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## Aggressive Presentation of an Innocent Nature Pigmented Villo-Nodular Synovitis - Diffuse Form: A Rare Case Report.

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### Case Report

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#### ABSTRACT

Pigmented villo nodular synovitis is an unusual benign disease of unknown cause producing circumscribed or diffuse thickening of the synovial lining of joints, tendon sheath and bursae. The clinical and pathological distinction between Pigmented villo nodular synovitis and malignant synovioma has in the past often posed difficult problems; there is no doubt that many amputations have been performed for what is now considered to be an entirely benign condition, mistakes may still be made by these who are unfamiliar with the two conditions. The purpose of the present report is to review the literatures on Pigmented Villo nodular synovitis and to present information with a case studied, in which histological material, detailed clinical history and aggressive presentation of benign condition are available.

#### INTRODUCTION

Chassaignac (1852) described lesions of the nodular form arising in relation to the flexor tendon sheaths of the middle and index fingers. Simon described a large pedunculated nodule in the knee. Both considered the lesions to be sarcomata. Moser reported the diffuse type of lesion: the ankle was affected and the patient was free of recurrence seven years after synovectomy. Dowd described a diffuse synovial lesion of the knee, was the first to question the malignant nature of this type of lesion. In the early literature the nomenclature is often confusing the terms Xanthoma, Giant Cell tumor of tendon sheath, Villous arthritis, benign synovioma and myeloxanthoma being used [1]. These names suggested a neoplastic origin, the nodular lesion was inflammatory. Jaffe et al introduced the term PVNS. They observed the nodular and diffuse lesions are histologically similar and suggested that they were part of the same disease process [2].

The benign course, as well as the histological appearance of the lesions, led to conclude the condition is not a tumor, but an inflammatory response to an unknown agent [3].

#### CASE

A 45 years male presented to the Orthopedic OPD with history of swelling and pain in the right knee joint since 8 years. Radiographs showed translucent, soft tissue mass with total destruction of distal femur and proximal tibia.

Fine needle aspiration cytology reported it as PVNS. Because of its locally aggressive presentation above knee amputation was done and sent for histopathological examination. The gross examination showed swelling of the amputated right leg with lower end of thigh. (Fig-1) Cut

section shows grey - brown, fleshy, partially circumscribed tumor mass measuring 12x 11x 10 cms, involving lower end of thigh and upper half of leg (sparing femur, tibia, fibula and joint space). (Fig-2) On microscopy, partly encapsulated, cellular tumor, cells in expansile sheets and lobular pattern. (Fig-3) Sub synovial tissue had nodules of round to oval cells with interspersed multinucleated giant cells. Groups of foamy cells and variable amount of pigment were seen. (Fig-4).



Figure 1



Figure 2

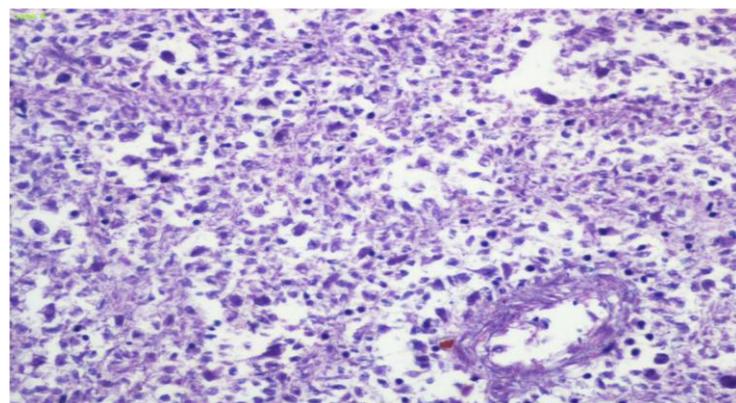


Figure 3

The treatment for PVNS is extensive synovectomy, the diffuse form of PVNS is known for recurrence following surgery. Radiotherapy following surgery causes disabling stiffness of the joint and possible carcinogenic effect [4].

In this case because of extensive involvement and joint destruction arthroplastic procedures are not amenable, amputation appears to be the only alternative “Fierce Facade with an innocent nature”.

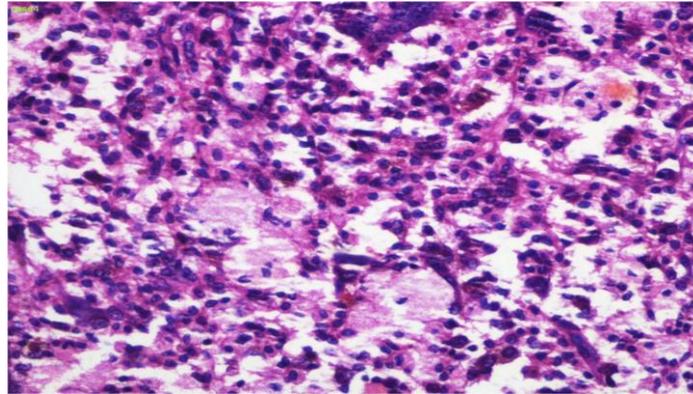


Figure 4

## RESULTS

Clinical evaluation showed no stigmata of systemic or congenital disease. A review of clinical, pathologic and radiologic features of PVNS considered following as differential diagnosis, Synovial Sarcoma - If the lesion is entirely or partly outside of the joint capsule, and presence of scattered irregular calcifications within the mass, when cystic bone changes are found in PVNS, Rheumatoid arthritis which is characterized by polyarticular involvement synovial fluid, biochemical analyses, elevated erythrocyte sedimentation rate and periarticular demineralization. Tuberculous arthritis, Osteoarthritis, Angiomas of osseous origin, Amyloidosis, Fibrous dysplasia, multiple enchondromatosis, Pseudogout [5] and chronic indolent Infectious synovitis may have similar appearance on arthrograms but multiple loose bodies within the joint space, secondary chondromatosis, Lipoma arborescens and Synovial hemangiomas mimick PVNS [6]. Correction of all the clinical aspects and the histologic features is usually required for definitive diagnosis. Tissue specimens can alleviate unnecessary surgical interventions and amputation.

## DISCUSSION

Relatively rare proliferative process found in the synovium and most commonly seen in the knee [7]. It is usually monoarticular and polyarticular involvement is rare [8]. In the knee joint it not only mimics internal derangement, but is also misdiagnosed as malignant lesions prompting needless amputations.[9] The arthroscopic features and histological characteristics are diagnostic. The nodular and diffuse lesions suggest tha they have common histogenesis, the exact pathogenesis remains unclear. The lesions are characterized by proliferation of fibroblastic and histiocytic mesenchymal cells below the synovial lining cells. Foam cells and iron deposits are secondary changes. The localized forms have a relatively high cure rate compared to diffuse forms. The histological features of stromal cells, abundant collagen and hyalinization led Jaffe to conclude the findings closely resemble an inflammatory process [10]. Cytogenetic data has shown various results. X-chromosome inactivation analysis showed that the lesion was polyclonal in origin and suggests PVNS is a reactive proliferation than a true neoplasm.

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