e-ISSN: 0975-1556, p-ISSN:2820-2643

Available online on www.ijpcr.com

International Journal of Pharmaceutical and Clinical Research 2024; 16(6); 1852-1854

Original Research Article

Necrotic Erythema Nodosum Leprosum versus Lucio's Phenomenon: A Striking Resemblance!

Vikas Anand[1], Ajoy Kumar Saha[2]

[1]Assistant Professor, Department of Skin and V.D., Jawaharlal Nehru Medical College & Hospital, Bhagalpur, Bihar

[2]Associate Professor and HOD, Department of Skin and V.D., Jawaharlal Nehru Medical College & Hospital, Bhagalpur, Bihar

Received: 25-01-2024 / Revised: 23-02-2024 / Accepted: 26-03-2024

Corresponding Author: Dr. Ajoy Kumar Saha

Conflict of interest: Nil

Abstract:

Vasculonecrotic reactions occour in leprosy as bullae and ulcers usually in BL and LL patients, either as Lucio's phenomenon or erythema nodosum (EN) with necrosis.

Keywords: Leprosy, Bullae, Ulcers.

Introduction

Lucio leprosy is a diffuse non-nodular form of multibacillary leprosy presenting rarely with Lucio's phenomenon, characterized by vascular thrombosis and invasion of blood vessel walls by leprosy bacilli, causing extensive skin ulcers. Type-II lepra reaction [erythema nodosum leprosum (ENL)] is a type III hypersensitivity reaction which commonly manifests as sudden appearance of crops of erythematous, edematous, evanescent, and tender nodules and plaques associated with fever and joint pain. ENL is associated with involvement of multiple organs leading to various complications.

Aim

The aim of our study is to document the clinical differences between the two vasculonecrotic reactions; i.e. Lucio's phenomenon and necrotic erythema nodosum leprosum that occur in leprosy patients and thereby establish clinical criteria as to conclusively differentiate between these two clinically indistinguishable scenarios.

Material and Method

The study was conducted on the patients of leprosy admitted in the in-patient of department of dermatology at Jawaharlal Nehru medical college and hospital, Bhagalpur, Bihar for a period of 6 months from November 2018 to April 2019. During this period, patients who developed bullae and ulcers during the course of treatment were enrolled in the study as subjects and were followed up closely on daily basis for evolution of lesions and systemic involvement so as to establish if the patients were suffering from Lucio's phenomenon or necrotic ENL.

Result

Differentiating these two forms of clinically resembling leprosy can at times be difficult, so we are discussing the differences between the two with the help of a prototype leprosy patient suffering from lepromatous leprosy.

Discussion

A 40 year old male suffering from lepromatous leprosy and on irregular multibacillary multidrug therapy for the past 2 years presented with complaints of multiple ulcers and bullous lesions on the upper and lower extremities for 20 days. The present complaints started with mild fever and joint pains followed by development of multiple erythematous, tender nodules on the extremities. Within a few days the nodular lesions became bullous; and over the next 3-4 days the bullae ruptured leaving ulcers covered with an eschar-like black hemorrhagic crust (as shown in figure 1). Cutaneous examination revealed nodular infiltration of the ear lobes, madarosis, and multiple flaccid bullae on the extremities. There were multiple ulcers of varying sizes with well-defined margins and necrotic bases. Nerves including the greater auricular, ulnar, radial and common peroneal were thickened bilaterally and non-tender. Motor examination was normal. The patient had glove-and-stocking anesthesia.

The patient had been suffering from recurrent episodes of ENL for the past 6 months for which he was on multidrug therapy with tapering doses of steroids. As the patient had developed deep necrotic painful ulcers after starting multidrug therapy associated with constitutional symptoms, visceral involvement, and neuritis; diagnosis of necrotic ENL was confirmed clinically.

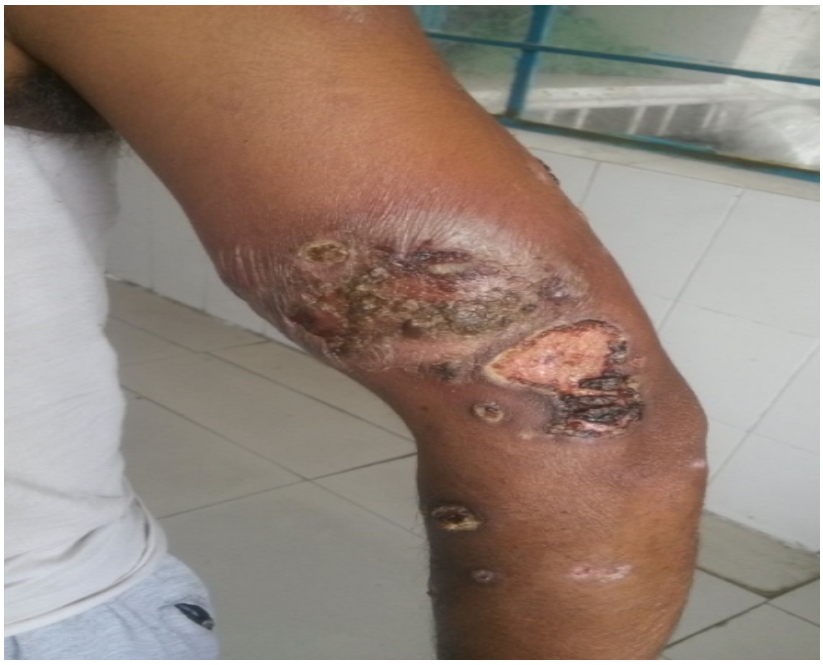

Figure 1:

Figure 1 **:** Proximal lesion over the arm shows a ruptured flaccid bullae forming a shallow ulcer with crusting and the distal lesion over the arm shows an ulcer with well defined edges and an eschar-like black hemorrhagic crust.

Hematological examination revealed leukocytosis with raised ESR. A slit-skin smear and smear from bullous fluid were sent to the Department of Microbiology which showed acid-fast bacilli (as shown in figure 2) with a Bacteriological Index (BI) of 2+ and 3+ respectively. Skin biopsy revealed epidermal spongiosis with neutrophilic infiltration of the dermis and neural and periadnexal infiltration by foamy macrophages, epithelioid cells and neutrophils. Both direct and indirect immunofluorescence studies did not show features of autoimmune bullous disorders. The patient was managed with high-dose steroid and antibiotics with subsequent improvement of the ENL lesions.

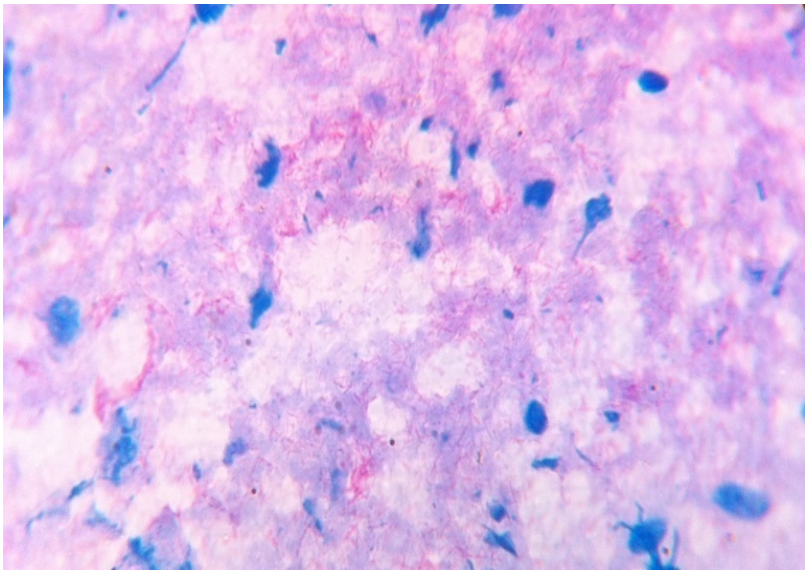

Figure 2: Slit skin smear showing presence of acid fast bacilli

ENL is an immune complex-mediated type III hypersensitivity reaction involving antigen, antibody, and complements. It manifests with erythematous, edematous, evanescent, and tender nodules in crops associated with systemic manifestations [1]. There are various morphological patterns of ENL such as nodular, bullous, pustular and necrotic, with nodular form being the most common [2]. In the present case, the patient presented with both bullous and nodular lesions which lead to necrotic ENL in the form of severe ulceration. The case is interesting both for necrotic ENL which is rarely found and because of formation of extensive scars as a complication.

Reactions in leprosy are divided into two types. Type-1 reaction, which is associated with alterations in cell-mediated immunity and is seen in tuberculoid and borderline groups; and type-2 lepra reaction which occurs in lepromatous leprosy. ENL is encountered during therapy of patients with lepromatous leprosy or borderline lepromatous leprosy and may occur as the presenting manifestation of the disease [3]. Vesiculobullous, pustular, ulcerated, hemorrhagic and erythema multiforme-like lesions have been reported in ENL [4].

Vasculo-necrotic reactions presenting as bullae and ulcers usually occur in BL and LL patients, either as erythema nodosum (EN) with necrosis or Lucio phenomenon [5]. Lucio phenomenon occurs in diffuse, non-nodular form of lepromatous leprosy, chiefly encountered in Mexicans and is seen in untreated patients. Bullae appear in the center of purpuric, painful, red patches as lucio phenomenon with no constitutional symptoms. In contrast our patient had fever and a past history of nodular lesions suggestive of ENL and there was no history of appearance of purpuric lesions at any stage of the disease ruling out Lucio phenomenon.

Lucio phenomenon usually appears in untreated or inadequately treated non-nodular lepromatous leprosy patients after a median of 1 to 3 years after the first manifestations of the disease and presents with erythematous mildly-painful areas over the extremities, which evolve into necrotic, geometric-shaped or jagged-edged ulcers of 0.5 to 1 cm in size [6,7]. There is usually no associated fever, constitutional symptoms, visceral involvement or neuritis. When the inflammation is severe, a hemorrhagic blister is formed, which breaks down to leave a deep ulcer with jagged edges surrounded by an inflammatory zone and usually heal in 2 weeks to form atrophic scars [8]. Vasculonecrotic erythema nodosum usually occurs in multibacillary LL and BL cases after starting multidrug therapy and the patients develop severe, deep painful ulcers associated with constitutional symptoms, visceral involvement, and neuritis and the ulcers heal with fibrotic scars [9] whereas lucio phenomenon presents as characteristic bizarre, geometric, erythematous lesions resulting in superficial ulceration and healing with scars in absence of any constitutional symptoms in a patient with untreated diffuse non-nodular form of leprosy.

Conclusion

Bullous lesions are rarely observed in leprosy and may occur in lucio phenomenon, lazarine leprosy, necrotic ENL or as presentation of sensory neuropathy. Other causes may be heat exposure, associated autoimmune bullous disorder; drug induced bullous reactions and other causes of necrotizing vasculitis. Lucio phenomenon is very rarely reported from India despite the huge load of multibacillary cases due to lack of clinical suspicion and inconsistent features on histopathology. We write this article to highlight the difficulties in differentiating Lucio phenomenon from Necrotic erythema nodosum and to help one and all in conclusively diagnosing between the two forms of leprosy.

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