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Case Report

A case of 'de novo' Histoid Hansen's disease

Lt Col Vikas Pathania^{a,*}, Col Ajay Kumar^b, Lt Col S.A. Hashmi^c,
Lt Col S.P.S. Shergill^c

^a Graded Specialist (Dermatology), Military Hospital Ambala, Ambala Cantt 133001, India

^b Classified Specialist (Dermatology), Military Hospital Ambala, Ambala Cantt 133001, India

^c Graded Specialist (Pathology), Military Hospital Ambala, Ambala Cantt 133001, India

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Introduction

Histoid Hansen's disease is an uncommon but established clinico-pathological entity in the post global leprosy elimination era, presenting as relapse following dapsone monotherapy and incomplete/irregular therapies or as resistance to dapsone. However, a high degree of suspicion is warranted in 'de novo' cases. The term 'Histoid' was originally coined by Wade in 1960 to describe the characteristic histopathology consisting of well circumscribed spindle-shaped cells with abundance of solid staining acid fast bacilli.¹ Clinically, it is characterized by discreet shiny, protuberant, firm nodules and plaques on normal appearing skin. We report a case of an elderly male presenting with 'de novo' Histoid Hansen's disease, with no history of leprosy or exposure to dapsone/Multi drug therapy, with characteristic clinical features and confirmatory histopathology.

Case report

A 50 years old male patient presented at the Dermatology outpatient department with history of multiple skin coloured and few red raised asymptomatic lesions over the trunk and all four extremities of 2 months duration. The onset was spontaneous and course progressive. There was no history suggestive of Leprea reactions, as also drug intake before onset of these lesions. There was no family history or history of close contact with a known case of Hansen's disease. Systemic and eye and mucosal examination was normal. Dermatological examination revealed widespread involvement of the trunk and extremities (Fig. 1), in the form of multiple, discrete, skin coloured, nodules and plaques, some of which showed overlying black adherent eschars (Fig. 2). Most nodules and plaques were cutaneous, few were subcutaneous, about 50 in number and distributed predominantly over the flanks, back and extensors of all four extremities. There were no anaesthetic/hypoesthetic patches or thickened/tender peripheral nerves. Histopathology from an intact nodule over the back revealed a stretched epidermis, over a thin Grenz zone and with well circumscribed nodular dermal infiltrates of spindle-shaped histiocytes arranged in whorls and bundles (Fig. 3). Ziehl–Neelsen stain revealed many slender, uniformly stained acid fast bacilli lying discretely and in clumps within these lesions (Fig. 4). Slit skin smears from the lesions were positive with bacterial index of 4+ (10–100 bacilli in an average oil immersion field). He was managed with Multi-bacillary MDT (Cap Rifampicin 600 mg and Cap Clofazimine 300 mg monthly supervised with Tab Dapsone 100 mg and Cap Clofazimine 50 mg daily) for one year. Regular follow-up showed satisfactory resolution of lesions after 6 months.

* Corresponding author. Tel.: +91 9812364747 (mobile).

E-mail address: vikascongo@gmail.com (V. Pathania).

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Fig. 1 – Multiple, cutaneous and subcutaneous, nodules and plaques.

Discussion

Histoid Hansen's disease is a rare lepromatous form of leprosy with reported incidence of 1.2% of all leprosy patients and 8.7% of Lepromatous leprosy by Singh et al.² Another study by Kaur et al showed the incidence of Histoid leprosy as 1.8%, of

Fig. 2 – Some of the nodules and plaques showing adherent eschars.

Fig. 3 – H&E section (10×) shows nodular dermal infiltrates of spindle-shaped cells arranged in whorls and bundles with a thin Grenz zone.

which 'de novo' Histoid lesions, i.e. lesions of Histoid leprosy developing without evidence of lesions of other types of leprosy, appeared in only 12.5% of all patients with Histoid leprosy.³ 'De novo' occurrence has been also reported sporadically elsewhere in literature.^{4,5} However, classically the condition has been described in relapse or resistant cases of Hansen's disease who were exhibited either Dapsone monotherapy or incomplete/irregular Multi drug therapy. They have been seen in patients from 10 to 84 yrs of age.⁶

Fig. 4 – Ziehl–Neelsen stain revealed numerous slender, uniformly stained acid fast bacilli lying discretely and in clumps.

There is a male preponderance with a male/female ratio of 5.7:1.³

The number of lesions is variable ranging from 3 to 50 in a single patient. Lesions are usually located over face, back, buttocks, extremities and bony prominences like elbows and knees, where they may occur discretely or in groups of protuberant skin coloured papules, nodules and plaques. As in our case, ulceration within these lesions has also been reported by various workers.⁶ In severe cases, mucosal involvement has also been reported.⁷ Other rare presentations of these lesions have been described including erythema nodosum leprosum, xanthomatous and umbilicated molluscum contagiosum like lesions.³

Histologically, several characteristic features have been described of which the hallmark remains the occurrence of well circumscribed elongated spindle-shaped histiocytes in sheets, whorls and storiform patterns. Lesions are usually located in the dermis and/or subcutis with a thin clear 'grenz' zone separating them from the epidermis. However no such clear zone was noted in our case. A pseudocapsule is often seen surrounding the lesion, formed by compression of adjacent tissue secondary to expansile nature of the lesion. Slit skin smears taken from these lesions always show abundance of acid fast bacilli which are uniformly stained, longer, slender and with tapering ends when compared to regular lepra bacilli. Clinically, differential diagnosis include neurofibromatosis, xanthomas, cutaneous sarcoidosis, post kala azar dermal leishmaniasis and molluscum contagiosum, while histologically, dermatofibroma, nodular subepidermal fibrosis and similar disorders may mimic Histoid Hansen's disease.⁷ In this case a differential diagnosis of dermatofibroma and lepromatous leprosy were considered. However, absence of thick collagen bundles interspersed between spindle-shaped cells and presence of acid fast bacilli within these lesions delineate the condition from dermatofibroma, while an exclusive collection of spindle-shaped Histoid cells in a storiform pattern with almost no lymphocytes and a very thin grenz zone, differentiate it from lepromatous leprosy. Treatment remains the same as for multi-bacillary leprosy as per current recommendations of the World Health Organization (WHO).⁸

Despite Wade making the retrospective diagnosis of this form of leprosy based on its unique histopathology in 1960,¹ the condition remained elusive with some workers reporting it as an entity by itself while others considered it to be a part of the lepromatous spectrum. Sehgal et al after their

extensive experience with these patients, formulated several prospective guidelines for delineation of the condition as distinct entity independent of Lepromatous leprosy in 1985.⁷ It has been speculated that focal defects in immunity as a consequence of partial elimination of bacilli due to selective drug resistance to dapsone results in these mutant organisms growing into resistant 'Histoid bacilli' in these patients and forming discrete circumscribed lesions of flattened histiocytes.⁶ It has been further postulated that 'de novo' cases may present in setting of depressed systemic cellular immunity, but enhanced local cellular and systemic humoral immunity.⁹ The case highlights the fact that in the post leprosy elimination era, a high clinical index of suspicion is solicited from the modern-day physician for diagnosing these emerging rare but unique variants of leprosy.

Conflicts of interest

All authors have none to declare.

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