


CASE REPORT

Combined type of nodular and diffuse forms of pigmented villonodular synovitis in a single knee joint of a child

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Abstract

There are two types of pigmented villonodular synovitis (PVNS), nodular and diffuse. Two types generally do not coexist, and only one type usually occurs. This report presents a rare case of 12-year-old boy with combined type of PVNS in the knee joint who was treated arthroscopically.

KEYWORDS

child, combined, knee, pigmented villonodular synovitis

1 | INTRODUCTION

Pigmented villonodular synovitis (PVNS) shows hyperplasia of the synovial tissue in the joints, tendon sheaths, or fibrous tissue. Although inflammatory and neoplastic causes have been hypothesized, the etiology of PVNS remains unknown. The estimated annual incidence of PVNS is about 1.8 per million in the worldwide population.¹ PVNS is usually found in patients between 30 and 50 years old, and rarely occurs in children.¹ It has two types, nodular or localized, and more frequently, the diffuse type of villous synovial proliferation is seen. The two types of PVNS show different characteristics. The nodular form appears as a single intra-articular nodule, which may be seen as a loose body. The diffuse form presents clinically as chronic monoarthritis. Hemarthrosis is a common finding in patients with diffuse PVNS. These two types of PVNS generally do not coexist. We present a very rare case of PVNS, in which both diffuse and nodular forms were found together in the right knee of a 12-year-old boy. To the best of our knowledge, there are no other reports of both types of PVNS occurring in a single joint of a child.

Written, informed consent was obtained from the parents of the patient to publish this case report, including the images.

2 | CASE REPORT

A 12-year-old boy presented with complaint of intermittent painful swelling on the right knee for the previous 3 months. He had a history of falling on the floor and hitting his anterior knee against the floor 3 months before the visit. His parents stated that bloody fluid was aspirated from the joint in a primary care unit at that time. A month later, painful swelling recurred in the same knee simply after standing for a short time. He denied fever, myalgia, or chills. His other medical and psycho-social history were not remarkable. He had no previous intervention on the involved knee. There was no notable family history. Physical examination revealed mild tenderness on the anterior knee, effusion, and limited range of motion from 0 to 130 degrees. There was no palpable mass, instability, redness, or local heat.

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Arthrocentesis revealed 85 ml of reddish and slightly turbid fluid with 1200 polymorphonuclear cells/ml in joint fluid analysis.



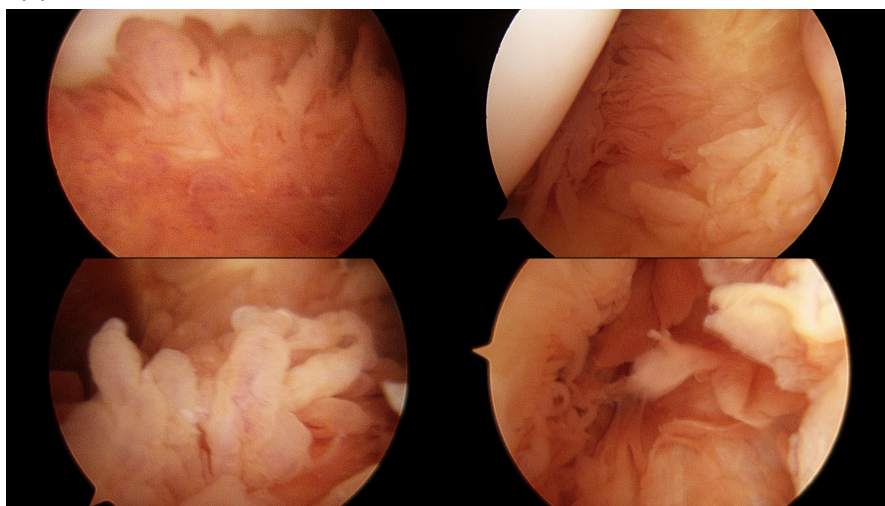
FIGURE 1 T2-weighted sagittal image of right knee: Arrowheads indicate diffuse form of irregularly marginated heterogeneous mass with low-to-intermediate signal intensity in the knee joint, and arrows indicate nodular form of well marginated mass with low signal intensity.

A magnetic resonance imaging (MRI) study was done ([Figure 1](#)). T2-weighted images showed both an irregularly marginated mass spreading through the suprapatellar pouch, posterior compartment, and intercondylar space, and a well-circumscribed mass confined to the anterior compartment with low-to-intermediate heterogeneous signal intensity. Hemosiderin-laden foci, which showed low signal intensity in both T1- and T2-weighted images, were observed to be scattered throughout the mass.

With the impression of localized PVNS and reactive synovial proliferation around the mass, arthroscopic examination, and synovectomy were recommended. However, surgery was delayed for 3 months due to personal reasons of the patient. On arthroscopy, a brownish nodule was found in the anterior compartment of the knee and there was extensive villi formation in the joint cavity including the suprapatellar pouch, medial and lateral gutters, anterior compartment, and posterior compartment ([Figure 2](#)). Excision of the nodular mass and total synovectomy were performed. For the differential diagnosis of other diseases such as synovial chondromatosis, nodular synovitis, and hemophilic arthropathy, a biopsy was also performed.

The histological findings in both the nodular mass and villi specimens showed giant cells and hemosiderin-laden macrophages ([Figure 3](#)). They both had similar histological appearances, but there was greater cellularity

(A)



(B)

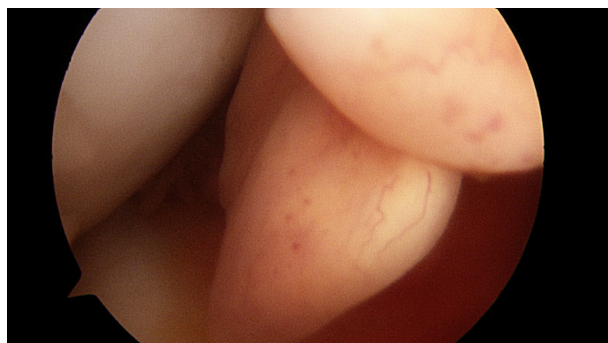


FIGURE 2 Multiple villi are showed in the suprapatellar pouch and the posterior compartment of the knee (A). Well marginated brownish mass is showed at the anterior compartment of the knee (B).

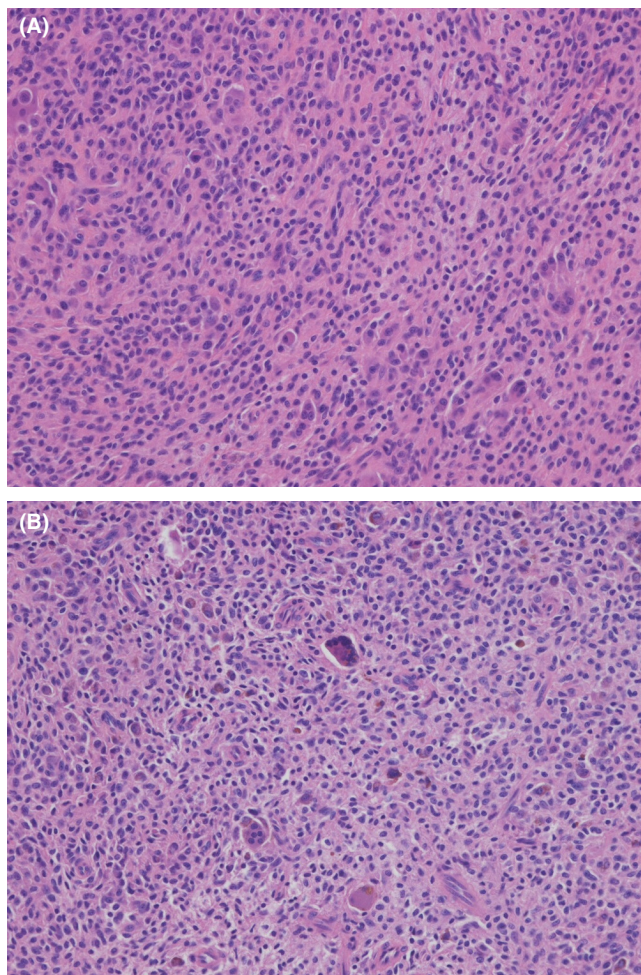


FIGURE 3 H&E stain (200 \times) of nodular form shows giant cells and hemosiderin-laden macrophages (A). H&E stain (200 \times) of diffuse form shows giant cells and hemosiderin-laden macrophages. It has higher cellularity and pigmentation than the nodular form (B).

and pigmentation in the diffuse form than in the nodular form. The mononuclear component was comprised of two types of cells, small histiocyte-like cells, and larger cells. Mononuclear stromal cells infiltrating the synovium, hyperplastic synovial cells, hemosiderin-stained multinucleated giant cells, and pigmented foam cells (lipid-laden histiocytes) were seen, which was consistent with PVNS.

Since the surgery, the patient has not had any symptoms such as swelling, pain, or limited motion for 3 years.

3 | DISCUSSION

Pigmented villonodular synovitis is an idiopathic, monoarticular reactive synovial disease characterized by the proliferation of synovial villi or nodules in a single joint. PVNS can be caused by a reactive response to trauma. About a third of the patients report previous trauma history in the

area of affected lesions.² However, gene fusion involving CSF 1 (1p13) and COL6A3 (2p35) has been found in PVNS patients,³ which means that PVNS is a neoplastic lesion rather than an inflammatory process. An immunohistochemical study by O'Connell et al.⁴ suggested that PVNS was a tumor of synovial surface cell origin. Radiographs do not usually show soft tissue abnormalities. However, radiographs and CT may sometimes show joint effusion or cystic erosion with sclerotic margins. If a patient's symptoms persist, arthroscopy and/or open synovectomy can be considered first as treatment.

There are two forms of PVNS, each with different characteristics. The localized form of PVNS is found mostly in the hands and large joints of patients, such as the ankle and the anterior compartment of the knee.⁵ The most common symptom is painless swelling of the involved joint. Radiographically, osseous erosion may be seen, and MRI shows a well-margined soft-tissue mass. Its recurrence rate after synovectomy is reported to be about 8%.⁶ The localized form is histologically identical to a giant cell tumor of the tendon sheath. Marginal excision is the recommended treatment. In contrast, the diffuse form of PVNS most commonly affects the knee (about 80%), but the hip, ankle, shoulder, wrist, and other joints can be involved. The symptoms are more severe than those of localized PVNS. Radiologically, degenerative changes in the joint are often seen. The recurrence rate is about 20%.⁷ The diffuse form is histologically similar to the localized form but usually involves the entire synovium. The diffuse form may show a more infiltrative pattern and less common hyalinization of the stroma, compared to the localized type. Hemosiderin deposition is more prominent in the diffuse form.⁸ As the recurrence rate is high, total synovectomy is often recommended for patients with the diffuse type, and adjuvant radiation therapy may also be employed. Since radiation may cause severe side effects such as permanent skin changes, joint stiffness, and radiation-induced sarcoma, new targeted drugs such as imatinib, which inhibits macrophage colony-stimulating factor receptors, have been introduced.⁵

It is important to distinguish between the two types of PVNS not only because of their different appearances, but because of their clinical differences, such as recurrence rates, treatment methods, and potential to become neoplastic. Perka et al.⁹ suggested that PVNS should be classified more strictly than before into a potentially neoplastic (diffuse) form and a reactive granulomatous (localized) form and that these two types of PVNS should be approached differently.

The child in this case uniquely had both diffuse and nodular forms in a single knee joint. MRI, arthroscopy, and gross findings showed different appearances of both

the diffuse and nodular forms. The microscopic findings were very similar except for the different degrees of pigmentation and cellularity. Although they seem to be different disease entities considering their distinctively different clinical characteristics, this case report suggests that they may coexist and develop from the same origin. We are not sure if there were any relationships between the two forms. Before encountering this case, we assumed that the two types were independent. Further studies are needed to examine how these two different forms of PVNS may be related.

In summary, PVNS is a rare cause of painful swelling of knees in children. The diffuse and localized types of PVNS may be related, although their clinical characteristics are distinctly different. Both forms of PVNS can co-exist in the same joint. MRI and arthroscopy are helpful for diagnosis.

AUTHOR CONTRIBUTIONS

Dong Hwan Kim: Writing – review and editing. **Jung Ho Noh:** Conceptualization; writing – original draft.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

ETHICAL APPROVAL

The authors have not obtained IRB approval as a case report of three or fewer patients is not considered to be human research according to IRB policy.

CONSENT

Written informed consent was obtained from the parents of the patient to publish this report in accordance with the journal's patient consent policy.

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