

Lucio Phenomenon and APLA in Hansen's Disease: A Rare Phenomenon

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ABSTRACT

Lucio phenomenon (LP) is a rare reactional state seen in cases of diffuse lepromatous leprosy. Lucio leprosy is a pure, primitive, and diffuse form of lepromatous leprosy. It is observed almost exclusively in Mexico and Central America and is considered a globally restricted phenomenon. However, isolated cases are being reported worldwide. Patients with Lucio leprosy often present with manifestations of LP, which includes purpuric macules with multiple and extensive areas of ulceration with bizarre-patterned, angulated borders mainly affecting the extremities. Lucio phenomenon is difficult to recognize, especially in nonendemic countries, which can lead to a delay in its diagnosis and management. We report a case of LP due to its occurrence in the classical form of lepromatous leprosy and rarity in Eastern India.

Keywords: Lepromatous leprosy, Lucio phenomenon, Purpuric macules, Systemic lupus erythematosus.

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INTRODUCTION

Lucio phenomenon is one such rare reactional state seen peculiarly in the pure and primitive diffuse form of lepromatous leprosy. This phenomenon was first described by Lucio and Alvarado in Mexico in 1852 after the identification of histopathological changes involving multiple acute, necrotizing cutaneous vasculitis. It was considered a globally restricted phenomenon endemic to Mexico and Central America until sporadic cases were reported from nonendemic areas of the world. It is clinically characterized by severe necrotizing cutaneous lesions, mainly on the extremities. Here, we present a rare case of a man with LP with secondary antiphospholipid antibody (APLA) syndrome in our hospital.¹

Antiphospholipid syndrome (APS) is a multisystem autoimmune disorder characterized by persistent presence of antiphospholipid antibodies directed against phospholipid-binding proteins and may present as venous and arterial thrombosis and or pregnancy loss. The most common sites of venous and arterial thrombosis are lower limbs and cerebral arterial circulation, respectively. The antiphospholipid antibodies are anticardiolipin antibody IgG and IgM, anti-beta2-gp1 antibody IgG and IgM, and lupus anticoagulant.²

Hansen's disease, or leprosy, a chronic infection caused by *Mycobacterium leprae*, is an old and widespread disease with a higher incidence in developing countries. It runs a chronic indolent course that can become complicated by acute, immune-mediated phenomena called lepra reactions.³

CASE DESCRIPTION

A 21-year-old boy with a history of Hansen's disease on multidrug therapy of dapsone, rifampicin, and clofazimine and previous history of type-1 reaction to antileprotic medications from Malda presented to our Emergency Department with complaints of fever for 22-days duration. Fever was insidious in onset, gradually progressive, intermittent, and low grade in nature. The patient also developed altered mental status, drowsiness, and restlessness for

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the last 7 days with one episode of loss of consciousness 3 days back, following which the family admitted the patient.

On further inquiry, the family also said that the patient had headache, occasional nausea, and vomiting for the same duration, and one episode of loss of consciousness 3 days back. The patient did not experience any episode of neck pain, photophobia, sore throat, chest pain, cough, palpitations, abdominal pain, yellowish discoloration of eyes and urine, reduced urinary output, or leg pains.

On examination, the patient was drowsy and was only obeying to simple commands. Pulse rate was 110 per minute, regular in rhythm, normal in volume, and normal in character. Respiratory rate was 22 per minute regular, thoracoabdominal in nature, and no accessory muscles of respirations were working. BP was 90/60 mm Hg. Temperature 100°F. Pallor was present, there was no cyanosis, icterus, clubbing, or edema. No lymph node was palpable.

Multiple hypopigmented patches with necrotic cutaneous lesions were present in both upper and lower limbs of varying



Figs 1A and B: Trunk, hand, and leg condition during admission

sizes along with multiple digital gangrenes in both upper and lower limbs (Fig. 1).

On neurological examination, the patient was only obeying simple commands. There was mild neck stiffness; Kernig's sign and Brudzinski's sign were absent. Cranial nerve examination was within normal limits, those which could be examined. There was reduced power in all the four limbs. Sensory examination only revealed decreased sensation in the hypopigmented areas of the body along with multiple digital gangrenes in both upper and lower limbs.

All the relevant investigations were sent for fever workup, and treatment was initiated on IV fluids, empirical IV antibiotics, and antipyretics. The patient did not improve with the following treatment. Fever was persisting. Magnetic resonance imaging (MRI) brain was done, which detected multiple thromboembolic lesions. The patient then underwent CSF studies that detected increased protein levels. Dermatological consultation was taken on several occasions in view of patient's Hansen's disease. Skin biopsy confirmed Hansen's disease along with colonization of endothelial cells with acid-fast bacilli and within macrophages, which were suggestive of LP.

Rheumatological evaluation was done and reports were suggestive of secondary antiphospholipid syndrome. Immunosuppressive therapy was deferred, in view of persisting fever, increasing trend of total leukocyte count, and nosocomial infection with *Candida tropicalis* in urine. IV Caspofungin was started. The patient's reports showed a decreasing trend of hemoglobin levels, and Direct Coombs' test was advised, which came out to be positive.

In view of persistence of fever and nonimprovement of the clinical condition, he was initiated with oral corticosteroids, oral hydroxychloroquine, subcutaneous (SC) unfractionated heparin, and dapsone, rifampicin, and clofazimine were continued. On day 17 of admission, the patient's fever subsided, skin rashes started fading, general condition improved, and was then discharged on tapering dose of steroids, hydroxychloroquine, warfarin, and multidrug therapy (MDT) in a hemodynamically stable and afebrile condition.

On follow-up after 14 days of discharge, he had remarkable improvement, no neurological deficit, and was walking and eating without assistance (Fig. 2, Table 1).

Day 1: MRI Brain: Multiple scattered T1 hypotense, T2/FLAIR hypertense intraparenchymal non-enhancing lesions of variable sizes showing intense diffusion restriction are seen in the bilateral basal ganglia, thalami, bilateral insular and fronto-parietal cortex, midbrain. Possible septic-embolic encephalitis or multiple cardioembolic infarct.

COVID 19 RT-PCR, MPDA, Dengue NS1 and IgM/IgG, Scrub Typhus IgM, Leptospira IgM, HBsAg, anti-HCV, HIV 1 and 2, was negative. Urine RE Normal.

EEG Brain: Suggestive of encephalopathic pattern.

Day 2: Trans-esophageal echo, no abnormality detected.

Day 3: CSF studies – Cell count 3, cell type 100% lymphocyte, Glucose 75, Protein 110, TB GeneXpert/Gram stain/AFB stain/CNS comprehensive panel, all negative.

Day 4: Blood CS and urine CS, negative.

Day 7: Skin biopsy: Suggestive of LP with underlying Hansen's disease.

Day 9: Blood CS negative; Urine CS *Candida tropicalis* growth.

Day 11: Lupus anticoagulant negative, anticardiolipin antibody IgM reactive, antiphospholipid antibody negative, beta2 GP1 antibody negative.

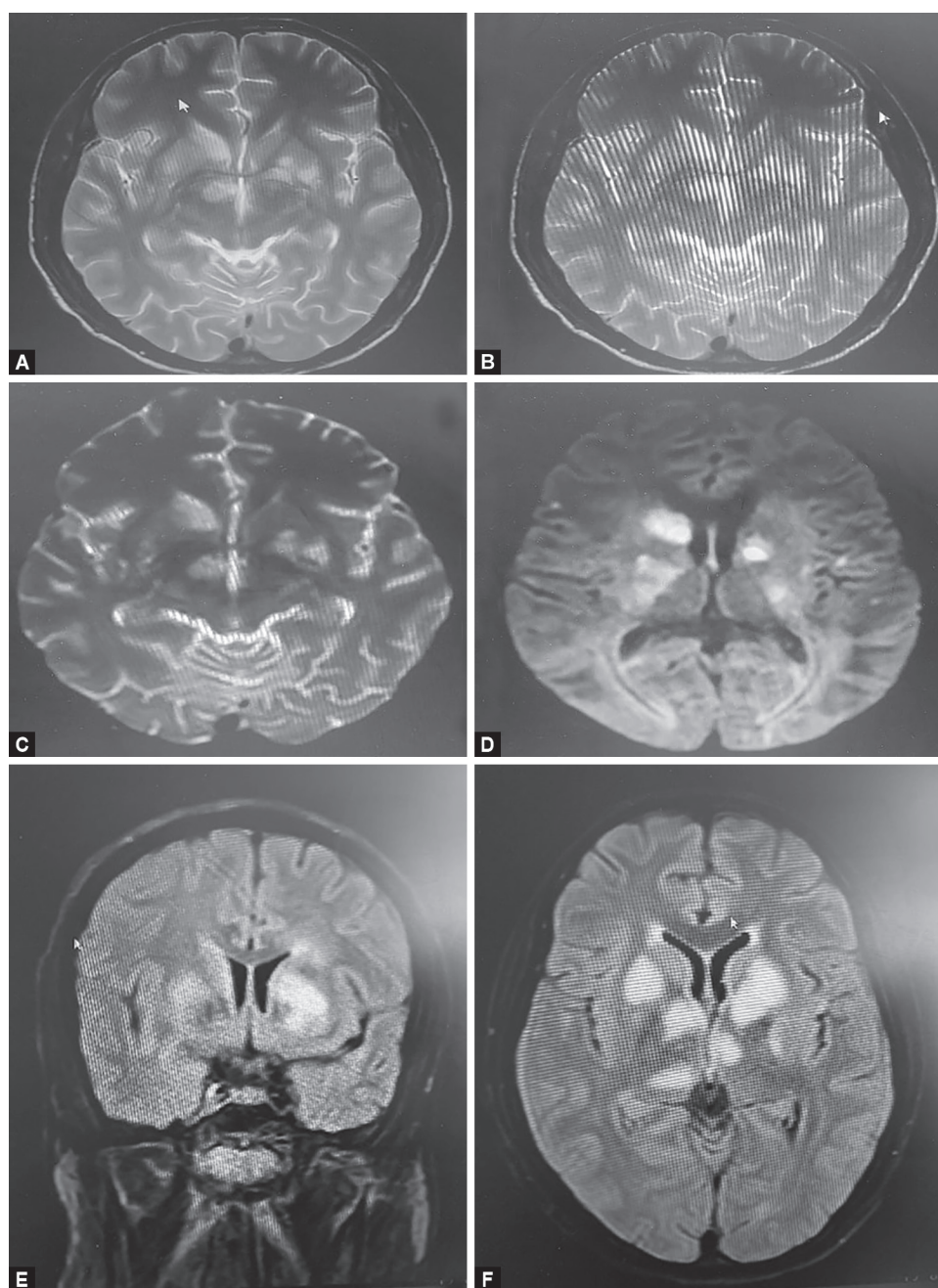
Day 13: Direct Coombs' test positive; stool for occult blood negative (Fig. 3).

CASE DISCUSSION

This patient of Hansen's disease on MDT with LP secondary APS with autoimmune hemolytic anemia was improved after administration of corticosteroids.

Diagnosis of APS is based on a combination of clinical features and diagnostic findings as per revised Sapporo APS Classification Criteria. Clinical criteria include (if anyone present): vascular thrombosis and pregnancy morbidity. Laboratory criteria include (if anyone present): IgG and/or IgM cardiolipin antibody, IgG and/or IgM anti-beta-2 glycoprotein, and lupus anticoagulant.⁴ This patient had clinical criteria of vascular thrombosis and laboratory criteria of positive IgM anticardiolipin antibody.

Hansen's disease is associated with secondary APS with previous documentations of case reports published previously as by Kaliyadan et al. in 2009 from India.⁵ Previous studies of Hansen's



Figs 2A to F: MRI brain on admission

Table 1: Blood investigation chart

Day of admission	Day 1	Day 3	Day 7	Day 10	Day 14	Day 17	Day 21
Hemoglobin	9.5	9.3	9.8	8	7.6	8.5	9.1
TLC	19800 N81L7	12400 N84L12	21300 N84L5	17600 N71L11	15000 N70L15	7600 N62L33	11800 N84L11
Platelet	1.98	1.84	1.69	2.06	3.73	3.24	3.73
CRP/Procalcitonin		8.1/0.14			9.3/0.11		
Urea/Creatinine	57/0.9	72/0.8	45/0.6	32/0.7	30/0.6	39/0.5	36/0.6
Sodium/Potassium	136/4.4	135/4.2	126/4.1	133/3.3	127/4.2	130/4.1	126/4.2
Total/Unconjugated bilirubin	0.8/0.6	0.6/0.5	1/0.7	1.4/0.9	1/0.7		0.6/0.5
Albumin/Globulin	2.7/3.3	2.6/3.2	1.9/2.3	2.5/2.4	2.7/2.8		2.9/3.1
SGOT/SGPT/ALP	175/65/80	66/71/44	36/38/51	29/38/74	27/28/830		25/40/87

L, live; N, neutral

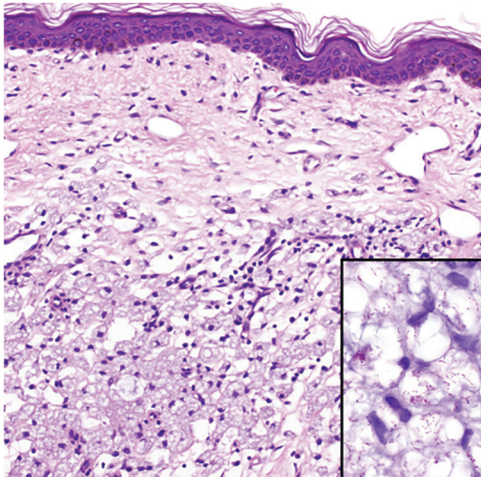


Fig. 3: Skin biopsy

disease-associated APS have shown predominance of IgM subtype of antibodies of APS.⁶

Lucio phenomenon is a sequelae of lepromatous leprosy, commonly more prevalent in neglected patients due to uninhibited multiplication and invasion of bacilli.⁷ Criteria for defining LP as per the international literature are skin ulceration, vascular thrombosis, and invasion of blood vessels by Hansen bacilli⁸ — all of those were present in this patient.

Lucio phenomenon has been previously reported as a rare presentation in patients of Hansen's disease by Sharma et al. and is treated by continuing the multidrug therapy with coverage of high-dose corticosteroids at the initial stages, as was done in this patient.⁹

CONCLUSION

Lucio phenomenon is an unusual presentation of Hansen's disease, a form of cutaneous vasculitis probably mediated by

immune-complex deposition and present as large, sharply demarcated ulcerative lesions and thrombosis of bigger vessels and dermis, and is treated by antileprotic medications, systemic glucocorticoids, and other supportive medications.

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