Erosive arthropathy in amyopathic dermatomyositis

Sirs,

Articular symptoms are reported in 25-50% patients with idiopathic inflammatory myopathies (IIM) (1,2). Arthropathy in IIM is typically symmetrical, non-deforming and non-erosive (1-4). A subluxing arthropathy, which may be associated with erosion, has been described in IIM patients with anti-synthetase antibodies (2, 4-9). We report a patient with amyopathic dermatomyositis who developed severe erosive arthropathy involving both shoulders during the course of his disease. An erosive arthropathy affecting the proximal joints, in the absence of overlapping features of other connective tissue diseases (CTDs),has hitherto not been reported in dermatomyositis.

A 45-year-old Chinese male presented in January 2001 with polyarthralgia, proximal myalgia and subacute onset of erythematosus skin rash over his face and hands. Examination revealed the typical heliotrope rash on his upper eyelids, and classic Gottron's papules on the knuckles of the hands. There was periungual vasculitis and Raynaud's phenomenon, but no proximal muscle weakness. Muscle enzymes were normal as were the electromyographic findings. The patient did not have any systemic abnormalities, sicca, dysphagia or skin thickening. There was no hyperkeratosis, scaling or fissuring of the palms and fingers (mechanic's hands). A biopsy of the lesional skin showed interface dermatitis without immunoglobulin deposits at the dermo-epidermal junction, which was compatible with dermatomyositis. Screening for common malignancies in our locality was negative. ANA, anti-dsDNA and rheumatoid factor were negative, as were anti-ENA (nRNP, Sm, Ro, La, Scl-70) antibodies on both counterimmunoelectropheresis and Western blotting. Anti-Jo-1 antibody was negative twice by immunoblotting. Chest X-ray showed minimal basal lung fibrosis; lung volumes and diffusion capacity were normal. There was no radiological evidence of erosive arthropathy of the wrists, hands and feet or periarticular calcification. There were also no overlapping features of other CTDs.

The patient was treated with topical corticosteroid and hydroxychloroquine (300 mg/day) and the skin rash gradually improved. In December 2001 he developed swelling of his right forearm and a fascial biopsy revealed eosinophilic fasciitis which responded well to prednisolone and azathioprine.

The patient remained well until March 2002 when he developed bilateral shoulder pain. Examination revealed bilateral shoulder arthritis with reduction in joint movement. The response to NSAIDs and intra-articular steroid was poor. Plain radiographs of the shoulder joints showed extensive symmetrical bony erosions of the glenoids and humeral heads. Subcutaneous calcinosis was also noted at the axillary regions. MRI of the shoulder joints confirmed severe erosive arthritis involving both gleno-humeral joints. The deltoid muscles and bone marrow of the humeral heads showed high signal intensity due to muscle and bone edema on T2-weighted images with fat saturation. Mild atrophy of the deltoid muscles was noted. Gadolinium enhanced images showed intense enhancement of the thickened synovium (Fig. 1). There was no evidence of avascular bone necrosis. As he is a chronic hepatitis B carrier with possible active hepatitis, methotrexate was not considered. Intramuscular gold therapy was given with partial response.

The arthropathy of IIM usually affects the distal peripheral joints. Hand arthritis with erosions, periosteal calcification, and interphalangeal thumb joint instability has been reported in patients with polymyositis and "overlap" features such as Raynaud's phenomenon, positive ANA, and LE cell phenomenon (8). Another study described 9 IIM patients with interstitial lung disease and inflammatory arthropathy (3). Eight had mild non-erosive arthritis while one had erosions and periarticular calcifications. The status of the anti-synthetase antibodies was unknown in these early studies.

The presence of anti-synthetase antibodies in IIM patients marks a distinct clinical subset. In a large series those with the antibodies were found to have significantly more frequent arthritis, fever, interstitial lung disease and "mechanic's hands" than those without (10); 33-57% had clinical arthritis (10). A subluxing or deforming non-erosive arthropathy has been reported in patients with the anti-Jo-1 antibody, with or without evidence of myositis (5, 9). Occasionally, the arthropathy was erosive and associated with periarticular calcification due to apatite deposition (4,6,7). Involvement of the distal joints such as the wrists and hands was usual.

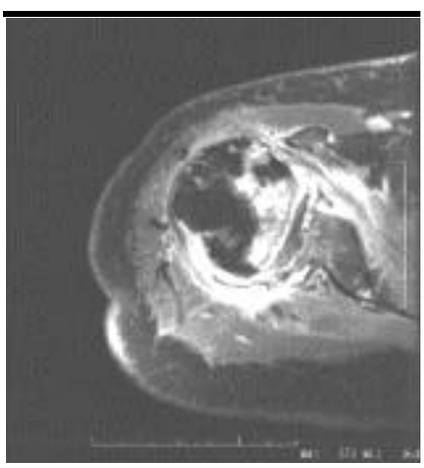

Fig. 1. Axial gadolinium enhanced image (TR 460, TE 9) of the right shoulder with fat saturation demonstrates intense enhancement in the thickened synovium. Contrast enhancement in the bone marrow of the humeral head due to inflammation can also be noted.

Our patient was unique in that he developed severe bilateral erosive arthropathy of both shoulders without involvement of other peripheral joints. He did not fulfill the ACR criteria for RA. There were no overlapping features of other rheumatic diseases that might have contributed to the articular erosion. Although his anti-Jo-1 was negative, it remains possible that other anti-synthetase antibodies such as anti-PL-7, anti-PL12 might have been present. Erosive arthropathy of the proximal joints should be recognized as a possible evolving feature in patients with dermatomyositis.

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Pulmonary and renal involvement in ankylosing spondylitis

Sirs,

Ankylosing spondylitis (AS), which is primarily a disease of the musculoskeletal system, can be accompanied by extraskeletal manifestations, but renal and pulmonary involvement are rare (1).

A 57-year-old man diagnosed with AS 30 years previously was admitted with the symptoms of dyspnea, back pain, polyuria, and swelling of the face. He had a spinal deformity and hip joint involvement. A systemic examination was normal apart from an increased expiration phase on auscultation. Laboratory investigations revealed: serum creatinine 1.6 mg/dl (normal 0.6-1.3), BUN 28 mg/dl (normal 6-20), total protein 5.8 gr/dl (normal 6.4-8.3), albumin 2.7 gr/dl (normal 3.4-5.0), erythrocyte sedimentation rate 75 mm/hr, and creatinine clearance 45 ml/min. Urinary analyses showed a density of 1008, 500 mg proteinuria, 8-10 leukocytes/hpf and 3 gr proteinuria/24 hr. Arterial blood gases were as follows: pH 7.46, PO_2 64 mm Hg, PCO_2 30 mm Hg, and oxygen saturation 94%. In pulmonary function tests (PFT) FVC was 3.16 (66%),FEV1 2.58 (68%),FEV1/FVC 81%, and there was a restrictive type ventilatory pattern. On chest x-ray there were bilateral interstitial infiltrative lesions in the middle and lower zones of the lung parenchyma. A Ppd test was done and found to be negative, as was sputum cytology repeated 3 times. High resolution computed tomography (HRCT) revealed a bilateral ground glass appearance in the lung parenchyma (Fig. 1). On renal ultrasound the dimensions of the left kidney were 144 x 64 mm,the thickness of the parenchyma was 15 mm,and the parenchymal echo was interpreted to be grade 2. In flow patterns obtained from the interlober arteries, RI was 0.69 in the spectral analysis, which was compatible with a renal parenchymal disease. A biopsy was scheduled but refused by the patient. Therefore a rectal biopsy was performed for possible amyloidosis but was negative. Serum IgA was 2.23 g/l (normal 0.7-4.0). The patient was started on pulse steroid therapy and sulphasalazine 2 gr/day.

Lung involvement occurs in 1% of AS patients approximately 20 years after onset of the disease. Although upper lobe fibrotic and infiltrative lesions are characteristic, in our patient chest x-rays showed middle and lower lobe infiltration (2, 3). There was no other explanation other than AS (such as methotrexate administration or occupational lung disease) for this finding.

There was a slight to moderate decrease in the patient's PFT, specifically the vital capacity and total lung capacity denoting a restrictive type ventilatory defect. Despite diminished thoracic mobility, a significant deterioration in PFTs is not usual in AS due to diaphragmatic compensation, as in our patient. We detected a ground glass appearance which is a strong evidence of interstitial lung disease in HRCT.

In AS, possible causes of renal dysfunction include; secondary amyloidosis, IgA and analgesic nephropathy (4). Regarding the absence of gross hematuria, a normal IgA level and proteinuria of 3 gr/24 hr, the possibility of IgA nephropathy was ruled out in our patient. In cases with analgesic nephropathy, there is usually colic type pain due to papillary necrosis, a sterile pyuria and proteinuria < 1 gr/24 hr, so we also ruled out analgesic nephropathy.

The incidence of renal amiloidosis in AS is reported to be 1-3% (5, 6). Proteinuria or nephrotic syndrome is the most common clinical manifestation where a biopsy should be performed (7), although unfortunately we could not do so as our patient refused. We were able to evaluate the rectal biopsy specimens, which were amyloid negative. However rectal biopsy has been proven to be positive in only 65% of systemic amyloidosis cases.

Here we present a case of AS with both renal and pulmonary involvement, which are rare clinical presentations. Extra-skeletal manifestations should be investigated in patients with longstanding AS in the presence of new symptoms.

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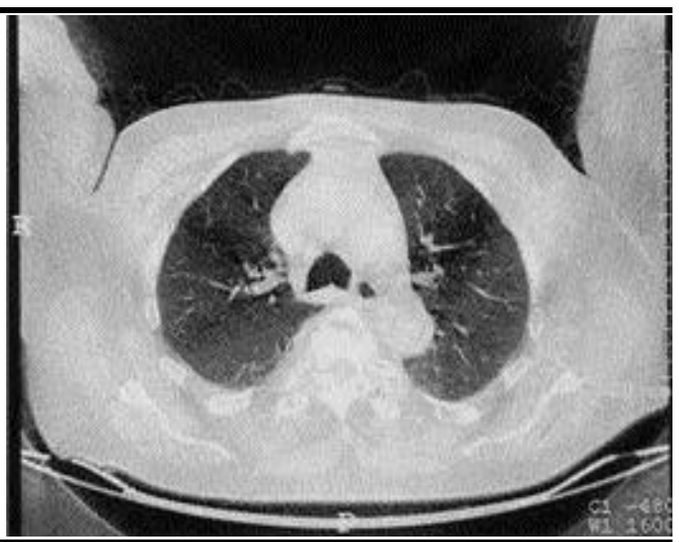

Fig. 1. Ground glass appearance on HRCT.