An unusual case of childhood leprosy

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Abstract

Leprosy is a chronic infectious disease which affects skin and upper respiratory mucosa. Disease is somewhat prevalent in most regions of Asia, Africa and South America. Leprosy is known to be associated with poverty, overcrowding and malnutrition. Childhood leprosy indicates ongoing transmission in the society. Majority of children with leprosy have single lesion leprosy and they rarely develop deformities. Unusual forms of childhood leprosy includes nodular leprosy and histoid leprosy. Here we are reporting a case of childhood histoid leprosy.

Introduction

Childhood leprosy should be considered when there are: 1. Longstanding skin lesions 2. Longstanding skin lesions with symptoms of peripheral nerve damage, 3. known leprous contact in the family, 4. Residing in a known leprosy endemic pocket, 5. Any combination of the above. Histoid leprosy is rare in children and is considered as a variant of lepromatous leprosy. It was originally described by Wade as a histological concept meaning bacillary rich leproma composed of spindle-shaped cells with the absence of globi formation. Clinically it is characterized by the presence of multiple, discrete, skin-coloured to yellow brown papules and nodules mainly distributed on posterolateral arms, lower back, buttocks, thighs and over bony prominences. Reactional episodes are uncommon in Histoid leprosy and Erythema Nodosum Leprosum has been reported rarely after initiation of treatment. Here we are reporting a childhood case of histoid leprosy with pronounced facial involvement and associated Erythema Nodosum Leprosum (ENL). Previously he has been misdiagnosed as neurofibromatosis.

Case history

A 11 year old boy presented with asymptomatic progressively increasing skin nodules on the face and limbs for 8 months duration. One of the nodules has been excised one month before in another hospital and the histology was that of a completely excised neurofibroma. He had no hypopigmented anaesthetic patches or numbness of hands and feet suggestive of leprosy or episodes of fever associated with erythematous skin nodules and arthralgia suggestive of lepra reactions. His grandmother had been treated for leprosy years before his birth. Examination revealed multiple discrete, skin coloured, shiny nodules mainly distributed on central face and forearms (Figure 1) and dactylitis involving both middle fingers (Figure 2). He had no characteristic facial changes of lepromatous leprosy except loss of eye brows. Both ulnar and common peronial nerves were thickened. Rest of the neurological examination was unremarkable.

The clinical diagnosis of histoid leprosy was made and confirmed by the positive slit skin smear with very high bacillary and morphological index.

Site	Bacillary Index	Morphological Index
Left ear lobe	5+	10%
Right facial nodule	6+	25%
Right forearm nodules	5+	25%
Left nostril	6+	10%

Following pre-treatment investigations, he was started on Multi Drug Therapy (MDT) - Multi Bacilary (MB) child pack. Routine family screening detected 02 positive family members (brother and father) with tuberculoid leprosy. By 3rd week of therapy he developed low grade fever, swelling of ankles, arthralgia and myalgia. He was pale and had pitting edema of both feet. Investigations revealed lowering of Hb (11.9-9.4g/dl), high reticulocyte count (4.8%) and drug induced haemolysis on blood picture with normal liver function tests. He was managed as dapsone induced haemolysis and low grade Erythema Nodosum Leprosum (ENL). Low dose of dapsone was continued and tapering course of steroids was commenced.

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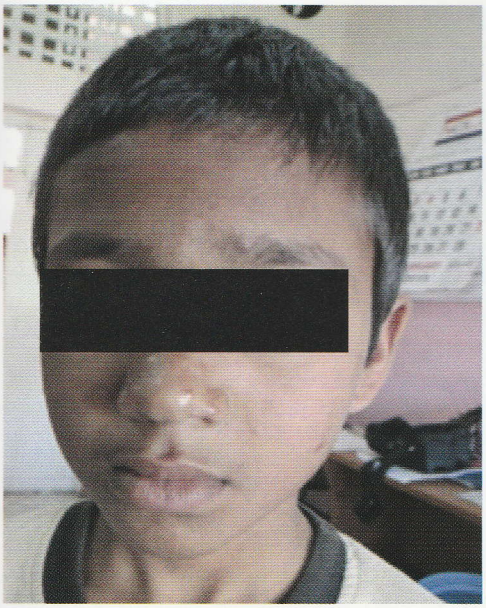

Figure 1.

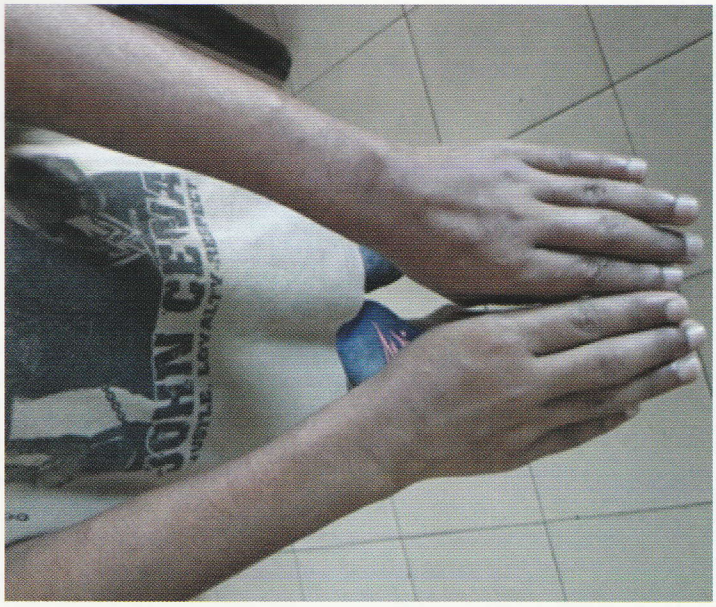

Figure 2.

Discussion

Histoid leprosy is considered as a rare variant of lepromatous leprosy with distinct clinical and histological features. The incidence is 2.79-3.65% of all leprosy cases[2] and has male preponderance. Majority of cases with histoid leprosy are adults and it has been rarely reported in children.

Predisposing factors to develop histoid leprosy are: lepromatous patients who relapse after dapsone monotherapy, dapsone resistance, irregular and inadequate treatment and mutant organism (histoid bacilli). Precise pathogenesis of histoid leprosy is not fully understood. It is thought to be interplay of genetic, immunological, environmental factors and previous therapy. Though strong genetic susceptibility is suggested, responsible genes have not been identified. Exaggerated humoral and cellular immunity to *M. leprae* with augmented local cell mediated immunity has been observed in immunological studies[5].

Characteristic clinical finding is presence of multiple, discrete, shiny, papules, nodules, plaques on apparently normal skin mainly distributed on posterolateral arms, lower back buttocks, thighs and over bony prominences. It may have characteristic features of lepromatous leprosy. Peripheral nerves may be normal or thickened[2,4]. Pronounced facial involvement has rarely been reported as in our case[1,4]. Other rare manifestations of histoid leprosy include: severe mucosal involvement, giant lesions, isomorphic koebnerization. Palms and soles are usually spared 1. Reactional episodes are rare in histoid leprosy and Erythema Nodosum Leprosum has been rarely reported after initiation of therapy.

Histoid leprosy should be considered in the differential diagnosis of dermatofibroma, neurofibroma, reticulohistiocytoma and xanthogranuloma particularly when they affect predilection sites of histoid leprosy. It can closely resemble neurofibromatosis. Positive family history with autosomal dominant inheritance, presence of other cutaneous features and certain differentiating histological findings help to distinguish the two conditions.

Diagnosis of histoid leprosy is mainly clinical, which can be confirmed by positive slit skin smear (SSS) test and histopathology. SSS is a simple bedside test which will be positive with very high bacillary index (5+/6+) and morphological index[4,8]. Fite stain of either SSS or FNAC of histoid nodule shows abundance of bacilli which are longer with tapering ends and arranged singly or in clusters. Histopathology of lesional skin will show normal or atrophic epidermis and cellular band (Unna band/ Grenz zone) located just beneath the epidermis. The leproma consists of fusiform histiocytes arranged in a whorled, criss-cross, or storiform pattern. Within the histiocytes, plenty of Acid Fast Bacilli (AFB) are seen. They are uniform in length and arranged in parallel bundles along the long axis of the histiocytes called historic habitués. Histopathology of neurofibromas closely mimic histoid leprosy and can be

easily misdiagnosed as in our case which indicate presence of histiocytes. Histoid leprosy can be successfully treated with MDT-MB[8].

Co-existence of histoid leprosy and neurofibromatosis is well documented[9]. In our patient almost all lesions subsided following treatment indicating that there are no associated neurofibromas.

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