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Case Report

"Lucio Phenomenon or Erythema Necroticans?"- A Diagnostic Dilemma

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Lucio phenomenon and erythema necroticans are a set of peculiar reaction patterns occurring most commonly in untreated or inadequately treated cases of primarily diffuse non nodular and nodular lepromatous leprosy respectively. A 32-year-old male, known case of lepromatous leprosy on irregular treatment for 17 months presented with sudden onset painful hemorrhagic blistering of bilateral distal extremities. There was no history of fever or any other constitutional symptoms. There was a history of similar episode six months back which healed with scarring. Histopathological examination showed focal areas of loss of epidermis with a collection of neutrophils and extravasation of RBCs were present. Dermis showed perivascular lymphocytic infiltrate and necrotic neutrophils and numerous foamy macrophages extending up to subcutis. The clinical features present in our patient showed features of both erythema necroticans and Lucio phenomenon, with histopathological findings suggestive of erythema necroticans, thus presenting to us with a diagnostic dilemma. Thus based on HP a final diagnosis of erythema necroticans was made. The patient was started on a combination of oral prednisolone (40mg/day) and thalidomide (100m g thrice a day), multibacillary multidrug therapy was continued. As the patient got himself discharged against medical advice, his therapeutic response could not be observed. There is need to develop criteria to characterize such cases.

Keywords : Lucio phenomenon, Erythema Necroticans, Leprosy

Introduction

In Leprosy, reactions tend to occur based on the pattern of immune response elicited. Lucio phenomenon/erythema necroticans are rare manifestations of such a response. Necrotic lesions in leprosy occur as a result of severe type 2 reaction. Differentiation between necrotic ENL and Lucio phenomenon can be difficult with cases of overlap being common. Though Lucio phenomenon is known to be endemic in Mexico, cases have been reported from the USA, Spain, Brazil (Furtado 1959) and Asia (Moschella1967). We are reporting a case showing overlapping features of both erythema necroticans and Lucio phenomenon.

Case Report

A 32-year-old male, known case of leprosy on multibacillary multidrug therapy for the last

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seventeen months presented with complaints of sudden onset painful hemorrhagic blisters of 2-3 days duration. Lesions were first noticed over the dorsum of the right hand followed by involvement of the left hand, bilateral lower limbs, and ears. Within two days, he developed ulceration over the same site. There was a history of pedal edema and joint pain restricting his limb movements. The patient developed watering from both eyes and stuffiness of nose for which ophthalmological and otolaryngology examination was done and was found to be normal. However, there was no history of fever or any constitutional symptoms. He was diagnosed with diffuse non-nodular lepromatous leprosy three years back and was started on MB-MDT, but was not regular to the treatment. He gave history of one episode of similar complaints six months back which slowly healed with scarring. There was no history of any major medical illness. On cutaneous examination, lesions were present symmetrically on his upper and lower distal extremities and ears with relative sparing of the trunk, in the form of multiple hemorrhagic bullae measuring 1-2cm in size, with surrounding purpuric patches. Two days later, these bullae ruptured and evolved into irregular ulcers and purpuric infarcts largest being 7x4 cm in size present on right lower limb, few covered with hemorrhagic crust (Figs. 1 and 2). Bilateral helices of the ear showed few vesicles with turbid fluid which evolved to form erosions covered with hemorrhagic crust (Fig. 3). Besides this, there was diffuse infiltration of the face and presence of scars on the buttock (Fig. 4). The peripheral nerves were symmetrically thickened and tender. Bilateral glove and stocking anesthesia was present. Fixed claw hand deformity of both hands was present (Fig. 5). The examination of cranial nerves was normal. Slit skin smear from both earlobes revealed the presence of broken bacilli with an average bacteriological index of +2 (Fig. 6). With the above findings, a provisional diagnosis of Lucio phenomenon with differential diagnosis of erythema necroticans in a known

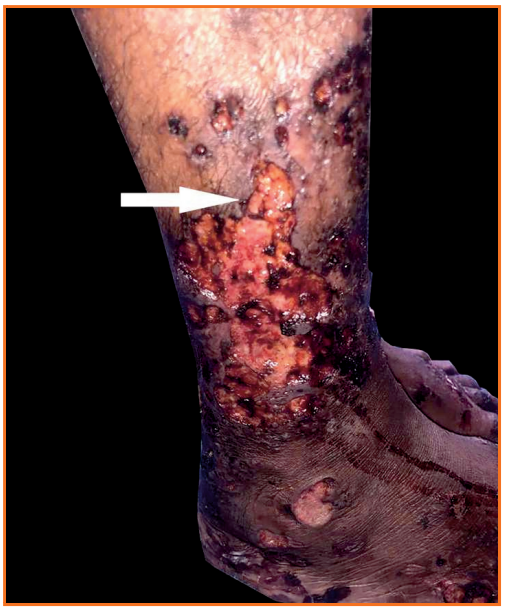

Fig. 1 : Right lower limb showing irregular purpuric infarcts and ulcers

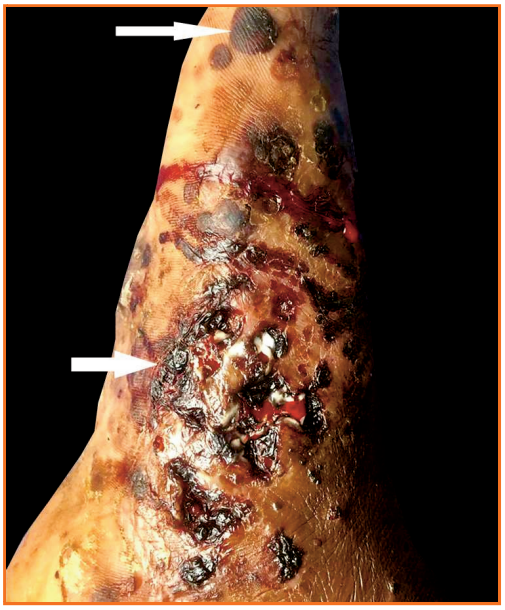

Fig. 2 : Right foot showing multiple hemorrhagic bullae with irregular purpuric infarcts

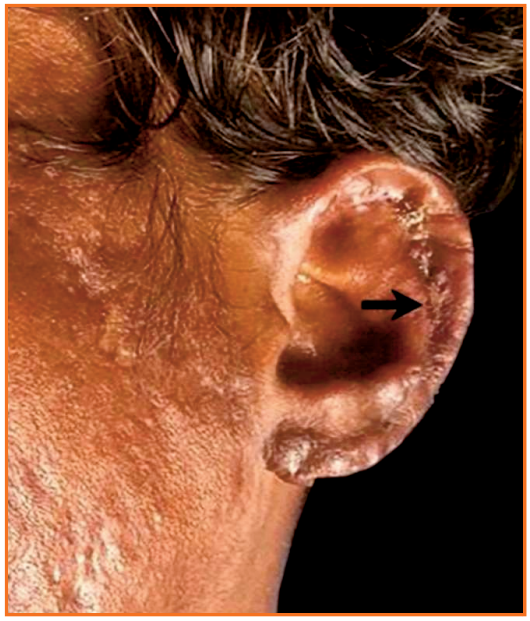

Fig. 3 : Few vesicles with turbid fluid covered with hemorrhagic crust on ears

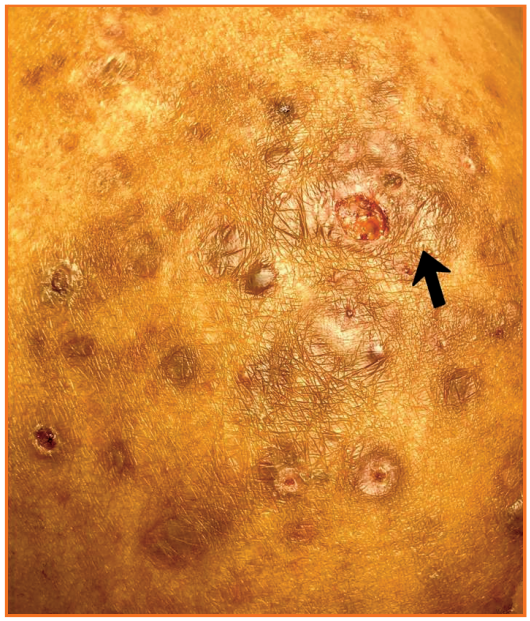

Fig. 4 : Atrophic scars on the buttock

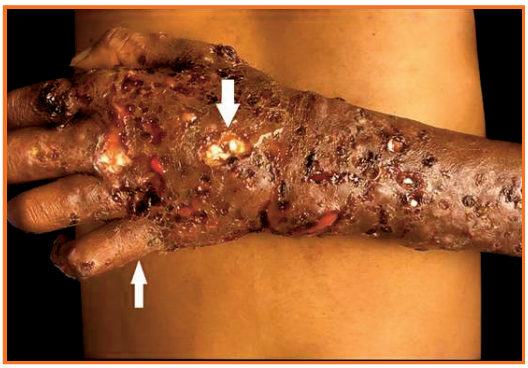

Fig. 5 : Fixed claw hand deformity with multiple purpuric ulcers and hyperpigmented papules

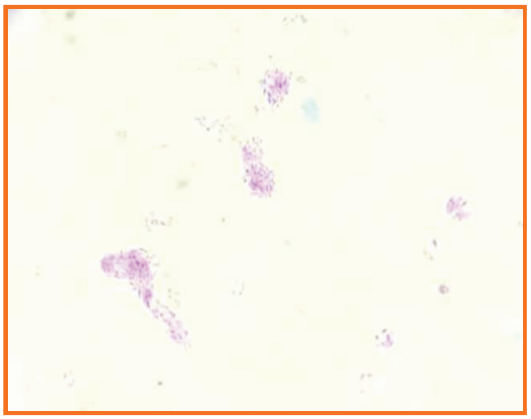

Fig. 6 : Slit Skin Smear showing acid fast bacilli with an average BI 2+

case of diffuse non-nodular lepromatous leprosy on irregular treatment with grade 2 deformity of limbs was made.

Complete blood count showed normocytic normochromic anemia with neutrophilic leucocytosis. Liver and renal function test, ELISA for human immunodeficiency virus, Hepatitis B surface antigen were negative. Pus culture was suggestive of MRSA. On histopathology, focal

areas of loss of epidermis with a collection of neutrophils and extravasation of RBCs were present. Dermis showed perivascular lymphocytic infiltrate and necrotic neutrophils (Fig. 7). Presence of foamy macrophages extending up to subcutaneous fat with some

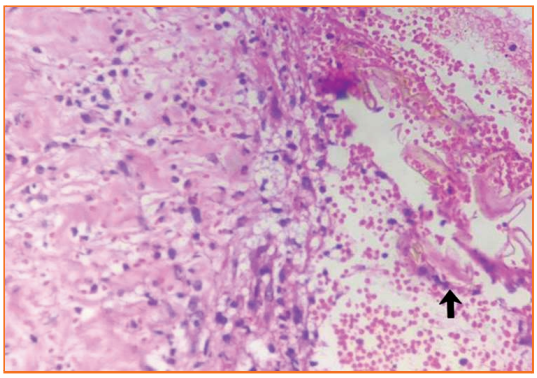

Fig. 7 : Histopathology showing extravasation of RBC's with perivascular lymphocytic infiltrate with necrotic neutrophils in the dermis (H & E ×40)

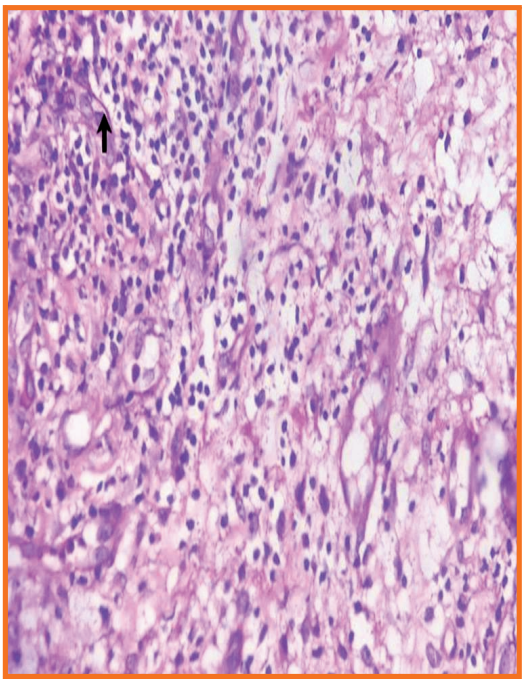

Fig. 8 : Presence of foamy macrophages throughout the dermis (H & E ×100)

degree of septal panniculitis (Fig. 8). However, Fite Faraco stain was negative. Clinically the morphology of lesions was suggestive of Lucio phenomenon but the histological features were suggestive of erythema necroticans. Thus a final diagnosis of erythema necroticans was made. The patient was started on oral prednisolone 40mg/day and thalidomide 100mg thrice a day. MB-MDT was continued. The patient got himself discharged against medical advice.

Discussion

The occurrence of most cases of Lucio phenomenon is limited to patients who are untreated or inadequately treated with diffuse non-nodular lepromatous leprosy (Costa et al 2005). It occurs after an average of 1 to 3 years after the first manifestation of the disease (Ranugha et al 2013, Mareen & Madhukara 2016). It was first described by Lucio and Alvarado in 1852, re-identified by Latapi in 1936, further elaborated by Latapi and Zamora as a form of reaction (Latapi & Zamora 1948, Furtado 1959). The classical description of Lucio leprosy is that of "laprabonita or beautiful leprosy" with no facial hair including eyebrows. Some consider it as a separate form of leprosy and define it as Lucio leprosy, due to its association with *Mycobacterium lepromatosis* in Mexico (Bhattacharjee et al 2020). It is characterized by a diffuse non nodular infiltration of skin with complete alopecia of eyebrows, eyelashes and body hair and the peculiar form of reaction(LP) is characterized by painful, hyperpigmented to bluish, or violaceous patches or plaques

surrounded by erythema which evolve into necrotic angular infarcts and ulcers (Costa et al 2005, Sehgal 2005, Bhattacharjee et al 2020) Severe inflammation can lead to the formation of hemorrhagic blister which breaks down to form deep ulcers as seen in our case (Ranughaet al 2013). Eventually, these ulcers heal with atrophic scarring or hypochromic scars with hyperchromic borders (Rea & Jerskey 2005, Mareen & Madhukara 2016). Constitutional symptoms are usually absent or mild (Mareen & Madhukara 2016). Necrotic lesions in leprosy can also occur as a result of severe type 2 reaction(ENL).

History and clinical findings in our case are not characteristic because of - pedal edema, joint pains to the extent of restricting his movements, watering of eyes and stuffiness of nose and cutaneous features of bullae and ulceration (especially on ears and buttocks). All these features are all more suggestive of necrotic ENL. The clinical features present in our patient showed features of both erythema necroticans and Lucio phenomenon. However, severe necrotic erythema nodosum presents with constitutional symptoms and deep round ulcers which heal with hypertrophic scars (Mareen & Madhukara 2016). It has also been recorded that some patients after the healing of Lucio phenomenon and on regular MDT are known to develop type 2 reactions. This could be one such case. The histopathological features in Lucio phenomenon may vary according to the site and timing of biopsy (Costa et al 2005). Dermal necrosis with the presence of fibrin thrombi with or without vasculitis in the upper dermal vessels can be seen in Lucio phenomenon. Necrotic ENL shows foamy histiocytes, neutrophils with or without vasculitis and panniculitis. Absence of AFB in histopthological specimen in our study is a finding which is difficulty to explain. A considerable amount of overlap in the histological features has been known (Bhattacharjee et al 2020). Lucio phenomenon and necrotic ENL respond to corticosteroids, in addition to MDT (Bhattacharjee et al 2020). Thalidomide is the treatment of choice for steroid nonresponsive ENL. Lucio phenomenon is usually said to be nonresponsive to thalidomide. However, few cases responding to thalidomide have been reported (Bhattacharjee et al 2020).

Our patient presented with histopathological and few systemic features of necrotic ENL, while the morphology of lesions favored Lucio phenomenon indicating a clinical overlap between two entities. This dilemma has been debated earlier also by Indian dermatologists (Sehgal 2005, Ranuga et al 2013). Though Lucio phenomenon is common in Mexico, South and Central America, cases are being increasingly reported from Asia too. Here we report a case showing overlapping features of both necrotic ENL and Lucio phenomenon thus presenting to us with a diagnostic dilemma. As the therapeutic assessment of our patient could not be done, his diagnosis remained partly unresolved. As such cases do occur in our settings, there is need to investigate these cases in detail and come out with appropriate guidelines.

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