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Case report

Histoid leprosy misdiagnosed as dermatofibroma: a case report

組織樣麻風被誤診為皮膚纖維瘤的一例報告

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Histoid leprosy is an uncommon variant of lepromatous leprosy. Its differential diagnoses include dermatofibroma, neurofibroma, molluscum contagiosum, xanthoma, keloids and can result in misdiagnosis in clinical practice. Herein, we report a female patient with histoid leprosy misdiagnosed as dermatofibroma, in order to illustrate the possibility of misdiagnosis, and give physicians an insight into histoid leprosy.

組織樣麻風是瘤型麻風的罕見變體。其鑑別診斷包括皮膚纖維瘤、神經纖維瘤、傳染性軟疣、黃瘤、瘢痕疙瘩等，故有可能在臨床治療中被誤診。本文中，我們報導了一名女性患者的組織樣麻風被誤診為皮膚纖維瘤，用來闡釋誤診的可能，並讓醫者深入了解組織樣麻風。

Keywords: Dermatofibroma, histoid leprosy, misdiagnosis

關鍵詞：皮膚纖維瘤、組織樣麻風、誤診

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Introduction

Leprosy, also known as Hansen's disease, is an uncommon, chronic, communicable disease caused by *Mycobacterium leprae* and has a diverse clinical presentation. Histoid leprosy (HL) is a rare form of lepromatous leprosy (LL), it is rarely observed in the patients who have not received treatment with anti-leprosy drugs.[1,2] Histoid leprosy is a great mimicker and can be easily misdiagnosed as the clinical features are similar to other dermatosis as in the present case.

Case report

A 41-year-old female patient presented to our department, with a 4-year history of generalised plaque, papules and nodules on her face, arms and legs. Two previous skin biopsies were reported as dermatofibroma. The lesions did not respond to treatment with Centella triterpenes cream for three weeks, and no oral medication had been given. Physical examination revealed a few red-brown infiltrating plaques, multiple skin coloured/red-brown papules and cutaneous/subcutaneous nodules scattered on the eyelids, face, forearms, hands and thighs, varying in size from 0.5 cm to 2 cm. The eyebrows were sparse but there were no ocular problems (Figures 1a & 1d). The nodules were mainly distributed on the lateral aspects of forearms and the back of hands, some nodules showed central depression and crust (Figures 1b & 1c), scattered pigmentation and red-brown papules on both thighs (Figure 1d). There was impairment of pain, touch and temperature sensation on the left face, left forearm and dorsum of the hands. The left ulnar nerve was thickened, with limited adduction of the left little finger, but the function of facial expression muscle was intact and there was no arthralgia. Skin biopsy was performed on her left forearm and revealed netlike stratum corneum, epidermal atrophy, a Grenz zone below the epidermis, collagen fibrosis around the lesions and in the dermis, there was a large number of spindle-shaped histiocytes and foam cells infiltrating the lesions, together with numerous acid-fast bacilli as showed by Wade-Fite stain (Figure 2). The diagnosis of HL was confirmed, and treatment with WHO MDT (take under supervision once a month: rifampicin 600 mg, clofazimine 300 mg; and dapsone 100 mg daily, clofazimine 50 mg daily) for leprosy was started.

Discussion

Histoid leprosy is an uncommon variant of LL, first reported by Wade in 1963. The exact position of HL along the immune spectrum and its relation to other subtypes is unclear.[3] At present, it is considered that HL is a form of lepromatous leprosy due to dapsone-resistant bacilli.[4] The prevalence of HL accounts for 1.1-3.6% of leprosy cases and predominantly occurs in multibacillary leprosy patients who have irregular or inadequate treatment, particularly dapsone monotherapy (due to dapsone resistance), but can also occur in relapse cases after successful treatment. The de novo occurrence of HL has also rarely been reported.[1] In HL the average age of infection is between 21 and 40 years although it can rare occur in children. It has a higher incidence in males than in females, which reinforces the importance of this de novo case report.[2,5]

Histoid leprosy is characterised by multiple infiltrated plaques, skin-coloured to red-brown papules and cutaneous, or subcutaneous painless, non-itchy, firm or soft, discrete, smooth, protuberant, globular infiltrated nodules on normal skin.[3] These lesions may resemble dermatofibroma, keloid, neurofibroma, molluscum contagiosum, xanthoma, reticulohistiocytosis, cutaneous metastasis.[5,6] The lesions are usually distributed on the posterior and lateral aspects of the arms, dorsum of hands, back, buttocks, thighs, legs and over bony prominences.[4,5] Moreover, some cases involve the eyebrows, earlobes, mucosae of

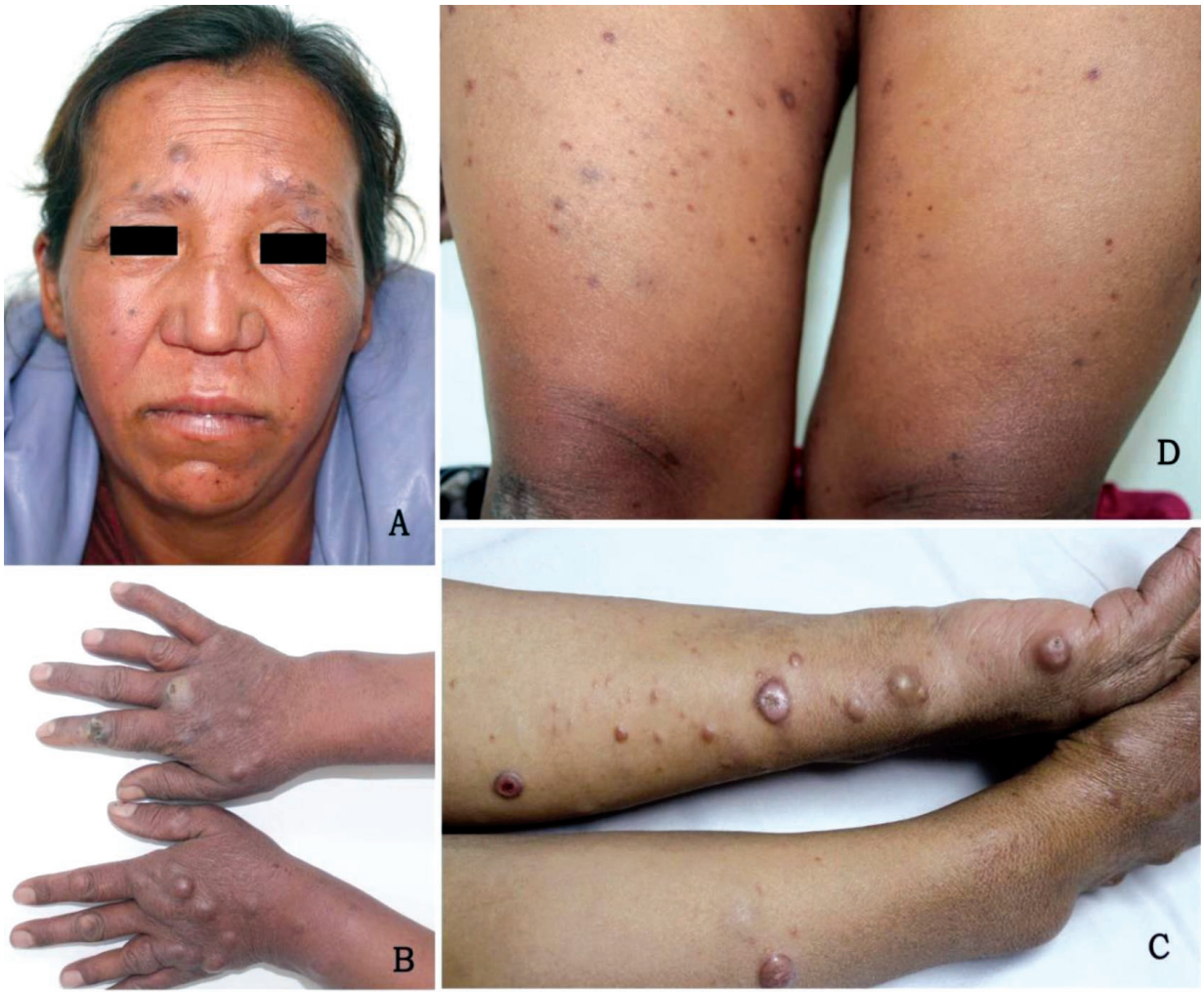

Figure 1. (a) Red-brown plaque, papules and nodules scattered on eyelid, frontal and cheek, with sparse eyebrows. (b) Skin coloured to red-brown, varying in size from 0.5 cm to 2 cm, dome-shaped, multiple nodules on the dorsum of hands. (c) Red-brown papules and nodules distribute on the lateral aspects of forearms, crusting on the surface of some nodules. (d) Pigmentation and red-brown papules scattered on both thighs.

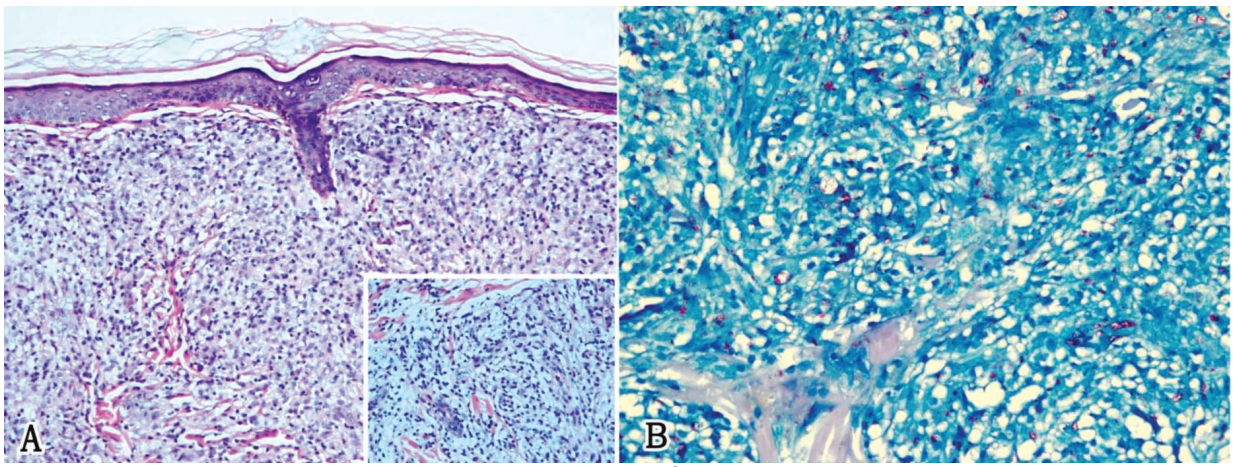

Figure 2. (a) Grenz zone below the epidermis, collagenous fibrosis in dermis around the lesions (Haematoxylin and eosin x 100). Inset: A large number of spindle-shaped histiocytes and foam cells infiltrated in the lesions (Haematoxylin and eosin x 400). (b) Numerous acid-fast bacilli (Wade-Fite stain x 400).

nasal cavity, and genitalia.[1,7] In addition, HL can also affect the peripheral nerves which sometimes show swelling or beading. The ulnar nerve has been reported as the commonest nerve involved. Nerve involvement leads to anaesthetic lesions and paresthaesia with varying degrees of impairment of temperature, touch or pain sensation. This will eventually lead to disability and deformity.[7]

Histoid leprosy is confirmed by histopathology and bacilloscopy. Typical histopathological findings include epidermal atrophy due to dermal expansion by the underlying leproma, with a subepidermal acellular band (Grenz zone) below the epidermis. Numerous spindle-shaped histiocytes and foam cells are present in the lesions. The spindle-shaped histiocytes form a storiform or whorled pattern with numerous acid-fast bacilli (AFB) seen on Ziehl-Neelsen stain which are consistent with multibacillary leprosy.[1,2] As the histopathological findings in HL are occasionally non-specific, early HL cases can be misdiagnosed as dermatofibroma as the clinical lesions resemble dermatofibroma and the histiocytes resemble fibroblasts. In addition, the typical histopathological features of HL (Grenz zone, spindle-shaped histiocytes generating in an intertwining, whorled, or storiform pattern) are often present in dermatofibroma.[8,9] As the clinical lesions and histopathology of HL resemble dermatofibroma, in the absence of bacilloscopy, HL may be misdiagnosed as dermatofibroma as in this case.

Today, in the post-global leprosy elimination era, leprosy has become uncommon especially in developed countries and non-endemic areas where physicians have become unfamiliar with this condition. Histoid leprosy in particular, poses a diagnostic challenge with many physicians unable to make the correct diagnosis in the early stages. We therefore report this unusual case to raise awareness of this entity and improve detection rates, ultimately to prevent transmission in the community and decrease the rate of deformity in patients.

Conflict of interest

The authors reported no conflicts of interest.

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