Indian J Lepr 2018, 90 : 313-318

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Case Report

Facial Lazarine Leprosy in Post-elimination Era: A Case Report

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Received : 26.03.2018 Accepted : 17.09.2018

Despite leprosy being eliminated worldwide, a reasonable number of new cases are encountered in many countries like India. Reactions in leprosy are quite common which contributes significantly to morbidity and disabilities in patients suffering from leprosy. Two types of reactions can occur in leprosy patients depending on host's immune response against *Mycobacterium leprae*. Lazarine leprosy is a rare ulcerating form of leprosy which is considered as an exaggerated type 1 reaction. We are reporting a case of facial lazarine leprosy in a normal immunocompetent person without any underlying malnutrition. This case presented with infiltrated, oedematous plaque with ulceration and crusting, features which are suggestive of Lazarine leprosy but can also be indicative of severe type 1 reaction. Aggressive nature of the lesion and ulceration were taken into consideration to diagnose this case as Lazarine leprosy.

Key words : Lazarine, Type 1 reaction, Leprosy, Ulceration

Introduction

Leprosy is a chronic infectious disease caused by *Mycobacterium leprae*, which primarily affects skin and peripheral nerves. The relatively uneventful chronic course of the disease is interrupted by immunologically mediated acute and sub-acute inflammations called as reactions. Lazarine leprosy is a rare form of leprosy characterized by spontaneous ulceration of skin lesions, commonly affecting trunk and extremities (Ramu& Dharmendra 1978). It is considered as an exaggerated type 1 reaction (Kumar & Dogra 2010). Though face is the commonest site for type 1 reaction, very few cases of Lazarine leprosy occurring over face has been reported (Sunandini et al 2015). Herein we report a case of facial Lazarine leprosy.

Case report

A 50 year old female presented to the Dermatology outpatient Department of our hospital

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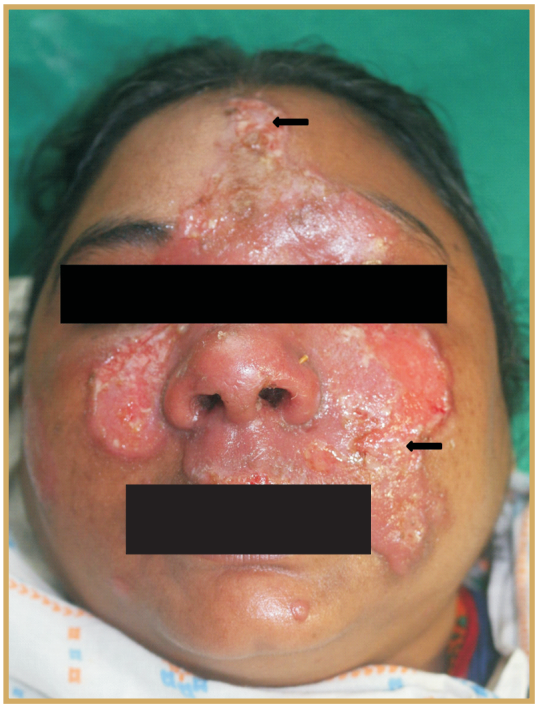

Fig. 1a : Erythematous, oedematous plaque with ulceration (black arrow) and crusting over central part of face.

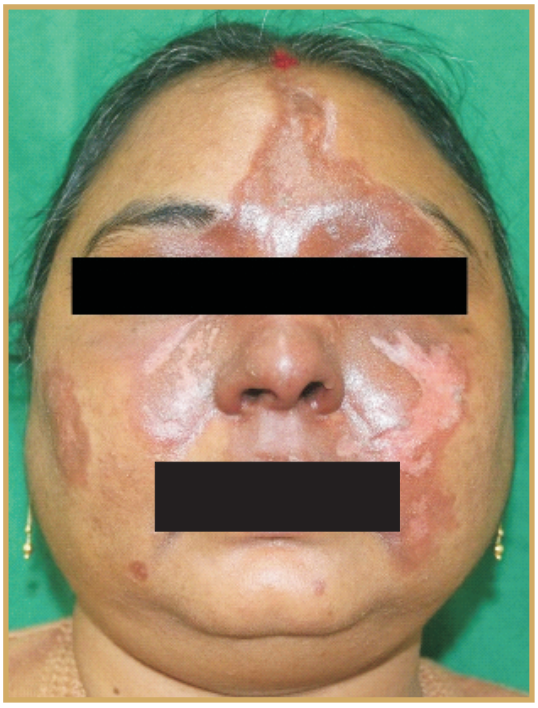

Fig. 1b : Complete healing of the lesion with atrophic scarring after 1 month of prednisolone therapy.

with large ulcerated plaque over centrofacial area for 10 days. Patient was apparently alright 10 months back. To start with she noticed two reddish asymptomatic patches over right cheek and forehead. She took some topical medication from nearby hospital but with no result. Gradually the patch over fore head enlarged to involve entire nose, paranasal area and portion of upper lip over 8 to 10 month period. Since last 10 days she had fever with myalgia along with painful ulceration of the preexisting lesion. She denies any history of infection, vaccination, physical or mental stress. Her medical and surgical history was uneventful. Family and personal history was non contributory. Patient was of average built and nutrition. Local examination showed a large, non tender, erythematous, ulcerated plaque with variable crusting over central part of face extending up to hairline above, upper lip below and malar area on both sides and encroaching left upper eyelid (Fig. 1a). Similar infiltrated erythematous papules were present over chin and left cheek. No feeding cutaneous nerve was present around the plaques. Peripheral nerves were not enlarged and there was no evidence of neuritis or any deformity. Nasal as well as oral mucosae were not involved. Regional lymph nodes were not enlarged. A differential diagnosis of Lazarine leprosy, Ulcerative Lupus Vulgaris, Cutaneous T cell lymphoma, Wegener's granu-

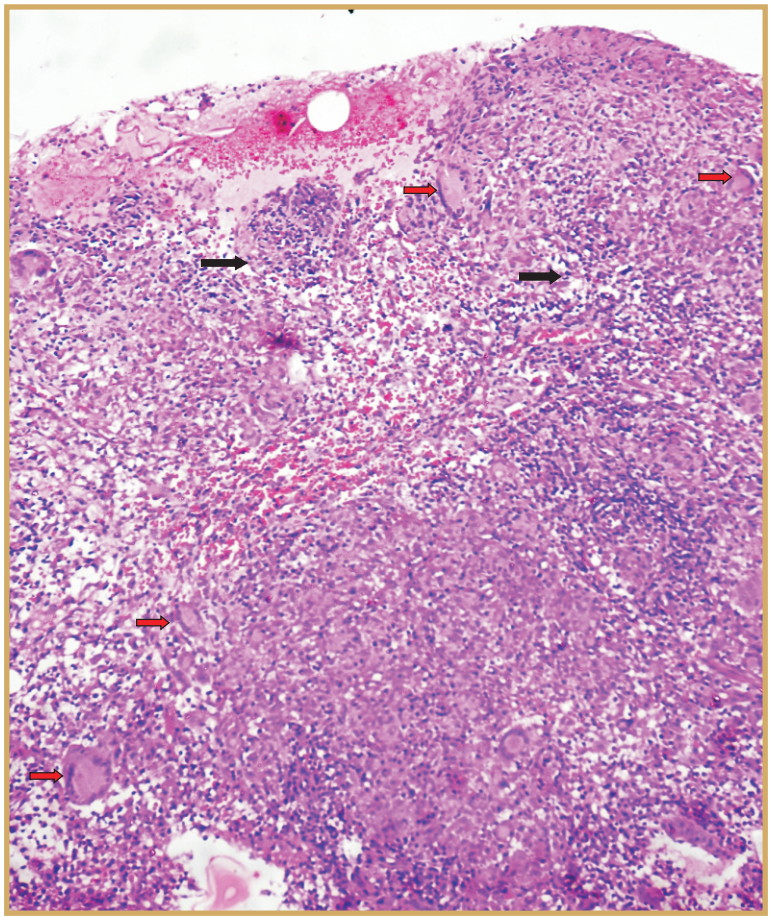

Fig. 2 : Denuded epidermis with upper dermal oedema and disorganized tubercular granuloma (black arrow) with plenty of giant cells (red arrow). (H & E, 10X)

lomatosis was made. Routine haematological and laboratory parameters were within normal limits. Mantoux test revealed an induration of 11 mm. ELISA test for HIV was negative. Tests for Anti-nuclear antibody (ANA) as well as cytoplasmic anti neutrophilic cytoplasmic antibody (c ANCA) were negative. X-ray chest was normal. X-ray skull did not reveal any abnormality in the underlying bony tissue. Gram stain and acid fast bacilli (AFB) stain from pus did not show any organism. Histopathology study of punch biopsy specimen revealed denuded epidermis, tuberculoid granuloma following neurovascular bundle with plenty of Langhans giant cell. The granulomas were disorganized due to intense dermal oedema (Fig. 2). Bacteriological index of granuloma was 1+. Based on clinical morphology showing ulcerated plaque and histopathology finding of intense oedema in the dermis along with granuloma following neurovascular bundle, a diagnosis of Lazarine leprosy was made. Patient was started on multidrug therapy for multi-

Table 1 : Distinguishing features of Lucio phenomenon and Lazarine Leprosy (Adapted from Salafia 1995)

	Lucio phenomenon	Lazarine leprosy
Onset	After 3-4 years	First few months
Patients general condition	Immuno-depressed	Usually healthy
BI at four sites	Highly positive	Negative/weakly positive
BI in the bullae	Positive	Positive
Spectrum of disease	LL	TT or BT
Lepromin reaction	Negative	Positive
Histopathology	Superficial leucocytoclastic vasculitis and necrosis	Denuded epidermis, intense dermal oedema and disorganized granuloma

bacillary leprosy as per world health organization guideline which consists of Rifampicin 600mg once monthly (supervised), Clofazimine 300 mg once monthly (supervised) and 50 mg daily and Dapsone 100mg daily (MDT MB) for 12 months duration along with prednisolone at a dose of 1 mg per kg body weight. After 1 month of oral prednisolone there was complete healing of ulceration with atrophic scarring (Fig. 1b). Steroid was tapered with continuation of MDT MB adult and she was advised for regular follow up.

Discussion

Though leprosy has been eliminated worldwide, it still remains as a major public health problem in many countries like India. Reactions in leprosy contribute to most of the deformities and disabilities in such patients. They can occur any time during treatment or after completion of therapy, even can be the first manifestation of the disease. Two major types of reactions exist depending on the type of immune response of an individual against *Mycobacterium leprae*. Type 1 reactions are delayed type hypersensitivity reactions occurring in borderline forms of leprosy i.e. borderline tuberculoid (BT), borderline borderline (BB), borderline lepromatous (BL) and can also be rarely seen in lepromatous leprosy (LL). Immune complex mediated type 2 reactions are commonly seen in lepromatous pole of leprosy (Kar & Sharma 2010). Risk factors implicated for type 1 reactions are older age, female gender, large facial patch, poor nutrition, various type of stress, hormonal imbalance, high bacillary load, high antibody titres to *Mycobacterium leprae* antigens, genetic defect in cell mediated immune response or cytokine production (Kar & Sharma 2010, Degang et al 2014). Lazarine leprosy is a rare form of leprosy characterized by extensive ulceration of the lesion. Though exact pathogenesis is not known, break down of local immunity, intense tissue oedema, increased proliferation of bacilli are considered as possible explanations for such type of reactions (Nanda et al 2004). According to some authors hypoproteinemia will lead to impaired cellular as well as humoral immunity leading to uninhibited proliferation of bacilli and intense tissue oedema secondary to decreased osmotic pressure that increases the probability of ulceration (Skinsnes & Higa 1976).

Lazarine leprosy was first described in 1852 by Raphel Lucio and Ignacio Alvadro. Previously this entity was thought to occur in lepromatous pole

of leprosy. Cochrane described Lazarine leprosy as a chronic progressive form of erythema nodosum leprosum where the patient develops subcutaneous nodules that ulcerate with a distressing general condition (Cochrane 1964). Strobel et al (1979) reported majority of cases of Lazarine leprosy in lepromatous pole with underlying malnutrition or other debilitating disorders. According to Ramu and Dharmendra (1978) lazarine leprosy occurs near tuberculoid end of borderline spectrum of leprosy in debilitated patients. Salafia (1995) reported a case of tuberculoid Lazarine leprosy. Later in 2004, Nanda et al reported two case of Lazarine leprosy both in Borderline Tuberculoid spectrum without any evidence of malnutrition or immune suppression. Bhat et al (2013) reported Lazarine leprosy in BT Hansen's as a manifestation of immune reconstitution inflammatory syndrome (IRIS) in an HIV patient. Sunandini et al (2015) reported two cases of Lazarine leprosy one in BT, and another in BL spectrum without any underlying debilitated condition. Though there are confusing reports of Lucio phenomenon as Lazarine leprosy the distinguishing features between these two entities given in Table 1 adapted from Salafia (1995).

This case was elderly female with a large facial patch with ulceration without any other risk factors like malnutrition or immune suppression. Lazarine leprosy is considered as an exaggerated type 1 reaction with extensive lesional ulceration. Usually lesional oedema with scaling on surface along with few satellite papules are considered as hall mark of type 1 reaction, ulceration is hardly encountered. The extent and severity of ulceration helped us to diagnose Lazarine leprosy. A strong index of suspicion prompted us to confirm the same by histopathology showing characteristic features of denuded epidermis, intense dermal oedema, disorganized granuloma and absence of vasculitis (Table 1). Accordingly the treatment was started promptly and patient responded well.

In the post elimination era of leprosy new cases with atypical presentation are not uncommon. Physicians should remain aware about it so that early treatment may be instituted to prevent permanent deformities.

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How to cite this article : Tripathy T, Panda M, Kar BR et al (2018). Facial Lazarine Leprosy in Post-elimination Era: A Case Report. *Indian J Lepr.* **90** : 313-318.