Histoid Leprosy: Case Report

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SUMMARY Histoid leprosy is a rare but well-defined entity with specific clinical, histopathologic, and bacteriologic features. We present a case of histoid leprosy in an 84-year-old Egyptian male in view of the rarity of this condition. The patient presented with erythematous itchy discrete and coalescent papules that were distributed bilaterally and symmetrically on the front and back of the trunk. Before approaching us, he was initially misdiagnosed as a case of pityriasis rosea. There was no mucosal or facial affection and the patient's general examination was normal. Routine hematologic investigations, urine analysis, liver and renal function tests were all normal. Slit skin smear revealed acid-fast bacilli of BI - 6+ and MI - 50-60%. Histopathologic examination of hematoxylin and eosin-stained section revealed atrophic epidermis with flattened rete ridges and dermal infiltration by nodular granulomata formed of spindle shaped histiocytes with pyknotic nuclei oriented in a storiform pattern. Fite's stain for lepra bacilli showed plenty of acid fast bacilli. So, the diagnosis of histoid leprosy was made. Therefore, ROM therapy (rifampicin 600 mg, ofloxacin 400 mg, minocycline 200 mg) was started and followed by multi-drug therapy for 2 years.

KEY WORDS: histoid, leprosy, multidrug therapy, ROM therapy

INTRODUCTION

Ever since it was first reported by Wade in 1963, histoid leprosy has remained enigmatic (1). It is a very rare form of multibacillary leprosy that may arise *de novo* or following an inadequate treatment with dapsone monotherapy or multidrug therapy (MDT). It commonly affects buttocks, back, face and extremities (2). The typical clinical features of histoid leprosy are numerous shiny, succulent, smooth, dome-shaped, non tender, soft to firm nodules overlying normal looking skin, along with papules and plaques. Histoid leprosy is treated as a multibacillary disease (3).

CASE REPORT

Eighty-four-year-old male patient presented with erythematous itchy papular skin lesions. Lesions were distributed bilaterally and symmetrically on the front and back of the trunk (Fig. 1 A, B) for 3 months. There was no mucosal or facial affection. Sensations were normal and peripheral nerves were not palpable. There was no history of any previous treatment taken and family history was noncontributory. The patient's general examination was normal. Before approach-

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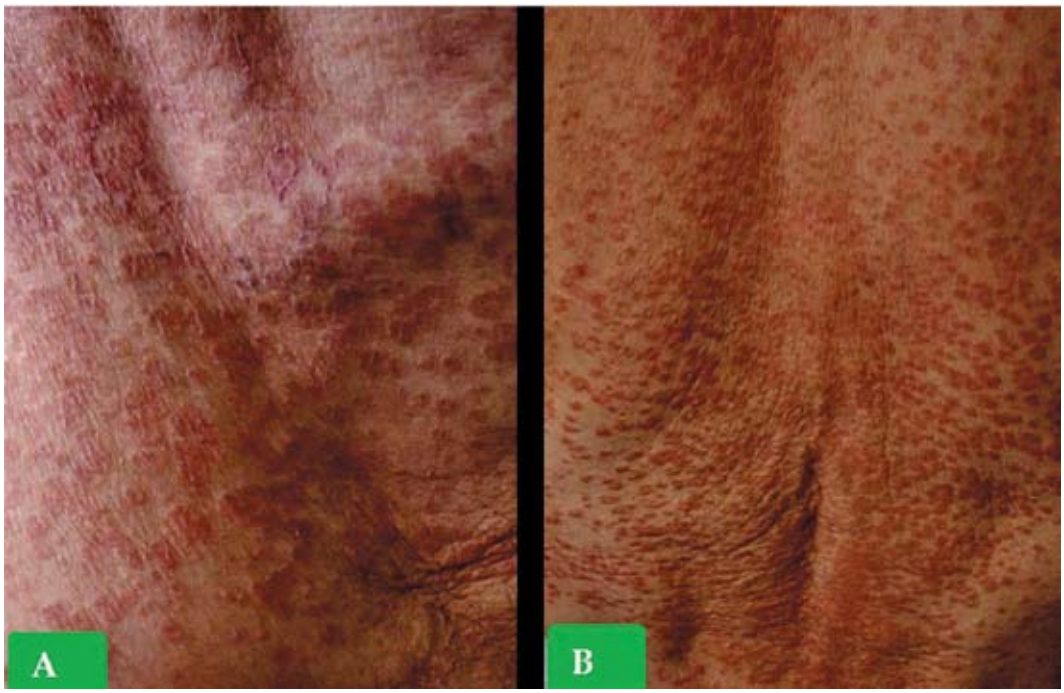

Figure 1. (A) Front of the trunk; (B) back of the trunk showing discrete and coalescent erythematous papules.

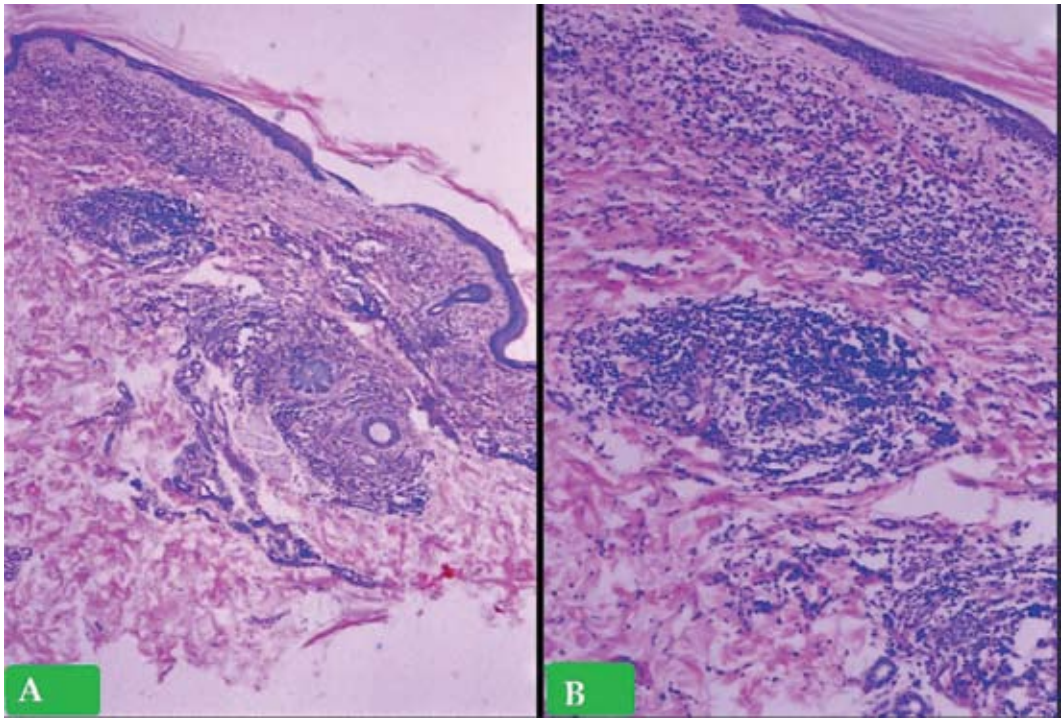

Figure 2. (A) Atrophic epidermis and flattened rete ridges. The dermis shows granulomatous infiltration (H&E, X40); (B) higher power magnification showing dermal granuloma formed of spindle shaped histiocytes with pyknotic nuclei (H&E, X100).

ing us, he was diagnosed as a case of pityriasis rosea and received topical soothing lotion without any improvement.

Routine hematologic investigations were normal. Urine analysis, liver and renal function tests were all normal. Bacteriological examination of slit skin smear revealed acid-fast bacilli of BI - 6+ and MI - 50-60%.

Biopsy was taken from one representative lesion, after getting patient's consent. Histopathologic examination of hematoxylin and eosin-stained section revealed atrophic epidermis with flattened rete ridges and dermal infiltration by nodular granulomata formed of spindle shaped histiocytes with pyknotic nuclei oriented in a storiform pattern (Fig. 2 A, B). Fite's stain for lepra bacilli showed plenty of acid fast bacilli. So, the diagnosis of histoid leprosy was made and the patient started ROM therapy (rifampicin 600 mg, ofloxacin 400 mg, minocycline 200 mg) and instructed to take MDT for 2 years.

DISCUSSION

Histoid leprosy is considered a variant of lepromatous leprosy (1) and by others as a distinct entity (2). The incidence has been reported to vary from 1% to 2% amongst total leprosy patients (3). It was originally described by Wade in 1963 as discrete, firm lesions and dome-shaped nodules which develop on apparently normal skin in patients with lepromatous leprosy (4).

Its exact etiopathogenesis is not well understood as it may arise *de novo* (as in the case presented), or may develop after an inadequate and irregular treatment with dapsone monotherapy or MDT (5).

There is a male preponderance. The increased incidence in males points to their greater exposure due to outdoor work. The average age affected is between 21 and 40 years (4). Kalla *et al.* (3) recorded the youngest patient aged 8 years. To the best of our knowledge, histoid leprosy has not been reported in a patient aged over 80 years, as in our case.

Histoid leprosy has characteristic clinical, histopathologic and bacterial morphological features (6). Clinically, it is characterized by cutaneous and/or subcutaneous nodules and plaques on apparently normal skin (2). The lesions are usually located on the posterior and lateral aspects of the arms, buttocks, thighs, dorsum of hands, and on the lower part of the back and over the bony prominences, especially over the elbows and knees (3).

Classical histopathologic findings include epidermal atrophy as a result of dermal expansion by the underlying leproma and an acellular band located immediately below the epidermis. The leproma consists of fusiform histiocytes arranged in a tangled or storiform pattern containing acid fast bacilli (6).

There are three histologic variants of histoid hansen: pure fusocellular, fusocellular with epitheloid component, and fusocellular with vacuolated cells. The third pattern is most commonly observed (7).

Histoid leprosy might represent an enhanced response of the multibacillary disease in localizing the disease process. An increase in both cell-mediated and humoral immunity against *Mycobacterium leprae*, as in lepromatous leprosy, has been hypothesized (3).

Clinical differential diagnoses include pityriasis rosea, drug eruption and cutaneous metastasis. Each of them can be differentiated from histoid leprosy on the basis of the characteristic histopathology and absence of mycobacteria in slit skin smear.

Nerve thickening was absent in our case, which probably was the cause due to which the diagnosis was missed.

Histoid leprosy is managed by initially administering ROM therapy once, followed by MDT for 2 years (6,8,9).

Today, only one year of therapy is recommended for multibacillary forms and attempts are being made to further reduce the duration. However, whether histoid should be treated as other multibacillary forms or other immunotherapies should be added to the treatment regimen deserves consideration (1).

CONCLUSION

Histoid leprosy is a distinct rare form of multibacillary leprosy with characteristic clinical, bacteriological and histopathologic features. It may arise *de novo* or as a relapse after inadequate leprosy treatment. It is treated as multibacillary forms.

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