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Case Report

Pigmented Villonodular Synovitis in a 15-Month-Old Boy Challenged the Physicians for Two Years: A Case Report and Literature Review

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Abstract

Background: Pigmented villonodular synovitis (PVNS) is a very rare pathology of the synovial membrane and even less common in the pediatric group. Rarity leads to misdiagnosis in pre-puberty cases, which may waste significant time for the patient before definite treatment. Reviewing such cases is useful for every pediatrician or orthopedic surgeon to avoid misdiagnosing possible cases.
Case Report: We report a 15-month-old boy who suffered pain and swelling in his knee for about two years before he was eventually diagnosed with PVNS and underwent surgery. Keeping this diagnosis in mind may have saved him and his family from two years of pain, several admissions, and unnecessary prescriptions.
Conclusion: PVNS in pediatrics is rare, but it can occur and be misdiagnosed for diseases such as juvenile rheumatoid arthritis (JRA) and septic arthritis which elongates the pain period. PVNS responds well to subtotal synovectomy, and symptoms are relieved after the surgery if well performed.

Keywords: Case Reports; Pigmented Villonodular Synovitis; Synovial Membrane

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Background

Pigmented villonodular synovitis (PVNS) is a rare pathology originating from the tissues of the bursa membrane, tendon sheath, or synovium. The estimated rate for this entity is 1.8 diagnoses in every one million people. The most common age for the occurrence of PVNS is in the third to fifth decades of age, and it rarely affects the pediatric group (1-5).

To date, no clear etiology has been defined for PVNS but genetic factors, trauma history, and chronic inflammation have been listed as potential predisposing agents.

This low incidence may mislead the patients' diagnosis. Many physicians forget to keep the PVNS diagnosis in mind when encountering a child presenting with joint pain and swelling (3, 4, 6-9). The most common issue in pediatric patients diagnosed with PVNS is the latency in the final diagnosis and delay in the treatment process, which wastes a remarkable amount of time and energy, both for the patient's family and the health care system (2-4, 6-8, 10).

In the current article, we will introduce a patient who has complained of occasional episodes of joint pain and swelling for about two years before final treatment. According to our literature review, so far, only around 50 pediatric PVNS cases have been reported worldwide. The number of patients under two years old is even less than ten (1-9). Reviewing these cases is necessary for every orthopedic surgeon and pediatrician in order not to miss the same cases.

Case Report

The current case is a boy who started to be symptomatic at 15 months of age and has been misdiagnosed and mismanaged for more than 19 months until, eventually, at about three years of age, he was referred to our clinic, diagnosed with PVNS, and scheduled for surgery. Since then, the patient has been symptom-free for 16 months, and no recurrence has been noted.

Very firstly, at 15 months of age, the patient complained of a three-day period of pain and swelling in his right knee joint and was diagnosed with bacterial inflammation. Therefore, he received antibiotic therapy. Symptoms have resolved, but six months later, once again, pain and swelling have made the patient visit a pediatric center. Due to the laboratory data and positive Coombs test, the patient was labeled for brucellosis and received an eight-week course of cotrimoxazole, rifampin, and ibuprofen, but the symptoms did not relieve.

The patient was admitted to a pediatric hospital afterward three times and underwent arthrocentesis and knee magnetic resonance imaging (MRI). Due to positive prepatellar effusion, he was diagnosed with juvenile rheumatoid arthritis (JRA) and received methotrexate and prednisolone for seven months. After seven months of treatment for JRA and the lack of pain relief, the patient was referred to our clinic. According to the MRI, a PVNS diagnosis was made, and the patient was scheduled for synovectomy surgery (Figures 1 and 2).

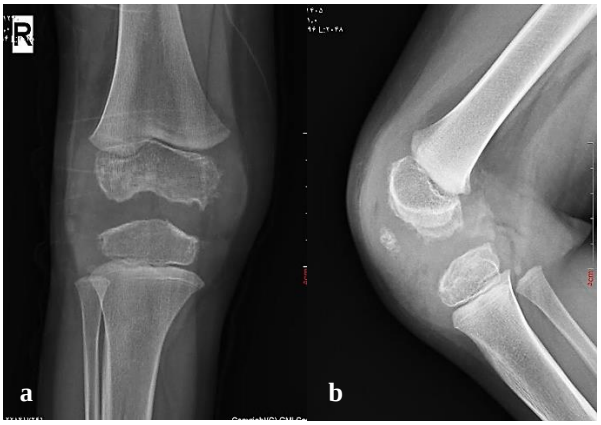

Figure 1. a) Anteroposterior (AP) X-ray of the knee of the patient before surgery; b) Lateral X-ray of the right knee before the surgery

During the surgery session, specimens were sent for histopathologic studies, and PVNS was confirmed. At 16-month follow-up, the patient was symptom-free and experienced no recurrence of symptoms since the surgery.

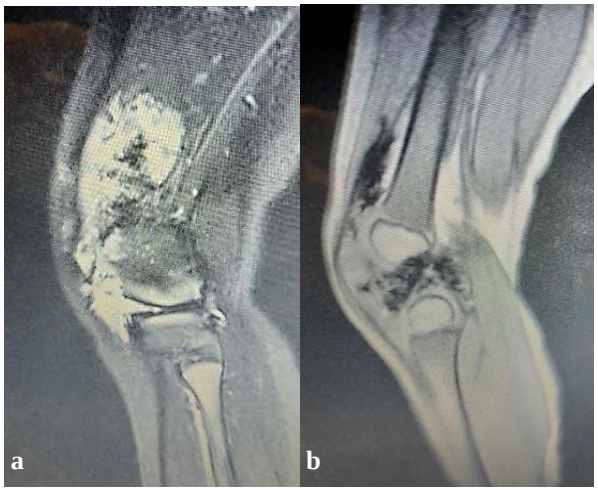

Figure 2. a) Coronal magnetic resonance imaging (MRI) of the patient's right knee before surgery; b) The sagittal cuts of the right knee MRI before surgery

Discussion

One common issue in all reported cases of PVNS in children is misdiagnosis. Therefore, it is necessary for every orthopedic surgeon or pediatrician to consider this possible diagnosis for patients experiencing pain, swelling, and limp, especially in the knee joint.

Due to our literature review, more than 80 percent of all reported pediatric cases with PVNS have experienced pain and swelling (2-4, 6-8, 10-12). The knee joint is the most common site for PVNS in both adolescence and the pre-puberty period. Other large joints, such as the shoulder, hip, and ankle can also be involved. Reported cases are mostly misdiagnosed for bacterial arthritis and JRA. Other misdiagnoses in the literature include tuberculosis (TB), familial Mediterranean fever (FMF), and hemophilic arthropathies (5, 7, 8, 10, 13-16).

JRA is justified as in the early stages of the disease, effusion is witnessed locally or diffusely in the knee and misleads the radiologists. Therefore, considering this alternative is mandatory for every unresponsive case to the common regimen prescribed for JRA, such as immunomodulators. On the other hand, bacterial arthritis, both chronic and acute forms, are common differential diagnoses that may mislead physicians to unnecessary prescriptions and ineffective arthrotomies that obligate ineffective admissions and antibiotic therapy.

The most accurate way to diagnose PVNS is MRI. Usually, plain X-ray imaging is inconclusive, and a definite diagnosis requires an MRI study. Low signal intensity areas in the synovial membrane, which reflect the hemosiderin deposition in this layer, are key to diagnosis (6, 8-10, 14, 17-19).

There are several options for treatment as radiotherapy and open or arthroscopic surgery. As radiotherapy has potential complications in the pediatric group, the most preferred treatment method is surgery and subtotal synovectomy. The cornerstone in the treatment process is removing all the abnormal tissues for pain elimination and avoidance from recurrence (10, 14, 15, 18, 19). We chose open subtotal synovectomy and requested a histopathologic study from the removed tissue, which approved the diagnosis. The patient became painless eventually three days after the surgery and experienced no same symptoms since the surgery.

Conclusion

PVNS, although rare in pediatrics, may lead to a significant waste of time and energy, both for the patient and the health care system. The similarity between symptoms of PVNS and other diagnoses such as JRA and septic arthritis misleads the health care providers and delays the definite treatment.

Reviewing this case and keeping PVNS diagnosis in mind when encountering patients experiencing pain and swelling, especially in the knee joint, benefit future patients and their families from the same story.

PVNS is diagnosed in the pediatric group as well and responds to the same standard protocol used in adolescence. Removing all the affected tissue relieves the patient's pain and limp.

Conflict of Interest

The authors declare no conflict of interest in this study.

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