

CASE REPORT

Denovo Histoid Hansen's Disease - "Bolt from the Blue"

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Abstract:

Histoid Hansen's is a rare form of multibacillary leprosy with distinct clinical and histopathological features. Histoid Hansen's is an uncommon variant of lepromatous leprosy that usually follows treatment failure. Occasionally it occurs de novo without any history of previous treatment. We report a case of 16 year old boy with multiple nodules on neck, chin and areolae with diffuse infiltration of both lips. Histopathology revealed Histoid Hansen's with typical spindle shaped histiocytes and bacillary index of 5+. Patient was started on WHO's multidrug therapy (MDT) for 12 months. A high index of suspicion is necessary especially when patients present with unusual skin lesions and estimation of bacterial index is highly indicated as proven in our case.

Key words: Histoid Hansen's, lepromatous leprosy, spindle shaped histiocytes

Introduction:

Histoid leprosy is an uncommon variant of lepromatous leprosy with characteristic clinical, immunologic, and bacteriologic findings. It occurs in lepromatous patients who relapse after dapsone monotherapy in presence of dapsone resistance or at times denovo.

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Case report:

A 16 year old male patient presented with asymptomatic, multiple, raised lesions over chin, neck and areolae since 2 years. Patient was apparently normal 2 years back, later he developed multiple, asymptomatic, raised lesions over chin, neck and areolae. These lesions were initially of pin head sized which gradually progressed to attain the present size.

History of asymptomatic diffuse swelling of lips since 6 months which progressed gradually. H/o decreased sensation over both feet and hands since 6 months which was gradually progressive. H/o nasal stuffiness and bleeding from nose (2 episodes) since 3 months. H/o recurrent, spontaneous blistering over fingers since 2 months.

There was no h/o slippage of foot wear, joint pains, myalgia, hoarseness of voice, watering/ redness of eyes, any light or red colored raised lesions or painful skin lesions over the body.

Cutaneous examination: Multiple, non-tender, skin colored, shiny, dome-shaped, poly-sized nodules (smaller-0.5x0.5 cm; larger-2x2 cm) noted over mental region, midline of neck and areolae (figure 1 & 2). Diffuse infiltration of both upper and lower lips was seen (figure 3). Multiple healed ulcers noted over ring, little finger (healing stage) and palm of left hand (figure 4). Xerosis seen on both lower limbs (figure 5).

Figure 1: Clinical photograph showing multiple, well defined, polysized, skin colored dome shaped nodules on chin and neck.

Figure 1



Peripheral nerve examination showed multiple nerve thickening which included bilateral supratrochlear, ulnar, radial cutaneous, lateral popliteal nerves and left greater auricular nerve (figure 6). Sensory examination showed glove and stocking type of anaesthesia noted. Motor examination showed wasting of thenar and hypothenar muscles of both hands with power 4+. Cranial nerve examination was normal.

Figure 2: Clinical photograph showing nodular infiltration of both areolae.

Figure 2



Figure 3: Clinical photograph showing diffuse infiltration of both lips.

Figure 3



Figure 4: Clinical photograph showing multiple ulcers at various stages of healing on the left hand.

Figure 4



Figure 5: Clinical photograph showing xerosis of both the legs.

Figure 5



Figure 6: Clinical photograph showing enlargement of the left greater auricular nerve.

Figure 6



Mucous membranes: Oral, genital and conjunctiva was normal;

Hair and Nails: normal. Systemic examination was unremarkable.

Based on history and clinical examination, a provisional diagnosis of HISTOID HANSEN'S was considered and subjected to necessary investigations.

Routine hemogram and other biochemical investigations were within normal limits.

Erythrocyte Sedimentation Rate – 50 mm/1st hr; G6PD – 9.49 U/g/Hb. Serology was negative.

Slit skin smear (SSS) - Bacteriological index (BI) – 5+; Morphological index (MI) – 96%

Skin biopsy - Two punch biopsy specimens (A & B) of size 5x5 mm were taken from lesional area over neck and left areola.

Both sections (A & B) showed thinned out epidermis with sub-epidermal Grenz zone. Dermis showed spindle shaped histiocytes, few polygonal cells arranged in intertwining pattern, admixed with occasional inflammatory cells (figure 7a, 7b). Fite-Faraco stain showed dense collection of acid fast bacilli with AFB- 5+ (figure 8).

Figure 7: Photomicrographs showing thinned out epidermis with sub-epidermal Grenz zone. Dermis shows spindle shaped histiocytes; 7a: (10x), 7b: (40x).

Figure 7 a

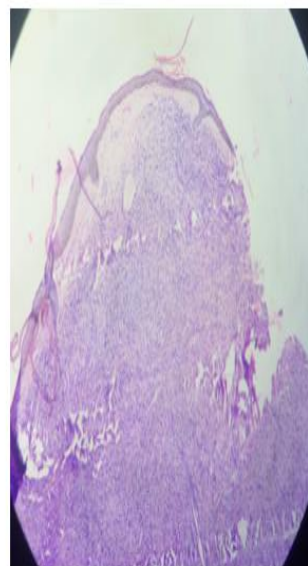


Figure 7 b

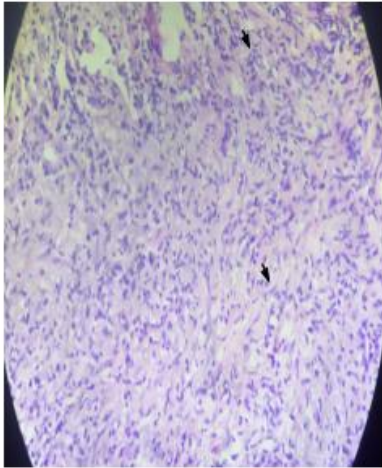
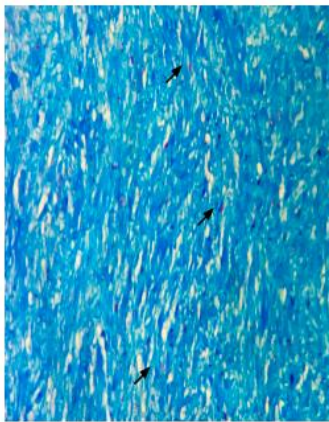


Figure 8: Photomicrograph showing dense collection of acid fast bacilli with AFB – 5+ in Fite-Faraco stain.

Figure 8



Based on history, cutaneous examination and histopathology, the diagnosis of Histoid Hansen's was confirmed.

Treatment:

Patient was started on WHO MB-MDT for 12 months with Rifampicin – 600 mg monthly supervised dose, Dapsone – 100 mg daily self-administered and Clofazimine – 300 mg monthly supervised dose and 50 mg daily self-administered

.Patient was advised about care of hands and feet.

Discussion:

Histoid leprosy is an uncommon variant of lepromatous leprosy and it was first described by Wade in 1960. Its incidence among leprosy patients in India is estimated to be 2.79 to 3.60%¹. There is a male preponderance with average age at diagnosis between 21 and 40 years¹. The incidence of Histoid leprosy is found to be 8.7% among lepromatous leprosy cases².

Clinically, Histoid leprosy is characterized by cutaneous and/or subcutaneous nodules or papules, which are usually painless, discrete, firm, smooth, globular and skin colored to yellowish brown, with normal appearing skin surrounding it. The lesions are usually found in the posterior and lateral aspects of the arms, thighs, buttocks, lower part of the back, dorsum of the hands, and over the bony prominences, especially over elbows and knees. Ears may be unaffected. Histoid lesions have also been reported to be present along the course of the peripheral nerve trunks and cutaneous nerves.³

Histoid leprosy clinically simulates dermatofibromas, xanthomas, reticulohistiocytosis, neurofibroma or cutaneous metastasis. They can be differentiated from Histoid leprosy on the basis of their characteristic histopathology, the absence of the acid fast lepra bacilli on slit skin smear and absence of nerve thickening.³

Slit skin smear from the lesions in Histoid leprosy show abundant acid fast bacilli in singles, clusters, or tightly packed in macrophages (globi). These bacilli tend to be longer with tapering ends when compared to the ordinary lepra bacilli. The bacillary index may be 4+ to 6+ and the morphological index may also be very high.⁴

Classical histopathological findings include epidermal atrophy as a result of dermal

expansion of the underlying leproma and a Grenz zone (band of Unna) located immediately below the epidermis. The macrophage reaction is unusual in that the cells frequently become spindle shaped or fusiform and oriented in a storiform or a whorled pattern. These histiocytes usually resemble fibroblasts and it has been suggested that fibroblast-like macrophages may have arisen from the tissue histiocytes rather than blood monocytes.⁴ Within these histiocytes, bacilli are arranged in groups or parallel bundles aligned along the long axis of the cell, described as histoid habitus. There are three histological variants of Histoid Hansen's, namely, pure fusocellular, fusocellular with epitheloid component, and fusocellular with vacuolated cells.⁴

The Histoid leprosy patients are usually treated with the World Health Organisation (WHO) multibacillary multidrug therapy for one year.¹

Conclusion:

This case has been reported to create awareness and to emphasize unusual clinical presentation of Histoid Hansen's. Due to high bacillary load, they can form potential reservoir for infection. Therefore, early diagnosis and complete treatment are necessary to achieve our goal of elimination of leprosy.

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