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Case Report

Bilateral coxitis in scleroderma-polymyositis overlap syndrome

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Abstract

Joint manifestations in scleroderma (Scl) and polymyositis (PM) are dominated by inflammatory arthralgia. Arthritis is less common and preferentially affects the hands, wrists, knees, and ankles. Involvement of the hip has been rarely reported in the literature. We report a case of coxitis diagnosed in a patient suffering from scleroderma-polymyositis overlap syndrome successfully treated by ultrasound-guided infiltration of triamcinolone hexacetonide

Key words: Overlap, scleroderma, polymyositis, coxitis, hip involvement

Introduction

Overlap syndrome is defined as an entity that satisfies the diagnostic criteria of at least two connective tissue diseases (1). The most common combinations are systemic sclerosis with Sjogren's syndrome, dermatomyositis or polymyositis, rheumatoid arthritis, and systemic lupus erythematosus.

Scleroderma is the most frequent connective tissue disorder associated with polymyositis (PM), with a prevalence of around 24% (2). Scleromyositis, a scleroderma/polymyositis (Scl/PM) overlap syndrome, belongs to heterogeneous syndromes of idiopathic inflammatory myopathies, which can be divided into groups on the basis of immunological and clinical features (3). Articular involvement is more frequent in patients where PM is a feature of overlap syndrome, with a prevalence between 25%-42% and preferentially affecting the hands, wrists, knees, and ankles (3, 4). The involvement of the hip is unusual within the connective tissue disease, especially Scl/PM, and has been rarely reported in the literature.

Case Presentation

A 33-year-old man was followed for 2 years for systemic sclerosis-polymyositis overlap syndrome, retained on clinical (skin sclerosis of the face, fingers and forearms, Raynaud's phenomenon , girdle muscular weakness), biological (positive antinuclear antibodies at 1280 IU/mL, creatine phosphokinase (CPK): 13 times the upper limits of normal, lactate dehydrogenase (LDH): 2.5 times the upper limits of normal, aspartate aminotransferase (AST): 2.5 times the upper limit of normal, alanine aminotransferase (ALT): 1.5 times the upper limits of normal), and histological evidence (muscle biopsy for polymyositis, renal biopsy objectifying focal segmental glomerulosclerosis in favor of scleroderma). The patient was treated with steroids, colchicine, and calcium channel blockers.

Two months before his admission, the patient presented with inflammatory pain in the right hip with reduced perimeter of walking (50-100 m). The pain intensity was assessed by the visual analog scale (VAS), which was 9/10.

Clinical examination evidenced an antalgic posture with flexion-abduction-external rotation of the right leg, a limp when walking, and restricted motion of both hips, mainly rotations and flexion (flexion 90° in both of hips, internal rotation 10° in the right hip).

Radiographs of the pelvis and Lequesne false profile* showed bilateral coxitis with global narrowing, especially of the right hip joint (Figure 1). Magnetic resonance imaging (MRI) of the pelvis showed bilateral synovitis more important on the right side without bone damage (Figure 2).

*Lequesne false profile is an oblique view of the edge of the acetabulum used to diagnose arthrosis affecting the anterior part of the joint and to measure the anterior coverage of the femoral head.

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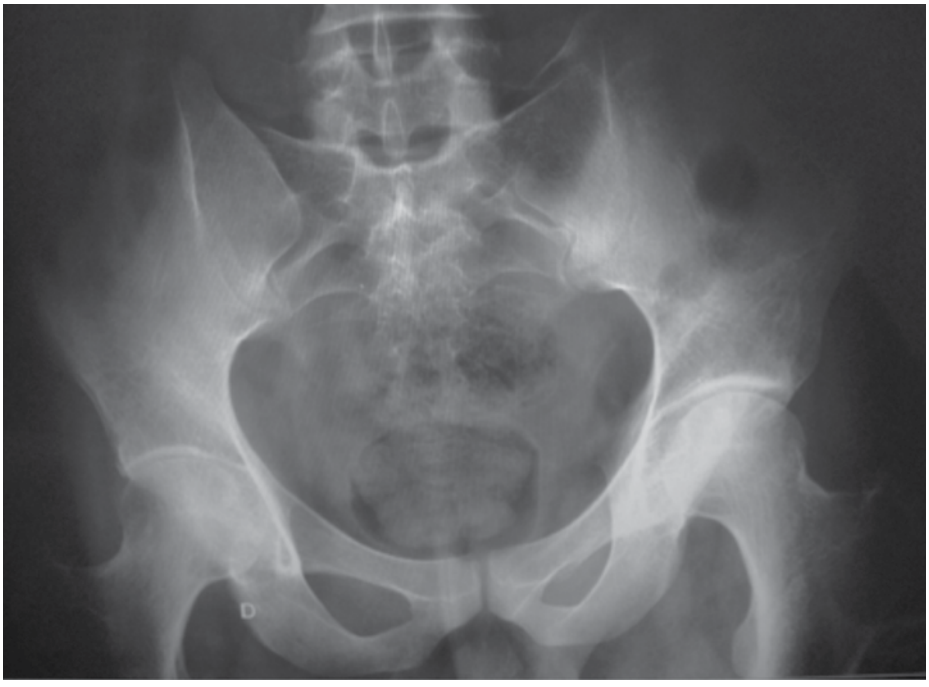

Figure 1. Radiograph of the pelvis showing global narrowing of the right hip joint

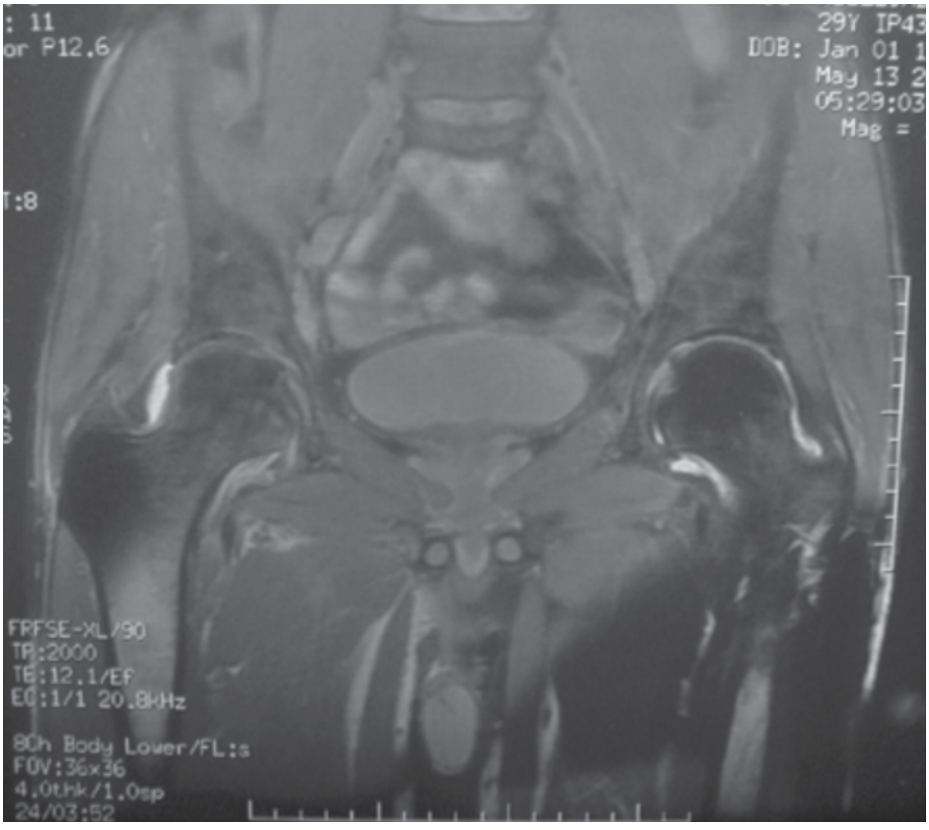

Figure 2. MRI of the pelvis showing bilateral synovitis of the hips

Laboratory tests found a moderate inflammatory syndrome (erythrocyte sedimentation rate (ESR) = 30 mm/h; C-reactive protein (CRP)=44 mg/L; 0-6 mg/L for normal range). The immunological tests were positive for antinuclear antibodies and PM/Scl. Anti-DNA and other soluble antigens (anti-U1 RNP, anti-Sm, anti-SSA/SSB, anti-Jo-1, and anti-centromere) were negative, ruling out the possibility of an anti-synthetase syndrome. Rheumatoid factor, anti-CCP, and X-rays of the hands and wrists were normal, ruling out the diagnosis of rheumatoid arthritis. The sacroiliac joints were normal in the radiographs and STIR MRI, eliminating the diagnosis of spondyloarthritis. The diagnosis of bilateral coxitis in the overlap syndrome was established. The patient underwent ultrasound-guided infiltration of triamcinolone hexacetonide (Hexatrione*, DEXO laboratories, Saint-Cloud, France) in the right hip, with good improvement in pain (VAS= 1/10), biologic inflammatory syndrome (ESR=12 mm/h, CRP=5 mg/L), range of motion, and unlimited perimeter of walking that was maintained after a 6-month follow-up.

Discussion

Older literature cases of severe diffuse scleroderma with extensive muscle involvement were described under the term scleromyositis. More recently, immunologic studies on scleroderma/polymyositis overlap syndrome disclosed a variety of antibodies associated with polymyositis and the scleroderma/polymyositis overlap. This overlap has been described in adults and adolescents. In a Canadian cohort of 100 patients with inflammatory myopathy, 29% had features of systemic scleroderma (SSc), which constituted 42% of 24 overlap syndrome patients in this cohort (1). Myositis may appear simultaneously with, before, or in already established SSc.

The sex ratio in the overlap group differed from that seen in idiopathic PM. A 9:1 female-to-male ratio occurred in the overlap group compared with a 2:1 ratio in idiopathic disease.

In contrast to idiopathic PM, patients with overlap syndromes often present with symptoms other than muscle weakness. Arthralgias and/or arthritis, sclerodactyly, Raynaud's phenomenon, and myalgias are the most common findings noted, being found in approximately 40%-50% of patients (4).

The study of Marguerie et al. (5), based on a series of 32 patients followed up for 8 years, showed that the main clinical features of scleromyositis are: Raynaud's phenomenon, scleroderma-like changes of the face and hands, arthralgia or arthritis, myositis or myalgia, interstitial lung disease (ILD), and calcinosis. Visceral involvement is not frequent and not severe, except for ILD. Joint involvement occurs in thirds of SSc patients during the course of the disease but may be the onset manifestation (6).

In polymyositis, articular involvement is characterized by arthralgias and arthritis. It is usually noted early in the course of disease, involving the wrists, knees, and the small joints of the hands. Joint involvement is classically non-erosive and frequently responsive to the treatment of the underlying inflammatory myopathy (7). Articular improvement has been noted in 25%-35% of patients with PM. It is more fre-

quent in patients where PM is a feature of overlap syndrome (4). In the retrospective study of Bohan et al. (8), true arthritis was not seen in idiopathic PM. Clinical information regarding frank arthritis in this disorder is rare, except in the anti-synthetase syndrome (especially with anti-PL7 and anti-PL12). Based on other studies of this disorder, true arthritis seems to be rare, especially in the hip joint localization. In fact, there are no studies describing involvement of the hip in connective tissue diseases, even less in overlap syndromes.

Joint involvement in scleroderma, outside of overlap syndrome, occurs frequently and may resemble rheumatoid arthritis in the early stages but is less destructive. Radiographic testing, when evaluated with RF and anti-CCP, will be a helpful tool to discriminate SSc arthropathy from RA-SSc overlap (9). In this disorder, the skeletal symptoms are usually symmetrical polyarthralgia and stiffness in the small joints, but the knees, shoulders, and wrists may also be involved (10). True arthritis, manifested clinically by joint pain, swelling, and erythema, occurs early in the disorder in approximately 10% of patients and at some time during the course in about 50% (11).

Biologically, scleroderma/myositis is associated with specific autoantibodies: anti-PM-Scl, anti-Ku, anti-U2 RNP, and anti-U5 snRNP (1). Positive antibodies to PM/Scl correlated with arthritis and a benign course of ILD. This immunological marker, PM/Scl, is highly characteristic although not specific for scleromyositis. Nevertheless, clinical-serological correlations are of diagnostic importance (12). The radiologic findings in the joints are nonspecific and include diffuse or periarticular osteopenia, soft tissue swelling or atrophy, joint space narrowing, subluxations, and tuft resorption (11).

For treatment, the joint symptoms respond readily to small doses of corticosteroids and do not require more intensive therapy, and the prognosis is usually favorable.

The occurrence of coxitis within the framework of polymyositis-scleroderma overlap syndrome is exceptional. Before attributing the joint damage to this syndrome, it is imperative to rule out chronic inflammatory rheumatism or an associated anti-synthetase syndrome.

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