, recurrence can occur after total synovectomy in diffuse forms of PVS and for that reason surgical treatment can be improved with radiotherapy. It is considered that in 25-50% of cases recurrence appears after the surgical treatment\(^{15}\). In our case the patient underwent partial synovectomy without signs of recurrence 3 months after the surgery. This presentation of diffuse pigmented villonodular synovitis which did not find to be communicating to the knee joint without any sign or evidence of knee effusion is rare. We recommend that PVNS should be included as a differential diagnosis when evaluating a young adult with non-traumatic persistent knee pain and swelling.
References-


