

Images in Rheumatology

Arthritis Mutilans in Jo1 Antisynthetase Syndrome

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Jo-1 antisynthetase syndrome (AS) is commonly associated with arthropathy, although joint erosions occur in the minority of patients. Arthritis mutilans, commonly associated with psoriatic arthritis, may rarely be seen in AS.

A 59-year-old female with a history of Jo1 antibody positive AS was referred for a second opinion on management. She

was diagnosed 6 years prior to review after developing dyspnea and cough. A high-resolution computed tomography scan of the chest showed lung involvement with bilateral mid-to-lower zone reticulations, in keeping with a nonspecific interstitial pneumonia pattern of interstitial lung disease. Her most recent lung function tests revealed a diffusing lung capacity for



Figure. Extensive osteolysis of the distal interphalangeal joints of the (A) ring, index, middle, ring, and little fingers bilaterally, (B) with involvement of the proximal interphalangeal joints and (C) involvement of the thumbs.

carbon monoxide of 55% predicted with total lung capacity 70% predicted.

The patient's joint symptoms manifested initially as stiffness, swelling, and pain of the small joints of the hands, which persisted despite treatment with mycophenolate mofetil and prednisolone. Clinically, there was dactylitis in all digits and the distal interphalangeal (DIP) joints on each digit were grossly swollen. Each distal phalanx was felt to float freely away from the joint. Radiographs of the hands showed extensive osteolysis of the DIP joints of the index, middle, and ring fingers bilaterally. Marginal erosions variably affected other small joints of the hands (Figure). A low positive anticyclic citrullinated peptide (anti-CCP; 13.0 U/mL [reference interval (RI) 0-7 U/mL]) and positive rheumatoid factor (80 IU/mL [RI 0-16 IU/mL]) were noted. Anti-CCP positivity has been previously demonstrated to be a marker of erosive arthropathy in Jo1 AS.¹

Findings were consistent with arthritis mutilans, which has been reported only extremely rarely in AS, although an erosive

arthropathy with subluxation is known to occur.¹⁻³ Pulsed monthly cyclophosphamide induction treatment with the National Institutes of Health protocol was initiated, with significant improvement of arthritis and stabilization of lung disease.

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