

Case Report

Histoid lepromatous leprosy presenting as keloid-like nodules

呈疤痕疙瘩樣結節的組織樣瘤型麻瘋

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A 26-year-old Phillipino lady presented with a one year history of multiple hypo- and hyperpigmented patches over the trunk and limbs with coppery-red nodules over her ear and limbs. A diagnosis of histoid lepromatous leprosy was made based on clinicopathological findings. The patient is currently treated with multidrug therapy.

26 歲菲籍女患者近一年於軀幹及四肢出現多發性高色素性及低色素性斑，並在其耳朵及四肢出現銅紅色丘疹。臨牀及病理診斷為組織樣瘤型麻瘋。患者現接受聯合化療。

Keywords: Histoid leprosy, lepromatous leprosy, multibacillary leprosy

關鍵詞：組織樣麻瘋、瘤型麻瘋、多菌性麻瘋

Introduction

Histoid leprosy is a rare form of multibacillary leprosy with distinct clinical and histopathological features. We report a Phillipino lady with histoid lepromatous leprosy presenting as asymptomatic keloid-like nodules with multiple patches of hyper-

and hypopigmentation. The diagnosis was made on the clinicopathological findings.

Case report

A 26-year-old Phillipino lady presented with several non-tender and non-itchy keloid-like nodules over her right ear, right thigh and trunk. She was also noted to have an asymptomatic patch of hyperpigmentation over her right knee for 1 year. Later in the course of the disease she developed faint hypopigmented lesions over her trunk. There was no preceding trauma. Her past health was unremarkable. She was born in the Philippines and has been living in Hong Kong for the past five years. None of her other family members were affected.

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Physical examination revealed a hyperpigmented erythematous-tan patch over her right knee extending to the lateral aspect of her knee (Figure 1). The lesion had an irregular border and was in some areas indistinct with varying lighter and darker shades of tan-brown. There were also three well-circumscribed firm nodules over the right ear, right thigh and right knee measuring 0.7 to 1.5 cm in diameter (Figures 2 & 3). The thigh and knee lesions were erythematous and coppery coloured spherical nodules while the lesion over the right ear was ellipsoid in appearance with a flesh-coppery colour. The later lesions consisted of faint hypopigmented patches over her trunk and limbs. Pain and light touch sensation were

reduced. Palpation of her peripheral nerves revealed thickening of her left ulnar nerve. Examination of her other systems was unremarkable. The constellation of clinical features suggested histoid Lepromatous leprosy as the most likely diagnosis. Other differential diagnoses of the nodular lesions include dermatofibroma, keloid scars, mycobacterial spindle cell pseudotumour, nodular xanthoma.

An incisional biopsy from the right knee lesion showed a nodular infiltrate of spindle and oval cells underneath a subepidermal Grenz zone (Figure 4). Among the spindle cell proliferation are the diagnostic round vacuolated cells stuffed



Figure 1. Hyperpigmented tan-brown patch over the left knee.



Figure 2. A flesh-tan coloured ellipsoid dermal nodule over the right ear.



Figure 3. A well demarcated erythematous dermal nodule over the right thigh.

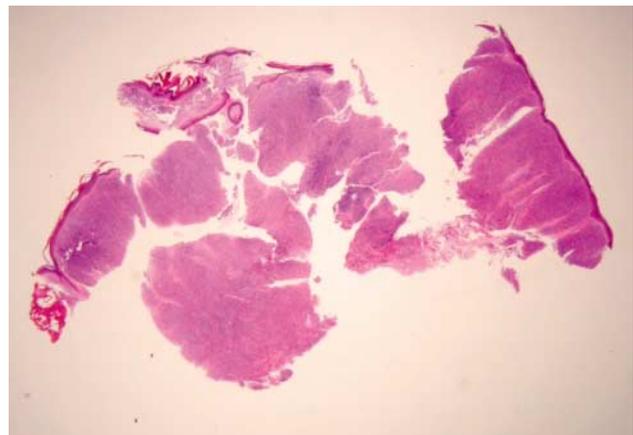


Figure 4. A nodular infiltrate of spindle and oval cells lies beneath the subepidermal Grenz zone. (H&E, Original magnification x 20).

with bacillary aggregates (so-called globi) (Figure 5). Myriads of acid fast lepra bacilli are evident with the Fite stain (Figure 6). A slit skin smear was positive with a Bacteriological Index (BI) of 0.16 and a Morphological Index (MI) of 0. The clinical and histological features were consistent with histoid lepromatous leprosy.

Other investigations showed iron deficiency anaemia whilst G6PD activity was normal. Liver and renal functions were normal. Both anti-nuclear antibody and syphilis enzyme immunoassay (EIA) were negative. The patient was treated with a multi-drug regimen consisting of rifampicin, clofazamine and minocycline.

Discussion

Histoid leprosy is a rare type of multibacillary leprosy with distinct clinical and histological features. It was first described by Wade in 1963. It has a reported incidence of 8.7% amongst lepromatous leprosy (LL) patients, and 1-2% amongst total leprosy patients.¹

Reports of histoid leprosy have occurred after inadequate and irregular treatment with dapsone

monotherapy or multidrug therapy (MDT), but may also occur *de novo* in as many as 68%.²

The aetiopathogenesis of this entity is not clear but an increased cell-mediated and humoral immunity against *M. leprae*³ and augmented local CMI demonstrated by necrosis and ulceration⁴ have been observed. These findings have been suggested to represent a hyperactive expression of multibacillary leprosy in an effort to restrict/localise or focalise the disease.⁵

Clinically, histoid leprosy occurs as sharply delineated cutaneous or subcutaneous papules, nodules, or plaques. These are usually firm, may be translucent and shiny, with an erythematous or coppery colour. The nodules in our patient felt soft during the excision suggesting a recently erupted lesions, whilst the chronic lesions were firmer and fibrotic. They are typically distributed over the backs, buttocks, face, extremities and over the bony prominences, especially the elbows and knees.⁶ Facies with relics of LL such as infiltration, loss of eyebrows and depressed nose may be present. These features were not present in our patient. The peripheral nerves may be thickened in the majority and usually affect multiple nerves. The ulnar nerve has been reported as the

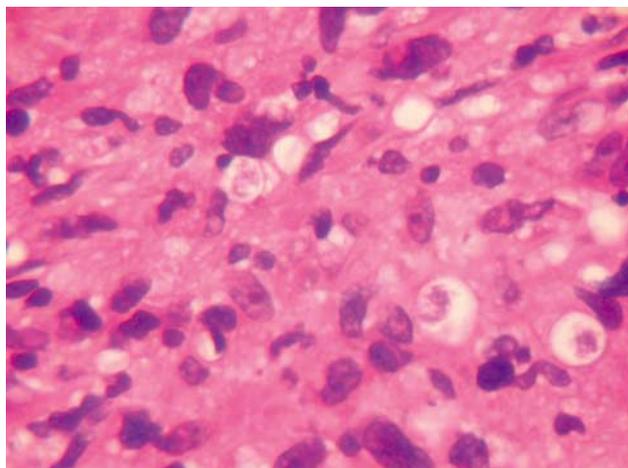


Figure 5. Diagnostic round vacuolated cells stuffed with bacillary globi. (H&E, Original magnification x 650)

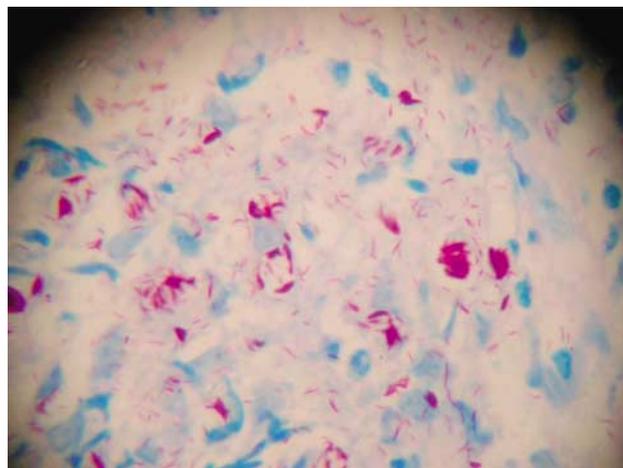


Figure 6. Myriads of acid fast lepra bacilli are present. (Fite, Original magnification x 650)

commonest nerve involved.² An erythema nodosum leprosum (ENL) reaction may occur during treatment and present as an acute exacerbation of existing lesions, softening, ulceration or discharge; these may occur in the absence of other features of the type 2 reaction.⁷

The differential diagnoses of the nodular lesions of histoid leprosy include dermatofibroma, neurofibroma, keloid scar, nodular xanthoma. A mycobacterial spindle cell pseudotumour may also have a similar appearance.⁸ A rare papulonodular variant of secondary syphilis sparing the palms and soles can also present with nodules mimicking those of histoid leprosy.⁹ Neurofibromatosis may also give features of multiple nodules, hyperpigmented (and less commonly hypopigmented) macules together with peripheral nerve enlargement.¹⁰ Clues to the diagnosis of histoid leprosy include the typical nodules together with the constellation of lesions present in multibacillary leprosy. The finding of altered or loss of sensation and peripheral nerve enlargement are particularly relevant.

Histologically, histoid leprosy is characterised by a granuloma formed by spindle-shaped histiocytes and/or polygonal cells arranged in a whorled, criss-cross or interlacing pattern. There is complete circumscription of the dermal granuloma in the majority of cases and may be pseudocapsule formation. A free subepidermal zone may occur. The epidermis may be normal or atrophic. Some cases may resemble a fibrohistiocytic tumour. A Ziehl Neilsen stain shows a striking abundance of uniformly stained long bacilli both intra- and extracellularly.^{2,5}

The slit skin smear performed in patients with histoid leprosy always reveal numerous solid-staining bacilli which often lie singly but occasionally in clusters or in globii. Most organisms are usually well preserved and long with tapering ends.⁵ Interestingly the BI was low and the MI was negative in our patient.

Treatment of leprosy in Hong Kong is based on the WHO recommended multi-drug therapy regimens. For multibacillary disease such as our patient the first line treatment includes monthly supervised treatment with rifampicin 600 mg and clofazamine 300 mg supplemented with daily self-administered clofazamine 50 mg and dapsone 100 mg. Minocycline 100 mg daily is a second-line drug and was substituted for dapsone in our patient because of anaemia.

Apart from pharmacological treatment, it is important to perform contact tracing, rehabilitation and educate patients to prevent long-term disability.

References

1. Sehgal VN, Srivastava G. *Histoid Leprosy*, 3rd edn. New Delhi: Jypee Brothers Medical Publishers, 2004.
2. Kalla G, Purohit S, Vyas MC. Histoid, a clinical variant of multibacillary leprosy: report from so-called nonendemic areas. *Int J Lepr Other Mycobact Dis* 2000;68:267-71.
3. Sehgal VN, Srivastava G. Status of histoid leprosy-- a clinical, bacteriological, histopathological and immunological appraisal. *J Dermatol* 1987;14: 38-42.
4. Sehgal VN, Srivastava G, Sharma VK. Immune responses in disseminated necrotizing histoid leprosy. *Int J Dermatol* 1988;27:413-4.
5. Sehgal VN, Aggarwal A, Srivastava G, Sharma N, Sharma S. Evolution of histoid leprosy (*de novo*) in lepromatous (multibacillary) leprosy. *Int J Dermatol* 2005;44:576-8.
6. Ramanujam K, Ramu G. Wade's histoid lepromatous leprosy; report of a clinical study. *Lepr India* 1966;41: 293-7.
7. Sharma SK, Rath N, Gautam RK, Sharma PK, et al. Histoid leprosy with ENL reaction. *Indian J Dermatol Venereol Leprol* 2002;68:342-3.
8. Liou JH, Huang PY, Hung CC, Hsiao CH. Mycobacterial spindle cell pseudotumor of skin. *J Formos Med Asso* 2003;102:342-5.
9. Pandhi D, Reddy BS, Khurana N, Agarwal S. Nodular syphilis mimicking histoid leprosy. *J Eur Acad Dermatol Venereol* 2005;19:256-7.
10. Thomas J, Wilson NC, Parimalam S, Augustine SM, Muthuswami TC. Multiple neurofibromatosis with histoid leprosy. *Int J Lepr Other Mycobact Dis* 1989; 57:695-6.