

## Case Report

## Lepromatous leprosy mimicking systemic lupus erythematosus: a case report

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**ABSTRACT**

The clinical manifestations of leprosy are highly variable, and the disease is notorious for being “a great imitator” of several other conditions. Leprosy may manifest with a variety of phenomena resembling those of autoimmune diseases. Herein, we report a 33-year-old male presenting with wounds on his left leg and hyperpigmented skin lesions all over his body. Six years earlier, the patient was diagnosed with systemic lupus erythematosus (SLE). However, therapy for SLE did not control his symptoms; instead, the patient developed features of leprosy, such as anesthetic skin lesions, nerve enlargement, and tenderness. Tests for antinuclear antibodies and anti-double stranded DNA antibodies were negative. Slit-skin smear showed a bacterial index of 6+ and morphological index of 10 %. Lupus band test results were negative. Histological findings were compatible with lepromatous leprosy. The clinical and serological similarities between leprosy and SLE may lead to erroneous diagnosis. Thus, clinicians should be aware of this characteristic for correct diagnosis.

**KEYWORDS** lepromatous leprosy, systemic lupus erythematosus

Leprosy, or Hansen’s disease (HD), is a chronic granulomatous disease arising from infection of the acid-fast bacilli, *Mycobacterium leprae*.<sup>1-3</sup> Leprosy is classically notorious for resulting in cutaneous and neurologic sequelae. As the clinical manifestations of leprosy are remarkably variable, the disease is famous for being a “great imitator” of several other conditions. Leprosy may easily be misdiagnosed as one of many diseases.<sup>1,4</sup> The bacterium has a long incubation period, and the disease may manifest with a variety of phenomena typical of autoimmune diseases, such as systemic lupus erythematosus (SLE) or rheumatoid arthritis.<sup>2</sup>

The clinical manifestations of HD are highly related to the host’s immune response to *M. leprae*

and can be classified into three forms, namely, tuberculoid, borderline, and lepromatous. Leprosy reactions involve T-cell reactivity to mycobacterial antigens, leading to spontaneous alterations in its clinical features.<sup>5,6</sup> This feature is particularly due to the disease’s unique and unconventional presentations and may lead to initial misdiagnosis and delayed treatment. Therefore, leprosy is frequently diagnosed at advanced stages, often with severe and extensive collateral damage.<sup>1</sup>

Herein, we report a leprosy case with the unusual clinical presentation of hyperpigmented patches over nearly the whole body. Clinical findings, such as malar rash, photosensitivity, arthritis, and lymphopenia, led to an initial diagnosis of SLE and, subsequently,

several years of SLE therapy. History taking, clinical examination, laboratory results, and histopathology examination proved the patient's condition to be HD, specifically lepromatous leprosy. The present case is unique for its unusual clinical presentation resembling that of SLE and leading to a delay in diagnosis and proper therapy. The patient has given the informed consent for the publication.

### Case report

A 33-year-old male, living for years in Demak, Central Java, an endemic region for leprosy, presented to the dermatology clinic of Dr. Sardjito Hospital with a wound on his left leg and brown skin lesions all over his face and body. Six years before this hospital visit, the patient reported reddish skin lesions over his face with a history of worsening redness and burning sensation after sun exposure. He also complained of pain on his leg. These initial clinical findings (i.e., malar rash, photosensitivity, and arthritis) are compatible with the clinical presentation of SLE, and the patient was diagnosed and treated for SLE with 32 mg/day methylprednisolone by a state hospital. This therapy failed to control symptoms, and the skin lesions spread to the patient's trunk and limbs over the next 3 years.

Three years before his hospital visit, the patient noticed a butterfly rash-like redness on his face that eventually spread to his trunk and limbs. This lesion became hyperpigmented, and the patient developed alopecia on his eyebrows and sensory loss in his legs. The patient was diagnosed with SLE at another regional hospital. However, after treatment, his arthritis and skin lesions recurred, and his neuropathic pain worsened. The patient treated himself with 8 mg/day methylprednisolone for the last 2 years.

Three weeks before hospital visit, he noticed wounds spreading over his left leg, and developed a fever. His previous complaints persisted. The patient presented to the Department of Internal Medicine of Dr. Sardjito Hospital and was clinically diagnosed with SLE once more; at this time, the patient complained of a malar rash, photosensitivity, arthritis, and leg ulcers, which was diagnosed as traumatic ulcers. Lymphopenia was also observed. The patient was treated with the SLE protocol therapy, including 16 mg of methylprednisolone and 160 mg of chloroquine daily, along with wound care using gentamycin ointment. The clinical appearance of a widespread hyperpigmented patch all over the patient's body led

a dermatology consultation for skin biopsy to support the diagnosis of cutaneous manifestation of SLE.

The patient denied the presence of other systemic diseases and any similar complaints in his family. His last travel was to East Kalimantan in 2006. While no history of familial cases of leprosy and SLE was reported, the region in which the patient resided is considered to be endemic for leprosy. The patient's overall physical status was good, and he was *compos mentis*. All vital signs were within normal ranges, and no lymph node enlargements were found.

Erythematous and hyperpigmented patches were observed on the patient's face. The patient reported a history of worsening redness and burning sensation upon sun exposure. These symptoms highly resemble the clinical appearance of a malar rash and photosensitivity, which are characteristic of SLE. However, further dermatological examination revealed erythematous and hyperpigmented patches all over the patient's face, not only the malar area. More importantly, madarosis (loss of the eyebrows) and solid infiltrate found on both earlobes were found, leading to our suspicion of leprosy (Figure 1a). Generalized erythematous and hyperpigmented patches with ill-defined borders all over the trunk and forelimbs and muscle wasting (thenar and hypothenar atrophy) were also present (Figure 1b). Multiple ulcers with granulated tissue at the base, ill-defined borders, and no secretion appeared on the legs (Figure 1c).

Further physical examination revealed significant sensory loss (e.g., touch, pain, and temperature sensation) over the skin lesions, as well as muscle weakness and thickening of peripheral nerves (ulnar nerve). Sensory and motor examination of the extremities demonstrated anesthesia on the tibialis posterior nerve and weakness on the peroneus communis nerve.

Differential diagnoses in this case included lepromatous leprosy and SLE. Based on history taking and clinical examination, a working diagnosis of lepromatous leprosy was made. Laboratory results showed leukocytosis (14–16 cell/ml) and lymphopenia (7%), with relatively normal liver and kidney functions. The patient's autoimmune profile revealed seronegativity for antinuclear antibodies (ANA), and other data were within normal limits, including anti-double stranded DNA (anti-dsDNA) antibodies. Direct immunofluorescence (IF) antibody staining revealed a negative lupus band test result (Figure



**Figure 1.** (a–c) Initial clinical features of the patient upon first presentation to the hospital; (d–f) Marked improvements after prompt leprosy treatment and 6 months following release from treatment

2a). Histological findings were compatible with HD, specifically lepromatous leprosy (Figure 2, b and c). Slit-skin smear for leprosy acid-fast bacilli from both arms and earlobes showed an average bacterial index of 6+ and morphological index of 10% (Figure 2d).

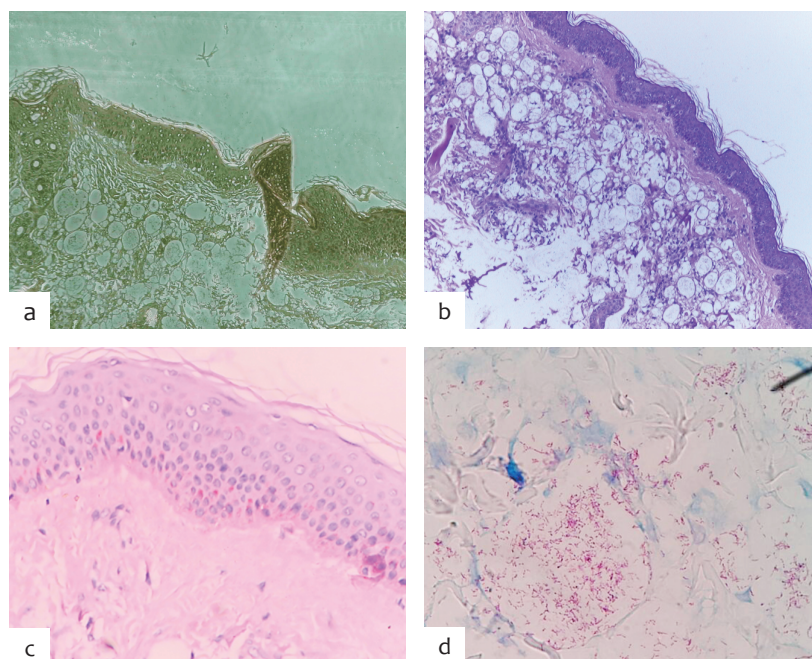
From history taking, clinical examination, laboratory results, and histological findings, a working diagnosis of lepromatous leprosy was made. The patient was treated with a multibacillary regimen for leprosy with three types of drugs (i.e., oral pulse of 600 mg of rifampicin and 300 mg of clofazimine once every 28 days and daily oral intake of 50 mg of clofazimine with 100 mg of dapson). This treatment lasted for 12 months. The patient was also treated with topical gentamycin cream twice a day for ulcers and supplemented with multivitamins. Symptoms of ulcer and arthritis were markedly improved within 1 month. Photographs showing marked improvements after prompt treatment and 6 months of follow up are shown in Figures 1d–1f.

## DISCUSSION

Leprosy or HD is a chronic granulomatous disease caused by infection with *M. leprae*. *M. leprae* is an acid-

alcohol fast obligate intracellular *Bacillus* with a tropism for peripheral nerves and reticuloendothelial cells. Thus, the *Bacillus* primarily affects peripheral nerves and skin.<sup>4,7</sup> As HD can imitate several diseases, diagnosis is especially challenging for patients with several clinical and serological rheumatic manifestations, particularly those residing in nonendemic regions.<sup>1</sup> Due to its diverse clinical manifestations (e.g., dermatological and neurological), leprosy can be confused with many other systemic autoimmune diseases, including SLE.

SLE is an autoimmune disease that poses an escalating susceptibility to infections and may trigger reactivation.<sup>7</sup> The American College of Rheumatology (ACR) classification criteria for SLE present sensitivity of 71%–96% and specificity of 90%–100%. These criteria are used in daily practice, and SLE is defined when a patient manifests four or more of the 11 conditions indicated.<sup>1</sup> Our patient showed a malar rash, photosensitivity, arthritis, and lymphopenia, leading to a diagnosis of SLE for 6 years. Sudden eruption of erythematous plaques on the face and body associated with photosensitivity in our patient was initially suggested as a symptom of lupus erythematosus. While the differential diagnosis of this case was mainly SLE, we excluded this diagnosis



**Figure 2.** (a) Negative result from a direct immunofluorescence study (lupus band test); (b) Hematoxylin-eosin staining of a skin biopsy specimen demonstrating epidermal orthokeratosis, focal epidermal atrophy, flattened rete ridges, and hypermelanization of basal cells. Dermal grenz zones with granulomas consisting mostly of foamy cells were observed. Lymphocyte infiltration within the papillary dermis, as well as the surrounding vascular and appendicular structures, was also found. Langhans cells were not found; (c) Thickening of the basal membrane zone was not found after periodic acid–Schiff staining; (d) Acid-fast bacilli were found in abundance after Fite-Faraco staining. The bacteria were widely spread and clumped into multiple globules

based on the American Rheumatism Association (ARA) criteria. Hyperpigmented lesions on the cheek, initially considered as a malar rash, were absent on the nasal bridge. Thus, these lesions were not a malar rash after all.

Certain manifestations of leprosy, such as anesthetic skin lesions, nerve enlargement, and nerve tenderness, should alert physicians. We propose that, due to the many similarities between SLE and leprosy, clinicians should be more aware of these diseases and conduct through physical and laboratory examinations to distinguish them; skin biopsy is also recommended. Misdiagnosis often leads to years of corticosteroid or immunosuppressant therapy, which may modify and worsen the clinical course of leprosy toward the lepromatous classification.<sup>2</sup> The clinical manifestations and course of HD highly depend on an individual's immune response to *M. leprae*, and a patient's T-cell immunity has a crucial protective role.<sup>1,8</sup>

The clinical presentations of leprosy vary and may include erythema nodosum, arthritis, fever, skin erythema, vasculitis, epididymitis, glomerulonephritis, pericarditis, and pleuritis.<sup>9</sup> The patient in this case showed generalized erythematous and hyperpigmented ill-defined patches all over his

face, trunk, and forelimbs. Physical, sensory, and motor examination results were compatible with the clinical finding of leprosy. Use of immunosuppression therapy by the patient probably led to a downgrading reaction, which often results in irreversible nerve damage.<sup>2</sup>

In the present case, the diagnosis of SLE was eventually excluded due to the patient's lack of response to immunosuppressant therapy and negative ANA and anti-dsDNA results. The presence of these autoantibodies may be related to chronically increased cell destruction and the release of sequestered antigens on account of tissue injury. Auto-antibody profiles and direct IF study results did not support the diagnosis of lupus. Circulating antibodies to DNA are almost always present in active disease and may occur without antinuclear factors.<sup>10</sup> IF assay or enzyme immunoassay can detect ANA.<sup>10</sup> A direct IF study showed a negative lupus band test result. Thus, we can exclude the differential diagnosis of SLE.

Considering our findings, clinicians should always include leprosy in their differential diagnosis of SLE and/or antiphospholipid syndrome with unconventional presentations, particularly in leprosy-endemic

areas. We also recommend targeted biopsy. Joint complications resembling rheumatoid arthritis may occur in leprosy.<sup>9</sup>

Osteoarticular manifestations are the third most common complication after skin and peripheral nerve system involvement.<sup>19</sup> Diagnosis can be established through full-depth skin or nerve biopsy smears.<sup>9</sup> Patients with the LL classification have very high bacillary loads and appear with many diffuse skin lesions without loss of nerve sensation.<sup>6</sup> The patient's slit-skin smear and histological findings were compatible with HD. Thus, a working diagnosis of lepromatous leprosy was made in accordance with history taking, clinical examination, laboratory results, and histological findings.

### Conclusions

HD is a great imitator of several other conditions, and diagnosing the disease is particularly challenging for nonendemic regions and patients who clinically and serologically present with some rheumatic manifestations. Our case shows an unusual presentation of leprosy initially diagnosed as SLE. We established a diagnosis of leprosy by history taking, clinical examination, laboratory results, and histological findings, all of which supported the diagnosis of HD, specifically lepromatous leprosy. Differential diagnosis of SLE was excluded via the diagnostic ARA criteria. Clinicians must be made aware of HD to enable its correct diagnosis and proper treatment.

### Conflict of Interest

The authors affirm no conflict of interest in this study.

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