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Pigmented villonodular synovitis associated with developmental dysplasia of the hip: A case report

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ABSTRACT

INTRODUCTION: Pigmented villonodular synovitis (PVNS) of the hip joint associated with developmental dysplasia of the hip joint (DDH) is an extremely rare co-presentation. The aim of this study is to report a case of PVNS associated with DDH.
PRESENTATION OF CASE: A 26-year-old women, a known case of DDH, presented with progressive right hip pain for one month duration. She was able to perform all the right hip movements with limitation due to pain. Plain radiographs showed a hip dislocation. Through a posterior incision, a mass of brown fibro-fatty soft tissue emerged. The specimen was sent for histopathological examination, The findings were variable mixture of giant cells, hemosiderin, and brown pigmentation in the synoviocyte cytoplasm.
DISCUSSION: Although both conditions were previously described separately, the significance of co-incidence of DDH and PVNS is not well understood due to the rarity of the association.
CONCLUSION: Co-incidence of DDH with PVNS is an extremely rare finding but could be safely managed if caught early in the beginning of the disease.

1. Introduction

Pigmented villonodular synovitis which was coined by Jaffe and associates for the first time in 1941 is a rare proliferative disorder that can affect synovium in young and middle-aged population [1,2]. The pathophysiology and etiology is not well understood. Some researchers think that chronic inflammation is the underlying cause. Others propose tumor-like disorder (uncontrolled proliferation) as the original pathology. The latter is maintained by chromosomal abnormalities and monoclonality [3]. Two types of PVNS have been described: a local nodular PVNS type, affecting tendon sheaths and smaller joints, and a diffuse PVNS pattern that is mostly intraarticular in the larger joints of the knee, hip, the ankle, shoulder, elbow, and wrist [4].

PVNS occurring in patients with DDH is an extremely rare finding, to date, only one case has been reported in the peer reviewed journals [5].

The aim of this study is to report a case of DDH whom intraoperatively found to have PVNS. The work has been written in line with SCARE 2018 guidelines [6].

1.1. Patient information

A 26-year-old women, known case of DDH, presented with progressive right hip pain for one month duration. She had limping since childhood, she did not get benefit from conservative treatment with physical therapy and oral analgesia. Past medical, past surgical, drug and family histories were negative.

1.2. Clinical findings

She was conscious, oriented, and was able to perform all the right hip movements with limitation due to pain, the range of motion was decreased gradually, she had short right lower limb with 6 cm limb length discrepancy.

1.3. Diagnostic assessment

Rheological tests were within the normal ranges, plain radiographs showed a hip dislocation secondary to DDH of the right hip (Crowe type IV) [Fig. 1], No mass or cystic lesion was found.

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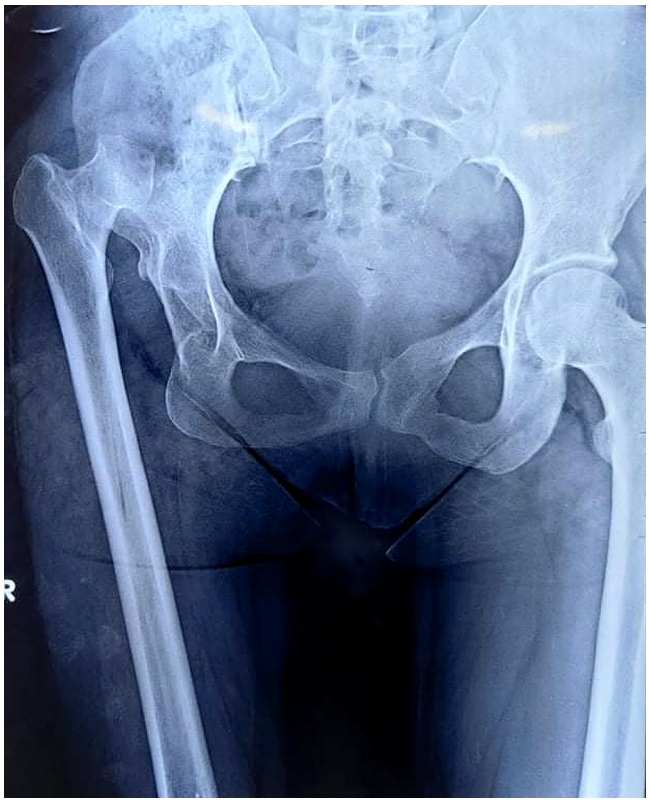

Fig. 1. Plain x-ray (anterio-posterior view) of the both hips, showing right hip dislocation (Crowe type IV), with right femoral head osteoarthritic changes.

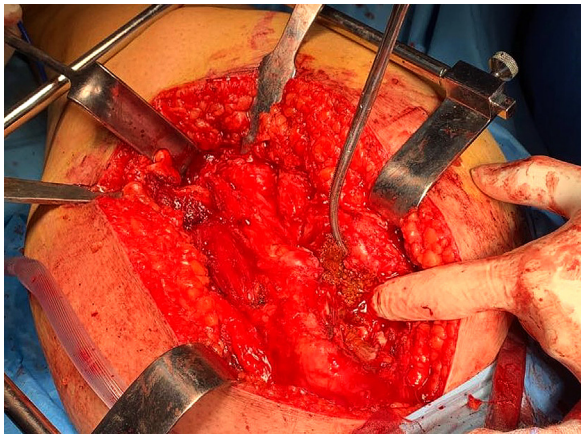

Fig. 2. Intraoperative photo showing a brownish mass emerging through the wound.

1.4. Therapeutic intervention

Based on the physical examination and radiological findings, a decision for total hip arthroplasty was met. Through a posterior incision, a mass of brown fibro-fatty soft tissue emerged. A growth of multiple brown nodular lesions were found inside the articular capsule of cystic nature, no extra-articular erosion was seen [Fig. 2]. Wide local excision was done. The specimen was sent for histopathological examination, a benign condition was suspected intraoperatively, so the procedure was resumed with subtrochanteric osteotomy, total hip arthroplasty was performed [Fig. 3], both limbs at end of operation was in same length, postoperatively microscopic section of the biopsy showed PVNS, the finding was variable mixture of giant cells, hemosiderin, and brown

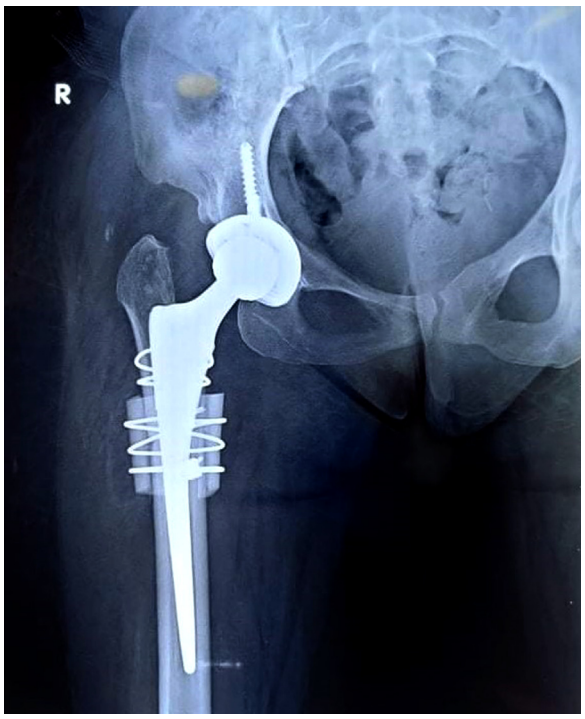

Fig. 3. Postoperative plain radiograph of right hip (anteroposterior view) showing femoral osteotomy with well aligned acetabular cup and femoral stem.

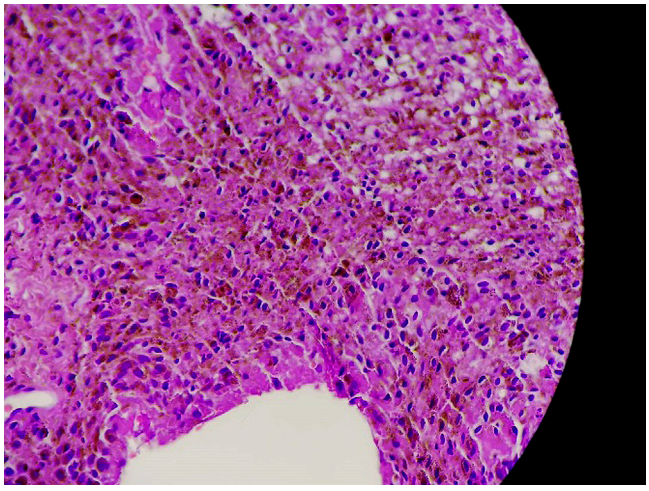

Fig. 4. Microphotography showing nodules with an abundant number of (pigmented) hemosiderin-laden macrophages.

pigmentation in the synoviocyte cytoplasm (Fig. 4). The procedure was performed by the first two authors.

1.5. Follow up

The post-operative course was uneventful. The patient was discharged from hospital on the first postoperative day. Six months after the intervention, the patient was found to be healthy, free from pain and limping. Plain hip x-ray showed a well-aligned ball and socket joint.

2. Discussion

Although both conditions were previously described separately, the significance of co-incidence of DDH and PVNS is not well understood due to rarity of the association [5].

In case of PVNS, gross histopathology may show diffusely thickened synovium, and larger lobulated masses inside the joint, occupying the joint cavity eroding the bone, also it may extend outside the joint via capsular perforation or through bursae communicating with the joint [1].

Although it may present with pain and swelling, the hallmark of the presentation is recurrent heamarthrosis (bloody effusion by arthrocentesis) [1]. The current case was admitted to the operating room for management of DDH.

Imaging is a part of the diagnostic workup, X-ray may show bony erosions with or without subchondral cysts in diffuse articular PVNS such as in the hip, ankle and tarsal joints. The cysts are round and lobulated, lined by thin sclerosis with sharp edges. Computed tomography (CT) scan may show thickening of the synovium, with intra or extra articular mass [7]. Magnetic resonance imaging (MRI) is the most sensitive imaging modality, in the hip joint, the typical findings include joint effusion, low signal intensity on both T1 and T2 weighted images due to hemosiderin deposition, lobulated synovial mass due to hyperplastic synovium, and bony erosions [4]. In this case preoperative work up failed to recognize the condition.

The target of treatment is complete arthroscopic synovectomy, followed by open posterior synovectomy if extraarticular extension is present, while local form could be treated by partial arthroscopic synovectomy [1]. In the present case, extra articular extension was not found.

Atilla and associates published their experience with a 55-year-old female presenting with an indolent swelling in the right upper thigh for one year duration. Preliminary evaluation discovered a well-circumscribed mass in the right hip joint. The patient's clinical assessment revealed bilateral DDH. The patient also complained from an insidious pain in the right hip joint and progressive buttock swelling with discomfort. A soft tissue 15 cm mass was evident from the greater trochanter extending to the ipsilateral iliac crest [5]. The current case was not similar in that the swelling was not evident preoperatively and findings were surprise for the surgical team although the operation proceeded owing to the benign nature of the lesion.

The role of complementary treatment is not known. Deep radiotherapy and isotope synoviorthesis have been suggested by some physician. Several isotopes have been used such as Rhenium 186, Yttrium 90, and Dysprosium 165. This complementary therapy has been recommended as the rate of recurrence is high in diffuse PVNS [3]. This case did not receive complementary therapy as the pathology was localized.

In conclusion, co-incidence of DDH with PVNS is an extremely rare finding but could be safely managed if caught early in the beginning of the disease.

Declaration of Competing Interest

The authors report no declarations of interest.

Sources of funding

No source to be stated.

Ethical approval

Approval is not necessary for case report in our locality.

Consent

Consent has been taken from the patient and the family of the patient.

Author contribution

Saywan Kaka Rash: Surgeons performing the operation, follow up the patient and final approval of the manuscript.

Fahmi Hussein Kakamad, Shvan H. Mohammed, Kardo M. Salih, Abdulwahid M. Salih: Writing the manuscript, final approval of the manuscript.

Registration of research studies

According to your previous recommendation, registration is not required for case report.

Guarantor

Fahmi Hussein Kakamad is the Guarantor of submission.

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