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SHORT COMMUNICATION

Typical Jaccoud's arthropathy in a patient with sarcoidosis

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Abstract Jaccoud's arthropathy (JA) is a deforming, non-erosive form of arthritis initially described in rheumatic fever but recently observed more frequently in patients with systemic lupus erythematosus. However, cases of JA have been described in association with other diffuse connective tissue diseases, neoplasias, and infection. We describe a rare case of sarcoidosis in a female subject who developed JA in her hands later in the course of the disease.

Keywords Jaccoud's arthropathy · Sarcoidosis · Deforming arthropathy

Introduction

Sarcoidosis is a systemic disease affecting several sites of the body, mainly the lungs, skin, and lymph nodes. Osteoarticular involvement has been observed in several case series and is characterized by acute arthritis in large joints in conjunction with fever, erythema nodosum, and mediastinal adenomegaly; chronic joint enlargement secondary to bone cysts; arthralgia and/or arthritis mimicking rheumatoid arthritis (RA), dactylitis, and sacroiliitis [1].

Jaccoud's arthropathy (JA) is a deforming, non-erosive form of arthritis that was initially described in rheumatic fever (RA) but recently has been observed more frequently in patients with systemic lupus erythematosus (SLE) [2]. In addition, JA has also been described in association with other diseases such as Sjögren's syndrome (SS) [3, 4], scleroderma [5, 6], dermatomyositis [6], psoriatic arthritis [7] vasculitis [8], ankylosing spondylitis [9], mixed conjunctive tissue disease [10, 11], pyrophosphate deposition disease [12], neoplasia [13, 14], and chronic pulmonary diseases [15]. To the best of our knowledge, the presence of JA in sarcoidosis has been described previously on only one occasion [16]. Herein, we describe another such case.

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Case report

In 2002, a previously healthy 51-year-old female patient began to experience intermittent pain and edema in her proximal interphalangeal joints, metacarpophalangeal joints, and wrists. The patient was prescribed benzathine penicillin, which she used for approximately 1 year with apparent clinical improvement. In October of 2009, erythematous and nodular lesions appeared in the region of the neck and upper limbs, in addition to the recurrence of joint pain. A skin biopsy was performed, the anatomical pathological result of which demonstrated chronic granulomatous inflammation without caseous necrosis. Further, search for acid-alcohol-resistant bacillus and fungus was negative. Having received a diagnosis of sarcoidosis, the patient began treatment with 40 mg per day of prednisone in February 2010. In March 2010, the patient was monitored in our clinic, showing persistent skin lesions and joint pain in the hands, despite the corticosteroid therapy. A physical examination revealed good general and nutritional health, and the patient was without fever and had normal blood pressure. Aside from the skin lesions mentioned previously, attention was called to the presence of reversible deformities of the hands type "swan neck"—consistent with a diagnosis

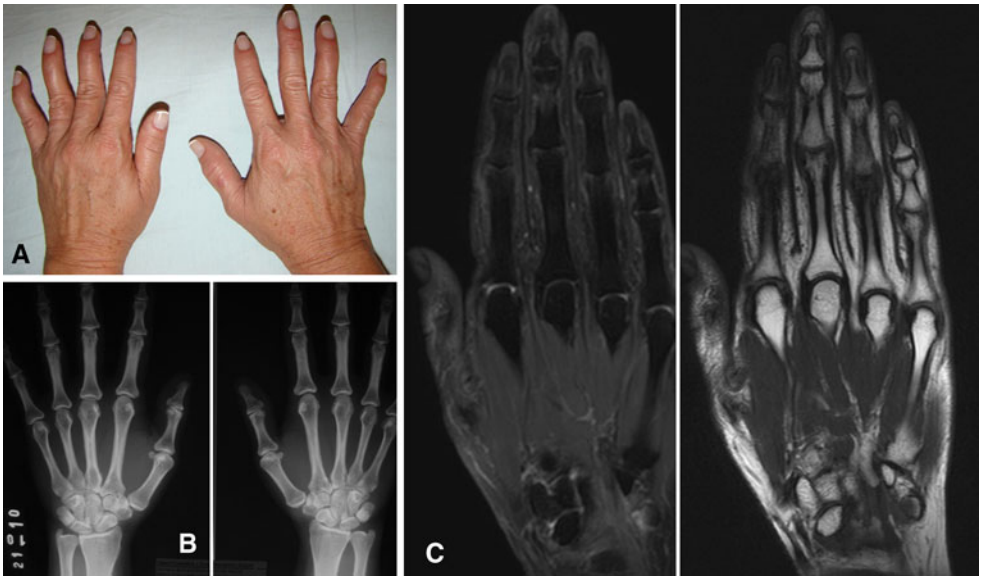

Fig. 1 **a** "Swan-neck" deformities of the hands. **b** Plain radiograph (*posterior–anterior view*) reveals no erosions. **c** Coronal fat-suppressed T2 and T1-weighted-SE magnetic resonance imaging demonstrates no signs of significant synovitis or erosions of the joints

of JA (Fig. 1). There were no signs of inflammation of the joints examined. The remainder of the physical exam was entirely normal. With respect to the complementary tests, the patient's hemogram, renal function, and liver profile were within normal parameters and the erythrocyte sedimentation rate and C-reactive protein were also normal. A plain radiological examination of the chest was normal. Rheumatoid factor, anti-CCP, ANA, anti-dsDNA, anti-SSA/Ro, anti-SSB/La, and anti-Sm antibodies were all negative; the Doppler echocardiogram was normal; plain X-ray, ultrasound, and magnetic resonance imaging of the hands did not present alterations (Fig. 1). The use of thalidomide was initiated in October 2010, a medication that brought relief to the skin and joint symptoms, while making possible the weaning from the corticosteroid.

Discussion

Clinically, JA is characterized by the presence of "reversible" joint deformities that mainly affect the hands, but that are also observed in other joints [17]. The joint deformities most frequently seen in JA are ulnar deviation, swan neck, "z"-thumb, and hallux valgus [2].

In cases of JA associated with RF, joint deformities are typically chronologically distant from the inflammatory articular manifestation, and sometimes patients with JA could even deny having experienced any previous articular symptom [19]. Similarly, at the moment of the evaluation, our patient had no sign of joint inflammation but only reversible deformities, reflecting a relationship to a previous joint injury as she described intermittent joint swollen in the past.

Other conditions that could lead to JA such as SLE, RF, SS, and HIV infection were ruled out based on the absence of clinical features and/or the negativity of the serological tests. Additionally, since rheumatic valvular heart disease is not rare in Brazil, an echocardiogram was performed, but this did not reveal any abnormality. In her case, sarcoidosis did not involve the lungs. Thus, chronic pulmonary disease was not the cause for her JA as has been previously reported [15]. The hand radiographs showed no erosions, and the absence of anti-CCP antibodies and rheumatoid factor excluded the presence of RA. While an "idiopathic" type of JA [20] cannot be excluded, it is unlikely as articular manifestation was a relevant feature of her disease; however, there is the possibility of coincidental yet not necessarily related conditions.

The mechanisms responsible for the development of JA are not yet well defined, although persistent synovitis and tenosynovitis and fibrotic retraction of the joint capsule may contribute [18]. In the only described case of JA associated with sarcoidosis [16], the patient had small subdermal nodules in her forearm and showed atypical deformities of JA characterized by flexion deformity of her fingers. An open biopsy of the patient's forearm revealed fibrosis of the tendons and granulomas of epithelioid cells in the muscles and tendon sheets. Our patient had no anatomopathological investigation of her joints, but both of her clinical and imaging findings seemed to be more typical of JA, similar to those seen in RF and SLE.

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Conflict of interest The authors have no conflicts of interest that are directly relevant to the content of this manuscript.

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