CASE REPORT

Palmoplantar, Genital and Lip involvement in a de novo case of Histoid leprosy

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Summary Histoid leprosy is a rare expression of mostly lepromatous leprosy, usually arising due to drug-resistant lepra bacilli in patients on long-term dapsone monotherapy or taking irregular and/or inadequate multibacillary multidrug therapy. De novo cases are also being reported. Although its clinical presentation is characteristic, lesions may not always be classical and on rare occasions, it may also involve some atypical sites. Here we are reporting a unique de novo case of histoid leprosy in a 24-year old male presenting with disseminated classical as well as uncommon cutaneous lesions which were also present on palms, soles, upper lip, penile shaft and scrotum. On slit-skin smear and histopathologic evaluation, lesions present over these atypical sites were confirmed to be of histoid variant of lepromatous leprosy. To the best of our knowledge, not a single case of histoid leprosy was reported previously with such a clinical presentation.

Introduction

Histoid leprosy (HL), first described by Herbert Windsor Wade in the Philippines in 1963, is a very uncommon but well recognised variant of multibacillary leprosy (mostly lepromatous), usually seen in patients on dapsone monotherapy or taking irregular therapy and/or inadequate antileprotic medications. The emergence of drug resistance and mutant bacilli is the basis of origin of this variant. Classically, HL presents with sharply defined, firm, hemispherical, dome
shaped, smooth, succulent, shiny, skin coloured or erythematous, papulonodular cutaneous and/or subcutaneous lesions characteristically arising from an apparently normal skin. The face, back, buttocks and extremities are most commonly involved sites. Here we report a de novo case of HL in a 24-year old male presenting with disseminated cutaneous lesions all over the body including palms, soles, upper lip and genitalia.

Case Report

A 24-year old male presented with asymptomatic multiple papulo-nodular and plaque type skin lesions all over the body including lips, genitalia, palm and soles for the last 1 year and diminished sensation in distal extremities for the last 10 months. Initially the lesions started developing as papules over the face, back, proximal extremities and buttocks. Over the next few months, it also appeared on lips, distal extremities and genitalia. In this period the patient noticed the reduction of touch sensation distal to forearm and knee bilaterally. He also noticed ulceration in a few lesions over his forearm and soles. There were no constitutional symptoms. A history of epistaxis and convalescent tender nodular eruptions were absent and he did not give a history of any chronic drug intake including dapsone. Family or other contact history was also not found.

Figure 1. (A) Skin colored, shiny, smooth and umbilicated nodules on upper lip. (B) ulcerated nodules on left forearm. (C) erythematous papules and nodules on palm. (D) plantar nodules. (E) infiltrated left ear.
Cutaneous examination revealed numerous (> 200 in number), non-tender, firm, skin and copper-coloured, shiny, dome shaped, papules and nodules distributed over the face, neck, lower back, buttocks, both upper and lower limbs, palms, soles, penile shaft and scrotum. Around seven to eight shiny nodules were present on his upper lip and a few of them were umbilicated (Figure 1A). Some of the lesions on forearm were ulcerated (Figure 1B). Lesions over the palm and soles were erythematous papules and nodules (Figure 1C) and (Figure 1D). A few plaques were also present over his left thigh and ear (Figure 1E).

A solitary skin coloured nodule was seen in the distal most part of penile shaft and scrotum (Figure 2A) and (Figure 2B).

Characteristically, all these lesions (except infiltrated ear lobes) appeared to be arising from apparently non-infiltrated skin. There was no madarosis (ciliary and superciliary) and the nasal bridge was also not depressed. Temperature, touch and pain sensations were intact over the cutaneous lesions. Glove and stocking type of hypoaesthesia was present. The ulnar, common peroneal, posterior tibial and great auricular nerves were bilaterally thickened, but non-tender.

There was no motor deficit and the patient was in good general condition. Systemic examinations, ophthalmoscopy, chest x-ray and abdominal ultrasound were not remarkable. The laboratory parameters including hemogram, HIV ELISA, VDRL, Hepatitis B and C, urinalysis, liver and renal function tests were within normal limits.

Slit skin smears (SSS) were prepared from the left ear lobe and genital papule and Ziehl-Neelsen (Z-N) staining was done. Both were positive for *Mycobacterium leprae* with a bacterial index of 6+(> 1000 bacilli/oil immersion field) and the morphological index of 70% (Figure 3A). Punch biopsy specimen taken from the erythematous plantar nodule, was sent for histopathologic examination (HPE). On hematoxylin-eosin (H&E) staining, it characteristically demonstrated epidermal atrophy, grenz zone and numerous spindle-shaped non-vacuolated histiocytes arranged in interlacing whorls. Wade-Fite staining further revealed parallel bundles of solid stained, long AFB inside these histiocytes (Figure 3B).
With all such clinical and distinct histopathological features, it was diagnosed as a de novo case of histoid leprosy and patient was started on WHO multibacillary multidrug therapy (MBMDT).

Discussion

Since the first description of HL, many cases have been reported over the time and are still being encountered worldwide. Based on very few observational studies from India, its incidence appears to lie between 2.79–3.60% of total leprosy cases.2 Of note, de novo cases are much rarer accounting to only 12.5% observed by a small retrospective study in India.3

Apart from the classical presentation, few cases of HL have been reported with some atypical morphological pattern such as keloidal, xanthomatous, neural, ulcerative, molluscoid and perforating forms.4–9 In extremely rare instances, sites of involvement can be atypical such as palms, hard palate, nasal mucosa, lips, penis and scrotum.4–9

We were unable to find a single case of HL having affection of three sites; palm and soles, lip and genitals in the same patient previously, as seen in our case. A review of atypical cases of HL with palmoplantar, lip or genital involvement has been presented in this article (Table 1).

Table 1. Previously documented cases of histoid Leprosy with palmoplantar and genito-mucosal involvement

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<th>Atypical Sites</th>
<th>Previously reported cases</th>
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<td>5. Kaliyadan F et al. (2012): Histoid lesion as single shiny erythematous plaque over distal part of penile shaft.</td>
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Additionally, presence of umbilication in some of the labial lesions in our patient is also an atypical feature as of note, there are only a handful of HL cases in which umbilicated cutaneous lesions have been described previously.\textsuperscript{4,9–12}

In view of such atypical cases and to avoid hasty misdiagnosis, a high index of suspicion is necessary. The common clinical differential diagnosis of HL includes neurofibromatosis, xanthomas, multiple eruptive dermatofibroma, lipomatosis, Kaposi’s sarcoma, diffuse cutaneous leishmaniasis, cutaneous metastasis, molluscum contagiosum, and Mycobacterium avium intracellulare lesion in HIV patients.\textsuperscript{13} The classical histoid lesions, sensory involvement with or without nerve thickening, presence of acid fast lepra bacilli on Z-N staining and characteristic HP findings rule out all the above mentioned entities.

Histopathological features of HL includes: presence of thinned out epidermis, subepidermal grenz zone (Unna’s acellular band) and spindle shaped histiocytes packed with AFB and arranged in interlacing bands or whorls in the dermis, often encircled by a pseudcapsule. Some polygonal cells, lymphocytes and plasma may also be present but foamy cells are absent. On Z-N and Wade-Fite staining, lepra bacilli are seen in parallel bundles along the axis of histiocytes, denoted as ‘histoid habitus’. Globi are usually not seen due to the lack of ‘gloea’. The bacilli appear as solid-stained, long, narrow with thin tapering ends.\textsuperscript{13}

The response of HL to the 12-month course of MB-MDT is not always adequate therefore it should preferably be recommended for 2 years.\textsuperscript{14} It has also been observed that addition of ofloxacin or Mycobacteria w vaccine (renamed as Mycobacterium indicus pranii) to the MB-MDT may improve the clearance of bacilli faster.\textsuperscript{14}

Being a potential reservoir of mutant bacilli, having a high bacillary index and a myriad of clinical presentations, early diagnosis, treatment and reporting of such cases is warranted.

References