Adult onset polyarticular pigmented villonodular synovitis

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Abstract

Pigmented Villonodular Synovitis (PVNS), typically, presents as a chronic monoarthritis affecting medium to large joints or tendon sheaths. Polyarticular presentation of PVNS is extremely rare, mostly reported in children and has not yet been reported in adults. We report a case of polyarticular PVNS, presenting in adulthood. We made the diagnosis based on characteristic MRI findings and typical histopathological changes. Patient was successfully managed with synovectomies and was not subject to inappropriate DMARDs. We suggest, that PVNS should be considered in the differential diagnosis of seronegative polyarthropathy, to avoid a delay in diagnosis and consequent delay in appropriate treatment. To our knowledge, this is the first case report of PPVNS presenting in adulthood, and serves as an example of atypical presentation of a rare condition.

Introduction

Symmetrical polyarthritis in the absence of rheumatoid factor and/or anti-CCP (cyclic citrullinated peptide) antibody may be diagnosed as seronegative arthritis. Seronegative arthritis encompasses several differential diagnoses including seronegative rheumatoid arthritis and inflammatory arthritis secondary to infections such as tuberculosis, hepatitis and human immunodeficiency virus. In children, pigmented villo nodular synovitis has been reported to manifest as polyarthropathy. Therefore, early diagnosis is of high importance in the management of seronegative arthritis.

Here we present a case polyarticular PVNS presenting for the first time in adulthood. We discuss useful investigations in the setting and emphasise on typical radiographic findings. Finally, we show that unjudicious use of Disease Modifying Anti-Rheumatic Drugs (DMARDs) could be avoided by careful consideration of differential diagnosis. Further, we show that even polyarticular presentation is amenable to succesful surgical intervention. Thus our case highlights the importance of considering PVNS in the differential diagnosis of seronegative arthritis.

Case Report

A 38 year old Bangladeshi gentleman was referred to our rheumatology department, with a 6 week history of bilateral painful and swollen knees. He experienced early morning stiffness in his knees, lasting for up to an hour. He had been involved in a road traffic accident seven years earlier, but could not recollect any injury to his knees. Systems enquiry was normal. There was no history of Raynaud's phenomenon, mouth ulcers, hair loss, photosensitivity, dry eyes or dry mouth. There was no history of weight loss, fever or night sweats. He had no symptoms suggestive of tuberculosis and had no contact with patients suffering from tuberculosis. His past medical history was unremarkable. He was not taking any medications. He worked as a chef in a restaurant.

On examination he looked systemically well. There was no lymphadenopathy, edema, pallor or icterus. Respiratory, cardiovascular and abdominal examination was unremarkable. Musculoskeletal examination revealed prominent swelling of the right knee. This was associated with reduced range of movements. He had no features suggestive of systemic lupus erythematosus or connective tissue disease.

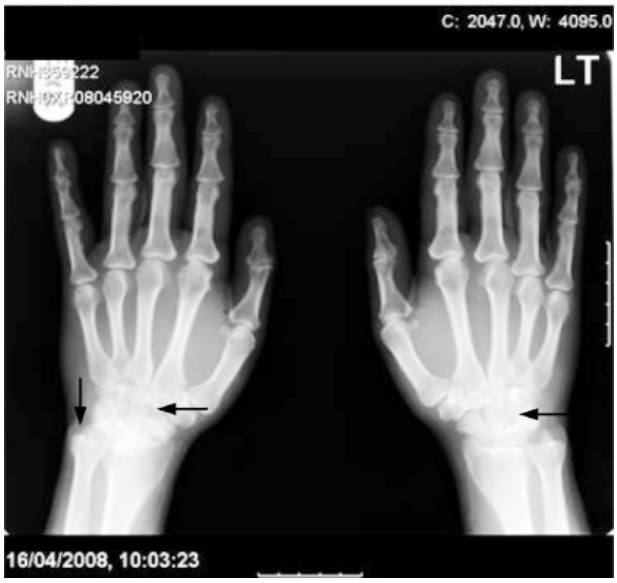
Full blood count, urea, electrolytes, liver function tests, erythrocyte sedimentation rate, c-reactive protein were normal. Rheumatoid factor and anti nuclear antibody were negative.

X-rays of the knees showed minor osteoarthritic changes. Analysis of his knee fluid aspirate, revealed a few pus cells, but no crystals. Cytology was negative. AFB was negative on both microscopy and cultures. MRI of his right knee demonstrated changes typical of PVNS (Figure 1). Arthroscopic examination of the right knee showed synovial thickening, moderate effusion, generalised extensive proliferative villous vascular synovium.

Medial menisectomy was performed. Biopsy findings were suggestive of pigmented villonodular synovitis. A year later, due to persistent pain, a total synovectomy was performed. Histology revealed changes of chronic synovitis.

A few months later, he presented with painful wrists. Both wrists were tender and movements were restricted. The left wrist showed advanced osteoarthritic changes with erosive changes of the proximal row of carpal bones.

Figure 1: X-ray images of hands demonstrate erosive changes (black arrow) of the carpal bones and radiocarpal and radioulnar joint

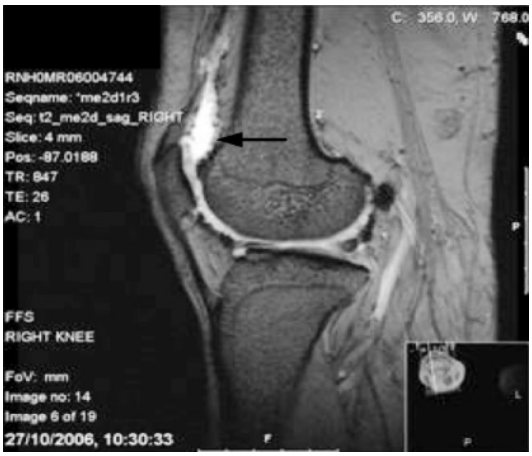


Two years later, he experienced severe pain, swelling and stiffness of right wrist. A total synovectomy was performed. Histological examination revealed diffuse chronic inflammatory cell infiltrate, rich in plasma cells, marked synovial hyperplasia with surface fibrin deposition. Collections of haemosiderin laden macrophages were present. Appearances were those of chronic synovitis with features suggestive of pigmented villonodular synovitis.

Seven years later, he experienced worsening pain in his right knee and left wrist. Magnetic resonance images of his right knee and left wrist showed changes consistent with pigmented villonodular synovitis in both knees and the left wrist.

Figure 2: Magnetic Resonance Images (MRI) of the right knee demonstrates an effusion with low signal areas (black arrow) due to haemosiderin (A). MRI of the left wrist demonstrates low signal areas (black arrow) due to haemosiderin and erosive changes.

A



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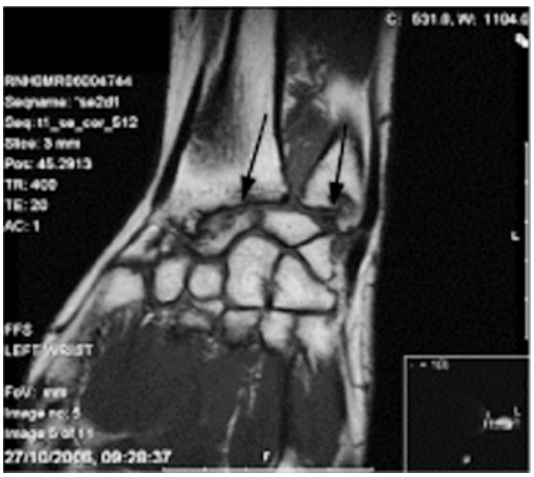

A year later, he was admitted with abdominal pain, when a diagnosis of pleural and peritoneal tuberculosis was made based on the clinical findings, pleural biopsy, computerised tomogram of chest and abdomen. Anti-tuberculous medication was started with good response.

In summary, our patient had chronic polyarticular PVNS, affecting multiple joints over a period of 7 to 8 years. On each occasion, a diagnosis was based on the characteristic radiographic (MRI) or histopathological features. Synovectomy proved to be effective, to treat both large and small joint disease. Immunosuppressive therapy was appropriately avoided.

Discussion

Pigmented Villonodular Synovitis (PVNS) is a benign proliferative disorder affecting synovial joints, bursae and tendon sheaths. Typically, it presents as a chronic monoarthritis of large joints, in the second to fifth decade, however, it has been described in paediatric as well as elderly patients. . A case of acute onset mono-arthritis has been described[1]. The annual incidence is estimated to be two cases per million population. There may be a long delay in presentation, particularly in children, as the joint pain is often mild and is disproportionate to the swelling[2]. Most commonly, it affects the knee (80%) and less frequently involves hips, ankles and shoulders. Multiple joint involvement has only been reported in younger patients and to our knowledge it has not yet been described in adults.

Aetiopathogenesis is poorly understood. A neoplastic aetiology has been suggested based on the findings of aneuploid DNA, chromosomal abnormality and malignant potential[3]. Reports of PVNS occuring in association with congenital anomalies raises a possible genetic link[4,5].

Diagnosis is made on the typical histopathological and radiographic (MRI) features. MRI findings of synovial proliferation, joint effusion and erosion of bone are common. Haemosiderin deposits in the synovial masses, appear as low signal areas, best seen on Fast Field Echo (FFE) sequence and are diagnostic of PVNS[6]. However, MRI may not differentiate PVNS from other causes of chronic hemorrhagic synovitis.

The prognosis is generally good, but may cause erosive arthritis. Localized lesions may be amenable to arthroscopic interventions, however, diffuse form may need total surgical synovectomy. Relapse is a frequent problem, and has been treated with either intrarticular radioactive Yttrium or anti-TNF agent, following athroscopic surgery[8,9] and Imatinib[10]. Anti-TNF therapy has been used to treat a case of PVNS, although it took few months to achieve disease control[11]. Therefore, surgical removal of the lesion appear to be associated with low risk of relapse whereas medical interventions including biological treatments appear to have variable benefit.

In conclusion, our case highlights, that atypical presentation of PVNS, needs to be considered in the differential diagnosis of a patient with seronegative arthropathy. We suggest that careful consideration of this rare condition may avoid unjudicious use of DMARDs and that even polyarticular PVNS is amenable to successful surgical intervention. The study shows that polyarticular pigmented villonodular synovitis may present as symmetrical seronegative arthropathy even in adults.

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