"Mutilans-type" Jaccoud Arthropathy

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Jaccoud arthropathy (JA) is a clinical complication most frequently seen in patients with systemic lupus erythematosus (SLE). It occurs at a prevalence of about 5%. JA is defined as the presence of reversible joint deformities, particularly in the hands, in the absence of articular erosions on plain radiographs[1]. A few small bone erosions may be identified by magnetic resonance imaging (MRI)[2]. The etiopathogenic mechanisms of the development of JA are unknown, but higher levels of serum interleukin 6 are observed in patients with concurrent SLE and JA than in patients without these conditions[3].

We herein present a case involving a 48-year-old woman with a 17-year history of SLE mainly characterized by malar rash, pleuritis, leukopenia, and intermittent joint inflammation in the hands, elbows, and knees. Despite the longterm use of hydroxychloroquine, azathioprine, methotrexate, and prednisone, she experienced progressive joint deformities that progressed to a severe deforming arthropathy (we named it as "mutilans type"), very different from classical JA, with luxation at several sites (Figure 1). Her hand deformities were not "reversible" like those seen in typical JA, probably secondary to a longstanding process of fibrosis in the soft tissues of the joints. This condition limited her articular functional capacity and decreased her quality of life.

Laboratory investigations showed an antinuclear antibody of 1/5120 homogeneous pattern with positivity of both anti-dsDNA and anti-SSA/Ro antibodies. Anticyclic citrullinated peptide antibodies and rheumatoid factor by ELISA were negative. Plain radiograph of the hands had no evidence of bone erosion; MRI revealed tenosynovitis, but also failed to show any erosion (Figure 2).

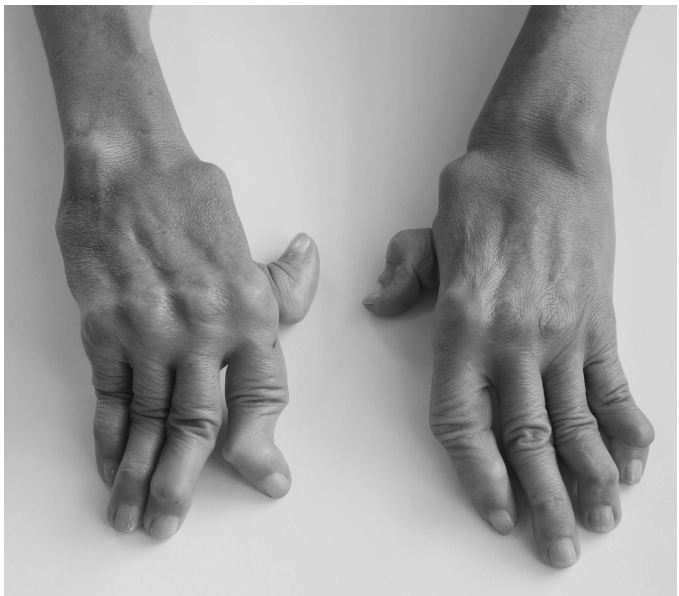

Figure 1. Severe hand deformities with joint luxations at several sites in a patient with Jaccoud arthropathy secondary to systemic lupus erythematosus.

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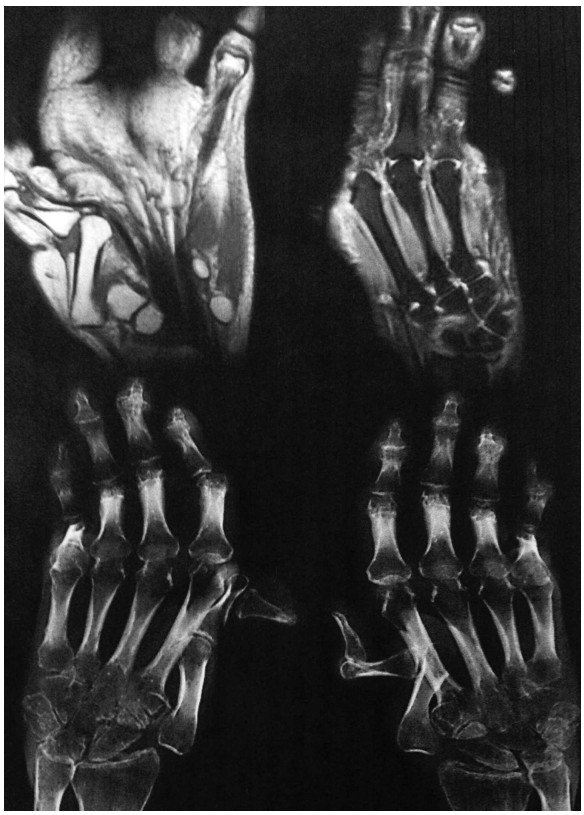

Figure 2. Coronal T1- and T2-weighted, fat-suppressed magnetic resonance images of the hands reveals mild synovitis and tenosynovitis without any erosion. Plain radiograph of the hands shows joint luxations, but no evidence of bone erosion.

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