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Case Report

De-novo Histoid Hansen's Disease: Case report with short review of literature

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Abstract Histoid Hansen's disease is a rare form of multibacillary leprosy. It has characteristic clinical and histopathological features. It is considered to be a form of lepromatous leprosy. It can arise in lepromatous patients with inadequate or irregular treatment, who relapse after dapsone monotherapy due to dapsone resistance or may rarely occur 'de-novo'. It may present as diagnostic enigma to the physicians. We present a case of de-novo histoid leprosy in a 20-year-old female.

Key words
Histoid hansen, lepromatous leprosy, multibacillary leprosy, spindle cells.

Introduction

Histoid leprosy is a well-recognized entity, characterized by cutaneous and subcutaneous nodules and plaques present over apparently normal skin, with unique histopathological and characteristic bacteriological morphology.

The term 'histoid leprosy' was first described by Dr. Herbert Windsor Wade in 1960s based on histological concept of bacillary-rich leproma. It is composed of spindle-shaped cells, along with the absence of globus formation.[1] Chronic form of Histoid Hansen's disease exhibits a fibromatoid tendency. [2] We report here a case of histoid leprosy in a 20-year-old female, presenting with multiple asymptomatic nodules as the main symptoms.

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Case Report

A 20-year-old female presented to dermatology outpatient with complaints of multiple asymptomatic nodules and plaques distributed over both forearms, dorsum of hands, chin, forehead and lower legs since 3 months' duration **(Figure 1)**. She had complaints of tingling and numbness over lower limb on and off since 1 year. There was no history of epistaxis.

There was positive family history and contact history with a known case of leprosy. The physical and systemic examination of the patient was all within normal limits. On cutaneous examination, there were multiple hyper-pigmented well-demarcated, shiny, non-tender nodules and plaques, firm to soft in consistency over both forearms, dorsum of hands, chin, forehead and lower legs **(Figure 2)**. Ulnar nerves on both upper limbs and the right common peroneal nerve were thickened and non-tender.

There was no lymphadenopathy, muscle

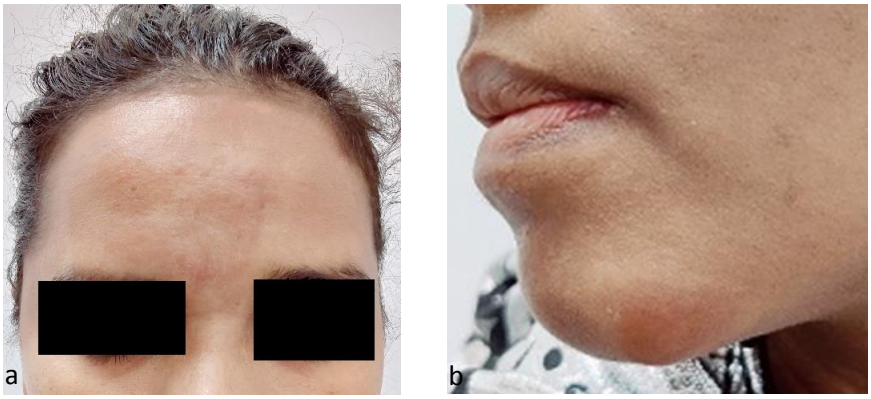


Figure 1 Plaque over forehead and chin.

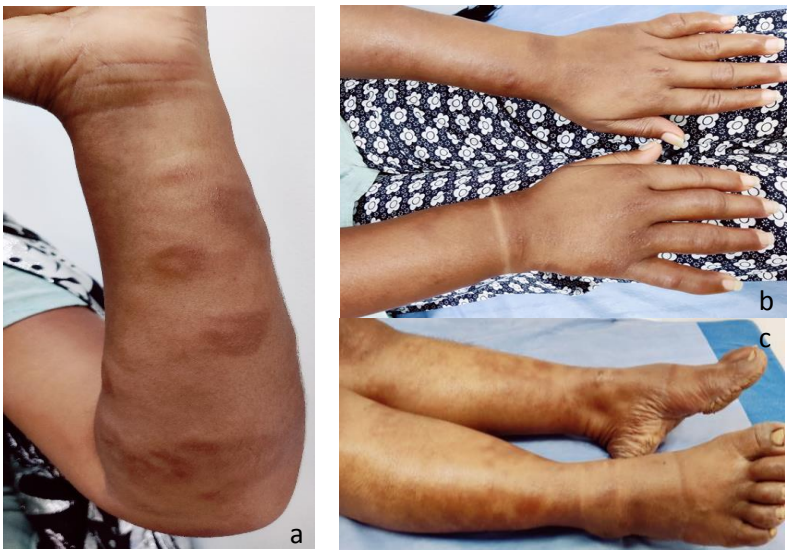


Figure 2 Well-demarcated, shiny nodules and plaques over left forearm, dorsum of both hands and lower legs.

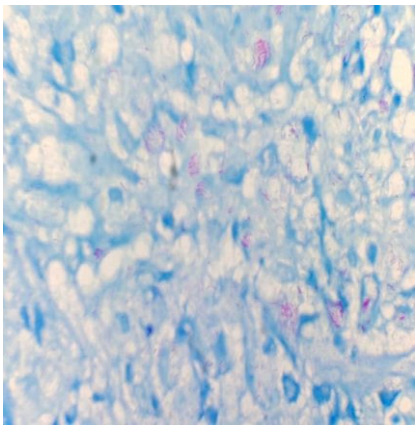


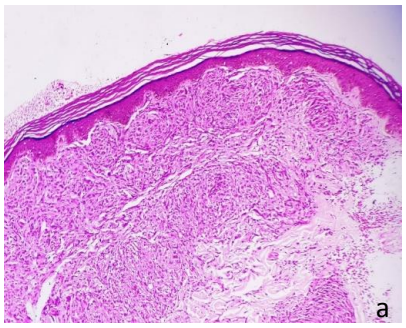
Figure 3 Slit-skin smear from nodule showing solid staining and granular lepra bacilli both singly and in groups (Ziehl–Neelsen, ×100).

weakness or wasting or trophic ulcer observed. Investigation revealed normal blood sugar levels and leukocytosis in blood picture. ELISA for HIV and Rapid Plasma Reagin (RPR) was negative. Clinically a differential diagnosis of histoid leprosy, xanthomas and neurofibroma was kept.

Slit skin smear was done and was stained with Ziehl-Neelsen stain which showed a lot of acid-fast bacilli (AFB) lying singly or in clusters. The AFB appeared as uniform solid stained, long rod shape with tapering terminals, with a bacterial index (BI) of 5+ and morphological index (MI) of >65% **(Figure 3)**. Histopathology of lesion from left forearm shows thinned out epidermis, presence of grenz zone and plenty of vacuolated macrophages, lymphocytes and histiocytes which are spindle shape arranged in storiform pattern in the dermis **(Figure 4 & 5)**. Thus diagnosis of histoid Hansen's disease was confirmed. The patient was put on multibacillary multidrug therapy (MB-MDT) and is in regular follow-up.

Discussion

Wade was first to describe 'Histoid' leprosy as an uncommon form of lepromatous leprosy in 1960.[1] The term 'histoid' is given for the



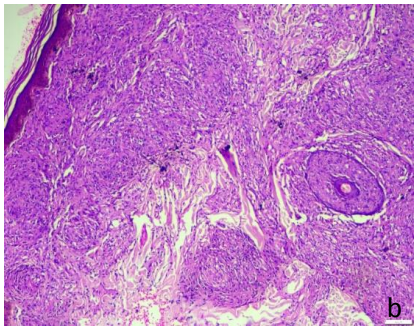


Figure 4 Histopathology of nodule showing grenz zone and dense collection of foamy histiocytes, spindle cells [hematoxylin and eosin (H and E), ×4].

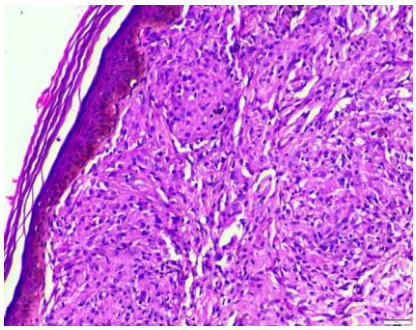


Figure 5 Histopathology of nodule showing dense collection of foamy histiocytes, spindle cells (H and E, ×10).

microscopic picture of the nodule showing spindle-shaped cells which resemble dermatofibroma. Clinically, it is characterized by cutaneous or subcutaneous nodules and papules, which are non-tender, firm, discrete, smooth, globular, skin colored to brown, with apparently normal skin surrounding it.[2] The lesions are usually located on the posterior and lateral aspects of the arms, buttocks, thighs, dorsum of the hands, lower part of the back, and over the bony prominences, particularly over elbows and knees.[3]

Histoid leprosy usually occurs in multibacillary patients who have irregular or inadequate treatment, particularly in patients who are on dapsone monotherapy. Cases which relapse after successful treatment as well as de novo cases have rarely been reported. In general therapy seems to impact the development of histoid leprosy in most cases. Histoid leprosy comprises only 1-2% of all leprosy cases and is noted to be more common in males.[3]

The pathogenesis of histoid leprosy still remains unresolved but it is known that the cell-mediated and humoral immune responses against Mycobacterium leprae in patients with histoid leprosy are enhanced as compared with classical lepromatous leprosy. Though macrophages are adequate they are lacking in functional ability and hence they cannot destroy the bacilli that exists in large numbers in histoid Hansen.[4]

Clinically the differential diagnosis for histoid leprosy includes xanthomas, neurofibroma, dermatofibroma, reticulohistiocytosis, or cutaneous metastasis. Each of them can be differentiated from histoid leprosy on the basis of its characteristic histopathological features, the absence of lepra bacilli on slit skin smear and nerve thickening.[5]

Classical histopathologic findings include epidermal atrophy as a result of dermal expansion by the underlying leproma and univolved band of papillary dermis (Grenz zone) located immediately below the epidermis. The leproma consists of fusiform histiocytes arranged in a whorled, crisscross or storiform pattern containing acid-fast bacilli.[2]

These AFBs are longer as compared with normal bacilli, uniform in length, and are arranged in parallel with the long axis of histiocytes. Tuberculoid granulomas can also be sometimes observed in the histiocytoid collections.

Three histological variants of histoid Hansen are described which are: Pure fusocellular, fusocellular with epitheloid component, and fusocellular with vacuolated cells. Fusocellular pattern with vacuolated cells is most commonly observed.[6]

Treatment for Histoid leprosy is Multi Bacillary Multi-Drug Therapy (MB-MDT) similar to that

in Lepromatous Leprosy.[7]

References

1. Wade H. The histoid variety of lepromatous leprosy. *Int J Lepr*. 1963;**31**:129-42.
2. Sengal VN, Srivastava G. Histoid leprosy A Review. *Int J Dermatol*. 1985;**24**:286-92.
3. Annigeri SR, Metgud SC, Patel JR. Lepromatous leprosy of histoid type: A case Report. *Indian J Med Microbiol*. 2007;**25**:70-1.
4. Manoharan R, Madhu R, Srinivasan MS. Histoid Hansen - A case reprt. *J Indian Soc Teledermatol*. 2008;**2**:12-6.
5. Kontochristopoulos GJ, Aroni K, Panteleos DN, Tosca AD. Immunohistochemistry in histoid leprosy. *Int J Dermatol*. 1995;**34**:777–81.
6. Mendiratta V, Jain A, Chander R, Khan A, Barara M. A nine-year clinico-epidemiological study of Histoid Hansen in India. *J Infect Dev Ctries*. 2011;**5(2)**:128-31.
7. Vora RV, Pilani AP, Mehta MJ, Chaudhari A, Patel N. De-novo histoid hansen cases. *J Glob Infect Dis*. 2014;**6(1)**:19-22.