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Pigmented Villonodular Synovitis of the Knee- A Cytologic Impression

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ABSTRACT

Pigmented villonodular synovitis (PVNS) of the knee is a benign but locally aggressive disease of synovial proliferation that occurs in localized nodular and diffuse villous growth patterns. Although inflammatory and neoplastic causes have been hypothesized, etiology remains unknown. PVNS usually presents as unilateral knee pain and swelling and mimics other knee ailments. Radiographs are often unremarkable, whereas magnetic resonance imaging may show characteristic intra-articular masses with signal dropout on T2-weighted sequences. The condition is surgically treated with open or arthroscopic total or partial synovectomy. High recurrence rates are associated with all treatments of diffuse PVNS. Intra-articular radioisotope injection and external beam radiation may be beneficial adjuvant therapy for extensive diffuse and recurrent PVNS of the knee. We report a rare case of pigmented villonodular synovitis in a 56-year-old woman, who presented with a painful mass in the posterior part of the left knee.

Keywords: Knee, Cytology, Pigmented villonodular synovitis.

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INTRODUCTION

Pigmented villonodular synovitis (PVNS) is a benign, hypertrophic synovial process characterized by villous, nodular and villonodular proliferation and hemosiderin pigmentation. PVNS occurs predominantly in the 2nd to 5th decades of life.[1,2] Intra-articular disease has no gender predilection, whereas extra-articular disease is more common in females.[3]

PVNS is a relatively uncommon disease. Previous terminology for PVNS included synovial xanthoma, synovial fibroendothelioma or endothelioma, benign fibrous histiocytoma, xanthomatous giant cell tumor of the tendon sheath, myeloplaxoma, chronic hemorrhagic villous synovitis, giant cell fibrohemangioma, fibrohemosideric sarcoma, sarcoma fusigiganocellulare, benign or malignant polymorphocellular tumor of the synovial membrane, and fibrous xanthoma of the synovial membrane.[3,4] Localized disease, whether extraarticular or intraarticular, represents 77% of cases compared with diffuse intraarticular involvement, which accounts for 23% in a ratio of 3.3:1.[4,5] Localized intraarticular form of disease is more common than extraarticular form and accounts for 6%-7% of all benign soft-tissue masses, whereas the diffuse intra articular form represents only 0.9%. [4–6]

CASE SUMMARY

A 56-year-old woman presented to the Orthopaedic Surgery clinics with a painful mass in the posterior part of the left knee for the last 6 months. Plain radiography revealed a soft tissue mass with slight osteolysis around both the femur and the tibia (Figure 1). MRI demonstrated abnormal masses around the knee joint with bone invasion. Fine needle aspiration cytology of the mass showed plasmacytoid synovial cells and pigment laden macrophages (Figure 2). The lobular mass was excised with curettage and artificial bone grafting. Histopatholgical biopsy showed marked proliferation of synovial tissue with nodular growth, scattered hemosiderin-laden macrophages and osteoclastic giant cells. No focal sarcomatous components or foci of necrosis was identified anywhere in the specimen. These findings suggested a diagnosis of benign pigmented villonodular synovitis (Figure 3).

RESULTS AND DISCUSSION

Pigmented villonodular synovitis is a synovial proliferative process. It is usually monoarticular; however, in the foot and ankle, it can affect more than one joint.[7] Pain with joint swelling and occasionally joint dysfunction are the presenting features.[7,8] A history of trauma has been found in 44%-53% of the patients.[7] It is unusual in the paediatric population and is more frequently poly-articular. It has also been described in association with cherubism, extremity lymphedema, jaw lesions, multiple lentigines syndrome, Noonan syndrome and vascular lesions.[9]

Macroscopically the synovium is diffusely thickened with multiple villous and nodular projections. These are typically dark brown and heterogeneous is colour with areas of yellow discolouration (xanthoma cells). Microscopic examination shows predominantly mononuclear histiocytes mixed with variable numbers of multi-nucleated giant cells in a diffuse infiltrative growth pattern.[2] It is important to note that histologic appearance may mimic aggressive neoplasms such as rhabdomyosarcoma, synovial sarcoma, or epithelioid sarcoma with the surmount role of imaging in guiding the pathologist to a diagnosis.[8,9]

Localized extraarticular disease typically reveals a multinodular, well delineated process embedded in a dense, partially collagenous pseudocapsule with its absence in diffuse intraarticular PVNS with cleft like spaces and discohesive zones.[4,7] All types of PVNS are composed of mononuclear histiocytoid cells with reniform nuclei and plump, eccentric eosinophilic cytoplasm, admixed with multinucleated giant cells and xanthoma cells.[1] The multinucleated giant cells contain a variable number of nuclei, from three or four to 50.[4,7] Hemosiderin deposition is usually apparent in all forms of PVNS, but is a much more prominent feature of diffuse intraarticular disease. In rare cases, chondroid or osteoid metaplasia with associated calcification is seen.[2,3] Mitotic activity may be as high as 10 mitoses per 10 high-power fields.[4,7] Both the mononuclear and multinucleated cells are positive for CD68 and CD163 (histiocytic markers), but the diagnosis is generally made from the hematoxylin and eosin stained tissue sections.[10,11]

The etiology of PVNS may be related to a myriad of conditions: either an inflammatory process, repeated hemorrhage into the joint, occult synovial hemangioma, neoplasia or a disorder of lipid metabolism. However, the capability of these lesions for autonomous growth, the identification of malignant transformation, and the more recent detection of cytogenetic aberrations all strongly support the hypothesis that PVNS has a neoplastic origin.[7,8] Trisomy of chromosomes 5 and 7 has also been reported in diffuse and malignant PVNS.[7,8] The diffuse intraarticular form of PVNS most frequently affects the large joints, with the knee involved in 66%-80% of cases. The hip is the second most commonly affected joint, accounting for 4%-16% of cases.[2,6]

Features on plain radiographic film are relatively non-specific with appearances mainly being those of a joint effusion. Marginal erosions may be present but it is not possible to distinguish PVNS from synovial chondromatosis. On CT Scan, joint effusions commonly co-exist.[8] The hypertrophic synovium appears as a soft brown tissue mass due to haemosiderin deposition. Calcification is very rare in the synovial mass. MRI typically shows mass-like synovial proliferation with lobulated margins. This may be extensive in the diffuse form or limited to a well-defined single nodule in the localized form of the disease.[9]

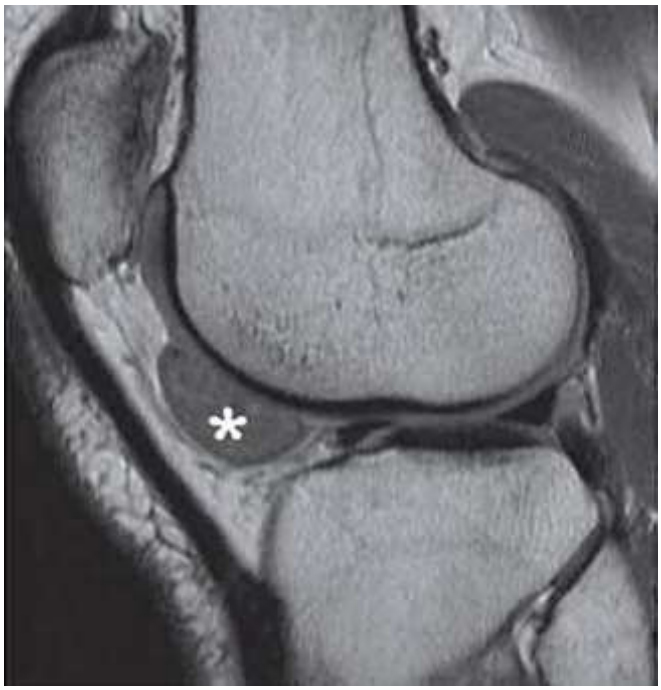

Figure 1: Plain radiography revealed a soft tissue mass with slight osteolysis around both the femur and the tibia.

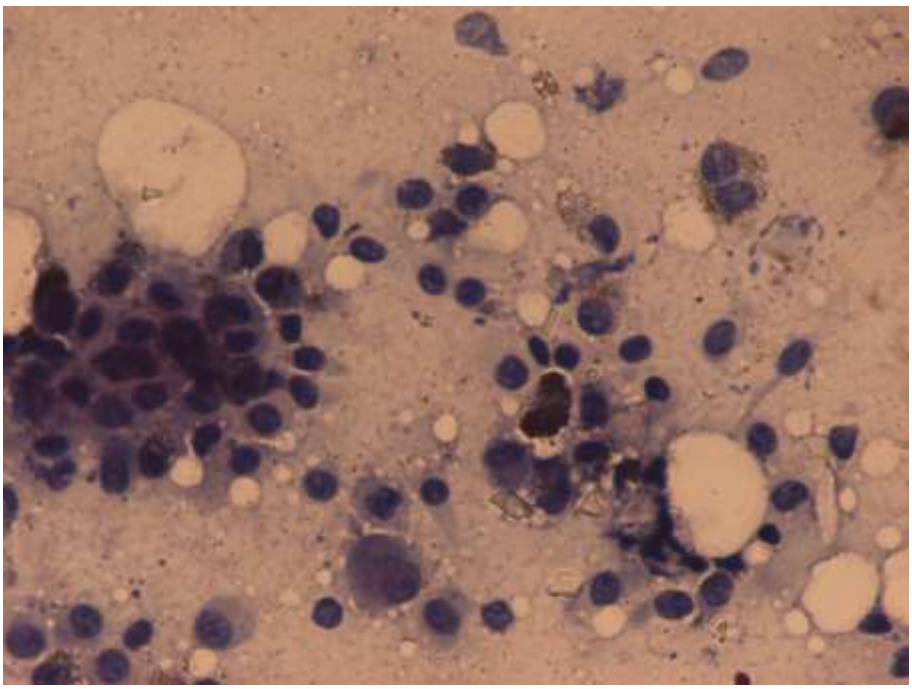

Figure 2: Fine needle aspiration cytology of the mass showed plasmacytoid synovial cells and pigment laden macrophages. Hematoxylin & Eosin x 40X.

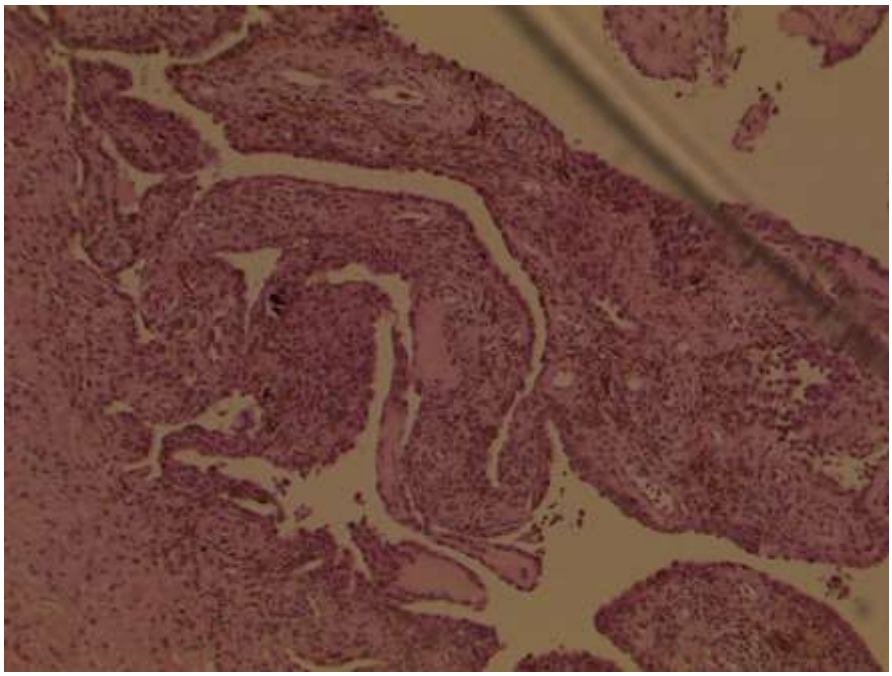

Figure 3: Histopathological biopsy showed marked proliferation of synovial tissue with nodular growth, scattered hemosiderin-laden macrophages and osteoclastic giant cells without any foci of necrosis. Hematoxylin & Eosin x 40X.

Complete synovectomy is the treatment of choice which offers a good prospect of cure. Adjuvant treatment is often employed with external beam radiotherapy and intra-articular injection of Yttrium 90.[11,12] In refractory cases, α-TNF (tumour necrosis factor) blockade and infliximab are advised.[12,13] Recurrence rates after total synovectomy is reported to be 7-20%. [12]

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