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Pediatric Pigmented Villonodular Synovitis Mimicking a Septic Hip

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Abstract

We describe a child with pigmented villonodular synovitis initially treated for a presumed hip infection. The correct diagnosis was not made until 2½ years later on a second admission. This is a rare disease with vague presenting symptoms that requires a high index of suspicion. Magnetic resonance imaging and tissue biopsy are usually needed for a definitive diagnosis. Surgery is the primary treatment option; however, the patient described was unusual in that she did well to date with conservative measures.

Keywords

pigmented villonodular synovitis; hip; pediatrics; MRI

Case

A 9-year-old girl presented with a 3-day history of left hip pain. The pain began after she fell on her left side while performing gymnastics. She reported a “pop” associated with sharp pain after the fall. This pain was located in the front of her hip/upper thigh region and did not radiate. The pain was not alleviated with over-the-counter pain relievers and worsened with weight bearing and any movement of the left lower extremity. The pain was described as progressive over 3 days and did not have variation in severity throughout the day. There was no morning stiffness, fever, rash, recent upper respiratory tract infection, or other joint symptoms reported.

Of note, the patient had been treated 2½ years earlier for a presumed left hip infection following a 1-week history of left-sided hip pain, decreased weight bearing, and 1 day of fever. At that time, she underwent fluoroscopic guided joint aspiration of the left hip. The synovial fluid was noted to be bloody with 45,540 red blood cells and 31,680 white blood cells. The cell differential of the synovial fluid showed 93% neutrophils. Due to the suspicion of infectious arthritis the area was surgically irrigated. In the operating room turbid brown fluid was noted at the site. Cultures of the tissue obtained from the site did not show any growth of organisms. Notable laboratory studies included a complete blood count of 12,100 white blood cells/mcL (normal: 4500–14,500), erythrocyte sedimentation rate of

16 mm/h (normal: 0–13), C-reactive protein (CRP) of 0.8 mg/dL (normal: 0–0.74), negative blood and urine cultures. She was treated with 3 weeks of vancomycin with resolution of her symptoms, and she remained symptom free until her presentation 2½ years later.

The patient's physical examination revealed normal muscle bulk and no obvious deformities. There was significant discomfort with extension, flexion, internal, and external rotation of the left hip. Range of motion was limited secondary to pain. There was significant pain with weight bearing and a limp on ambulation. Patrick test and Schober test were limited due to pain. There was no synovial thickening, warmth, effusion, limitation, or tenderness of any other joints.

Notable laboratory studies included a complete blood count with 8900 white blood cells/mL, erythrocyte sedimentation rate of 35 mm/h, and a C-reactive protein of 2.99 mg/L. An x-ray showed a normal right hip and an ossified body near the left hip joint. Magnetic resonance imaging (MRI) was used to further evaluate the region (Fig. 1).

The MRI showed a lobulated, large, intermediate signal intensity mass distending into the left hip joint and filling the entire joint cavity with the presence of moderate joint fluid. There were focal areas of low signal intensity within this mass. Blooming artifacts were seen on the spoiled gradient recalled sequence indicating presence of either calcification and/or hemosiderin. The bone marrow signal intensity was normal and no bony erosions were noticed. The postcontrast sequence showed heterogeneous enhancement of the lobulated mass along with intense enhancement of the synovium which was nodular in appearance. Bone marrow signal intensity was normal. The differential diagnosis at this point included inflammatory arthritis, pigmented villonodular synovitis (PVNS), or synovial chondromatosis.

A left hip tissue biopsy was performed by interventional radiology. The pathology was limited and showed fibroconnective skeletal muscle and synovial fragments rich in foamy macrophages (Fig. 2). An iron stain demonstrated a high hemosiderin content (Fig. 3). Based on signs, symptoms, and diagnostic findings, PVNS was diagnosed. Due to the fact that she was asymptomatic with recovery of range of motion after conservative treatment that included physical therapy and nonsteroidal anti-inflammatory drugs, surgery was not pursued. Her pain decreased over the next few months with full recovery of range of motion. X-rays were repeated 5 months later and showed no erosions. She remains asymptomatic 1 year after the MRI and tissue biopsy, with physical examination showing painless range of motion. Surgical intervention has not been warranted.

Discussion

PVNS is a rare disease caused by proliferation of intra- or extra-articular synovial tissue and presents in 2 forms: diffuse or localized. The synovium can contain hemosiderin, macrophages, and giant cells. The areas affected include the joint, tendon sheath, or bursa. Localized PVNS occurs when a single mass is present in the synovium, involving a smaller section. In most cases the entire synovium is effected and this represents diffuse disease.¹

There is controversy as to whether PVNS is inflammatory or neoplastic. Some studies support hemarthrosis or repetitive trauma as a cause, however, this does not explain recurrence after treatment. Clonal abnormalities have been found pointing to a neoplastic process though uncertainty remains as to the significance of these findings. Previous reports show both a monoclonal type cell proliferation and a polyclonal origin supporting both a neoplastic process and a reactive process.^{1–3} A recent finding of a chromosomal translocation at 1p13 has linked overexpression of macrophage colony stimulating factor 1 and its receptor in the pathogenesis.⁴

This disease has a peak incidence in the fourth and fifth decade of life, can occur at any age, and is less common in childhood. It can affect both sexes and almost always involves a single joint. Large joints are usually involved, most commonly the knee.¹ In one study of 250 patients, the hip was affected in 3.6% of individuals.⁵ The second or third most common joint varies by report as either hip, shoulder, ankle, and finger involvement.^{1,2,6} Synovial tissue of any joint can be affected and even axial skeleton involvement has been reported.⁷ The overall incidence is about 1.8 cases per million people per year.⁸

The presentation can be vague and one must have a high index of suspicion to make the diagnosis of PVNS. A history of trauma is reported only occasionally. Based on the region that is involved there can be local pain and range of motion limitation. Warmth and swelling of joints mimics arthritis. The symptoms can be intermittent as there are exacerbations and symptom-free periods.^{1,2}

Many different radiologic imaging methods have been used in diagnosing PVNS. These include x-ray, arthrography, bone scan, ultrasound, computed tomography (CT), and MRI.² X-rays can be normal or show erosions in chronic disease.⁸ Arthrography will show filling defects and is not widely used due to the availability of MRI. Bone scans have proved to be nonspecific. Ultrasound is technician dependent thus limited, however, it can be useful in locating and differentiating PVNS from other conditions. The high iron content in lesions is easily seen as a high density mass with CT and the synovial proliferation enhances with contrast. CT is limited by reduced sensitivity for inflamed tissue and poor resolution of soft tissue.^{1,2}

MRI has proved to be the most useful imaging modality. A low signal on T1- and T2-weighted images can be seen due to the hemosiderin. Hyperplastic synovium represented as a mass as well as bone erosions are visible at times. Lipid content, congested synovium, and effusion are seen as high signal intensity. A contrast study is recommended as the lesions will enhance.^{1,2,8} Review of MRI imaging modalities has shown that gradient-echo imaging and enhanced imaging in pediatric patients is valuable in diagnosing the extent of disease.⁹

MRI findings are not pathognomonic as other diseases such as synovial chondromatosis, monoarthritis, fibromatosis, tumor, amyloidosis, and hemophilia are in the differential.^{1,2} In confirming the suspected diagnosis tissue biopsy should be obtained. A joint aspiration will yield brown or blood stained fluid. Biopsy of the PVNS lesion will reveal macrophages, multinucleated foreign body giant cells with yellow-brown hemosiderin pigment.^{8,10}

Treatment of this disease is important, however, recurrence after intervention can occur. The overall recurrence of diffuse PVNS is reported in 10% to 56% of cases.¹ A meta-analysis of 53 patients with only hip involvement showed that 19% had recurrence after treatment.⁵ The average time for recurrence is around 5 years with increasing risk with time.³ Data on pediatric patients are limited due to the rarity of disease.

The cornerstone of PVNS treatment is surgery. For localized disease arthroscopic excision of the lesion is possible and yields low recurrence rates. For diffuse disease a total synovectomy is indicated as clinical studies have shown higher recurrence from incomplete synovectomy. A total synovectomy can be performed arthroscopically or as an open procedure. It is thought that recurrence is less with open procedures; however, postoperative morbidity is higher. Arthroscopic surgery is more complex in joints other than the knee such as the shoulder and hip.^{1,11,12} There are no stringent guidelines regarding the hip, however, most feel that a total synovectomy is warranted. If there is any damage to the joint cartilage then the synovectomy is combined with total hip arthroplasty or arthrodesis.⁵

Radiotherapy has been used for diffuse PVNS but with caution as malignant transformation of the lesion has been reported.¹ The current recommendation is that radiotherapy be used as an adjunct to total synovectomy to prevent recurrence when there is remaining tumor. It is also mainly used in recurrent cases. In one study 40 of 41 patients who received a mean radiotherapy dose of 38 Gy maintained control of disease for an average follow-up time of 77 months. The recommendation for residual disease is a moderate dose of radiation after complete removal of PVNS.¹³

The use of radioactive isotopes such as yttrium 90 for PVNS showed some promising results. There are risks of local and systemic spread of radiocolloids which can result in chromosomal damage and infertility.¹⁴ A recent article suggested the use of the tumor necrosis factor blocking agent, infliximab, for PVNS; however, there is limited experience.¹⁵

In the patient discussed herein, in retrospect the initial presentation was most likely related to PVNS and not infection. Her tissue culture grew no bacteria and the synovial fluid was noted to be bloody, which is consistent with the diagnosis of PVNS. Two and a half years later her symptoms were similar; infection was suspected but MRI and tissue biopsy studies revealed PVNS. In a group of 55 patients with PVNS it took an average of 4.5 years before a histologic diagnosis was found.¹⁵

The patient described in this case did not undergo surgery and only participated in physical therapy with improvement in her symptoms. Other conservative approaches include ambulation assistive devices and nonsteroidal anti-inflammatory drugs. Although unusual, there are reported cases of PVNS being treated conservatively and resolving without the use of surgical measures.¹⁰ The patient is followed clinically and with radiography in the anticipation of surgical treatment.

Conclusions

Pediatric PVNS is a rare but important cause of joint pain as the symptoms are vague and can mimic more common diseases. The diagnostic test with the most yields is a contrast-enhanced MRI as it shows a typical heterogeneous signal appearance. A tissue biopsy is needed for a definitive diagnosis and will demonstrate giant cells, macrophages, and hemosiderin pigment. There are many treatment options with complete synovectomy being the current optimal choice. Tumor necrosis factor blocking agents and other novel therapies may play a role as the pathogenesis is further investigated.

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Figure 1.

(Left) Coronal magnetic resonance image using short T1 inversion recovery demonstrates mass, synovial enhancement, and low signal intensity (arrows). (Right) Axial T1 magnetic resonance image of pelvis using spin echo and fat saturation demonstrates lesion distending the left hip with low signal intensity (arrows).

Figure 2.
(Left) Hematoxylin-eosin stain, 40×. Resected tissue composed predominantly of numerous foamy macrophages and rare pigment-laden macrophages. (Right) CD68 immunostain, 40×. Abundance of macrophages highlighted by a macrophage immunostain.

Figure 3. Prussian blue iron stain, 40 $\times$. Synovial tissue with grainy hemosiderin stain in superficial tissue.