Pigmented Villonodular Synovitis Presenting as a Baker Cyst

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Abstract

Pigmented villonodular synovitis (PVNS), a rare hyperproliferation of the synovial lining, 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 PVNS that presented with a popliteal cyst as the chief problem report. In other cases PVNS presented as a popliteal cyst, and evidence was reported of PVNS within the cyst stalk and presumable intra-articular involvement. Our case did not present this way.

We recommend that the diagnosis of PVNS be considered in young adults who present with a popliteal cyst.

First described by Chassaignac[1] in 1852 and then by Jaffe and colleagues[2] in 1941, pigmented villonodular synovitis (PVNS) has remained a diagnostic challenge as a consequence of its variety of presentations and similarities with other pathologies. Granowitz and colleagues[3] recognized 2 forms of the disease: local and diffuse. Twenty percent of cases are local and 80% are diffuse.[4] 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.[4,5]

PVNS is an exceedingly rare condition (1.8 cases per 1,000,000 people) that has not been strongly associated with a specific risk factor, such as occupation, environment, ethnicity, or sex.[4,5] However, age correlates with the disease. PVNS typically presents between the second and fourth decades of life, though the range is 11 years to 82 years.[6,7] The highest incidence of PVNS (up to 80%) involves the knee joint.[8]

Presentations of PVNS may vary, but pain and swelling are common patient reports. Cases of PVNS have been reported to present as a Baker cyst, but biopsy results revealed the histologic features of PVNS within the cyst and presumably with intra-articular involvement.[9-13]

In this article, we describe a case of diffuse PVNS that initially did not present with the histologic features of PVNS within the cyst stalk. The diagnosis was confirmed only after arthroscopic biopsy of the synovium. The patient provided written informed consent for print and electronic publication of this case report.

Case Report

A 25-year-old woman presented with an approximate 6-month history of posterior knee fullness and generalized knee pain. She had seen her local orthopedist, who ordered radiographs, which were normal, and magnetic resonance imaging (MRI), which suggested a ruptured Baker cyst. Aspiration of the joint yielded 90 mL of blood-tinged fluid. The orthopedist performed an arthroscopic evaluation for presumed "synovitis," and noted a "large amount of synovial tissue" with a hemorrhagic effusion. The synovial tissue was cauterized with an arthroscopic thermal device. The patient was then turned to the prone position, and the orthopedist excised the presumed "Baker cyst" through a curved posterior, but laterally based incision. The patient said

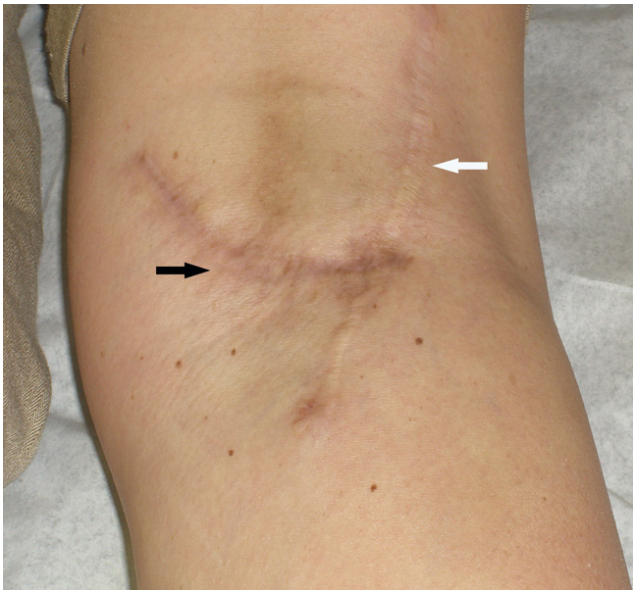

Figure 1. Healed incision sites on right knee show initial, laterally based incision (white arrow) and subsequent, medially based incision (black arrow), which afforded complete excision of cyst.

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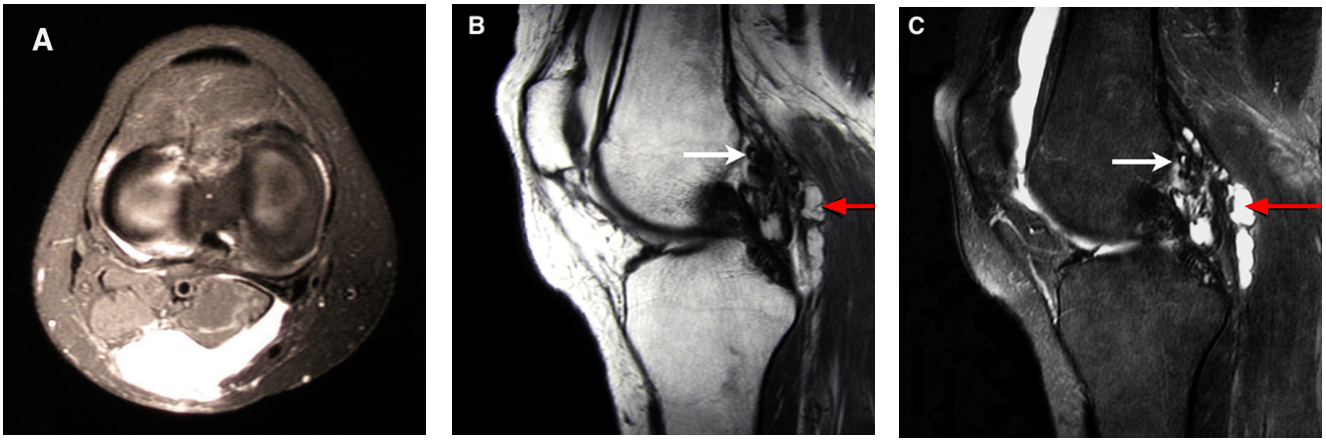

Figure 2. Preoperative magnetic resonance imaging. (A) Axial T_2 image shows posterior fluid collection communicating with posterior medial joint. Sagittal T_1 (B) and sagittal T_2 (C) images of intercondylar notch show Baker cyst posteriorly (red arrow) and synovitis with small foci of hypointense signal abnormality consistent with hemosiderin deposition (white arrow).

that, after surgery, the posterior knee fullness returned rapidly and became limiting within weeks. During the interim, she had multiple ultrasound-guided aspirations of the cyst with steroid injections, which offered no relief. After being referred to a rheumatologist and another orthopedist, she sought consultation at our institution.

On our initial examination of the patient, we found a minor knee effusion and a large popliteal cyst. There was a healed posterior lateral incision on the posterior fossa. As the patient already had undergone arthroscopy, and the posterior mass was her chief problem report, a repeat open cyst excision was scheduled.

At surgery, a posterior medial incision (Figure 1) was made, and dissection was carried between the semimembranosus and the medial head of the gastrocnemius. The cyst was traced down to the capsule, which was partially excised. Pathology revealed "fragments of tenosynovial-type soft tissue showing histiocytic reaction with focal hemosiderin deposition." There was no mention of PVNS in the pathology report, though a persistent knee effusion prompted us to perform a knee aspiration, which revealed blood-tinged fluid. T_1- and T_2 -weighted MRI showed diffuse areas of synovitis with diminished signal intensity, consistent with PVNS (Figure 2). As the patient was experiencing recurrent effusions, a sub–total synovectomy was performed. Using anterior, posterior, and "trans-septal" posterior portals, we performed an essentially complete excision of the red, inflamed synovium (Figure 3). A shaver was used for excision while a thermal wand was used to help "coagulate" the bloody tissue. Specimen findings were consistent with PVNS (Figure 4).

Convalescence was uneventful, and the patient was last seen approximately 12 weeks after surgery. Findings were no significant effusion, essentially full range of motion, and no recurrence of the popliteal mass.

Discussion

PVNS can imitate many other conditions, both on physical examination and imaging studies. As a result, mean time between presentation and diagnosis is 4.4 years.[14] A review of 2 studies revealed that 40% to 54% of PVNS cases were not diagnosed correctly before surgery.[7,15] Some of the most common misdiagnoses are extensor mechanism malalignment, meniscal lesion, inflammatory arthritis, and popliteal cyst.[7]

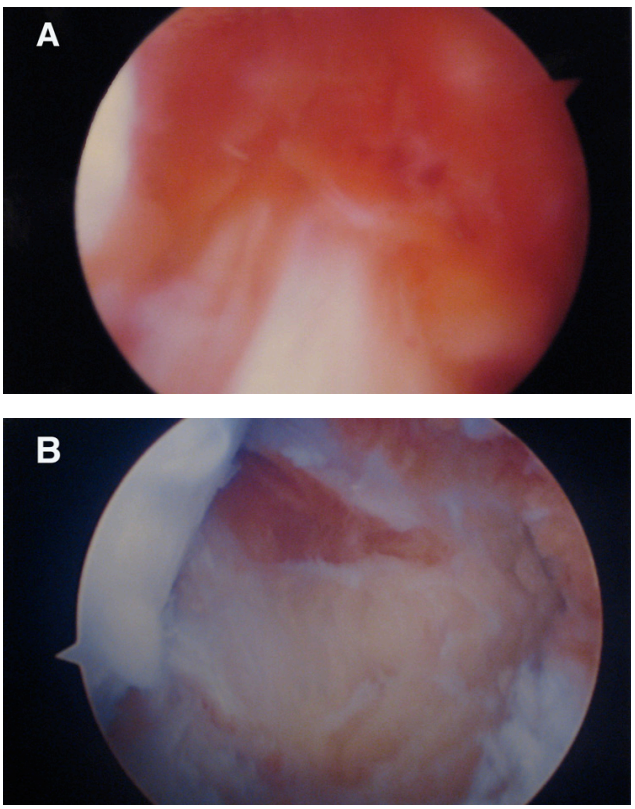

Figure 3. Arthroscopy images show (A) diffuse involvement of synovium before resection and (B) status after synovectomy.

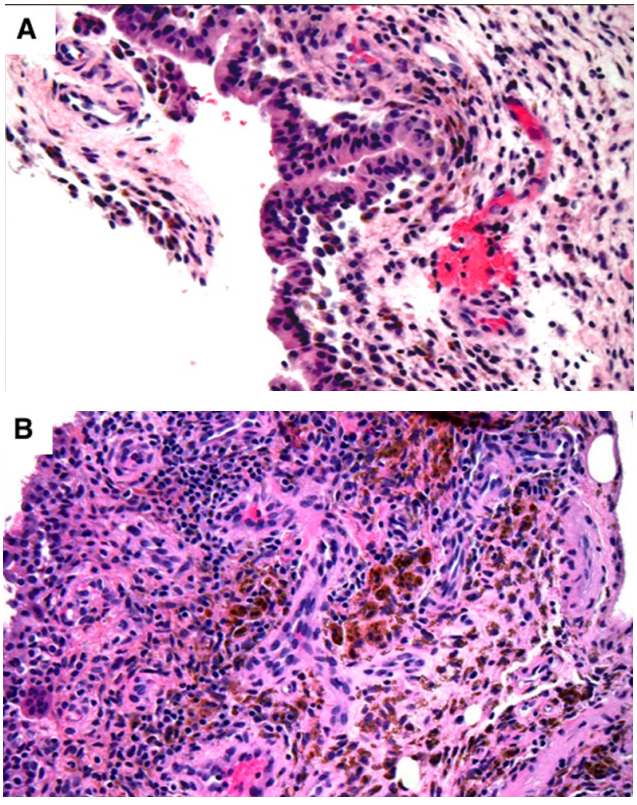

Figure 4. Histologic sections. (A) Baker cyst shows no evidence of pigmented villonodular synovitis. (B) Synovium shows classic findings of pigmented villonodular synovitis: hemosiderin deposits, histiocytes, and giant cells.

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.[7] There has been some debate about the reliability of the finding of a bloody aspirate on arthrocentesis, as the incidence of this finding has ranged from 44% to 69%.[4,7] Our patient presented with all these diagnostic features.

Plain radiographs of a knee with PVNS may be normal in 54% of cases, and nonspecific soft-tissue swelling is the most common finding.[16] Our patient's radiographs did not show any bony changes, though soft-tissue fullness was appreciated. Although reports of bony erosions with PVNS are common, 1 investigator estimated incidence within the knee to be as low as 26% to 32%.[17] 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.[16,18] In the knee, this usually occurs only when the disease is in its late stages, as the knee capsule allows for decompression into adjacent spaces, such as the gastrocnemius/semimembranosus bursa. In our patient's case, the initial pathology of popliteal cyst likely arose from this mechanism.

MRI is the superior tool for synovial imaging. The classic histologic hemosiderin deposition of PVNS shows as low T_1 and T_2 signal intensity.[16,18,19] One investigator found these features in all pathologically confirmed PVNS cases, but it should be noted that hemosiderin deposition is not specific to PVNS, and synovial proliferation may vary.[16] Regardless, MRI is of significant use in the diagnosis of PVNS, and, in our patient's case, it provided valuable evidence.

For many surgeons, arthroscopic synovectomy is the preferred treatment for PVNS, as recovery is faster and there are fewer functional complications when compared with open arthrotomy.[20,21] Newer techniques for refractory cases include augmenting surgery with radiation or infliximab (tumor necrosis factor α inhibitor).[18,22] No studies have examined the usefulness of thermal energy in inducing remission.

As the diagnosis of PVNS may be mistaken for an assortment of other conditions, we recommend that this diagnosis be considered when younger adults present with a popliteal cyst, regardless of cyst stalk pathology. The absence of chondrosis or meniscal pathology—2 other known risk factors for popliteal cyst formation[23]—in the presence of a popliteal cyst should raise suspicion for PVNS.

As most popliteal cysts originate near the semimembranosus muscle insertion, open excision should include a medially based incision so that the cyst/capsular communication may be excised.

Authors' Disclosure Statement

The authors report no actual or potential conflict of interest in relation to this article.

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This paper will be judged for the Resident Writer's Award.