

Lepromatous leprosy presenting clinically as De-novo Histoid Leprosy

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Abstract Leprosy (Hansen's disease) is a chronic granulomatous disease caused by *Mycobacterium leprae* that can present with cutaneous manifestations and affect peripheral nerves over two major polar forms (lepromatous and tuberculoid) as well as several intermediate forms. We hereby report a case with features suspect for *de novo* Histoid leprosy that was ultimately diagnosed as lepromatous leprosy.

Key words

Histoid leprosy, lepromatous leprosy, chronic granulomatous disease.

Introduction

Leprosy (Hansen's disease) is a chronic granulomatous disease caused by *Mycobacterium leprae* that can present with cutaneous manifestations and affect peripheral nerves over two major polar forms (lepromatous and tuberculoid) as well as several intermediate forms. A rarer variant known as Histoid leprosy exists with specific clinical and histopathological features. Its exact etiology and pathogenesis are not well understood as it may arise after inadequate or irregular treatment with dapsone or multidrug therapy (MDT) or arise *de novo*.¹ Histoid leprosy presents significant challenges to the global disease burden and eradication of leprosy.² We hereby report a case with features suspect for *de novo* Histoid leprosy that was ultimately diagnosed as lepromatous leprosy.

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Case report

A thirty-six-year-old Pakistani male, an agriculturalist by occupation, presented to the Marie Adelaide Leprosy Centre in Saddar, Karachi with complaints of multiple nodules across his body for the past two-and-a-half years. The numerous, smooth, dome-shaped, soft to firm papulo-nodular lesions were distributed across his face, limbs, and trunk (**Figure 1-3**). Loss of sensation across the patient's left hand was noted for the past five to six years and loss of sensation across bilateral feet was noted for the past three years. Additionally, weakness of the left hand was noted, particularly of the deformed fifth finger and wasting of the hypothenar compartment (**Figure 4**). The patient also complained of chronic nasal stuffiness and intermittent epistaxis over the course of three years. Additional findings included bilateral superciliary and ciliary madarosis and bilaterally enlarged ulnar, radial, lateral popliteal, posterior tibial and great auricular nerves. There was no history of fever, pain or long-term drug intake.



Figure 1-3 Scattered, soft to firm papules and nodules across arms, legs and trunk.



Figure 4 Claw-hand deformity of fifth finger of left-hand and hypothenar wasting.

Skin smears were positive for acid-fast bacilli and the patient was diagnosed with multibacillary leprosy. Biopsy was undertaken out of concern for Histoid leprosy. The patient was initiated on anti-leprosy multi-drug therapy (MDT). Histopathology revealed epidermal atrophy, an uninvolved grenz zone, and sheets of foamy histiocytes which allowed for Histoid leprosy to be ruled out for a final diagnosis of lepromatous leprosy (**Figure 5 & 6**). Subsequently the patient was continued on MDT.

Discussion

Histoid leprosy is named because of its distinct histological appearance showing spindle-shaped cells resembling those in a dermatofibroma. This variant of leprosy is more common in males and in India its incidence among leprosy patients is estimated to be between 2.79-3.60%.³ Previous cases of Histoid leprosy have been associated with dapsone resistance, with relapse after dapsone monotherapy or multidrug therapy or without any treatment altogether. Histoid leprosy is characterized clinically by multiple smooth, painless, succulent, firm, cutaneous or subcutaneous nodules, papules and plaques, on otherwise normal skin.⁴ These lesions are classically located on extremities and the trunk with some reports showing pronounced involvement of the face.^{1,5} Our patient matched the clinical phenotype of Histoid leprosy.

Histoid leprosy can mimic dermatofibromas, xanthomas or neurofibromas but can be differentiated based on its unique histopathology. Under the microscope, a leproma or a lesion with a circumscribed nature can be seen often surrounded by a pseudocapsule, along with spindle-shaped histiocytes forming interlacing bands or whorls, and a large number of acid fast bacilli² (**Figure 7a, 7b**).

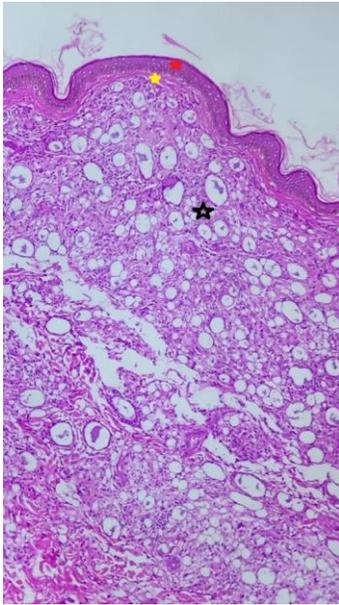


Figure 5 Histopathology with H & E stain revealed atrophic epidermis (red star), grenz zone (yellow star) and diffuse foamy histiocytes in dermis (black star).

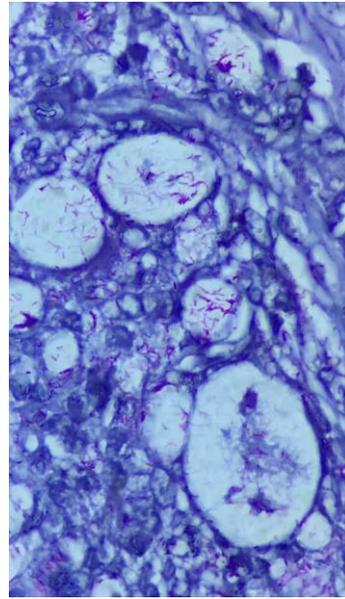


Figure 6 Lepra Stain (Wade-fite staining) showing pink acid-fast bacilli in sheets of foamy histiocytes.

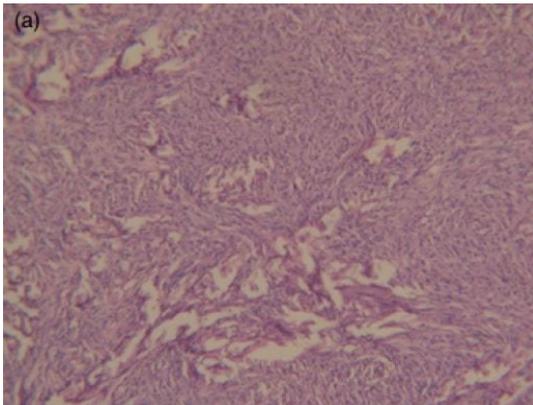


Figure 7a Histopathology with H & E stain of Histoid leprosy reveals fusiform (spindle-shaped) histiocytes in a storiform pattern. (x40)

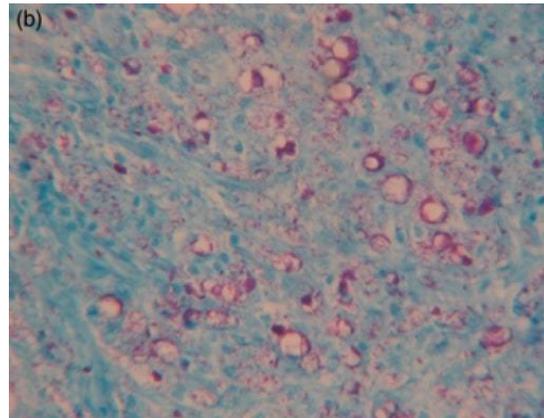


Figure 7b Ziehl-Neelson stain of Histoid leprosy revealing multiple acid-fast bacilli. (x100)

Additionally, epidermal atrophy can also be expressed as a result of dermal expansion of the underlying leproma and a grenz zone immediately below the epidermis.⁵ Our patient's histopathology showed sheets of foamy histiocytes in the dermis, supporting a lepromatous rather than Histoid leprosy diagnosis.

Histoid leprosy presents a significant challenge in the efforts for global eradication of leprosy due to its potential association with multi-drug therapy and dapsone. Histoid leprosy is initially managed with rifampicin 600 mg, oxofloxacin 400mg and minocycline 200 mg once, which is followed by the multibacillary leprosy multi drug therapy of rifampicin, clofazimine and dapsone.⁵

Here we report a unique case of lepromatous leprosy that presented with concern for *de-novo* Histoid leprosy.

References

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