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Case Report

Histoid Leprosy Presenting with Borderline Tuberculoid Leprosy in Type 1 reaction: An Uncommon Shift of Spectrum

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Histoid leprosy is an uncommon variant of leprosy with characteristic clinical, immunological and bacteriological features and is considered to be a polar variant of lepromatous leprosy. Coexistence of other immunologically diverse forms of leprosy in histoid end of spectrum is very rare. We report a rare case of histoid leprosy on multi-drug therapy for last 7 months shifting to borderline tuberculoid spectrum in type 1 reaction or Wade's contamination, spectral shift.

Keywords : Histoid Leprosy, Borderline Tuberculoid, Type 1 Reaction

Introduction

Leprosy is a chronic disease caused by *Mycobacterium leprae* affecting the peripheral nervous system, the skin, and other tissues. There are a wide spectrum of manifestations varying from clinically stable tuberculoid and lepromatous forms to clinically unstable borderline spectrum, mid-borderline being the least stable. Apart from these, there are rare variants like Indeterminate, Pure Neuritic, Histoid, Lucio and Lazarine leprosy. Earlier histoid leprosy (HL) was reported as a result of inadequate treatment in Dapsone monotherapy era, but now there are increasing data of de novo HL. The term "histoid" is derived by the histological finding of the dermal infiltrate composed by a predominance of spindle shaped "histiocytes" (Maroja et al 2016). It is usually considered a stable multibacillary form, and shifting to another spectrum of disease or exhibiting lepra reaction has rarely been reported.

It is known that leprosy is a complex spectral disease and there is no cut-and-dried clinical presentation. At a given time, there can be leprosy lesions of varied morphology fitting in the description of any or all stages of the disease even with clinico-pathological discordance depending on the spectrum of the lesion biopsied. However, BT and Histoid lesions together are rare. We report a case of de novo HL developing lesions of borderline tuberculoid (BT) leprosy in type 1 lepra reaction after 7 months of multi-drug therapy (MDT).

Case Report

A 32-year-old married female presented to us at Dr RP Govt Medical College, Kangra at Tanda

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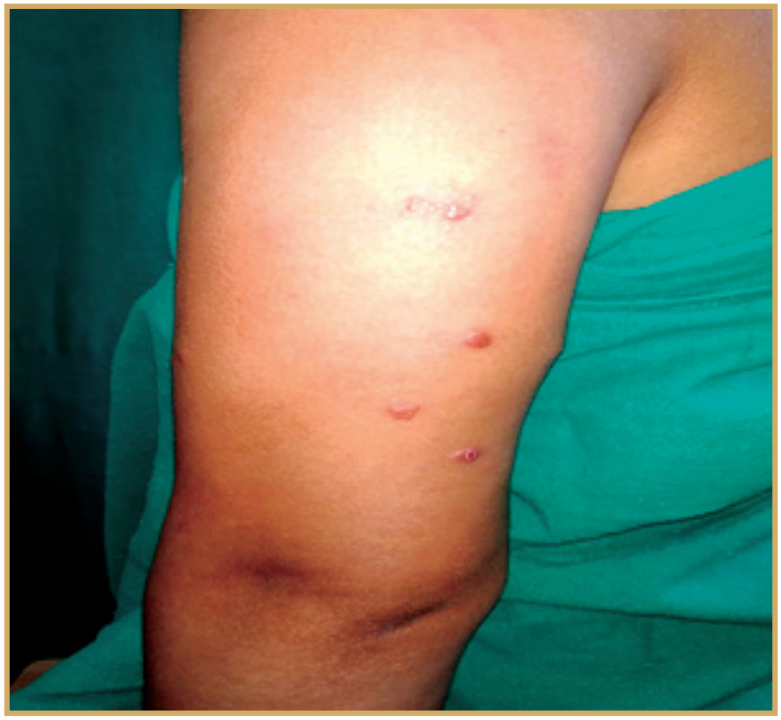

Fig. 1a : Skin coloured dome shaped papules on posterior aspect of left arm with normal surrounding skin.

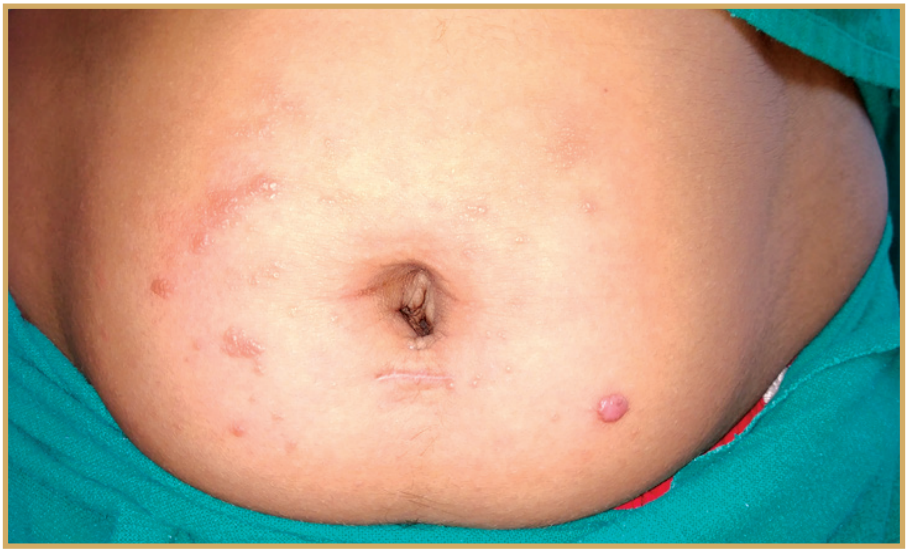

Fig. 1b : Skin coloured tiny papules and erythematous dome shaped plaque in periumbilical region with normal surrounding skin.

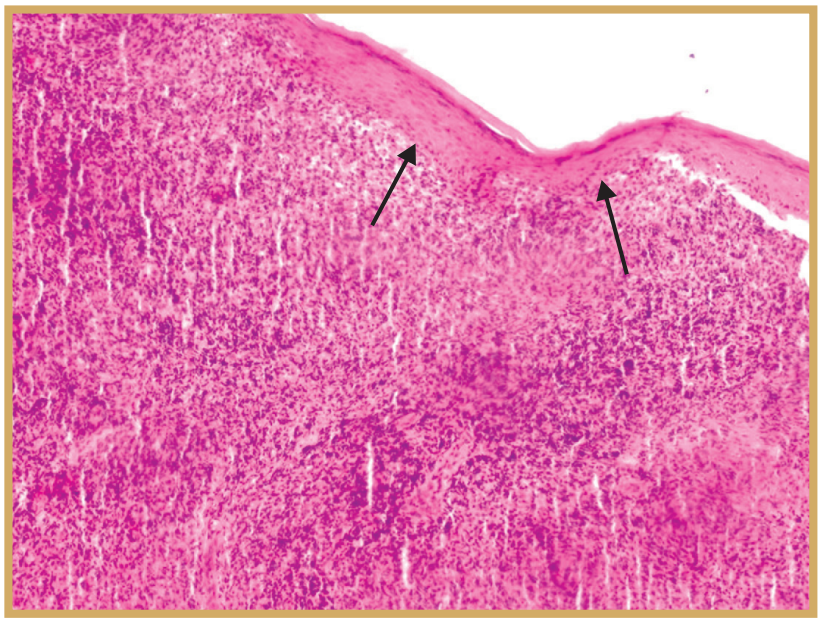

Fig. 2a : Loss of rete ridges and epidermis with spindle cells in dermis (Hematoxylin-Eosin, x100).

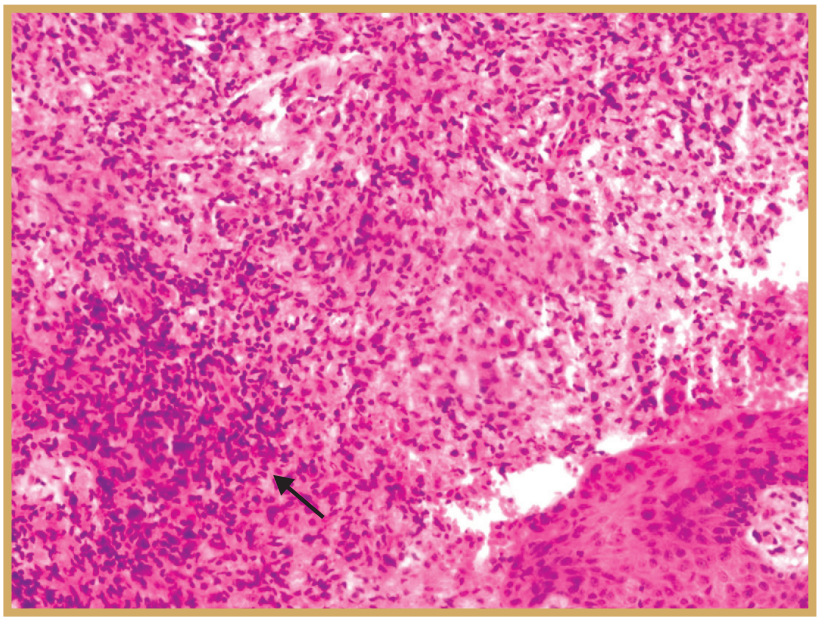

Fig. 2b : Sheet of spindle cells in entire dermis, completely replacing the adnexal structures (Hematoxylin-Eosin, x400).

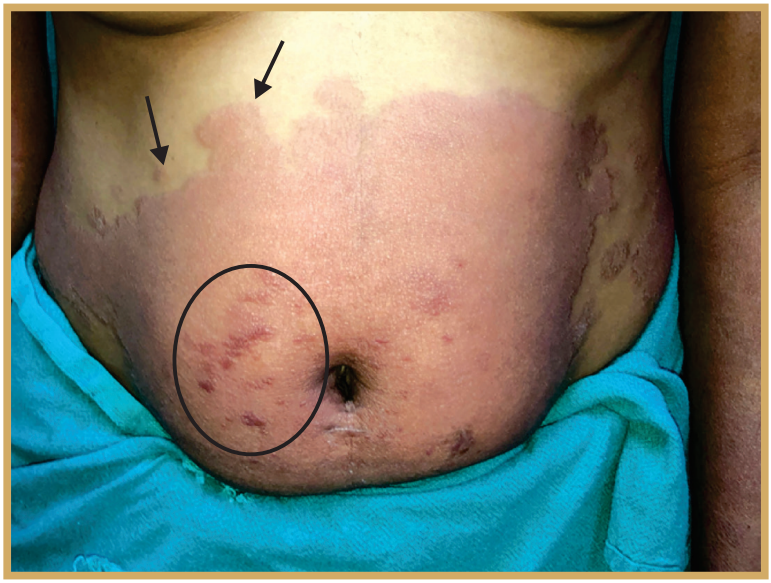

Fig. 3 : Erythematous, oedematous plaque with pseudopodia and satellite lesions (arrows) over abdomen with previous histoid lesions (circle) in periumbilical region.

with asymptomatic skin coloured to reddish raised lesions over face (chin), ears, left arm and abdomen from 7 months. There was no history of spontaneous blistering, slipping of chappals, difficulty/weakness in hands while carrying out routine activities, pedal edema, no history of fever, epistaxis, any ulcer or deformity of hands and feet. There was no history of contact with leprosy patients and no history of any drug intake for leprosy. Cutaneous examination revealed multiple shiny, firm, non-tender, skin coloured to erythematous papules and nodules present on chin, bilateral ears, posterior aspect of left arm (Fig. 1a) and abdomen (Fig. 1b) over a background of normal skin. Bilateral ulnar, radial cutaneous, common peroneal and post tibial nerves were thickened. Sensory and motor examination was normal. All other routine investigations were normal. Slit skin smear showed a full field of acid fast bacilli with a Bacteriological Index of 6+ and Morphological Index of 70%. A lesional biopsy revealed loss of rete ridges and epidermis being stretched over a sheet of spindle cells in dermis, completely replacing the adnexal structures (Fig. 2a). Presence of characteristic spindle cells (Fig. 2b) led to the diagnosis of histoid leprosy, Fite Faracco stain was positive and bacillary index was 6+. Clinical and histopathological findings were consistent with HL and patient was started on MDT-MB (multibacillary). After 7 months of treatment, patient presented to us again with new onset reddish raised lesions all over the body of 10 days duration. The previous nodular lesions were still persistent and there was no history of fever or pain. History of pedal oedema was present, not subsiding on rest. On examination, there were well defined, non-tender, erythematous, oedematous plaques with pseudopodia and satellite lesions present over anterior aspect of neck, lower abdomen

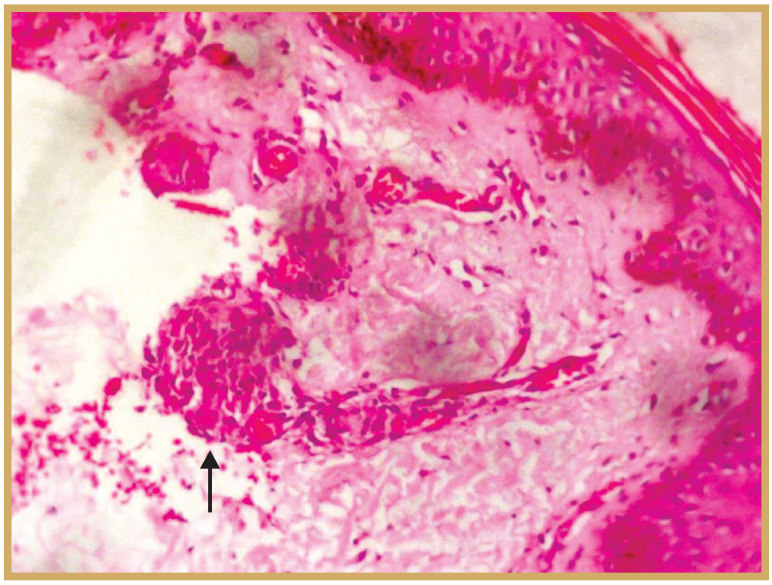

Fig. 4 : Perivascular ill formed epithelioid granulomas along with perineural lympho-histiocytic infiltrate (Hematoxylin-Eosin, x400).

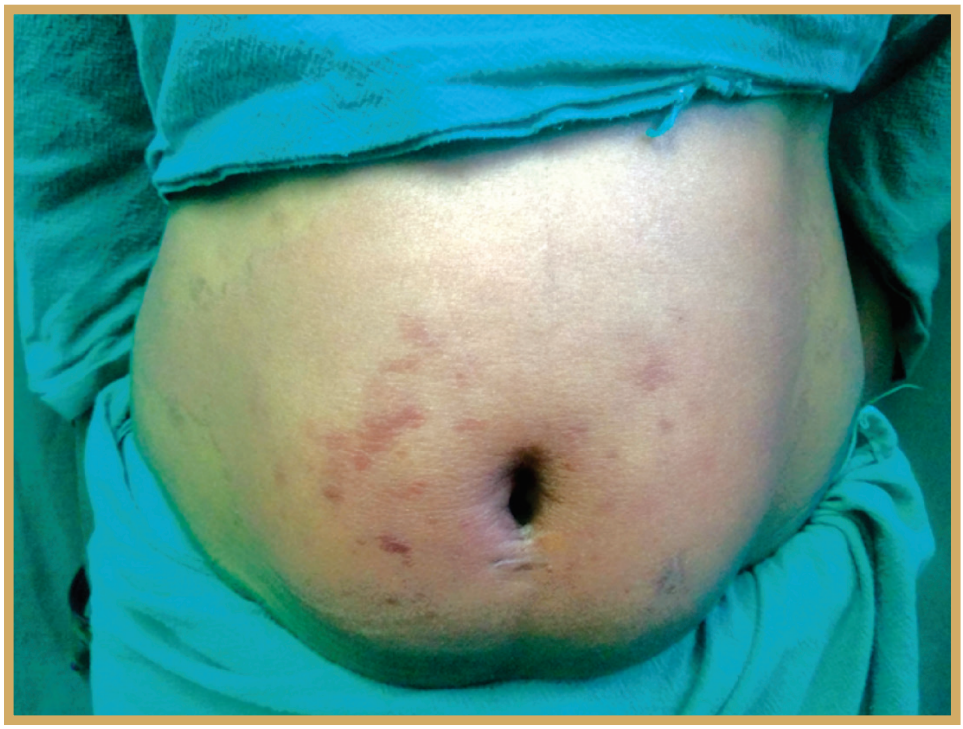

Fig. 5 : Resolution of lesional erythema and oedema after 2 weeks of steroids along with MB-MDT.

(Fig. 3), back and bilateral upper limbs. There were ill defined erythematous plaques over the background of xerosis present over bilateral buttocks and lower limbs. There was no neuritis or any new onset sensory motor loss. Slit skin smear from the new lesions was negative. Bacteriological Index was still 6+ from the HL lesions. Biopsy from the new lesions revealed peri-appendageal and perivascular ill formed epithelioid granulomas along with perineural lympho-histiocytic infiltrate (Fig. 4). Fite Faracco was positive and Bacteriological index was 3+. Based on the clinical and histopathological findings, diagnosis of BT leprosy was made. Although fever and neuritis were absent, lesional erythema and oedema was prominent. MDT-MB was continued and patient was started on Prednisolone 40 mg for type 1 lepra reaction following which the erythema and oedema subsided in 2 weeks (Fig. 5).

Discussion

Histoid leprosy (HL) is an uncommon variant of leprosy first described by Wade in 1960 (Wade 1960). There is no estimate of global prevalence but in India the incidence is estimated to be 2·79–3·60% among patients with leprosy (Gupta 2015). There is male preponderance and the average age affected is between 21 and 40 years (Kalla et al 2000). HL exhibits clinically multiple symmetric, discrete, firm shiny, painless succulent, globular, protuberant, skin coloured dome-shaped or oval papules with a smooth bright surface. There can be cutaneous, or subcutaneous nodules and plaques over apparently normal skin. The nodules may involve face, back, limbs and bony prominences, as well as mucous membranes in severely affected patients (Rodriguez 1969). The histological finding includes epidermal atrophy and an acellular band (Unna band) located immediately below the epidermis and expanding dermal granulomas, composed by spindle-shaped histiocytes, in a whorled, storiform pattern. AFB are abundant and the bacilli have elongated morphology (Shaw et al 2000). HL was originally described as a manifestation of drug resistance after irregular or inadequate treatment with Dapsone monotherapy or multi-drug therapy. However, it has also been reported with relapse in the presence of supervised monthly multidrug therapy and de novo in patients without any treatment. It can also occur in patients with relapsing disease because of stoppage of treatment (Ramanujam & Ramu 1963). It is considered as a variant of lepromatous leprosy by some researchers while others consider it as a distinct entity (Price & Fitzhebert 1966). The exact cause of association of HL with BT leprosy remains unexplored. It is proposed that HL, usually considered a stable disease, may have spectral shift depending upon cell mediated immunity. However, we found no case report of spectral shift in literature other than BT leprosy (Das et al 2014, Rao 2016). In both the cases reported, lesions of HL developed few months after onset of BT lesions, either due to stoppage or not taking treatment, which can probably be explained by downgrading of spectrum after stopping treatment. Likewise, in our case the lesions of BT leprosy developed due to upgrading after continuous MDT-MB treatment for 7 months. One plausible explanation could be the peculiar 'tuberculoid contamination' phenomenon observed by Wade in some of his specimens of HL. He reported 'tuberculoid foci' in histoid nodules with epithelioid cells in centre, almost free from bacilli, in contrast with the normal abundance of AFB in the surrounding tissue (Wade 1960). Histoid transformation from unstable forms is also known (Ramanujam & Ramu 1963). This coexistence of 'tuberculoid foci' and histoid nodules could be the reason of shift in spectrum from HL lesions to BT lesions

after starting of treatment in our case or vice versa in absence of treatment in the two cases reported previously. Leprosy reactions in HL have been only rarely reported and are mainly Type 2 reactions (Kaur et al 2009, Mendiratta et al 2011, Nair & Kumar 2013). Type I lepra reaction, driven by delayed hypersensitivity to *M. leprae*, predominantly affect borderline leprosies, but is rarely reported in HL (Singh et al 2015, Sun et al 2017). Possible explanation could be that the cell-mediated immunity is good in HL as substantiated by increased CD36 expression by the keratinocytes, predominance of CD4 lymphocyte over CD8 lymphocytes, and increased number of activated lymphocytes and macrophages in the lesion (Sehgal et al 1985). Our patient might have upgraded to BT leprosy in type 1 reaction due to activation of cell mediated immunity in HL lesions following MDT-MB. Reactional erythema and edema subsided within 2 weeks of steroids. This case report of BT lesions in type 1 reaction with histoid lesions, raises the possibility of some unknown mechanisms which link HL both to coexistence with BT lesions as well as the tendency to go in type 1 reaction rather than it being considered a polar disease.

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