



Histoid Leprosy-How Is It Different From Lepromatous Leprosy?-A Case Report

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ABSTRACT-

Histoid leprosy, a rare variant of lepromatous leprosy has characteristic clinico- pathological manifestations .We present this case due to their rarity, atypicality of dermal lesions, failure in early diagnosis, and higher bacillary load thereby high lighting the differentiating factors between the two.

Keywords-histoid, eradication, leprosy,dapsone,Ziehl Nelsen

Introduction

Histoid leprosy is an uncommon variant of lepromatous leprosy with characteristic clinical, bacteriological and histopathological findings. It occurs in lepromatous patients who relapse after dapsone monotherapy in presence of dapsone resistance or at times *de novo*.^[1]

CASE REPORT-A 45 years old female presented with asymptomatic raised shiny skin coloured nodules on trunk, back and hands. There was history of dapsone monotherapy 10 years back. Nerve thickening and lymph node enlargement were absent. There was no impairment of pain, touch, or temperature sensation. No history of

epistaxis/insectbite/trauma/long-term drug intake was given. Histopathology revealed focal epidermal atrophy with an underlying cellular band. The dermis revealed a large, circumscribed collection of benign spindle cells resembling fibroblasts, arranged in a whorled and storiform pattern, with entrapped but preserved small dermal nerve and adnexal structures. Ziehl Nelsen (ZN) staining (5% sulphuric acid) revealed abundant, mostly solid, acid fast bacilli (AFB) within the histiocytes and endothelial cells, as well as lying extracellularly. These bacilli were longer than the

normal bacilli, scattered singly as well as arranged in clusters.



Figure- 1- a) Nodules on the dorsum of hand.



Figure 1-b) small nodules on gluteal region extending to thighs.

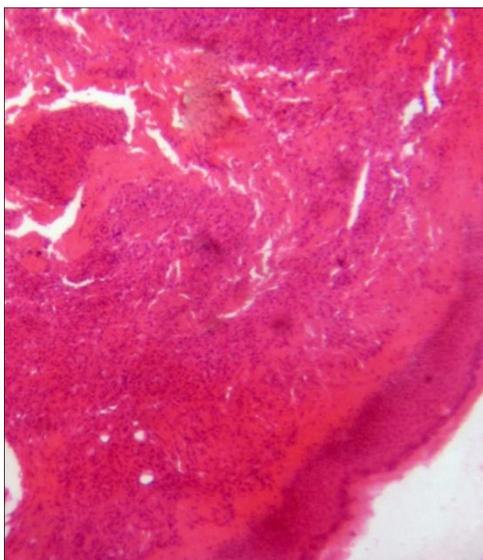


Figure-2-a)epidermal atrophy,underneath is a grenz zone and a histiod nodule mimicking fibrous lesion.(H&EX10)

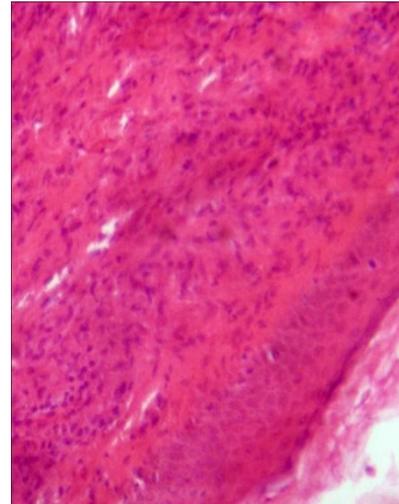


Figure2-(b)-high power view of histiod nodules arranged in whorls and storiform pattern (H&EX400)

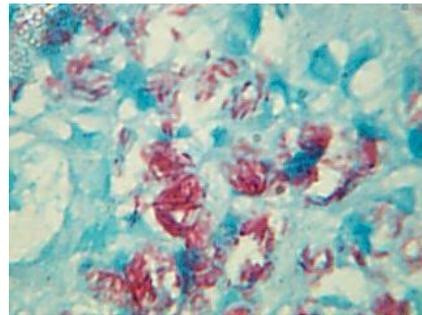


Figure -2-(c)- Myriads of AFB-clusters and singly scattered (Ziehl Neelsen, $\times 1000$).

DISCUSSION- Histoid leprosy is a rare type of lepromatous leprosy which has a prevalence of 2.79% to 3.6% among Indian leprosy patients.^[2] The average age at diagnosis is between 21 and 40 years.^[3] This kind of leprosy is mostly diagnosed in patients who have taken long-term single drug therapy of dapsone.^[4] However, it has also been reported in patients who relapsed even on supervised monthly dose of multidrug therapy and in patients without any treatment ^[5]. Histoid leprosy has specific clinical, histological findings

and bacterial morphology. Dermal lesions include dermal and subdermal, firm, oval or dome-shaped, red or skin-coloured nodules and plaques over an apparently normal skin. These lesions have regular pattern and the overlying skin is shiny and stretched^[4]. Lesions are usually located on the posterior and lateral aspects of arms, buttocks, thighs, dorsum of hands and on the lower part of the back and over the bony prominences, especially over the elbows and knees^[3]. Dermatofibromas, xanthomas, neurofibromas, reticulohistiocytosis, and cutaneous metastasis are the differential diagnosis. It may even resemble acute sarcoidosis, keloid, molluscum contagiosum, mycobacterial spindle cell pseudotumor, and papulonodular variant of secondary syphilis sparing the palms and soles.^[6,7,8] Slit skin smear from histoid lesions shows abundant acid fast bacilli occurring in clusters, singly or tightly, packed in macrophages. The bacilli appear long with tapering ends, when compared to ordinary lepra bacilli. Bacteriological index may be 5+ to 6+ and morphological index may be very high too.^[9] Histopathology is unique in histoid leprosy. Classical histopathological findings include epidermal atrophy as a result of dermal expansion by the underlying leproma and an acellular band (Unna band) located immediately below the epidermis. This dermal expansion of histiocytes pushes aside the dermal collagen resulting in the formation of pseudo capsule. The leproma consists of fusiform histiocytes arranged in a whorled, criss-cross or storiform pattern. These histiocytes resemble fibroblasts and it is suggested that these fibroblast

-like macrophages may have arisen from tissue histiocytes rather than from blood monocytes. There are three histological variants of Histoid Hansen's namely pure fusocellular, fusocellular with epithelioid component, and fusocellular with vacuolated cells. The third pattern is most commonly observed. Differential diagnosis of histopathology of Histoid Hansen's includes dermatofibromas and neurofibromas and fibrohistiocytoma.^[8] This case is a classical presentation of histoid Hansen's, occurring after dapsone monotherapy. The slit skin smear and histopathological examination confirmed the diagnosis of histoid Hansen's. The pathogenesis of this rare and unusual variant of leprosy still remains unresolved. Although histoid leprosy is considered to be a variant of lepromatous leprosy there exists an enhanced immune response against *Mycobacterium Leprae* in these patients compared with lepromatous leprosy with respect to both cell mediated immunity and humoral immunity. Despite the presence of adequate number of macrophages it has been claimed that they lack the functional property to kill bacilli that exists in high numbers in histoid.^[11] Management is entirely different from lepromatous leprosy. Rifampicin 600 mg, Oloxacin 400 mg and Minocycline 200 mg is followed by WHO MBMDT therapy for 2 years.^[3] The present case is a classical presentation of histoid Hansen's, occurring after dapsone monotherapy. The slit skin smear and histopathological examination confirmed the diagnosis of histoid Hansen's. The patient is responding to the treatment, till 6-month followup.

CONCLUSION-

It is very important to diagnose histoid leprosy because of its rarity, different histopathological findings and treatment. Also they carry very high bacillary load, so forming a barrier in the eradication of leprosy.

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