Diffuse type of pigmented villonodular synovitis of knee masquerading as a soft tissue sarcoma

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ABSTRACT

Pigmented villonodular synovitis (PVNS) is a rare benign neoplasm involving the synovial joints and tendon sheaths. It can be classified into localised and diffuse type. The diffuse type of PVNS is larger and very aggressive, clinically mimicking as soft tissue sarcoma. Radiological investigation can aid in knowing the relation of the mass to the joint cavity and invasion of surrounding structures. A biopsy can pose a diagnostic challenge for pathologists. Surgical excision is the treatment of choice. Histopathological examination is necessary for a definite diagnosis.

Keywords: Pigmented villonodular synovitis; diffuse; soft tissue sarcoma; histopathology.

INTRODUCTION

Pigmented villonodular synovitis or tenosynovial giant cell tumour is a rare, benign neoplasm associated with synovial proliferation and hemosiderin deposition inside the joints, tendon sheaths, and bursae (1, 2). An average incidence of 1.8 cases per 1,000,000 population has been reported. It typically presents between the second and fifth decades of life and has a slight female predominance (1, 3, 4). PVNS presents in two forms: localised and diffuse-type, with the latter being less frequent, more destructive with a higher rate of recurrence (1, 5). Localised PVNS are smaller masses commonly involve fingers (85%), followed by wrist, ankle, and knee joint. In contrast, diffuse PVNS is more massive and often occurs in the knee joints (80%), followed by hip, ankle, shoulder, and elbow joint (5). Histologically, the mononuclear component consists of two cell types, small histiocyte-like cells and large cells. When the tumour is large, and the large cells are predominant, they can be easily mistaken for a sarcoma (1). Diffuse PVNS has a poor prognosis as compared to localised type owing to increased risk of recurrence. Herein, we present a rare case of diffuse PVNS in a 55-year-old female who presented with swelling over the right calf.

CASE REPORT

A 55-year-old female presented with swelling on the right calf region for four years. The swelling was insidious in onset, initially the size of a small lemon, which gradually progressed in size. She gave a history of warmness over the swelling and intermittent pain. There was no history of numbness or multiple joint pains. However, recently she felt difficulty in doing her daily activities and hence presented to the hospital seeking medical advice. On examination, a solitary firm, mobile swelling measuring 10cm X 8cm (Fig. 1), was seen over the right calf. Mobility was restricted to the horizontal plane on contracting the gastrocnemius muscle. Sensation and power were normal. Provisional clinical diagnosis of soft tissue sarcoma was made. X-ray of the right knee showed soft tissue density in the posteromedial aspect of the distal third of the thigh and upper third of the leg without any bone erosions (Fig. 2). Magnetic resonance imaging (MRI) showed a well-defined, lobulated, enhancing lesion measuring 20cm X 11cm X 9cm in the posteromedial aspect of the upper third of the leg and lower third of the thigh, involving the intermuscular plane between semimembranosus and biceps femoris muscles, which is communicating with the knee joint (Fig. 3A & 3B). After making a clinical diagnosis of soft tissue sarcoma, incisional biopsy from the mass was done, which showed sheets of foamy histiocytes, plasma cells, lymphocytes, and occasional giant cells. Extensive areas of fibrosis, fibrinoid necrosis, and hemorrhage were noted. As definite diagnosis was not possible, open excision of the right knee mass was done. The excised mass was well-circumscribed, partly encapsulated, pale brown, and nodular, measuring 20cm X 13cm X 7cm with attached skin tissue measuring 1.5cm X 2cm. Adipose tissue and tendon were also seen. The cut surface of the mass showed a solid tumor with pale white to yellow-brown areas (Fig. 4). Focal glistening areas with cystic degeneration and hemorrhage were also noted.

Histopathology showed numerous villous fronds lined by hyperplastic synovial epithelial cells and histiocytes (Fig. 5). Villous fronds showed a good amount of hemosiderin-laden macrophages and hemorrhage (Fig. 6). Stroma was very cellular, showing closely packed, medium-sized polyhedral cells containing round to oval, plump nucleus with

fine chromatin, and pale to eosinophilic cytoplasm. Osteoclast-like giant cells were also seen (Fig. 7). Hypocellular areas showed hyalinised collagen. Xanthomatous change showing histiocytes were seen at the periphery (Figure 8). Extensive inflammation composed of lymphocytes and plasma cells was observed. The periphery of the tissue showed tendon sheath. Diagnosis of diffuse-type- Pigmented villonodular synovitis- right popliteal fossa was made. The post-operative period was uneventful and is doing well during nine months follow-up.

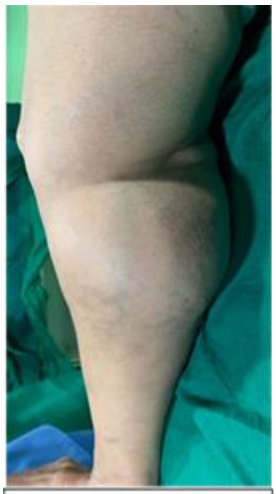


Fig. 1: Clinical image showing swelling in the right calf region, posteromedial to the knee joint

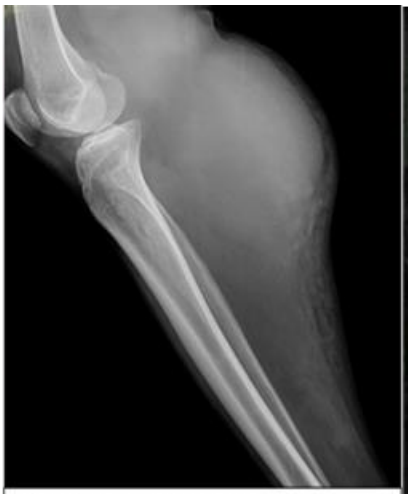
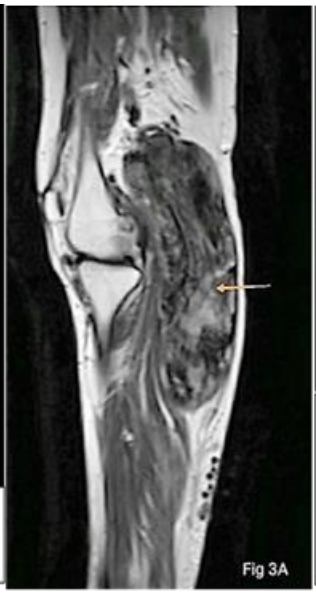


Fig. 2: X-ray of the right knee showing soft tissue density in the posteromedial aspect of the distal third of the thigh and upper leg.



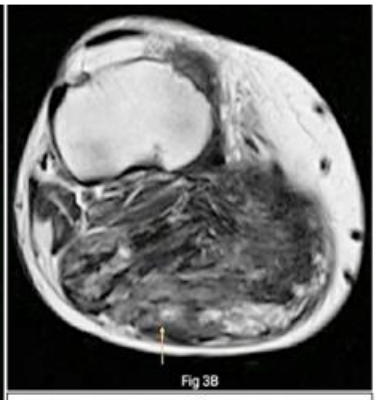


Fig. 3A: MRI- Sagital T2 view of the right knee showing multilobulated high-signal intensity lesion (arrow) in the posterior compartment communicating with the knee joint.
Fig. 3B: MRI- Axial view (T2) of the right knee showing multilobulated lesion (arrow) posterior to the distal femur.

Fig. 4: Gross specimen - the cut surface of the mass showing pale white and grey-brown areas.

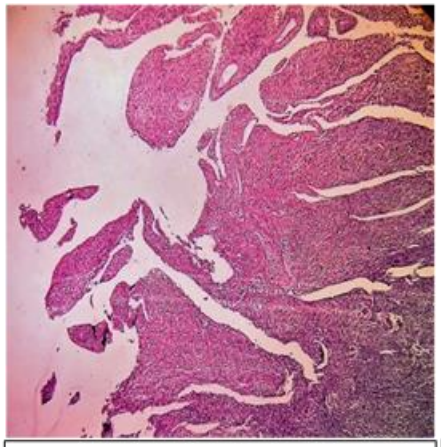


Fig. 5: Synovial tissue showing villous fronds lined by hyperplastic synovial cells (H&E, 10x)

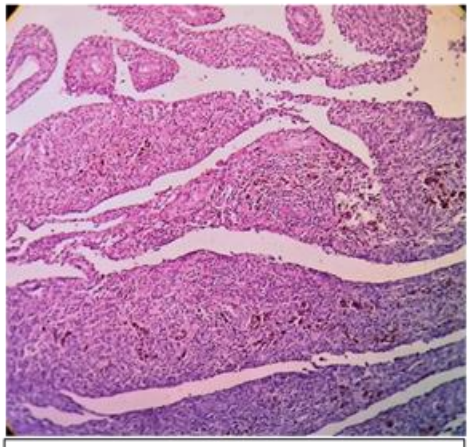


Fig. 6: Villous fronds showing hemosiderin-laden macrophages and chronic inflammatory cells (H&E, 10x)

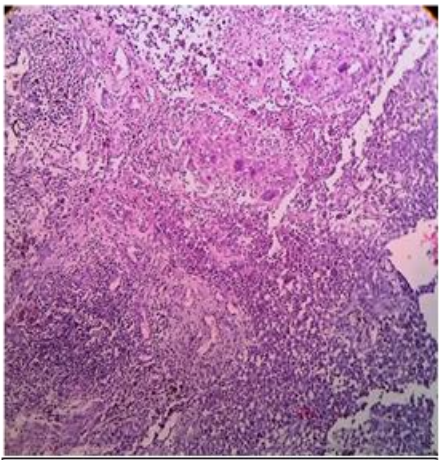


Fig. 7: Closely packed polyhedral cells containing round to oval, plump nucleus along with osteoclast-like giant cells (H & E, 10x)

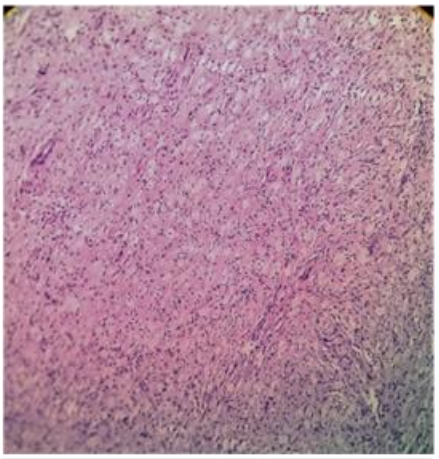


Fig. 8: Xanthomatous reaction (H & E, 10x)

DISCUSSION

Pigmented villonodular synovitis or tenosynovial giant cell tumour was first described in 1852 by Édouard-Pierre-Marie Chassaignac as nodular damage of synovial membrane which was subsequently termed as "pigmented villonodular synovitis" by Jaffe *et al.,* in 1941 (6). It typically occurs between the second and fifth decade of life with a peak incidence in the second and third decade and has a female preponderance [3, 4]. PVNS can present in two forms: Local and diffuse type. Localised PVNS most commonly involves the fingers (85%) and the wrist joint. Rarely, it may affect the anterior compartment of the knee. Diffuse PVNS is less common and has a destructive growth pattern (1, 5). It most commonly involves the knee joints (80%) followed by the hip, ankle, shoulder, and elbow joint (5). Unlike localised PVNS, diffuse PVNS is larger and involve the entire synovial tissue of the knee and can extend extra-articularly and present as soft tissue mass. Cases involving small joints of the spine and temporomandibular joints have been reported (6). Diffuse PVNS is usually monoarticular, with sporadic cases involving multiple joints. Patients with diffuse PVNS present with pain and edema of the involved joint, which increases with the disease advancement, further progressing to joint stiffness and limited mobility (7). Our patient was a 55-year-old female who presented with swelling in the right calf with intermittent pain of 4-year duration with a recent history of difficulty in performing her daily activities.

Imaging such as X-ray, Ultrasonography (USG), CT-scan, and MRI can aid in diagnosis. Plain radiograph of knee helps to detect bone erosions; however, have less sensitivity. USG can detect the solid or cystic nature of the mass. CT scan will show increase density due to hemosiderin deposition and hyperplastic synovium. A low signal- blooming effect in the T2 sequence of MRI is highly suggestive of PVNS (3, 6). In our patient, X-ray of the right knee showed soft tissue density in the posteromedial aspect with no bony erosions. MRI showed a well-defined, lobulated, altered signal intensity lesion in the posteromedial aspect of the upper third of the leg extending to the lower third of the thigh communicating with the knee joint. The lesion appeared hypointense on T1 and heterogeneously hyperintense in T2 sequences with heterogeneous post-contrast enhancement. These features were suggestive of soft tissue neoplasm favouring soft tissue sarcoma was, which prompted incisional biopsy from the mass, histopathology of which showed features of inflammatory pseudotumour.

Surgical excision is the treatment of choice for Diffuse PVNS. Open surgical excision is performed for tumours with extra-articular extension, while arthroscopic synovectomy is preferred for intra articular mass. The best results have been shown by combining both procedures followed by radiotherapy with a local recurrence rate of only 12% (7).

Radiotherapy can be considered in patients with local relapse and in whom complete resection is not possible. Radiation doses of 20–50 Gy are supplied in 15–25 fractions, six weeks–8 weeks following surgery (6). In our case, as there was a strong suspicion of sarcoma clinically and radiologically owing to the huge size, the surgeons decided to perform open excision of the mass to relieve the extra-articular compression effects.

Grossly, diffuse PVNS are large nodular, partially capsulated mass measuring >5cm (8). On cut section, the mass is usually solid and firm with yellow-brown discolouration due to hemosiderin deposition giving a variegated appearance (8). Microscopically, the hyperplastic synovium shows finger-like papillary processes with fibrous stroma. The stroma comprises of compact hypercellular areas alternating with pale, loose discohesive zones. Hypercellular areas show two types of cells: small histiocytes like cells with round to ovoid nucleus with longitudinal groove and larger cells with abundant eosinophilic cytoplasm, reniform nucleus, and a peripheral rim of hemosiderin granules. Sheets of foam cells, osteoclastic giant cells, and hemosiderin-laden macrophages are also seen (5, 9).

Although PVNS and diffuse type of tenosynovial giant cell tumor have a close resemblance histologically, the presence of synovial tissue thrown into villous fronds having hemosiderin pigment favours PVNS. Increased mitotic figures, marked nuclear hyperchromasia, spindling of mononucleated cells, abundant eosinophilic cytoplasm, and stromal myxoid change favours malignancy (1).

Other differentials include deep benign fibrous histiocytoma and giant cell tumor of soft tissue. Deep benign fibrous histiocytoma occurs in the extremities. They are well-circumscribed tumors, although deep-seated ones are larger. Histologically, they are cellular tumours with a storiform architecture having hemangiopericytoma like areas, composed of spindle to plump, ovoid to elongated cells with vesicular nuclei and indistinct pale to eosinophilic cytoplasm. Nearly half of the cases lack foamy histiocytes and giant cells. Giant cell tumour of soft tissue usually occurs in the superficial soft tissue of upper and lower extremities. They have a multinodular architecture with fibrous septa dividing the nodules containing hemosiderin-laden macrophages. The nodules contain mononuclear stromal cells and osteoclast-like giant cells. Mitosis, foamy histiocytes, and aneurysmal bone cyst-like areas may also be seen (1).

In our case, following the open excision of the mass, the patient is doing well for nine months.

CONCLUSION

Diffuse pigmented villonodular synovitis is a rare benign tumor involving the synovium and tendon sheaths around the knee joint. As it is a large tumor causing compression effects, it is mistaken for a malignant lesion. The radiological investigation, including MRI aids, to delineate the mass with surrounding structures. An adequate incision biopsy would help in knowing the nature of the mass. Surgical excision is the treatment of choice. The histopathological examination will give a definite diagnosis.

CONFLICT OF INTEREST

Authors declare no conflict of interest.

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