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Case Series

A Case Series of Histoid Leprosy with a Brief Comparison of the Clinical Features with that of Lepromatous Leprosy

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Histoid leprosy (HL) is a rare highly bacilliferous variant of lepromatous leprosy (LL). We are describing here a case series of HL in a tertiary care centre along with a comparison of the clinical features with cases of LL encountered in the same centre. There were 6 cases of HL in our centre for the past 10 years accounting for 1.86% of the total number of leprosy cases. HL constituted 11.54% of the total LL cases. 4 cases were de novo HL and 2 cases due to relapse. Papules, plaques and nodules were the commonest primary skin lesions. The distribution was localised in HL, mainly confined to the upper and lower limbs, while in LL it was symmetrical and generalised and mainly localised to the trunk. Superciliary madarosis, ear lobe infiltration, glove and stocking type of anaesthesia were found in the majority of LL, while lacking in HL patients. Type 2 lepra reaction and Grade 2 disability was much more common in LL, than HL. A very important finding in this case series is that the mean BI and MI of HL was more than in LL. All the patients were given of MB-MDT for 12 months and in patients who had initial BI of 4+ or more were given 24 months treatment and there were no cases of relapse after release from treatment.

Keywords : Histoid Leprosy, Lepromatous Leprosy, Comparison

Introduction

Histoid leprosy (HL) is a rare highly bacilliferous variant of lepromatous leprosy (LL) first described by Wade in 1963 (Wade 1963). Wade first described this in patients who were on long term dapsone monotherapy and attributed it to drug resistance. However, de novo cases are now being reported (Nair & Kumar 2013). HL patients may harbour drug resistant mutant strains. The clinical presentation of HL is smooth, shiny, succulent dome shaped papules, nodules and plaques arising from an apparently normal looking skin in contrast to lesions of LL which arises from an infiltrated skin. Moreover HL presents with localised lesions, when LL presents with generalised symmetrical lesions. We are reporting here a case series of HL and comparing the clinical features with that of LL.

Case Series

In this 10 year retrospective case series there were 6 cases of HL and 46 cases of LL. HL constituted 1.86% of the total leprosy cases (322) and 11.54% of the total LL cases. 4 cases were de novo HL and 2 cases were relapse cases, one from histoid leprosy itself and one from a case of LL subpolar. The relapse in both cases occurred within 6 months after RFT. The salient clinical and smear features of the HL are given in Table 1,

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Table 1 : Salient features of the histoid leprosy cases included.

No.	Gender	Age (years)	Type	Duration taken to detect (months)	Primary skin lesions	Distribution	Nerve thickening	Type 2 reaction at onset	Type 2 reaction at follow up	Grade 2 disability	Mean BI	MI (%)
1	Female	35	De novo	6.5	Papules, Nodules	Upper limb, Posterior trunk	Both ulnar	Nil	Nil	Ulnar palsy	6	70
2	Female	34	De novo	4	Nodules	Upper limbs, Lower limbs, trunk	Both ulnar, Both radial cutaneous, Superficial peroneal	Nil	Nil	Nil	5.67	70
3	Male	30	Relapse	13	Papule, Nodule, Umbli-cated papules	Trunk	Both ulnar, Both radial cutaneous, Both com-mon peroneal, Both tibial	Present	Nil	Nil	4	70
4	Male	38	De novo	8.25	Papule, Nodule	Upper limbs, lower limbs	Both great auricular, Both ulnar, Both radial cutaneous	Nil	Nil	Nil	6	60
5	Male	57	De novo	6	Papule, Plaque	Upper limbs, Posterior trunk	Both ulnar, Both radial cutaneous, Both com-mon peroneal, Both supercial peroneal	Nil	Nil	Nil	5	15
6	Male	48	Relapse	11	Papule, Plaque	Upper limb, Lower limb	Both ulnar, Both Radial cutaneous, Both com-mon peroneal	Nil	Nil	Nil	6	20

Abbreviations : BI-Bacteriological Index, MI-Morphological index

Table 2 : Comparison of salient features of histoid leprosy with lepromatous leprosy.

Type of leprosy	Commonest primary skin lesions	Distribution Commonest sites	Ear lobe infiltration	Madarosis	Nerve thickening	Glove and stocking anaesthesia	Type 2 reaction	Grade 2 disability	Mean BI (+)	Mean MI (%)
Histoid (n=6)	Papules, nodules, all cases, asymmertrical	Upper and lower limbs, all cases	1 (16.67%)	2 (33.33%)	5 cases symmetrical (83.33%)	Nil	1 (16.67%), ENL	1 (16.67%)	5.45	50.83
Lepromatous leprosy (n=46)	Papules, Plaques (32 cases, (69.57%), symmetrical	Trunk, (39 cases, (84.78%)	42 (91.30%)	29 (63.04%)	39 cases (84.79%)	31 cases (67.39%)	9 (19.57%)	13 cases (28.26%)	3.67	43.33

while the comparison of the salient features with LL is given in Table 2. There were 4 males and 2 females in the HL series with a male/female ratio of 2:1, while there were 37 males and 9 females in the LL group with a male/female ratio of 4.1:1. The mean age in the HL group was 40.33 years, while the mean age in the LL group was 43.67 years. The mean duration of illness was 8.13 months in the HL group and 7.56 months in the LL group. There was no history of contact in the HL series while 9 cases (19.57%) in the LL group had positive history of contact, out of which 4 cases had family contacts. All the HL cases did not have any family members involved. The commonest clinical presentation was asymmetrical papules and nodules in the upper and lower limbs in all the 6 cases of HL (100%), while 4 cases had lesions on the trunk also, while in the LL group it was papules and plaques distributed on the trunk in 84.78% of the cases, while symmetrical distribution was seen in 69.57% of the cases. 5 cases of HL had symmetrical nerve thickening,

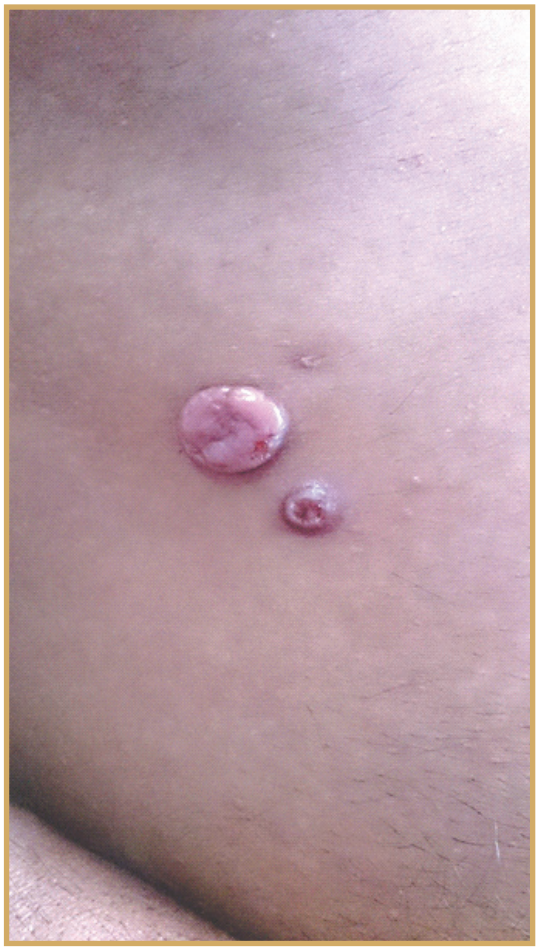

Fig. 1 : Smooth shiny succulent plaques and papules arising from a normal skin.

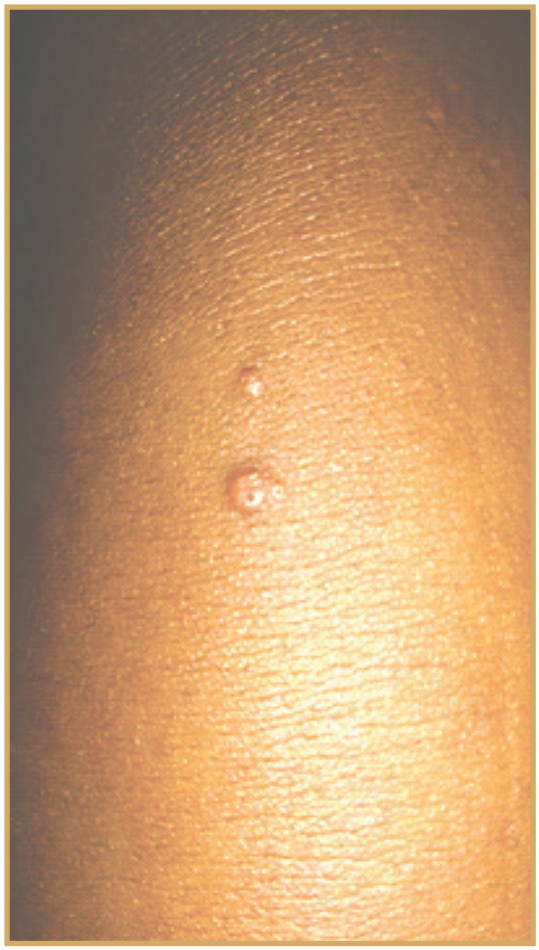

Fig. 2 : Umblicated papules of histoid leprosy.

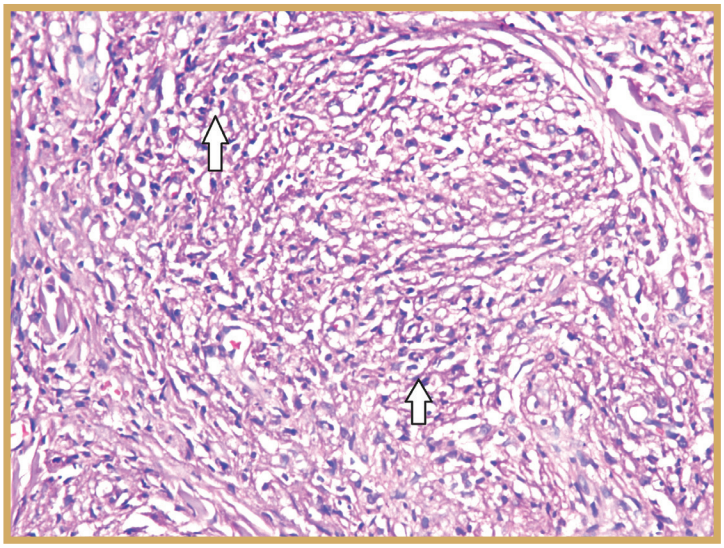

Fig. 3 : Histopathology showing spindle shaped histiocyes arranged in whorls and circles (H & E, x 400).

while in the LL group symmetrical nerve thickening was seen in 39 cases. Ear lobe infiltration was seen in one case of HL and 42 cases of LL, while madarosis was seen in 2 cases of HL and 29 cases of LL. No HL case had glove and stocking type of anaesthesia, while 31 cases in the LL group had glove and stocking anaesthesia. 1 case of HL had ENL, while 9 cases of LL had ENL. 1 case of HL had Grade 2 disability (ulnar palsy), while 13 cases of LL had Grade 2 disability. There was no systemic involvement in the HL group, while 6 cases in the LL group had generalised lymphadenopathy and 2 cases hepato-splenomegaly. 5 cases of HL had 24 months MB-MDT, out of which 2 had ofloxacin also added in the regimen. 1 case of HL which was a relapsed case of HL was given alternate regime. In the LL group 14 cases were given MB-MDT, FDT for 12 months while 32 cases were given MB-MDT for 24 months. Fourteen cases of LL were given the standard 12 month WHO regimen, whereas 32 cases of HD LL were given 24 months treatment since there initial BI was 4+ or more and hence extended treatment according to recent WHO recommendations. WHO only recommends standard MB-MDT for HL, but in our department protocol we add 400 mg ofloxacin also in HL cases as there are reports that adding ofloxacin rapidly reduces the bacillary load (Vora et al 1995, Bartos et al 2020). Both groups showed good response to MB-MDT. In the 12 cases of LL who were given 12 months FDT, showed a mean BI of 2.13 after 12 months of FDT, while MI was 0%, while in the LL group who was given 24 months MB-MDT mean BI after 24 months of MB-MDT was 0.53, MI 0%. In the HL group the mean BI after 24 months MB-MDT was 0.77, MI 0%, while in the 2 cases who were given ofloxacin also, the mean BI was 0.46. Both groups were followed upto 1 year after RFT. There were no relapse cases in the follow up period in both groups, while 4 cases in the LL group developed ENL lesions.

Discussion

In this 10 year retrospective case series we had 6 cases of HL. This shows the rarity of HL. In a previous study done in the same institute we could detect only 17 cases of HL in a 20 year period (Nair & Kumar 2013). However, eventhough HL is rare, they are highly bacilliferous and form a significant stumbling block in the era of elimination of leprosy. There was not much difference in the mean age, duration of illness and gender ratio in both the HL and LL group and this was consistent with other studies (Sehgal et al 2009, Raheja et al 2022). However, we would find significant differences in the clinical presentation, smear findings and histological features of HL compared to LL in this case series. HL mainly presented with asymmetrical lesions, while LL presented in the majority with symmetrical involvement. Morphology of the lesions also showed differences, with nodules more commonly seen in HL than LL. The extremeties were more commonly involved in HL, while in LL it was the trunks. Ear lobe infiltration and madarosis was seen in LL cases more. However symmetrical nerve thickening was seen both in HL and the LL groups. Glove and stocking anaesthesia was seen only in the LL group. ENL was more common in LL than HL which is consistent with other studies (Pandit & Sumathi 2021). Grade 2 disability was also more frequent in the LL group. HL is also characterised by different morphological presentations compared to LL. The papules and nodules are smooth, shiny, succulent arising from an apparently normal skin in HL, in contrast to LL where the lesions arise from an infiltrated skin (Fig. 1). Molluscum contagiosum like lesions were the the only atypical variant seen in this case series (Fig. 2). Other case series reported keloid like, xanthoma like, sarcoid like and tumor like presentations and this can lead to misdiagnosis unless a strong clinical suspicion is maintained (Thappa et al 2001, Nair et al 2006 & 2016, Mohapatra et al 2018).

An important smear finding in this case series of HL was the mean BI and MI was more than in cases of LL, and the mean MI of 50.83% in HL cases is much higher than other reported case series and indicates the highly bacilliferous nature of HL (Sehgal et al 2009, Pandit & Sumathi 2021, Raheja et al 2022). There are significant histopathological differences between HL and LL. In fact HL is always diagnosed in conjunction with biopsy findings. All our cases had the classical spindle shaped histiocytes arranged in whorls, circles and curlicues packed in the dermis (Fig. 3), while LL cases had the classical macrophage granulomas with foamy changes. A recent study (Da Costa et al 2013) using immunocytochemistry in both HL and LL demonstrated that both the spindle cells in HL and foamy macrophages in LL stained for CD68 indicating common macrophage lineage. However, the exact reason why there is spindle histiocyte response in HL and foamy macrophage response in LL in the same disease caused by *M. leprae* is not known. There are a few studies indicating that the spindle shaped histiocytic response may be due to the mutant strains of *M. leprae* residing in HL (Da Costa et al 2013). However, the so called “tuberculoid contamination” was not seen in our series. This denotes the presence of epitheloid cells among the spindle shaped hsitiocytes indicating a downgrading from a tuberculoid spectrum to HL (Seghal & Srivastava 1987).

The HL case series and the LL group showed good response to MB-MDT. In 2 cases of HL we also added ofloxacin 400 mg as there are reports that there can be rapid reduction in the BI and MI with ofloxacin in the regimen (Vora et al 1995, Bartos et al 2020). An interesting finding in this study was that the mean BI in LL cases was much less in cases given 24 months MB-MDT than in

cases with 12 month MB-MDT after completing therapy. Thus WHO advocates that if the initial BI is 4+ or more, it is advisable to continue MB-MDT even after 12 months to prevent relapse. We currently follow these guidelines for cases with initial BI of 4+ or more (Bartos et al 2020).

In conclusion, HL is a rare variant of LL with differences in the clinical and histopathological presentations compared to LL. The variable clinical presentations can confuse the dermatologist and misdiagnosis is possible. They are higly bacilliferous and form a major hurdle for the control programmes. Further case reports and studies may throw light on this unique and fascinating form of leprosy.

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