International Journal of Research in Medical Sciences
Kaur N. Int J Res Med Sci. 2020 Dec;8(12):4509-4511
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012
DOI: https://dx.doi.org/10.18203/2320-6012.ijrms20204971

Case Report

Pathogenesis of bullous erythema nodosum leprosum: a case report

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Received: 27 October 2020
Revised: 01 November 2020
Accepted: 02 November 2020

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ABSTRACT

A 54 years old male, presented with acute onset of fever, malaise and body ache and multiple painful reddish swellings and fluid filled lesions in different parts of body. He gave history suggestive of several earlier episodes of type 2 lepra reactions with erythema nodosum leprosum lesions which were managed with corticosteroids. Dermatological examination revealed multiple erythematous tender nodules and plaques on face, extremities and trunk. He also had multiple bullous lesions on trunk. Investigations revealed polymorphonuclear leukocytosis and raised ESR. Biochemical investigations were normal. Slit skin smear examination showed fragmented acid fast bacilli with bacteriological index of 5+.

Keywords: Bullous, Erythema nodosum leprosum, Leprosy

INTRODUCTION

Leprosy is a chronic, slowly progressive, granulomatous infection caused by *Mycobacterium leprae*. Reactions in leprosy, also known as lepra reaction, are not uncommon. They are of two types; type 1 lepra reaction (occurring in borderline disease) and type 2 lepra reaction (occurring in lepromatous disease).[1]

Skin lesions in lepra reaction generally manifest as exacerbation of existing skin lesions which become more erythematous and edematous as well as appearance of fresh similar lesions (type 1) or appearance of crops of numerous evanescent, erythematous, tender nodules and plaques (type 2). Bullous lesions are a rarity in lepra reactions. Recently we came across a case of leprosy with type 2 lepra reaction having bullous lesions which prompted us to report the case.[2]

CASE REPORT

A 54 years old male, presented with acute onset of fever, malaise, body ache and multiple painful reddish swellings and fluid filled lesions in different parts of body of 12 days duration. He gave history suggestive of several earlier episodes of type 2 lepra reactions with erythema nodosum leprosum (ENL) lesions which were managed with moderate to high doses of corticosteroids with improvement. However attempts to taper off the steroids were never successful. For the first time on this occasion he had also developed multiple fluid filled lesions. There was no history of having any precipitating factor for the lepra reaction.

Dermatological examination revealed multiple bilateral almost symmetrically distributed erythematous tender nodules and plaques on face, extremities and trunk. He also had multiple bullous lesions on trunk. In the beginning the bullae were tense containing clear fluid but latter they became flaccid, and ruptured to form erosions and crusts. Nodules and plaques were evanescent and recurrent, individual lesions lasting for few days, later healing with hyper pigmentation. Nikolsky's sign and bulla spread sign were negative. Infiltrations were present over earlobes and in eyebrow region with supraciliary madarosis. Bilateral ulnar and common peroneal nerves were thickened

uniformly but non-tender. There was no mucosal involvement. There was no feature suggestive of neuritis, iridocyclitis, orchitis or any other systemic involvement.

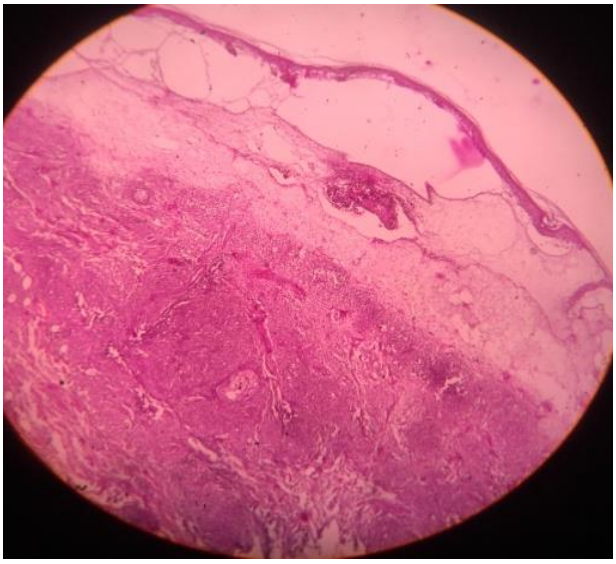

Figure 1: Bullous ENL of subepidermal bulla and sheets of foamy macrophages, madarosis, facial infiltration seen in leprosum lesions.

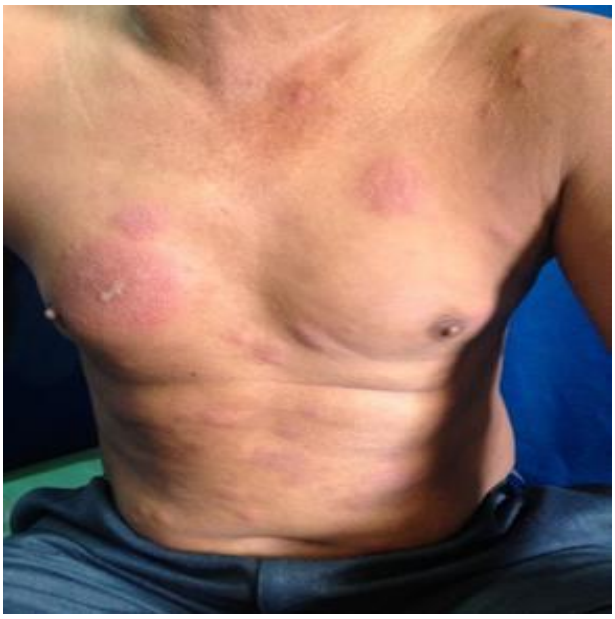

Figure 2: Type 2 erythema nodosum leprosum.

Investigations revealed polymorphonuclear leukocytosis and raised ESR. Biochemical investigations were normal. Slit skin smear examination showed fragmented acid-fast bacilli with bacteriological index (BI) of 5+. Skin biopsy from the bullous lesion showed focally thinned out epidermis and shows sub-epidermal bulla containing neutrophils. Dermis showed diffuse sheets of foamy macrophages with sprinkling of neutrophils.

Patient was managed with a course of systemic corticosteroid starting at 60 mg/day and tapered over a period of month, antileprosy drugs were continued. He showed good response to this treatment. Both bullous and nodular lesions started healing in about a week's time. He was started on thalidomide 200 mg twice daily for a month and later on 100 mg twice daily for 3 months and steroids were tapered off. There was no recurrence of the ENL lesions during the follow up period of about 6 months.

DISCUSSION

Less than 10 cases of bullous ENL have been reported in literature.[3] The mechanism of bulla formation has been described as due to leukocytoclastic vasculitis or severe dermal oedema. Sethuraman et al reported severe bullous ENL in a 35 year old male which was controlled by intra-venous hydrocortisone.[4]

Lepra reactions reflect abrupt changes in the host parasite immunologic balance and are associated with acute clinical exacerbation.[2] Type 2 reaction is an immune complex reaction and is seen mostly in lepromatous (multibacillary) cases.[5]

During type 2 lepra reaction these antibodies combine with *M. leprae* antigen to form immune complexes which circulate and get deposited in various tissues, activate complement and damage these tissues.[5] Bullous lesions in leprosy may be manifestations of severe ENL reaction in patients having very high bacillary load.[6]

Bullous eruptions have been reported during treatment with rifampicin and dapsone.[6,7] Though pustular, ulcerated, erythema multiforme lesions have been reported in lepromatous leprosy, necrotic bullous lesions are infrequent. Generalized bullous eruptions during treatment with rifampicin and dapsone have been reported in the past. Dharmendra and Ramu have described rare incidence of bullous type lepra lesions like the presentation in our case.[8]

In current case the use of systemic naproxen, prednisolone, and clofazimine were able to control the manifestations of bullous ENL. Naproxen is one of the cheapest NSAID useful in ENL to control moderate degree of pain and inflammation.[9]

Controlled use of systemic corticosteroid like prednisolone rapidly control the manifestations of ENL and it is particularly indicated when ENL is associated with neuropathy like that of our case.[10] But long term use of systemic high dose of corticosteroid is associated with adverse effects and it should be tapered off gradually to avoid further exacerbation. Clofazimine is useful to reduce the dose and duration of therapy with prednisolone and helpful in weaning patients off corticosteroid. Though variously described as drug of choice in ENL therapy use of thalidomide in controlling the disease manifestation is not be encouraged as per WHO guidelines.[11]

CONCLUSION

Bullous lesions are a rarity in lepra reactions. Bullous lesions in leprosy may be manifestations of severe ENL reaction in patients having very high bacillary load. Controlled use of systemic corticosteroid like prednisolone rapidly controls the manifestations of ENL.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Kaur N. Pathogenesis of bullous erythema nodosum leprosum: a case report. Int J Res Med Sci 2020;8:4509-11.