

Case Report

An Unusual Presentation of Type 2 Lepra Reaction: Mimicking Sweet's Syndrome

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Abstract

Type-2 lepra reaction or Erythema nodosum leprosum (ENL) is a type-III hypersensitivity reaction, which usually occurs in cases of lepromatous and borderline lepromatous leprosy. Classic ENL reaction presents as widespread crops of tender, erythematous and evanescent nodules. Atypical presentations of ENL have been documented. Here, we report an unusual presentation of type 2 lepra reaction simulating Sweet's syndrome, in a twenty seven year old man who had no preceding diagnosis of leprosy, which was diagnosed by slit skin smear and histopathological examination.

Key-words: ENL, lepra reaction, Sweet's syndrome

Introduction

Leprosy is a classic example of a broad spectrum disease with varied clinical manifestations that mimics various other chronic dermatoses. Type 2 lepra reaction, otherwise termed erythema nodosum leprosum [ENL] is an acute inflammatory reaction seen in patients with lepromatous leprosy and borderline lepromatous leprosy. Classic ENL is characterized by the appearance of crops of tender subcutaneous nodules found on face and outer surface of limbs, and usually spontaneously resolves leaving behind hyperpigmented macules.^[1] However it may also present in unusual ways.

Case History

A 27 year old male presented to the OPD with fever of sudden onset and multiple painful skin lesions over both the forearms since past 2 weeks. He initially developed red, painful elevated lesions, few of which rapidly progressed into blisters and superficial ulcers. The lesions initially appeared over the forearms, then spread out on upper arms and the trunk. Patient had associated multiple joint pains. He did not complain of any muscle weakness or sensory loss. He had no preceding diagnosis of leprosy and did not give history of sim-

ilar skin lesions in the past. However there was a history of sore throat one week prior to the onset of these skin lesions.

On clinical examination, patient was febrile, his vitals were stable, and there was absence of pallor, icterus, clubbing, cyanosis, lymphadenopathy and oedema. Cutaneous examination showed multiple erythematous, edematous, tender plaques studded with pseudovesicles as well as frank vesicles. Few of the lesions over the forearms had progressed to form superficial ulcers covered with loosely adherent, brownish-black hyperpigmented crusts. Base of few of the ulcers were necrotic (Fig. 1-3). Few asymptomatic skin coloured papules were found on the posterior aspect of the right earlobe and pinna, without any earlobe infiltration and few normo-aesthetic atrophic plaques were present on the right shoulder. There were no other features suggestive of leprosy. A complete blood count showed leukocytosis and a raised ESR. Rest of the haematological investigations, urine analysis and other biochemical parameters were within normal limits. Slit skin smears for acid fast bacilli (AFB) was taken done from the skin lesions, eyebrows, earlobes, and normal skin and the bacteriological index was found to be 6+, 6+, 4+ and 3+ respectively (Fig. 4). Biopsy taken from the skin lesion showed dense infiltration of the upper dermis and periadnexal structures with histiocytes and lymphocytes. Subcutis showed infiltration by histiocytes, lymphocytes, along with occasional neutrophilic aggregates and necrotic dust. Few dermal blood vessels show-

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ed features of vasculitic features. Fite stain was positive for AFB (Fig. 5).

Discussion

Reactions in leprosy represents episodes of acute hypersensitivity to antigens of *Mycobacterium leprae* brought about by a disturbance in the pre-existing immunological balance. Type 1 reactions are associated with changes in cell mediated immunity and are seen in patients with borderline leprosy whereas Type 2 reactions or ENL are associated with immune complex deposition and occur in lepromatous and borderline lepromatous leprosy.^[2] These reactions are most often seen during the anti-leprosy treatment or after completion of the treatment.^[3] However the reaction can be the initial presentation in untreated Leprosy patients suggesting the need to suspect leprosy in such scenario.

Among type 2 reactions of leprosy rare and atypical presentations described Includes Pustular, bullous, ulcerated and erythema multiforme-like lesions^[4] ENL mimicking Sweet's syndrome is a rare presentation and has been reported as early as 1987 by Kou and Chan.^[5, 6]

This patient presented with an abrupt onset and rapid progression of skin lesions with a history of antecedent upper respiratory tract infection, fever, arthralgia, presence of leukocytosis, and a raised ESR and hence a likely differential diagnoses. of sweet's syndrome was considered.^[7] On further examination the asymptomatic skin coloured papules on the ears and atrophic plaques over right shoulder were noted, which led to the suspicion of leprosy in a reactional state and a Slit skin smear and histopathological examination confirmed this diagnosis. Most of the reported cases simulating Sweet's syndrome have had a prior diagnosis of borderline or lepromatous leprosy but this patient had no such preceding diagnosis of leprosy.^[8]

Fig 1. Erythematous edematous plaques with pseudovesicles and frank vesicles.



Fig 2. Superficial ulcers covered with brown hyperpigmented crust.



Fig 3. Ulcer with a necrotic base



Fig 4. Slit-skin smear showing AFB lying singly and in globi

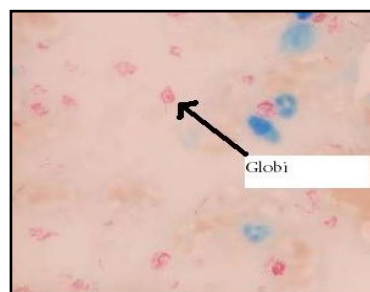
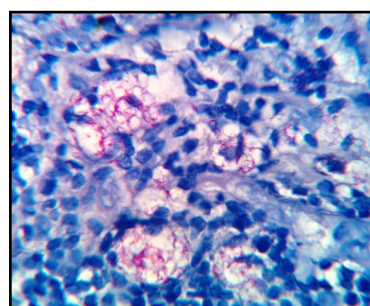


Fig 5. Fite stain showing numerous acid fast bacilli



Conclusion

ENL may some times present in unusual ways. A high index of suspicion and awareness of the various atypical presentations of lepra reactions by clinicians is required to make an early diagnosis to prevent severe morbidity and also mortality due to the disease.

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