

## CASE REPORT

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# Erythema nodosum leprosum as presenting feature of lepromatous leprosy

Marilyn Arosemena, Carlos I. Pacheco, Ankitha Lingamaneni, Ankur Aneja

## ABSTRACT

**Leprosy is known to be a great mimicker and can present with a wide variety of clinical signs and symptoms. Erythema nodosum leprosum mostly occurs after treatment but rarely can develop as the initial feature of the disease. We report a case of lepromatous leprosy presenting as systemic painful maculopapular rash. This case illustrates the challenges in diagnosing leprosy in a non-endemic country.**

**Keywords:** Erythema nodosum leprosum, Lepromatous leprosy, Maculopapular rash

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## INTRODUCTION

Leprosy is an infectious chronic granulomatous disease caused by the acid-fast bacillus *Mycobacterium leprae*. It is known to be a great mimicker of rheumatologic disease and can present with a wide variety of clinical signs and symptoms resembling rheumatoid arthritis (RA), seronegative spondyloarthropathies (SA), lupus, or even systemic necrotizing vasculitis. The lack of knowledge about this condition in countries where the disease is considered eradicated, contributes to late diagnosis [1, 2].

Leprosy can be classified as tuberculoid, borderline tuberculoid, mid-borderline, borderline lepromatous, and lepromatous. Reactions are divided into type 1 reaction (or reversal reaction, RR), type 2 reaction, or Erythema nodosum leprosum. Erythema nodosum leprosum (ENL) is a humoral response that leads to inflammatory and painful nodules that result in nerve and organ damage. ENL occurs most commonly during the first year of multidrug therapy, but rarely can occur as an initial presentation. Signs and symptoms include bright red raised, evanescent plaques of varying sizes, fever, neuritis, arthritis, epididymo-orchitis and iridocyclitis, all which lead to a misdiagnosis of this entity with other rheumatologic diseases [3, 4].

Although the numbers of new cases detected globally have been declining, this condition still remains a major problem in some developing countries. Furthermore, reported cases of this disease amongst immigrants in developed nations have been on the rise. The annual leprosy statistics in 2015 from 121 countries indicate that a small number of leprosy cases still exist in about 12 countries. Because of the aforementioned resemblance to rheumatologic conditions, lepromatous leprosy presenting with polyarthritis, muscle weakness and diffuse maculopapular rash can be very challenging for physicians to diagnose, especially in a developed nation with low incidence. We report a case of lepromatous leprosy presenting with painful maculopapular rash and polyarthritis [1, 2].

## CASE REPORT

A 42-year-old Hispanic male presented to the hospital with a 2-year history of remitting and relapsing fever, diffuse maculopapular rash, and persistent generalized pain. Upon arrival, his vital signs were stable except for a fever of 39.3 C. He was alert and oriented, but complained of severe numbness and tingling in his hands and feet, chills, and night sweats accompanied by a diffuse, non-pruritic, painful maculopapular rash. Of note, he had been diagnosed with dermatomyositis four years ago given the constellation of findings. His medication history only consisted of prednisone 20 mg daily in order to manage his 'dermatomyositis'. He reported similar episodes over the past two years and required an average of six hospitalizations per year to manage his symptoms. He characterized his pain as a burning sensation 8/10 in severity located across all of his joints, bones, and muscles that improved transiently with prednisone and morphine but was exacerbated by palpation and maintaining the same position for an extended period of time. His pain was accompanied by dyspnea and episodic vision loss 2–3 times per day that initially began as blurriness and progressed to excessive tear production and complete vision loss. These episodes would last 2-3 minutes and spontaneously resolve. On physical examination, the patient was in moderate distress and was found to have madarosis of bilateral eyebrows, sparse eyelashes, and ear helix with several non-tender flesh-colored and light brown papules (Figure 1). Numerous painful red indurated dermal papules and nodules were noted on his arms, forearms, lower abdomen, and upper back (Figure 2). Additionally, he had non-pitting edema in both hands, and his left foot had an overlying fourth metatarsal digit with a hyperkeratotic plantar plaque of approximately 1 x1.5 cm (Figure 3).

Due to his previous diagnosis of dermatomyositis, the patient was initially continued on his prednisone dose of 20 mg daily along with medication to manage pain. During his hospitalization, he had laboratory tests performed and was found to have leukocytosis, microcytic hypochromic anemia with iron level <10 mcg/dl, C-Reactive Protein 22.4 mg/dl, erythrocyte sedimentation rate 63 mm/Hr, elevated aldolase at 20.8 U/L, and a creatine phosphokinase (CPK) within normal limits at 49 units/L. Special immunology laboratory tests were sent and returned with negative results for ANA, Anti Jo 1, and Rheumatoid Factor. Given the patient's poor response to prednisone, absence of typical dermatomyositis manifestations of heliotrope rash, shawl sign and gottron papules, normal CPK, and negative antibodies, dermatology service was consulted for further work up. The dermatology service reported palpable enlarged ulnar nerves bilaterally and performed biopsy of the right ear lobe, right temple, and right chest. Biopsy showed extensive leukocytoclastic vasculitis with fibrinoid degeneration, pandemic perivascular and interstitial leukocytoclastic reactions with histiocytic infiltrates, focal



Figure 1(A and B): (A) Madarosis of bilateral eyebrows, sparse eyelashes. (B) Ear helix with several non-tender flesh-colored and light brown papules.

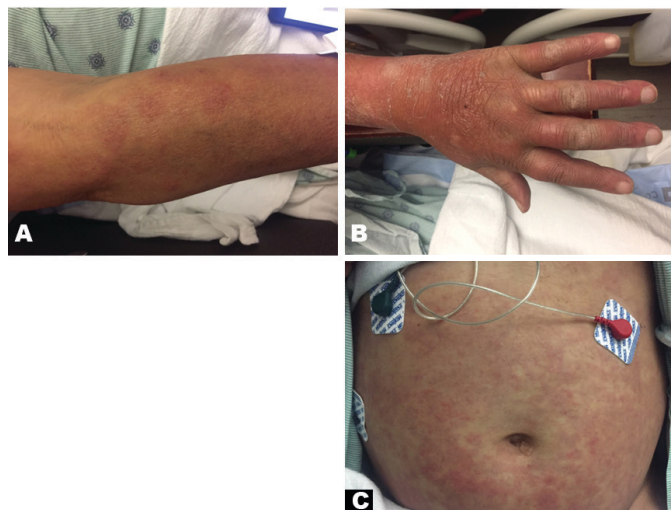


Figure 2(A–C): Red indurated dermal papules and nodules on (A) forearms, (B) hands and (C) abdomen.



Figure 3: Left foot with overlying 4th metatarsal digit.

involvement into the subcutis, prominent papillary edema and large number of acid-fast *M. leprae* bacilli within small vessels highlighted with FITE stain, histiocytes and interstitium. The patient was started on prednisone, dapsone and minocycline for disease management as well as morphine and gabapentin to control neuropathic pain. When the ophthalmology service was consulted for blurry vision, the patient was diagnosed with keratitis and given the appropriate medication. His painful maculopapular rash was not under control, so methotrexate and rifampin were subsequently added to the disease-modifying regimen, which provided some relief. The patient was then discharged to a skilled nursing facility to continue physical therapy.

## DISCUSSION

Leprosy is a great mimicker of several rheumatologic diseases, and as a result, can be very challenging for primary physicians to diagnose [3]. This patient had been previously diagnosed with dermatomyositis. However, his diffuse maculopapular rash and pain did not respond to corticosteroids, whereas rheumatologic diseases typically respond to steroid therapy. The absence of gottron papules, shawl's sign and heliotrope rash, negative antibodies and a normal CPK level decreases the likelihood of dermatomyositis in this setting. Establishing the true diagnosis of leprosy was complicated because the patient had atypical findings for leprosy, including painful maculopapular rash with no lepromas or hypoesthetic plaques. All findings were secondary to a rare presentation of leprosy known as ENL, which generally occurs in lepromatous leprosy cases and rarely in borderline cases. Young patients with high bacillary index and skin infiltration like the patient presented above are more prone to developing these reactions [5].

Although a vast majority of leprosy cases present with dermatologic and neurologic features, musculoskeletal manifestations are also quite common. Skin findings vary from macules to hypopigmented or hypoesthetic plaques and nodules, while neurologic manifestations can range from mononeuropathy to mononeuritis multiplex and distal symmetric polyneuropathy. The prevalence of rheumatic features in leprosy varies across case series, ranging from 1–2% described in large dermatology series to 60–80% in cases from rheumatology clinics. Charcot's arthropathy is the classic rheumatologic manifestation of leprosy described in textbooks. It is usually found in longstanding cases with neurologic involvement; findings range from subluxation, dislocation, or pathologic fractures to complete joint destruction. In this case, the patient had an overlying fourth metatarsal digit [6, 7].

Leprosy may also mimic vasculitis, especially in cases with high bacillary load in the vascular endothelium. Commonly known as "lucio leprosy", this variety is more common in Mexico and Costa Rica. Our patient had biopsy findings indicative of leukocytoclastic vasculitis,

another factor contributing to difficulty differentiating from rheumatologic disease [8].

Leprosy is a great masquerader with manifestations resembling several different connective tissue diseases. Biopsy-proven cases of leprosy can present with heliotrope rash, muscle weakness, elevated muscle enzymes, oral ulcers, malar rash, and photosensitivity, but can also resemble scleroderma with Raynaud's phenomenon, pitting scars, and skin thickening. In cases where findings don't present with typical rheumatologic disease, skin biopsy should be performed to rule out destructive diseases such as leprosy and rare presentations such as ENL should be part of the differential. Demonstration of acid-fast bacilli in the dermis is the cornerstone of diagnosis [7–9].

## CONCLUSION

Though leprosy poses a more significant disease burden in developing nations, it continues to have prevalence in developed regions. Diagnosis is often complicated by lack of awareness as well as atypical signs and symptoms. Therefore, it is critical that clinicians acknowledge these varied presentations and keep leprosy in their differential when faced with unusual findings reminiscent of rheumatologic disease.

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## Author Contributions

Marilyn Arosemena – Conception of the work, Design of the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Carlos I. Pacheco – Conception of the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Ankitha Lingamaneni – Acquisition of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Authors declare no conflict of interest.

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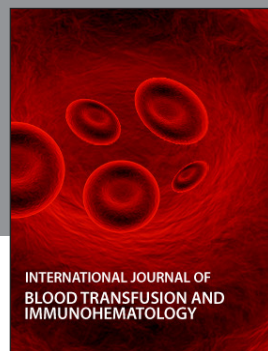
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