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Lucio Phenomenon: A rare phenomenon in Asian subcontinent

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Abstract

Lucio phenomenon (LP) is an uncommon leprosy reaction seen in individuals with a distinct, primary and generalized manifestation of lepromatous leprosy known as Lucio leprosy. Lucio Phenomenon is characterized by the presence of purpuric macules, necrosis, extensive ulceration and distinct, angular borders primarily affecting the limbs. Lucio leprosy is mainly seen in Mexico and Central America and only few countable cases have been reported in the Asian and Middle East countries[1]. Awareness of this condition is crucial for early detection and timely management. This case report highlights a rare instance of LP diagnosed in Western India; a region not traditionally endemic for leprosy. Delay in accurate identification of this condition can result in significant disability and potential disease transmission within the community.

Keywords: Lucio Leprosy, Lucio Phenomenon, Vasculopathy, Vasculitis, Buergers Disease.

Introduction

Lucio phenomenon is a necrotizing panvasculitis and is classified as a variant of type 2 leprosy reaction. It is clinically characterized by necrotic-hemorrhagic lesions predominantly affecting the extremities. Though primarily observed in Mexico and Central America, limited occurrences have been reported in India. Here we came across this phenomenon in a patient who was not presenting with symptoms of leprosy

Case Presentation

A 63-year-old male, chronic smoker presented with painful ulcers on his upper and lower limbs persisting for one month. He also reported symptoms of malaise, weight loss, and asthenia for the same duration. On examination widely distributed necrotic, ulcerated lesions with serous discharge on both lower limbs, elbows and the dorsum of the hands were present. The lesions exhibited purpuric margins, sloughing, areas of necrotic eschar. He had pitting edema with shiny xerotic skin on both lower limbs, and diminished arterial pulses in all four limbs. Pallor was noted, with a lean and emaciated build. A provisional diagnosis of small and medium vessel vasculitis was considered, with Berger's disease as a differential due to his smoking history.

The hemogram showed microcytic hypochromic anemia (hb 7.4 g/dL), reduced serum iron (20 µg/dL), TIBC (101 µg/dL), and raised serum ferritin (841 ng/dL), hypoalbuminemia (1.8 g/dL), and total protein (5.2 g/dL) with normal liver enzymes. Tests to rule out coagulopathy or connective tissue disorders, including ANA, BT, CT, PT/INR, aPTT, and lupus anticoagulant, were within normal limits. Urinalysis, blood sugar levels, chest X-ray, and Mantoux test were normal for his age. Serology for viral markers was negative.

USG Doppler of of both lower limbs revealed diffuse extensive intimal calcification of CFA, SFA, DFA, PA, ATA, PTA & DPA with reduced flow and mild subcutaneous edema, with no evidence of deep vein thrombosis.

The H& E stain of biopsy showed epidermal necrosis and ulceration, superficial & deep perivascular infiltrates of lymphocytes and neutrophils, few small and medium sized vessel in subcutaneous tissue showed features of vasculitis with complete lumen occlusion. Interestingly there were many foamy macrophages filled with eosinophilic material seen in the dermis and subcutaneous tissue pointing towards a diagnosis of leprosy. Fite-Faraco stain revealed slender eosinophilic bacilli in macrophages scattered in the dermis and vessel walls confirming the diagnosis of leprosy.

Upon further questioning, the patient reported diffuse numbness extending distally from his knees and elbows, slipping of chappals, and difficulties with fine motor tasks.

Sensory examination showed symmetrical hypoesthesia on both distal extremities, with thickened, non-tender ulnar, radial, and common peroneal nerves. Motor examination revealed weakness of the palmar interossei in both hands.

Slit skin smear from earlobes and eyebrows revealed numerous uniformly stained acid-fast bacilli with a bacteriological index of 4+. Nerve conduction velocity (NCV) revealed asymmetrical sensory-motor axonal neuropathy affecting both upper and lower limbs.

As the patient had no plaques or patches of leprosy, he was diagnosed with diffuse leprosy. He developed a vasculitic reaction with bacilli present in the vessel walls, consistent with Lucio phenomenon in Lucio leprosy. Patient was started on multidrug therapy with

rifampicin, dapsone and clofazimine along with supportive wound care. Patient showed good recovery after starting these medications with complete repethelisation of the ulcers in 3-4weeks

Discussion

Diffuse Lepromatous Leprosy (DLL), also known as Lucio leprosy, is a severe form of leprosy characterized by a high multibacillary load. Although Mycobacterium lepromatosis is often associated with DLL, cases involving Mycobacterium leprae or dual infections have also been reported[3].

Clinically, DLL manifests as diffuse, non-nodular lesions and can develop into Lucio Phenomenon(LP). LP results from uncontrolled replication of lepra bacilli that invade blood vessel walls and endothelial cells. This process induces endothelial proliferation and constriction of the vascular lumen. Inflammatory responses and alterations in the coagulation system result in vascular thrombosis, ischemia, infarction, and tissue necrosis, culminating in the histopathological characteristics of Lucio phenomenon[6].

Lucio phenomenon presents with multiple painful erythematous violaceous macules and hemorrhagic blisters that progress into necrotic and ulcerated lesions, typically affecting the upper and lower extremities[5].

A systematic review by Frade MA et al. collected 49 cases, of which 9 (18.36%) were from Indian case reports. Most patients presented with LP before being diagnosed with leprosy.

In our case, the patient did not have symptoms suggesting leprosy and presented with LP, which mimicked vasculitis. This underscores the importance of recognizing diffuse lepromatous leprosy to avoid diagnostic delays. Notably, madarosis was present in almost all reviewed cases, an important finding that was initially overlooked even in our patient.

Diagnosing LP requires three criteria: skin ulceration, vascular thrombosis, and infiltration of blood vessels by leprosy bacilli. Late diagnosis can lead to severe complications such as sepsis, amputations, and fatalities due to coagulation disorders.

Treatment with anti-leprosy regimens containing rifampicin is highly effective, along with a brief course of high-dose corticosteroids to manage the immune response to M. leprae antigens.

Conclusion- Lucio Phenomenon can manifest as the initial presentation of Lucio leprosy even in non-endemic regions. Diagnosing and timely management of this condition is essential in reducing mortality, morbidity and community transmission.

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Legend Figures

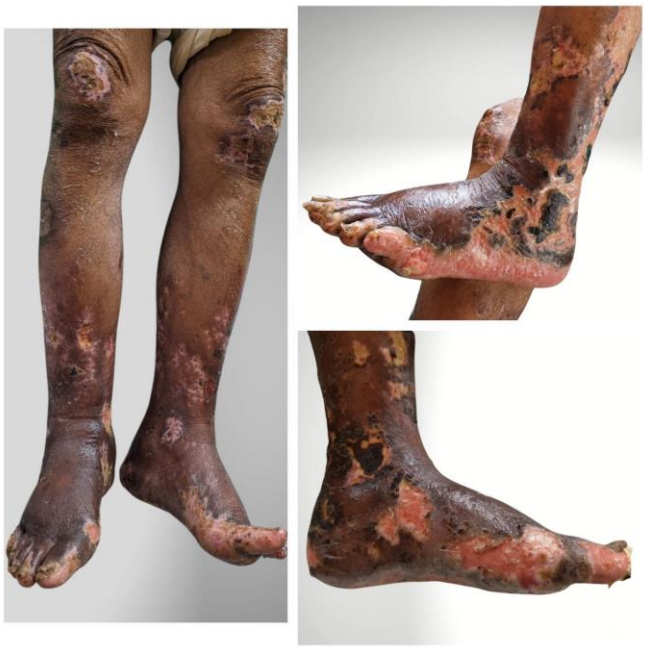

Figure 1: Extensive ulcers and necrosis with serous discharge over knees, ankles and soles.

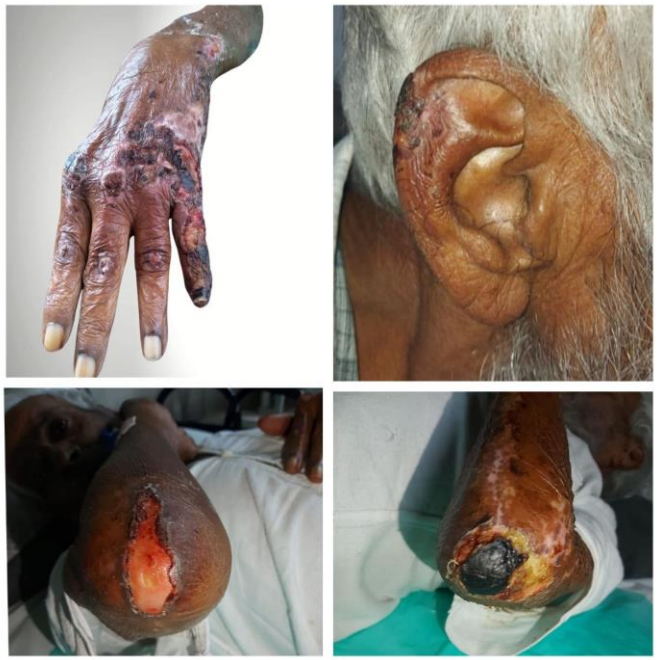

Figure 2: Diffuse erythematous purpuric and ulceronecrotic lesion over hands and elbows. Necrotic lesion over pinna too.

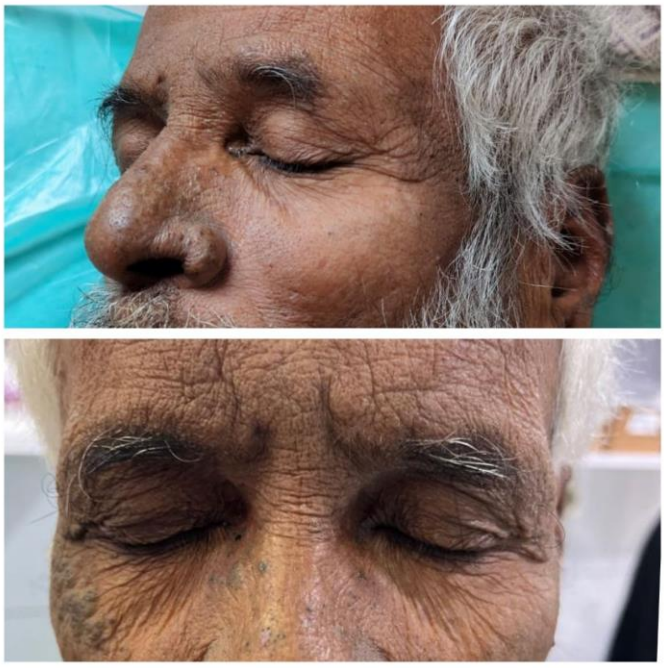

Figure 3: Bilateral superciliary madarosis present

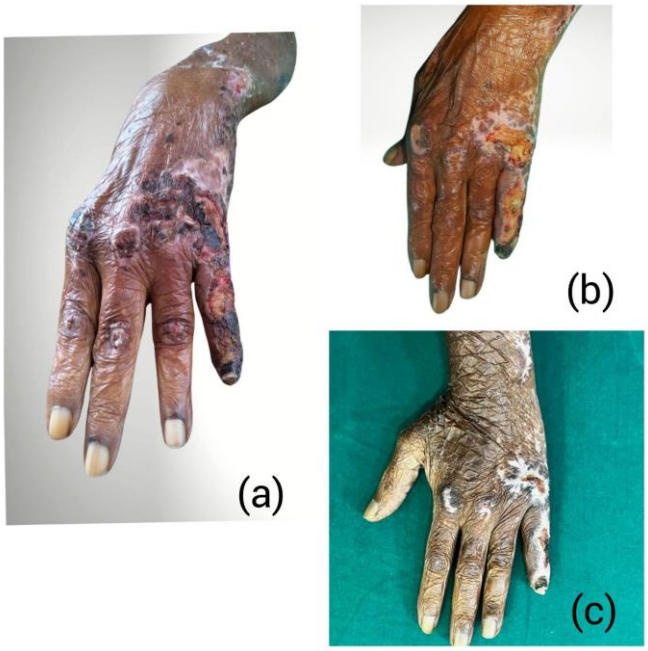

Figure 4: Image (a) shows ulceronecrotic lesions progressing (b) to ulcer with yellowish adherent crust and finally (c) after 4 weeks of treatment healing with irregular scars and depigmentation.

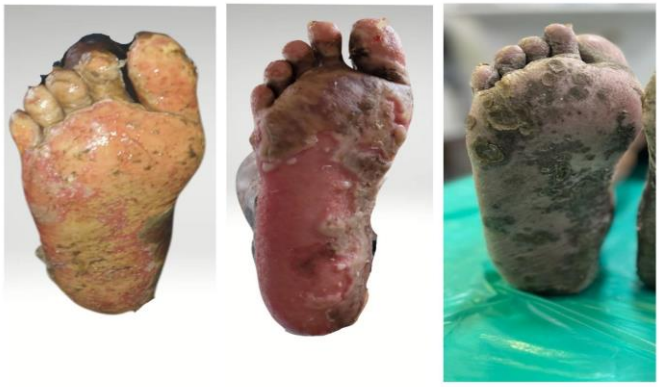

Figure 5: (left to right) Presenting with complete sloughing of sole with loss of substance in toe tips and serous discharge. After starting treatment showing reepithelization with healthy granulation tissue to complete sole reepithelization in 4weeks.

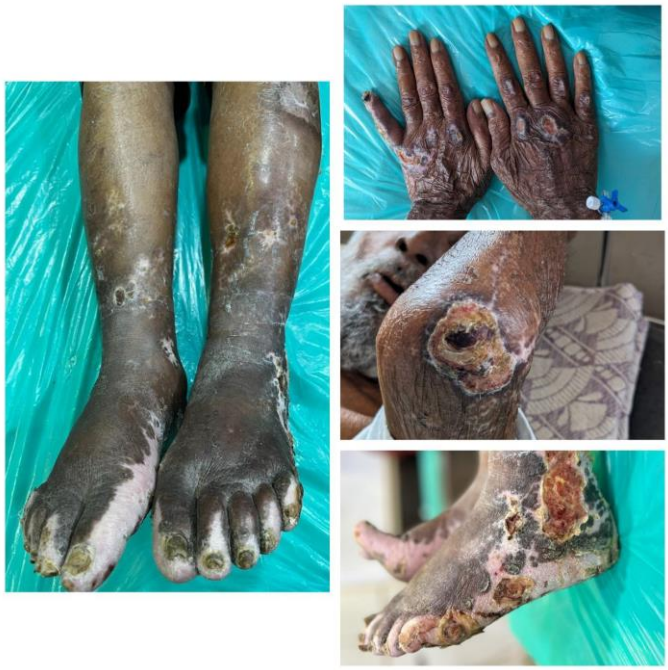

Figure 7: After 1month of admission and commencement of treatment. Healing with ulcers and scars at site of necrotic eschars.

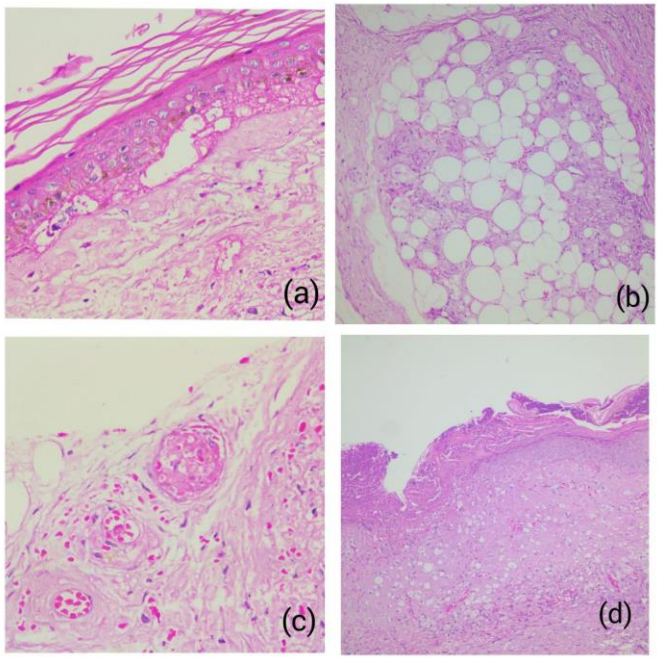

Figure 8: H&E Stain (a)Epidermal and Dermal edema with bulla formation (b) lobular panniculitis with foamy macrophages in subcutaneous tissue(10x); (c) vasculitis with vessel lumen obliterated by thrombus (10x) (d)Epidermal necrosis and ulceration(10x).

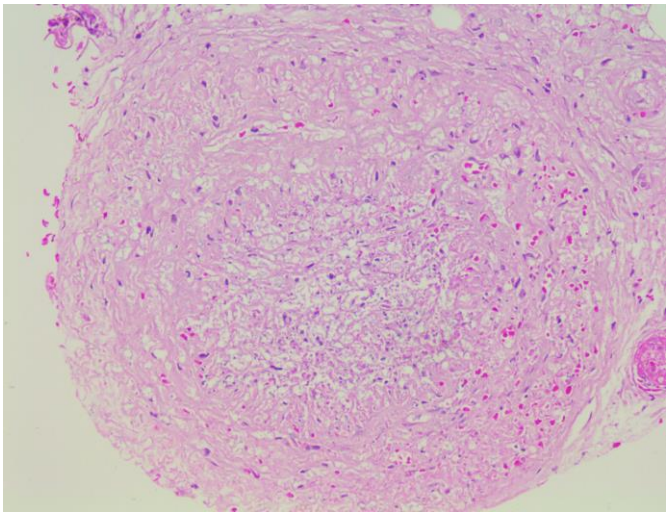

Figure 9: H&E stain (10x) Infiltration of vessel wall. Thickening of vessel wall and narrowing of lumen.

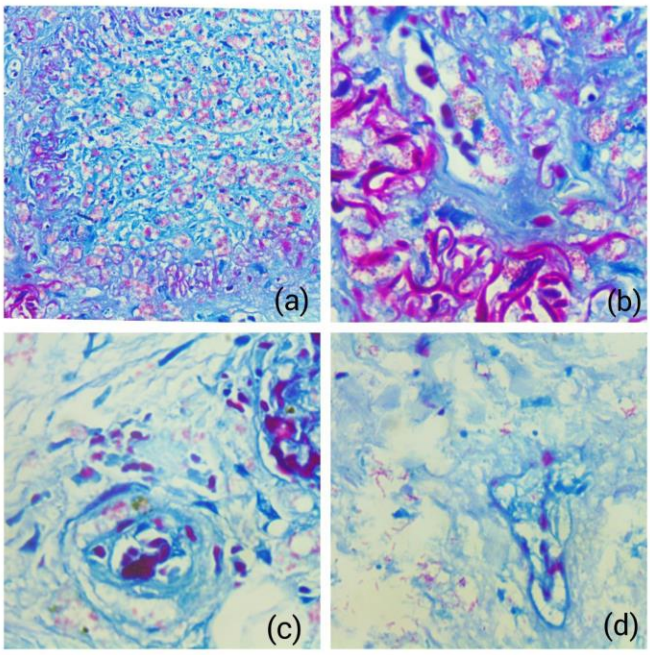

Figure 10: Fite Faraco stain(x100) showing Lepra bacilli in endothelial cells of vessels and in foamy macrophages.