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Lucio's phenomenon in a non-endemic region: A case report

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Abstract

Leprosy remains a persistent health challenge in endemic regions with cases rising in non-endemic regions such as North America. Patients with leprosy present with a variety of symptoms including limited skin lesions in tuberculoid leprosy to extensive lesions and high bacterial proliferation in lepromatous leprosy. This case report details a 77-year-old Canadian man of South Asian descent with lepromatous leprosy and Lucio's phenomenon in Western Canada. The patient exhibited widespread retiform purpura on the limbs with localized ulcerations, erosions, and necrosis on the left hand and feet, peripheral neuropathy, and digit shortening. Histopathological examination and PCR confirmed *Mycobacterium leprae*. Management involved a 24-month multidrug therapy, leading to significant symptom reduction. This report highlights the diagnostic challenges of leprosy in non-endemic regions and the importance of a multidisciplinary approach for accurate diagnosis and treatment.

Keywords

Lucio's phenomenon, lepromatous leprosy, *Mycobacterium leprae*, Canada

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Introduction

Leprosy, once declared "eliminated" by the World Health Organization in 2000, continues to pose a significant health challenge due to its persistence in endemic regions such as South America, Asia, and Africa. Despite the official elimination status, global reports indicate there have been 200,000 new cases of leprosy since 2017, with 80.2% of these cases occurring in India, Brazil, and Indonesia.[1] Increased globalization has led to a notable rise in leprosy cases in non-endemic regions such as North America. In Canada, the prevalence is now approximately 0.6 cases per 100,000 people.[2]

Leprosy manifests along a spectrum, ranging from tuberculoid leprosy, characterized by a limited number of skin lesions and a robust cell-mediated immune response, to lepromatous leprosy, marked by extensive skin lesions, diminished cell-mediated immunity, and high bacterial proliferation.[3]

Lepra reactions are a notable feature of leprosy and can be categorized as type 1 reactions, type 2 reactions, or Lucio's phenomenon (type 3). Type 1 reactions are a delayed type of hypersensitivity, while type 2 reactions are an acute immune complex vasculitis, encompassing erythema nodosum leprosum.[3]

Lucio's phenomenon is a very rare and acute vascular necrotic reaction in patients with the diffuse lepromatous form of leprosy that can be life threatening. This reaction is characterized by diffuse lesions, predominantly on the extremities, which may include nodules that heal into atrophic stellate scars.[3] Untreated Lucio's phenomenon can cause various complications, including nerve damage, systemic inflammation, deformity, amputation, and resulting disability.[4] Due to its rarity in non-endemic regions and its clinical similarity to other rheumatic diseases and various forms of vasculitis and vasculopathies, lepromatous leprosy and Lucio's phenomenon can lead to diagnostic uncertainty and delayed treatment.

Here we report a case of lepromatous leprosy with Lucio's phenomenon in Western Canada. This case report aims to highlight the clinical features of this condition, increase

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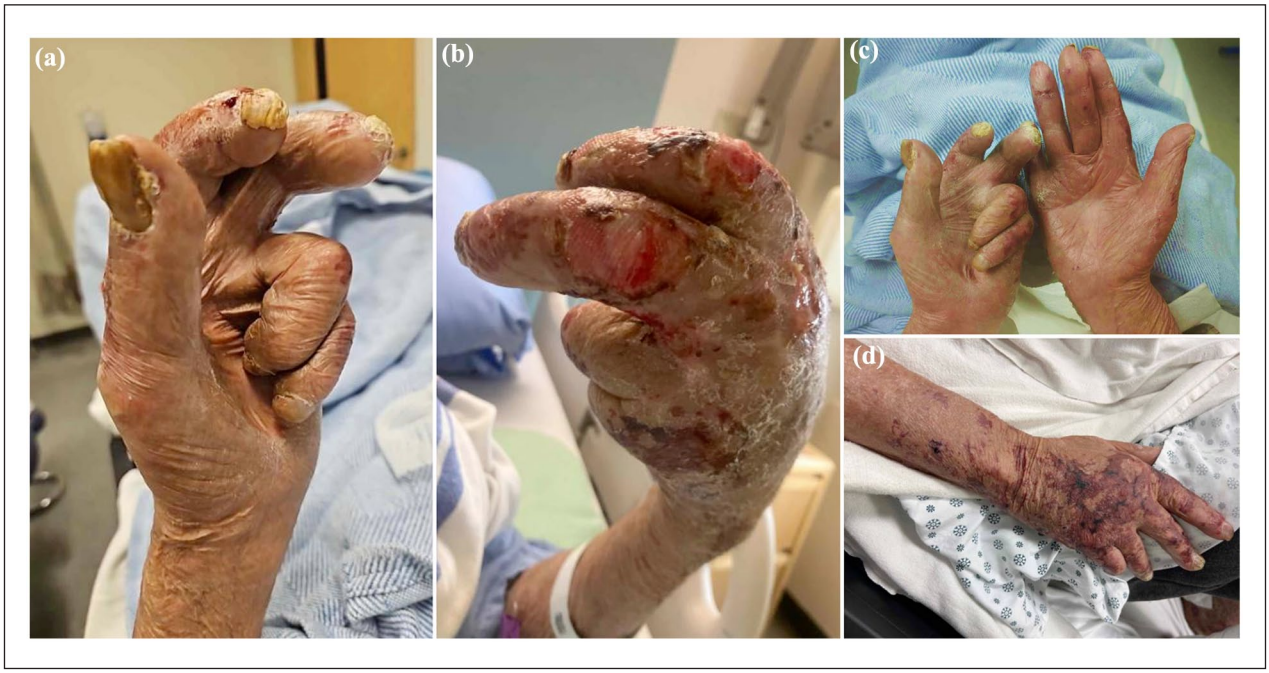

Figure 1. (a) "Ulnar Claw" deformity with flexion contractures of the fourth and fifth digits of the left hand, also known as the Papal Benediction Sign. (b) Painless ulcers and erosions on the dorsal aspect of digits 2 and 3 of the left hand. (c) Digital shortening of the left hand's digits 2 and 3, along with an "Ulnar Claw" flexion contracture of digits 4 and 5. (d) Inflamed retiform purpura on the extensor right forearm, dorsal right hand, and dorsal aspects of digits 2–4.

awareness of this rare manifestation of leprosy, and improve diagnostic accuracy in non-endemic regions.

Case report

A 77-year-old Canadian man of South Asian descent presented to dermatology for consultation with a 1-week history of widespread retiform purpura on the limbs with localized ulcerations, erosions, and necrosis on the left hand (Figure 1) and feet (Figure 2). Additional features were noted including ichthyosis on his torso, saddle nose deformity, digit shortening, and madarosis. His past medical history included hypothyroidism, hypertension, a previous stroke, and a diagnosis of infected psoriasis 1 year previously. He had a significant travel history, spending 6 months annually in India for many years. He reported no preceding medications or viral illnesses. A neurological assessment revealed significant sensory loss to his feet and exhibited a "Papal Hand" flexion contracture (or "Ulnar Claw hand") of the fourth and fifth fingers of the left hand. The initial clinical presentation led to a differential diagnosis, including livedoid vasculopathy, vasculitis, and nutritional deficiencies.

Initial laboratory testing revealed positive antinuclear antibody, anti-Jo-1, and pANCA (MPO-positive), along with elevated rheumatoid factor. To clarify the diagnosis, a skin biopsy was performed. Histopathological examination revealed granulomatous dermatitis and panniculitis (Figure 3). Fite staining of the biopsy specimens showed innumerable acid-fast bacilli, which was highly suggestive of *Mycobacterium*. Further analysis revealed diffuse histiocytic infiltrates involving the entire dermis and extending into the subcutis, with focal perineural and intraneural involvement. The histiocytes had a bubbly, vacuolated cytoplasm with the presence of globi. Acid-fast bacilli were found within the endothelial cells and lumen of blood vessels. On direct immunofluorescence, granular C3 (2+) and IgA (1+) positivity was detected at the dermoepidermal junction. PCR testing showed the presence of *Mycobacterium leprae*, confirming the diagnosis of lepromatous leprosy with Lucio's phenomenon.

Management involved a 24-month multi-drug therapy (MDT) consisting of clofazimine, rifampin, dapsone, and prednisone. After initiating treatment, the patient had a notable reduction in their initial symptoms and did not experience any new symptoms such as swollen painful nerves, new skin lesions, hyperpigmentation, reduced sensation, or eye pain. Informed consent was obtained from the patient.

Discussion

Leprosy has a long and complex global history, affecting millions of people across different regions for centuries.[5] This case highlights a rare instance of lepromatous leprosy with the even rarer presentation of Lucio's phenomenon in a non-endemic region. On review of the literature, prior to this case report, there have been no published reports of Lucio's phenomenon occurring in Canada, highlighting the rarity of this reaction.

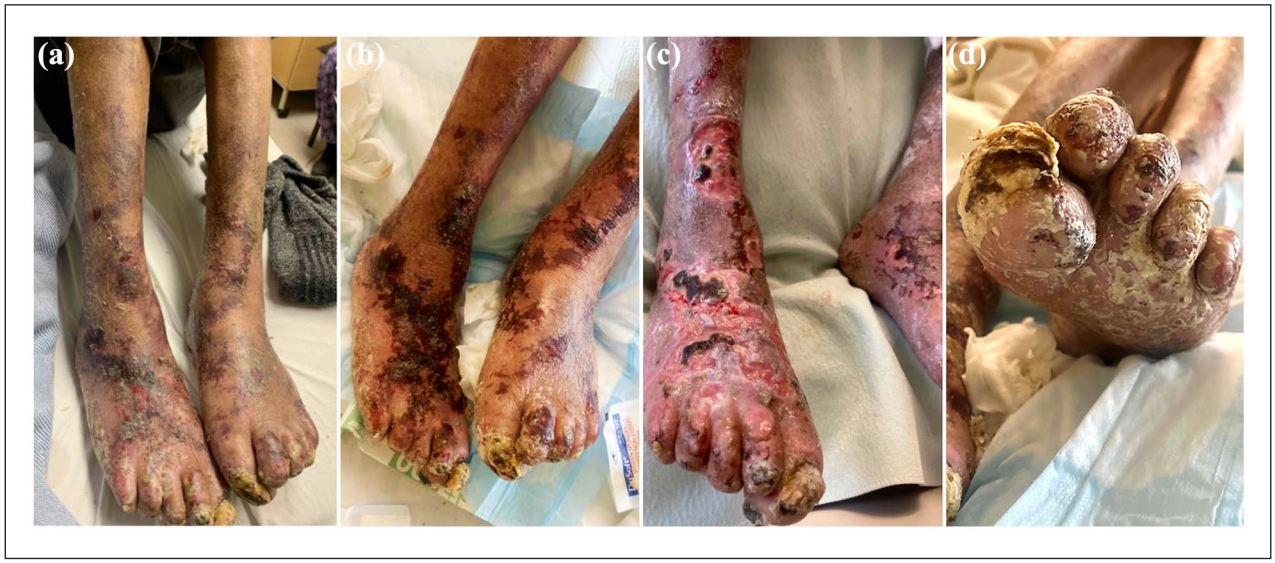

Figure 2. (a) Retiform purpura on the anterior lower legs and dorsal feet, with erosions on the right dorsal foot and toes. (b) Progression of skin necrosis in areas affected by retiform purpura on the dorsal feet and lower legs. (c) Reticulated ulceration on the dorsal right foot and lower right leg in areas previously affected by retiform purpura. (d) Hyperkeratosis and digital shortening of the toes on the left foot.

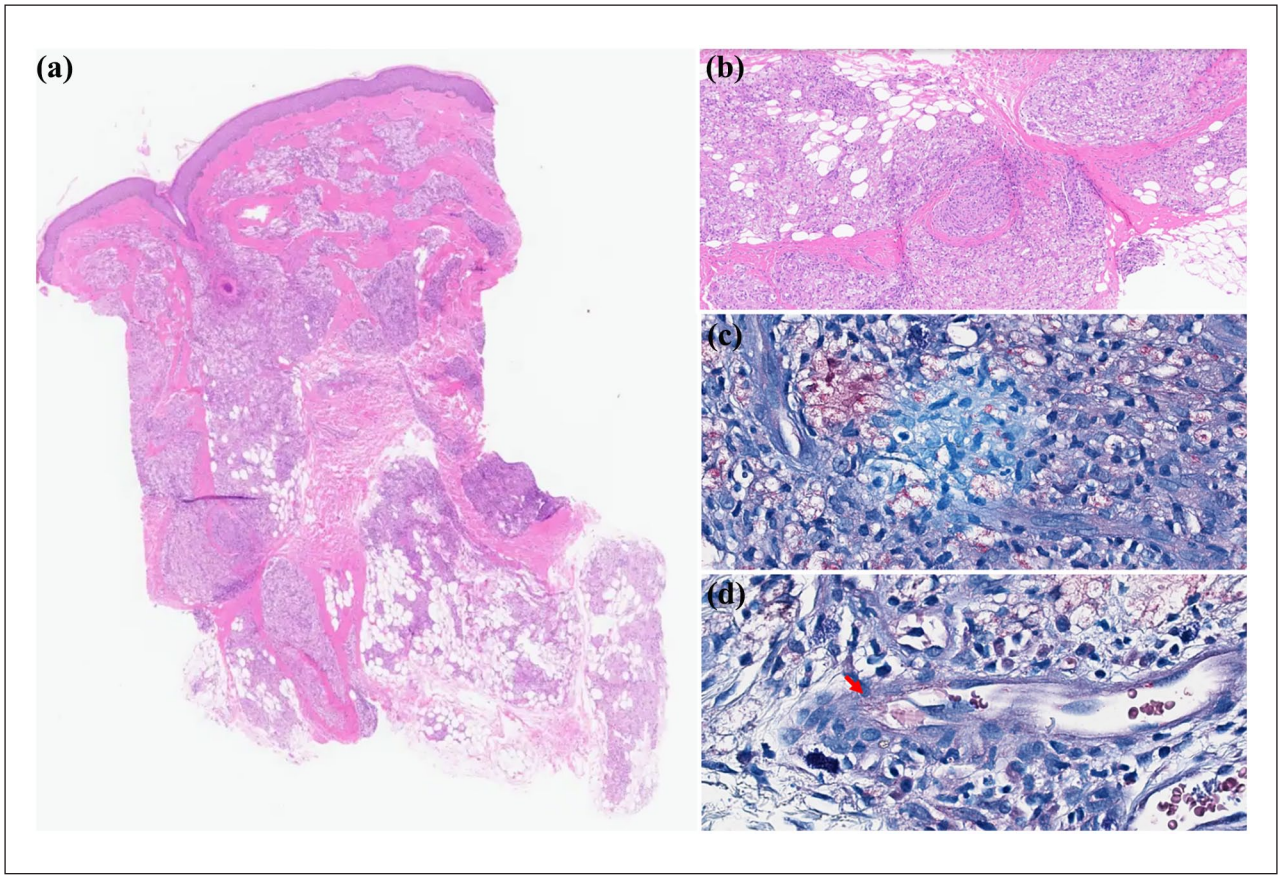

Figure 3. (a) Grenz zone with diffuse granulomatous dermal inflammation extending into the subcutis and focal perineural inflammation (H&E, ×2). (b) Perineural inflammation (H&E, ×10). (c) Numerous acid-fast bacilli present within histiocytes (Fite stain, ×66.5). (d) Numerous acid-fast bacilli within the blood vessel lumen (red arrow; Fite stain, ×70).

M. leprae are obligate intracellular pathogens that preferentially infect and replicate in peripheral nerves and Schwann cells, leading to neuronal damage explaining the neurological symptoms observed in this patient.[6] Additionally, in leprosy patients, bone lesions may occur due to trauma and secondary bacterial infections affecting denervated tissues. This can result in bone atrophy, improper remodeling, and ultimately digit shortening.[7]

The patient's history reveals a protracted and complicated journey toward diagnosis. His travel history to India suggests the likely source of his leprosy infection. However, the exact onset of the infection remains undetermined.

An important differential in the workup for this patient's retiform purpura including autoimmune etiologies, and the positive antibodies were a significant confounder in the diagnosis. However, upon discussion with Rheumatology and review of the literature, it was determined that significant inflammation in the context of lepromatous leprosy could also cause increased rheumatic biomarkers, posing a diagnostic challenge.[8,9] In conjunction with Infectious Diseases and a positive PCR result for *M. leprae*, we were able to arrive at the correct diagnosis. The infection was likely misdiagnosed multiple times due to the rarity of leprosy in non-endemic regions, demonstrating the critical role of a multidisciplinary approach in the accurate diagnosis and treatment of this infection.

Despite significant advancements in the understanding and management of leprosy, the primary treatment of MDT has remained the same. This regimen has been effective in reducing the incidence of leprosy and preventing the transmission of *M. leprae*. While MDT was originally implemented to prevent the development of resistant strains, reports have shown a steady increase in antibiotic resistance.[10] Additional antibiotics such as minocycline, clarithromycin, and fluoroquinolones have shown promise in treating drug-resistant cases of leprosy.[10]

In terms of Lucio's phenomenon, there is currently no consensus on the treatment. Various approaches have been used with differing outcomes, including MDT exclusively, corticosteroids, anticoagulants, systemic antibiotics, surgical debridement, and skin grafting.[11]

In conclusion, leprosy remains a significant health challenge in endemic regions and is still observed in non-endemic areas. This case report highlights a rare instance of lepromatous leprosy with Lucio's phenomenon in Western Canada and the importance of multidisciplinary management, including Dermatology and Infectious Diseases.

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Patient consent

Consent was obtained from the patient for the anonymized images and information to be published in this article.

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