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Case Report

# Pigmented Villonodular Synovitis of the Ankle: A rare localization

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**Abstract:** Pigmented villonodular synovitis (PVNS) is a rare benign proliferative growth of the synovium of obscure aetiology. PVNS may present as a localized nodule, usually arising in the hand, or as a diffuse process, usually found in the knee. The optimal treatment is surgery. As surgery may not eliminate all affected tissue, postoperative radiosynoviorthesis is discussed to remove residua of the synovial membrane. There is a low rate of malignant change but the lesion has the ability for reactivated growth and a tendency to recur. This paper reports the case of a rare localizated Pigmented villonodular synovitis of the ankle.
**Keywords:** pigmented villonodular synovitis, ankle, synovectomy.

## INTRODUCTION

Pigmented villonodular synovitis (PVNS), first termed by Jaffe et al in 1941, is a rare benign proliferative growth of the synovium of obscure aetiology [1]. The estimated annual incidence is 1.8 patients per million with no significant gender predisposition. It occurs most commonly in the knee but rarely in the ankle [1, 2].

Histology of PVNS reveals hypertrophic synovial process characterized by villous, nodular, and villonodular proliferation and pigmentation from hemosiderin [1-3].

There are two forms of PVNS: diffuse and localized. When the entire synovium is affected, the condition is referred to as diffuse PVNS (DPVNS). When a single discrete mass is present in the synovium, it is called localized PVNS. Both forms may arise from intra-articular or extra-articular synovial tissues [3-6].

Therapeutic options involve radical synovectomy or arthroplasty if joint destruction has occurred. There is a low rate of malignant change but the lesion has the ability for reactivated growth and a tendency to recur.

## CASE REPORT

A 33-year-old woman admitted with a ten-month history of soft tissue swelling in the left ankle. On review of symptoms, the patient reported severe pain enough to disturb sleep and to hinder physical activities. She denied any trauma, fever, chills or weight loss. No other acute symptoms were reported by the patient. The physical examination of the left ankle revealed 2 cm × 4 cm painful and firm mass lying anterior to the lateral malleolus (Figure 1). The patient had no warmth, erythema or effusion. The ankle moved normally and her neurovascular status was intact. Laboratory tests revealed normal white blood cell counts 9100/mm and a slight increase of erythrocyte sedimentation rates 33mm/h. Plain x-ray images revealed a loss severe joint space of the talocalcaneal and subtle soft tissue swelling inferior to the lateral malleolus (Figure 2). Ultrasound showed hypoechoic formation with an average abundance of articular effusion (Figure 3). The MRI revealed a well-defined lytic lesion situated in the talus and an intermediate signal mass extending laterally from the subtalar joint (Figure 4). Biopsy was performed and histological examination revealed the presence of multiple histiocytes, some of them containing brown siderin pigment, as well as of numerous multinucleated giant cells these findings are compatible with Pigmented Villonodular Synovitis of the Ankle. A transverse lateral ankle incision centered over the mass was performed. Surgical exploration brought to light the mulit nodular, red, brown and thickened synovium with diffuse haemosiderin staining characteristic of PVNS. The patient underwent tumor wide excision with a talocalcaneal arthrodesis (Figure 5). No adjunct radiation therapy or chemotherapy was given. At his

first out-patient follow-up visit at our clinic he has shown good clinical improvement, the patient remained pain-free. After 3 years of active follow up the patient is free of local or distant recurrence (Figure 6).

**DISCUSSION:**

Pigmented villonodular synovitis (PVNS) is a rare disease characterized by idiopathic proliferation of synovial tissue in the joint, tendon sheath, and bursa [6, 9]. It has been postulated that PVNS may be because of an inflammatory process or a disturbance of lipid metabolism [3, 4]. PVNS of the ankle is much rarer than that of the knee. It was found in the foot and ankle in approximately 2.5% of the cases of Rao et al and 25% of those of Ushijima [16, 14]. Typically, the tumor appears between the third and fifth decades of life, there is no racial or gender preponderance [6, 8].

The symptoms of PVNS are nonspecific and can mimic those of any type of soft-tissue mass. Patients will generally present with pain, joint effusions and swelling. The duration of symptoms is variable (mean duration of symptoms of 27 months) [11]. Laboratory investigations are usually unrewarding as they are almost invariably normal.Plain radiology may show in the localized nodular form of PVNS, a marginal loss of bone density may be found in areas where pressure is applied on bone tissue [9,14]. Resonance imaging is the most valuable modality in identifying PVNS in the ankle, as the soft tissue mass and possible bone infiltration may be monitored.

It displays local nodular PVNS of soft tissue with hypo- and hyperintense nodular portions in T2-weighted images with corresponding enhancement after i.v. administration of gadolinium in T1-weighted images. The presence of a haemosiderin Causes a doserelated decrease in the relaxation time of T1 and T2 images [3, 6]. The extent of bone erosion can be depicted with the same pattern of signal intensity alteration compared to soft tissue [16].

To confirm the diagnosis either a needle or an open biopsy (often excisional) is necessary. Histological examination of the specimen appears as a broad sheet of histiocytes with scattered lymphocytes, lipid and hemosiderin-laden foam cells, and multinucleated giant cells [1,3]. The multinucleated giant cells and the foam cells distinguish PVNS from other arthropathies.

Surgery is the corner stone of treatment. Therapeutical difficulties may arise from high rates of recurrence due to incomplete removal. All PVNS cells need to be removed to prevent a recurrence. Function and stability of the joint involved should be preserved.

The treatment of localized forms of PSVN of the hind foot rests on the widest possible synovectomy, before the installation of the osteocartilaginous lesions. When these are present, the arthrodesis Then becomes necessary in order to ensure the articular indolence and stability [12, 16].

The radiotherapy has proven good results as a postoperative treatment. In current treatment approaches, radiotherapy is recommended as an adjunct to total synovectomy for preventing recurrences in the presence of residual tumor or as second-stage treatment in recurrent cases. The synoviorthesis Isotopic with Yttrium 90 may be indicated in the extensive PVNS form. It was performed under radioscopic control. During the synoviorthesis, a corticosteroid (triamcinolone hexacetonide) was injected with the isotope [8, 11, 13]. Malignant transformation after radiation is a worrisome side effect, especially in young patients.

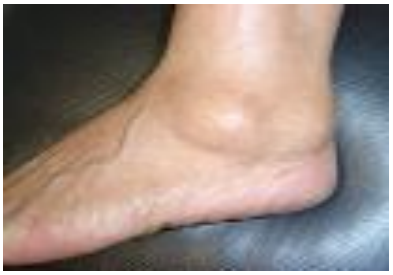

**Fig-1: masse on the left ankle**

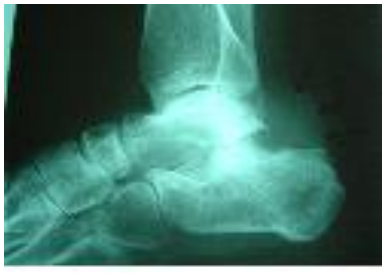

**Fig-2: Lateral X-ray of the ankle showing severe joint space of the talocalcaneal and soft-tissue masse**

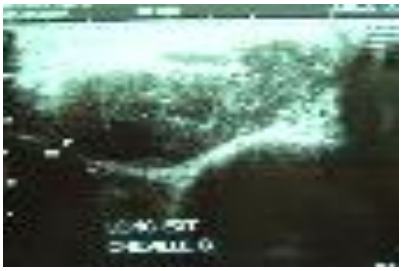

**Fig-3: Ultrasound showing hypoechoic formation with an average abundance of articular effusion.**

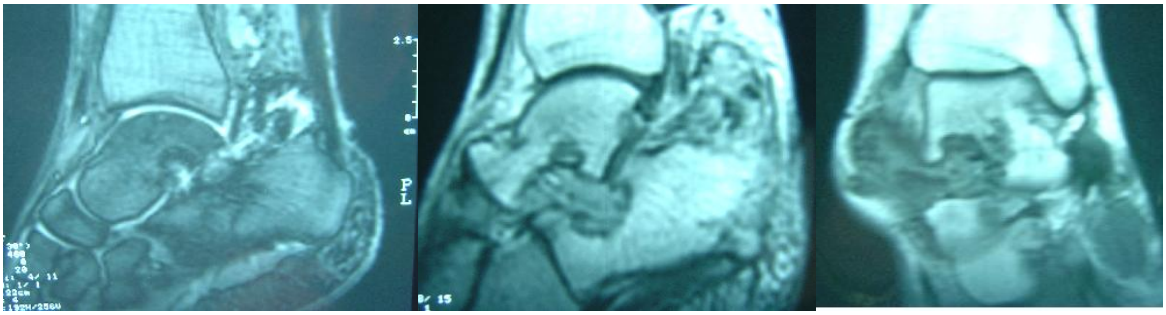

**Fig-4: (A-B) MRI scan (T1-weighted image) showing an inhomogeneous low-signal soft tissue lesion predominantly involving the subtalar joint.**
(C) MRI scan (T2-weighted image) showing a lowsignal lobulated soft tissue lesion affecting the ankle joint

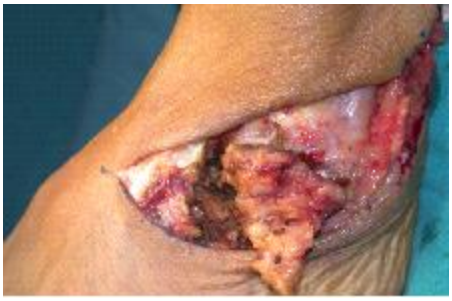

**Fig-5: Intraoperative findings thickened synovial layer with brownish and yellowish villonodullar masses**

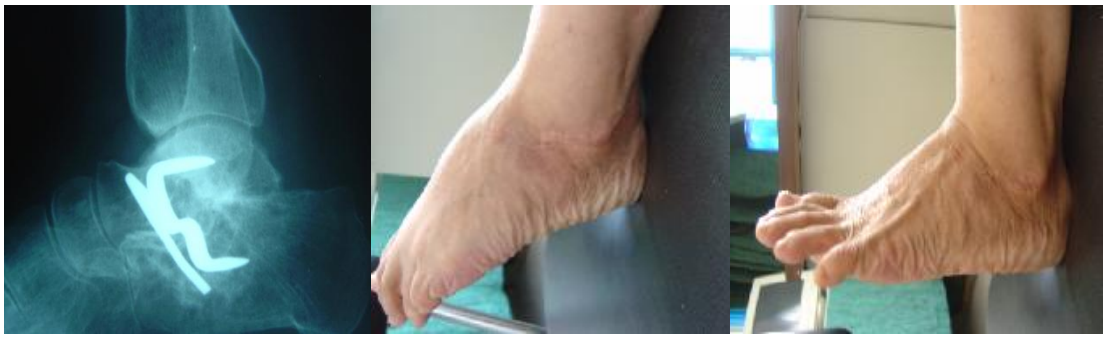

**Fig-6: left ankle After 3 years**

**CONCLUSION:**

PVNS is a rare benign but locally destructive disease with significant potential for severe joint morbidity. MRI is very useful for diagnosis and for planning the extent and the technique of synovectomy. Complete surgical removal of a localized form or total synovectomy is the first line of treatment. The PVNS should be kept in mind in the differential diagnosis of soft tissue and bone tumors.

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