Journal of Pakistan Association of Dermatologists.2023;**33(1)**:331-334.

Case Report

A fatal case of Lucio phenomenon in untreated Lucio leprosy

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Abstract Lucio phenomenon (LP) is a rare and potentially fatal reaction that exclusively occurs in diffuse lepromatous leprosy or Lucio leprosy. It is characterized by purpuric macules, which progressively develop into extensive painful ulceration, primarily affecting the lower extremities. We report a case of 56-year-old with purpuric patches, blisters, and erosions on his face, mouth, arms, hands, legs, and feet that started to appear for three days without any hypopigmented or erythematous patches prior to the complaint. Slit skin smear examination revealed a bacterial index of 6+ with a morphological index of 13.66% and the Ziehl-Neelsen stains for acid fast bacili (AFB) were positive. He was diagnosed with LP complicating with sepsis. He was managed with multidrug therapy for multi-bacilary leprosy, corticosteroid, and antibiotic. However, his clinical condition was deteriorated and he died due to septic shock. Early recognition and prompt management are the key to successful therapy and preventing complications. Until now, there is no standard guideline for LP treatment due to its rare incidence. Multi-drug therapy for multibacillary leprosy is recommended for the management of LP. The use of corticosteroids and thalidomide is still controversial.

Key words
Acid-fast bacilli, Lucio leprosy, Lucio phenomenon, multi-drug therapy, necrosis.

Introduction

Lucio phenomenon (LP) is an exclusive reaction found in Lucio leprosy, manifesting as purpuric macules, ulceration, and necrosis that primarily affects the lower limbs.[1,2] Histologically, LP shows necrotizing vasculitis, colonization of acid-fast bacili (AFB) within endothelial cells, and thrombosis. It commonly occurs after 1 to 3 years of the onset of untreated Lucio leprosy.[3] Previously, LP is frequently documented in Central America, however, recently similar cases have been reported from other countries.[4-6] Without adequate treatment, LP can be complicated with sepsis.[3] Currently, there is no standard guideline on the management of LP. We report a case of fatal Lucio leprosy with LP complicated by diabetes mellitus and sepsis treated in a tertiary referral hospital in Indonesia.

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Case

A 56-year-old man complaints of painful purpuric patches, blisters, and erosions on his face, mouth, arms, hands, legs, and feet that started to appear for three days. He never noticed any hypopigmented or erythematous patches with paresthesia prior to the incidence. He regularly took metformin 1 x 500 mg for diabetes mellitus since six years ago. There was no history of herbs, supplements, or other drugs consumption. Over the last three months, his eyebrows, eyelashes, and hair started to fall out,

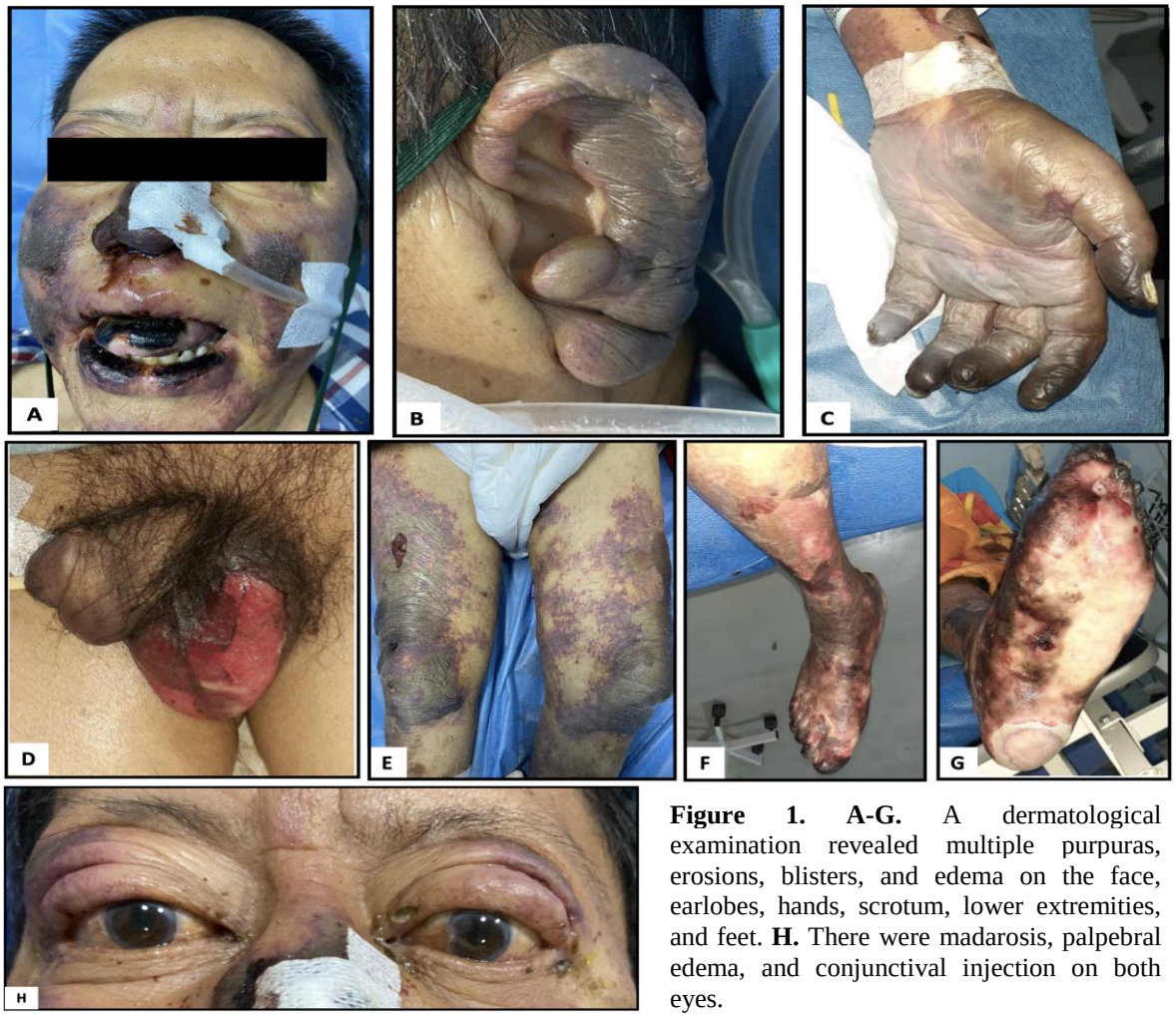

Figure 1. A-G. A dermatological examination revealed multiple purpuras, erosions, blisters, and edema on the face, earlobes, hands, scrotum, lower extremities, and feet. **H.** There were madarosis, palpebral edema, and conjunctival injection on both eyes.

accompanied by frequent flu-like symptoms. However, he did not complaint of visual disturbances.

Upon examination, he was severely ill with normal blood pressure, tachycardia, and tachypnea. Dermatological examination revealed multiple purpuras, erosions, blisters, and edema on the face, earlobes, scrotum, and extremities. (**Figure 1A-1G**) with negative Nikolsky sign. There were madarosis, palpebral edema, and conjunctival injection in both eyes (**Figure 1H**). Laboratory investigations showed anemia (haemoglobin 6.4 g/dL); thrombocytopenia 60,000x103/µL; leukocytosis 29,980x103/µL with neutrophilia; impaired renal function (ureum 81.6 mg/dL, creatinine 2.0 mg/dL, glomerular filtration rate 36.2 mL/min/1.73m2); hyperglycemia 453 mg/dL; hyponatremia 121 mEq/L; hypoalbuminemia 1.87 g/dL; increased marker of inflammation (C-reactive protein 193.8 mg/L and procalcitonin 89.1 ng/mL); increased d-dimer 7200 µg/L; prolonged prothrombin time (PT) 23.8 seconds and activated partial thromboplastin time (aPTT) 71.6 seconds; and normal liver enzymes. Serology for anti-HIV, hepatitis B, and hepatitis C were non-reactive. Polymerase chain reaction (PCR) for COVID-19 was negative. Gram stain from skin erosions on the scrotum showed scarce leukocytes and gram-positive cocci with no fungal element. Microbial culture of skin

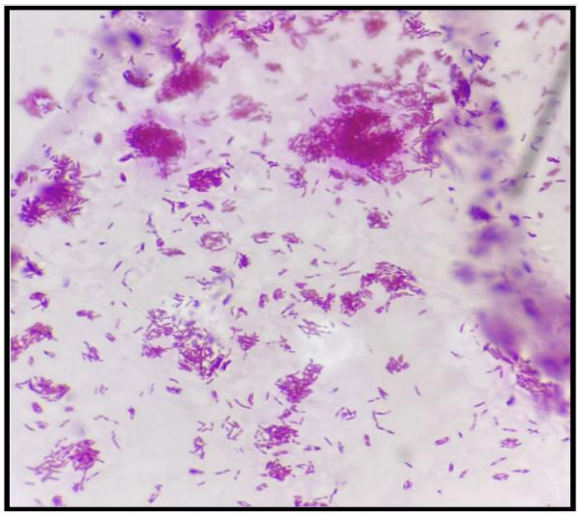

Figure 2. Abundant acid-fast bacilli from skin slit smear of the right cheek. Ziehl-Neelsen, 400x.

erosion showed *Proteus vulgaris* isolate sensitive to chloramphenicol, gentamycin, kanamycin, ampicillin/sulbactam, aztreonam, ceftriaxone, ceftazidime, cefotaxime, cefepime, doripenem, cefepime, tigecycline, meropenem, and levofloxacin. Both urine and blood cultured were sterile. Slit skin smear from right cheek indicated a bacterial index of 6+ with a morphological index of 13.66%. The Ziehl-Neelsen stains for AFB were positive (**Figure 2**). He was diagnosed with LP complicated with sepsis, diabetes mellitus, and disseminated intravascular coagulation. He received systemic medication of multi-drug therapy consisting of rifampicin 600 mg monthly, dapsone 100 mg daily, and clofazimine 300 mg monthly followed by 50 mg daily; IV methylprednisolone 48 mg; IV meropenem 3 x 1 gr; insulin 4 unit/hour; heparin 20,000 unit/day, and blood transfusion. We treated skin erosions with fusidic acid ointment and wound dressing, including the interdigital area, to prevent fusion of the fingers. Skin biopsy and bone marrow puncture (BMP) were planned to assess the damage of vascular endothelial and involvement on the bone marrow, respectively. However, because his condition progressively worsened and there were contraindications (thrombocytopenia and prolonged PT and aPTT), the skin biopsy and BMP were postponed. His clinical condition deteriorated with decreased consciousness and extensive cutaneous infarcts **(Figure 3)**. He died due to the consequent septic shock after 19 days of hospitalization.

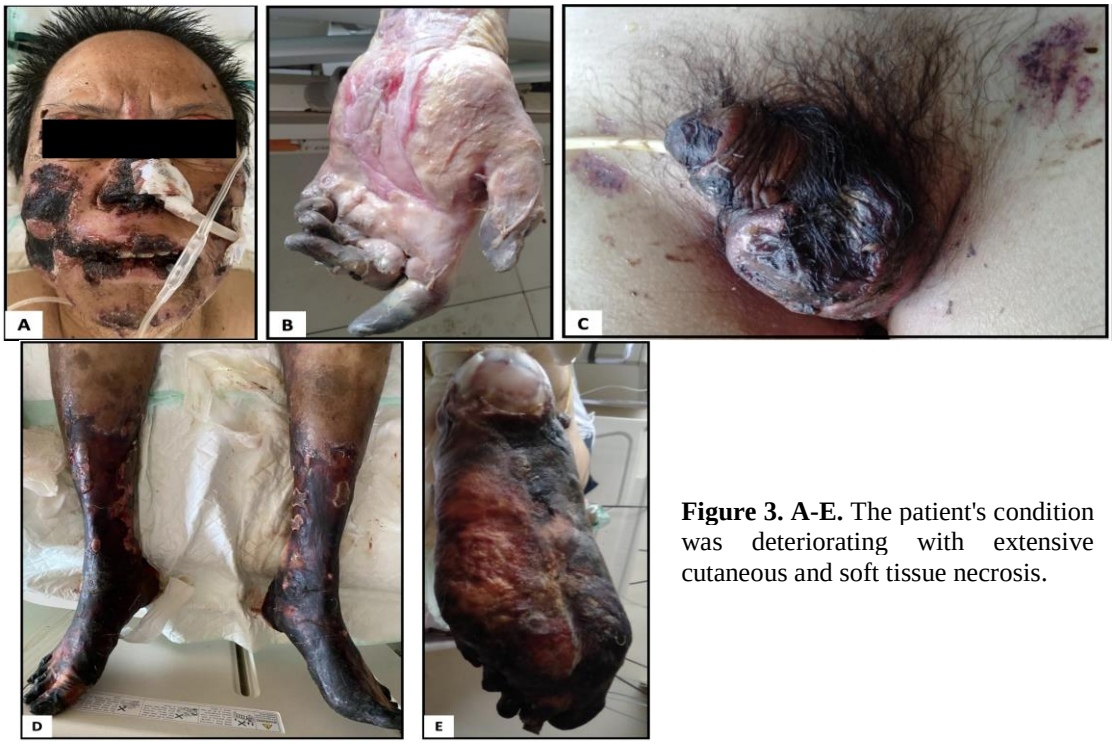

Figure 3. A-E. The patient's condition was deteriorating with extensive cutaneous and soft tissue necrosis.

Discussion

The clinical manifestations in our patient were identical to Choon *et al*.'s report,[5] which found ulcers, blisters, and hemorrhagic purpuric lesions predominantly in the lower extremities. The main distinctive histologic features of LP are AFB colonization of endothelial cells, proliferation with noticeable thickening of vascular walls, angiogenesis, vascular ectasia, and thrombosis.[7] The laboratory findings in our patient revealed anemia and thrombocytopenia, which prompted a possibility of bone marrow involvement. A systematic review by Frade *et al*.[4] found the positive result of AFB in two out of three LP patients whose BMP was examined.

Although there is no standard guideline for managemment of LP, most reports recommend treatment with antileprosy drugs (MDT) and systemic steroids.[4] Contrary, Peixoto *et al*.[8] found that MDT for multibacilary alone were effective for treating LP. Misra *et al*.[9] treated LP with the combination of MDT MB, prednisone, and thalidomide with marked improvement in five months after therapy initiation. In our case, the patient received MDT MD, methylprednisolone, antibiotics for sepsis, and daily wound care. He died after 19 days of hospitalization due to septic shock. Sepsis is a potentially fatal complication in LP. Our case demonstrated the importance of early diagnosis and appropriate treatment to decrease morbidity and mortality.

Conclusion

Lucio phenomenon is a rare leprosy reaction that commonly occurs in untreated Lucio leprosy. Clinicians should have a high index of suspicion for LP, especially in patients with diffuse lepromatous leprosy. Understanding the clinical appearance and histopathologic features of LP is necessary for proper management and prevent complications.

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