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Case Report

Atypical and bilateral pigmented villonodular synovitis of wrist in an adolescent patient: case report and literature review

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Abstract: Background: Pigmented villonodular synovitis (PVNS) is a rare benign proliferative disorder of the synovium. It rarely occurs in adolescents, particularly in immature patients with bilateral manifestation. Case presentation: We present a case of atypical and bilateral PVNS of wrist in a 14-year-old boy. Initially, the patient presented with left wrist pain and swelling without the history of trauma. Physical examination revealed an obvious lesion in the dorsal part of left wrist. Radiographs, computed tomography (CT) and magnetic resonance imaging (MRI) showed multiple abnormal signal shadows and arthroedema in the left wrist. Arthroscopy operation was performed, and histologic examination suggested the diagnosis of PVNS. Only 10 months later, the patient presented with the similar symptoms and signs in the right wrist. But MIR and histologic examination were atypical. In this article, we also review and summarize 26 studies on 30 adolescent patients with PVNS. Conclusions: This study provides an example of atypical and bilateral PVNS in adolescents.

Keywords: Pigmented villonodular synovitis, adolescent, wrist

Introduction

Pigmented villonodular synovitis (PVNS) is defined as a rare benign proliferative disorder of the synovium with unknown etiology and pathogenesis. Chassaignac first described PVNS in 1852 [1]. In 1941, Jaffe et al. found that PVNS was characterized by a progressive synovial proliferative lesion with the formation of villi or lobulated nodules in the joint, bursae, and tendon sheath [2]. PVNS can be divided in two forms: diffuse and localized. The diffuse form has an effect on the whole synovium of the joint. The localized form commonly involves the synovium as a pedunculated nodule. The isolated and discontinuous lesion that occurs only in tendon sheath is also known as giant cell tumor of tendon sheath.

The incidence of PVNS is approximately 1.8 patients per million population each year [3]. It usually can be found in adult patients aged 20 to 50 years, and the distribution between men and women is equal [4, 5]. A patient commonly presents with insidious symptoms of pain and swelling. PVNS is difficult to distinguish from other synovial diseases of inflammation and neoplasms, including rheumatoid arthritis, hemophilic arthropathy, and tuberculosis. The knee is the most common site of pathologic changes, followed by other large joints such as hip.

Although several case studies have reported PVNS occurs in children [6], it has rarely been documented in adolescents, especially bilateral PVNS of wrist. Here, we report a case of atypical and bilateral PVNS in an immature boy, and present a review of the published literature among adolescents.

Case presentation

A 14-year-old boy presented with a 5-month history of pain and swelling in the left wrist, that could not be relieved by drug treatment at

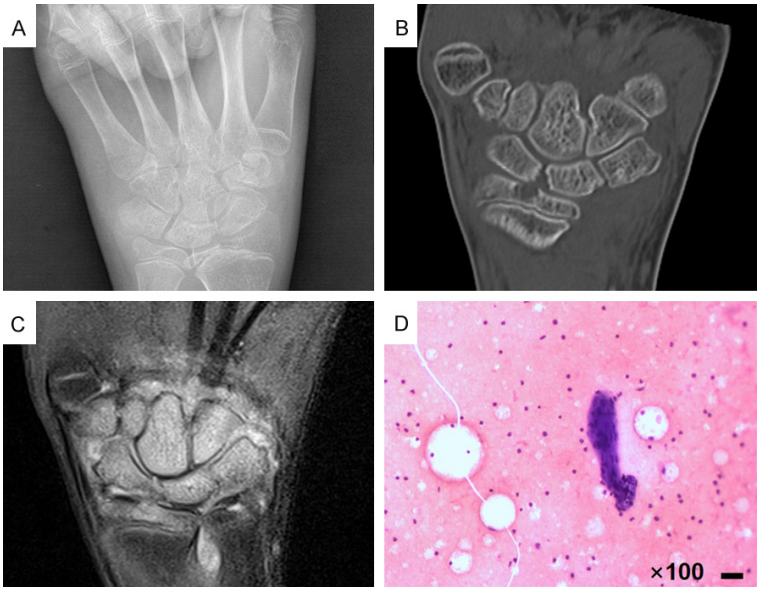

Figure 1. Anteroposterior radiograph (A) and CT image (B) of the left wrist demonstrated multiple bone destruction foci; (C) MRI of the left wrist showed multiple abnormal signal shadows and arthroedema; (D) Cytologic examination indicated a few short spindle cells and inflammatory cells in the swelling of the left wrist. Scale bar, 200 μm. Magnification, 100 ×.

his local hospital. There was no preceding history suggestive of trauma. Physical examination revealed a 3 cm × 2 cm size lesion in the dorsal wrist without overlying skin changes. Radiographs of the left wrist were obtained, which indicated multiple bone destruction and soft tissue swelling (**Figure 1A**). Computed tomography (CT) also demonstrated multiple bone destruction of the left wrist (**Figure 1B**). Magnetic resonance imaging (MRI) was performed, which showed multiple abnormal signal shadows and arthroedema in the left wrist (**Figure 1C**). Hence, puncture examination in the swelling of the left wrist was performed. 0.5 ml gray-red liquid was extracted and then was sent to the pathology department for cytologic examination. As shown in **Figure 1D**, there were a few short spindle cells and inflammatory cells under the microscope. From the above, the reason for left wrist bone destruction was not clear, and the final diagnosis still neededpathologic biopsy. Arthroscopy for left wrist lesion exploration and a removal operation was performed.

Histopathologic examination showed villous and nodular configuration (**Figure 2A-C**), hyperplastic synovium (**Figure 2D-F**), and pigmented area (**Figure 2G-I**), suggesting a diagnosis of PVNS. Due the hyperplastic synovial spread into adjacent joint spaces, it was difficult to remove completely. Radiotherapy with 3400 CGy dose was performed after operation.

Only 10 months later, the patient returned because of pain and swelling in the right wrist. Physical examination showed a solid mass with a diameter of 3 cm. MRI suggested an infectious lesion in the right wrist (**Figure 3A**, **3B**). Again, arthroscopy for the right wrist lesion exploration and removal operation was performed. Histologic examination showed only proliferative and degenerate fibrous adipose tissue and focal granulation tissue (**Figure 3C**, **3D**). Because of the lack of nodular appearance, the right wrist lesion failed to be diagnosed as PVNS. However, in consideration of the similar clinical features of bilateral wrist lesions, the diagnosis of PVNS was the most likely suggested. During the 2-year follow-up period, the patient did not show any signs of recurrence in the bilateral wrists.

Discussion and conclusions

This study gives an overview of PVNS in adolescents. A comprehensive literature search was performed prior to September 2th, 2019. As shown in **Table 1**, a total of 26 studies with 30 adolescent patients were obtained in this review [7-32]. The mean age at presentation was 14.3 years. PVNS in adolescents occurred most commonly in the knee, similar to adult patients. Among them, there were 9 male patients (30.0%) and 20 female patients (66.7%). Pain and swelling were the most common clinical presentation in adolescents. Arthroscopic synovectomy (AS) and open synovectomy (OS) were the main treatments for PVNS patients. After operation, most adolescent patients showed no evidence of disease, and only two patients recurred [8, 19].

This case we reported provides an example of atypical and bilateral PVNS in adolescent. The patient showed similar symptoms and signs

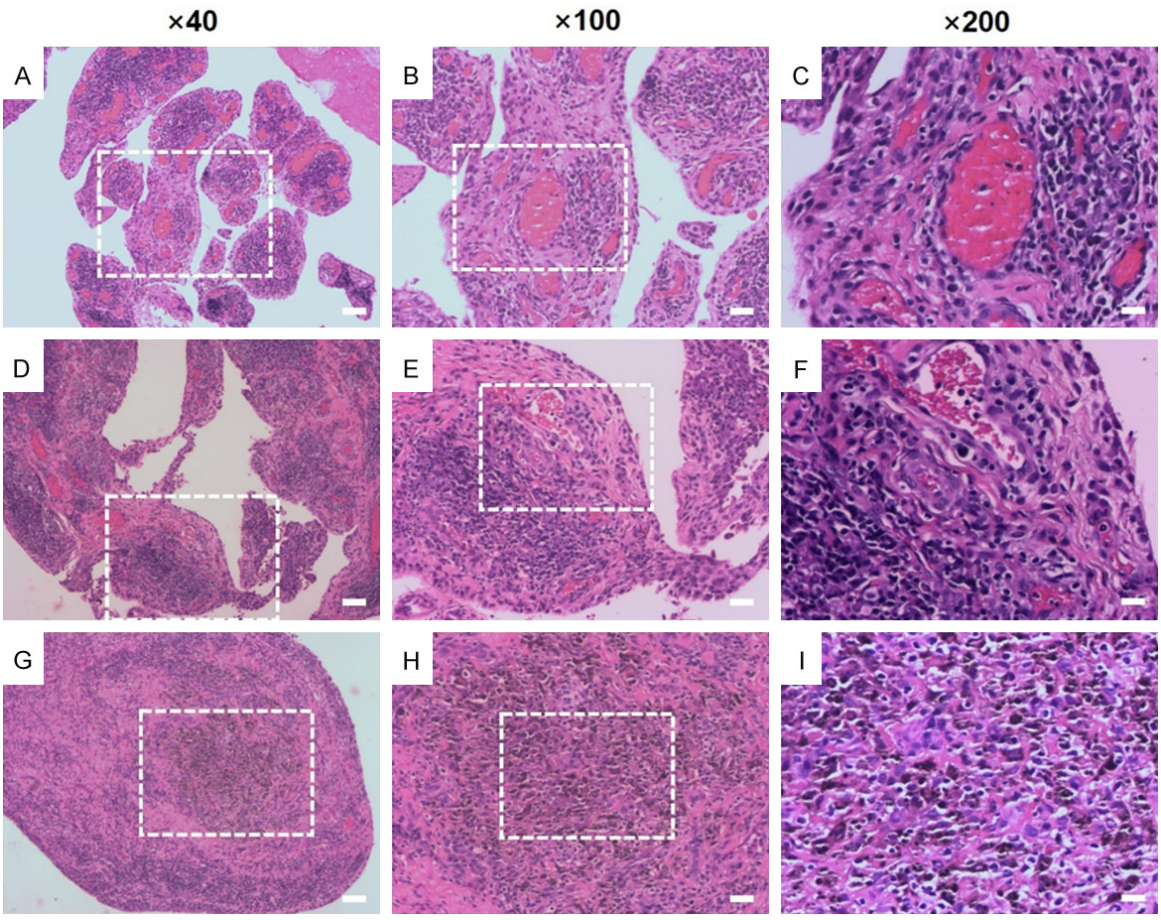

Figure 2. Histopathologic examination of the left wrist showed villous and nodular configuration (A-C), hyperplastic synovium (D-F), and pigmentation area (G-I) at different magnifications. Scale bar, 200 μm.

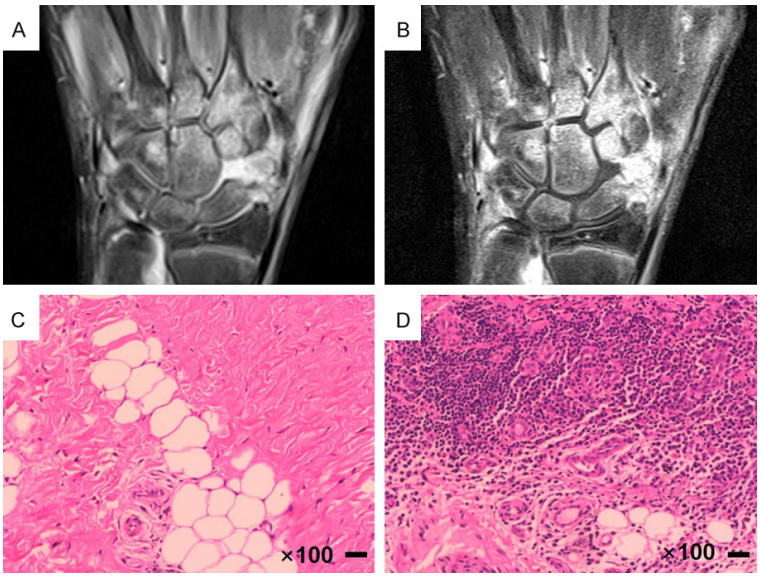

Figure 3. T2 (A) and T1-weighted (B) MRI image showed an infectious lesion of the right wrist; Histologic examination of the right wrist showed proliferative and degenerate fibroadipose tissue (C) and focal granulation tissue (D). Scale bar, 200 μm. Magnification, 100 ×.

with PVNS in the left wrist, but the result of MRI examination was atypical. We can confirm the diagnose of PVNS through the gold standard of histopathology. Then, the patient also presented with pain and swelling in the right wrist after the operation on the left wrist. We only suggested the right wrist as PVNS through clinical features, due to the atypical results of MRI and pathologic examinations.

Until now, the etiology of PVNS has not been identified. Whether PVNS is a chronic inflammatory lesion or a neoplasm of synovial tissue is controversial [33-35]. Moreover, the cellular origin of PVNS is not clear.

Table 1. Summary of clinical characteristics of pigmented villonodular synovitis in the adolescent literature

Author (year)	Age (years)	Sex	Location	Clinical presentation	Treatment	Clinical result	Follow up
Atik et al. 2017	14	M	Knee	Intermittent swelling	OS	NED	NA
Baroni et al. 2010	12	F	Knee	Gonalgia	NA	NA	90 months
	14	F	Knee	Gonalgia	NA	NA	78 months
	15	F	Knee	Gonalgia	NA	NA	108 months
Bessette et al. 1992	15	M	Knee	Recurrent swelling	OS	NA	NA
Brenner et al. 2007	17	F	Knee	Swelling	AS+OS	NED	1 month
Duncan et al. 2015	12	M	Calcaneocuboid	Swelling	OS	NED	12 months
Hoeffel et al. 1997	16	F	Wrist	Pain and swelling	OS	NA	NA
Hulailah et al. 1998	12	F	Knee	Pain and swelling	AS	NA	NA
Jerome et al. 2009	16	NA	Elbow	Pain and swelling	OS	NED	48 months
Kaneko et al. 2000	14	F	Ankle	Swelling	NA	NED	3.4 years
Kang et al. 1992	12	F	Knee	Pain and limping	NA	NA	NA
Kim et al. 1997	16	M	Ankle	Pain and swelling	NA	NED	25 months
Kim et al. 2005	12	M	Knee	Pain	AS	NED	14 months
Kimura et al. 2015	14	F	Lumbar spine	Pain	OS	NED	NA
Klammer et al. 2013	16	F	Knee	Pain	AS+OS	Recurrence	3.5 years
Lunawat et al. 2012	14	M	Great toe	Swelling	NA	NA	NA
Mascheroni et al. 2008	13	F	Ankle	Pain and swelling	AS	NA	NA
Matsui et al. 2001	13	M	Knee	Haemarthrosis	AS	NA	NA
Müller et al. 1999	16	M	Shoulder	Pain and swelling	NA	NA	NA
Novikov et al. 2018	11	F	Ankle	Discomfort and swelling	NA	NED	5 years
Pannu et al. 2014	17	F	Metatarsophalangeal	Pain	OS	NED	1 year
Sanchis-alfonso et al. 2000	16	F	Ankle	Pain and swelling	AS	NED	NA
Spritzer et al. 1987	16	M	Knee	Pain and swelling	NA	NA	NA
Willimon et al. 2018	15	F	Hip	Pain	AS	NED	12 months
	12	F	Hip	Pain	AS	NED	53 months
	17	F	Hip	Pain	AS	NED	14 months
Wyllie et al. 1969	13	F	Knee	Pain and swelling	NA	NED	12 months
Yamashita et al. 2012	14	F	Knee	Pain	AS	Recurrence	2.6 years
Zampeli et al. 2015	16	F	Ankle	Pain and swelling	AS	NED	1 year

Note: M: male; F: female; AS: arthroscopic synovectomy; OS: open synovectomy; NA: not available; NED: no evidence of disease.

Physical examination of PVNS patients may present with pain, swelling, hemarthrosis, or erythema [24]. The typical MRI result of a patient with PVNS may show low signal areas on T1- and T2-weighted images, suggesting signal attenuation by hemosiderin [25]. The histologic characteristics of PVNS reveal a villous and/or nodular proliferation of the synovium including three cellular components: mononuclear stromal cells, multinucleate giant cells, and xanthoma cells [17]. A patient with PVNS requires complete open or arthroscopic synovectomy for treatment [7]. Adjuvant chemotherapy and radiation depend on the severity level of PVNS [36].

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Disclosure of conflict of interest

None.

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