LUCIO’S PHENOMENON: A RARE CASE REPORT

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ABSTRACT
Lucio’s phenomenon/erythema necroticans is a reaction pattern that occurs in untreated pure primitive diffuse lepromatous leprosy (PPDL) or lepromatous leprosy. Majority of the case reports are from Central America and the condition is extremely uncommon in the Indian subcontinent. Its clinical features are fairly characteristic and consist of extensive, bizarre, painful ulcerations of the skin with absent constitutional symptoms. The clinical diagnosis is confirmed by histopathology characterized by colonization of the endothelial cell by acid fast bacilli, ischemic epidermal necrosis and necrotizing vasculitis of the small vessels of the superficial dermis. Its precise pathogenesis is still unclear but most accepted is free replication of M. leprae in endothelial cells, and enhanced exposure of mycobacterial antigen to circulating antibodies, resulting in vasculitis and infarction. We report a case of lepromatous leprosy with Lucio’s phenomenon from India which unfortunately progressed to gangrene of the limbs requiring amputation.

Key Words: Necrotizing Erythema, Lepromatous leprosy, Vasculitis

INTRODUCTION
Lucio phenomenon also known as necrotizing erythema is believed to be a type 2 reactional states that complicates lepromatous leprosy or borderline leprosy that have not received specific treatment or have been irregularly treated. It was first described in 1852 by Lucio and Alvarado as severe, necrotizing cutaneous reaction in diffuse, nonnodular leprosy. It was named as “Lucio phenomenon” in 1948 by Latapi and Zamora and was noted to occur solely in “pure and primitive diffuse lepromatosis.” Lucio leprosy is a variant of Virchowian leprosy (VL) characterized by erythematous-violaceous macules and hemorrhagic blisters, but its main characteristic feature is diffuse cutaneous infiltration, without nodule formation, generating a brilliant, moist and myxedematous complexion, imparting a healthy aspect to the patient. As a result of these characteristics it is also called as “Pretty Leprosy”. Very few cases have been reported in the International literature which emphasizes the rarity of this event. We report an undiagnosed lepromatous leprosy case which progressed to Lucio’s phenomenon culminating in death of the patient.

CASES
A 60 year old woman presented with on and off multiple skin rashes, fever, and joint pains since 4 years. The skin lesions initially appeared over both upper limbs which gradually progressed to involve the trunk and lower limbs in a span of one year. The lesions were erythematous and asymptomatic. Fever was of intermittent type, not associated with chills and rigor. Patient also had joint pain involving both knee and elbow joints. She had received treatment at a local private hospital as a case of vasculitis, with intermittent oral prednisolone 30 mg and NSAIDS. Following which there was temporary relief of the symptoms, with flare up of skin lesions when she discontinued treatment. On cutaneous examination there were multiple erythematous papules and infiltrated plaques distributed bilaterally symmetrical over chest, back (Figure 1&2) abdomen, trunk, both upper arms and thighs including palms. Superficial brown atrophic scars were also seen in between the plaques (Figure 3). There was no thickening of the peripheral nerves or sensory loss. Systemic examination was unremarkable.
Case Report

Figure 1: Multiple erythematous papules over chest

Figure 2: Bilateral symmetrical distribution of plaques and patches over back

Figure 3: Superficial atrophic scar over right thigh
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Figure 4: Irregular necrotic ulcer with jagged edges

Figure 5: Showing gangrenous changes of tip of the toes
The case was admitted for thorough evaluation & management. The differential diagnosis included vasculitis and chronic granulomatous diseases like Sarcoidosis & Hansen’s. The patient was on oral prednisolone 30 mg since 2 weeks which was stopped on admission. Within two weeks of admission she developed haemorrhagic bullae initially over dorsum of right leg followed by left leg. They broke open to leave behind large necrotic, irregular shaped ulcers over both lower limbs covered with yellow slough (Figure 4). Gradually the tips of the toes developed gangrenous changes (Figure 5).

Laboratory investigations showed anaemia (haemoglobin 8g/dl), ESR 52 mm/hr, CRP was positive and ultrasound abdomen revealed mild hepatomegaly. Rheumatoid arthritis factor, antinuclear antibody, cryoglobulins, coagulation studies, blood sugar, renal and liver function tests, mantoux and chest X-ray were normal. Serology for HIV, HCV, HbsAg and syphilis was negative.

Slit skin smear showed a bacterial index of 6+ with globi from bilateral ear lobes and from the infiltrated plaque. Skin biopsy from the infiltrated plaque showed features of lepromatous leprosy that is atrophic epidermis, grenz zone and clusters of foamy histiocytes infiltrating the subcutis (Figure 6). The ulcerated lesion showed leucocytoclastic vasculitis and invasion of the endothelial cells by AFB both in isolation and globi formation (Figure 7).

**Figure 6:** H & E section showing intact epidermis with subepidermal grenz zone and foamy macrophages in the dermis

**Figure 7:** Fite faraco stained section showing globi of acid fast bacilli within endothelial cell
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The diagnosis of lepromatous leprosy with Lucio’s phenomenon was made based on the highly characteristic necrotic and ulcerated skin lesions and was confirmed by histopathological examination of skin specimens. Multidrug therapy (MDT) for the lepromatous form of leprosy as per WHO along with systemic antibiotics was initiated but within one week the lower limb lesions progressed to gangrenous changes requiring amputation. But the patient refused for amputation and was discharged against medical advice.

RESULTS AND DISCUSSION

In leprosy, the term “reaction” is defined as the appearance of symptoms and signs of acute inflammation superimposed on relatively eventful lesions of leprosy. Pfaltzgraff Re and Bryceson (1985) quoted they represent acute hypersensitivity reactions to antigens of M. leprae. Three types of leprosy reactions have been described by Jopling. They are
1. Type I reactions- is a delayed hypersensitivity reaction which corresponds to the type IV reaction of Coombs and Gell.
2. Type II reactions- antigen antibody mediated reaction involving complement which corresponds to the type III reaction of Coombs and Gell.
3. Lucio’s phenomenon - whose pathogenesis is least, understood. Lucio’s phenomenon itself is not a form of leprosy, but a reactional state that complicates LL or BL leprosy which has been not treated or irregularly treated. Costa et al describes it is endemic in Mexico but has also been reported in the USA, Spain, South and Central America and East and West Asia. According to Pursley and Jacobson (1980), it usually occurs 3 to 4 years after the onset of leprosy. Patients present with manifestations of Lucio leprosy, LL or BL leprosy concomitant with specific Lucio’s phenomenon lesions. Lucio leprosy presents with diffuse skin infiltration which resembles myxedema, loss of eyebrows and eyelashes and thickening of the upper eyelids with shiny eyes giving the patient a sleepy or melancholy look. In addition to these features there may be epistaxis due to ulceration of nasal mucosa and hoarse voice due to laryngeal involvement (Monteiro et al., 2012). However, there was no features of Lucio leprosy in our case.

Lucio phenomenon lesions are characterized by the onset of erythematous-violaceous, ill-defined and infiltrated plaques, breaking down into jagged-edged skin ulcerations that develop fine dark brown scars. The lesions vary in shape, number and size, averaging 0.5–1 cm, with a necrotic, geometric irregular-shaped aspect which is the hallmark lesions of Lucio’s phenomenon. In descending order of frequency the sites involved are the feet, legs, hands, forearms, thighs, arms and, rarely, trunk and face. Our patient presented with infiltrated plaques mainly distributed over trunk and abdomen and there was also involvement of palms which was not found in earlier case reports.
The systemic manifestations of Lucio’s phenomenon include hepatomegaly which was observed in our patient and may be due to the bacillary infiltration of liver observed in LL or BL leprosy. Other manifestations observed were splenomegaly, fever, arthritis and nephritis.

The laboratory findings observed in Lucio phenomenon are anemia, an elevated erythrocyte sedimentation rate, hypoalbuminemia, hypergammaglobulinemia, false-positive serological reactions for syphilis, positive rheumatoid factor, mixed type cryoglobulins, circulating immune complexes and hypo-complementemia. Lucio phenomenon alone might not be responsible for all the laboratory alterations described earlier but several comorbid conditions such as secondary bacterial infections, malnutrition and erythema nodosum may also be responsible.

Rea et al., and Costa describes histological characteristics consistent with Lucio phenomenon. They include colonization of the endothelial cell by acid fast bacilli, ischemic epidermal necrosis, necrotizing vasculitis of the small vessels of the superficial dermis, endothelial proliferation of the medium-sized vessels of the mid-dermis with passive venous congestion, and neutrophilic infiltration. In our case we noticed only vasculitis and endothelial cell infiltration by AFB.
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Among the various hypotheses proposed for the pathogenesis of Lucio’s phenomenon, most accepted is free replication of *M. leprae* in endothelial cells, and enhanced exposure of mycobacterial antigen to circulating antibodies, resulting in vasculitis and infarction (*Calux*).

Others are Arthus phenomenon, mediated by deposition of immune complexes as described by Ouismorio and Shwartzman phenomenon which is bacterial hypersensitivity precipitated by a synergistic bacterial association between *M. leprae* and pyogenic cocci (Moschella).

The diagnosis is basically made on clinical grounds aided by characteristic histopathological findings. However it has to be differentiated from erythema nodosum presenting as necrotizing lesions and leprosy reactional states.

The ulceration and necrosis in erythema nodosum are round, regular lesions and occur always over the nodules. They usually occur after specific leprosy treatment and associated with constitutional symptoms like fever and malaise.

In short, according to the international literature, three criteria are adopted as the rule for Lucio phenomenon definition: cutaneous ulceration, vascular thrombosis and blood vessel walls invasion by Hansen’s bacillus.

There is no specific treatment for Lucio phenomenon. There are reports of using thalidomide, steroids, dapsone, MDT (multidrug therapy) and rifampicin with variable response. Lucio phenomenon is a rare event that may reach severe proportions and cause death by disseminated intravascular coagulation and/or septicemia. Of the few cases reported worldwide, very few are from India, i.e., by Saoji and Kumari (2001). Whether Lucio phenomenon is restricted to Lucio leprosy or it is a reactional manifestation of nodular forms of leprosy needs further study. Although Lucio phenomenon is more common in Central America, in Indian subcontinent also we should have high index of suspicion to diagnose and to differentiate from erythema necroticans in leprosy reactional states.

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