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Case Reports

Uncommon Presentations of Leprosy

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Leprosy mainly presents typical features like hypopigmented, hypo anesthetic patch(es) with or without involvement of nerve(s). But in hyperendemic areas, the presentation may vary greatly, ranging from nodular, keloidal, or pustular forms of disease. We are reporting here three cases with uncommon presentations of leprosy which mimicked a wide range of dermatological conditions. The first case had a plethora of pustular lesions all over the body, resembling subcorneal pustular dermatosis (SPD). In the second case, a young male patient had just keloid-like plaques on his face; histology revealed that the condition was Histoid Hansen's. The third case had two patches and a widespread, asymptomatic nodular eruption; the histology of this patient revealed the simultaneous presence of two distinct disease poles. All these cases developed de novo Hansen's disease. In our cases, diagnosis was confirmed by bacteriological and histopathological assessment of patients. One of our patients presented with 'Pustular erythema nodosum leprosum' and another with atypical nodules and plaques mimicking other dermatological conditions. Thus, leprosy can be a "great mimicker". Though such uncommon manifestations can also occur in low-endemic locations, these are likely to have higher incidence in hyperendemic places.

Keywords : Pustular ENL, Atypical Histoid Hansen's, SPD, Foamy Histiocytes, Downgrading Leprosy, Unusual Presentations, Leprosy

Introduction

Leprosy is one of the oldest diseases known to man. Despite advances in all spheres of medical science, leprosy continues to be a public health challenge in countries like India (Rao & Suneetha 2018). Currently, seven states in India namely Bihar, Uttar Pradesh, Chhattisgarh, Jharkhand, Maharashtra, Odisha, and West Bengal contribute almost 3/4th of the total leprosy burden (NLEP 2015-16). Although India has achieved its elimination goal still disease is great concern in hyperendemic areas.

The prevalence rate of leprosy has come down from 0.69 per 10,000 population in 2014-15 to 0.45 per 10,000 in 2021-22 (NLEP 2022). Further, the annual new case detection rate per 100,000 population has come down from 9·73 in 2014-15 to 5·52 in 2021-22. Common presentations, such as a visible hypopigmented/erythematous, anesthetic/ hypoesthetic skin lesion, or an area of varying sensory loss on the skin. (Kumar & Dogra 2016). Rarely, the patient may present with nasal stiffness, pustular crops, epistaxis, and/ or keloid-like plaques without significant sensory impairment. Uncommon presentations may mimic a wide range of dermatological conditions which may further cause delay in diagnosis and treatment. Although precise numbers are

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unknown, we were able to record approximately 400 new cases of Hansen's disease in 2022. Theoretically, every 40th - 50th of the cases that were brought to us had uncommon features that needed to be diagnosed with histopathological examination.

In this case series, we reported three uncommon presentations of leprosy in the OPD of the Department of Dermatology, Venereology, and Leprology of our college, coming from Bihar and eastern Uttar Pradesh. Informed consent has been taken from all the patients to publish the clinical information and images anonymously.

Case reports

Case 1:

A 35-year-old male patient presented to Dermatology OPD in November 2022 with a complaint of sudden eruption of pustular lesions all over the body (Figs. 1A, 1B). On thorough examination, ear lobe infiltration and a few necrotic lesions were also found over the limbs. He had gloves and stocking anesthesia along with a bilateral thickened ulnar nerve with mild neuritis. Necrotic lesions were present from the last 6 months for which he was taking oral steroids from a non-registered medical practitioner. Pustular lesions developed on sudden stoppage of oral steroids. The patient was not taking MDT as the diagnosis was not established by the local practitioner. There was no family history or contact history. He also complained of on-and-off fever along with arthralgia. On slit skin smear (SSS) examination the bacteriological index was 6+ from ear lobes (Fig. 1C). On histopathological examination (Figs. 1D and 1E), nodular granulomatous inflammation centered around neurovascular bundles of superficial and

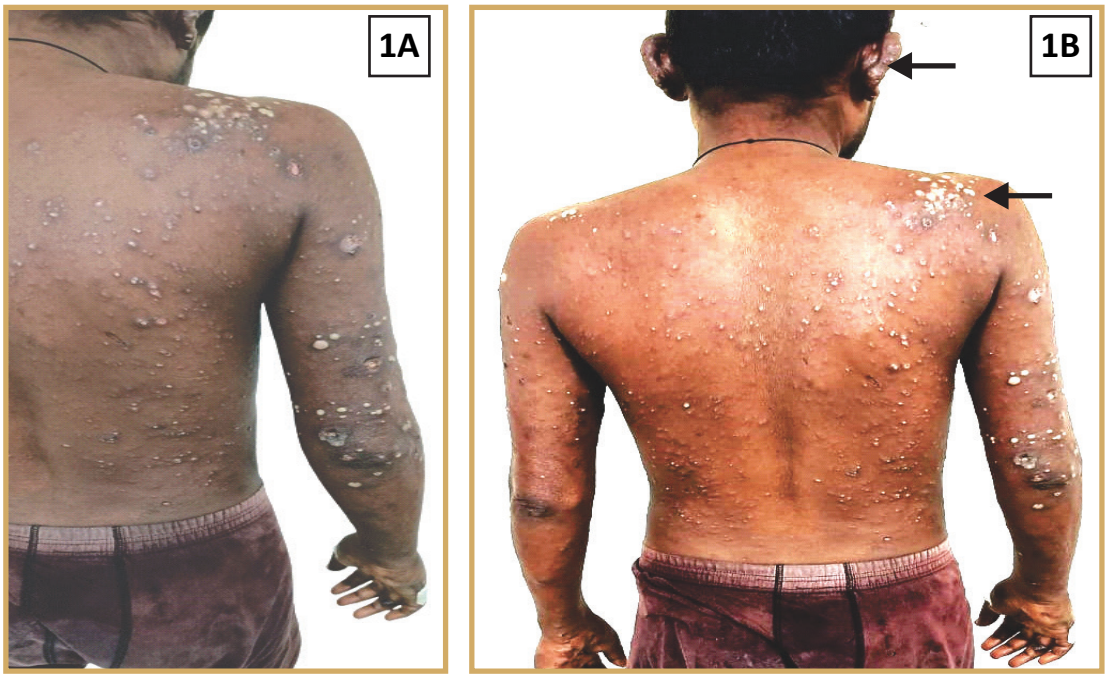

Fig. 1A : Multiple pustules along with necrotic nodules present over the trunk and upper limb.

Fig. 1B : Generalized eruption of pustular lesions more numerous on extensors and ear lobe infiltration.

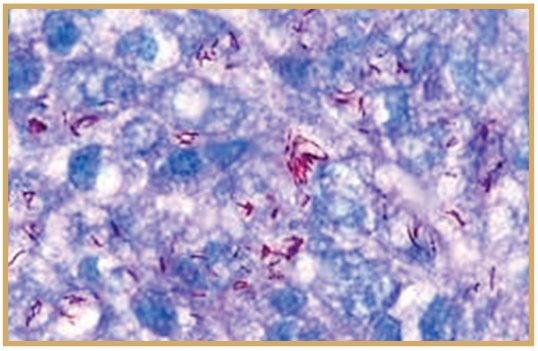

Fig. 1C : Slit skin smear (SSS) shows globi of bacilli present all over the smear.

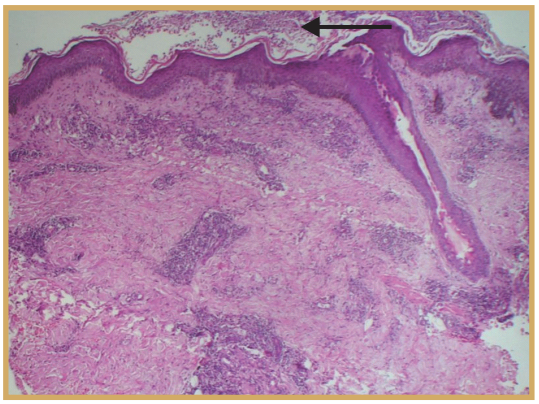

Fig. 1D : Overlying epidermis shows an intraepidermal pustule, a few neutrophils can be seen within the epidermis with spongiosis, 10x magnification.

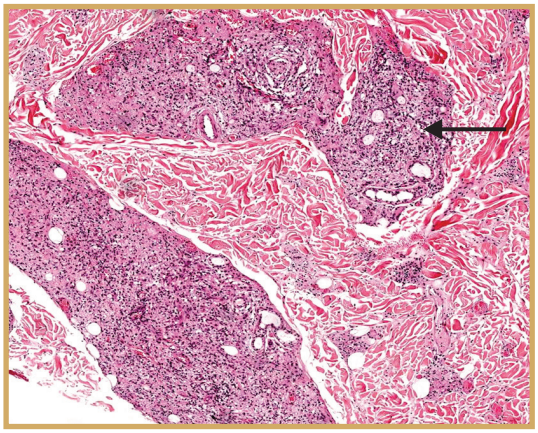

Fig. 1E : Nodular granulomatous inflammation consisting of foamy macrophages and lymphocytes, 10x magnification.

deep dermis. The granuloma consists of foamy macrophages and lymphocytes. Numerous neutrophils dot the granulomas accompanied by moderate amounts of nuclear dust and fibrin. The overlying epidermis shows an intraepidermal pustule. A few neutrophils can be seen within the epidermis with spongiosis. A final diagnosis of Pustular Erythema nodosum leprosum (Pustular ENL) was made based on clinic-pathological correlation. The patient was admitted to the Dermatology department, and MDT was started along with steroids and thalidomide. The patient responded very well to treatment. The patient is on regular follow-up (monthly for one year) and has an occasional recurrence of similar lesions.

Case 2:

A 23-year-old male patient from eastern U.P. visited our Dermatology OPD in October 2022 for complaints of gradually progressive nodular lesions over his face for the last 6 months (Fig. 2A). Lesions were asymptomatic according to the patient. The patient has not taken any treatment because the lesions were asymptomatic. There was no history of Hansen's disease in the family or contact. On examinations, multiple nodules and plaques (Fig. 2B) were seen all over his face including infiltration of nasal alae and vermilion border of lower lip. Hot, cold, and touch sensations were normal, and no nerve thickening or neuritis was found. Based on clinical findings, a differential diagnosis of cutaneous sarcoidosis, Histoid Hansen's, multiple facial cylindromas, and cutaneous histoplasmosis was suspected. On histopathology, dense monomorphous granulomatous infiltrate was seen comprising of foamy histiocytes (Fig. 2C) with a monotonous appearance involving the reticular dermis and

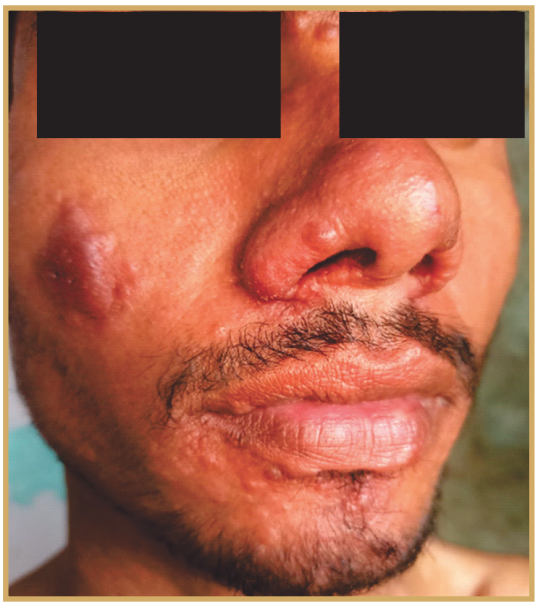

Fig. 2A : Showing multiple nodules infiltrating nasal alae, lower lip and keloidal plaques over cheeks.

Fig. 2B : Showing plaque and nodules over forehead and cheek.

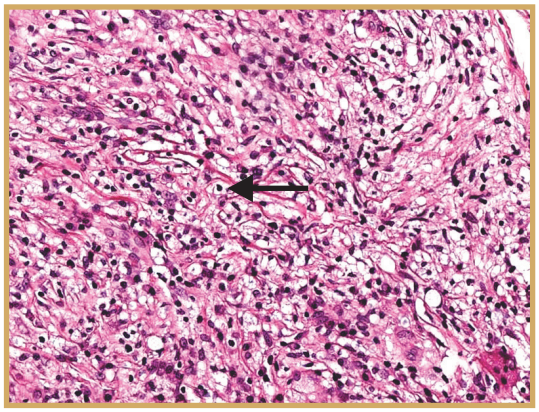

Fig. 2C : Foamy histiocytes and elongated and spindle-shaped cells forming large parallel bundles, magnification 40x.

sparing appendages and upper epidermis. In large parts of the section, the foamy histiocytes and elongated and spindle-shaped cells form large parallel bundles. The subepidermal papillary dermis (grenz zone) was spared by the infiltrate. The macrophage granuloma followed neurovascular bundles in the deep reticular dermis. So, a final diagnosis of Histoid Hansen's was concluded based on clinical-pathological correlation. The patient was started on MB-MDT and after 12 weeks, maximum resolution of lesions was seen and has been followed for 6 months.

Case 3:

A 50-year-old male patient from Bihar visited the Dermatology OPD of our college in September 2021 with complaints of the gradually progressive eruption of painless, non-tender, skin-colored nodules all over the body including the face and limbs. The patient does not have any history of Hansen's disease in family or contact. He did not have any history of pruritus, anorexia, or weight loss. On examination firm, non-tender, non-fluctuating nodules were seen all over the body including the face and limbs (Fig. 3A). On closer examination two hypopigmented, hypo-

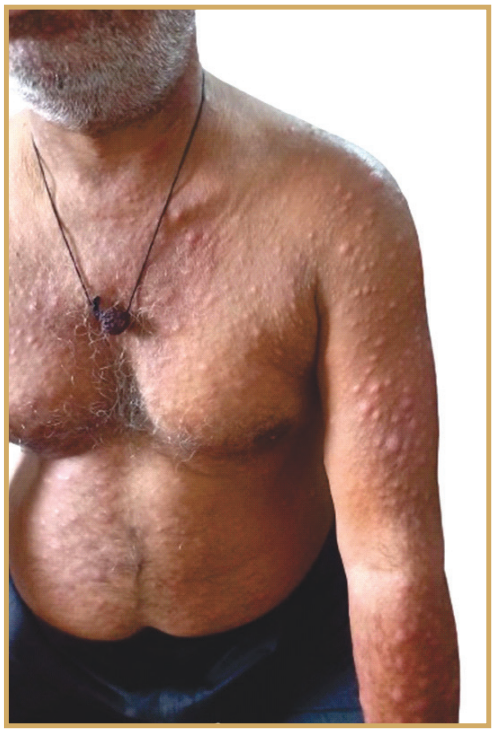

Fig. 3A : Crops of skin-coloured nodules showing all over the body.

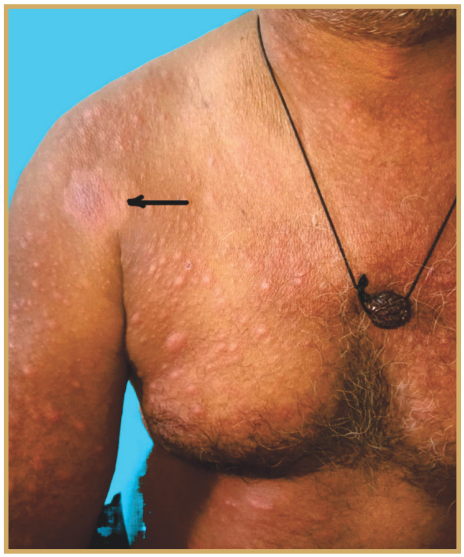

Fig. 3B : The black arrow shows a hypopigmented patch over the right shoulder.

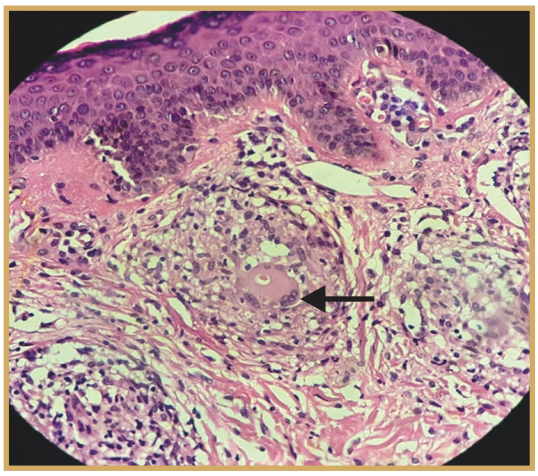

Fig. 3C : Shows well-defined epithelioid cell granuloma with Langerhans giant cell, in patch lesion (black arrow), 40x magnification

anesthetic patches were also found on the trunk (Fig. 3B). No palpable lymphadenopathy was found. Sensations were impaired over patches only, was normal rest all over the body. There was no nerve thickening or neuritis. Based on findings, a clinical differential diagnosis of aleukemic leukemia cutis, cutaneous T cell lymphoma, and Hansen's disease was suspected. Two 5mm punch biopsies were taken, one from the nodule and another from the patch.

Histopathology of the patch lesion showed tuberculoid granulomatous inflammation centered around neurovascular bundles of superficial and deep dermis. There was flattening of rete ridges and thinning of the epidermis. The infiltrate of lymphocytes, epithelioid cells, and giant cells followed appendages and involved arrector pili muscles. The dermo-epidermal junction was found to be focally infiltrated by lymphocytes. A few nerve bundles in the deep reticular dermis were also surrounded by lymphocytes (Fig. 3C). This is consistent with borderline tuberculoid Hansen's disease (BTHD). At the same time, histopathology of nodules

showed nodular granulomatous infiltrate cantered around neurovascular bundles of superficial and deep dermis and sparing the subepidermal papillary dermis (Grenz zone). The granulomas were elongated and were made up of immature epithelioid cells, pale staining foamy histiocytes, and lymphocytes with occasional plasma cells. The overlying epidermis was thin and flat which was consistent with borderline lepromatous Hansen's disease (BLHD). So, the final diagnosis of borderline tuberculoid Hansen's disease (BTHD) downgrading to borderline lepromatous leprosy (BLHD) was made. MB-MDT was started and the patient responded very well to MDT.

Discussion

Although the presentation of leprosy is distinct it has cardinal features like a hypopigmented patch along with sensory impairment and nerve thickening. However, in hyperendemic areas, leprosy is a great mimicker of a wide range of dermatological conditions. Even though rare manifestations can also occur in low-endemic locations, prompt detection and management are crucial to preventing disability and transmission because of the greater incidence in hyperendemic places.

Usually, diffuse pustular crops as a presentation of ENL are very rare, and to date, we have encountered very few such cases. In the literature, one similar case reported by Sirka et al (2021) shows the diagnostic dilemma posed by such uncommon presentations. Pustular ENL may mimic the secondary impetization of ENL and varicella. ENL may present with various atypical forms, pustular forms have been described rarely (Maymone et al 2020). Higher bacillary load and, in a few instances, bactericidal drugs like ofloxacin, have been implicated in the pathogenesis of pustular form (Dave et al 2003). In our case 1, sudden withdrawal of systemic steroids was the cause for pustular lesions. Inadequate diagnosis and treatment delays, exemplified by this case under the care of an unqualified practitioner, highlight the importance of educating the public about early symptoms and accessing quality medical care.

Lesions of Histoid Hansen's can also be variable from dome-shaped nodules to keloidal plaque. Sometimes other features like sensory impairment and neuritis may be absent (Lupi et al 2006). As in our case 2, no other features of Hansen's disease were seen except nodules and plaques over the face. Although most cases of Histoid Hansen's arise in pre-existing lepromatous leprosy cases, they may occasionally present de novo without any history of treatment. Histological features are striking with the presence of circumscribed nodular lesions with spindle-shaped histiocytes, sub-epidermal grenz zone, and abundant lepra bacilli. The cells are arranged in an intertwining pattern, and some cases may resemble fibrohistiocytic tumors (Gupta et al 2022). In our case-2 subepidermal grenz zone along with infiltration of foamy histiocytes and elongated spindle-shaped cells were seen in a storiform pattern. This case of Histoid Hansen's disease responded well to MB-MDT. Thus, Histoid Hansen's may manifest with diverse clinical presentations, even in the absence of cardinal signs of the disease.

Leprosy is a complex spectrum disease, and its clinical presentation lacks a straightforward, cut-and-dried characterization (Dhattarwal et al 2023). In our case 3, the patient has mainly nodular lesions of lepromatous leprosy along with two patches of the tuberculoid spectrum. The histology of both, patch and nodules is consistent with two different poles of leprosy. This was mainly a case of downgrading tuberculoid leprosy. MB-MDT was given to the patient which showed good improvement in 3 months. Another differential diagnosis Leukaemia cutis may be easily differentiated by the presence of pruritus, and the absence of acid-fast bacilli on SSS. The

challenge in diagnosis lies in the widespread distribution of asymptomatic nodules. Therefore, it is crucial to thoroughly examine the patient under optimal lighting conditions and in full exposure. In our case, the presence of two patches served as a diagnostic clue.

Conclusion

Leprosy can be a 'great mimicker' as seen in the three cases described in this article. All types of Hansen's disease, across the spectrum, may present with atypical presentations. In one of our case reports described above, the patient presented with pustular ENL due to the sudden withdrawal of systemic steroids by a non-registered medical practitioner. Histoid Hansen's can also present in atypical variable forms. The simultaneous presence of lesions of two poles of leprosy is rare but was present in our case. Such unusual presentations of leprosy may not only pose diagnostic challenges but may also lead to delays in treatment, which may cause further damage to nerves and other organs.

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