

Pigmented Villonodular Synovitis Presenting as a Popliteal Cyst

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What to Learn from this Article?

Any suspicion of a rare cause of a common symptom should warrant detailed investigation and thorough management.

Abstract

Introduction: Pigmented villonodular synovitis is an uncommon disease characterized by hyperplastic synovium, large effusions and bone erosions. It commonly mimics other conditions. As a result, the diagnosis may remain elusive, as the classic signs of tenderness and effusion are not specific. Occasionally, PVNS presents as a popliteal cyst, which may divert attention from intra-articular pathology. In this article, we describe a case of pigmented villonodular synovitis that presented with a popliteal cyst as the chief problem.

Case Report: A twelve year old boy presented to us with a history of a gradually enlarging mass in the left popliteal fossa since eighteen months. He complained of restriction of terminal knee flexion, but he had no pain. After clinical examination and imaging the mass was interpreted as being either a popliteal cyst or chronic hematoma without ruling out the possibility of soft tissue sarcoma. An open excisional biopsy of the lesion was performed which revealed a reddish-brown cystic-nodular mass communicating with the knee joint. . The histology was consistent with that of pigmented villonodular synovitis. We performed an arthroscopic synovectomy 3 weeks later. Abnormal synovium found in all compartments of the knee was removed. Histology from the intra-articular lesion was also consistent with a diagnosis of pigmented villonodular synovitis.

Conclusion: A swelling in the popliteal region may be due to various etiologies. The cause should be thoroughly investigated. Along with clinical examination and imaging the tissue must be subjected to histopathological examination. Pigmented villonodular synovitis can rarely present as a popliteal cyst. In such a situation, treatment consists of dealing with intra-articular and extra-articular pathology.

Keywords: Pigmented villonodular synovitis; Popliteal cyst; Arthroscopic

Introduction

Pigmented villonodular synovitis is an uncommon disease characterized by hyperplastic synovium, large effusions and bone erosions. The disease was first described by Jaffe et al. as a distinct entity in 1941. PVNS has an incidence of 1.8 cases per 1,000,000 people and has not been strongly associated with a specific risk factor, such as occupation, environment, ethnicity, or sex. [1,2] PVNS typically presents between the second and fourth decades of life, though the range is 11 years to 82 years.[3,4] Joint most

commonly is the knee (up to 80%).[5] PVNS can present in 2 forms: local and diffuse. 20% of cases are local and 80% are diffuse.[3] The local form presents as a pedunculated or sessile lesion, and has a low rate of recurrence, whereas the diffuse form presents as a rust-colored villonodular hyperproliferation extensively involving the synovial lining, and has a higher rate of recurrence.[1,2] The extra-articular spread of pigmented villonodular synovitis (PVNS) has been reported scarcely in the literature and treatment appears to be controversial. Occasionally, PVNS presents as a

Author's Photo Gallery



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Figure 1: Swelling in the popliteal region. Swelling in the left popliteal region 10cms in diameter and about 3 cms raised from the surrounding tissues occupying the entire popliteal fossa. Lentigenes on the lateral side of the knee.

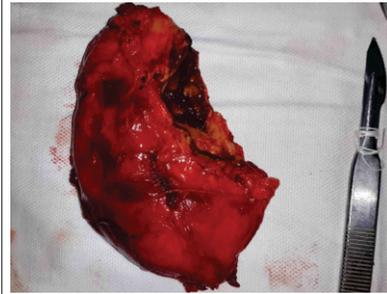


Figure 2: Specimen after excision. Reddish-brown cystic-nodular mass measuring 12x5x8 cms, obtained after the first surgery.

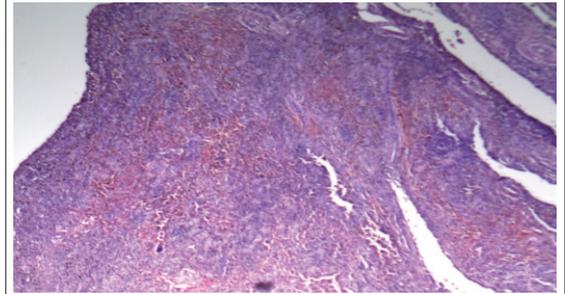


Figure 3: Histology of the lesion at 10x. Histology of the lesion showing papillary projections lined by hyperplastic synovial cells infiltrated by lymphocytes and few plasma cells with hemosiderin consistent with a histology of pigmented villonodular synovitis. Histology specimen at 100x in the left upper corner.

popliteal cyst, which may divert attention from intra-articular pathology. In this article, we describe a case of PVNS that presented with a popliteal cyst as the chief complaint..

Case report

A twelve year old boy presented to us with a history of a gradually enlarging mass in the left popliteal fossa since eighteen months. He complained of restriction of terminal knee flexion, but he had no pain. On general examination our patient had cutaneous lentigenes over the back of the neck and on the skin over the back of left knee. Local examination revealed a palpable mass around 10cms in diameter and about 3 cms raised from the surrounding tissues occupying the entire popliteal fossa. The mass was not warm or tender and was not associated with any skin changes. It was non-pulsatile. The flexion of the left knee was terminally restricted as compared to the right knee.

The anteroposterior and lateral radiographs of the knee were normal. Magnetic resonance imaging revealed a well defined lobulated STIR/ T2 hyperintense lesion measuring 12.8 x 4.8 x 8.8 cms with multiple thick septae within it. Superiorly, the lesion was lying in the soft tissue between semimembranosus, gracilis and sartorius muscles posteriorly and gastrocnemius muscle anteriorly. Inferiorly it was lying posterior to gastrocnemius muscle. The lesion was lying posterior to neurovascular structures in the popliteal fossa. There was minimal effusion within the joint which was thought to be sympathetic in nature. The mass was interpreted as being either a complicated popliteal cyst or chronic hematoma without ruling out the possibility of soft tissue sarcoma.

We decided to perform an open excisional biopsy of the lesion which revealed a reddish-brown cystic-nodular mass. The mass was seen to communicate with the knee joint. The cyst was removed and sent for histopathological examination. The communication with the knee joint space was sealed with purse-string sutures. The histology revealed papillary projections lined by hyperplastic synovial cells infiltrated by lymphocytes and few plasma cells with hemosiderin deposits and was consistent with a histology of pigmented villonodular synovitis.

As we suspected that the disease had originated in the knee and spread to the popliteal space, the first surgery (posterior only) could not have removed the intra-

articular origin of the lesion. So we decided to do arthroscopic synovectomy.

We performed an arthroscopic synovectomy 3 weeks later. Brownish-red hypertrophied synovium with papillary projections was found in all compartments of the knee was removed and sent for histological examination. The histology from the intra-articular lesion was also consistent with a diagnosis of pigmented villonodular synovitis. At 6 months follow-up, there is no relapse.

Discussion

Popliteal cysts are believed to represent an enlargement of the gastrocnemio-semimembranosus bursa.[6] It has been shown that this bursa communicates with the joint space of the knee in 50% of normal adult subjects. Studies of the pathogenesis of popliteal cysts have shown that they are dependant on a valvular mechanism. The presence of a valve, along with the existence of an effusion, allows a unidirectional flow of the synovial fluid from the articular cavity to the bursa, determining the appearance and persistence of the cyst.[6,7] In our case, the effusion was caused by an altered synovial membrane, typical of PVNS. The histologic examination of the cyst removed during the first surgery showed the presence of typical alterations compatible with PVNS, indicating a spread of the pathology into the popliteal cyst. Our patient also had cutaneous lentigenes over the back of the neck and on the skin over the back of left knee. There have been reports on association of cutaneous lentigenes with pigmented villonodular synovitis.

As PVNS can mimic many other condition, there is generally a delay in the

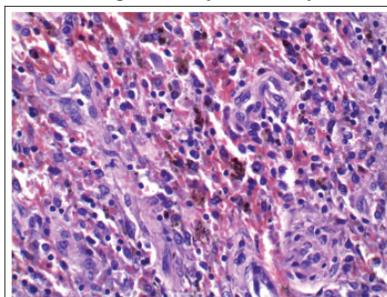


Figure 4: Histology specimen at 100x



Figure 5: Arthroscopic appearance of the intra-articular lesion. Brownish-red hypertrophied synovium with papillary projections was found in all compartments of the knee

diagnosis of this condition. A review of 2 studies revealed that 40% to 54% of PVNS cases were not diagnosed correctly before surgery.[4,8] Of patients with PVNS of the knee, 96% present with a large effusion and distention of the suprapatellar pouch, 40% have a palpable mass, and nearly all have limited flexion and extension.[4]

Radiographic examination of the knee may be normal (54% of cases) or may show a non specific soft tissue swelling.[9] The radiograph may show bony erosions. Bony erosions vary according to joint location and ultimately joint volume capacity. Compared with the hip, the knee, by way of its capacious capsule, has a much lower incidence of bony changes. The pressure exerted by the space-occupying lesion is postulated to apply compressive forces to the articular surface and to induce atrophy and erosive changes.[9,10]

MRI is the current imaging technique of choice. Although the MR findings in PVNS are not pathognomonic, they are highly suggestive of the diagnosis. The MRI features of PVNS depend on the fat, fibrous tissue and iron present. The presence of haemosiderin within tissue causes shortening of both T1 and T2 relaxation times. Haemosiderin has magnetic susceptibility properties. This is manifested as low signal "blooming effect" best appreciated on gradient echo sequences. Areas of high signal on T1 sequences represent either lipid laden macrophages or haemorrhage. Areas of bright signal on T2 weighted images may be present within the abnormal synovial membrane and are believed to represent loculated areas of joint fluid trapped within the synovial membrane. The lesions of PVNS show intense enhancement after administration of gadolinium [11].

The management of PVNS of the knee with extra articular involvement includes open excision of the extra articular mass

and arthroscopic synovectomy. This can be done in 2 stages i.e. open excision followed by arthroscopic synovectomy after 6 weeks allowing the posterior capsular defect to heal. [12] Or both these procedures can be carried out in a single sitting, as there is a postulated risk of spread of the intra-articular lesion extra-articularly in the interim increasing the probability of recurrence. [13] If the lesion lies anterior to neurovascular structures in the popliteal fossa, one can attempt to do both the procedures of synovectomy and decompression of the cyst arthroscopically.

Radiotherapy can be considered in patients with local relapse and in patients with a large amount of disease in whom complete resection is not possible.[14] It has been used in the management of recurrences with varying success. However it is generally avoided in children because of growth issues and the concern about radiation associated malignancy later in life.

TNF- blockade with infliximab has been reported as an effective therapy for refractory pigmented villonodular synovitis.

Conclusion

A swelling in the popliteal region may be due to various etiologies. The cause should be thoroughly investigated. Along with clinical examination and imaging the tissue must be subjected to histopathological examination. Pigmented villonodular synovitis can rarely present as a popliteal cyst. In such a situation, treatment consists of dealing with intra-articular and extra-articular pathology.

Clinical Message

A rare manifestation of a rare disease may present as a common symptom. Thorough investigation and comprehensive treatment is the key to successful management.

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