Arthroscopic Management of Pigmented Villonodular Synovitis of the Hip in Children and Adolescents

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Background: Pigmented villonodular synovitis (PVNS) is a benign proliferative synovial disorder most commonly described to affect the knee in adults. Literature describing PVNS in the pediatric population is limited to 2 small case series and a handful of single-patient case reports. Within these studies, only 2 patients with PVNS of the hip are described.

Purpose: To describe the presentation, management, and outcomes of a single-center series of pediatric patients with PVNS of the hip treated with arthroscopic synovectomy.

Study Design: Case series; Level of evidence, 4.

Methods: A retrospective review of consecutive pediatric patients treated for PVNS at a single institution was performed. Inclusion criteria consisted of patients younger than 19 years with surgically treated PVNS of the hip.

Results: Five pediatric patients with a mean age of 11.0 years were treated for PVNS of the hip from 2011 to 2016. The mean duration of symptoms from onset to surgical treatment was 247 days (range, 3-933 days). Upon review of magnetic resonance imaging (MRI) results, radiologists included PVNS in their differential in 3 patients. Seven surgeries were performed in 5 patients. All therapeutic procedures were arthroscopic synovectomies. Nodular PVNS was present in 4 patients, and diffuse disease was present in 1 patient. At a mean 32-month follow-up (range, 12-63 months), all patients were considered to be free of recurrence based on clinical examination and/or follow-up MRI. Four patients were asymptomatic and returned to all of their previous sports activities.

Conclusion: Young age at the time of diagnosis is a point to be highlighted in this cohort, and symptoms may be present for many months prior to diagnosis due to the failure to consider PVNS in children. Therefore, for patients with "atypical" presentations or lack of improvement with treatment for rheumatologic, bleeding, or infectious disorders, PVNS should be strongly considered. MRI with gradient echo sequences is the diagnostic imaging study of choice. One patient with diffuse involvement and preoperative degenerative changes showed progressive changes postoperatively. This type of PVNS may have a worse prognosis, but more diffuse cases are needed before the prognosis can be determined. Arthroscopic synovectomy following a timely diagnosis of PVNS produces good outcomes in nodular cases, with no evidence of symptomatic or radiographic disease persistence among these patients.

Keywords: hip arthroscopy; synovectomy; PVNS

Pigmented villonodular synovitis (PVNS) is a benign proliferative synovial disorder characterized by hemosiderin-laden villous and nodular lesions.[11] First described by Jaffe et al[11] in 1941, PVNS has historically been described as a monoarticular disease in adult patients aged 20 to 45 years, affecting men and women equally. The estimated annual incidence is 1.8 patients per million population.[18] The typical patient presents with an insidious onset of swelling and pain localized to a single joint, most commonly the knee. The cause of PVNS is unclear, with postulated mechanisms ranging from inflammatory[8] to neoplastic.[21]

Magnetic resonance imaging (MRI) findings include hyperplastic synovium with nodular or diffuse proliferations that have low signal on T2-weighted and proton density–weighted images. This signal dropout, related to the

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TABLE 1
Study Group[a]

Patient No.	Age at Surgery, y	Sex	Preoperative Duration of Symptoms, d	PVNS Type	Index Surgery	Additional Surgery	Duration of Follow-up, mo	Recurrence
1	15	F	933	Nodular	Excisional biopsy and arthroscopic synovectomy	—	12	No
2	12	F	131	Diffuse	Excisional biopsy and arthroscopic synovectomy, chondroplasty, microfracture	—	53	No
3	4	M	38	Nodular	Excisional biopsy and arthroscopic synovectomy	—	17	No
4	17	F	130	Nodular	Excisional biopsy and arthroscopic synovectomy, labral repair, femoroplasty, removal of hardware	—	14	No
5	7	M	3	Nodular	Diagnostic arthrocentesis prior to PVNS diagnosis	Excisional biopsy and arthroscopic synovectomy	63	No

[a]F, female; M, male; PVNS, pigmented villonodular synovitis. Dashes (—) indicate no additional surgery.

paramagnetic effect of hemosiderin, is most prominent on T2-weighted gradient echo sequences.[6,15] A large joint effusion may also be present. Biopsy is the gold standard for diagnosis and reveals hemosiderin-laden macrophages, vascularized villi, mononuclear cell infiltration, and sporadic mitotic figures.[16]

The knee is by far the most common site of involvement in PVNS, followed second by the hip. Large retrospective reviews describe PVNS as affecting the hip in 3% to 18% of their total cohorts.[4,13,18] Historically, multiple treatment strategies have been used for PVNS of large joints, including synovectomy (open and arthroscopic), arthrodesis, irradiation, and arthroplasty.

Literature describing PVNS in the pediatric population is limited to 2 small case series[1,19] and a handful of single-patient case reports.[5,9,10,12,20,22,24,26] Within these studies, only 2 patients with PVNS of the hip are described.[9,20] In addition, Byrd et al[3] described a case series of 13 patients (ranging in age from 14 to 46 years) with PVNS of the hip, but the majority of patients were adults. The purpose of the current study was to describe the presentation, management, and outcomes of a single-center series of pediatric and adolescent patients with PVNS of the hip treated with arthroscopic synovectomy.

METHODS

All patients diagnosed and treated with PVNS between January 2011 and December 2016 at Children's Healthcare of Atlanta at Scottish Rite were retrospectively identified in this institutional review board–approved study. Patients were identified by International Classification of Diseases 9th Revision (ICD-9), ICD 10th Revision (ICD-10), and Current Procedural Terminology (CPT) codes as well as by keyword search of radiology and pathology reports. Inclusion criteria were patients younger than 19 years with a histologic diagnosis of PVNS of the hip treated surgically. Patients with PVNS of joints other than the hip were excluded. Chart review, including clinical, imaging, and operative reports, was performed for all patients to determine age at the time of presentation, joint involvement, duration and description of symptoms, previous diagnoses, physical examination findings, MRI characteristics, surgical intervention, histologic diagnosis, clinical follow-up, and disease persistence and/or recurrence. The primary outcome was disease persistence and/or recurrence.

RESULTS

A total of 18 pediatric patients with a diagnosis of PVNS were identified. Five of these patients had PVNS of the hip and are described in Table 1. Patients with PVNS of other joints were excluded, including the knee (n = 11), shoulder (n = 1), and elbow (n = 1). The mean age at first symptoms was 11.0 years (range, 4-17 years). The 5 included patients were 3 females and 2 males. The left hip was affected in 3 patients and the right in 2 patients. Two patients described a traumatic origin leading to the onset of symptoms. One patient presented acutely to the emergency department with a presentation mimicking septic arthritis. All patients described groin and anterior hip pain as their primary symptom. Two patients reported difficulty with weight bearing. Physical examination was similar in all patients, with painful range of motion and positive anterior impingement testing. The mean duration of symptoms in the affected joint prior to surgical treatment was 247 days (range, 3-933 days).

In no patient was the initial working diagnosis PVNS. The diagnoses made prior to the diagnosis of PVNS included transient synovitis (n = 2), femoroacetabular

impingement (n = 1), femoral neck stress fracture (n = 1), and a bleeding disorder (n = 1).

Plain radiographs and MRI were the most commonly used diagnostic studies in the workup of localized joint pain. Figure 1 provides select preoperative and postoperative radiographs and MRI sequences in a patient with PVNS of the hip. Initial radiographs were normal in 3 patients. Of the 2 patients with abnormal radiographs, 1 patient had joint space narrowing with sclerosis and calcified loose bodies and the other patient had in situ screw fixation of a slipped capital femoral epiphysis 5 years previously. Preoperative imaging, including MRI in 4 patients and computed tomography (CT) in 1 patient, was available for review. The CT was obtained in the patient with previous screw fixation of a slipped capital femoral epiphysis as preoperative planning for hip arthroscopy for femoroacetabular impingement. In this patient, PVNS was an incidental finding at the time of hip arthroscopy. Radiologic interpretation of these advanced imaging studies included PVNS in the differential in 3 patients. Other differential diagnoses based on advanced imaging included a hemorrhagic effusion (n = 2), synovial chondromatosis (n = 1), and nonspecific effusion (n = 1).

A total of 7 surgeries were performed on 5 patients. Four patients underwent a single surgery. The patient who initially presented to the emergency department with an examination mimicking septic arthritis had an MRI consistent with an effusion; this resulted in an arthrocentesis of the hip under anesthesia that yielded a large hemarthrosis. Repeat arthrocentesis with steroid injection was performed for pain control while a hematologic workup was pursued. Ultimately, a repeat MRI at 3 months following the first scan showed evidence for nodular PVNS, and an arthroscopic synovectomy was performed. Of the 4 patients treated with a single surgery, all were arthroscopic synovectomies. In addition to having a synovectomy, 1 patient had a chondroplasty and microfracture of the femoral head for chondral injury secondary to chronic diffuse PVNS resulting in degenerative changes. Another patient had treatment of hip impingement with acetabular rim trim, labral repair, and femoroplasty prior to nodular PVNS being identified during the peripheral compartment examination. Histologic diagnosis was consistent with PVNS in all 5 patients. Arthroscopic evaluation revealed nodular disease in 4 patients, all within the peripheral compartment, and diffuse disease in 1 patient.

At a mean follow-up of 32 months (range, 12-63 months), 4 patients were asymptomatic and had returned to their regular activities. One patient had decreased range of motion with anterior impingement symptoms but minimal pain with daily activities. This patient was noted to have diffuse PVNS at the time of arthroscopy and had degenerative changes and joint space narrowing noted on preoperative radiographs. In this patient, follow-up postsurgical imaging over the course of more than 4 years demonstrated progressive joint space narrowing and a noncongruent femoral head. Postoperative MRI was performed in 3 patients and showed no evidence for residual or recurrent PVNS. Two patients did not undergo postoperative MRI. One of these patients was asymptomatic at 1-year follow-up, and family and physician preference was to defer repeat imaging. The other was the patient with diffuse PVNS, and repeat MRI was not believed to be beneficial in the setting of advanced degenerative changes.

DISCUSSION

PVNS of the hip is a well-described entity in the adult population, but prior to this study it had rarely been reported in children and adolescents. In this study, we described the presentation, surgical management, and outcomes of a single-center series of 5 children and adolescents with PVNS of the hip treated with arthroscopic synovectomy.

Only 2 series of pediatric patients with PVNS have previously been published.[1,19] In neither study did any of the patients have PVNS of the hip. In 2010, Baroni et al[1] published a retrospective case series of 9 patients younger than 16 years with PVNS of the knee. At the time of presentation, the mean age was 9.8 years (range, 2-15 years) and symptom duration was a mean of 18 months. In nearly half of the patients, joint pain had been misdiagnosed as a rheumatologic disorder (juvenile idiopathic arthritis), and those patients had a mean 34-month delay to diagnosis. Synovectomy (5 open and 4 arthroscopic) was performed in all patients, and the authors reported no recurrences at a mean follow-up of 8.5 years.

The second pediatric case series was published by Neubauer et al[19] in 2007 and included 6 patients aged 7 to 15 years. Five of the patients had PVNS of the knee, while 1 patient had PVNS of the hand. Similar to the findings by Baroni et al,[1] the majority of patients were misdiagnosed as having rheumatologic disorders, with a delay in diagnoses of more than 1 year. The diagnosis of PVNS had been considered in only 1 patient prior to surgery. Following synovectomy, recurrence was reported in 2 of the 6 patients.

Although the knee has historically been described as the most common location of PVNS, pediatric case reports suggest that any joint can be involved. In addition to noting PVNS of the knee,[2,10,22] authors have described PVNS in the sacroiliac joint,[12] elbow,[24] hip,[9,20] and foot.[5] A more recent case report described a 7-year-old child with 2 years of hip pain and radiographs showing extensive bony erosive changes in the femoral head.[9] This patient underwent open synovectomy, followed by repeat open synovectomy for recurrence 2 years later. Follow-up radiographs showed narrowing of the joint space and early closure of the proximal femoral physis. Similar to the presentation of one of our patients is a case report of a 9-year-old child with an acute onset of hip pain that was treated 2 years prior to representation for presumed septic arthritis of the hip despite negative cultures.[20] MRI showed a large mass in the hip joint with blooming artifacts, and biopsy by interventional radiology was diagnostic of PVNS. At 1-year follow-up, the patient had complete resolution of symptoms with nonsteroidal anti-inflammatory medications and physical therapy, and the family elected to continue nonoperative treatment.

The most important point illustrated by our study, combined with these small pediatric case series, is the lengthy

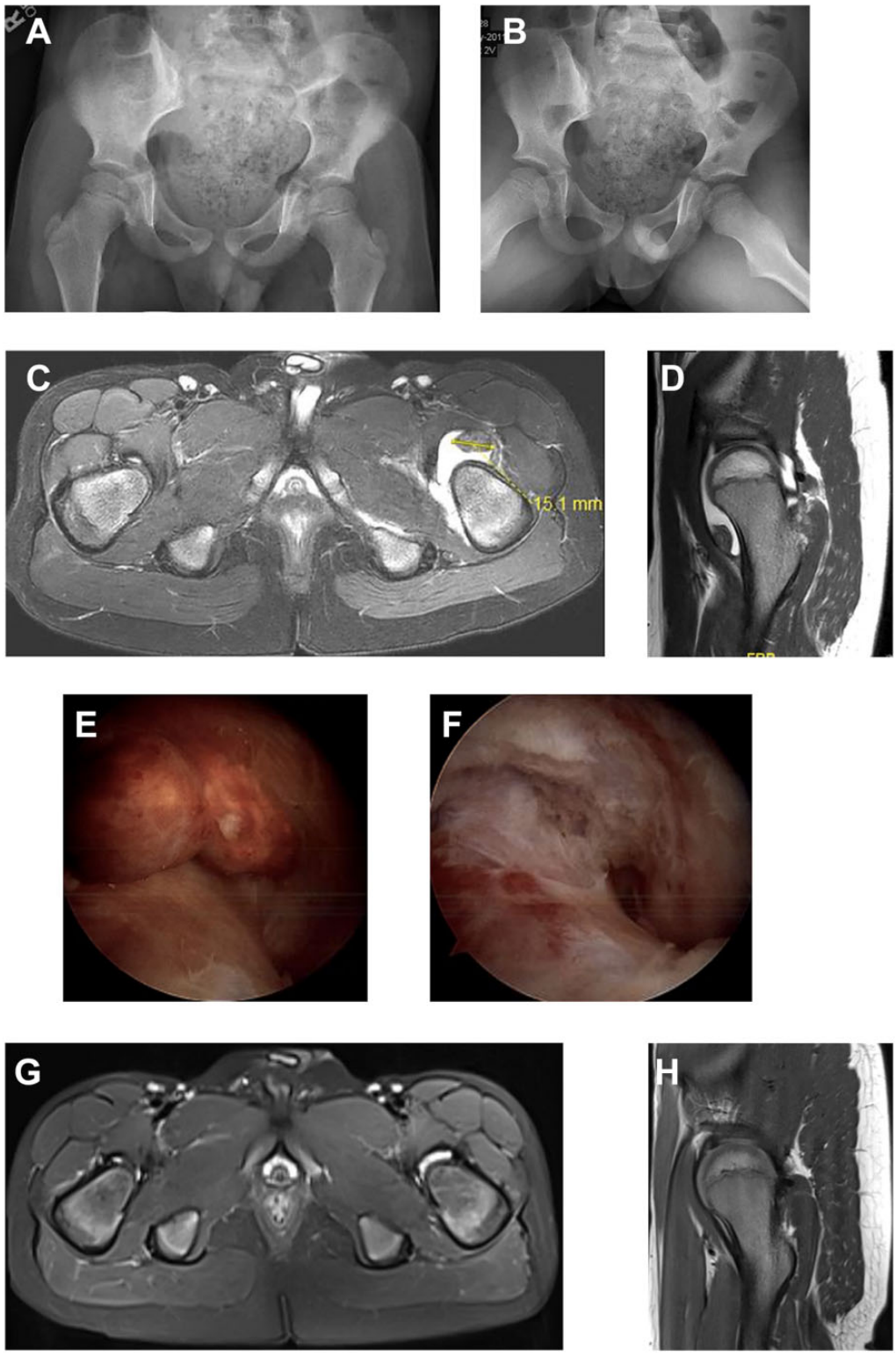

Figure 1. Pigmented villonodular synovitis (PVNS) of the left hip in a 4-year-old male. Preoperative (A) anteroposterior and (B) frog-leg lateral radiographs of the pelvis with evidence of effusion. (C) Preoperative T2 axial and (D) sagittal magnetic resonance imaging (MRI) sequences with joint effusion and hypointense 1.5-cm mass in the anterior recess. Intraoperative arthroscopic images of the peripheral compartment PVNS nodule (E) prior to and (F) after resection. Postoperative (G) T2 axial and (H) proton-density sagittal MRI sequences 6 months after surgery, with no effusion and interval excision of mass.

delay in diagnosis of PVNS in young patients. Children with an acute onset of swelling, pain, and refusal to bear weight frequently are subject to workup for septic arthritis, transient synovitis, or a bleeding disorder. More chronic joint symptoms receive a rheumatologic diagnosis, frequently juvenile idiopathic arthritis, with prolonged pharmacotherapy with minimal or transient symptomatic improvement. PVNS must be included in the differential of children and adolescents with acute or chronic hip pain that does not have a clear infectious origin or has failed to respond appropriately to pharmacotherapy and physical therapy. MRI with gradient echo sequences should be performed early to evaluate for this disease process. Failure to consider PVNS as a differential diagnosis may result in a delayed diagnosis, which may lead to progressive functional impairment and degenerative changes in the affected joint.

With respect to treatment of PVNS of the hip using joint-preserving surgeries, the literature describes open,[4,7,25] mini-open,[7] and arthroscopic[3,13,17] approaches. Some authors[3,7,13,17,23,25] advocate that mini-open or open procedures allow for more complete excision and potentially lower rates of recurrence. In our series, nodular PVNS of the hip was found most often in the medial recess of the peripheral compartment. Therefore, careful attention and arthroscopic skills are necessary to visualize both the central and peripheral compartments of the hip. Additionally, we believe that the arthroscopic approach may yield lower morbidity in the setting of revision surgery in select patients with disease persistence.

If performing hip arthroscopy, the surgeon must have complete central and peripheral compartment visualization, which requires a 70° arthroscope, flexible instruments such as electrothermal probes, multiple portals, and a dynamic examination to evaluate the most medial and lateral aspects of the peripheral compartment. Some authors have described specific arthroscopic techniques to aid in visualization of the central and peripheral compartments, including extension of the capsulotomy length[13] and t-capsulotomy.[17] Surgeons should be facile with these techniques prior to undertaking hip arthroscopy. No studies have directly compared outcomes of arthroscopic versus open approaches. It seems that surgical approach should be dictated by surgeon experience and skill with open versus arthroscopic techniques.

The only series of patients with PVNS treated exclusively with arthroscopic hip synovectomy is a cohort of 13 consecutive patients with a mean age of 26.8 years reported by Byrd et al.[3] Disease type included nodular in 3 patients, diffuse in 3 patients, and mixed in 7 patients. At a mean follow-up of 63 months, all patients had significant improvement in their Harris hip score, and none had postoperative complications. One patient had evidence of residual PVNS on follow-up MRI, and 1 patient required total hip arthroplasty 6 years following the initial procedure because of progressive osteoarthritis. Byrd highlighted the importance of complete peripheral compartment examination, as arthroscopy of the central compartment alone resulted in failure to diagnose PVNS in 2 patients referred to him after their initial procedure.

Synovectomy combined with arthroplasty is described in adult patients with good outcomes and lower recurrence than synovectomy alone.[4] In contrast, a recent systematic review performed by Levy et al[14] found no difference in recurrence after synovectomy versus synovectomy plus arthroplasty for the treatment of PVNS in adults. We believe that in the pediatric patient, synovectomy in isolation results in good outcomes with low rates of recurrence. Furthermore, repeat arthroscopic synovectomy, if necessary for recurrent or persistent disease, is a viable and low-risk option in the pediatric and adolescent patient. In contrast, significant concerns arise regarding the need for revision arthroplasty when it is performed as the primary treatment for PVNS in a young patient. Furthermore, arthroplasty is not a viable surgical option for young children with significant growth remaining.

We acknowledge several limitations of this study, the most significant of which is low power related to a small number of patients. However, considering that the yearly incidence of PVNS is 1.8 cases per million and that PVNS of the hip represents a very small number of these cases, larger studies are difficult to perform. Of the total 18 pediatric patients with PVNS treated at our center over 6 years, 5 (28%) involved the hip. This small number of patients makes it difficult to draw conclusions regarding rates of persistence and/or recurrence. A second limitation is the heterogeneity of diagnostic imaging, treatment, and follow-up that is inherent in retrospective studies. Although some physicians obtain serial postoperative MRIs to evaluate for persistence or recurrence, other physicians obtain advanced imaging only in the setting of new or progressive symptoms.

CONCLUSION

This case series of children and adolescents represents a single center's experience with the treatment of PVNS of the hip with arthroscopic synovectomy. Although the adult literature suggests that PVNS is most commonly identified in the second decade of life,[3,4] age alone should not exclude PVNS from the differential diagnosis in children. PVNS should be considered in pediatric patients with an insidious onset of atraumatic hip pain with laboratory studies and imaging that do not support infectious or inflammatory origin. Symptoms may be present for months to years prior to diagnosis, and patients are frequently misdiagnosed with rheumatologic disorders. MRI with gradient echo sequences is the diagnostic imaging modality of choice. The 1 patient in our study with diffuse involvement and preoperative degenerative changes showed progressive degeneration postoperatively, as expected given the degree of articular cartilage injury present diffusely throughout the joint. This type of PVNS may have a worse prognosis, but more diffuse cases are needed before the prognosis can be determined. In patients with a timely diagnosis of nodular PVNS, hip arthroscopy with examination of the central and peripheral compartments, nodule excision, and synovectomy

produces good outcomes with no evidence for recurrence among the nodular cases in our small series.

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