Clinical Case Report

Medicine®

OPEN

Multifocal pigmented villonodular synovitis in a child

A case report

Liang Zhao, MD[a,b], Kaiyu Zhou, MD[a,b], Yimin Hua, MD[a,b], Yifei Li, MD[a,b], Dezhi Mu, MD[a,b,*]

Abstract

Introduction: Pigmented villonodular synovitis (PVNS) is a rare, benign proliferative disorder of the synovial membrane that typically presents in adults and affects a single joint. Multifocal PVNS is very rare, particularly in childhood. We reported a rare case of multifocal PVNS affecting over 20 joints in a child.

Clinical procedure: A 7-year-old female patient had a 6-month history of multifocal joints swelling with mild pain. She was diagnosed as polyarticular juvenile idiopathic arthritis at a local hospital. Naproxen, methotrexate, infliximab, and pavlin were used to treat the patient for 2 months. However, the treatment had no effect, the joints swelling remained. The patient was then transferred to our hospital. Physical examination revealed multiple joints swelling, especially in the shoulders joints. Puncture fluid from a shoulder joint was bloody. Magnetic resonance imaging (MRI) revealed synovial thickening and hemosiderin deposition. Biopsy of joint synovium found villous nodules, the invasion of foam cells, and hemosiderin deposition. By collecting all of the evidence, the diagnosis of PVNS was confirmed.

Conclusions: PVNS was easily misdiagnosed as rheumatoid arthritis and the formal treatment was usually delayed. This case described here is the first case of PVNS involving such a large numbers of joints that has been reported in the literature.

Abbreviations: HE = hematoxylin and eosin, MRI = magnetic resonance imaging, PVNS = pigmented villonodular synovitis.

Keywords: child, multifocal, pigmented villonodular synovitis

1. Introduction

Pigmented villonodular synovitis (PVNS) is a rare, benign proliferative disorder of the synovial membrane that typically presents in adults and affects a single joint. Multifocal PVNS is rare, particularly in childhood.[1,2] We report a rare case of multifocal PVNS affecting over 20 joints in a 7-year-old female to highlight the importance of considering PVNS as a differential diagnosis in children presenting with long-term joint swelling and to reduce the occurrence of misdiagnosis.

2. Case presentation

A 7-year-old female had a 6-month history of multifocal joint swelling with mild pain but without joint dysfunction, fever, or rash. She had experienced no trauma and had no relevant family history. The initial suspected diagnosis determined at a local hospital was polyarticular juvenile idiopathic arthritis with negative rheumatoid factor. After undergoing treatment with naproxen, methotrexate, infliximab, and pavlin for 2 months, the joint swelling remained, although pain had been relieved. The child was then referred to our hospital. Physical examination revealed a 4.5 cm × 4.5 cm symmetric cystic mass in the region of the bilateral shoulder joints (Fig. 1) without signs of redness, tenderness, or high skin temperature. Slightly swollen knees, wrists, and fingers were also noted, and the fluctuation test was positive. Results within normal ranges were obtained for all laboratory examinations, including routine blood tests; assessments of C-reactive protein, erythrocyte sedimentation rate, biochemistry, coagulation function, autoantibodies, rheumatoid factor, transfusion immunity, mycoplasma antibody, and bone marrow; rheumatism screening; the fungal G test; and the tuberculosis skin test. No abnormal signs were found in thoracic and abdominal enhanced CT examinations. Joint effusion and synovial thickening in the bilateral hips, knees, ankles, shoulders, elbows, and metatarsophalangeal joints 1–5 were observed via ultrasonography. Erosion of the humerus bone was visible on the right shoulder x-ray (Fig. 2). Magnetic resonance imaging (MRI)

Editor: Stuart Barry Polisner.

Consent: Ethical approval was obtained from the Ethics Committee of West China Second University Hospital, Sichuan University. Written informed consent was obtained from the parents of the patient for publication of this case report and any accompanying images.

Funding: This work was supported by grants from the National Science Foundation of China (No. 81330016) and the Subject of Sichuan Provincial Department of Health (No. 130124).

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

[a] Department of Pediatrics, [b] Key Laboratory of Ministry of Education for Obstetric & Gynecologic and Pediatric Diseases and Birth Defects, West China Second University Hospital, Sichuan University, Chengdu, China.

[*] Correspondence: Dezhi Mu, Department of Pediatrics, West China Second University Hospital, Sichuan University, Chengdu, Sichuan, China (e-mail: mudz@scu.edu.cn).

Medicine (2016) 95:33(e4572)

Received: 30 May 2016 / Received in final form: 17 July 2016 / Accepted: 19 July 2016

http://dx.doi.org/10.1097/MD.0000000000004572

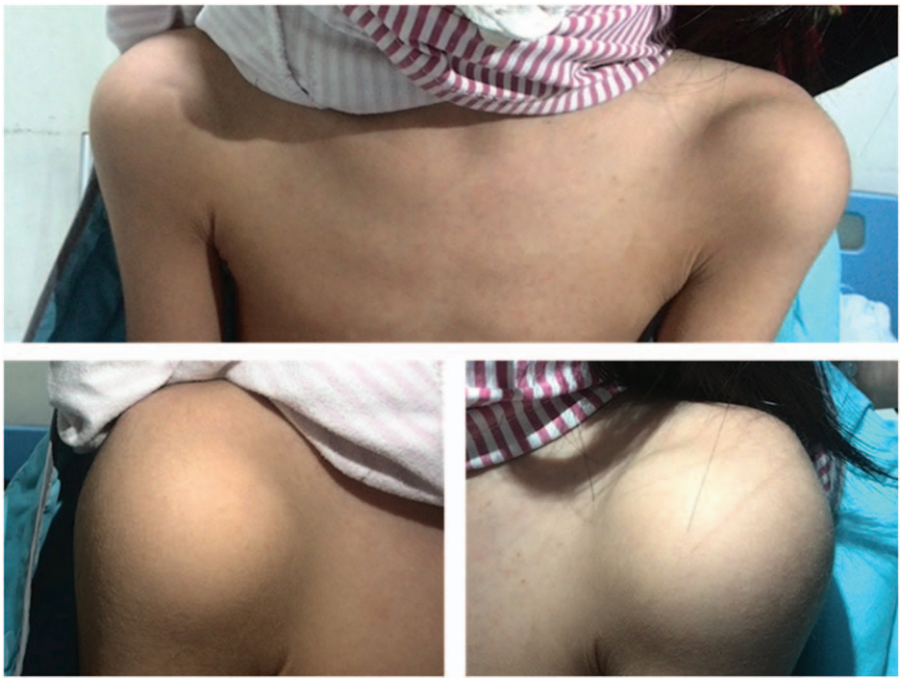

Figure 1. Obvious swelling of the bilateral shoulder joints.

revealed obvious joint effusion, diffuse synovial thickening, and hemosiderin deposition within synovial masses (Fig. 3). Puncture fluid from the left shoulder joint was bloody, and its smear and culture were negative. Pathological examination of the joint fluid demonstrated the presence of many lymphocytes and phagocytic cells but no tumor cells. Biopsy of the left knee joint synovium and capsular tissue indicated the obvious expansion of villous nodules, the invasion of foam cells, and hemosiderin deposition (Fig. 4), confirming the diagnosis of PVNS.

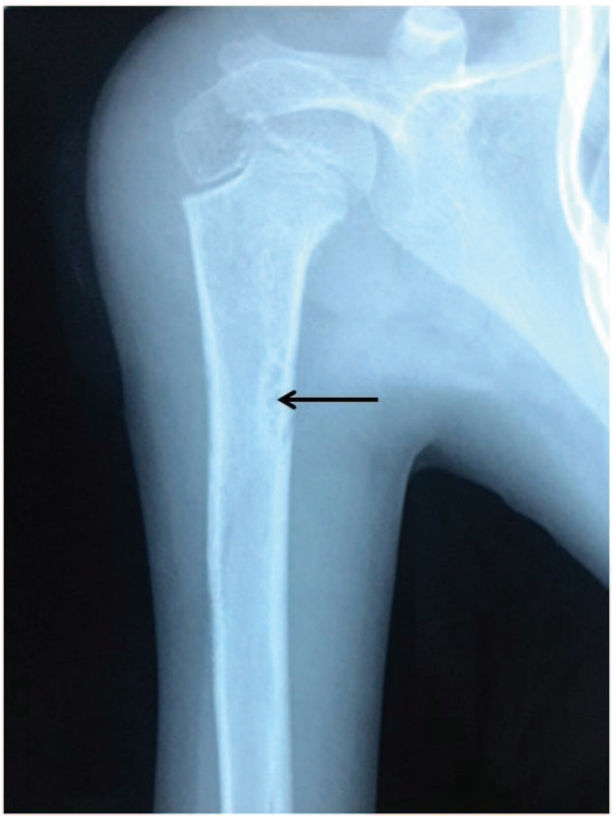

Figure 2. X-ray of the humeral bone that shows the erosion area.

3. Discussion

PVNS is a rare, benign proliferative disorder of the synovial membrane.[3] Its annual incidence rate is ~1.8 cases per million.[4] PVNS usually presents in adults between 30 and 40 years of age and is rarely observed in childhood.[5] The main clinical manifestations of PVNS include repeated bleeding, swelling, and sometimes pain in the affected joints. In late-stage PVNS, joint dysfunction occurs because bone and cartilage may be

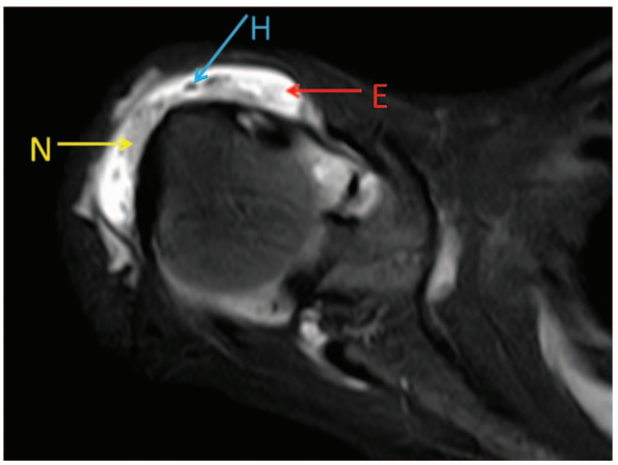

Figure 3. MRI of the right shoulder that shows asynovial nodule (N), joint effusion (E), and hemosiderin deposition (H) within synovial masses.

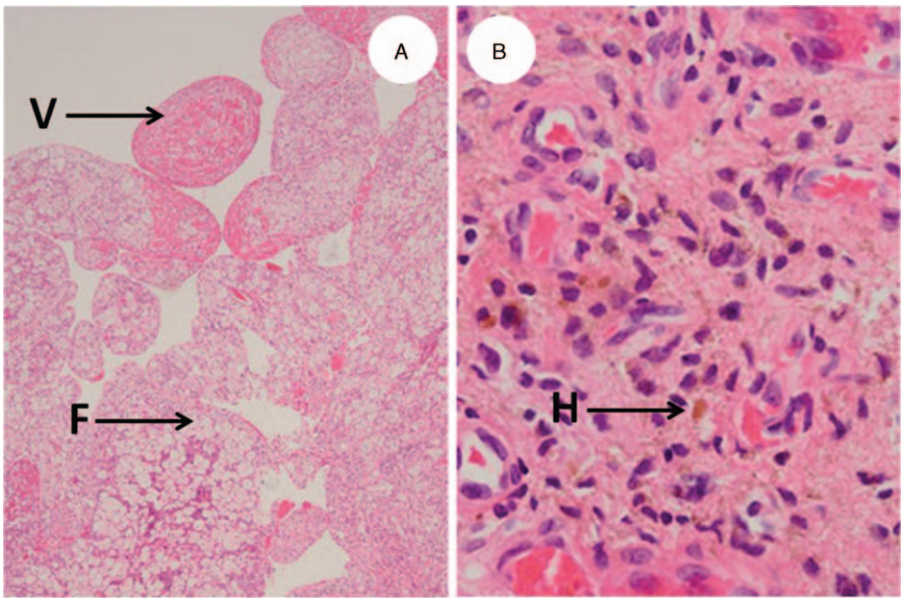

Figure 4. (A) Low-power photomicrograph that shows a villous nodule (V) and foam cells (F) (HE × 40). (B) High-power photomicrograph that shows hemosiderin (H) (HE × 400). HE = hematoxylin and eosin, MRI = magnetic resonance imaging.

invaded.[5–7] Although multiple joints can be involved, PVNS most commonly affects only a single joint.[3]

MRI has been recognized as the best imaging method for diagnosing PVNS.[8,9] MRI features of PVNS include variable synovial proliferation, joint effusion, and bone erosion; 1 particularly notable characteristic is hemosiderin deposition within synovial masses.[9] Diffuse or nodular synovial hyperplasia, multinucleated giant cells, foam cell infiltration, and hemosiderin pigmentation are typical findings from histopathological analyses of the joint synovium and capsular tissue.[10]

The differential diagnosis of PVNS includes infection, tumor, hemarthrosis, and rheumatoid arthritis.[11–14] PVNS is easily misdiagnosed because of its rarity. In extant literature reports, PVNS diagnoses have almost always been delayed.[14–16] In the largest report, which involved 237 patients, the median delay from initial clinical symptoms to final diagnosis was ~18 months.[16] Because PVNS has a much lower incidence in pediatrics, this disease is more easily misdiagnosed in children. PVNS requires radical treatment that combines prosthetic arthroplasty and synovectomy.[3] However, frequent recurrence after synovectomy has been observed.[17]

The child described in the present study mainly manifested with long-term multifocal joint swelling. After the possibilities of infection, tumor, and systemic bleeding disorders were excluded via multiple examinations, the initial suspected diagnosis was polyarticular juvenile idiopathic arthritis. However, antirheumatic treatment was ineffective. The eventual diagnosis of PVNS was not confirmed until joint MRI and biopsy were performed. Due to her multiple lesions, the child refused to undergo surgery; instead, traditional Chinese medicine was orally administered.

We suggest that PVNS should be considered when a child has unexplained long-term joint swelling with bloody joint puncture fluid and no remarkable routine examination findings. Joint MRI and histopathological assessment of the joint synovium and capsular tissue should be performed to confirm this diagnosis. To our knowledge, the described case is the first case of PVNS involving such a large number of joints in a pediatric patient that has been reported in the literature.

Acknowledgments

The authors would like to thank Dr. Liang Xie for his help with this case.

References

[1] Safaee M, Oh T, Sun MZ, et al. Pigmented villonodular synovitis of the temporomandibular joint with intracranial extension: a case series and systematic review. Head Neck 2015;37:1213–24.
[2] True VL, Monsell FP, Smith TA, et al. A severe systemic presentation of pigmented villonodular synovitis in a child with underlying Chediak–-Higashi syndrome. J Pediatr Orthop B 2015;24:526–9.
[3] Botez P, Sirbu PD, Grierosu C, et al. Adult multifocal pigmented villonodular synovitis-clinical review. Int Orthop 2013;37:729–33.
[4] Myers BW, Masi AT, Feigenbaum SL. Pigmented villonodular synovitis and tenosynovitis: a clinical epidemiologic study of 166 cases and literature review. Medicine (Baltimore) 1980;59:223–38.
[5] Duncan N, Rajan R. Case report of pigmented villonodular synovitis arising from the calcaneocuboid joint in a 12 year old male. Foot (Edinb) 2015;25:59–61.
[6] Oh SW, Lee MH, Eoh W. Pigmented villonodular synovitis on lumbar spine: a case report and literature review. J Korean Neurosurg Soc 2014;56:272–7.
[7] Sung KS, Ko KR. Surgical outcomes after excision of pigmented villonodular synovitis localized to the ankle and hindfoot without adjuvant therapy. J Foot Ankle Surg 2015;54:160–3.
[8] Lang N, Yuan HS. Computed tomography and magnetic resonance manifestations of spinal pigmented villonodular synovitis. J Comput Assist Tomogr 2015;39:601–6.
[9] Cheng XG, You YH, Liu W, et al. MRI features of pigmented villonodular synovitis (PVNS). Clin Rheumatol 2004;23:31–4.
[10] SaxenaA , Perez H. Pigmented villonodular synovitis about the ankle: a review of the literature and presentation in 10 athletic patients. Foot Ankle Int 2004;25:819–26.
[11] Meena S, Gangary SK. Knee tuberculosis masquerading as pigmented villonodular synovitis. J Res Med Sci 2014;19:1193–5.
[12] Van Der Heijden L, Gibbons CL, Dijkstra PD, et al. The management of diffuse-type giant cell tumour (pigmented villonodular synovitis) and

giant cell tumour of tendon sheath (nodular tenosynovitis). J Bone Joint Surg Br 2012;94:882–8.
[13] Ramesh B, Shetty S, Bastawrous SS. Pigmented villonodular synovitis of the knee in a patient on oral anticoagulation therapy: a case report. J Med Case Rep 2009;3:1212009.
[14] Neubauer P, Weber AK, Miller NH, et al. Pigmented villonodular synovitis in children: a report of six cases and review of the literature. Iowa Orthop J 2007;27:90–4.
[15] Coutinho M, Laranjo A, Casanova J. Pigmented villonodular synovitis: a diagnostic challenge. Review of 28 cases. Acta Reumatol Port 2012;37:335–41. 2012.
[16] Xie GP, Jiang N, Liang CX, et al. Pigmented villonodular synovitis: a retrospective multicenter study of 237 cases. PLoS One 2015;10: e0121451.
[17] Sharma V, Cheng EY. Outcomes after excision of pigmented villonodular synovitis of the knee. Clin Orthop Relat Res 2009;467:2852–8.