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Original Research Paper

Orthopaedic

PIGMENTED VILLONODULAR SYNOVITIS OF HIP: A RARE CASE

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ABSTRACT Pigmented villonodular synovitis (PVNS) is a rare disease that can affect any joint, bursa or tendon sheath. The hip is less frequently affected than the knee.1 We report a case of a 46 years old male presented with history of pain and limp over right hip since 6months associated with difficulty in sitting cross legged and squatting to performing day today activity. There was no history of fever, weight loss, and long term drug intake. Radiological assessment showed secondary arthritis of right hip with features of avascular necrosis (AVN). The provisional diagnosis was Arthritis of right hip secondary to AVN/ Inflammatory arthritis and the patient underwent uncemented right total hip replacement(THA), Intra operatively the articular erosion was noted with reddish gray abnormal synovial growth. Histological assessment showed diffuse expansive sheets of histiocytes admixed with hemosiderin laden macrophages and alveolar spaces lined by synovial cells and collagenized stroma. A short term follow up of 18months did not show any clinical recurrence. We conclude that, from our observation, complete excision of lesion combined with THA is an effective treatment for PVNS of Hip.

KEYWORDS : Pigmented villonodular synovitis; Hip; Hyperplasia, Total hip replacement.

INTRODUCTION

Pigmented Villonodular Synovitis (PVNS) is a rare benign condition that affects the synovial lining of joints, tendon sheaths, and bursae.[2] PVNS is a locally aggressive musculoskeletal disorder characterized by hemosiderin deposition within the tendon sheaths and bursae of joint. The incidence is 80% over the knee followed by hip and ankle joints.[3]

It is characterized by the growth of abnormal tissue called synovial proliferations that can invade and destroy surrounding tissues, leading to joint dysfunction and pain. PVNS is most commonly found in the knee joint, but it can also occur in other joints such as the hip, ankle, and shoulder.

In intra-articular forms, the large joints are the most affected: the knee (70%), hip (15%) and then the ankle, shoulder and elbow.[1] Hip is one of the largest and most important joints in the body, responsible for bearing weight and enabling mobility. In this case, we will focus on a rare and unusual presentation of PVNS in the hip joint.

Case Description

A 46 years male presented to our OPD with history of pain in right hip joint for 6 months. Pain was insidious in onset, continuous and aggravated with walking. Pain and swelling were gradually progressive in nature. There was no h/o trauma, fever, weight loss and long term drug intake. Pain was associated with limp and difficulty in sitting cross legged and squatting affecting day today activities.

Physical examination revealed restricted range of movements in all planes and tenderness over the right hip joint. Plain radiograph of the right hip (Fig. 1) showed features of secondary arthritis of right hip with avascular necrosis as follows:

1. Joint space narrowing: This occurs due to the loss of cartilage and joint destruction, which results in a reduced space between the femoral head and the acetabulum.
2. Subchondral bone erosion: This is the destruction of the underlying bone beneath the cartilage.
3. Osteophyte formation: This is the growth of new bone at the margins of the joint as a response to joint degeneration and inflammation.
4. Changes in joint alignment: Corresponding to advanced arthritis.

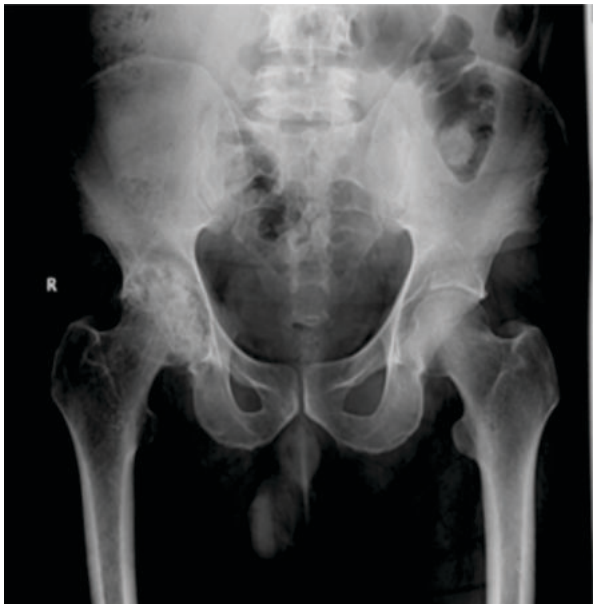
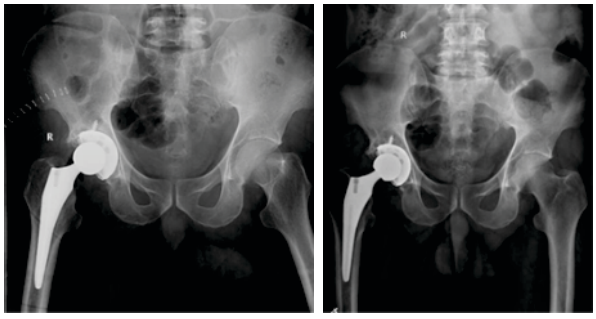


Figure 1: Pre- Operative Anteroposterior Radiography Of The Hip

Based on radiological features it was classified as classified as stage 4 of Ficat and Arlet[4]. Considering clinical and radiological features he was planned for uncemented THA.

Intra operatively we observed unusual synovial hypertrophy with reddish/gray discoloration(Fig. 3). Total synovectomy was done and proceded with uncemented THA as per plan. The synovium was sent for histopathological assessment, which showed papillary hyperplasia with numerous histiocytes & hemosiderin laden macrophages(Fig. 4).

Post operatively course was uneventful and rehabilitation was done as per standard protocols[4]. Patient is being followed up regularly till date and 18months follow up did not show any signs of recurrence(Fig. 2).



Postoperative Day1 18 Months Follow Up X-ray

Figure 2: Post Operative Anteroposterior Radiography Of The Hip After THA

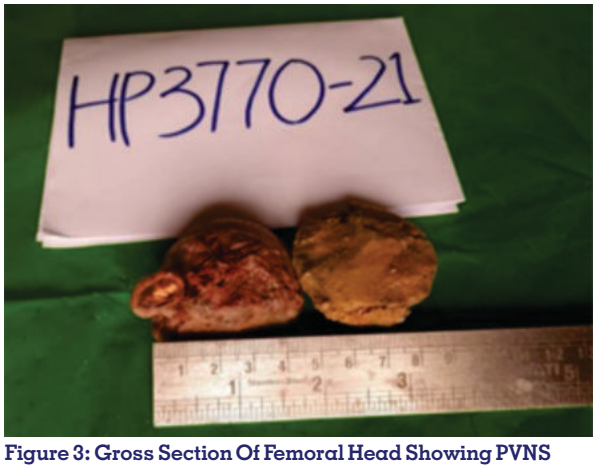
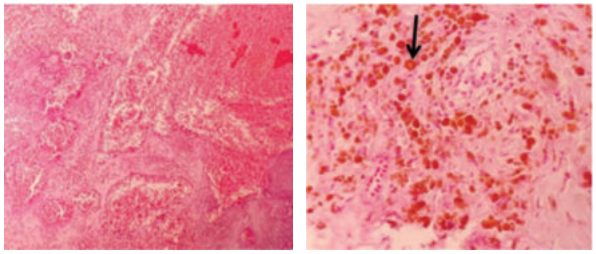


Figure 3: Gross Section Of Femoral Head Showing PVNS

Macroscopic Findings

External surface noted an attached nodule to the femur head measuring 2.2x1.2x0.8cm. External surface of nodule congested, grey brown. Cut section of nodule grey white to grey brown. Also multiple grey yellow to grey brown tissue bits largest measuring 2x1x0.8cm, smallest measuring 0.5cm in diameter. Cut section of all tissue bits grey brown.



10X H&E Shows Papillary hyperplsia 40X H&E shows hemosiderin macrophages

Figure 4: Histology Of PVNS Of Right Hip

Microscopic Findings

Sections form the femur head shows the bony trabeculae with the marrow depicting fatty spaces and with sparse hemopoeitic elements.

Multiple sections from the soft tissue depict a lesion composed of diffuse expansive sheets of histiocytes admixed with hemosiderin laden macrophages. The synovium depicts papillary hyperplasia with numerous histiocytes & hemosiderin laden macrophages seen in the stroma. Also seen are alveolar spaces lined by synovial cells with collagenized stroma. No evidence of giant cells. No atypia. No granulomas. No evidence of necrosis.

Impression: Features suggestive of Pigmented Villonodular Synovitis –Hip

DISCUSSION

PVNS is a rare condition which usually affects young individuals between the ages of 20 and 40.Our patient was a 46yr old male not in typical age group with pathology in unilateral hip, One joint is typically affected by PVNS similar to our case; involvement of multiple joints is highly uncommon.[5]

The prevalence of PVNS is slightly higher in female patients, according to several earlier investigations.[6] However we reported a male case. Several cyst-like lesions or erosive areas in the non-weight-bearing regions of the acetabulum, femoral head, and/or femoral neck are the most common roentgenogram findings of PVNS in the hip joint. In 95% of the patients studied by Cotton et al. (cortical erosion, 43%; cyst-like erosion, 90%), these bony lesions were present.[7]

Uncertainty surrounds PVNS's aetiology. The pathophysiology of PVNS is the subject of numerous ideas, including etiopathological mechanisms including inflammatory reactions[8], defective cellular and humoral immunity, hereditary susceptibility, or recurrent bleeding after trauma. The most popular idea at the moment, nevertheless, appears to have a neoplastic origin.[9] PVNS local recurrence and metastasis have been described in numerous publications.[10] Aggressive PVNS are being treated with adjuvant radiation treatment.[10] Nevertheless, arthroplasty and total synovectomy have also been utilised as PVNS alternate treatments. Results from synovectomy with THA have been found to be superior to those from synovectomy alone in prior trials.[11,12]In their eight cases that were treated with cementless THA over an average follow-up of 8.9 years, Yoo et al. observed no recurrent PVNS.[13] They came to the conclusion that THA combined with synovectomy is a suitable therapy option for PVNS patients who exhibit end-stage joint degradation and that it appears to significantly enhance clinical outcomes and avoid disease recurrence.

The histology of Pigmented Villonodular Synovitis (PVNS) of the hip joint typically shows synovial proliferation, which is the abnormal growth of synovial tissue. This tissue can appear as either a diffuse, thickened lining or as nodular masses within the joint. The synovial tissue is composed of different types of cells, including macrophages, fibroblasts, and multinucleated giant cells. The synovial tissue in PVNS is characterized by an abundance of hemosiderin-laden macrophages, which give the tissue a brownish color and is responsible for the term "pigmented." This hemosiderin deposition is due to bleeding within the synovium, which is a common feature of PVNS. Under the microscope, the nodular masses in PVNS may appear as well-circumscribed lesions composed of synovial lining cells, histiocytes, and multinucleated giant cells. These nodules may invade and destroy surrounding joint structures, leading to joint damage and pain.

CONCLUSION

PVNS of the hip joint can be challenging to diagnose due to its

non-specific symptoms and rare occurrence. Early recognition and appropriate treatment can lead to improved outcomes and prevent long-term joint damage. In this case with atypical presentation, THA with total synovectomy was a successful treatment option for PVNS of the hip, as it allowed for removal of the abnormal synovial tissue and reconstruction of the joint.

In conclusion, this case report highlights the importance of considering PVNS in the differential diagnosis of hip pain and stiffness in young adults, especially in the absence of a clear history of trauma or prior surgery. THA with total synovectomy can be an effective treatment option for PVNS of the hip, leading to resolution of symptoms and improved joint function. Long-term follow-up is necessary to monitor for recurrence of the condition.

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