

LOCALIZED NODULAR SYNOVITIS OF THE ANKLE – A CASE REPORT



Orthopaedic

KEYWORDS: Pigmented Villo Nodular Synovitis (PVNS), Ankle joint, excision biopsy, localized nodular synovitis.

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ABSTRACT

Pigmented villonodular synovitis (PVNS) is a synovial proliferative disorder which is often a diagnostic difficulty because of its nonspecific presentation and subtle radiographic findings. It is of two types – nodular and diffuse. Its occurrence in the ankle joint, especially of the nodular type is extremely rare. We report a case of localized nodular synovitis of the ankle in a middle aged female treated by radical excision alone.

INTRODUCTION:

Pigmented Villo Nodular Synovitis (PVNS) represents a benign, hypertrophic synovial process characterized by villous, nodular and villo-nodular proliferations and pigmentation from hemosiderin. These components vary in prominence from lesion to lesion. The knee joint followed by hip are the common sites of villo nodular synovitis. The reported annual incidence of villo nodular synovitis in the joints is only 1.8 cases per 1 million. Of this only 21% is of nodular type and is almost always seen in the knee joint. Even 82 to 90% of the diffuse type cases are seen in the knee (66 to 74%) and hip (16%) joints. We came across only two case reports of nodular PVNS in the ankle, one from the USA and another from Germany. We report an extremely rare case of nodular type of PVNS in the ankle joint for the first time in the Indian literature.

CASE REPORT:

A 45 year old female presented with a long standing and slowly growing swelling in the left ankle for 6 months, associated with pain during walking for one month. There was no history of antecedent injury and the patient denied any fever, chills, weight loss, malaise and other joint pains.

The patient had effusion, joint line tenderness and painful restriction of movements in the left ankle. No distinct mass was palpable but the patient had a more prominent fullness in the anterior aspect of the ankle.

Arthrocentesis revealed a hemorrhagic fluid. Ankle radiographs were normal. MRI revealed a nodular soft tissue mass with low signal intensity in T1 and T2 weighted images (Fig 1 & 2).



Fig 1: MRI ankle, T1 weighted image.



Fig 2: MRI ankle, T2 weighted image.

The patient was taken up for excision biopsy through the anterior approach to ankle. A 3*4 cm nodular mass was found arising from the synovium in the anterior aspect of the joint (Fig 3). The pedicle was ligated and the mass excised in toto. Histopathology showed multiple histiocytes containing haemosiderin pigment and numerous multinucleated giant cells, confirming it as nodular type of pigmented villonodular synovitis (Fig 4).

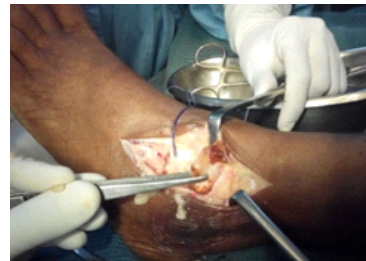


Fig 3: Intra op picture showing nodular mass.

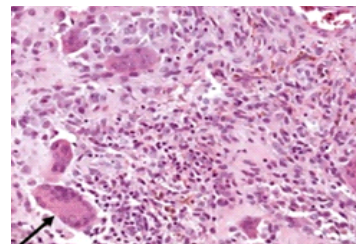


Fig 4: Histology slide showing multiple haemosiderin laden histiocytes.

Post operatively we splinted the ankle using a below knee plaster slab until suture removal. Then the patient was mobilized with a removable ankle brace and she resumed full weight bearing by three weeks. The brace was continued for six weeks. The patient was reviewed fortnightly until six weeks after which she was reviewed at 3,6,12 and 24 months. After six weeks the patient resumed full functional capacity with the ankle having a painless full range of motion. Now after two years of follow up there is no clinical evidence of recurrence and the patient is walking without pain.

DISCUSSION:

Pigmented Villo Nodular Synovitis (PVNS) is a locally destructive fibrohistiocytic proliferation, characterized by many villous and nodular synovial protrusions, which affects joints, bursae, and tendon sheaths. PVNS was first described by Jaffe, Lichtenstein, and Suto in 1941. "The yellow-brown pigmentation is caused by excessive deposits of lipid and hemosiderin"[1].

"This condition can be diffuse or localized"[2]. When the entire synovium of the joint is affected, and when there is a major villous

component, the condition is referred to as diffuse pigmented villonodular synovitis (23%). When a discrete intraarticular mass is present, the condition is called localized pigmented villonodular synovitis (6%). When the process affects the tendon sheaths, it is called localized giant cell tumor of the tendon sheaths (72%).

Both the diffuse and the localized forms of villonodular synovitis usually occur as a single lesion, mainly in young and middle-aged individuals of either sex. One of the most characteristic findings in PVNS is the ability of the “hyperplastic synovium” to invade the subchondral bone, producing cysts and erosions”[2]. Although the cause is unknown and is often controversial, some investigators have suggested an autoimmune pathogenesis. Trauma is also a suspected cause.

Clinically, PVNS is a slowly progressive process that manifests as mild pain and joint swelling with limitation of motion. Occasionally, increased skin temperature is noted over the affected joint. “The knee joint is most commonly affected and 66% of patients present with a bloody joint effusion”[2]. In fact, the presence of a serosanguinous synovial fluid in the absence of a history of recent trauma should strongly suggest the diagnosis of PVNS. The synovial fluid contains elevated levels of cholesterol, and fluid reaccumulates rapidly after aspiration. Other joints may be affected, including the hip, ankle, wrist, elbow, and shoulder. “There is a 2:1 predilection for females”[1]. Patients range from 4 to 60 years of age, with a “peak incidence in the third and fourth decades”[3]. The duration of symptoms can range from 6 months to as long as 25 years.

Radiography reveals a soft-tissue density in the affected joint, frequently interpreted as joint effusion. However, the density is greater than that of simple effusion, and it reflects not only a hemorrhagic fluid but also lobulated synovial masses. A marginal, well-defined “erosion of subchondral bone with a sclerotic margin”[3] may be present (incidence reported from 15% to 50%), usually on both sides of the affected articulation.

Arthrography reveals filling defects in the contrast-filled joint. CT effectively demonstrates the extent of the disease. The increase in iron content of the synovial fluid results in high Hounsfield values; a feature that can help in the differential diagnosis. MRI is extremely useful in making a diagnosis, because on T2-weighted images the intra-articular masses demonstrate a combination of high-signal-intensity areas, representing fluid and congested synovium, interspersed with areas of intermediate to low signal intensity, secondary to random distribution of hemosiderin in the synovium. In general, “MRI shows a low signal on T1- and T2-weighted images because of hemosiderin deposition and thick fibrous tissue”[2].

The other common diagnostic possibilities include “hemophilic arthropathy, synovial chondromatosis, and synovial hemangioma” [3].

“Local recurrence is uncommon in the nodular type and is around 50% in the diffuse type” [6]. The treatment for localized form is marginal excision and for the diffuse form is total synovectomy. “Radiotherapy (radiosynoviorthesis) in the diffuse type may be justified if synovectomy alone fails to control the disease process” [4]. Following total synovectomy the patients are usually subjected to a radiation of “34 Gy in 15 doses over a 3-week period” [4]. Local skin problems like hyperpigmentation, pruritus and eczema are common which usually heal with local care.

CONCLUSION:

Considering the rarity of villo nodular synovitis in the ankle, its diagnosis warrants strong clinical suspicion and thorough investigation. Though Ganse et al recommended “radiosynoviorthesis in addition to excision”[5] we treated our patient with excision alone since the joint was completely normal except for the solitary nodule which we could excise in toto. Close followup of our patient for 2 years has shown no clinical signs of recurrence and she remains

symptom free. Hence radical excision alone can be curative in cases of localized nodular synovitis while in cases of diffuse PVNS a radiosynoviorthesis following total synovectomy may be needed to eradicate the persisting PVNS cells and thereby prevent further recurrence.

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