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Case Report

De-Novo Presentation of Histoid Leprosy on an Unusual Site

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ABSTRACT

Histoid Leprosy (HL) is a rare variant of Lepromatous Leprosy, occurring in long-standing cases, mostly in a background of acquired drug resistance. Patients usually present with sudden onset multiple nodules and plaques, most often involving the skin and subcutaneous tissue of trunk and lower limbs. Here we report an unusual case of de novo (without any history of prior anti-leprotic therapy) HL, arising as multiple nodules in the face. The histopathology of the lesion showed collection of spindled macrophages in the dermis, oriented in a storiform-like pattern. A possibility of HL was considered and the diagnosis was confirmed by performing a modified ZN stain (Wade-Fite stain) on the biopsy material, which revealed the presence of acid-fast bacilli (AFB) with a bacillary index of 6. Then the patient was put on multibacillary multi-drug chemotherapy and was thereby cured.

Keywords: **Lepromatous Leprosy, Histopathology, India**

Introduction

Histoid Leprosy (HL) is a rare, but well-defined entity with specific clinical, histopathological and bacteriological features. It was originally described by Wade in 1963 as discrete, firm nodules developing on apparently normal skin in patients with lepromatous leprosy (LL) (1). Further observation revealed that it usually develops after an inadequate and irregular treatment with dapsone monotherapy or multidrug therapy. Patients usually present with sudden onset multiple nodules, involving the trunk and lower limbs. The incidence has been reported to vary from 1-2% among total leprosy patients (2). Histoid Leprosy possibly represents an enhanced response of the multibacillary disease in localizing the disease process (3). Because

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of its rarity, variable clinical presentation and uncertainty in its etiopathogenesis HL remained an enigmatic disease.

Herein we report a rare case of de novo HL, affecting the face of a 25-year-old patient.

Case Report

A 25-year-old man, from the sub-himalayan tribal community presented to the Out Patients Department of Dermatology, North Bengal Medical College, Siliguri, India in 2012 with few innocuous tiny nodules in the face. The lesions measuring around 0.5-1 cm in diameter and were present for past one year. The nodules were present in lip, chin and over the pinna (Fig. 1). There was no history of fever, pain, altered sensations or long-term drug intake. No other similar lesions were present in any parts of the body. There was no loss of sensation, hypopigmentation, nerve thickening or regional lymph node enlargement. The patient had no history of kala-azar, tuberculosis or leprosy. The study was approved by the institutional Ethical Committee and informed consent was obtained from the patient.

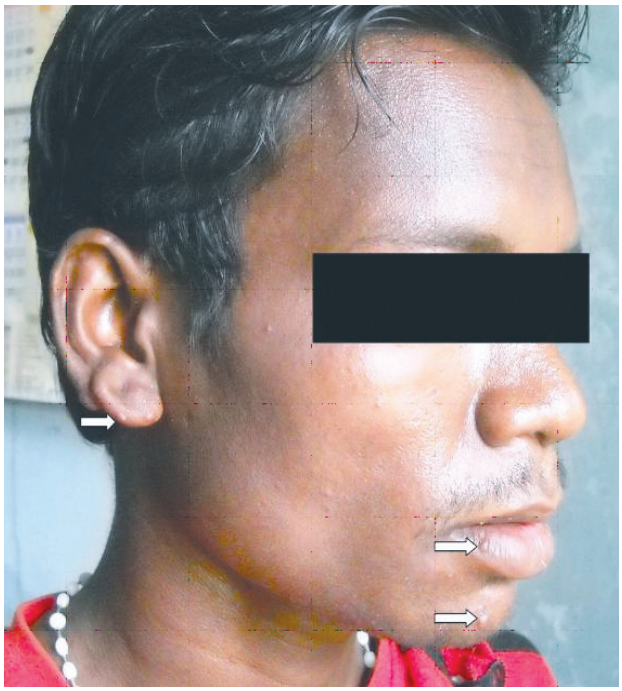

Fig. 1: Histoid leprosy: tiny papules over the face (arrows)

Clinically the lesions mimicked various dermatologic conditions like acne vulgaris, seborrhoeic keratosis, xanthomas, neurofibroma, dermatofibroma, lupus vulgaris, cutaneous leishmaniasis etc. The dermatologists could not come to a definite diagnosis clinically and they advised for biopsy examination. Finally, biopsy from one of the nodule was done and subjected to histopathological examination.

The routine microscopic picture showed dense infiltration of the dermis by histiocytes, which often tend to be spindle shaped and oriented in whorled, storiform-like pattern, beneath the thinned-out epidermis and a well-delineated subepidermal grenz zone (Fig. 2A,2B). Dermal appendages were eventually destroyed. Provisional diagnosis was histoid leproma. The differentials were post-Kala-azar dermal leishmaniasis and dermatofibroma. Modified ZN stain revealed, high bacillary loads with a bacillary Index of 6, the majority were solid staining, arranged in clumps and also in singles. The final diagnosis of 'de-novo' HL was rendered. The patient was put on multibacillary multidrug therapy (MDT) and the lesions were disappeared by three months.

Discussion

Histoid Leprosy is a common disease in India, more prevalent in the southern part of the country. In India, its incidence among leprosy patients is estimated as 2.79% to 3.6%. The average age of diagnosis is between 21 to 40 years (2). Initially, it was thought to result from dapsone resistance and relapse after dapsone monotherapy. However, reports of patients on multidrug chemotherapy and of patients without any treatment are also available now (4).

Histoid Leprosy was known to occur in patients, previously manifested with LL lesions. However, similar to our patient, rare cases have been reported, where patient developed de novo HL without any previous history or features of LL or any form of leprosy. For this reason Sehgal *et al*. has rightly suggested that histoid leprosy

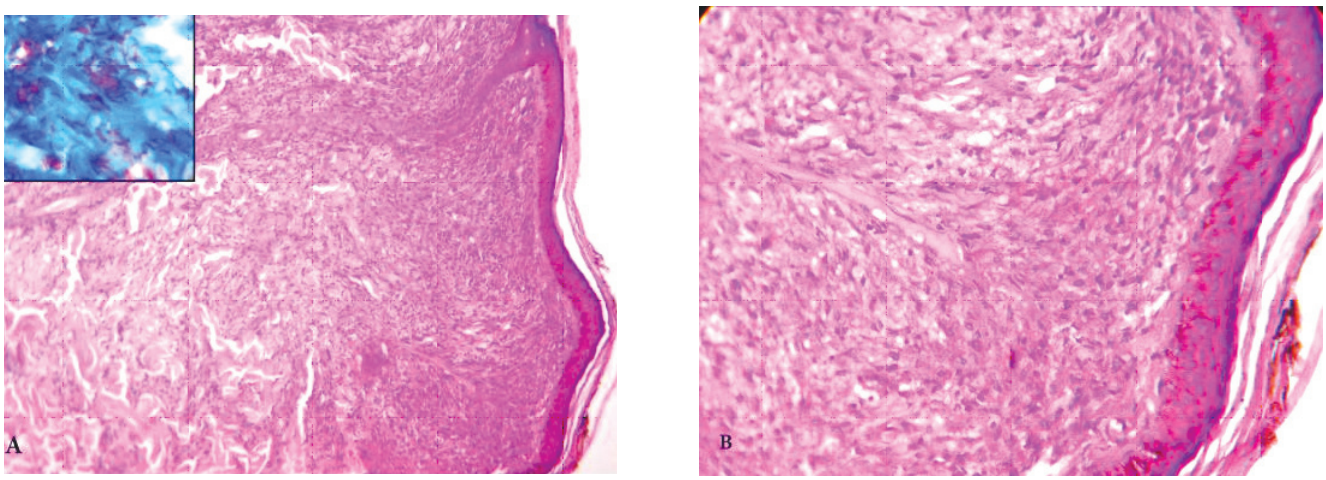

Fig. 2: A) Histoid leprosy: thinned out epidermis with mass of fusiform histiocytes in the dermis and a clear subepidermal grenz zone (H&E stain, ×40 magnification). Clumps of acid-fast bacilli(inset) (Modified Zhiel-Neelsen stain, ×1000 magnification). **B-** Proliferation of spindle shaped macrophages in upper dermis. (H & E stain, ×400 magnification).

should be considered as a distinct form of leprosy (5). Several previous studies have shown male predominance of the disease (3, 4). The increased incidence in males probably is due to their greater exposure during outdoor work. Our patient was also a manual labourer by profession.

Clinically, histoid lepromas present as painless, firm, discrete, smooth and skin-coloured to yellowish-brown cutaneous and/or subcutaneous papules and nodules, located in arms, buttocks, thighs, dorsum of hands, lower part of back and over bony prominences. Unlike our patient face is usually not affected. Histoid lesions have also been reported along the course of the peripheral nerve trunks and cutaneous nerves (6). According to Kaur *et al*. buttock is the commonest site of HL (7). Similar to our case, Mendiratta *et al*. reported most of the HL patients of their series presented with central facial lesion (3). Genital and mucosal lesions have been described by some authors (8).

Unusual presentation and location in the absence of anaesthesia simulates a variety of dermatologic lesions, like xanthoma, neurofibroma, dermatofibroma, lupus vulgaris, herpetic nodules, milker's nodule, cutaneous leishmaniasis etc. Each of them can be differentiated from histoid leprosy based on its characteristic histopathology, absence of lepra bacilli on ZN stained section (6). Histiocytic markers such as CD68 and MAC387 may be helpful along with ZN stain. Slit skin smears from histoid lesion are very helpful for rapid diagnosis. It usually show abundant AFB occurring in clusters packed in macrophages. The bacilli appear long with tapering ends, when compared with ordinary lepra bacilli and having a high bacteriological as well as morphological index (3). In our case, the dermatologists were not certain about the nature of the lesion, so they advised urgent biopsy instead of slit skin smear examination.

Classical histopathological findings include epidermal atrophy overlying subepidermal acellular grenz zone. Dermis is filled with fusiform histiocytes arranged in a whorled or storiform pattern. The histiocytes resemble fibroblasts, within packed with numerous acid-fast bacilli, arranged in parallel bundles along the long axis of the histiocyte (7).

Reactions are uncommon in histoid cases. Erythema Nodosum Leprosum (ENL) can be a commonly observed phenomenon in a leprosy patient during transition to manifest histoid (7).

Histoid Leprosy is initially managed by range of motion (ROM) therapy with rifampicin 600 mg, ofloxacin 400 mg and minocycline 200 mg;

followed by WHO multibacillary MDT therapy (4).

In January 2007, India was declared to have eliminated leprosy. However, on the contrary still cases of leprosy are being reported from different corners of this country including those areas with low endemicity. As suggested by Palit and Inamadar, histoid form could serve as a reservoir of leprosy and as a source of new cases (9). This could pose a serious threat to our elimination program. Early diagnosis and complete treatment of such patients is very important to achieve our goal of elimination of leprosy.

In conclusion, the HL can occur in-patient without previous history of leprosy. Therefore, any long-standing nodular lesion particularly in the face should be viewed with suspicion. Slit skin smear examination and histopathological examination of the lesion are necessary investigations. Histopathology of HL may be some time confusing. Any dermal lesion having storiform spindle cell proliferation should be supplemented by ZN stain to exclude histoid Hansen.

Acknowledgement

The authors declare that there is no conflict of interests.

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