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Case Report

A Rare Case of Erythema Nodosum Leprosum Presenting Clinically as Type 1 Reaction

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Leprosy reactions represent acute immunologically mediated inflammatory episodes, which may interrupt the otherwise chronic course of Leprosy. They are of two major types - Type 1 Lepra reaction, or Reversal reaction, and Type 2 Lepra reaction or ENL. Reactional states in leprosy are the most common cause of permanent nerve damage, deformity, and disability. However, atypical presentations of reactions may pose a diagnostic challenge for the treating dermatologist. Here we report an unusual case of Lepra reaction clinically presenting as Type 1 reaction, and having histological features of ENL. The patient presented with erythematous, edematous and infiltrated plaques as the predominant lesions, with a history of recurrent episodes of such lesions, associated with constitutional symptoms. The clinical picture was in favor of BT downgrading to BL leprosy in Type 1 reaction. On the basis of the histopathological findings, a diagnosis of ENL was suggested. However, absence of typical ENL lesions, and clinical resemblance of the lesions to Type 1 reaction makes this case unusual and hence it is being reported.

Key words : Leprosy, Lepra Reaction, Type 1 Reaction, ENL, Atypical Presentation

Introduction

Leprosy is a chronic granulomatous disease, caused by *M. leprae* that affects the skin, nerves and other organs. Reactions in leprosy are acute, immunological exacerbations that occur during the natural course of infection, precipitated by factors like stress, concurrent infections, pregnancy, lactation, and institution of drug therapy. They are of two major types - Type 1 Lepra reaction, or Reversal reaction, which is a type IV hypersensitivity reaction, and Type 2 Lepra reaction or Erythema Nodosum Leprosum (ENL), which is a type III hypersensitivity reaction. Rarely, a third type of Leprosy reaction may be seen, called Lucio phenomenon.

Reactional states in leprosy are the most common cause of permanent nerve damage, deformity, and disability. However, atypical presentations of reactions may pose a diagnostic challenge for the treating dermatologist. A number of unusual presentations of ENL have been reported previously in literature. Awareness of these atypical variants and prompt diagnosis and treatment is of utmost importance to prevent complications in

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potentially treatable patients.

Here we report an unusual Lepra reaction clinically presenting as Type 1 reaction, and having histological features of ENL.

Case Report

A 55 year old male came with complaints of red, painful, diffuse lesions over face, trunk and extremities since 15 days. The lesions had initially started 3 years back, as episodes of red, painful, nodular lesions, recurring every 2-3 months, associated with fever, chills and generalized body ache during each episode. There was history for slippage of footwear while walking, and difficulty in buttoning of shirt since 1.5 years. The patient had previously taken treatment from some local doctor for 2 years, details of which were not available.

General Physical Examination : On general examination, patient was febrile (101° F). Diffuse edema was present over face and pitting edema on bilateral feet.

On Cutaneous examination, a single, large, well defined, erythematous plaque with central sharp margin and peripheral sloping margins was present over the left side of the back, with few satellite lesions (Fig. 1). Multiple erythematous, infiltrated plaques were present in symmetrical distribution over rest of the back, shoulders, chest and abdomen. (Figs. 2, 3, 4). There was diffuse infiltration and thickening of the skin over face (Fig. 5). Multiple hyperpigmented patches were present on back and extremities.

Other important findings were :

(i) A single deep ulcer was present on the left lateral malleolus, with a raised hyperkeratotic rim and reddish granulation tissue.

(ii) Sensory examination revealed hypo aesthesia over dorsum of both hands and loss of sensation to pin prick over both lower limbs, extending up to knees.

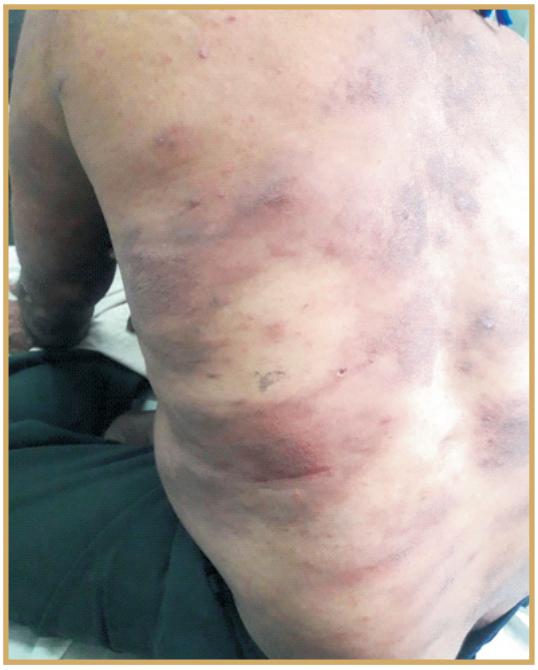


Fig. 1 : Showing a large erythematous plaque with central sharp margin and peripheral sloping margins; suggestive of borderline leprosy

(iii) Nerve examination showed bilaterally thickened and tender ulnar nerve, of grade 2 severity. Other peripheral nerves were not palpable.

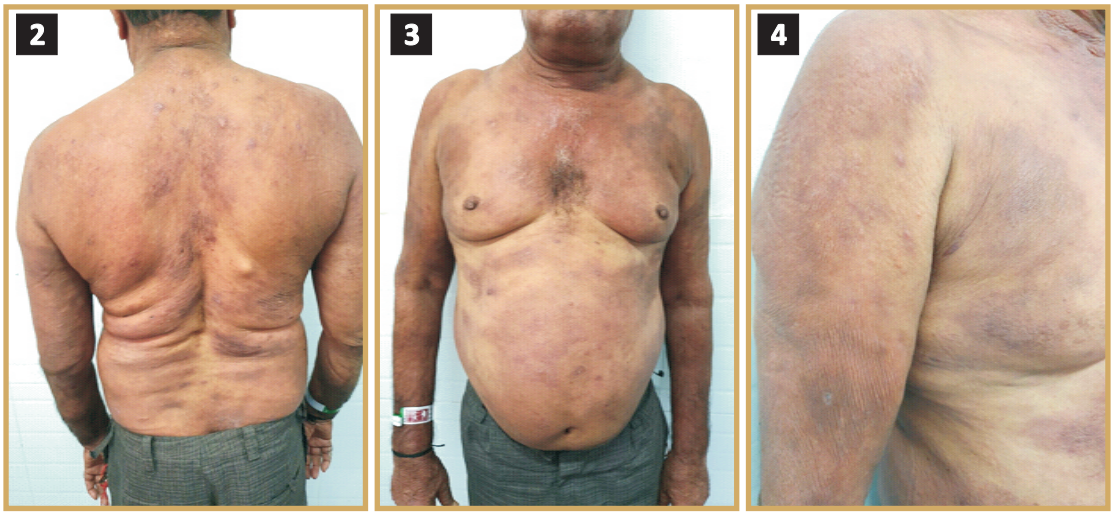
(iv) Earlobes showed infiltration, and madarosis was also present.

(v) On ocular examination, mild watering and conjunctival congestion in both eyes was noted. Slit lamp examination and fundoscopy revealed no other abnormalities.

(vi) Card Test and Book test in this patient were positive.

On the basis of history and clinical examination, a diagnosis of Hansen's in type 1 reaction (downgrading) was made.

Histopathological examination : Skin biopsy was taken from one of the infiltrated lesions over



Figs. 2, 3, 4 : Showing multiple erythematous, infiltrated plaques over back, shoulders, chest and abdomen

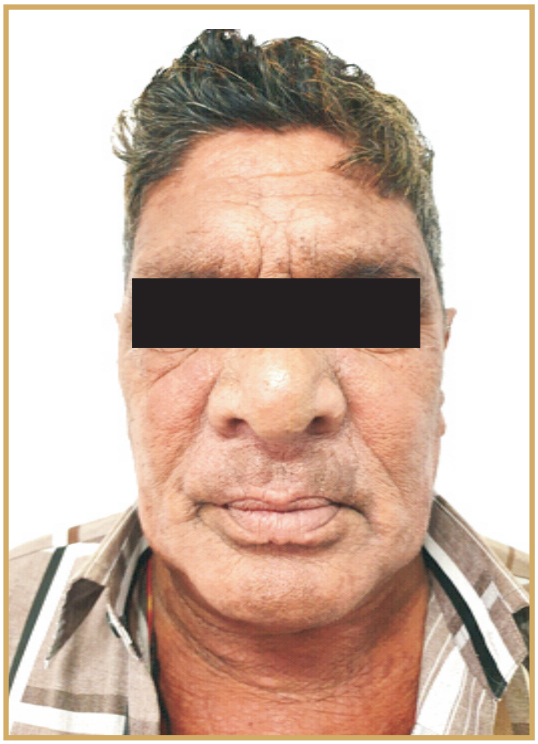


Fig. 5 : Diffuse infiltration and thickening of the facial skin

back, which showed presence of acid fast bacilli. Nodular granulomatous inflammation was seen, centered around the neurovascular bundles of superficial and deep dermis. The granuloma consisted of foamy macrophages and lymphocytes. Numerous neutrophils were seen dotting the granulomas accompanied by moderate amounts of nuclear dust and fibrin (Fig. 6). A diagnosis of ENL was suggested on the basis of histopathological findings.

Hematological examination showed leucocytosis (17000/cmm) with neutrophilia (88%).

Patient was started on Multibacillary Multidrug therapy for leprosy, along with systemic corticosteroids and Thalidomide. He was followed up after a duration of 4 weeks, and showed good response, with decreased infiltration of lesions. No new lesions were observed.

Discussion

Leprosy reactions represent acute immunologically mediated inflammatory episodes, which may interrupt the otherwise chronic course of Leprosy.

Table 1 : Differences between Type 1 and Type 2 Reactions (Job 1994)

Features	Type 1	Type 2
Type of immunological reaction	Delayed type hypersensitivity reaction	Antigen antibody immune complex reaction (Arthus reaction)
Type of patients affected	Borderline types (BT, BB, BL, rarely subpolar LL)	LL, rarely BL
Constitutional signs and symptoms like fever, malaise, arthralgia, myalgia	None or rare	Common
Type of the skin lesions	Existing skin lesions (few or many or all) suddenly becomes reddish, swollen, warm, painful, tender	Fresh red, painful, tender, cutaneous nodules, plaques. The existing skin lesions remain unchanged
Nerve involvement	Nerves close to skin lesions may be enlarged, painful and tender due to acute neuritis with loss of nerve functions (loss of sensation and muscle weakness). Neuritis may also appear suddenly	Nerves may be affected but not as common or as severe; as in type 1 reaction
Eye involvement	Corneal anesthesia and lagophthalmos (weakness of eyelid closure) may occur due to nerve involvement (5th and 7th cranial nerves)	Internal eye diseases like iritis, iridocyclitis, glaucoma, cataract, are common
Other organs	Not affected	May be affected: (lymphadenitis, epididymoorchitis, painful dactylitis, periosteal pain, myositis, arthritis, and glomerulonephritis, are common

Abbreviations: BT, borderline tuberculoid; BB, borderline borderline; BL, borderline lepromatous; LL, lepromatous leprosy.

Features of Type 1 and Type 2 reactions have been summarized in Table 1.

The histopathological features of acute Type 1 reaction are dermal edema, lymphocytic infiltration in the dermis and disruption of normal granuloma organization (Job 1994). In type 2 (ENL) reactions, the typical histologic changes include neutrophils in the deeper dermis, infiltrating the granulomas and preexisting lepromatous lesions. Neutrophilic infiltrate may extend into the sub cutaneous fat, producing panniculitis, which may be seen in 60-70% of

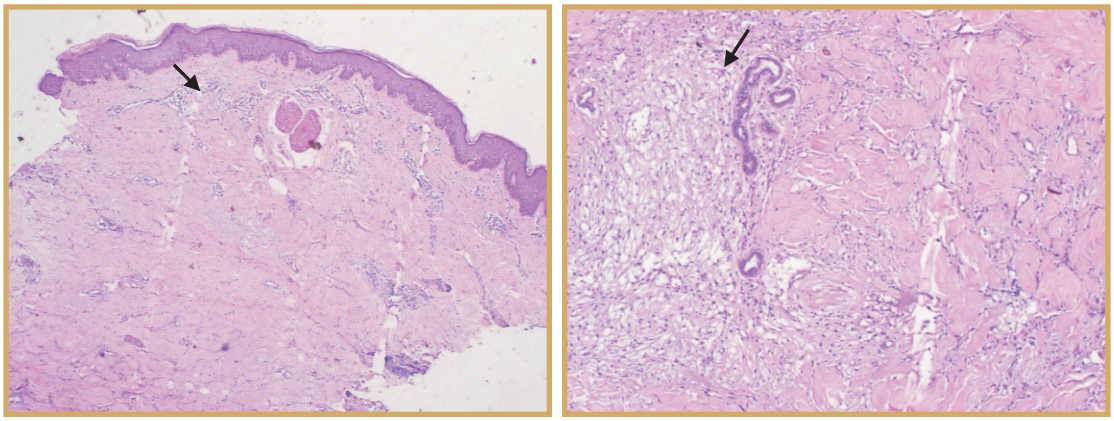


Fig. 6 : Histopathology of skin lesion from back, H& E stain, 10X magnification for (a) nodular granulomatous inflammation in superficial and deep dermis, 100X for (b) Granuloma consisting of foamy macrophages and lymphocytes. Numerous neutrophils dot the granulomas.

ENL cases (Mabalay et al 1965, Adhe et al 2012). There is also associated vasculitis and macrophage degeneration with the breakdown of foam cells (Pandhi & Chhabra 2013).

In a diagnosed case of leprosy, the classical presentation of ENL is easy to diagnose on the basis of clinical findings. However, atypical presentations of ENL may be seen rarely, which poses a diagnostic challenge for the treating Dermatologist/Leprologist. These atypical features may include pustular, ulcerated, hemorrhagic, vesicular or bullous, necrotic, Erythema multiforme-like, Sweet's syndrome-like, or PLEVA - like presentations (Vijendran et al 2014).

In the case being reported here, there were erythematous, edematous and infiltrated plaques present as the predominant lesions. The clinical picture was in favor of downgrading Type 1 reaction.

On the basis of the histopathological findings, a diagnosis of ENL was suggested. However, absence of typical ENL lesions, and clinical resemblance of the lesions to Type 1 reaction makes this case unusual and hence it is being reported.

Similar to our observations, such an overlap between type 1 and type 2 reactions in leprosy has been reported in a clinico-histological study of ENL cases done by Chatterjee (2017) at PGIMER, Chandigarh. They observed 2 patients of BL leprosy with overlapping clinical features, and 3 patients with overlapping clinical as well as histologic features of both type 1 and type 2 reactions. There were 7 cases, in which ENL was not suspected clinically, but biopsy findings led to diagnosis of leprosy with early ENL changes.

In our patient, multidrug therapy for leprosy, systemic corticosteroids along with supportive treatment was started at the time of admission, on the basis of clinical diagnosis of Hansen's in type 1 reaction; Thalidomide was added after the histopathology report suggested a diagnosis of ENL. Corticosteroids, being one of the first line treatment options of ENL, given alone could also show improvement in this case. However, since the patient had a history of recurrent reactional

episodes, Thalidomide was added along with steroids, as it could lead to better improvement and less frequent recurrences.

Conclusion

In conclusion, atypical presentations of type 2 reactions in leprosy are a diagnostic challenge, and may often lead to misdiagnosis and delayed intervention. A high index of suspicion is required for correct diagnosis of such cases. Confirmation with the help of biopsy, and subsequent clinico-histological correlation is essential.

References

1. Adhe V, Dongre A, Khopkar U (2012). A retrospective analysis of histopathology of 64 cases of lepra reactions. *Indian J Dermatol*. **57**: 114-117.
2. Chatterjee D (2017). The diagnostic dilemma of erythema nodosum leprosum - a clinico-histological study. *Lepr Rev.* **88:** 217-226.
3. Job CK (1994). Pathology of leprosy. In: Leprosy, 2nd edn., Hastings RC (Editor) Edinburgh: Churchill Livingstone, pp. 193-234.
4. Mabalay MC, Helwig EB, Tolentino JG et al (1965). The Histopathology and Histochemistry of Erythema Nodosum Leprosum. *Int J Lepr*. **33**: 28-49.
5. Pandhi D, Chhabra N (2013). New insights in the pathogenesis of type 1 and type 2 lepra reaction. *Indian J Dermatol Venereol Leprol*. **79**: 739-49.
6. Vijendran P, Verma R, Vasudeyan B et al (2014). Rare atypical presentations in Type 2 lepra reaction: a case series. *Int J Dermatol*. **53**: 323-326.

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