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Case Report

Exacerbation Reaction in a Case of Lepromatous Leprosy: An Intrigue !

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A 24 years old male presented with multiple, shiny, infiltrated papules, plaques with central crusting and ulceration without any systemic features. Histopathological examination of the skin nodule showed perivascular and perineural collection of foamy histiocytes, few neutrophils and lymphocytes. Superficially below the epidermis there was localised area of necrosis with macrophages and infiltration of neutrophils. Fite stain was positive for AFB thus proving a diagnosis of lepromatous leprosy with exacerbation reaction. In rare instances in lepromatous leprosy exacerbation reaction occurs locally in hyperactive granulomas which may mimic erythema nodosum leprosum clinically.

Key words : Lepromatous leprosy, Exacerbation reaction, Erythema nodosum leprosum.

Introduction

Lepromatous leprosy (LL) presents with multiple hypopigmented macules, localised or diffuse infiltration, papules, nodules or plaques, symmetrically distributed over the face, trunk and extremities (Pfaltzgraff and Brycesson 1985). Nodular leprosy is a result of progressive deterioration of the macular, diffuse or infiltrated forms of leprosy. Exacerbation reactions (ER) are acute reactions occurring locally in histoid or other highly active lepromatous lesions with an exceptionally heavy bacterial load. Clinically, they are almost silent although they may cause ulceration and the release of viable bacilli (Ridley and Ridley 1984).

Here we are presenting a case of acute ER in a patient of LL without any systemic affection.

Clinical

A 24 years old male presented in dermatology outpatient department with asymptomatic, skin colored, shiny elevated pea sized lesions of gradual onset started from the chest area as

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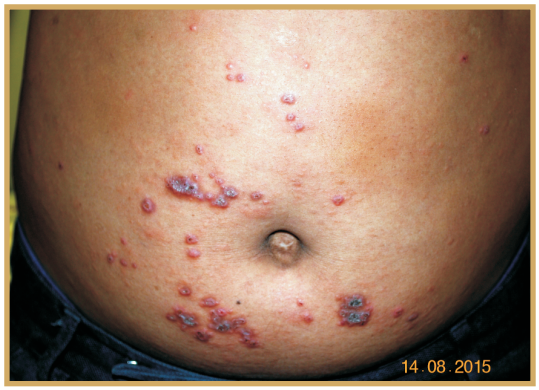

Fig 1 : Multiple, bilateral, waxy, skin coloured, non- tender, shiny, infiltrated papules and plaques with central ulceration and crusting over few lesions on abdomen.

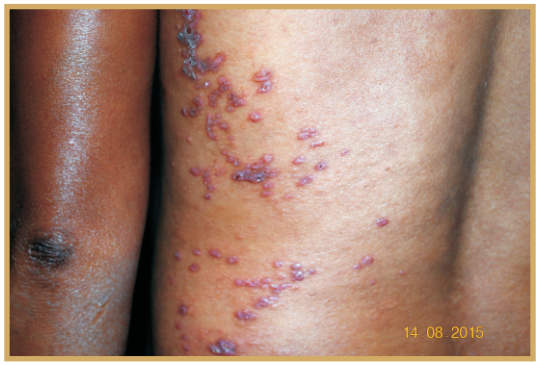

Fig 2 : Multiple, waxy, skin coloured, non-tender, shiny, infiltrated papules and plaques with central ulceration and crusting over few lesions on the trunk.

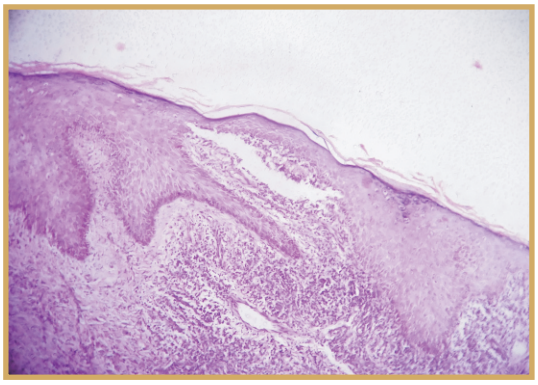

Fig 3 : Unremarkable epidermis, dermis showing collection of foamy histiocytes, few neutrophils and lymphocytes. Superficially below the epidermis localized area of necrosis with macrophages and infiltration of neutrophils. (H & E stain, 10X)

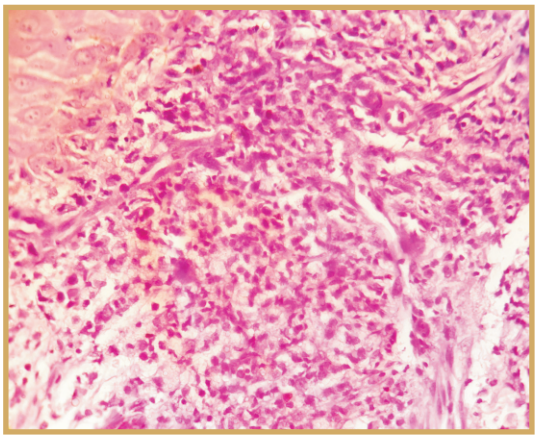

Fig 4 : Dermis showing perivascular collection of foamy histiocytes, few neutrophils and lymphocytes. (H and E stain, 40X)

reddish spots and then developed on the back, upper arms and buttocks in two months duration. There was central necrosis with ulceration in the centre of few of them. He had no history of fever, malaise, bodyache, joint pains, appearance of any evanescent painful nodules or edema of extremities. He also had not taken any medication for the same nor gave history suggestive of chronic disease like tuberculosis or leprosy or episodes of reactions in the past. Family history was not contributory.

Skin examination revealed multiple, bilateral, symmetrical waxy, skin coloured, non-tender, shiny, infiltrated papules and plaques with central ulceration and crusting over few lesions on the

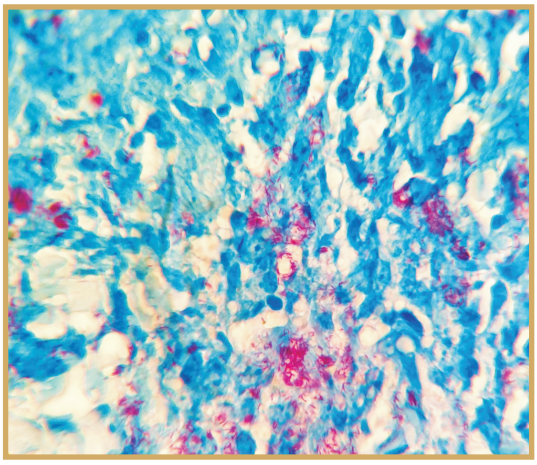

Fig 5 : Fite-Faraco stained section showing lepra bacilli in 100X oil emulsion lens.

shoulders, trunk, back and buttocks (Figs 1 and 2). In addition, he had nontender, bilaterally thickened great auricular nerves, ulnar nerves and common peroneal nerves with glove and stocking anesthesia. Supraciliary madarosis was seen along with bilateral earlobe infiltration. Eye examination and genital examination was within normal limits. No lymphadenopathy. We reached the provisional differential diagnosis of Leprosy with type 2 reaction, papulonecrotic tuberculids, Secondary syphilis, Leishmaniasis. Laboratory investigations of hemogram, blood sugar, liver and renal function tests were within normal limits. VDRL was negative with titres < 1:8. HIV ELISA was negative. Slit-skin smears from four different sites including from two lesions over the forehead and back revealed bacterial index(BI) of 4+ with Ridley's logarithmic index (Average of all four sites) and Morphological index (MI) of 20% on AFB staining. Histopathological examination of the skin nodule over the back showed unremarkable epidermis, dermis showed perivascular and perineural collection of foamy histiocytes, few neutrophils and lymphocytes. Superficially below the epidermis there was localized area of necrosis with macrophages and infiltration of neutrophils (Figs 3, 4). Vasculitis was not seen. Fite stain was positive for AFB (Figs 5). Patient was started on WHO recommended multidrug therapy for multibacillary leprosy (MDT MB). On follow up after one month, the ulcerations and crusting over the papules and nodules had subsided. At the same time papules and nodules had also started to subside. Patient is on tenth pack of MDT MB and there are no new lesions and existing lesions have subsided with hyperpigmentation.

Discussion

Lepromatous exacerbation is described in older literature as a type of reaction. It is seen in advanced lepromatous patients with nodular and plaque lesions. It is characterised by red, painful, swollen and tender LL lesions with or without constitutional symptoms. The lesions have a tendency to develop necrosis and ulcerate at times. These reactions can be severe histologically but may not be accompanied by any systemic disturbance, and their nature is not so far understood. Histologically there are localised areas of necrosis in middle of macrophages along with localised infiltration of neutrophils as seen in our case. Vasculitis is rarely seen. Macrophages contain a relatively large amount of leprosy bacilli (AFB). This may be due to sudden burst of bacterial multiplication which leads to localised cell necrosis and acute inflammation (Kar and Chauhan 2016).

Sometimes these ER may mimic ENL. The difference between ER and ENL is that ER occurs in hyperactive lepromas whereas ENL in regressing

Table 1 : Exacerbation reaction vs ENL vs Reversal reaction

	Exacerbation Reaction	Type 2 Reaction (ENL)	Type 1 reaction (Reversal Reaction)
Type of immunological reaction	Sudden burst of bacillary multiplication in hyperactive lepromas with acute inflammation	Antigen antibody immune complex reaction	Delayed type hypersensitivity reaction
Types of patients affected	Lepromatous leprosy patients with nodular or plaque type lesions and histoid leprosy patients	LL type rarely BL	Borderline types (BT, BB and BL)
Constitutional signs and symptoms	None or rare	Common	None or rare
Type of skin lesions	Red, painful, tender LL lesions with tendency to develop necrosis and ulceration	Fresh red, painful, tender cutaneous nodules and plaques. Existing skin lesions remain unchanged.	Existing skin lesions (few or many or all) suddenly become reddish, warm, painful and tender
Histopathology	Macrophages containing large number of lepra bacilli (AFBs). Localised areas of necrosis in middle of macrophages.	Foci of acute inflammation superimposed on chronic inflammatory infiltrate. Neutrophilic infiltrate. Foamy macrophages with fragmented bacilli. Necrotizing vasculitis may be seen.	Edema within and about the granulomas and proliferation of fibrocytes in dermis. In upgrading reactions granuloma becomes more epitheloid and activated and Langhans giant cells are larger.
Treatment	Start and Continue MDT.	Continue MDT and Depending on severity of ENL, NSAIDS and general measures in mild cases. Thalidomide and/or systemic corticosteroids in severe cases	Continue MDT and NSAIDS and general measures in mild cases Systemic corticosteroids in severe cases.

granulomas. Each ER is a specific event that is usually localized to hyperactive lepromas. The reason for this is not clear. But this localization may be an explanation for lack of systemic complication. Hyperactive lepromas are few in number, they are characterised by high cell turnover with heavy bacterial load, a classic example being histoid leprosy where many exacerbations take place. Clinically they are silent inspite of ulceration and release of viable bacilli as in our case. Rarely ENL reactions have also been documented in histoid lesions (Sharma et al 2002). The difference among ER, ENL and reversal reaction are summarised in Table 1.

Histologically both ER and ENL commence with polymorph infiltration in areas of macrophage cell death. The macrophages in ER are large and cytoplasmically active, containing increased numbers of solid organisms, while those in ENL lesions are effete, with poorly detectable degraded bacterial debris (Ridley and Ridley 1984). Later In ER there may be increased capillary permeability, small vessel necrosis and mast cell degranulation. IgE and C1q are commonly involved.

In our case, the papules showed necrosis and ulceration, without any other feature of type 2 leprosy reaction. Skin biopsy from these lesions confirmed exacerbation reaction. Through this case report we attempt to focus light on exacerbation reaction which has of late been forgotten and hence few reports are reported in literature. Though many studies are available on reactions, these intriguing exacerbations still remain poorly understood.

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