

De-novo Histoid Leprosy

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ABSTRACT

Histoid leprosy is a rare form of multibacillary leprosy with distinct clinical and histopathological features. It is a variant of lepromatous leprosy. It occurs in lepromatous patients, who relapse after dapsone monotherapy, in the presence of dapsone resistance or at times '*de-novo*'. We describe here a case of histoid leprosy, clinically mimicking neurofibromatosis.

Keywords: Fine needle aspiration cytology, histoid Hansen, lepromatous leprosy

INTRODUCTION

Histoid leprosy is a well-recognized entity, characterized by cutaneous and / or subcutaneous nodules and plaques present over apparently normal skin, with unique histopathological and characteristic bacterial morphology. The term 'histoid leprosy' was originally coined by Wade as a histological concept of bacillary-rich leproma composed of spindle-shaped cells, along with the absence of globus formation (so conspicuous in ordinary leproma). It exhibits a fibromatoid tendency in the chronic form.^[1] Since then, there have been many reports, with variable findings.^[2] We report here a case of histoid leprosy in a 45-year-old man, presenting with itching of the skin and multiple nodules as the main symptoms.

CASE REPORT

A 45-year-old man, an agriculturist by occupation, complained of multiple nodules all over the body, associated with itching over the skin since one year. The nodules were distributed on his low

back, buttocks, arms, and abdomen. There was no history of fever, pain, epistaxis, pedal edema, altered sensations or long-term drug intake for the same. Family history was non-contributory. On clinical examination, there were multiple nontender nodules, firm-to-soft in consistency, measuring 0.5 cm to 2 cm. Papules were also present over the extensor surface of the arm [Figure 1], abdomen, lower back, and buttocks [Figures 2a and 2b]. There was no impairment of pain, touch or temperature sensation. Nerve thickening and lymph node enlargement were also absent. A clinical diagnosis of neurofibroma / xanthoma was made.

Routine investigations were within normal limits. Fine needle aspiration cytology (FNAC) of the back and arm nodule showed round-to-elongated, benign, spindle cells, with cytoplasmic vacuolation. The Ziehl-Nielsen (ZN) stain showed numerous acid-fast bacilli (AFB). The slit skin smear revealed plenty of acid-fast bacilli, with a bacterial index of 4+. Histopathology of the lower back nodule showed atrophic epidermis with a subepidermal grenz zone. The dermis showed sheets of round-to-spindle-shaped histiocytes compressing the skin appendages. These spindle cells were arranged in whorled, criss-cross / storiform patterns [Figure 3]. The nuclei were pyknotic and the cytoplasm foamy and vacuolated. The Fite-Faraco stain showed numerous uniformly stained bacilli arranged in singles and in clumps [Figure 4] with a bacterial index of 4+. A final diagnosis of '*de-novo*' histoid leprosy was made. The

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Figure 1: Photograph showing multiple nodules on the arm



Figure 2a: Photograph showing multiple nodules on the lower back



Figures 2b: Photograph showing multiple nodules on the buttock

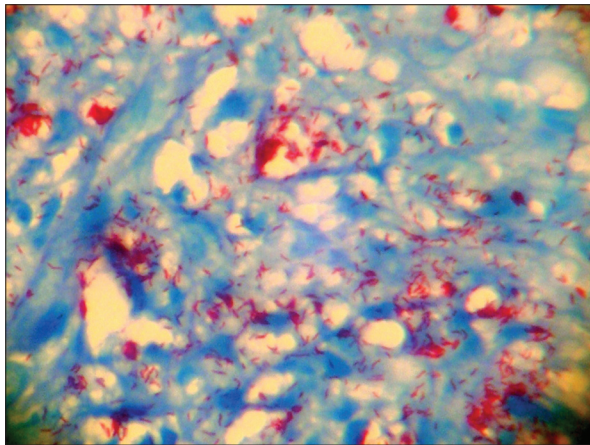


Figure 4: Photomicrograph showing acid-fast bacilli in singles and in clumps (Modified Fite-Faraco stain, $\times 1000$)

patient has been put on multibacillary multidrug therapy (MBMDT) and is on regular follow-up.

DISCUSSION

Histoid leprosy is not uncommon in South India. It is considered by some as a variant of lepromatous leprosy and

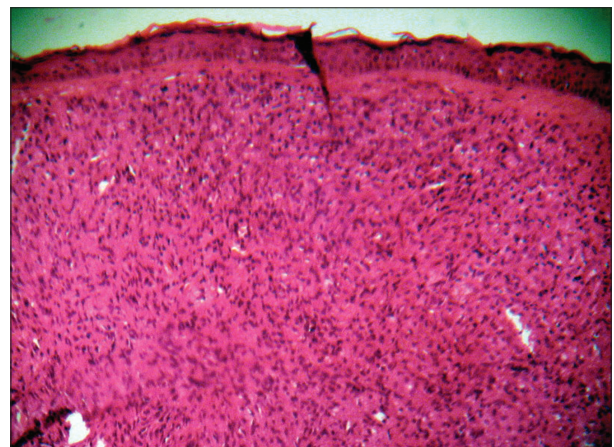


Figure 3: Photomicrograph showing epidermal atrophy and grenz zone. The dermis is replaced by spindle-shaped histiocytes arranged in interlacing bundles, whorls or crisscross patterns (H and E, $\times 100$)

by others as a distinct clinical entity. In India, its incidence among leprosy patients is estimated to be 2.79 to 3.60%. There is a male preponderance, and the average age at diagnosis is between 21 and 40 years.^[2] The incidence of histoid leprosy was found by Singh *et al.* to be 8.7% among lepromatous leprosy and 1.2% among all leprosy patients. Rodriguez found that, of 72 relapsed patients, 28 [39%] developed histoid lesions, which occurred much more frequently in men than in women. They have been reported in patients of age 10 – 84 years.^[1]

Clinically it is characterized by cutaneous and or subcutaneous nodules and papules, which are painless, firm, discrete, smooth, globular, skin-colored to yellowish-brown, with apparently normal skin surrounding it. 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, especially over elbows and knees.^[2] Ears may be unaffected. Histoid lesions have also been reported to present along the course of the peripheral nerve trunks and cutaneous nerves.^[3]

Erythema nodosum leprosum rarely occurs in histoid Hansen.^[3] Bhutani *et al.*, in 1974, found classical ENL reaction in three of his 20 patients of histoid leprosy, which occurred during sulfone therapy.^[4] Histoid leprosy clinically simulates xanthomas, neurofibroma, dermatofibroma, reticulohistiocytosis or cutaneous metastasis. Each of them can be differentiated from the histoid leprosy on the basis of its characteristic histopathology, the absence of lepra bacilli on slit skin smear and nerve thickening.^[3]

Slit skin smear from the histoid lesions show abundant acid-fast bacilli in clusters, singles or tightly packed in macrophages. These bacilli appear longer with tapering ends when compared to the ordinary lepra bacilli. The bacillary index may be 5+ to 6+ and the morphological index may be very high too. The enormous bacillary population in histoid lesions is suggested to be due to focal loss of immunity.^[3]

Classical histopathological findings include epidermal atrophy as a result of dermal expansion of the underlying leproma and a grenz zone located immediately below the epidermis.^[1] The lesion consists of fusiform histiocytes arranged in a whorled, criss-cross or storiform pattern. These histiocytes resemble fibroblasts and it is suggested that fibroblast-like macrophages may have arisen from the tissue histiocytes rather than blood monocytes.^[5] Within these histiocytes, an abundance of acid-fast bacilli can be seen.^[5]

The AFB are not found in globi formation, as they do not secrete any glial substance. They are longer than the normal bacilli, uniform in length, and are arranged in parallel bundles along the long axis of histiocytes. Within the histiocytoïd collections, there can also be islands of tuberculoid granulomas, which are called by Wade as, 'contaminating tuberculoid bacilli'.^[3] The Fernandez-Mitsuda reaction is negative.^[3]

Histoid leprosy is managed by initially giving the range of motion therapy, with Rifampicin 600 mg, Ofloxacin 400

mg, and Minocycline 200 mg, which is followed by WHO MBMDT therapy.^[2]

Fite's stain from FNAC is also positive in case of histoid leprosy, which must stimulate interest for further studies. FNAC is a simple cost-effective method of investigation that is more useful in tuberculoid, lepromatous, and histoid leprosy patients.^[6] In our case, a correct cytological diagnosis was possible on the basis of a positive Fite-Faraco stain. This case highlights the importance of FNAC in all similar cases where spindle cell nodular lesions of the skin are encountered. It was good to carry out a Fite-Faraco stain for ruling out or confirming the diagnosis, especially as there was no clinical suspicion in our case.

CONCLUSION

In this phase of the National Program of Leprosy Eradication, a high index of suspicion is essential to continue the surveillance for new and relapse cases, rather than to wait for voluntary reporting. Early diagnosis and complete treatment is very important to achieve our goal of elimination of leprosy.

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