

## Case Report:

# Pigmented Villonodular Synovitis (PVNS) – A Case Report

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### Abstract:

Pigmented villonodular synovitis (PVNS) is benign proliferative disorder of the synovium of unknown origin. PVNS is divided into intra-articular and extra-articular form which may present as i) diffuse or ii) localised/nodular form. It is commonly seen in knee joint but hip, ankle, shoulder, wrist & other joints can be involved. It is usually seen in young adults from 20-50 years of age. It is seen more in men than women. A 26 years old female presented to our OPD with chief complaints of pain & swelling over right knee since 1 year. MRI right knee was suggestive of synovial thickening & collection in anterior aspect of knee with peripheral hemosiderin deposits which is suggestive of PVNS. Arthroscopic resection of this lesion was performed & histological report was revealed presence of giant cells & hemosiderin laden macrophages which are suggestive of PVNS. The case is presented because of unusual involvement in a female. Milwaukee Shoulder Syndrome (MSS) may be associated with PVNS.

**Keywords:** Pigmented Villonodular Synovitis (PVNS), Arthroscopy, Milwaukee Shoulder Syndrome

### Introduction:

Pigmented villonodular synovitis (Synovial Xanthoma / Villous Synovitis) is characterised by idiopathic villous overgrowth & pigmentation of synovial membrane. This condition is usually seen in young adults from 20 – 50 years of age.<sup>[1]</sup>

Pigmented villonodular synovitis (PVNS) may be of diffuse or localised form. The diffuse form occurs in 2nd & 3rd decades and the localised / nodular form occurs in 3rd & 5th decades. About 80% cases are diffuse and 20% cases are localised form.<sup>[2]</sup>

PVNS is divided into types -

1. intra-articular.....Pigmented villonodular synovitis

2. extra-articular form .....Pigmented villonodular bursitis

Depending on location of synovial membrane involved. Pigmented villonodular tenosynovitis for lesions of tendon sheath is known as Giant Cell Tumour (GCT) of tendon sheath.<sup>[3]</sup> It is more commonly seen in men.<sup>[4]</sup> Incidence of 1.8 per 1 million per year.<sup>[2,5]</sup> The treatment of choice is synovectomy either open or arthroscopic. Local recurrence rate is 18 to 46%.<sup>[6,7]</sup> Post surgical external radiotherapy may be required in some cases, as recurrence rate is high.<sup>[8]</sup>

### Case report:

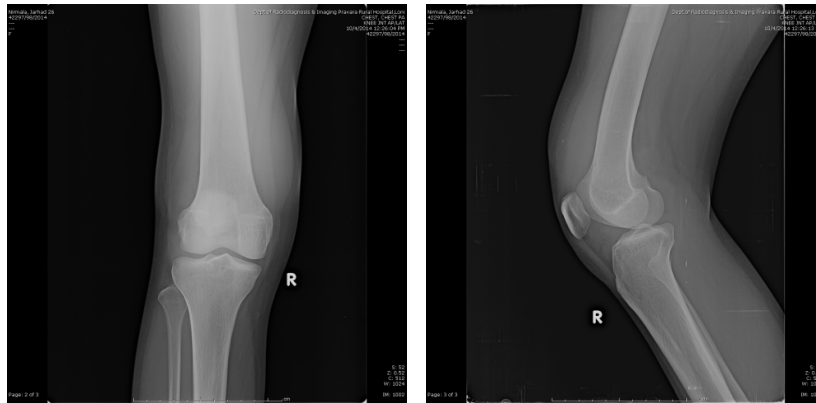
A 26 years old female presented to our hospital with 1 year history of gradually increasing

swelling & mild pain in right knee without symptoms of mechanical obstruction. There was no history of trauma / fever.

On clinical examination, she had normal gait, moderate right knee effusion, and mild tenderness over medial joint line with no

restriction of movements. There was no evidence suggestive of with meniscal, or ligament injury or joint instability. Thickened synovium was palpable. Family history was not significant. Blood investigations were within normal limits.

Conventional radiography of right knee (AP & LAT view) was within normal limits (As shown in Fig.1).



**Fig 1:** X-ray Rt. Knee AP & Lateral view

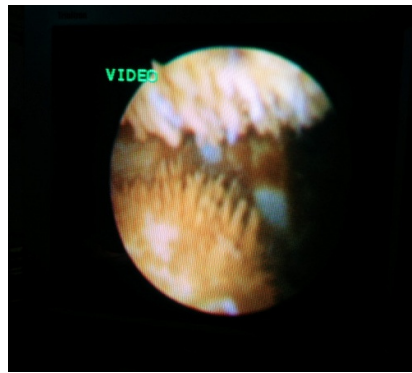
MRI right knee was suggestive of synovial thickening & collection in anterior aspect of knee appearing hyperintense on STIR & T2W images and peripheral hemosiderin deposits appearing hypointense on GRE, STIR & T2W

images. Bony erosions noted on medial and intercondylar region of tibia. Nodular structure measuring 2.9 x 2.2 cm noted in posterior aspect of knee (As shown in Fig. 2).



**Fig 2:** MRI images of Rt. Knee suggestive of PVNS

On arthroscopic examination of right knee, we found hemorrhagic effusion & pigmented proliferation with villous appearance of synovial tissue (As shown in Fig.3).

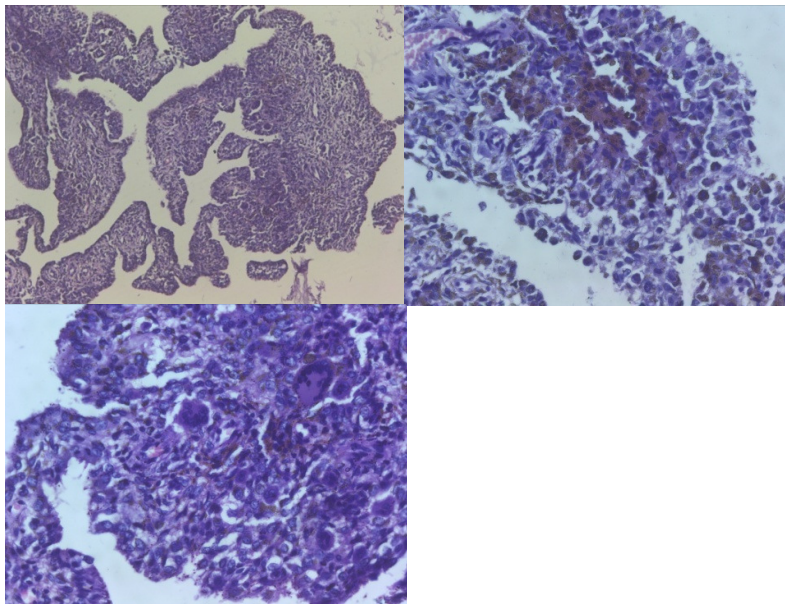


**Fig 3:** Arthroscopic image of Rt. Knee of PVNS

Resected material was sent for histopathology. Histopathological report revealed -

- i) Villous hypertrophy.
- ii) Synovial proliferation with histiocyte like cells.
- iii) Cells are small & round to oval in shape.
- iv) Coffee-bean shaped nuclei seen.
- v) Diffuse presence of hemosiderin laden macrophages.
- vi) Few Giant cells are also seen.

All these features were consistent with Pigmented villonodular synovitis (As shown in Fig.4).



**Fig 4:** Histopathological slides showing giant cells & hemosiderin laden macrophages

Post-operatively patient recovered well & is ambulatory without walking aids and full resolution of symptoms. She is under follow up.

**Discussion:**

PVNS is benign proliferative disorder of the synovium of unknown etiology.<sup>[9]</sup> The probable theories regarding proposed are-

- i) repeated hemarthrosis,
- ii) benign neoplastic processes,
- iii) reaction to unknown stimuli & to repeated episodes of trauma<sup>[6,10]</sup>

PVNS is diagnosed on an average, 4-5 years after presentation.<sup>[11]</sup> Incidence of PVNS is 1.8 per 1 million population per year.<sup>[2,5]</sup> PVNS presents as monoarticular lesion & knee joint is most commonly involved but it is also seen in hip, ankle, shoulder, wrist & other joints. This condition is usually seen in young adults from 20-50 years of age.<sup>[1]</sup> PVNS may be diffuse or localised form. The diffuse forms occur in 2nd & 3rd decades & the localised / nodular forms occur in 3rd & 5th decades. Ratio of diffuse to localised form is 80:20.<sup>[2]</sup> PVNS is broadly divided into intra-articular & extra-articular form depending on location of synovial membrane involved. Usually PVNS term is used for intra-articular form, Pigmented villonodular bursitis for localised lesions of bursas & pigmented villonodular tenosynovitis for lesions of tendon sheath which is also known as Giant Cell Tumour (GCT) of tendon sheath.<sup>[3]</sup> Prevalence is higher in men.<sup>[4]</sup>

On physical examination of patient with PVNS of knee will reveal-

- One / more palpable nodules or diffuse joint swelling.
- Around 96 % of patients present with a large effusion & distension of suprapatellar pouch

- 40 % of patients have diffuse palpable synovial mass.
- Mild to moderate tenderness over medial patellofemoral area.
- Aspiration of joint is reveals serosanguineous or blood tinged fluid.

Plain X-ray shows no abnormality but may show bony erosion. Bony erosions vary according to location of joint & its volume. The pressure exerted by the space occupying lesion leads to atrophy & erosive changes.<sup>[12,13,14]</sup> MRI is superior investigation for diagnosis & for planning treatment. Areas of low signal are observed with T<sub>1</sub> & T<sub>2</sub> weighting in the synovial membrane which are due to classic hemosiderin deposition of PVNS.<sup>[12,14]</sup>

Histological findings show presence of polyhedral cells surrounded by collagen. Giant cells, foam cells, fibrous tissue & hemosiderin are also present. The treatment of choice for localised / nodular form is marginal excision & diffuse form is total synovectomy. Synovectomy can be either open or arthroscopic. Arthroscopy causes less injury to structures & leads to earlier recovery.<sup>[6,10]</sup> Recurrence is common but are adequately handled by adjuvant radiation therapy following primary resection. Malignant transformation is rare. Differential diagnosis of PVNS are- DJD, tuberculosis, haemophilia, amyloidosis, gout, synovial chondromatosis.<sup>[15]</sup>

**Conclusion:**

PVNS is rare entity in orthopaedic practice. Commonly seen in males of middle age, but we suggest considering PVNS in females of middle age with no history of trauma and knee effusion.

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