Indian J Lepr 2023, 95 : 139-146

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Case Series

Histoid Leprosy: Are We Still Harbouring Potential Community Menace? A Case Series from North-East India

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Received : 25.11.2022 Revised :17.03.2023 Accepted : 26.03.2023

Leprosy is a chronic granulomatous disease caused by *Mycobacterium leprae*. Histoid leprosy is considered an uncommon multibacillary form of leprosy characterized by presence of histoid lepromas which present as erythematous round or oval shaped glistening nodules. Most commonly this form of leprosy is seen in patients on irregular therapy with MDT. In this series we report a total of three cases with their clinical, histopathological and dermoscopic findings.

Keywords : Histoid Leprosy, Multibacillary, Spindle, Vacuolated, AFB, North-East India

Introduction

Histoid leprosy is a multibacillary variant of leprosy, which may occur at anytime before starting MDT or during the course of illness. However, *de novo* cases have also been reported (Sehgal & Srivastava 1985). The pathogenesis and etiology of histoid leprosy is not entirely clear but the numerous bacilli in lesions are probably due to focal immunity loss (Kaur et al 2009, Kalla et al 2000). In an effort to restrict the spread of infection there is a hyperactive and exaggerated multibacillary expression. Due to local ulceration and necrotic changes there is increased overall cell and humoral response supplemented by local immune response (Chan et al 2006). Clinically there are multiple painless, discrete, round to oval smooth and glistening papules, nodules which are subcutaneous or/ in skin. They are usually of amber yellow colour, varying in size from 1-3cm, although giant lesions have also been reported. The surrounding skin is apparently healthy and normal in appearance. Lesions are usually distributed over posterolateral aspect of thighs, buttocks, arms, back, dorsum of hands and bony prominences like back of elbow and knees. Sometimes, genital and oral mucosa may also be involved but palms and soles are generally spared (Sehgal & Srivastava 1985, Annigeri et al 2007). Histoid leprosy is commonly seen in the age group of 21-40 years. As per the review of literature male to female ratio has been reported as 2:1 in most parts of the world (Noordeen 1985, Manoharan et al 2008).

We report a case series of three patients who presented with cutaneous lesions in the OPD of dermatology department of our hospital and were diagnosed as cases of histoid leprosy later during the course of evaluation. The study

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Table 1 : Profiles of patients of histoid leprosy included in the study.

S No	Age, sex and area of stay of the patient	Type of lesions	SSS with BI and MI	Nerve involvement	Treatment	Reaction: type 1,2
1	26 years Male, resident of Assam	Nodules	3+,50%	Ulnar, radial cutaneous, common peroneal, posterior tibial nerve	Single dose ROM +MB-MDT	No
2	36 years, Male , resident of Bihar	Nodules, ulcer on sole and clawing of digits of hands and legs	4+,50%	Ulnar, common peroneal, posterior tibial nerve	Single dose ROM +MB-MDT	No
3	42 year, Male, resident of West Bengal	Papules and nodules	6+,60%	Ulnar, greater auricular, common peroneal, posterior tibial nerve	Single dose ROM +MB-MDT	No

includes cases reported during the year 2022 in the hospital.

Synopsis of Cases studied:

Profile of cases included in this series is summarized in Table 1.

Case 1

A 26 years old male patient, local resident of Assam, district Tejpur and farmer by profession presented with multiple large sized skin lesions (nodules) over elbow and legs (Figs. 1, 2) of 6 months duration. Some of the nodules had developed ulceration and pain from past 10-12 days. On examination all the sensations to superficial touch, temperature pain and pressure were normal. Nerve examination revealed thickened and non tender ulnar, radial cutaneous common peroneal and posterior tibial nerve. No evidence of any systemic involvement in form of changes in eyes, lymph nodes, deformities or

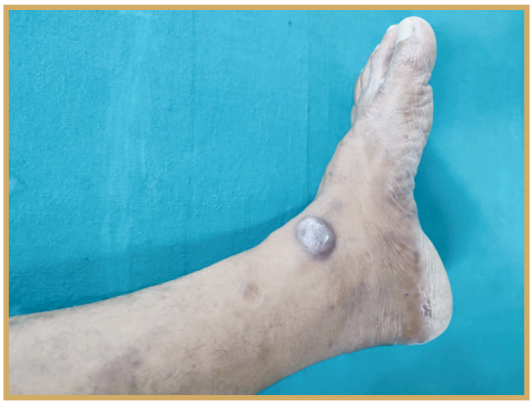

Fig. 1 : Nodules of Histoid Leprosy on the foot.

disabilities seen. The nodules were distributed over right lateral malleolus and right knee and measured 2x2cm and 3x3cm respectively, with superficial erosions (Fig. 1). No history of any close contact or similar lesions in family members could be elicited. There was no prolonged treatment history in the past in the patient as well as his close contacts in family. The patient was evaluated after detailed history with slit skin smear and biopsy for histopathology from the nodule. Differential diagnosis of nodular lesions like cutaneous leishmaniasis, eruptive xanthoma, kaposi sarcoma, neurofibroma, leiomyomas, sarcoidosis, histoid leprosy and atypical mycobacterial infection were considered.

Skin smears were taken using the standard slit and smear method (SSS) from these lesions showed abundance of organisms occurring singly as well as in clusters. and globi. Many bacilli were solid staining but fragmented forms were also seen. His bacillary index was 3+, morphological index was 50%. Numerous bacilli were also seen on Fite Faraco and ZN (Ziehl Neelsen) staining of biopsy specimen (Fig. 3). Histopathology revealed atrophied stratum epithelium with loose band of connective tissue in upper dermis. Fibro-histiocytic cells were arranged in sheets and storiform pattern. These cells had spindle to oval shaped nuclei and histiocytes cells with round nuclei and vacuolated cytoplasm was seen in dermis. Few foamy macrophage granulomas were also seen (Fig. 4). The features are distinct and characteristically significant of histoid form of leprosy. This was in contrast to presence of well-formed granulomas with differentiated epithelioid, giant cells and macrophages in tuberculoid form of leprosy with very few or nil acid fast bacilli on Fite Faraco and ZN staining. The patient was diagnosed as a case of histoid leprosy.

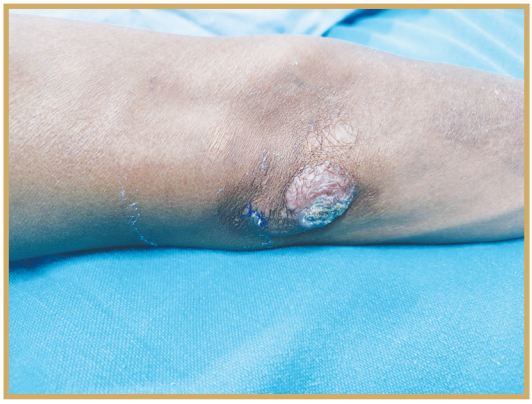

Fig. 2 : Showing histoid lesions on arms.

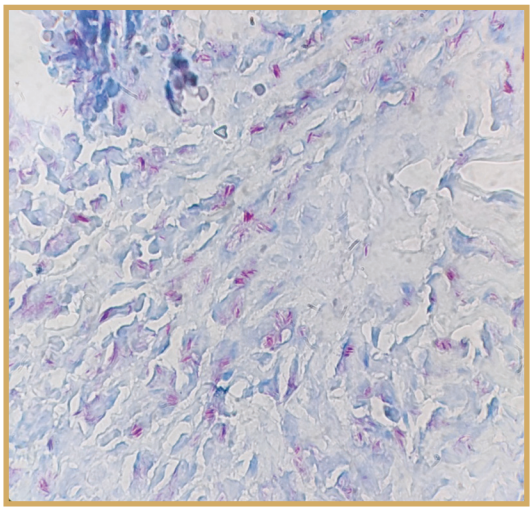

Fig. 3 : Histopathology of the skin lesion biopsy [Oil emersion, 100 X], using ZN stain - showing histiocytes with multiple acid fast bacilli.

Case 2

A 36 years old male married patient, clerk by profession and resident of Bihar reported with chief complaints of repeated bleeding from nose of 6 months duration and associated multiple skin coloured raised lesions over body predominantly over face and lower limbs. On examination

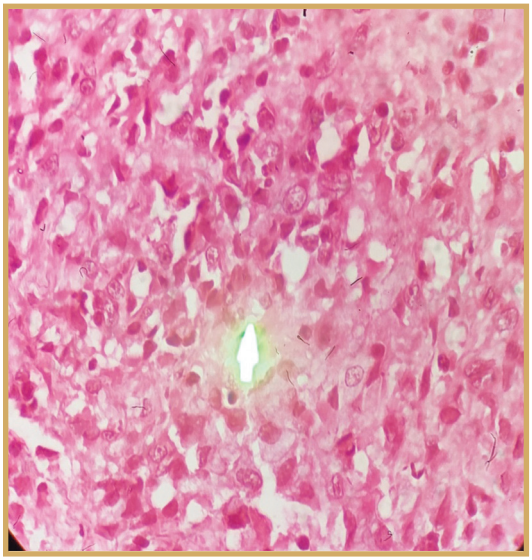

Fig. 4 : H & E-stained skin biopsy sections (oil immersion, 100X) showing macrophage granuloma.

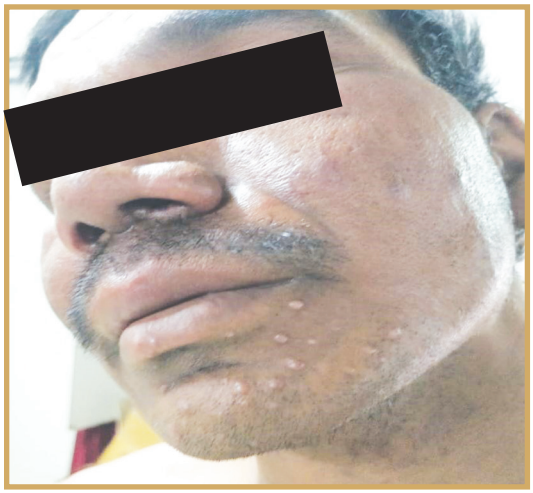

Fig. 5 : Face nodules in a patient of histoid leprosy.

multiple skin-coloured nodules of size 0.5x1 cm were noticed over face with surrounding normal skin (Fig. 5). No history of any past treatment

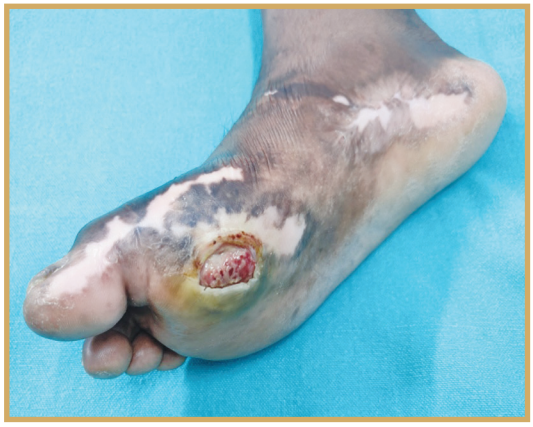

Fig. 6 : Trophic leg ulcer in a patient of histoid leprosy.

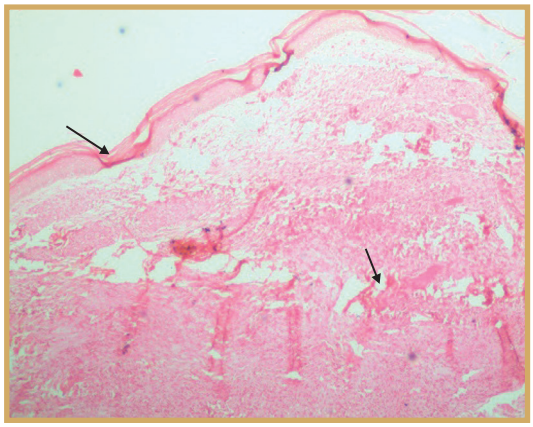

Fig. 7 : H & E- stained skin biopsy sections (40X) showing atrophied stratified squamous epithelium, loose connective zone in papillary dermis.

was identified in patient or his family members, history of close contact was also negative. All the sensations including superficial touch, temperature, pain and pressure were found intact. Nerve examination revealed thickened and non-tender ulnar, common peroneal and posterior tibial nerve. He had a solitary trophic

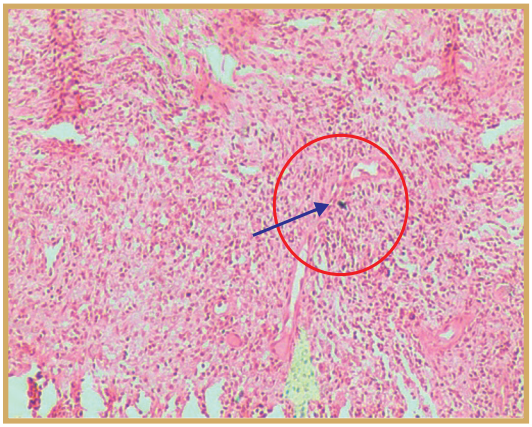

Fig. 8 : H&E sections (100x) showing spindle cell histiocytes arranged in whorls, storiform pattern with scattered lymphocytes and plasma cells.

ulcer on right foot and deformities over fingers of both hands in the form of mild clawing of digits (Fig. 6). Wasting of small muscles of hands, thenar, hypothenar eminence was present. There was associated hypoaesthesia in glove and stocking distribution. Systemic causes of peripheral neuropathy and other differentials of trophic ulcers were ruled out. SSS was done and biopsy sample was sent for histopathology. Numerous lepra bacilli were noticed from the SSS and nodules of the patient. SSS was positive for AFB with a BI of 4+ and MI was 50%. Histopathology revealed thinning of epidermis with granulomas of spindle shaped cells arranged in whorls within dermis (Figs. 7, 8). Few foamy cells were also appreciated. The patient was started on 3 drugs MB-MBT and single dose ROM, care of hypo-aesthetic foot was explained and collagen dressing done for the trophic ulcer of the foot. The patient was advised regular physiotherapy and hand splints were provided to the patient for correction of deformity of upper limb which included clawing of digits. He was diagnosed as a case of lepromatous leprosy primarily who developed histoid lesions in the absence of timely treatment. The course of treatment was uneventful with no reported reactions or ENL observed for a period of one year .

Case 3

A 42 years old male patient resident of West Bengal, Hubli presented with chief complaints of multiple asymptomatic shiny flat topped skin lesions of 2 months duration. The patient had

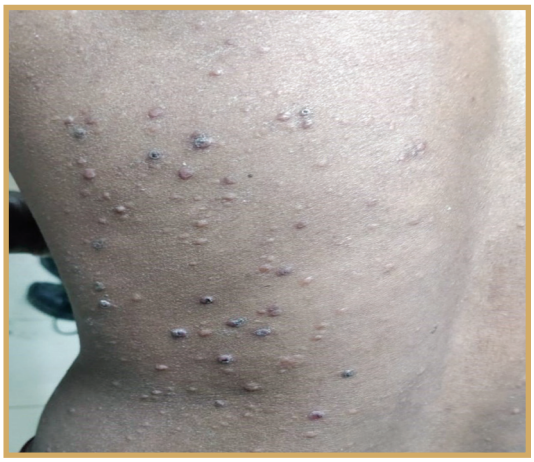

Fig. 9 : Multiple papules and nodules of histoid leprosy.

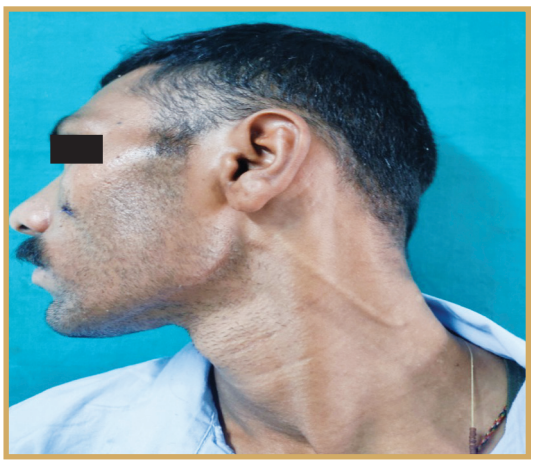

Fig. 10 : Thickened greater auricular nerve.

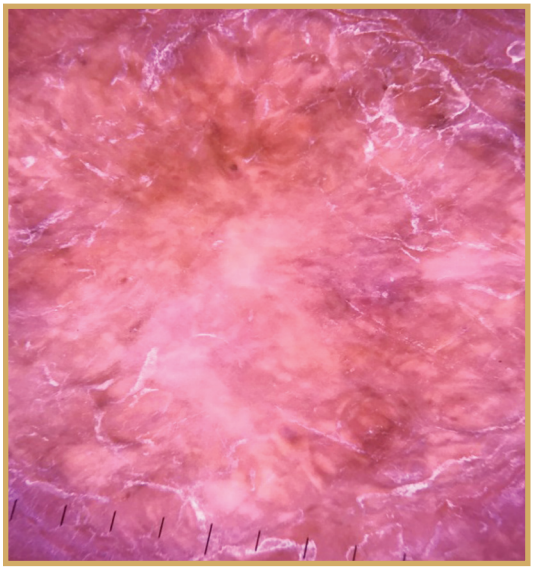

Fig. 11 : Dermoscopic appearance of the lesions, revealing brown structureless area surrounded by white streaks suggestive of scaling and accentuation of pigmentation at the periphery of the lesion.

been seen by many specialists before he was referred for skin consultation. There was no associated history of loss of sensation, weakness, pain and deformities anywhere on the body. There was no history of past treatment or any contact with a patient of leprosy in self and family members. There were multiple skin coloured papules and nodules distributed on trunk of the individual (Fig. 9). On nerve examination greater auricular nerve, bilateral ulnar and common peroneal nerves were palpably thickened and non-tender (Fig. 10). He was suspected as a case of *de novo* histoid leprosy. The other differentials of papulonodular lesions were suspected and ruled out. SSS from lesion was positive for AFB, bacterial index was 6+ and morphological index was 60%. Biopsy sample was taken from a skin lesion and sent for histopathology. Histopathology showed findings typical of histoid variant of leprosy. Dermoscopy of the lesions revealed brown structureless area surrounded by white streaks suggestive of scaling and accentuation of pigmentation at the periphery of the lesion (Fig. 11). The patient was started on 3 drugs MDT and single dose ROM.

Discussion

Histoid leprosy (HL) was earlier predominantly seen in patients treated with dapsone monotherapy as relapse or in the settings of absent, inadequate, or irregular treatment. *De novo* cases of HL are also observed and reported, though rare. In our study two cases were of *de novo* HL out of the three cases reported. One patient histoid leprosy occurred in the setting of absence of treatment, trophic ulcer, hypoanaesthetic feet and recurrent epistaxis of six months duration. He also had clawing of fingers and claw hand. The other two patients presented with asymptomatic skin lesions and nerve thickening. Numerous AFB were seen in SSS from the lesions of all the three patients. On histopathologic examination, features specific to histoid leprosy were seen.

Histoid leprosy has a prevalence of about 1.2% of all leprosy patients and 8.7% of lepromatous leprosy. (Pathania et al 2019). Reactions are rare in histoid leprosy. Erythema nodosum leprosum has been reported in case reports during the era of sulfone treatment (Bhutani et al 1974, Kalla et al 2000). Histopathologically three variants are described, these are pure fusocellular, fusocellular with epithelioid component, and fusocellular with vacuolated cells. The third pattern is the most commonly seen (Patnaik et al 2008). Numerous globi are seen which are teeming with many acid-fast bacilli, and glial substance is missing. Bacilli are longer than normal, uniform, and are stacked in parallel bundles lying along the long axis of histiocytes.

Wade has described contaminated tuberculoid granulomas which are isolated collections of these granulomas within the histiocytes (Wade 1960).

Histoid leprosy usually responds well to standard MB-MDT, however, in a case report by Pathania et al 2019) a modified MDT with daily minocycline, ofloxacin and clofazimine was administered for the patient who was hepatitis B positive to avoid risk of hepatotoxicity. Regimens for treatment of all three patients of histoid leprosy in our cases included single dose of ROM (Rifampicin 600mg+ Ofloxacin 400mg+ Minocycline 200 mg) as rifampicin rapidly reduces bacillary load and ofloxacin with minocycline have strong bactericidal action and standard MB-MDT for 12 months (Pathania et al 2019). All three patients responded to standard treatment in terms of clearance of preexisting lesions, no new lesions and decrease in bacillary load. Hence there arose no need to ponder over possibilities of antibiotic resistance or their testing. Studies have demonstrated that data on complete clearance or relapse of histoid cases was limited and there remains no consensus on treatment duration for histoid leprosy, however standard MB-MBT therapy is a safe and efficacious tool for treatment of histoid leprosy (Gregory et al 2020). In our cases none of the patients developed type1 or type 2 reactions. The bacillary load in histoid lesions is very high and untreated cases remain as reservoirs of infection, potential vectors of spread and source of new cases. This makes undiagnosed and untreated sporadic cases of histoid leprosy a community menace (Palit & Inamadar 2007)

Conclusion

Histoid leprosy is a rare form of lepromatous leprosy which earlier used to present as relapse following dapsone monotherapy and now also in case of incomplete or irregular MB-MDT. In view of rare atypical variants of leprosy which may go unnoticed and untreated a constant high degree of suspicion is required for reporting of such cases so as to aid in early diagnosis and prompt treatment of these cases especially the bacilliferous types.

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How to cite this article : Sharma L, Pathania V, Singh A et al (2023). Histoid Leprosy: Are We Still Harbouring Potential Community Menace? A Case Series from North-East India. *Indian J Lepr.* **95** : 139-146.