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Case Report

De-novo Ulceration - Rare Lazarine Leprosy - like Presentation of Borderline Lepromatous Hansen's Disease

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Leprosy is caused by *Mycobacterium leprae*, which has a distinct predilection for cooler parts of the body. There are many forms of leprosy that range from the mildest indeterminate form to the most severe lepromatous type. The type of disease an individual develops depends on the host immune status; with tuberculoid type being seen in those with good immunity and lepromatous form in individuals with poor immunity to leprosy bacillus. Deep ulcers may occur in association with erythema nodosum leprosum, Lazarine leprosy and Lucio phenomenon. We report a male who presented with multiple acute onset foul smelling stellate ulcers over bilateral extremities. He was diagnosed as Borderline lepromatous leprosy (not in reaction) on the basis of histopathology. This type of presentation has not been reported till date.

Keywords : Lazarine leprosy, Lucio phenomenon, Borderline lepromatous leprosy

Introduction

India achieved elimination of leprosy in December 2005 with the initiatives of the National Leprosy Eradication Program (NLEP) and active participation of governmental as well as non-governmental sector, however, according to the latest statistics of the World Health Organization (WHO), South-East Asia including India, remains the highest contributor to the global burden of leprosy (Tayshetye et al 2013, Kumar 2015). The different manifestations of leprosy such as numb hypopigmented or erythematous patches, localized paraesthesia, shooting pains, blisters of hands and feet, motor weakness, nasal stuffiness, epistaxis, synovial swelling of wrist, pedal edema, painless nodules, diminished/excessive sweating have been reported since time immemorial. But even as we strive to eradicate leprosy, newer forms of presentation continue to emerge – laryngeal dyspnoea, bullae with hematoma, xanthoma like presentation etc. (Das et al 2007)

Borderline lepromatous (BL) leprosy presents as slightly infiltrated plaques with coppery hue and not-so-symmetrical distribution with areas of

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apparently normal skin in between. With progression, papules or nodules may develop, with sloping margins. Signs of nerve damage may appear sooner in BL than LL Hansen's. Type 2 reactions are known to occur in BL Hansen's, occurring as tender crops of nodules associated with systemic symptoms and occasionally arthritis, ocular and renal involvement. However, BL leprosy presenting as de-novo ulcers is hitherto unreported.

Case report

A 46 years old male, welder by occupation presented with multiple foul-smelling, asymptomatic ulcers over extremities since 15 days. Lesions appeared as erythema followed by central blackish discolouration and ulceration progressing rapidly up to elbows and knees. Fever, chills and constitutional symptoms were absent. He denied history of any preceding hypopigmented, hypoaesthetic patches or erythematous plaques, joint pain, tingling, numbness, nasal stuffiness, epistaxis, or swelling of hands and feet.

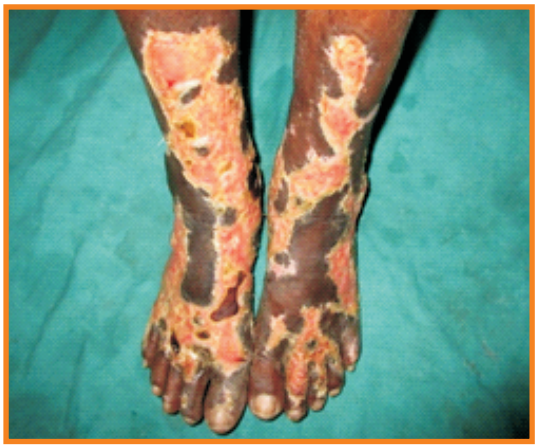

Fig. 1 : Stellate ulcers with irregular border and sloughing at base with central eschar over bilateral dorsum of foot.

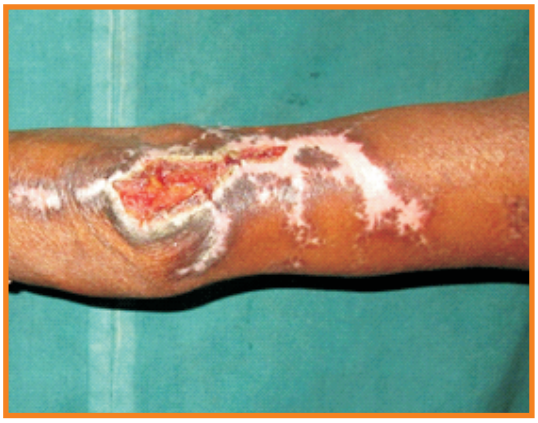

Fig. 2 : Stellate ulcers with irregular border and sloughing at base with central eschar over bilateral elbows.

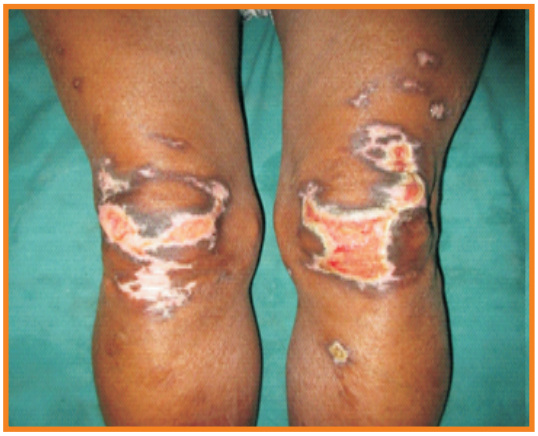

Fig. 3 : Stellate ulcers with irregular border and sloughing at base with central eschar over bilateral knees.

Clinical examination revealed multiple polysized stellate non-tender ulcers with irregular border, sloughing at base with central eschar over bilateral elbows, dorsae of hands, knees and ankles extending up to sole (Figs 1, 2, 3). Pinnae showed small ulcers along periphery. Bilateral ciliary and supra-ciliary madarosis was noted. Peripheral nerve examination could not be

performed because of overlying ulcers. Mucosal and sensory and motor system examination was unremarkable.

Differential diagnosis of necrotising vasculitis, Rickettsial fever and ulcerated erythema nodosum leprosum (ENL) were considered. Routine investigations were within normal limits except haemoglobin of 8 gm%. Indirect Coombs test was negative. Nerve conduction velocity study was not feasible due to extensive ulceration. Histopathology from periphery of ulcer showed perivascular and periadnexal collections of foamy macrophages admixed with lymphocytes in dermis and subcutaneous tissue. Scattered ill formed granulomas with rim of lymphocyte cuffing was seen without evidence of necrosis, neutrophilic infiltrate or vasculitis (Fig. 4). Ziehl-Neelsen stain was positive for acid fast bacilli clinching diagnosis of borderline lepromatous leprosy. Slit-skin smear (Fig. 5) revealed solid-staining acid fast bacilli with bacteriological index of 0.6. There was no evidence of foamy macrophages or granuloma on bone marrow biopsy.

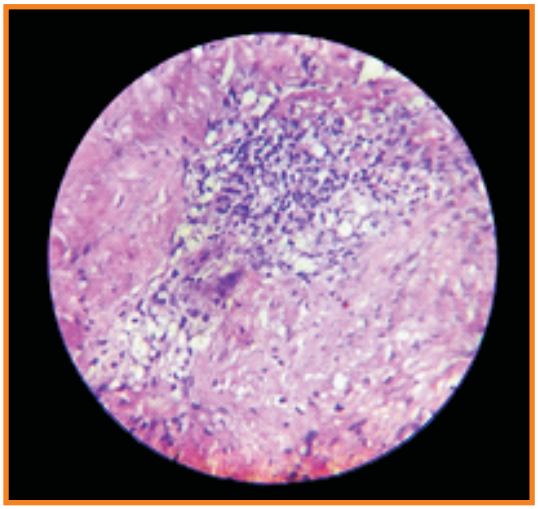

Fig. 4 : Perivascular and periadnexal collections of foamy macrophages admixed with lymphocytes in dermis and subcutaneous tissue with scattered ill formed granulomas with rim of rich population of lymphocyte cuffing. H & E (40 X)

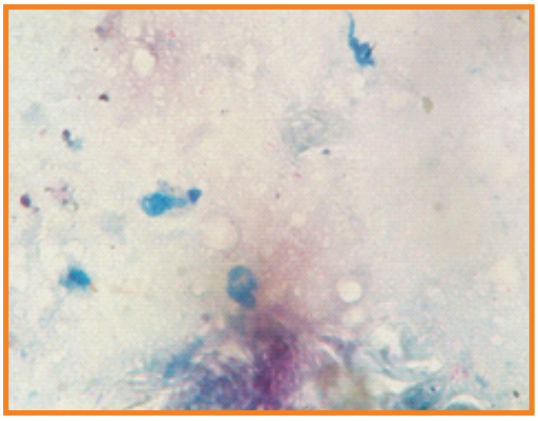

Fig. 5 : Acid fast bacilli on slit skin smear.

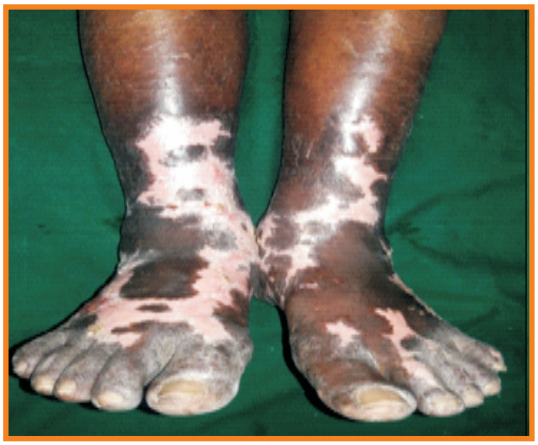

Fig. 6 : Ulcers healed with hypopigmentation

Patient was started on WHO multidrug therapy for MB Hansen's (Cap. Rifampicin 600 mg once a month, Cap. Clofazimine 300 mg once a month and 50 mg daily). Dapsone was omitted on account of anaemia. Topical Fusidic acid cream and injection Ceftriaxone 1 gm intravenous infusion for 10 days followed by oral amoxicillin /

clavulanate was administered. To hasten re-epithelisation of ulcers, autologous platelet rich plasma was injected weekly around the edges. A significant reduction in ulcer dimensions was observed after four to five treatment sessions. (Fig. 6)

Discussion

Leprosy is a chronic granulomatous disease caused by *Mycobacterium leprae* with a wide spectrum of clinical, histopathological and immunological characteristics. Although, it can be diagnosed fairly accurately based on three cardinal signs, leprosy is a great imitator and can have varied clinical presentations. This propensity for unusual presentations can confuse and delay diagnosis, as in our case.

Classically in BL leprosy, lesions range in number from solitary to numerous and widespread. Generally, the annular and plaque lesions are asymmetrically distributed, but the lepromatous like nodules, if numerous, are symmetric. Skin lesions are often (not necessarily) hypoaesthetic or anesthetic. Nerve palsies are most prevalent in BL disease. Lesions range in number from solitary to numerous and widespread (Lee et al 2012). Despite extensive search through Indian and world literature, we were unable to find any report of BL leprosy presenting as spontaneous ulcerations similar to our case.

Ulcerating Type 1 reaction is called Lazarine leprosy, first described in 1852 by Rafael Lucio and Ignacio Alvarado. Later, Pardo Castello described a series of 23 cases, proposing that this type of reaction may occur in borderline tuberculoid or borderline lepromatous disease as a result of high inflammation or high bacillary load respectively (Sundandini et al 2015). Breakdown of local immunity, severe tissue reaction and protein energy malnutrition have been implicated in pathogenesis. Lazarine leprosy presents with spontaneous ulceration even without pre-existing skin lesions. However, our patient lacked histopathological evidence of reaction and necrosis which are the pathognomonic features of this rare form (Kumar and Dogra 2016). Therefore the present case was classified as Borderline Lepromatous leprosy and not as Lazarine leprosy.

ENL is an acute inflammatory reaction in lepromatous leprosy or occasionally in borderline lepromatous leprosy. Vesiculo-bullous, pustular, ulcerated (erythema nodosum necroticans), lesions have been reported in ENL (Panda et al 2012, Kar and Chauhan 2016). Histopathology of classical ENL lesions shows increased vascularity with intense infiltration of neutrophils invading the vessel walls. In case of erythema nodosum necroticans there is obliterative angitis and endarteritis (Kar and Chauhan 2016) which was not seen in our case.

Absence of typical features of acute neutrophilic vasculitis such as fibrinoid necrosis, extravasation of erythrocytes and neutrophils with release of nuclear debris (leukocytoclasia) ruled out necrotising vasculitis in the present patient (Merkel and Monach 2012).

Conclusion

Early diagnosis and prompt therapy of leprosy hold the key to achieving the elusive goal of eradication. The absence of clinical cardinal features can delay diagnosis and treatment initiation with potentially disastrous deformities and disabilities. In such clinically confounding situations, slit skin smear and histopathology come to the rescue by providing vital clues aiding accurate and timely diagnosis.

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