Bilateral pigmented villonodular synovitis of the knee

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Abstract

Pigmented villonodular synovitis is a disorder resulting in a villous, nodular, or villonodular proliferation of the synovium, with pigmentation related to the presence of hemosiderin. These lesions are almost exclusively benign with rare reports of malignancy. Pigmented villonodular synovitis can occur in a variety of joints and at any age but most often occurs within the knee in the young adult. Pigmented villonodular synovitis is a rare disease entity, and bilateral synchronous or metachronous involvement of a joint is even more uncommon, with few reports previously described in the literature. We present a case of pigmented villonodular synovitis involving both the right and left knee in the same patient, with radiographic imaging, magnetic resonance imaging, photograph and video intraoperative imaging, and pathologic correlation.

Case report

A 62-year-old man, with a medical history of HLA-B27-positive ankylosing spondylitis and asthma, initially presented to his primary care provider in July 2001 with left knee pain and a sensation that something was “popping out” of the anteromedial aspect of the joint. There was no instability of the knee on physical examination. Radiographs of the left knee were obtained, which demonstrated mild patellofemoral degenerative osteophyte formation with a small knee joint effusion (Fig. 1). He was diagnosed with patellofemoral osteoarthritis and subsequently made repeated visits to his primary care provider and physical therapist over the next several years because of persistent, nonremitting pain.

In September 2009, because of worsening pain, as well as new symptoms of locking and a palpable, mobile lump along the anteromedial joint line, a magnetic resonance imaging (MRI) scan was obtained. MRI revealed a joint effusion, patellofemoral osteoarthritis, and a multilobulated lesion in continuity with the synovium in the anteromedial patellofemoral joint space. The lesion exhibited homogeneous enhancement and “blooming” on gradient echo images (Fig. 1) and was suspicious for pigmented villonodular synovitis (PVNS). He was referred to the orthopedic surgery service at our institution, where he underwent excision of the intra-articular lesion in January 2010. Pathologic examination (Fig. 2) confirmed the diagnosis of PVNS. After postsurgical rehabilitation, he has not experienced recurrent left knee pain.
A 62-year-old man with bilateral pigmented villonodular synovitis of the knee. Frontal (A) and lateral (B) radiographs of the left knee demonstrate minimal patellofemoral osteophyte formation and small joint effusion. Sagittal T2 fat-saturated (C) and sagittal (D) and axial (E) postcontrast T1-weighted magnetic resonance images of the left knee reveal a lobulated, enhancing mass (arrows) in the anteromedial knee, which on excision was consistent with pigmented villonodular synovitis.

Fig. 2 – A 62-year-old man with bilateral pigmented villonodular synovitis of the knee. (A) Low- and (B) high-power photomicrographs demonstrate a sheet-like growth of mononuclear cells, primarily histiocyte-like, admixed with multinucleated giant cells, some of which demonstrate a peripheral rim of hemosiderin, consistent with a diffuse-type tenosynovial giant cell tumor.
In October 2010, he reported a painless lump above the right knee to his primary care provider, which was diagnosed clinically as a ganglion cyst. In April 2012, he developed discomfort at the site of the lesion, and radiographs as well as MRI scans were obtained in an effort to further characterize (Fig. 3). The MRI demonstrated intra-articular lesions similar to those detected within the contralateral knee and most consistent with PVNS. The patient opted to defer surgical intervention to avoid postoperative rehabilitation.

Years later, the patient returned because of increasing discomfort associated with the suspected PVNS lesion within his right knee, at which time he was referred to the orthopedic surgery service at our institution for definitive management. At this time, in November 2014, a follow-up MRI was obtained. This demonstrated multiple intra-articular lesions consistent with PVNS, similar in appearance to those on the prior MRI scan acquired in April 2012, along with multiple sites of cartilage loss in the medial and patellofemoral compartment. In January 2015, he underwent arthroscopic removal (Fig. 4) of the lesions along with synovectomy and chondroplasty. Pathology was consistent with PVNS (Fig. 5). He expressed resolution of his symptoms in the early postoperative period.

Discussion

PVNS was initially described in the tendon sheath by Chassaignac in 1852, with Simon later describing a nodular, intra-articular form of the disease in 1865 [1,2]. As such, PVNS and giant cell tumor of the tendon sheath are terms often used interchangeably to describe these mostly benign proliferations of the synovium affecting joints, tendon sheaths, and bursae. Moser elucidated a diffuse form of the disease in 1909 [3]. In 1941, Jaffe consolidated the various forms of PVNS and giant cell tumors of the tendon sheath, subclassifying the disease based on diffuse or focal or nodular presentation and intra-articular versus extra-articular location [4–6].

Fig. 3 – A 62-year-old man with bilateral pigmented villonodular synovitis (PVNS) of the knee. Frontal (A) and lateral (B) radiographs of the right knee demonstrate a small joint effusion. Axial (C) and sagittal (D) T2 fat-saturated and sagittal gradient echo (E) images of the right knee demonstrate multiple hypointense nodules (arrows) with focal areas of internal low signal intensity on the gradient echo sequence consistent with susceptibility artifact related to hemosiderin deposition in these areas of intra-articular PVNS.
The World Health Organization uses a separate classification system based on prognostic implications. The tumors are divided into 2 categories dependent on location and behavior. In specific terms, the diffuse-type giant cell tumor is defined as a “destructive proliferation of synovial-like mononuclear cells admixed with multinucleate giant cells, foam cells, siderophages, and inflammatory cells” [7]. This form typically involves a joint, but can involve extra-articular soft tissues, and is characterized by an infiltrative, sheet-like growth pattern. The localized type, which can be seen in the tendon sheath, bursa, or joint space, demonstrates the same cell types as the diffuse type but is typically smaller, more well circumscribed, and less destructive than its diffuse-type counterpart. Although both forms of the disease can recur after surgical excision, rates are higher in the diffuse type [6,7].

Intra-articular PVNS is an uncommon disease with an incidence of 1.8 cases per 1 million, typically presenting in the fourth decade without a sex predilection. With respect to the intra-articular form of the disease, the knee is the most commonly involved joint, and the hip, elbow, and shoulder are the next most common sites [5,6,8]. Absence of a palpable lesion, which is less common with the diffuse intra-articular form of disease, and the insidious onset of pain, swelling, and motion restriction are the typical presenting symptoms. Because of these relatively nonspecific symptoms, diagnosis often requires months or years to establish [6,9].

Initial radiographs can be normal or reveal a joint effusion, soft-tissue mass, and/or bony erosion. These extrinsic erosions are well defined, often with rim sclerosis, and occur on both sides of the joint. Erosions are likely due to contact or mass effect on the adjacent bone and as such, more commonly present in smaller, lower-capacity joints. Bone mineralization and joint spaces are typically preserved [5,6]. Similar findings are seen with computed tomography (CT). CT may also reveal synovial thickening, often exhibiting increased attenuation owing to the presence of hemosiderin. The intra-articular lesions are unlikely to be calcified, allowing for differentiation from post-traumatic or degenerative intra-articular bodies or synovial osteochondromatosis [6].

Fig. 4 – A 62-year-old man with bilateral pigmented villonodular synovitis of the knee. Intraoperative photographs during arthroscopy taken before (A) and after (B) resection of intra-articular mass (arrow). The supplementary video shows the arthroscopic resection of the intra-articular mass from the right knee.

Fig. 5 – A 62-year-old man with bilateral pigmented villonodular synovitis of the knee. (A) Low- and (B) high-power photomicrographs demonstrate a sheet-like distribution of mononuclear and multinucleated giant cells with peripheral hemosiderin deposition and collagenized stroma consistent with diffuse-type tenosynovial giant cell tumor.
MRI, with or without arthrography, demonstrates the aforementioned findings of erosions, joint effusion, synovial thickening, and soft-tissue masses in greater frequency and detail than radiographs or CT. Diffuse-type PVNS appears as a multinodular synovial thickening of low or intermediate signal intensity on all pulse sequences. “Blooming” or accentuation of signal reduction on gradient echo sequences due to magnetic susceptibility artifact from hemosiderin is typical of PVNS [5,6,8]. Localized-type or focal intra-articular PVNS manifests as a well-defined soft-tissue mass with low to intermediate signal intensity on T1-weighted images and heterogeneous appearance on T2-weighted images. Blooming on gradient echo images is commonly present but is less extensive than in the diffuse form [6]. If intravenous gadolinium-based contrast is administered, enhancement of the lesions is common, more so in the diffuse than the localized form, and likely because of the hypervascularity of the lesions. The hypervascular nature of the lesions is also evident with angiography or the blood flow and blood pool images of 3-phase Tc-99m nuclear scintigraphy [6].

Polyarticular or multifocal disease, as in our case, is rare and has been most frequently described in children, an age group that otherwise rarely develops PVNS. There are only scant previous reports of multifocal PVNS in adults. Many of these reports predate the advent or widespread availability of MRI. Of these cases, there are examples of bilateral disease at presentation, as well as of macronchondral lesions, as in our case [8–16].

Surgical excision is the preferred management for both localized and diffuse giant cell tumor of the joint, with success being dependent on complete resection with clear margins. In the setting of extensive diffuse disease, where complete resection is unlikely, surgery is performed to debulk before radiation or medical therapy, as well as to provide symptomatic relief. Radiation therapy, both external beam and radiosynoviorthesis, can be used in diffuse intra-articular disease, preferably in conjunction with surgical management. Medical therapy consists of immunomodulating agents such as the tumor necrosis factor inhibitor infliximab [6,8,9]. Widely varying recurrence rates have been reported both for localized (0%-44%) and diffuse (8%-56%) disease, with increased recurrence rates in cases in which complete synovectomy could not be obtained, resulting in residual disease at the resection margin. Positive outcomes with surgery for localized disease and with surgery and adjunct therapies have been described, highlighting the value of making the diagnosis to allow for optimal and timely management [5,6,8,9].

Although PVNS is a rare disease entity, and bilateral disease even more uncommon and only sparsely described in the literature, it remains an important and treatable consideration in a patient with chronic, progressive joint pain. Our case of PVNS involving both the right and left knee in the same patient demonstrates the radiographic and MRI appearance of the disease, with photograph and video intra-operative imaging and pathologic correlation.