

CASE REPORT

An atypical case of a massive extra-articular pigmented villonodular synovitis

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Abstract

Pigmented villonodular synovitis (PVNS) is an idiopathic thickening of the synovium in form of nodules and villi. It commonly affects the knee. The synovium has fat and haem deposits. On MRI it appears heterogenous and its treatment is almost always a total synovectomy

A 62-year-old female presented with a painless right knee swelling that was predominantly on the posterior aspect with some occasional mechanical symptoms of locking and catching with transient pain. MRI reported a large heterogenous hypointense mass that was 17.4cm by 9.0cm by 6.7cm. Biopsy confirmed PVNS and on surgery over and above the intra-articular component and a single large encapsulated posteriorly-located mass excised which was extra-articular.

This particular case of a massive extra-articular PVNS is extremely rare and an important learning point while managing these cases with arthroscopy.

Keywords: pigmented villonodular synovitis, extra-articular, synovectomy, arthroscopy, knee

Introduction

Pigmented villonodular synovitis (PVNS) is a rare, benign, monoarticular, proliferative neoplastic condition that presents with synovial thickening in the form of nodules and villi, with pigmentation due to haemosiderin deposits. It was previously thought to occur secondary to trauma and chronic inflammation but recent literature suggests that it is a neoplasm because of evidence of chromosome 5q33 rearrangement (1). It typically is mono-articular and intra-synovial, commonly affecting the knee(2)(3). PVNS is either localized or diffuse affecting synovium of a joint or bursa and on MRI it appears heterogenous on T1 and T2 weighted images(4). Its treatment is almost always synovectomy whether its intra or extra-articular with a recurrence rate of about 30%(2). Synovectomy can be done open or arthroscopic depending on the extent and location of disease. There is usually an intra-articular focus of the disease in presence of extra-articular disease(2).

The authors report a case whereby the pathology is both intra and extra-articular but predominantly extra-articular and it's extremely rare to encounter such a large extra-articular PVNS mass.

Case presentation

A 62-year-old female patient had been having a relatively painless right knee swelling that is progressively increasing in size over the last 4 years. She had been experiencing discomfort and reduced motion of the knee but had no history of trauma. She was able to walk but with difficulty climbing stairs due to the limited flexion. She had no instability but had occasional locking. She also did not have any numbness or altered sensation in the right lower limb.

Her general examination was normal. The knee swelling was predominantly on the posterior aspect of the knee - approximately 15cm by 15cm in size, rounded, asymmetrical, smooth, normal overlying skin, not pulsatile and no transillumination. The inguinal lymph nodes were not enlarged with normal neurovascular examination. The right (affected) knee had reduced range of motion of 5-100 but the left was Normal (0-130)

The patient had normal blood works which included CBC, UEC, Alkaline phosphatase and LFTs. An MRI was ordered and showed the below picture.

MRI findings suggested a gross synovial thickening with posterior capsular distension that gives a heterogenous signal that is principally hypointense on T1 and T2 images. There is also an extra-capsular extension of the mass. The

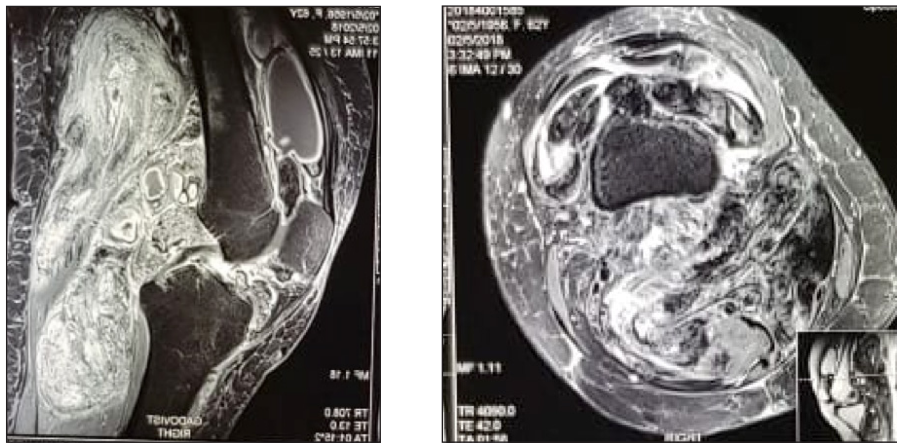


Figure 1. Magnetic resonance images of the right knee

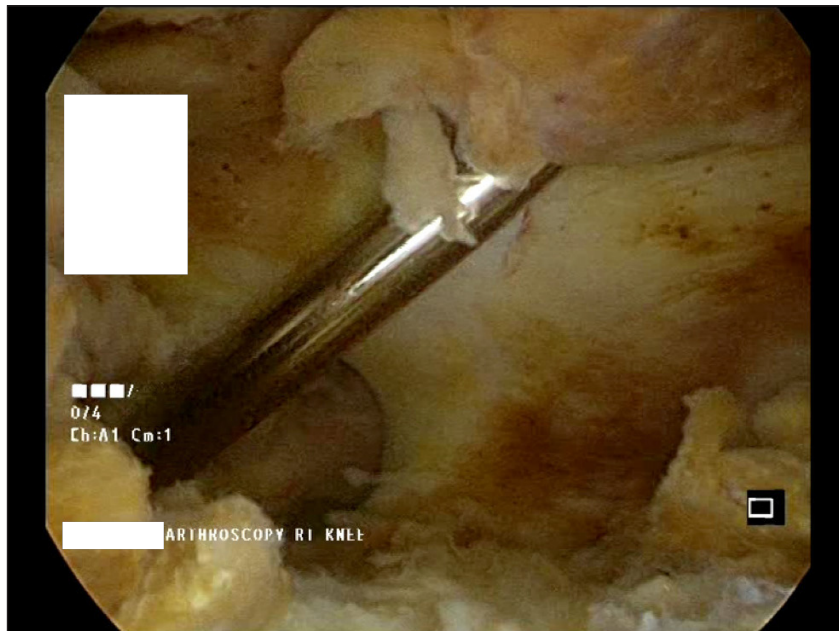


Figure 2. Right knee arthroscopy

mass is measuring 17.4cm (CC) by 9.0cm (W) by 6.7cm (D). This was found to be suggestive of a pigmented villonodular synovitis with extra-capsular extension.

A core biopsy was done at 4 different sites on the posterior aspect of the mass and the pathologist confirmed the mass to be pigmented villonodular synovitis.

Consent for surgical management was sought for an anterior arthroscopic synovectomy and posterior open synovectomy. Intra-operatively on arthroscopy a hemarthrosis was drained at first and the synovium was noted to be brownish and inflamed. Synovectomy was then performed.

The patient was then put in a prone position and a posterior approach to the knee was done and a large encapsulated villo-nodular lesion was carefully dissected out and excised

completely. All neurovascular structures were found intact and no obvious communication noted into the joint space. The mass excised was approximately 20cm by 10cm. Haemostasis was meticulously achieved after letting the tourniquet down, closure was done in layers and no drain was put.

Post-operatively, the patient started ambulating the next day and did not have any complications. The range of movement was at 0-110 and mobility improved greatly. There was no wound or neurovascular compromise. At 3 months, post-op the patient reported minimal discomfort of the knee but there was no clinical evidence of recurrence. Long term follow up is planned both clinically and radiologically. The excised mass was sent to the pathologist for further examination and histology, and was reconfirmed as pigmented villonodular synovitis.

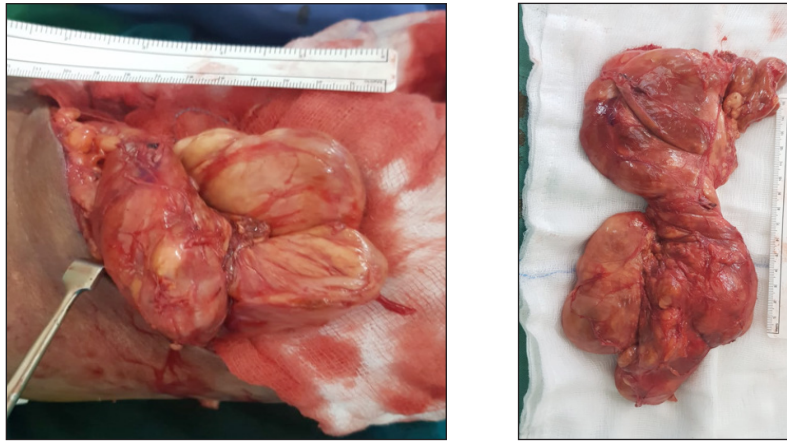


Figure 3. Images of the excised mass from the posterior aspect of the right knee

Discussion

The incidence of extra-articular PVNS is reported to be 1.8 cases per year per million individuals (3)(5) which is extremely rare. It commonly occurs in the younger age group (20 to 50 years)(4,6) and the ratio between male to female is 1:1.5-2 for the extra-articular disease(5,7,8). PVNS was described by Jaffe et al(1) in 1941 and described it as a benign proliferative inflammatory manifestation. It is predominantly found in the knee joint (80%)(3,5,9–11). PVNS can extend extra-articular in a tendon sheath or bursa(2). It is characterized by hemosiderin deposition with multinucleated giant cells. Patients present with vague symptoms like discomfort, pain, swelling and reduced range of motion.

The pathogenesis of this disease is uncertain but the theories surrounding it suggests that it is associated with repeated hemarthrosis due to multiple mild trauma or a lipid metabolism disorder(4). PVNS can be localized or diffuse depending on the extent of hemosiderin deposition. If hemosiderin deposition is in a localized area then the synovial lining thickens with hemosiderin deposits forming a discrete nodule. In the diffuse form the entire synovial lining is involved. The genetic theory suggests a 5q33 chromosomal rearrangement due to increased expression of CSF1 gene.(12)

Clinical presentation can often be non-diagnostic hence imaging is the choice of investigative modality. MRI often shows characteristic features of low signal intensity in both T1 and T2 images due to hemosiderin deposits. There is also a signal loss on gradient echo sequences (2). The nodular form can cause impingement within the joint and cause mechanical symptoms like locking or catching. The diffuse form can cause reduced range of motion, pain, occasional hemarthrosis and joint degenerative changes.

Treatment of this condition is almost always total synovial resection. Adjuvant medications is still under research with the aim to reduce recurrence. Radiotherapy has not been very successful which ends up in treatment failure and has complications like wound breakdown and osteonecrosis(5).

There is a high recurrence rate after operative treatment but it rarely becomes malignant(10). Surgical excision is the treatment of choice and an arthroscopic technique is recommended for anterior and an open technique for the posterior disease. However, recurrence rate is higher for the arthroscopic approach (46%)(2).

This particular case is atypical because it is a massive posteriorly based extra-articular lesion that is well capsulated with an intra-articular component that is anterior. We believe that this neoplasm occurred posteriorly in a bursa which would justify the term pigmented villonodular bursitis (PVNB) in this case.

Conclusion

PVNS is a rare benign neoplasm can present in an atypical manner around the knee and in this case predominantly in the posterior aspect that can be excised with its capsule safely.

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