Arthritis mutilans as a radiographic feature of systemic sclerosis

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A 47-year-old man presented to our Rheumatology department with a three-year clinical picture of Raynaud's phenomenon, symmetric and additive polyarthralgias of the wrists, hands, knees and ankles, as well as progressive skin thickening of the fingers, face, trunk and proximal limbs. He was previously diagnosed with anti-topoisomerase I antibody positive diffuse cutaneous systemic sclerosis (SSc) and treated with subcutaneous methotrexate 25mg/weekly and low-dose prednisolone (<10mg/daily). His disease remained stable during the year following the diagnosis with no evidence of arthritis, pulmonary, cardiac or renal involvement, after which he began complaining of inflammatory symmetric arthralgia and swelling of both wrists and hands.

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Physical examination revealed, apart from the sclerodactyly, *de novo* polyarthritis of the wrists, metacarpophalangeal (MCP), proximal and distal interphalangeal (IP) joints (Figure 1), as well as tenosynovitis of the extensor tendons of the fourth compartment of the wrists, bilaterally. Conventional radiography (Figure 2) showed a bilateral destructive arthropathy of the distal IP and trapezium-first metacarpal joints, with "pencil--in-cup" deformities and subluxation of the left first metacarpal. Ultrasound exhibited grey-scale synovitis of the radiocarpal, intercarpal, MCP and IP joints, with power-Doppler signal of the MCP joints. Figure 3 depicts the ultrasound appearance of the right second "pencil-in-cup" distal IP joint, with marked architectural distortion and heterogeneous grade 3 grey-scale synovitis.

The patient reported no personal or familiar history of psoriasis. Rheumatoid factor and anti-citrullinated protein antibodies were negative. A diagnosis of arthri-

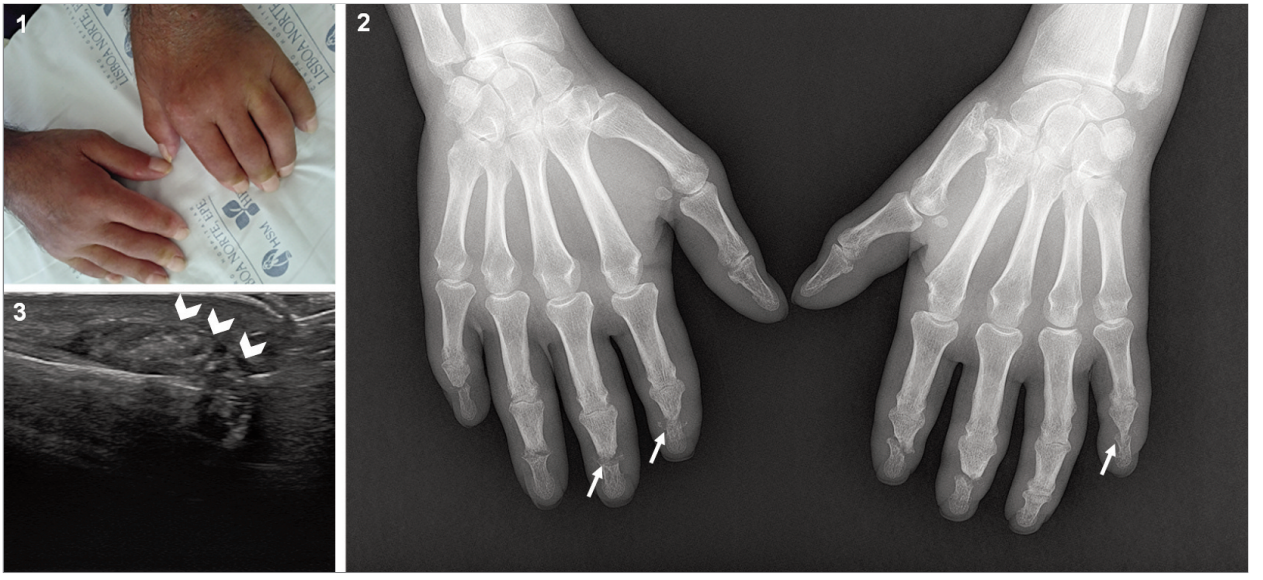

FIGURE 1. **1)** Sclerodactyly and polyarthritis of the wrists and hands; **2)** Conventional radiography of the hands showing destructive arthropathy and "pencil-in-cup" deformities (**arrows**); **3)** Right second "pencil-in-cup" distal interphalangeal joint ultrasound showing exuberant grey-scale synovitis (**arrow-heads**).

tis mutilans secondary to SSc was established. Subcutaneous tocilizumab 162mg/weekly was added to the patient's treatment regimen but withdrawn three months later due to inefficacy and severe respiratory infection ultimately leading to the patient's death.

While typically associated with psoriatic arthritis, arthritis mutilans may seldom be a feature of SSc itself[1-3]. Its prevalence remains unknown and no clinical or laboratory predictors have been identified so far[1,4]. Although some observational studies have displayed the efficacy of tocilizumab over articular manifestations[5], the optimal management of arthritis in SSc remains an unmet clinical need.

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