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Case Series

De-novo Histoid Leprosy: A Series of Four Cases

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Histoid leprosy, an unusual variant of lepromatous leprosy, originally thought to manifest following dapsone monotherapy, is being increasingly reported 'de-novo'. We hereby report four de-novo cases; one mimicking lupus pernio presenting with an uncommon lesion (plaque) at an extremely rare site (nose). Of the remaining three classical cases, one exhibited a not-so-frequent development of erythema nodosum leprosum (ENL) reaction that needed modified multidrug therapy for two years, other three responded well to standard multibacillary multidrug therapy which was also given for two years.

Keywords : Histoid, Leprosy, De-novo, Reaction

Introduction

Histoid leprosy, a rare variant of lepromatous leprosy, presents with distinct isolated nodules, high bacillary index and characteristic histopathological findings of interlacing bands of spindle-shaped fibroblast-like macrophages, likely arising from tissue histiocytes, simulating dermatofibroma (Job et al 1977, Sardana et al 2020, Sehgal et al 2009). The nodules, cutaneous and/or subcutaneous, may rarely form plaques. Histoid leprosy has been reported from India, it has been clinically, bacteriologically, histologically and immunologically characterised in pre-MDT era also (Sehgal et al 1985, 1987). This variant can appear de-novo in cases of lepromatous leprosy, or in inactive cases thereof with or without relapse, unlike in the era prior to WHO-MDT when majority of its cases emerged among patients with lepromatous leprosy on dapsone monotherapy (Meena et al 2017, Nair & Kumar 2013).

All these four cases seen recently in our outpatient department and being presented herein were de-novo. A solitary lesion (plaque) on an exceedingly rare site (nose) in case 1 confounded us with the diagnostic probability of lupus pernio. Case 4 developed a not-so-commonly seen reaction, in histoid leprosy, of erythema nodosum leprosum (ENL) (Kalla et al 2000, Sharma et al 2002, Nair & Kumar 2013, Pandit & Sumathi 2021). None gave history of contact with a known case of leprosy, or of unusual signs, symptoms such as pain, fever, numbness, weakness, swollen hands and feet, nasal

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stuffiness, difficulty in buttoning and slipping of chappals or previous treatment of leprosy.

Case Reports

Demographic details, clinical presentation, disease duration, bacillary and morphological indices of slit skin smear for AFB (L) and skin biopsy findings of all four cases are presented in Table 1.

Case 1

A Nepalese male, aged 44 years, presented with a gradually increasing swelling of nose due to an overlying erythematous infiltrated firm plaque with telangiectatic surface (Fig. 1). He also complained of wheezing and difficult breathing. He was provisionally diagnosed as a case of lupus pernio and differentials of histoid

Table 1 : Demographic, clinical and laboratory details of cases of histoid leprosy

Case No.	Age	Gender	Domicile	Site of lesions	Primary lesion	Duration (months)	SSS		Biopsy index
							BI	MI	
1	44	Male	Nepal	Nose	Plaque	3	5+	50%	5+
2	68	Female	Maharashtra	Face, dorsum of hands	Plaques, nodules	6	4+	40%	4+
3	40	Female	Bihar	Upper limb	Papules, nodules	9	5+	40%	5+
4	27	Male	Maharashtra	Face, lower limbs	Papules, nodules	13	6+	60%	6+

Abbreviations : BI= Bacillary index, MI= Morphological index, SSS= Slit skin smear

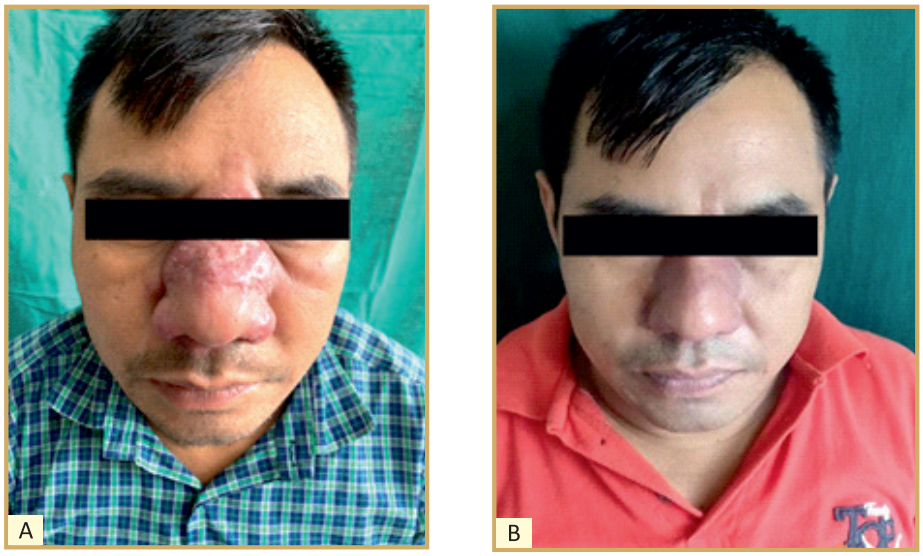

Fig. 1 : Case 1 - (A): Well-defined erythematous, infiltrated plaque over the nose sparing the tip, (B)- After 3 months of treatment.

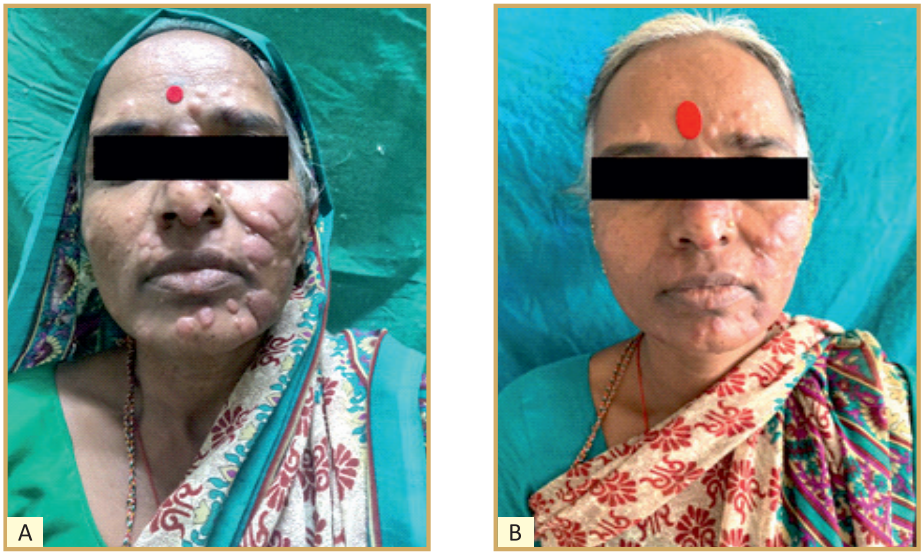

Fig. 2 : Case 2 - (A): Multiple, well-defined, erythematous, infiltrated plaques over the face, (B) After 3 months of treatment.

leprosy, lupus vulgaris and granulomatous rosacea were considered. Ziehl-Neelsen stain of slit skin smear revealed bacillary index (BI) of 4+ and thereby confirmed multibacillary leprosy.

Cases 2, 3 and 4

Cases 2, 3 and 4 had classical findings of histoid leprosy— isolated, dome-shaped, firm papulonodules over apparently normal skin (Fig. 2), high BI on slit skin smears and characteristic histopathological findings described below. Case 4 revealed thickened but non-tender bilateral ulnar and common peroneal nerves without any sensory or motor deficit; other three cases had no evidence of neuritis.

Histopathological findings in all four cases revealed atrophic epidermis, perivascular and periadnexal mononuclear cell infiltrate with loads of bacilli, majority solid staining acid fast, within diffuse collection of foamy macrophages and spindle shaped histiocytes arranged in tight bands, curlicues and whorls (Fig. 3).

All four were initially put on multibacillary multidrug therapy (MB-MDT); three responded well (Figs. 1 and 2). However, case 4 who soon after receiving the first supervised dose developed high grade fever with chills, sore throat and breathlessness was given modified MDT regimen comprising once daily clofazimine (50mg), ofloxacin (400mg) and minocycline (100mg) for 6 months followed by clofazimine (50mg) and minocycline (100mg) for additional 18 months. One month after commencing treatment, he developed swollen hands and feet, fever, pain in the (thickened) ulnar and common peroneal nerves, and crops of painful evanescent papulonodules. Diagnosed to have developed ENL reaction, he was treated initially with systemic glucocorticoids with satisfactory response; however, this flared up on tapering thereof over 3 months and was administered additional thalidomide. Regular follow up – monthly till on MDT and 6 monthly thereafter- showed satisfactory response in all four cases.

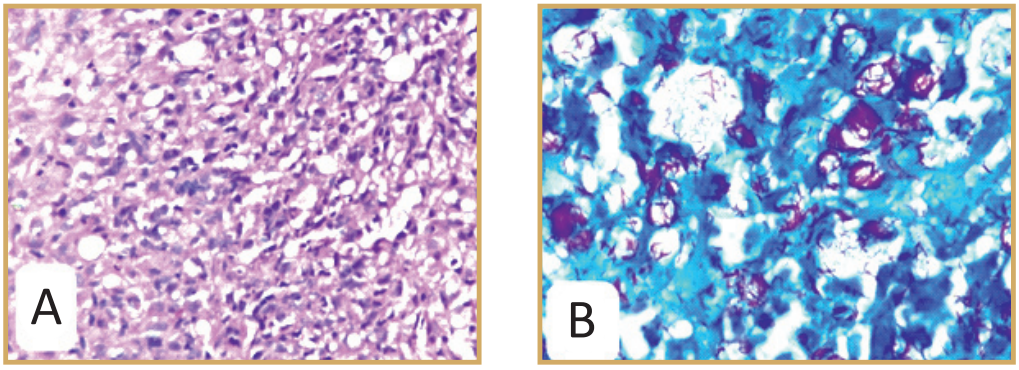

Fig. 3 : (A) - Numerous spindle shaped histiocytes and few foamy macrophages (H&E,40X). (B) - Acid fast bacilli with a BI=6+ (ZN stain, 100X).

Table 2 : Indian studies of histoid leprosy cases in post-WHO multi-drug therapy era

Study, Year of publication with reference	Number of histoid/total leprosy cases with percentage of cases of histoid leprosy	Number with solitary nasal swelling/ facial involvement	Cases with Lepra reaction (Type 1)	Cases with Lepra reaction (Type 1)	Remarks
Kalla et al (2000)	25/893 (2.8%)	0/13	Nil	2	Cases spanning over 11 years
Nair & Kumar (2013)	17/829 (2.05%)	0/3	Nil	4	Cases spanning over 10 years
					De-novo= 11;
					Originally lepromatous leprosy= 5;
					Indeterminate on dapsone monotherapy relapsed as histoid leprosy= 1
Singh et al (2015)	1	0/1	1	Nil	
Pandit & Sumathi (2021)	3	0/3	Nil	1	
Swain et al (2015)	1	Nasal mucosa without any swelling/ plaque	Nil	Nil	Presented an ENT specialist with nasal mucosal lesion only
Tiwary et al (2017)	1	0/1	Nil	Nil	
Sharma et al (2002)	1	0/1	Nil	1	
Total	49	0/23	1	8	

Discussion

Histoid leprosy, first described by Wade in 1963, is an expression of multibacillary leprosy (Wade & Tolentino 1963). Sehgal et al (2009) from India emphasized the need to pay special attention to this exquisite form of multibacillary disease in the post-elimination era of leprosy. Originally believed to be associated with dapsone mono-therapy or inadequate treatment, de-novo histoid leprosy is being increasingly reported (Wade & Tolentino 1963, Sehgal et al 2009, Bhat et al 2015, Mohapatra et al 2018).

Clinically, unlike diffuse infiltrated skin of the usual cases of lepromatous leprosy, histoid leprosy presents with well-defined smooth shiny dome-shaped firm to hard, nontender and freely movable, papules and nodules over apparently normal skin of extensor extremities, back and buttocks (Bhat et al 2015). Facial involvement seen in three of our cases was particularly noteworthy in case 1 being extremely unusual presentation with a solitary nasal cutaneous plaque mimicking lupus pernio (Fig. 1).

Contrary to the common perception of rarity of ENL reaction in histoid leprosy (Kalla et al 2000, Sharma et al 2002, Nair & Kumar 2013, Singh et al 2015, Swain et al 2015, Tiwary et al 2017, Pandit & Sumathi 2021), case 4 was undergoing ENL reaction during his visit for second monthly dose of MDT. Ramanujam and Ramu (1969) postulated that the nodules in histoid lepromatous cases tend to soften from the centre of nodule which in some instances can precede the acute exacerbation of existing lesions — classical type I lepra reaction –- with pustules and ulceration of nodules with or without constitutional symptoms.

Few useful pointers that emerge from juxtaposition of this series of our cases vis-a-vis the reports for histoid leprosy from India including two larger Indian studies (Kalla et al 2000, Nair & Kumar 2013); latter analysing 42 cases of histoid leprosy from among all cases of leprosy (n= 1722) spanning over a decade in the post-WHO MDT era (Table 2) are: first, nasal cutaneous swelling seen as a presentation in one of our cases, was not reported in any of the 23 cases with facial lesions— the solitary case reported by an ENT specialist (Swain et al 2015) involved only the nasal mucosa; second, while ENL was seen uncommonly (16.32%) in these larger studies (Kalla et al 2000, Nair & Kumar 2013), type 1 reaction was even rarer (2.08%), thirdly, percentage of histoid leprosy among all cases in the above mentioned two larger Indian studies was 2.8% and 2.05% only and finally, percentage of de-novo (100% of ours) cases was 64.70% among the large study by Nair & Kumar (2013) whereas Kalla et al (2000) having not analysed the same.

Currently there are no clear recommendations regarding the treatment of histoid leprosy. Most clinicians follow the WHO regime for multibacillary therapy (MB-MDT) extended for a minimum of 2 years, preferably till smear negativity (Bartos et al 2020). Three of our cases received multibacillary therapy for 2 years whereas case 4 was given modified multidrug therapy comprising clofazimine, ofloxacin and minocycline for a total duration of 24 months following non-tolerance to supervised dose of multibacillary therapy (MB-MDT).

To conclude, the highly bacilliferous, though less common, histoid variant of lepromatous leprosy, is increasingly occurring de-novo; can occasionally give a clinical surprise occurring at an exceedingly rare site and with less common lesion and may sometimes be in reaction. Early recognition of de-novo cases with prompt institution of sufficient duration of WHO MB-MDT therapy under prolonged follow-up is important to ensure favourable outcome.

References

1. Bartos G, Sheuring R, Combs A et al (2020). Treatment of histoid leprosy: A lack of consensus. *Int J Dermatol*. **59**: 1264-1269.
2. Bhat YJ, Hassan I, Yaseen A, Wani R (2015). De novo histoid leprosy: A case report from a post-elimination area. *Indian J Dermatol*. **60**: 214.
3. Job CK, Chacko CJG, Taylor PM (1977). Electron microscopic study of histoid leprosy with special reference to its histogenesis. *Lepr India*. **49**: 467–471.
4. Kalla G, Purohit S, Vyas MC (2000). Histoid, a clinical variant of multibacillary leprosy: report from so-called non endemic areas. *Int J Lepr Other Mycobact Dis*. **68(3)**: 267-271.
5. Meena DK, Sharma S, Chauhan P (2017). De-novo Histoid Leprosy Masquerading as Cryptococcosis: A Case Report. *Indian J Lepr*. **89**: 45-48.
6. Mohapatra PS, Ray R, Khandare M (2018). A rare presentation of histoid leprosy: A case report. *Indian J Lepr*. **90**: 75-78.
7. Nair SP, Kumar GN (2013). A clinical and histopathological study of histoid leprosy. *Int J Dermatol*. **52(5)**: 580–586.
8. Pandit VS, Sumathi S (2021). Histoid Leprosy: Case Series from Non-Endemic Region in Post-Elimination Era. *Indian J Lepr*. **93**: 415-422.
9. Ramanujan K, Ramu G (1969). Wade's histoid lepromatous leprosy. Report of a clinical study. *Lepr India.* **41**: 293–297.
10. Sardana K, Bhushan P, Khurana A (2020). Clinical Leprosy. In: Jopling's Handbook of Leprosy, 6th edn (Sardana K, Khurana A, eds), CBS Publishers & Distributors, New Delhi, pp6-58.
11. Sehgal VN, Srivastava G, Saha K (1985). Immunological status of histoid leprosy. *Lepr Rev*. **56**: 27–33.
12. Sehgal VN, Srivastava G (1987). Status of histoid leprosy – a clinical, bacteriological, histopathological and immunological appraisal. *J Dermatol (Tokyo).* **14** : 38–42.
13. Sehgal VN, Srivastava G, Singh N, Prasad PVS (2009). Histoid leprosy: the impact of the entity on the postglobal leprosy elimination era. *Int J Dermatol*. **48**: 603-10.
14. Sharma SK, Rath N, Gautam RK et al (2002). Histoid leprosy with ENL reaction. *Indian J Dermatol Venereol Leprol*. **68**: 342-343.
15. Singh N, Kumari R, Gupta D et al (2015). Type 1 lepra reaction in histoid leprosy. *Int J Dermatol.* **54(5)**: 564–567.
16. Swain SK, Jain AK, Panda M et al (2015). Isolated case of mucosal histoid Hansen's disease of the nasal cavity in a post global elimination era. *J Infect Public Health*. **8(6)**: 630-3.
17. Tiwary AK, Kumar P, Agarwal S (2017). Palmoplantar, Genital and Lip involvement in a de-novo case of Histoid leprosy. *Lepr Rev*. **88(2)**: 280–284.
18. Wade HW, Tolentino JG (1963). The histoid variety of lepromatous leprosy. *Int J Lepr*. **31**: 608–609.

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