

## De novo' Histoid Leprosy in a Dwarf with Lesions at Unusual Sites

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Histoid leprosy is an uncommon variant of lepromatous leprosy with characteristic clinical, bacteriological, immunological and histological findings (Jopling and McDougall 1966). Clinically, it is characterized by multiple, smooth, shiny, dome shaped, non tender, discrete, firm, waxy, yellowish papules and nodules over normal looking skin of lepromatous patients on dapsone monotherapy or received inadequate as well as irregular treatment and rarely it may present de novo (Bopp and Bakos 1975, Kaur et al 2009). It was initially reported by Wade in 1963 from Philippines (Wade 1963). Later, sporadic episodes of this unusual variant of leprosy have been published occasionally from different parts of the world, mostly from India (Sehgal and Srivastava 1985, Mendiratta et al 2011, Vasavi and Reddy 2012).

Herein, we report an interesting episode of *denovo* histoid leprosy in a 16 years old male dwarf with lesions occurring at a typical sites for its clinical interest. A 16 years old male patient presented with the complaints of asymptomatic, skin colored, shiny papules and nodules of



Fig 1A : Involvement of Chin

appearing over face, trunk, back and extremities for preceding one month duration.

Dermatological examination of the patient

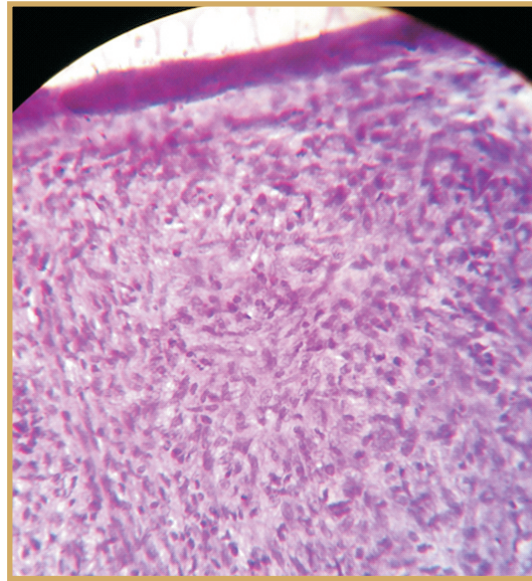
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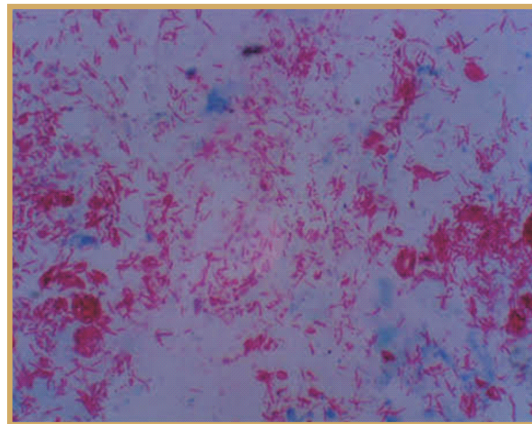
**Fig 1B : Involvement of ear lobe**



**Fig 2 : Histological findings in lesional biopsy showing histoid characteristics**



**Fig 1C : Involvement of back and upper limb**



**Fig 3 : AFB in the lesion**

revealed multiple, waxy, non tender, shiny, dome shaped, yellowish infiltrated papules, nodules and plaques of varying sizes and shape distributed symmetrically over face, ear lobes, chin, trunk, back, extremities and over genitalia. (Figure 1 (A,B,C)). Sensations over the lesions were not impaired but peripheral nerves are bilaterally

thickened and non tender.

Routine hematological and biochemical investigations were within normal limit. The slit skin smears from nodule on the ear lobe on AFB staining revealed plenty of solid, pink, acid fast bacilli with a bacillary index of 6+. (Figure 3).

Histopathological examination of a nodule over the back on H & E staining showed atrophy of the epidermis and a clear grenzzone and diffuse collection of foamy macrophages and spindle shaped histiocytic granulomas with heavy bacillary load in the dermis. (Figure 2). The Fite-Faraco stain showed numerous uniformly stained acid-fast bacilli arranged in clumps (Figure 3). The patient was diagnosed as a case of histoid leprosy and advised multibacillary multidrug therapy (MBMDT) and asked to come for follow-up.

His development of milestones and IQ to the age except delay in the onset of puberty. No history of consanguinity in the family. He had not taken any medication nor given history suggestive of chronic disease like tuberculosis or leprosy, sensory or motor disturbances and as well episodes of reactions in the past. Family history was not contributory except that his only brother who is younger to him having normal stature with no evidence of leprosy. The patient is a dwarf with short stature (height 107 cms and weight of 17 Kgs,) and his X-rays of wrist and elbow showed incomplete fusion epiphysis process and non formation of pisiform bone suggestive of decreased velocity of bone growth (Figure 4).



**Fig 4 : X-ray of hands of dwarf patient**

Exact etiopathogenesis of histoid leprosy is not well understood but because of the fact that it may arise *de novo* or may develop after inadequate and /or irregular treatment with dapsone, it is believed that the interplay of genetic factors, immune response and treatment received by the patient may influence the manifestations of the disease (Bopp and Bakos 1975, Kaur et al 2009)

However, we were intrigued to note that the disease manifested '*de novo*' for the first time with no previous episode of leprosy or history of taking anti-leprosy treatment or evidence of leprosy in the family. Histoid leprosy occurring *de novo* was also reported in lepromatous leprosy patients occasionally (Vasavi and Reddy 2012, Murthy et al 2011).

Association of dwarfism and histoid leprosy in our patient may be simply a chance factor and may not have any etiological significance. Our patient had demonstrated classical histoid leprosy clinical features. However, it will be of interest to consider the impact of hormonal factors and immunological imbalance (s) if any.

The lesions in histoid leprosy 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 (Annigeri et al 2007). In contrast to this, our patient presented lesions over the face, trunk, back, both upper limbs and legs. This variation is of clinical interest. Erythema nodosum leprosum may occur rarely in histoid leprosy (Vasavi and Reddy 2012, Hastings and Opromola 1994).

Classical histopathological findings in histoid leprosy similar to the ones seen in our patient include epidermal atrophy as a result of dermal expansion by the underlying leproma, a grenzzone and acellular bands located immediately

below the epidermis. The leproma consists of fusiform histiocytes arranged in a whorl or criss-cross pattern with abundant acid fast bacilli occurring in clusters or singly. The bacilli appear longer with tapering ends, when compared to ordinary lepra bacilli, usually arranged parallel to the long axis of cells (Hastings and Opromola 1994).

Histoid leprosy clinically simulates xanthomas, neurofibroma, dermatofibroma, reticulohistocytosis, or cutaneous metastasis, molluscum contagiosum and papulonodular variant of secondary syphilis sparing the palms and soles.

Histoid leprosy is an unusual distinct form of multibacillary leprosy and it requires early detection and prompt drug therapy. It would thus be relevant to be aware of all possible variations in the clinical presentation of histoid form of leprosy such as seen in site distribution observed in this study.

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