Mechanic's hands and hiker's feet in antisynthetase syndrome

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A previously healthy 64-year-old man presented to hospital after five weeks of progressive shortness of breath on exertion, proximal muscle weakness, symmetric polyarthralgias in the hands, and thickening of the skin over both palms and soles (Figure 1). He had substantial hyperkeratosis over his palms and soles, particularly on the distal and lateral aspects of the phalanges (Figure 1), and a diffuse erythematous, nonpruritic macular rash in a shawl distribution on his upper chest. He had 4/5 strength in his hip flexors and deltoids.

The patient required high-flow oxygen and was admitted to the intensive care unit. High-resolution computed tomography and bronchoscopy suggested interstitial lung disease. Further investigation showed that he had elevated serum levels of creatinine kinase (4100 [normal range 57–208] U/L) and C-reactive protein (55 [normal range 0–1] mg/L), and an elevated erythrocyte sedimentation rate of 55 (normal range less than 15) mm/h. We included dermatomyositis, polymyositis, antisynthetase syndrome, scleroderma, systemic lupus erythematosus and mixed connective tissue disease in the differential diagnosis. This patient's presentation was typical of antisynthetase syndrome, which was confirmed by a positive test result for an antisynthetase antibody, anti-Jo-1.

Antisynthetase syndrome is an uncommon idiopathic inflammatory muscle disease characterized by a constellation of symptoms, including hyperkeratosis ("mechanic's hands"), interstitial lung disease, myositis, polyarthralgia, fever and Raynaud phenomenon in patients with a positive test result for antisynthetase antibodies. In one study, 17 of the 18 patients were women, with a mean age of onset of 39 years.[2] Several antibodies have been found to be associated with antisynthetase syndrome, of which anti-Jo-1 is the most common.[1,2] It is a heterogeneous disease, and the clinical manifestations can be linked to the specific antisynthetase antibody involved.[1] The term "mechanic's hands" describes hyperkeratosis over the palms predominantly in the distal and lateral aspects of the phalanges, and about 90% of cases will have similar findings on the soles ("hiker's feet").[3] A good history will differentiate pathologic from physiologic hyperkeratosis.

Treatment of antisynthetase syndrome usually includes high-dose corticosteroids for at least four weeks with a gradual taper. If long-term treatment is required, cyclophosphamide, azathioprine, mycophenolate mofetil and rituximab are commonly used.[1,2] Although prognosis has not been well studied, antisynthetase syndrome is believed to be a chronic disease. One study found that 87% of patients experienced disease relapse.[2] Our patient was treated with high-dose corticosteroids. Within three days, the arthralgias and rash resolved, and both the hypoxia and hyperkeratosis improved. A corticosteroid taper and an outpatient referral to rheumatology for consideration of immunomodulatory therapy were planned.

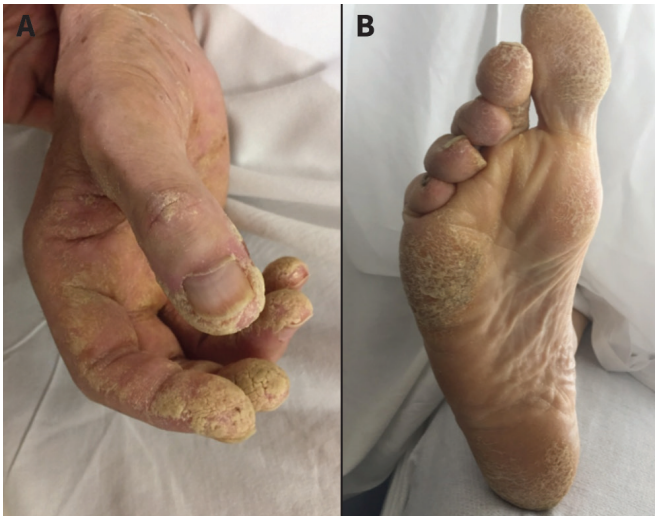

Figure 1: A 64-year-old man with anti-Jo-1-positive antisynthetase syndrome. Hyperkeratosis visible on (A) his right hand, consistent with "mechanic's hands" and (B) his right foot, consistent with "hiker's feet."

References

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