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Research Article

IMAGING SPECTRUM OF PIGMENTED VILLONODULAR SYNOVITIS: A CASE SERIES

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ABSTRACT

Pigmented villonodular synovitis is abenign proliferative disease affecting the synovialjoints resulting in villous or nodular changes inthe synovial tissue, large effusions and bonyerosions.PVNS affects young adults in theirthird or fourth decades with an incidence of 1.8per 1 million per year.Pigmented villonodular synovitis (PVNS) is a slowgrowinglesion of uncertain etiology arising from thesynovial membrane. It is characterized by villous andnodular overgrowths of the synovial membrane of thebursa or the tendon sheath. The appendicular skeleton,especially large joints such as the knee and hipjoints are frequently involved.Here, we highlight the spectrum of findings in histopathological proven cases of PVNS, in particular the varied signal intensity characteristics of PVNS at MR imaging.

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INTRODUCTION

Pigmented villonodular synovitis (PVNS) is abenign proliferative disorder primarily occurring in thelarge joints of the appendicular skeleton such as theknee and hip joints.Many other terms have been applied to it, includingnodular tenosynovitis, fibrous histiocytoma of synovium, giant cell synovioma, and fibrous xanthoma ofsynovium (Gezen *et al*., 1996). The term "pigmented villonodular synovitis" was first proposed by Jaffe in 1941 (Gezen *et al*., 1996). PVNSresults in various degrees of villous and/or nodularchanges in these affected structures. Two primaryforms are described, including a diffuse form thataffects the entire synovial lining of the joint, bursa, ortendon and a smaller localized form. The diffuse formtypically involves the large joints such as the knee andhip joints. The localized (focal) form affects the smalljoints of the hand/feet. The focal form is sometimesseen around the tendon sheaths and is then calledgiant cell tumor of tendon sheath (Titelbaum *et al*., 1992). In practice, the term "PVNS" is generally used when the conditionaffects the joints, regardless of whether it isdiffuse on localized.

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PVNS lesions typically contain areasof intermediate and/or low signal intensity on T1-and T2-weighted images (Jelinek *et al*., 1989). The decreased signalintensity becomes more pronounced on the longTR/TE images, because of the preferential shorteningof T2 relaxation times of hemosiderin pigment (Spritzer *et al*., 1987).

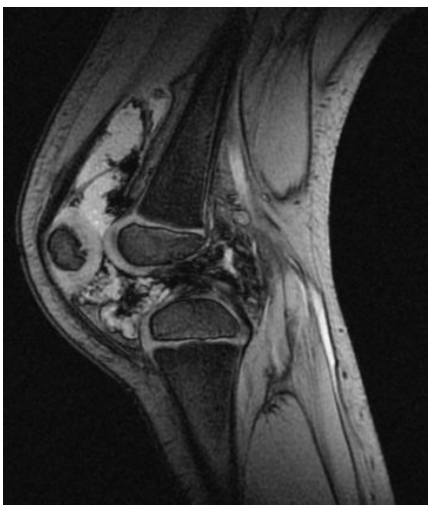

Figure 1: Coronal T2 Weighted Magnetic Resonance image showing hypointense, thickened diffuse synovial proliferation with moderate joint effusion

Case 1: 9 year old boy with complaints of left knee swelling since 1 year presented to orthopedics OPD. On examination, joint line tenderness was elicited with synovial thickening.

Case 2: 22 year old young adult with long standing left knee swelling 6 months with decreased range of movement.

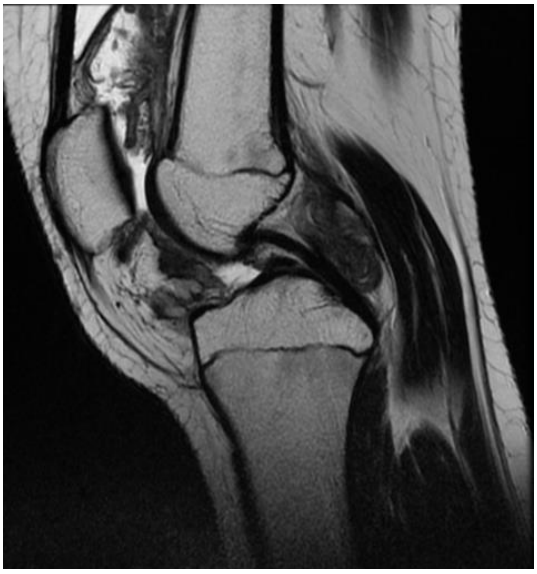

Figure 2. Sagittal Gradient echo Magnetic Resonance image reveals prominent diffuse hypointense synovial thickening with blooming artefact suggesting hemosiderin deposition in the proliferating synovium

Case 3: 3 year old child with chronic history of left knee swelling with recurrent effusions and mild pain without any mechanical symptoms presented to orthopedics OPD. No history of trauma, loss of weight, other joint involvement or bleeding disorders.

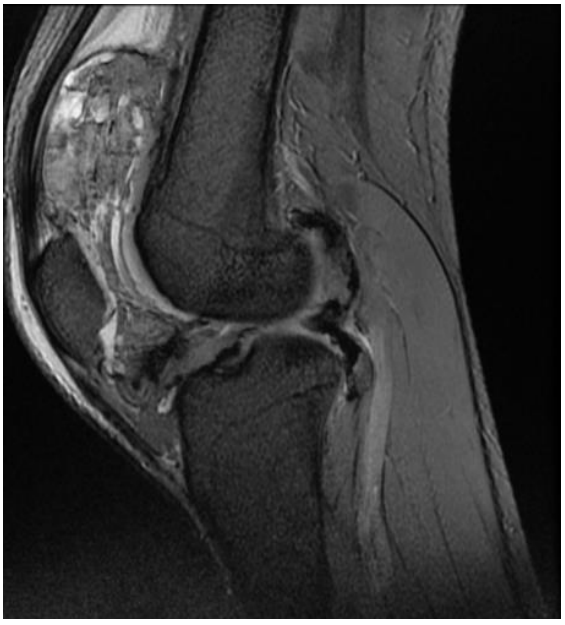

Figure 3: Coronal Gradient recalled echo Magnetic Resonance image shows large amount of low signal intensity tissue, findings that represent the blooming artefact from hemosiderin with posterior extension that replaces the entire knee joint

Case 4: 28 year old young adult with history of right knee swelling since 4 months with inability to perform activities of daily routine.

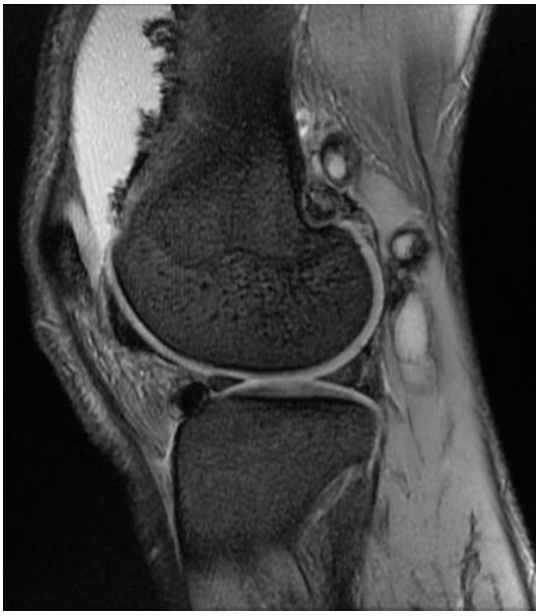

Figure 4: Coronal Gradient recalled echo Magnetic Resonance image showing blooming artefact in the proliferating synovium – suggesting focal nodular pigmented villonodular synovitis

Case 5: 18 year old young adult with history of left knee swelling since 6 months with joint tenderness and synovial hypertrophy on examination.

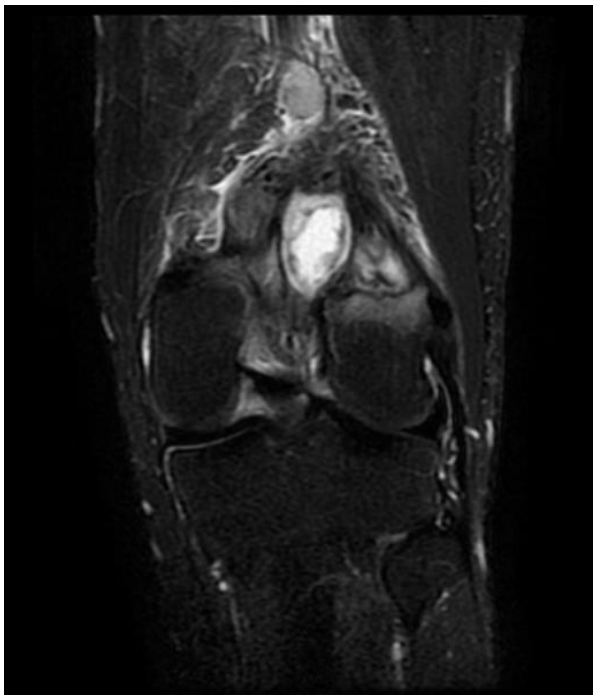

Figure 5: Coronal STIR Magnetic Resonance image showing hyperplastic low intensity synovium due to hemosiderin deposition

Case 6: 32 year old gentleman with history of left knee swelling since 2 years with stiffness and tenderness.

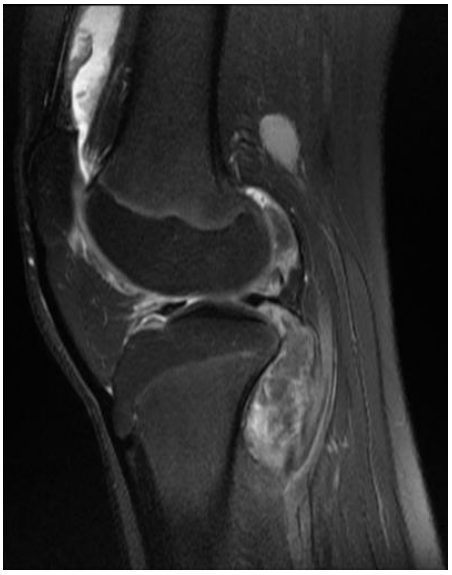

Figure 6: Sagittal T2 Weighted Magnetic Resonance image of the knee with a large effusion and villous proliferation arising from the synovial lining

DISCUSSION

PVNS is a locally aggressive proliferative disordermost commonly affecting large synovium-lined joints.In some instances, the disease may show extraarticularsoft tissue involvement, representing extraarticular extension of a primary intraarticular process.Rarely, the disease may reside completely outside thejoint, in which case the origin maybe from the synoviumof the bursa or the tendon sheath (Spritzer *et al*., 1987; Retrum *et al*., 1987). The exact etiology of PVNS is ill understood. Inflammatoryreaction, neoplasia, hyperplasia, metabolicderangement, and recurrent hemorrhages dueto trauma have all been considered possible etiologies (Gezen *et al*., 1996; Weidner *et al*., 1986; Granowitz *et al*., 1976). PVNS is seen to affect a wide age range of patients, with no definite sex predilection (Giannini *et al*., 1996). PVNS is a monoarticular process and may appear as either a localized or diffuse form within the joint. Patients present with insidious onset of progressive joint swelling and discomfort. Because clinical signs and symptoms are typically non-specific and laboratory tests are unremarkable, imaging plays a key role in the diagnosis and treatment of these patients. Furthermore, PVNS spares joints with hemophilic arthritis and Charcot arthritis (Giannini *et al*., 1996). Recurrent swelling is caused by joint effusion, which is out of proportion to the mild degree of pain. In general, no history of related trauma is reported. Aspiration often reveals xanthochromic or serosanguinous joint fluid. However, the lack of bloody effusion does not exclude the diagnosis of PVNS (Nicholas *et al*., 2016; Nelson *et al*., 2014). It also has a significant tendency for localrecurrence and this has been related to the degree ofresection, lesion cellularity, and mitotic activity (Gezen *et al*., 1996; Granowitz *et al*., 1976). The pathologic findings of spinal PVNS are similarto those seen in the appendicular skeleton. Synovialcells lined villous fronds containing polygonalmononuclear cells, multinucleated giant cells, fibroblasts, and foamy macrophages containing lipid andhemosiderin may be noted. Expansive sheets ofmononuclear cells interrupted by cleft like spaces arealso seen; however, extra-articular lesions usually donot have grossly discernible villous patterns, and hemosiderinstaining is less evident than in their intraarticularcounterparts. The presence and amount ofhemosiderin is related to the extent of mechanicaltrauma that the lesion undergoes.

Routine radiographs and CT scan of PVNS often demonstrate pressure erosion with scleroticmargins of the bone associated with nodular softtissue masses. The matrix of the tumor is rarely calcified.Reactive bone formation is also typically absent.On CT scans, the lesion may appear hyperattenuatedsecondary to the presence of intracellularand extracellular hemosiderin (Titelbaum *et al*., 1992). At MR imaging, PVNS show areas of intermediate to low signal intensityabnormality due to the presence of hemosiderin, which is accentuated on the T2 gradient echo sequences (Jelinek *et al*., 1989; Spritzer *et al*., 1987), but it may show variable appearance, depending on the composition of the lesion and relative proportion of hemosiderin, lipid, fibrous tissue,cyst formation and cellular elements. In the absence of typicalimaging findings of PVNS, image-guided biopsy orexcision of the lesion may be considered. The presence of mononuclear cells, multinucleated giant cells, and hemosiderin deposition, if present, are typicalhistologic features, which indicate the diagnosis ofPVNS.

MR imaging is useful for preoperative, non-invasive diagnosis of PVNS in many cases. MR findings reflect the histologic composition of the tissues comprising the PVNS lesions. The most characteristic finding is nodular intraarticular masses of low signal intensity on T1, T2 and proton-density weighted sequences, the low signal intensity is a result of hemosiderin deposition. The effects of the ferromagnetic properties of hemosiderin are accentuated on T2- weighted sequences, especially gradient recalled echo sequences, because of the differences in magnetic susceptibility between hemosiderin and adjacent tissues. PVNS lesions characteristically show prominent contrast enhancement with the administration of gadolinium.

The treatment of choice for PVNS of all typesis surgical resection, with complete synovectomy being particularly important in cases of diffuse intra articular disease. Recurrence is morefrequent with diffuse intraarticular PVNS, andadjuvant radiation therapy may also be employedfor treatment in these cases. Malignant PVNS israre and difficult to distinguish, both pathologicallyand radiologically, from multiple local recurrencesof benign disease unless there is metastaticinvolvement of the lungs or lymph nodes.Understanding and recognizing the spectrum ofradiologic appearances and their pathologic bases allow improved patient assessment and are importantto optimize clinical management.

Conclusion

Pigmented villonodular synovitis (PVNS) is an uncommon benign proliferative disorder of unknown cause that may involve the synovium of the joint diffusely or focally. The knee followed by hip is the most common location for PVNS. In this case series we reviewed the range of imaging features in histopathologically proven cases of PVNS involving the knee joint.MR imaging findings of prominent low signal intensity

seen with T2 imaging and blooming artefact from hemosiderin are nearly pathognomonic of PVNS.

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