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TYPE 2 LEPRA REACTION WITH ULCERATION (ERYTHEMA NECROTICANS) A CASE REPORT

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HOW TO CITE THIS ARTICLE:

Anila Sunandini, Dhanyasree, Suryanarayana, Subhasini, Padmasree. "Type 2 Lepra Reaction with Ulceration (Erythema Necroticans): A Case Report". Journal of Evolution of Medical and Dental Sciences 2015; Vol. 4, Issue 15, February 19; Page: 2630-2634, DOI: 10.14260/jemds/2015/379

ABSTRACT: Leprosy is a chronic granulomatous disease caused by mycobacterium leprae with wide spectrum of clinical, histopathological and immunological characteristics. Leprosy reactions mainly encountered are type 1 and type 2 lepra reactions. Type 2 lepra reaction, otherwise termed erythema nodosum leprosum, is an acute inflammatory reaction seen in patients with lepromatous leprosy or occasionally in borderline lepromatous leprosy. Rarely in severe type 2 leprosy reaction nodular lesions breakdown to form ulcers. This severe type 2 reaction with ulceration is called erythema necroticans. We are reporting a case with type 2 reaction with ulceration that have responded to thalidomide with no recurrences during 6 month follow up.

KEYWORDS: Type 2 lepra reaction, erythema necroticans, erythema nodosum leprosum.

INTRODUCTION: Type 2 lepra reaction, otherwise termed erythema nodosum leprosum, is an acute inflammatory reaction seen in patients with lepromatous leprosy or occasionally in borderline lepromatous leprosy. Though it is usually seen during the course of treatment it may occur in previously untreated patients as well. The lesions described in type 2 reactions are erythematous, painful, tender papules and nodules. In mild reaction nodules are small in number and spontaneously resolve leaving behind hyperpigmented macules. In severe reactions, nodules tend to increase in size and ulcerate. Ulcerations heal with scarring. Sometimes erythema nodosum leprosum may be the presenting manifestation of leprosy.^[1] Vesiculobullous, pustular, ulcerated, and hemorrhagic and erythema multiforme-like lesions have been reported in ENL (erythema nodosum leprosum).^[2] We are reporting this rare severe type 2 leprosy reaction with ulceration, who was started on thalidomide, prednisolone and chloroquine. He responded well with healing of the ulcers with scarring and without any recurrence of ENL lesions during 6 months of follow up.

CASE REPORT: A 40 year old male presented with multiple ulcers over both upper and lower extremities since 10 days. There was history of recurrent episodes of raised nodular lesions over the skin since 2 years. Each episode is occurring every 15- 30 days, associated with intermittent high grade fever with chills, rigors and arthralgias and the nodular lesions were healing with hyperpigmentation without any ulceration. Present episode there was ulceration of nodular lesions associated with pain and burning sensation. He was taking MB-MDT since 2 years.

On examination patient was febrile. Cutaneous examination [Fig. 1] there are multiple well defined superficial ulcers, irregular, round to oval in shape, size varying from 1x1 cm to 3 x4 cm, well defined margins, floor is covered with dry necrotic crusts and base is mobile. Surrounding skin there was hyperpigmentation, hair loss and decreased sensations. Ulcers were distributed over upper limbs and lower limbs. Few nodular lesions were present on upper limbs and face, tender and firm in

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consistency. Few hypopigmented depressed scars were present over body. Peripheral nerve examination there was bilateral thickening and tenderness of nerve trunks.

Fig. 1: Multiple ulcers over both upper and lower limbs ulcers with dry necrotic crusts.

Fig. 1

Haematological investigations shows mild anemia, elevated total leucocyte counts (TLC- 21, 900/mm³), differential leucocyte count there is polymorphonuclear leucocytosis, blood glucose levels were normal, urine routine examination shows albumin in traces. VCTC & VDRL tests were non-reactive, ESR was elevated. Slit skin smear showed granular forms of bacilli. Biopsy from the nodular lesion showed orthokeratosis, flattened rete ridges, dermis and subcutaneous tissue showed several inflammatory cell infiltrate of lymphocyte, neutrophils and few histiocytes. Fat tissue showed mostly intralobular inflammatory cell infiltrates.

Fig. 2: 10x Dermis & subcutaneous tissue – several inflammatory cells – lymphocytes, neutrophils, few histiocytes No well-defined granulomas.

Fig. 2

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Fig. 3: 40xDermis showing perivascular inflammatory cell infiltrate.

Fig. 3

Patient was diagnosed as severe ENL reaction - erythema necroticans and started on Thalidomide, chloroquine and oral prednisolone. After 2 weeks of treatment ulcers healed with atrophic, hypochromic, angulated depressed scars and no recurrence of nodular lesions during 6 months of follow up.

Fig. 4: Before treatment

Fig. 5: After 2 weeks of treatment

In the post elimination era of leprosy and with effective implementation of MBMDT, we are coming across this rare type of ulcerative leprosy reactions type 1 leprosy reaction which is a very rare entity. Early management of these reactions will decrease disease burden and complications - ENHANCED GLOBAL STRATEGIC plan 2011-2015. This will necessitate the role of dermatologists at tertiary level, in the effective management of leprosy reactions that can even occur with effective implementation of MBMDT at primary health care level.

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DISCUSSION: Leprosy is a chronic granulomatous infectious disease caused by *Mycobacterium leprae*. There is no other human infectious disease in which the clinical picture is as varied as leprosy. Type 2 lepra reactions are a type 3 Coomb and Gell hypersensitivity reaction usually seen in lepromatous and borderline lepromatous patients. It usually occurs later during the course of treatment and in longstanding untreated patients.^[2] Antigen-antibody complexes are formed during the treatment in multibacillary leprosy and in patients with longstanding untreated disease with high bacillary load due to the death of bacilli; deposition of these complexes in various tissues causes inflammatory response with constitutional symptoms.

Over half of Lepromatous leprosy and quarter of borderline lepromatous patients can develop type 2 reaction. In skin it presents most commonly with crops of multiple red dusky brown tender papules and nodules over the face and extensor aspects of the limbs. Lesions tend to recur at the same sites and if they do not resolve completely a chronic painful panniculitis develops which may persist for months to years. Exacerbating factors for ENL are stress, pregnancy, lactation, concurrent illness and medication.^[3] Arthritis, iritis, iridocyclitis and nephritis are other known features of ENL.

ENL is more severe in caucasians and mongolians than in negroes. Ulceration occurs more readily and rapidly and unusual forms are seen. Various rare and atypical variants of ENL have been described in literature. Verma KK et al have reported necrotic erythema nodosum leprosum as a presenting manifestation of leprosy.^[4] A rare variant of type 2 reaction characterised by pustular lesions on switching from WHO MDT to ofloxacin aided MDT was reported by Dave et al.^[5] A persistent and localized variant of ENL was reported by S Prabhu et al.^[6] Galvez et al reported an atypical evolution of BL as well as type 2 reaction primarily involving multiple nerves than skin.^[7]

A case of ENL clinically presenting as urticarial vasculitis that was then diagnosed to have LL has been reported by Funk WK et al.^[8] Kou et al have reported a male with underlying lepromatous leprosy presenting with atypical reactional state simulating sweet syndrome.^[9] A fatal case of erythema necroticans, the cause of death being septicemia secondary to skin ulcers and URTI ENL has been reported by Sethuraman et al and Petro et al.^[10,11] This case is presented due to rarity of ENL leprosy presenting with such severe ulcerations, especially at a time when we have eliminated leprosy as a major health hazard in our country.

Although we encounter type 2 lepra reactions after initiating the multibacillary leprosy patients on MDT-MB, cases of ENL presenting with extensive cutaneous ulcerations are a very rare entity. We are reporting a case of type 2 reaction with ulceration -erythema necroticans with good response to thalidomide, as effective management is crucial to decrease the morbidity of leprosy in this post elimination era.

In the post elimination era of leprosy and with effective implementation of MBMDT, we are coming across this type of ulcerating type 2 leprosy reaction which is a very rare entity. Early management of these reactions will decrease disease burden and complications - ENHANCED GLOBAL STRATEGIC plan 2011-2015. This will necessitate the role of dermatologists at tertiary level, in the effective management of leprosy reactions that can even occur with effective implementation of MBMDT at primary health care level.

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FINANCIAL OR OTHER

COMPETING INTERESTS: None

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Date of Submission: 03/01/2015.
Date of Peer Review: 04/01/2015.
Date of Acceptance: 10/02/2015.
Date of Publishing: 19/02/2015.