

Case Report

Pigmented villonodular synovitis combined with developmental hip dysplasia: a case report

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Abstract: Hip dislocation secondary to developmental dysplasia of the hip (Crowe type IV DDH) in adults is a debilitating condition, and associated with increased postoperative complication rate. Pigmented Villonodular Synovitis (PVNS) is an uncommon benign but locally aggressive disease, which mostly affects the large, synovium-lined joints. PVNS of hip joint accounts for 15% of all PVNS cases. But rarely, it may extend beyond the articular capsule and present as a periarticular mass. Here, we report a 67-year-old woman, who has PVNS 50 years after she was diagnosed with Crowe type IV DDH, complained of acute and persistent pain with a buttock mass coming from the right hip. Her initial presentation, diagnostic workup, and treatment will be discussed, along with a brief review of the literature.

Keywords: Developmental hip dysplasia, pigmented villonodular synovitis, total hip arthroplasty, synovectomy

Introduction

First named by Jaffein 1941, Pigmented Villonodular Synovitis (PVNS), mostly affecting large joint, is a relatively uncommon benign disease, which is characterized by pigmentation with villous transformation of synovial lining. Myers and Masi estimated that the overall prevalence of PVNS is 1.8 per million people, and that the hip accounting for 15% cases [1, 2]. However, there are no data in the literature of any patient with developmental hip dysplasia (DDH) and PVNS. Here, we present a 67-year-old female patient with coincidental presence of Crown type IV DDH and PVNS.

Case report

A 67-year-old woman diagnosed developmental dysplasia of the right hip about fifty years ago presented to our department of orthopedics on May 18, 2016, complained of acute and persistent right hip pain and difficulty with activities of daily living for 20 days. Due to her poor financial status, she has walked with a crutch for fifty years, without any regular treatment for her hip. Now, despite conservative treatment with physical therapy and 60 mg of eloricoxib twice a day, she still suffered disabling pain and limitation in daily life.

Physical examination revealed a nontender swelling in the right gluteal region with a soft-to-firm density (**Figure 1**). An area of the local skin was covered with some red spots induced by the Chinese herbs plaster, there was no pain or itch in this area, no distal neurovascular deficit was found either. All movement of the right hip was painful and she could not stand still without axillary crutch. Significant loss of range of motion (ROM) of her right hip was revealed (flexion 60°, extension 0°, internal rotation 0°, external rotation 10°, abduction 10°, adduction 5°). When in the upright posture, she presented a mild tilting of the pelvis. In addition, the patient had apparent shortening of the affected extremity with a leg length discrepancy of 3.5 cm.

All laboratory examination results were in the normal range, except for an elevated sedimentation rate of 40 mm/hour (0-20 mm/hour, normal range), and an elevated C-reactive protein of 18.4 mg/L (0-8.0 mg/L, normal range).

Plain radiographs showed a hip dislocation secondary to developmental dysplasia of the right hip (Crowe type IV DDH), with some cystic lesions of the femoral neck and dysplasia of the lesser trochanter (**Figure 2A**). Considering the



Figure 1. Preoperative photo. A buttock mass in the right hip, with some red spots on the skin.

soft tissue mass in the right hip, a magnetic resonance imaging (MRI) of the pelvic was conducted. Both T1-weighted images (T1WI, **Figure 2C**) and T2-weighted images (T2WI, **Figure 2D**) showed a spotty low signal, well-circumscribed, proliferative, nodular lesions around the femoral head. However, the mass showed intermediate high signal intensity in T2WI images.

Based on the medical history, physical examination, laboratory studies, and imaging evidence, we thought that the first diagnosis was DDH for sure. However, the lesion around the femoral head should be differentiated from PVNS, chronic infection, inflammation, and soft tissue tumors. A biopsy is needed for definitive diagnosis. After fully explanation to the patient and got her consent, she received a surgery treatment using the posterolateral hip joint approach on May 21, 2016. After incising the fascia lata, a mass of brown soft tissue emerged. Diffuse growth of multiple brown nodular lesions and blood effusion were found inside the articular capsule, severely eroding the femoral head, neck and true acetabula [Supplementary Figure 1](#). The frozen-section test revealed synovial nodular proliferation suggestive of PVNS. An extensive synovectomy followed by sub-trochanteric osteotomy and total hip arthroplasty were performed by our team, using a cemented acetabulum (Link, Germany) and a modular cement-less femoral stem (Link, Germany). About 1 cm of limb-lengthening of the affected side was found after installing the prosthesis. Fortunately, the sciatic nerve showed no signs of tension with the knee in flexion or extension. Postoperative pathologic examination confirmed the PVNS diagnosis,

based on the variable mixture of giant cells, hemosiderin, and brown pigmentation in the synoviocyte cytoplasm (**Figure 3**). Postoperative plain radiographs of the pelvic showed the reduction of hip dislocation (**Figure 2B**). She was discharged at 5 days postoperatively without pain and complication.

Discussion

Pigmented Villonodular synovitis (PVNS) is a benign, slowly progressive lesion of unclear etiology, characterized by villous or nodular proliferation of any synovial lined structures, with pigmentation or hemosiderin deposits. There are two types of PVNS: localized and diffuse, which are histologically identical to giant cell tumor of the tendon sheath.

Early diagnosis of PVN is necessary because high percentage of bony erosion and joint destruction will occur with no appropriate treatment. Patients with PVNS commonly presented as insidious pain, local swelling and activity limitation of affected joint. Imaging features of PVNS have been well documented in literature. Soft tissue swelling around the joint, cortical erosions with cystic degeneration in the acetabulum, femoral head and neck, are often featured in plain radiographs [3]. Magnetic resonance imaging (MRI) is the most sensitive and precise modality for diagnosing PVNS, which is characterized by joint effusion, nodular synovial proliferation, bony erosions and bulging of soft tissues. Hemosiderin deposited on proliferative synovium results in a spotty low or extensive low signal area within the proliferative synovial masses on T1- and T2-weighted images. Significant gadolinium enhancement can be seen on T1-weighted images, but this finding is not specific [4, 5].

Literature reported various potential causative factors of PVNS, including surgery, trauma, neoplasia, hemorrhagic effusion, disorders of lip metabolism and chronic inflammation [6]. However, the true etiology of PVNS remains unknown. Our patient has suffered the Crowe type IV hip dysplasia for 50 years, which is a major cause of secondary hip osteoarthritis (OA). Besides, some sub-clinical chronic bleeding episodes caused by her daily activities may lead to blood retention in her hip joint capsule. Given this condition, we postulated that a prolif-

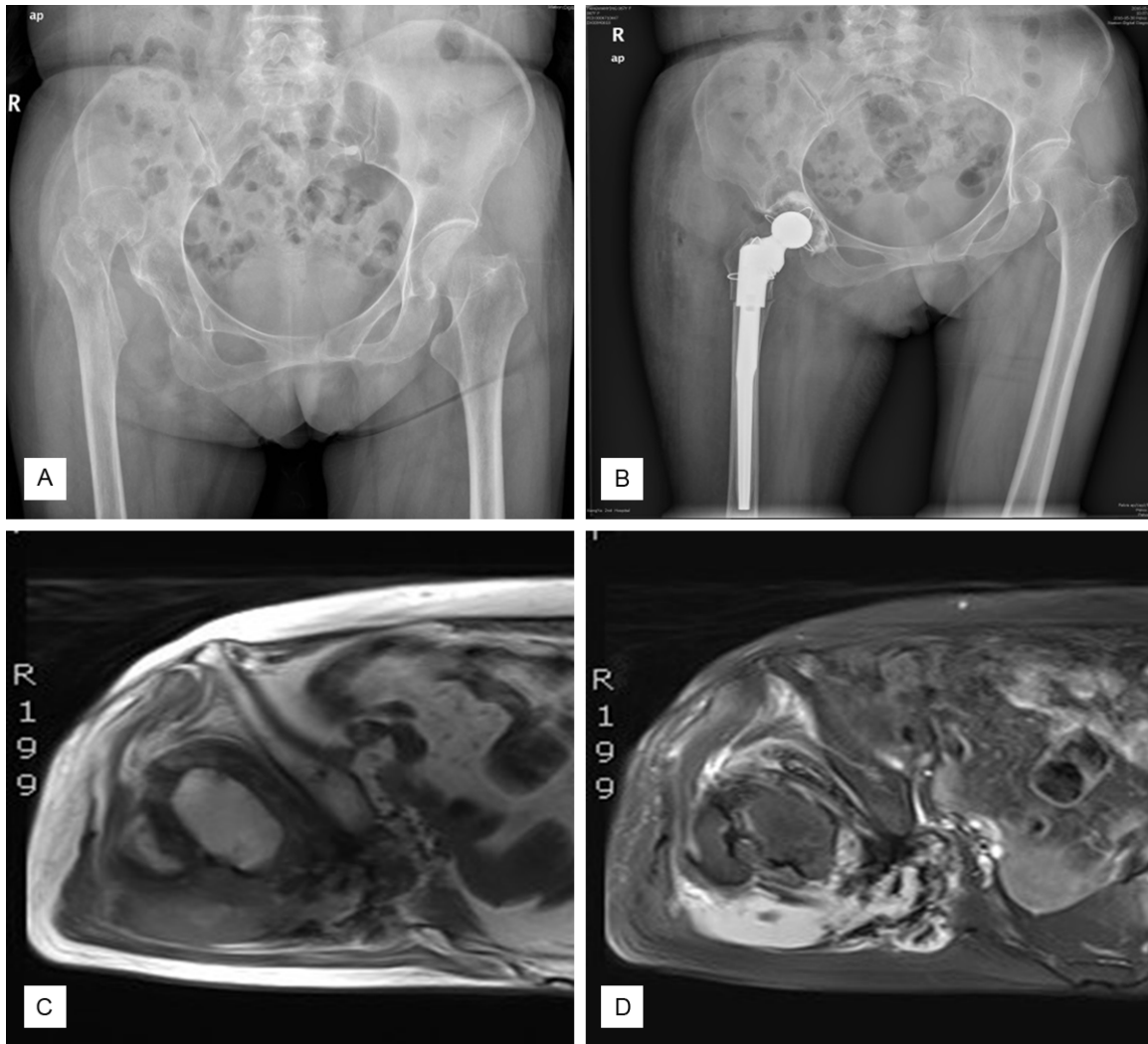


Figure 2. Preoperative and postoperative imaging data. A. Pre-op pelvic plain radiography shows a complete posterior dislocation of the right hip joint secondary to hip dysplasia. B. Post-op pelvic plain radiography shows reduction and reconstruction of the right hip. C. Axial T1-weighted MRI. D. Axial T2-weighted MRI shows an slightly low single intensity and an intermediate hyperintense signal mass lesion located posterior to the hip joint, respectively. Both of them show a spotty low signal, proliferative, nodular lesions around the femoral head.

erative synovitis may have occurred in response to osteoarthritis or the chronic inflammation it induced, and that the hemosiderin deposition within the synovial tissue may result from the chronic bleeding episodes. This observation is supported by a previous report [7].

PVNS is typically a monoarticular process which mostly involves large joints like knee, hip, ankle, shoulder and elbow [1]. Rovner reported a rare case of PVNS occurring in zygapophyseal [8]. It often appears in the third and fourth decades of life. However, several reports have shown that PVNS can also be a threat to children [9, 10]. Normally, the proliferative synovial lesion is confined to joint capsule. However, some-

times it may extend beyond it and present as an extra-articular soft tissue mass, mimicking soft tissue sarcoma, invading the pelvis or the thigh and impinging on the sciatic or femoral nerves [11, 12]. Kyung S. first described a PVNS of the hip presented as a buttock mass [13]. They thought that rapidly proliferating synovial tissue increases intra-articular pressure and that joint capsules already weakened by the disease process give way to the synovium. It was a valuable view, but in our case, we believed that the long-time posterior dislocation of her right hip was the primary cause of weakened articular capsule which led to proliferative synovium extension to form a mass beneath the fascia lata.

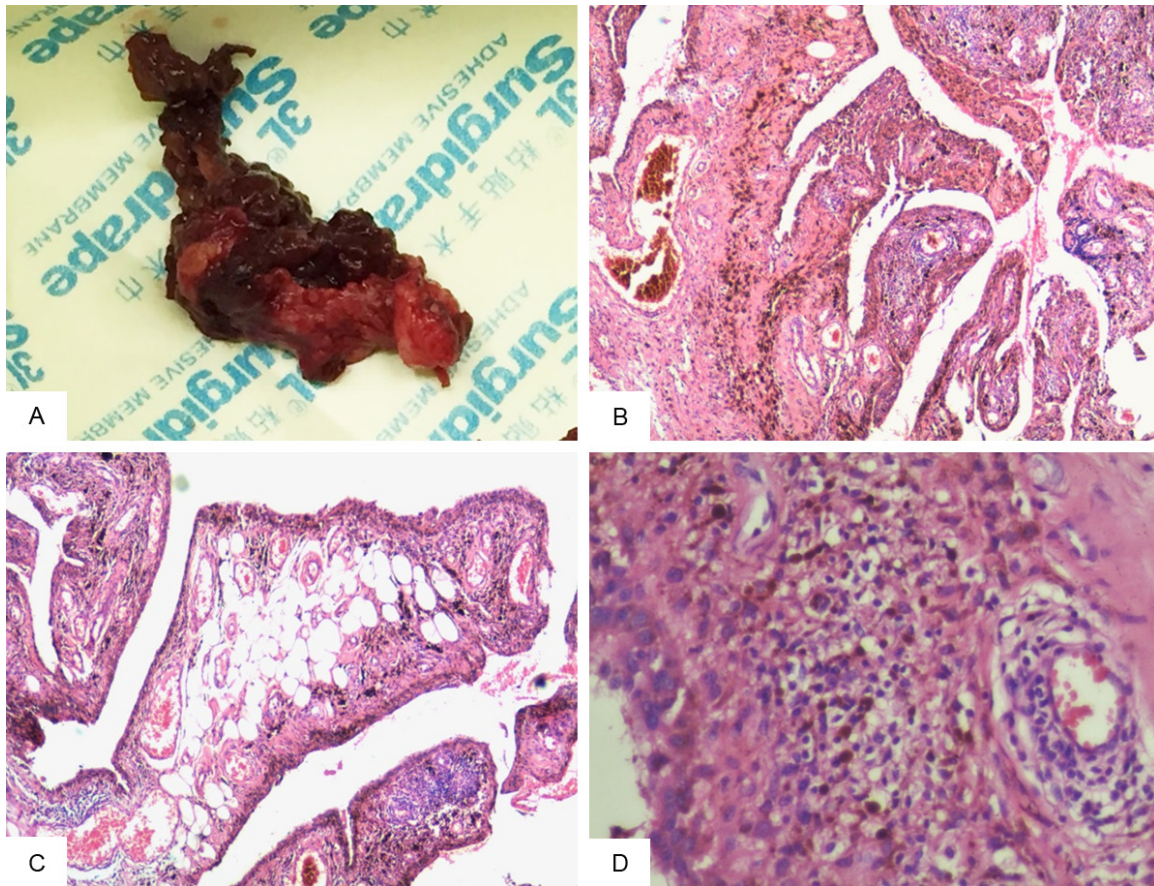


Figure 3. Pathologic results of the lesion. A. A part of gross specimen of the lesion extending beyond the hip joint capsule. The brown nodular lesion attached tightly to the proliferative synovium. B. Synovial tissue with typical papillary projections and brown pigmentation or hemosiderin in the cytoplasm (Hematoxylin and eosin, magnification $\times 40$). C. Papillary projection contains fat and hemosiderin (Hematoxylin and eosin, magnification $\times 40$). D. An area of multinucleated giant cells, foamy cells and brown pigmentation or hemosiderin deposits. (Hematoxylin and eosin, magnification $\times 100$).

Synovectomy alone or combined with arthroplasty is suitable treatment for PVNS. Unfortunately, although synovectomy is effective to alleviating pain and restoring joint range of motion, it fails to protect the joint against osteoarthritic progression, patients often need a joint replacement in the near future [2]. Arthroplasty conducted after dislocation and removal of the femoral head is a favorable option in cases presenting end-stage arthritis, which can also provide a better access for performing a completely synovectomy and lower local recurrence rate consequently. In cases of recurrent pigmented villonodular synovitis, local radiotherapy and intra-articular yttrium-90 (Y-90) may be helpful for improving clinical prognosis [6].

Hip dislocation secondary to developmental dysplasia of the right hip (Crowe type IV DDH) is

a debilitating condition, usually causing apparent shortening of the affected leg and end-stage arthritis. It is pathologically characterized by flattening or inversion of the labrum and capsular structures combined with dysplasia of the femoral ossification center and abnormal acetabula development, resulting in a high inclination and insufficient femoral head coverage [14]. Anteroposterior pelvic plain radiography is the cornerstone of initial assessment for DDH. Abnormal Tönnis angle (normal, $>10^\circ$) and lateral center-edge (CE) angle of Wiberg (normal, $>25^\circ$) are diagnostic criteria of hip dysplasia. Computed tomography (CT) scans can help surgeons assess acetabulum conditions and make preoperative planning for acetabulum re-directional osteotomy [15]. MRI is not regularly used in hip dysplasia, which is limited for checking suspicious lesions with no signs of structural

abnormality on radiographs. Joint-preserving surgeries including periacetabular osteotomy and femoral osteotomy are limited to young patients (<30 years old) with minimal cartilage degeneration and good hip-joint congruency before undergoing a surgery. Total hip arthroplasty is the standard treatment choices in adults with end-stage osteoarthritis or disability develop. However, complications including sciatic nerve palsy, post-op hip dislocation, intra-operative femoral fracture and infection rate are higher in patients with hip dysplasia than with osteoarthritis. In addition, considering the incidence of symptomatic periprosthetic inflammatory reactions of metal-on-metal bearing, current evidence supports the use of a metal-on-polyethylene bearing [16].

In conclusion, we report an especial case of PVNS, which occurred in fifty years after the patient got Crowe type IV developmental hip dysplasia. To the best of our knowledge, this is the first case report of PVNS combined with Crowe type IV DDH. Hip pain exacerbated with physical activity and joint swelling usually present at DDH patient. However, we hope to highlight that PVNS of the hip, despite being uncommon, should be considered as a possible cause of disabling pain in patient with hip dysplasia, especially when it is combined with an abnormal soft-tissue component. We speculate that PVNS in our patient results from a reaction to the inflammation factors induced by chronic injury or hemarthrosis and osteoarthritis. Further research and discussion are required to clarify the true etiology of PVNS. Synovectomy is a primary procedure for PVNS which can be effective in preservation of the hip joint, but fails to reduce the recurrence rate and progression of osteoarthritis. THA along with synovectomy is currently the best choice for advanced arthritis or hip dysplasia with PVNS. Long term follow-up is needed for the final result of this case.

Disclosure of conflict of interest

None.

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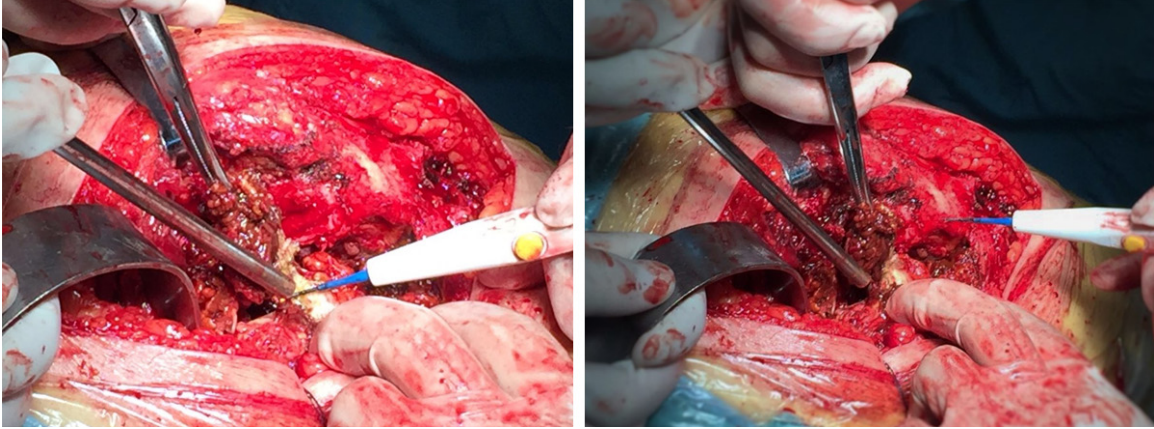
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Supplementary Figure 1. Intraoperative photo. Diffuse growth of multiple brown nodular lesions were found inside the articular capsule.