

## Case report:

# Bullous erythema nodosum leprosum : a case report

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### ABSTRACT:

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. He gave history suggestive of several earlier episodes of Type 2 lepra reactions with erythema nodosum leprosum (ENL) 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. Blood investigations revealed polymorphonuclear leukocytosis and raised ESR. Fitefaraco stain was positive and bacteriological index (BI) of 5+ was reported. He 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 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.

**Keywords:** Leprosy, Erythema nodosum leprosum, Bullous

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### 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 borderlinedisease) and the Type 2 lepra reaction (occurring in lepromatous disease).<sup>1</sup> 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.<sup>2</sup>

### 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.

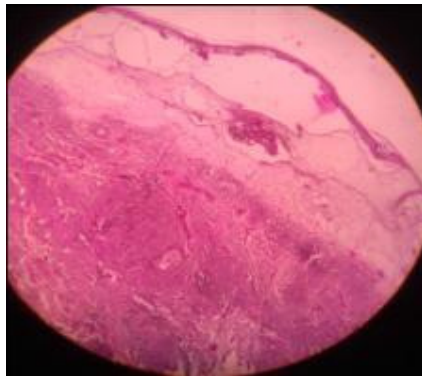
Dermatological examination revealed multiple bilateral almost symmetrically distributed erythematous tender nodules and plaques on face, extremities and trunk. He also had multiple bullous lesions on trunk. In the beginning the bullae were tense containing clear fluid but later they became flaccid, and 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.

Blood Investigations revealed polymorphonuclear leukocytosis and raised ESR. Fitefaraco stain was positive and bacteriological index (BI) of 5+ was reported. Skin biopsy from the bullous lesion showed focally thinned out epidermis with sub-

epidermal bulla containing neutrophils. Dermis showed diffuse sheets of foamy macrophages with sprinkling of neutrophils.

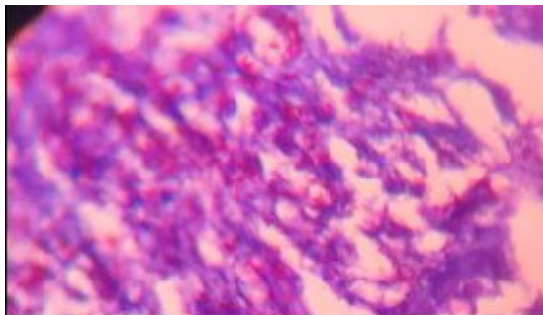
He was managed with a course of systemic corticosteroid starting at 60 mg/day and tapered over a period of month, antileprosy drugs were continued. He showed good response to this treatment. Both bullous and nodular lesions started healing in about a week's time. He was started on thalidomide 200 mg 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.



**Fig 1 : Bullous ENL showing subepidermal bulla and sheets of foamy macrophages in dermis .H &E (10X)**



**Fig 2 : Madarosis, facial infiltration**



**Fig 3 : Lepra bacilli with Bacillary index 5+ Fitefaraco stain**

## DISCUSSION:

Less than 10 cases of bullous ENL have been reported in literature.<sup>3</sup>The mechanism of bulla formation has been described as due to leukocytoclasticvasculitis or severe dermal oedema. Sethuraman et al.reported severe bullous ENL in a 35 year old male which was controlled by intravenous hydrocortisone.<sup>4</sup>

Lepra reactions reflect abrupt changes in the host parasite immunologic balance and are associated with acute clinical exacerbation.<sup>5</sup>

Type 2 reaction is an immune complex reaction and is seen mostly in lepromatous (multibacillary) cases.<sup>6</sup>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.<sup>7</sup>

Bullous lesions in leprosy may be manifestations of severe ENL reaction in patients having very high bacillary load.<sup>8</sup>Bullous eruptions have been reported during treatment withrifampicin<sup>9</sup>and dapsone.<sup>10</sup>

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.<sup>11</sup>

In this 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.<sup>12</sup>Controlled use of systemic corticosteroid like prednisolone rapidly control the manifestations of ENL.<sup>13</sup> 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.<sup>14</sup>

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