Pigmented Villonodular Synovitis in a 63-year-old Adult: A Case Report

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Abstract

Pigmented villonodular synovitis (PVNS) is a rare proliferative disease of the synovium, most commonly seen in young and middle-aged adults. We report a 63-year-old man presenting an acute onset of swelling, a feeling of tightness in the right knee, and difficulty in squatting and standing from squatting position. The patient received conservative treatment, which was ineffective. Subsequent magnetic resonance imaging results led to a diagnosis of PVNS, and extensive arthroscopic synovectomy was performed but recurrent knee swelling developed gradually since the following month. The symptoms eventually subsided after a second arthrotomy followed by total synovectomy. However, the patient suffered from consequent atrophy of quadriceps muscles after months of immobility. He received regular rehabilitation until he could walk without a cane one year later. PVNS is a rare disease commonly seen in young or middle-aged adults; when occurring in an elderly patient, it can be easily neglected or confused with other joint conditions. Although favorable in general, local control rates and functional outcomes of PVNS could be far less desirable in older adults. Early diagnosis, intervention, and rehabilitation are important to achieve better functional outcomes.

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Key words: pigmented villonodular synovitis (PVNS), older adult, functional outcomes

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**Introduction**

Pigmented villonodular synovitis (PVNS) is a rare proliferative process that affects the joint synovial tissue, tendon sheath, and bursa. It is a locally aggressive lesion that may invade and destroy surrounding soft tissue and bone, resulting in functional deterioration of the joints and extremities. The estimated annual incidence is 1.8 per million population in United States [1,2]. PVNS often appears in the third and fourth decades of life [3] with an average age of 35 (14-68) years [4]. Diagnosis of this condition is frequently delayed or mistaken as osteoarthritis, a meniscal tear, or other ligamentous injury [3]. PVNS could cause diagnostic confusion in older adults since it has a relatively low incidence and can be easily misinterpreted as osteoarthritis, which has high prevalence among older adults.

**Case report**

This 63-year-old patient is a physician with a history of osteoarthritis of both knees and type 2 diabetes mellitus controlled by medications. He does not carry heavy weights in daily life nor does he have a regular exercise habit. He does not smoke or regularly consume alcohol.

The patient had felt occasional soreness in both knees for several years but did not pay much attention to the symptom nor did he take any pain killers. He had fallen into a ditch and injured his right knee 20 years ago, but there was no episode of acute inflammation on the knee after the incident.

He has had mild swelling in the right knee since March 2009, especially in the suprapatellar area. The lesions did not show changes in skin color; there was no pain or heat sensation; the range of motion was not limited. The symptoms did not cause much problem during the following 3 months. Acute progressive right knee swelling developed on June 12, 2009 and the patient began to have a tight sensation in his right knee, along with difficulty on squatting and standing from squatting position. There was no pain or heat sensation in the knees at the time.

Initial X-ray (Figure 1) revealed asymmetric joint space narrowing and spur formation. The suprapatellar effusion and surrounding soft tissue swelling were found only from the lateral view. Conservative treatment was attempted at first, but the symptoms were not relieved. Magnetic resonance imaging (MRI) was then performed on June 15, 2009 which showed a severe chronic tear in the bucket handle, wear in the lateral meniscus, hyaline cartilage in the lateral femoral condyle and lateral tibial plateau. Frond-like synovial proliferation with moderate effusion was noted in the suprapatellar bursa and the joint
Figure 1  Right Knee X ray (AP + lateral view)

A: AP view
The AP view only showed asymmetric joint space narrowing with spurs formation. The suprapatellar effusion and soft tissue swelling were obscure here.
B: Right lateral view
The suprapatellar effusion and surrounding soft tissue swelling were noted.

Figure 2  MRI of right knee

Fig. 2A, 2B
(Sagittal view, a: T2 weighted, b: T1 weighted-fat suppression with contrast)
1. Moderate effusion was noted in the suprapatellar bursa and the joint space of the right knee.
2. Frond-like synovial proliferations (arrow) were best observed on T2 weighted image (2A). They were low signal on T2 weighted image, and were strongly enhanced by contrast on T1 weighted-fat suppression image (2B).
Other findings included severe chronic bucket-handle tear and wearing of lateral meniscus and hyaline cartilage of lateral femoral condyle and lateral tibial plateau.

Fig. 2C, 2D
(Coronal view, c: proton weighted, d: proton weighted-fat suppression)
Other findings included severe chronic bucket-handle tear and wearing of lateral meniscus and hyaline cartilage of lateral femoral condyle and lateral tibial plateau.

space of the knee, and this characteristic was consistent with the symptom of pigmented villonodular synovitis (Figure 2).

The patient underwent extensive arthroscopic synovectomy on July 6, after 3 weeks of ineffective conservative treatment. However, recurrent knee swelling had developed gradually in the following month. He was further treated with total synovectomy via an arthrotomy on October 14, 2009. The histopathological report confirmed the diagnosis of PVNS.

After the operation, the joint condition improved but marked quadriceps atrophy developed after months of immobility. The patient spent much time in rehabilitation. The circumferences of his right and left thighs were 38 cm and 41 cm, respectively, after 6 months of muscle-strengthening exercise. He could finally walk without a cane one year later.

**Discussion**

The etiologies of PVNS remain unclear. Some researchers found the cause in chronic inflammation [5] while others treat it as a tumor-like disorder [6,7]. Histology of PVNS reveals hypertrophic synovial process characterized by villous, nodular, and villonodular proliferation and pigmentation from hemosiderin [8]. PVNS most commonly affects the large joints but may involve any synovial joint. The knee is the
most frequently affected joint, followed by the hip, shoulder, and other joints [9]. It is usually monoarticular. Patients with this disease often report accompanying pain, joint effusions, and swelling. There are two forms of PVNS. The diffuse form of PVNS (DPVNS) affects one compartment or the entire synovium of a given joint, while the localized form of PVNS (LPVNS) affects a single discrete mass in the synovium [3,10].

Result of plain radiography may be normal or may reveal nonspecific changes depending on the severity of the disease. In the knee joint, it shows an increased soft tissue density at the anterior or posterior synovial sac with or without a lobulated mass, which indicates synovial hypertrophy and joint effusion [10]. MRI is currently the preferred imaging modality for diagnosis [3,10]. Characteristic findings include hyperplastic synovium and heterogeneous signal intensity in all the imaging sequences in combining low signal intensity areas (created by paramagnetic effects of hemosiderin deposits and fibrous tissue) and high signal intensity areas (representing congested synovium and fat content) [10]. The low signal blooming effect of hemosiderin is best seen on T2-weighted gradient-echo images. Joint effusion at the periphery of the lesion is noted, demonstrating low- to intermediate-signal intensity on T1 and proton density images and high signal on T2-weighted sequences [10]. MRI could also help to determine the extent of disease involvement and to distinguish DPVNS from LPVNS [3].

The optimal treatment of LPVNS is marginal excision [11]. Primary cases of DPVNS are treated by complete synovectomy, which can be accomplished by arthroscopic or open techniques if the lesion is located within the joint [10]. In cases of extra-articular involvement, open synovectomy should be considered. Incomplete synovectomy is associated with a high incidence of recurrence [10]. Postoperative radiotherapy with low to moderate doses may be beneficial if there is residual tumor or recurrence. MRI appeared to be highly sensitive but less specific for detecting residual pigmented villonodular synovitis postoperatively because of the confounding presence of surgical changes within the knee [4]. The local control rates after treatment were relatively high (78–98%), and functional outcomes were good or excellent for most patients (86–93%) according to previous case series [11].

PVNS is a rare disease which is commonly seen in young or middle-aged adults, and it could be easily neglected or confused with other joint conditions when happening to older adults. Although local control rates and functional outcomes are good in general, they could be far less favorable in older adults. Early diagnosis, intervention, and rehabilitation are
important for better functional outcomes.

References


老年色素絨毛結節滑液膜炎病例報告

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摘要

色素絨毛結節滑液膜炎是一種罕見的增生性良性關節病變，通常發生於年輕人或中年人。我們報告一個 63 歲男性個案，初始表現為急性右膝腫脹無力。他在接受保守性症狀治療無效之後，才經由核磁共振檢查診斷出色素絨毛結節滑液膜炎。個案在接受第一次膝關節鏡清除手術後，關節腫脹仍持續並有許多滲出液。第二次開放性滑液膜全切除術後關節症狀終於緩解，但由於長期靜止不動造成右大腿萎縮無力，他被迫進行為期一年的復健才能自由行動。色素絨毛結節滑液膜炎是一種通常發生於中壯年的罕見關節病變，此種疾病若發生在年長者身上容易與其他關節疾患混淆以致延誤治療。此外，年長者術後功能回復也可能較一般人來得差。早期診斷治療以及術後復健，對於良好預後以及術後日常生活功能的維持甚為重要。

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關鍵詞：色素絨毛結節滑液膜炎、年長、日常生活功能