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Pigmented villonodular synovitis of the hip in a recreational runner: a case report

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Objective: *To present the diagnostic, clinical, and radiological features of pigmented villonodular synovitis (PVNS), create awareness of this rare condition, and provide guidance for conservative healthcare practitioners for further referral and appropriate management.*

Case presentation: *We present the case of a 41-year-old recreational runner who presented to the clinic with anterior hip pain of one year duration. Following a clinical history and examination, the patient was diagnosed with clinical femoroacetabular impingement. Radiographs taken at that time displayed mild degenerative joint disease of the left hip joint with coxa profunda. After four weeks of conservative care, the patient reported no improvement in symptoms. The patient was then referred for an MRI, while conservative care continued. Ten weeks later, the patient's symptoms and functional abilities had worsened. The MRI was obtained and the diagnosis of PVNS was made.*

Summary: *PVNS is a rare disease that can mimic mechanical hip pain. A high index of suspicion should*

Une synovite villonodulaire pigmentée de la hanche chez un coureur amateur

Objectif : *Présenter les caractéristiques diagnostiques, cliniques et radiologiques de la synovite villonodulaire pigmentée (PVNP), sensibiliser le public à cette maladie rare et fournir aux praticiens de santé conservateurs des conseils pour le renvoi des patients et la prise en charge.*

Présentation du cas : *Nous présentons le cas d'un coureur amateur de 41 ans qui s'est présenté à la clinique avec une douleur antérieure de la hanche depuis un an. Après une anamnèse et un examen clinique, on a diagnostiqué un conflit fémoro-acétabulaire. Les radiographies prises à ce moment-là révélaient une légère maladie dégénérative de la hanche gauche avec coxa profunda. Après quatre semaines de soins conservateurs, les symptômes du patient n'étaient pas soulagés. On lui a demandé de subir un examen par IRM et on a poursuivi les traitements conservateurs. Dix semaines plus tard, les symptômes et les capacités fonctionnelles du patient s'étaient aggravés. On a obtenu les résultats de l'examen par IRM et on a diagnostiqué une PVNP.*

Résumé : *La PVNP est une maladie rare qui peut imiter une douleur mécanique de la hanche. L'indice de*

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be utilized when symptoms worsen despite conservative care. Referral for advanced imaging is critical for appropriate diagnosis of PVNS.

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KEY WORDS: pigmented villonodular synovitis, FAI, hip, mechanical, pain, chiropractic

suspicion est élevé lorsque les symptômes s'aggravent malgré des soins conservateurs. Un examen par imagerie avancée est essentiel pour un établir un diagnostic de PVNP.

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MOTS CLÉS : synovite villonodulaire pigmentée, conflit fémoro-acétabulaire, hanche, mécanique, douleur, chiropratique

Introduction

Pigmented villonodular synovitis (PVNS) is a rare condition that is characterised by a proliferation or hyperplasia of the synovial membrane within joints, bursae, or tendon sheaths.[1] Due to the non-specific nature of the clinical presentation of PVNS, diagnosis is usually delayed.[2,3]

We report a case of PVNS of the hip in a recreational runner. The purpose of this case report is to present the diagnostic, clinical, and radiographic features of PVNS in order to create awareness of this rare condition amongst health care practitioners. It is important for healthcare practitioners to be aware of the clinical presentation and appropriate management for PVNS as it can mimic more common musculoskeletal conditions, as it did in this case.

Case presentation

A healthy 41-year-old architect and recreational runner presented to the clinic in November 2019 with a complaint of insidious onset intermittent left anterior hip pain of one year duration. The patient is female and stood 5 feet 9 inches (175.25 cm) and weighed 135 lbs (61.24 kg). Aggravating factors included running, prolonged sitting for more than thirty minutes, and yoga poses that require hip external rotation. Movement helped to relieve their pain. The patient did not report any external or internal hip clicking or clunking. The patient denied any red flags. Radiographs were ordered in May 2019 by the referring physician which displayed mild degenerative joint disease of the left hip joint with coxa profunda, mild facet arthrosis from L4-S1 and minimal degenerative joint disease of the left sacroiliac joint. (See Figures 1 and 2).

Upon physical exam, hip range of motion was full however, pain was elicited at end range internal and external rotation. Flexion Adduction Internal Rotation (FADDIR) and

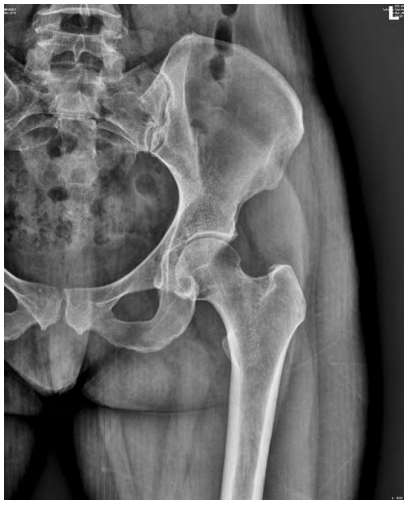


Figure 1. *Anterior posterior hip radiograph. Mild degenerative joint disease of the left hip joint with coxa profunda, mild facet arthrosis from L4-S1 and minimal degenerative joint disease of the left sacroiliac joint.*

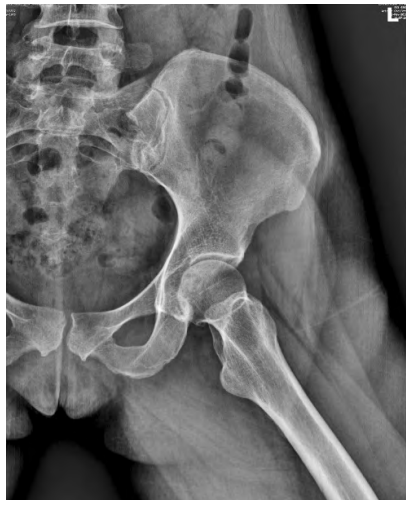


Figure 2. *Frog-leg lateral hip radiograph. Mild degenerative joint disease of the left hip joint with coxa profunda.*

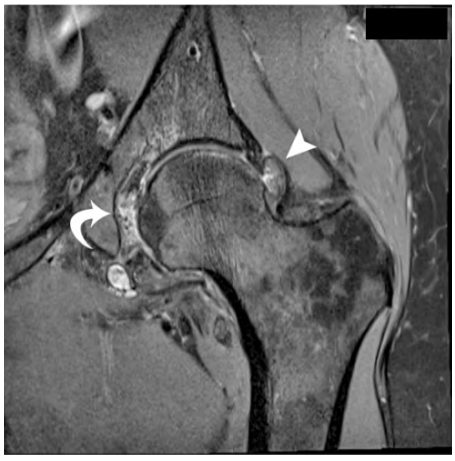
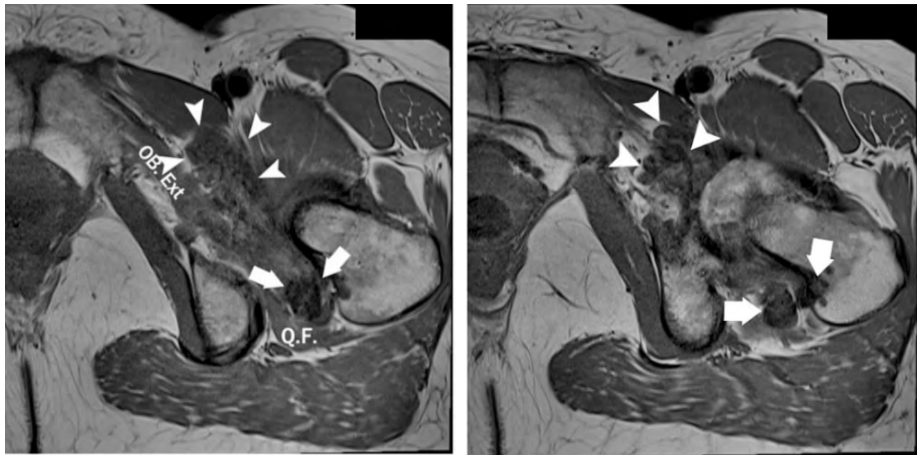


Figure 3.
Coronal Oblique Proton Density Fat Sat sequence (TR/TE: 4090/24) of left hip shows multiple frondlike foci of low signal intensity in the perilabral recess, above the femoral neck (white arrowhead) and in the acetabular fossa extending inferiorly (curved white arrow).



4A *4B*

Figure 4.
4A, 4B. Axial T1 weighted sequence (570/15) images at the level below the acetabular fossa show hypointense nodular masses displacing the obturator externus (OB. Ext.) muscle medially (arrow heads) and quadratus femoris (Q.F.) muscle posteriorly (arrows).

Flexion Abduction External Rotation (FABER) tests were positive and hip distraction was relieving. The hip pain was recreated with the left leg falling into abduction while in right lateral recumbent position. The following tests were found to be negative: hip scour, log roll, Thomas test, cross-legged sitting, resisted hip ranges of motion, sacroiliac joint provocation tests, and an examination of the lumbar spine was normal. Lower limb neurological examination, which included deep tendon reflexes, sensory and motor testing, was found to be within normal limits. A working diagnosis of clinical femoroacetabular impingement was made and the patient was treated one to two times per week for four weeks. The treatment plan included soft tissue therapy, hip joint mobilizations, ergonomic modifications, and a comprehensive rehabilitation program focusing on core endurance, gluteal and hamstring strength.

In this case, the working diagnosis of clinical femoroacetabular impingement was diagnosed based on the Warwick International Consensus Agreement, that defines FAI as a clinical disorder of the hip with a triad of symptoms (motion- or position-related pain in the hip or groin), clinical signs (positive hip impingement tests, such as FADDIR), and imaging findings that show evidence of cam or pincer morphology on plain-film radiographs.[4]

A re-evaluation was conducted after four weeks of care and the patient reported no improvement in symptoms. A letter was sent back to their medical doctor suggesting that further diagnostic imaging may be necessary. Subsequently, an MRI was ordered in January 2020. Meanwhile, care was continued with progression to rehabilitation and modifications to passive treatment. However, by late January 2020 the patient reported worsening of pain post-treatment and the inability to sit for longer than five to 10 minutes. They also reported an uncomfortable clicking sensation with walking and the pain was interrupting their sleep. By February 2020, they reported that the hip felt "out of place" and that the pain was occurring with walking. Care was ceased at this time and the patient awaited the MRI.

The MRI was scheduled for late April 2020, but due to the COVID-19 lockdown, the patient obtained their MRI in October 2020. Based on the MRI findings the patient was diagnosed with pigmented villonodular synovitis (see Figures 3, 4A, 4B, and 5). This patient was then re-

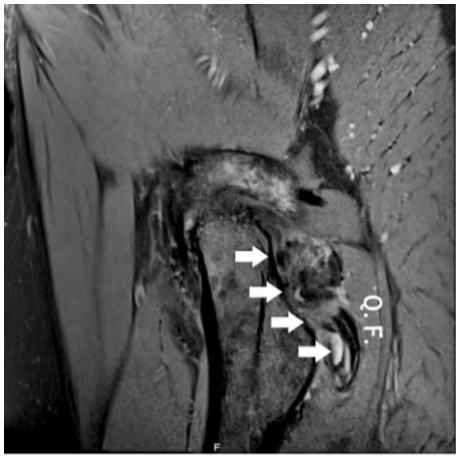


Figure 5.
Sagittal Proton Density fat-suppression sequence (2630/23) shows predominantly hypointense nodular masses (white arrows) posterior to the femoral neck and greater trochanter displacing quadratus femoris (Q.F.) muscle posteriorly.

ferred to an orthopedic specialist and is currently awaiting intervention.

Discussion

Pigmented villonodular synovitis (PVNS) is a type of tenosynovial giant-cell tumour that is defined by the proliferation of the synovial membrane in joints, bursae, or tendon sheaths.[1] It affects 1.8 per million people and the intra-articular form is predominantly found in large joints such as the hip (70% of cases) and knee (15% of cases).[1,5] PVNS of the hip typically occurs between the second and third decade of life.[1]

The etiology of PVNS is not well known, with trauma, neoplasia, genetic predisposition, and chronic inflammation secondary to hemarthrosis suggested as possible causes.[1] There may be a connection to chromosomal anomalies, which has been suggested more recently.[6]

The clinical presentation of PVNS of the hip is non-specific, which requires clinicians to have a high index of suspicion for the diagnosis.[1] Diagnosis is often delayed due to slow progression of symptoms, pain tolerance, and symptom variation, with an average of four years between onset of symptoms and diagnosis.[2,3] The main symptom is pain, which may be localized to the anterior hip or groin, that increases in intensity and duration as the disease progresses.[3] Other signs and symptoms may include painful intra- or extra-articular swelling, which can result in associated limitation in mobility and stiffness of the joint.[1,3]

Imaging studies are critical for the diagnosis and treatment planning of PVNS. Specifically, plain film radiographs, CT, and MRI can be used to assess the presence and severity of the disease. Radiographs are often unremarkable in early stages, as they were in our case. However, in the advanced stage of the disease, radiograph will display osteolytic changes, specifically erosions of bone and subchondral cysts around the axis of the joint capsule or at the acetabulum near the fovea.[3] CT can also be used to visualize smaller erosions and subchondral cysts that are not visible on radiograph, however it is less helpful in evaluating the surrounding soft tissues and synovium.[7,8] Lequesne *et al*.[8] proposed a radiologic classification system for PVNS:

1. "Evocative" – initial stage of disease with or without joint space narrowing as well as large subchondral cysts. This form represents 62% of cases.[1]
2. "Pseudo-coxitis" – localized joint space narrowing with deep erosions of the head of the femur and/or acetabulum in weight bearing surfaces. Possibility of subchondral cysts outside the weight bearing area. This form represents 16% of cases.[1]
3. "Pseudo-coxarthrosis" – localized joint space narrowing is secondary to the expansion of subchondral cysts into the joint space. This form represents 14% of cases.[1]

The preferred imaging modality for examination of PVNS is MRI.[9] It allows for precise visualization of the lesion(s), which allows for accurate diagnosis as well as pre-intervention planning and post-intervention review.[7] Features of PVNS on MRI include: (1) variable extent of synovial proliferation and thickening, (2) intra-articular effusion and erosion of bone, and (3) the deposit of hemosiderin within lesions in the synovium.[7,10] The lesions containing hemosiderin are pathognomic of PVNS.[9,11,12] Hemosiderin is a protein compound that stores iron. Due to the ferromagnetic properties of this protein, when deposited on synovial tissue, it results in a hyposignal on

T1- and T2- weighted images.[10] Fast field echo (FFE) sequence MRI images are best to visualize hemosiderin deposits, which can be visualized as small spotty areas, or diffuse larger areas of hypointense signal.[10]

The current consensus based on large case studies in the literature, is that operative management is the standard for treatment for PVNS.[2,3,9] However, the surgical approach is poorly defined in the case of PVNS of the hip. Joint preservation is preferred as much as possible for younger patients.[1] Total synovectomy of the diseased synovium is typically the approach when the articular cartilage is preserved, in order to prevent further destruction and loss of function of the joint.[2,9] This is typically done with open surgery and surgical hip dislocation which exposes all the synovial recesses.[8,13] However, this procedure does not seem to prevent the development of secondary hip osteoarthritis in the future.[9] Total hip arthroplasty is considered when there is significant joint destruction present.[1,2,9] More recently, arthroscopic excision of the diseased synovium has been studied as a treatment procedure. Byrd and colleagues described good outcomes in their study, including improvements in patient-reported outcome measures, and only one out of thirteen patients undergoing a revision after six years.[14,15] External radiotherapy has been proposed, but not validated in the literature as an effective treatment modality.[1,2,16] There is some literature that supports external radiotherapy post-operative management to help prevent recurrence and address any residual disease.[17]

Given its vague signs and symptoms, PVNS of the hip often masquerades as mechanical hip pain, which is particularly common in runners. This may lead to a delay in diagnosis and appropriate treatment. It is critical for conservative healthcare practitioners, including chiropractors, to have a high index of suspicion for this diagnosis, particularly when there is a lack of improvement with conservative care and a progression of pain intensity and duration, and decrease in functional abilities. If radiographic features are associated with uncharacteristic clinical symptoms, PVNS should be considered as a differential diagnosis.[3] When PVNS of the hip is suspected, referral for diagnostic imaging (specifically, MRI) and subsequent surgical referral are prudent.

Summary

The clinical presentation of the PVNS of the hip in this case was typical of clinical femoroacetabular impingement that presents often to healthcare providers such as chiropractors and physiotherapists. It is prudent for therapists to have an index of suspicion for this diagnosis when symptoms do not improve or worsen with conservative care. Referral for advanced diagnostic imaging is important to accurately diagnose this condition, and further surgical referral is required for appropriate management. Early diagnosis and intervention for PVNS results in better outcomes for the patient.

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