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Case Report

Bullous Erythema Nodosum Leprosum: A Rare Entity

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Erythema Nodosum Leprosum (ENL) usually presents in patients with Lepromatous Leprosy. It commonly presents as tender erythematous nodules but other rare forms like pustular, vesicular, bullous or necrotic have also been reported. We report a case of a 30 year old male who initially presented with erythematous tender nodules and plaques over both lower limbs with a past history of being treated for Lepromatous Leprosy. He further developed bullous lesions over the same sites. Biopsy specimen of a bulla was sent for histopathology which was suggestive of ENL. This presentation of ENL makes it an interesting case to be reported.

Keywords : Bullous ENL, Lepromatous Leprosy, Erythema Necroticans

Introduction

There are two types of reactions seen in Leprosy, Type-1 Lepra Reaction (usually seen in borderline leprosy) and Type-2 Lepra which is also known as Erythema Nodosum Leprosum (ENL) and is usually seen in patients with Lepromatous Leprosy (Meyerson 1996). It generally presents after the start of Multi Drug Therapy (Rea & Levan 1975). ENL commonly presents with tender nodular lesions but other forms like pustular, vesicular, bullous or necrotic lesions have been reported in adults (Sethuram et al 2002). Very few case reports of Bullous ENL (Jopling 1984, Periaswami & Rao 1985, Rijal et al 2004) are found in literature, thus, enticing us to report this case. The presentation can be misdiagnosed with other immunobullous conditions like pemphigus or bullous pemphigoid, bullous impetigo, drug reaction, contact dermatitis, arthropod reaction, bullous diabeticorum.

Case Report

A 30 year old male patient presented in the Skin & V.D. Out patient Department in MGM Medical College & Hospital, Aurangabad with multiple erythematous tender nodules associated with swelling over both lower limbs. There was history of fever and joint pain as well. Similar complaints were present since last the four years. Both his Ulnar nerves were tender and thickened and sensations were reduced in the lower limbs. He

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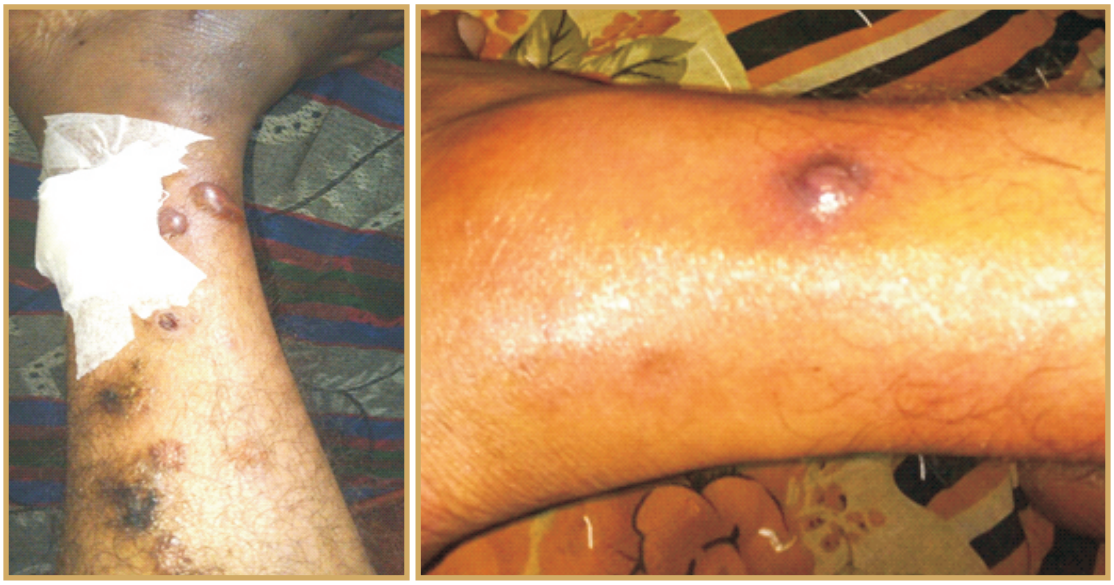

Figs 1 and 2 : Showing bullous lesions over lower extremity.

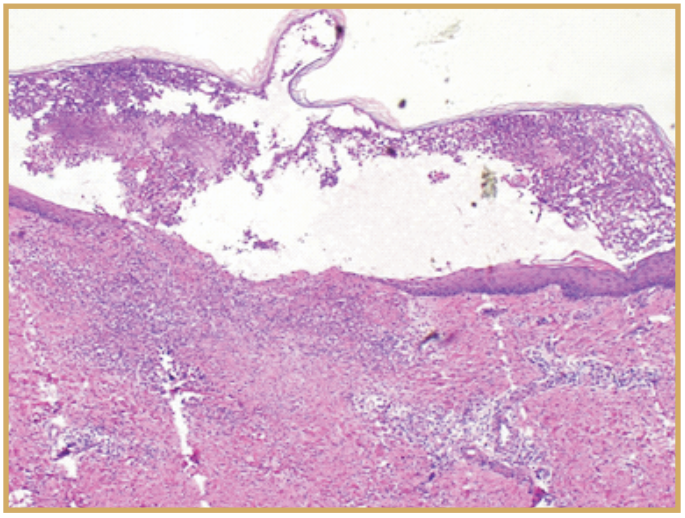

Fig 3 : Epidermis showing marked ballooning with focal areas of necrosis and intraepidermal pustule with granuloma consisting of foamy macrophages and lymphocytes.

then gave us history of being treated with MDT for Lepromatous Leprosy approximately eight to ten years back. In the last four years, patient had taken treatment from outside in the form of steroids and hydroxychloroquine but the symptoms weren't relieved. Biopsy of the lesion confirmed ENL. A few days later, he developed bullous lesions over the same sites with no other

lesions in the oral cavity and no associated complaints of pruritus (Figs 1 and 2). Tzanck Smear showed no acantholytic cells either. A repeat biopsy specimen of a bulla was sent for histopathology which revealed a granuloma consisting of foamy macrophages and lymphocytes with surrounding neutrophils (Fig 3). Patient was started on oral Prednisolone 30mg but new crops continued to present. Patient was then started on Thalidomide 100mg QID while prednisolone was gradually tapered, after which the symptoms began to subside. MDT was started in this patient again.

Discussion

The different types of leprosy are based on the immunological responses of the body to the infection. Lepromatous leprosy represents poor Cell - mediated immunity to *Mycobacterium leprae.* Lepra reactions reflect the changes seen in host - bacteria immunological balance and are associated with acute clinical exacerbations (Pflatzgraff & Ramu 1994). Type 2 Lepra Reaction is an immune complex mediated reaction during which antibodies combine with the bacterial antigen to form complexes. These complexes further get circulated and get deposited in various tissues causing damage. Though multi-drug treatment (MDT) kills most of the bacilli, the elimination of the dead bacilli is very slow (Ridley & Ridley 1983) and this can continue to cause immune perturbations.

ENL is characterized by crops of tender erythematous plaques and nodules over face, trunk and extremities. Other features included are arthritis, synovitis, iridocyclitis, lymphadenitis, epididymoorchitis among others.

The mechanism of bulla formation may be due to leukocytoclastic vasculitis or severe dermal oedema. They may also indicate a high bacillary load. This condition needs to be differentiated from other bullous conditions including bullous drug eruptions, bullous pemphigoid, pemphigus vulgaris.

The sudden appearance of the lesions along with the absence of lesions in the oral mucosa and the absence of acantholytic cells in Tzanck smear help us rule out conditions like pemphigus and bullous pemphigoid. Bullous lesions have been reported during treatment with Rifampicin (Goel & Balachandran 2001) and Dapsone (Dutta 1980), however, our patient was not on either prior to appearance of lesions. The possible trigger in this patient could be stress due to the disease itself and also its effect on his professional life. Thalidomide and Corticosteroids form one of the best lines of treatment for patients with recurrent and chronic ENL. In a case of primary attack of severe ENL, prednisolone alone forms the 1st line of treatment followed by Clofazimine and Thalidomide. (Kar & Gupta 2015) However, in this case, since the patient presented to us as a chronic or recurrent type of ENL and did not show improvement with Prednisolone alone, we chose to start him on Thalidomide while tapering the dose of Prednisolone. The other option in a case like this is substituting Thalidomide with Clofazimine. A combination regimen is always preferred for such cases.

This case is interesting because of its rare presentation of bullous ENL and its improvement on being treated with Prednisolone and Thalidomide.

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