Bilateral, asymptomatic scaly and fissured cutaneous lesions of the fingers in a patient presenting with myositis

A 69-year-old woman was admitted because of sudden onset of fever and symmetrical arthralgias, predominantly in the upper limbs. She also had global muscle weakness and wasting, as well as dyspnea and loss of appetite. On clinical examination, active mobilization of the joints was impossible, except for the elbows, hands and wrists. On thoracic auscultation slight bilateral crackles were revealed over the lower chest. Unusual cutaneous findings were noted, consisting of scaling and fissuring on the lateral sides of the second and third finger of both hands, a picture similar to that produced by manual labor (Figure 1). The dermatological examination was otherwise unremarkable. A skin biopsy specimen was obtained from an involved finger and stained with Hematoxylin-Eosin (Figures 2). Histological examination showed an interface psoriasiform dermatitis consisting of parakeratotic hyperkeratosis, epidermal hyperplasia, necrotic keratinocytes with focal exocytosis and vacuolar degeneration of the basal layer, perivascular lymphocytic infiltrate and telangiectasias. ESR was 18 mm in the first hour. Muscle enzymes were raised: creatine kinase 3712 U/l (normal range 15-210), aspartate transaminase 108 U/l (normal range 5-44), alanine aminotransferase 114 U/l (normal range 5-48), lactate dehydrogenase 1848 U/l (normal range 230-460), aldolase 40 U/l (normal range 2-7.6).

WHAT IS YOUR DIAGNOSIS?
Diagnosis: Anti-synthetase syndrome (ASS) with mechanic’s hand

Immunological tests revealed the presence of anti-extractable nuclear antigen (ENA) antibodies with anti-Jo-1 specificity. Electromyography showed small amplitude, brief actions potentials and increased insertional activity was consistent with irritable myopathy. A muscle biopsy was not performed. Lung disease was present with interstitial lesions on CT scans and a pulmonary function test showing an obstructive disease. The following tests were normal or negative: urinalysis, serum creatinine, rheumatoid factor, antinuclear antibodies, anti-dsDNA, ANCA and anticardiolipin antibodies, cryoglobulins, nailfold capillary microscopy and echocardiography. Search for an occult neoplasm was negative.

A high dose of corticosteroids, methylprednisolone 2.5 mg/kg/day, was required and cyclophosphamide 2 mg/kg/day was added for the presence of an interstitial lung disease. Myalgias and muscle weakness improved significantly as the cutaneous lesions gradually disappeared.

DISCUSSION

Antibodies binding to aminoacyl-tRNA synthetases have been identified in the serum of 25% to 40% of adults with inflammatory myopathies.[1] The first to be discovered and the commonest of these antibodies, anti-Jo-1, (in 15% to 30% of adults with polymyositis or dermatomyositis) is directed against histidyl t-RNA synthetase.[2] At least four other anti-synthetase antibodies have been described.[1] The anti-synthetase antibodies are specific for a clinically distinctive syndrome characterized by myositis along with non-erosive arthritis, interstitial lung disease, skin changes (Mechanic’s hand, Raynaud’s phenomenon) and fever.[3] Isolated clinical manifestations such as interstitial lung disease have been reported in about 10% of cases.[4] Compared to classic dermatomyositis (DM), in which such reported similar lesions were sporadically described in association with other skin manifestations, mechanic’s hand seems to be a distinctive feature of the ASS,[5] present in up to 70% of patients,[6] though sporadically reported in classic DM in association with other cutaneous findings.[7]

Light microscopy findings[7] are similar to those observed in DM. In the present case, the distinguishing cutaneous feature is the presence of mechanic’s hand in the absence of any other skin lesion generally observed in typical DM (heliotrope rash, malar rash and Gottron’s papules). The differential diagnosis includes irritant contact dermatitis, tinea manuum and dry dishyrdotic eczema. Since myositis and cutaneous manifestations may not follow a parallel course, the finding of mechanic’s hand should prompt a search for anti-synthetase antibodies and may serve as a clinical diagnostic aid in patients with an underlying idiopathic inflammatory myopathy.

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