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CASE REPORT

Leprosy mimicking lupus erythematosus

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ABSTRACT

Leprosy, a contagious and chronic granulomatous disease caused by *Mycobacterium leprae*, is classically known to have cutaneous and neurologic sequelae. Leprosy usually has a long incubation period and may manifest with a variety of autoimmune phenomena reminiscent of autoimmune diseases, such as systemic lupus erythematosus (SLE) or rheumatoid arthritis. We describe a case of a 40-year-old man presenting with a long history of recurrent skin rashes and hand numbness, initially diagnosed as carpal tunnel syndrome and SLE, who was later proven to have borderline leprosy. This suggests that this underappreciated disease should still be considered in the differential diagnosis of granulomatous skin rashes with rheumatic manifestations, even in nonendemic regions.

Introduction

Leprosy, also known as Hansen's disease, is a chronic granulomatous disease affecting mainly the skin and nerves caused by *Mycobacterium leprae*. The course and clinical manifestations of the disease are largely dependent on an individual's immune response to *M. leprae* and can be roughly classified into tuberculoid, borderline, and lepromatous forms. It has long been known that numerous clinical and serological similarities exist between patients with leprosy and connective tissue diseases. These varying clinical manifestations, as well as a tendency of the disease to have a protracted course, often lead to a delay in early recognition and diagnostic confusion. We report a patient with borderline leprosy who had initially been misdiagnosed for several years.

Case report

A 40-year-old Myanmar-born man, living in Taiwan for 25 years, presented to our hospital for acute appendicitis. During the admission period, an in-patient dermatology consultation was called to evaluate skin lesions and joint deformities. According to the patient, he had suffered from bilateral hand and wrist pain, swelling, and deformities for 4 years. Worsening fingertip tingling, numbness, and pallor, especially during cold weather, were noticed around the same time. Decreased thermal sensation on the left palm and fingers ensued, which had resulted in burn injury several times. He had received open carpal tunnel release surgery twice 4 years earlier, once at the elbow and the other time at the wrist, but the symptoms still progressed. In addition, a malar rash was observed at that time, which had become even worse after sun exposure. Laboratory findings at that time showed positive antinuclear antibody [ANA; 1:1280 (+), speckled pattern] and positive antiribonucleoprotein antibody (9.9 U/mL; normal value <5.0 U/mL). He was previously diagnosed with systemic lupus erythematosus (SLE) at a regional hospital 3 years earlier based on four of the 11 American Rheumatologic Association criteria for the diagnosis of SLE (malar rash, photosensitivity, arthritis, positive ANA test). Treatment with prednisolone 20 mg daily and hydroxychloroquine 200 mg daily were initiated, followed shortly by the addition of cyclophosphamide 50 mg daily to control his disease. However, the therapy did not control the symptoms, and skin lesions spread to trunk and limbs in these 3 years. This patient had stopped using the immunosuppressive agents 1 year before presentation.

On examination, the patient had generalized erythematous annular patches and plaques with ill-defined borders on the face, trunk, and forelimbs (Figure 1). Hand swelling, muscle wasting, claw deformity of fingers, and drop hand were observed evidently in the left hand (Figure 2). Neurologic examination at these areas showed significant sensory loss (touch, pain, and temperature sensation) and muscle weakness.

Laboratory examination showed reduced hemoglobulin (94 g/L) with increased reticulocyte production index (7.0%), a normal white

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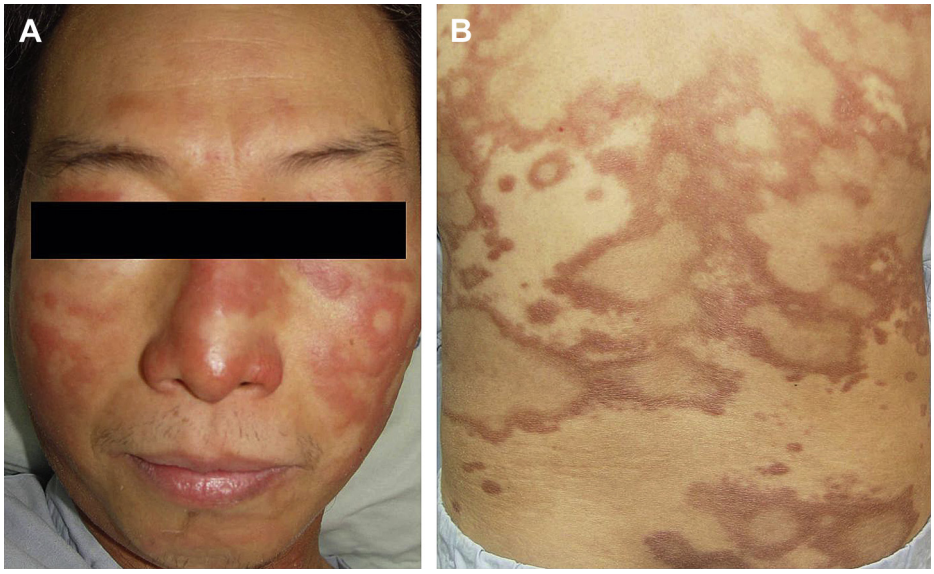

Figure 1 Generalized erythematous annular patches and plaques with ill-defined borders on the (A) face and (B) back.

blood cell count, normal liver and kidney function tests, and elevated erythrocyte sedimentation rate (71 mm in the first hour; normal value <12 mm/hour). His autoimmune profile revealed seropositivity for ANA [1:160 (+), speckled pattern], but other data were within the normal limit, including complement fractions 3 and 4 (C3 and C4), antidouble strand DNA antibody, anticardiolipin antibody screen, extractable nuclear antigen autoantibodies screen, anti-Smith, antiribonucleoprotein, anti-Sjogren's syndrome A antibody (anti-SSA), and anti-Sjogren's syndrome B antibody (anti-SSB). This patient denied having other systemic disease, recent travel history, smoking, or substance abuse. The initial impression was subacute cutaneous lupus erythematous with a sensorimotor polyneuropathy. A skin biopsy from the right forearm revealed diffuse granulomatous infiltrate in the reticular dermis (Figure 3A,B) composed of histiocytes, plasma cells, a few lymphocytes, and multinucleated giant cells. Some globi were also noted in the granuloma. An acid fast stain showed an aggregate of bacilli within the globi in keeping with lepromatous leprosy (Figure 3C,D). The direct immunofluorescence study was all negative. Slit-skin smears from bilateral forehead and earlobes showed an average bacterial index of two. On nerve conduction studies, a decrease in motor and sensory conduction velocity combined with a prolonged motor distal latency and a decreased compound muscle action potential were noted in the left ulnar and radial nerve. These findings suggested sensorimotor polyneuropathy superimposed with multiple entrapment neuropathies.

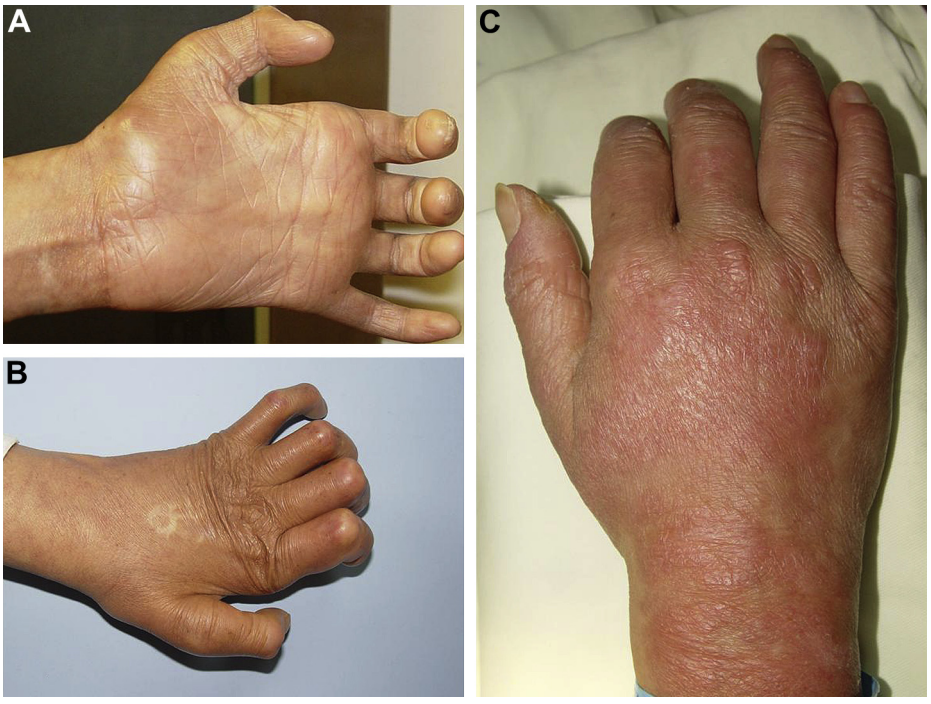

Figure 2 Bilateral hand lesions of our patient: (A,B) Muscle wasting and claw deformity on the left hand; (C) erythematous swelling on the right hand.