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Case Report

Unusual Presentation of Necrotic Erythema Nodosum Leprosum on Scalp: A Case Report

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Lepra reactions are acute episodes occurring during the disease process of leprosy and are of 2 types: type 1 or reversal reaction and type 2 reaction or erythema odosumleprosum (ENL). In the episodes of lepra reaction several parts are affected including face and extremities like oral cavity. In the present case report we reported a rare case of lepromatous leprosy with necrotic ENL involving scalp apart from the usual sites. A 58 year old married male presented to us with complaints of spontaneous onset, recurrent eruption of multiple reddish raised painful lesions. Biopsy from the infiltrated skin over the back showed atrophic epidermis, free Grenz zone, diffuse and periadnexal macrophage granulomas with predominant mononuclear infiltrate, appandageal atrophy, fibrosis around the neural structures and leukocytoclastic vasculitis. Fites stain showed strong positivity for *M. leprae.* His routine blood investigations showed anemia (Hb=7.8 gm%), neutrophil leukocytosis (TLC=17,600, DLC=P66L28M4E2) and raised ESR (80 mm in the first hour). These bullous and necrotic lesions in leprosy may be a manifestation of severe type II reactions in patients with very high bacillary load.

Key word: Lepromatous Leprosy, Erythema nodosumleprosum, scalp

Introduction

Lepra reactions are acute episodes occurring during the disease process of leprosy and are of two types - type 1 or reversal reaction and type 2 reaction or erythema nodosumleprosum (ENL). Type 2 reactions are seen in lepromatous leprosy and borderline leprosy and classically present as evanescent crops of tender plaques and nodules; however vesicular lesions, bullous lesions, necrotic and ulcerative lesion have also been described predominantly from Mexico and South America. Few reports of necrotic and bullous ENL (Dogra et al 2002, Barman et al 2005) have also been published from India. Though face and extremities are commonly affected, it has been reported from unusual sites like oral cavity (Swain et al 2008). We reported a rare case of lepromatous leprosy with necrotic ENL involving scalp apart from the usual sites.

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Case

A 58 yr old married male presented to us with complaints of spontaneous onset, recurrent eruption of multiple reddish raised painful lesions since 20 days. Patient was apparently well 20 days back when he noticed sudden appearance of painful reddish raised pea sized lesions on abdomen and chest. Lesions soon turned dark black in the centre, followed by ulceration. New lesions subsequently appeared in crops over face, neck, scalp, chest, abdomen bilateral upper limbs and lower limbs. The size of lesions varied from 0.5x 0.5 to 15x 20 cm with largest lesion being present on scalp. Patient also had high grade fever associated with chills and rigors. There was no history of any sensory loss, spontaneous blistering, light coloured lesions over body, nasal stuffing or epistaxis, swelling over feet, bending of fingers of hands, slippage of chappals from foot. There was no history suggestive of intercurrent infections like sore throat, pain abdomen, burning micturition or joint pains. No history of intake of anti leprosy drugs. No past history of similar complaints in self or family.

On general examination, patient was febrile with 101F temperature and had discrete, nontender, 1.5 to 2 cm bilateral inguinal lymphadenopathy. Pitting pedal edema was also present.

Cutaneous examination revealed multiple tenderulcers of size 1x1 cm to 14x8 cm, with irregular border and black necrotic adherent base, present over face, scalp, lower back, upper limb, lower limb (Fig 1). Facial examination revealed madarosis and infiltration over face and eyebrows. Scalp showed extensive necrotic ulcerative lesions present diffusely over frontal and occipital areas. There was patchy loss of sensation in glove and stocking distribution. Card test was positive in right hand. Peripheral nerves examination revealed symmetrically thickened, non tender ulnar and common peroneal nerves. There was no evidence of testicular or ocular involvement. Slit skin smear showed clumps of acid fast bacilli with bacteriological index 5+.

Biopsy from the infiltrated skin over the back showed atrophic epidermis, free Grenz zone, diffuse and periadnexal macrophage

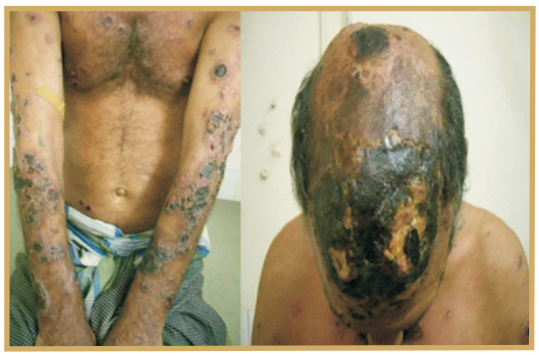

Fig 1 : Cutaneous examination revealed multiple tender, necrotic ulcers of size 1x1 cm to 14x8 cm, with irregular border and black necrotic adherent base, present over face, scalp, lower back, upper limb, lower limb

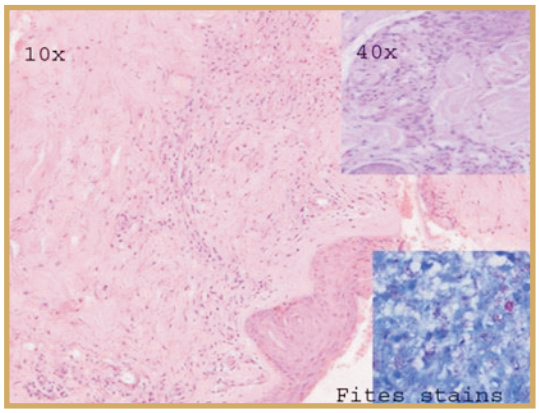

Fig 2 : Biopsy from the infiltrated skin over the back showed atrophic epidermis, free Grenz zone, diffuse and periadnexal macrophage granulomas with strong fites stain .

granulomas with predominant mononuclear infiltrate, appandageal atrophy, fibrosis around the neural structures and leukocytoclastic vasculitis. Fites stain showed strong positivity for M. *leprae.* (Fig 2). His routine blood investigations showed anemia (Hb=7.8 gm%), neutrophil leukocytosis (TLC=17,600, DLC= P66L28 M4E2) and raised ESR (80 mm in the first hour). Other laboratory investigations including liver and kidney function test, blood sugar, urine examination, throat swab, ASLO, ultrasound abdomen and chest X ray were within normal limits. Based on clinical findings and examination a diagnosis of lepromatous leprosy with type 2 reaction was made. Patient was admitted and started on multibacillary multi drug therapy without dapsone in view of anemia and steroids 60 mg per day along with thalidomide 100 mg tds. Thalidomide was started early in treatment in view of severe necrotic ENL. New lesions stopped appearing and older lesion started healing within 2 weeks of starting treatment and subsequently thalidomide and steroids were tapered. Patient received MB MDT without dapsone for initial one month, after which his hemoglobin increased and dapsone was started.

Discussion

Typr II lepra reactions and type III hypersensitivity reactions occurring due to high release of bacterial antigen load from dying bacilli, leading to immune complex formation and TNF-α secretion. It usually occurs later during the course of treatment or in longstanding untreated patients (Meyerson 1996). Though classically lesions present on face, extremities and trunk; as plaques and nodules haemorrhagic, vesicular, erythema multiforme like, bullous, necrotic and ulcerated lesions have also been reported rarely (Dogra et al 2002) .Vashisht et al reported a rare presentation of bullous ENL reaction on trunk in a multibacillary leprosy case on MBMDT (Vashisht and Das 2013). Ramesh et al (1996) reported the occurrence of multiple ulcers, necrotizing type 2 reactions in a 57 year old patient. Verma et al (1993) also described necrotic erythema nodosum leprosum as one of the presenting manifestation in lepromatous leprosy. Swain et al in 2008 described a case of necrotic erythema nodosum leprosum lesions in oral cavity along with involvement of usual sites. Rare occurrence of necrotic erythema nodosum leprosum in a child was reported by Pandhi et al (2005) .These bullous and necrotic lesions in leprosy may be a manifestation of severe type II reactions in patients with very high bacillary load.

Our case also had this rare presentation of necrotic and ulcerated ENL lesions over the body, other than plaques and nodules. Also, in addition to the involvement of typical sites, he had ulcerative ENL lesions over the scalp which is a very rare site of involvement in leprosy.

Scalp is considered as one of the absolutely immune zones of leprosy, however, there are few cases reported in literature with the involvement of leprosy in scalp, but no case have been reported of involvement of scalp with necrotic/ ulcerative type 2 reactions of leprosy to the best of our knowledge.

The ulcerative lesion some time may mimikewith cuntaneous necrotizing vasculitis. They can be differentiated by the absence of other features of leprosy, negative slit skin smears, and negative findings in tissue biopsy.

This case has been presented for rarity of necrotic ulcerated ENL reaction in a case of multibacillary leprosy and its presence on scalp along with other usual sites.

Thus though incidence of leprosy is showing a decline, the clinical manifestations of the disease are changing, and thus a clinician should always

bear in mind these unusual presentations and unusual site involvement in leprosy to avoid missing the new cases and ensuring proper treatment to all cases.

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