

CASE REPORT

Zoster-like segmental presentation of lepromatous leprosy

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Introduction

The Ridley-Jopling classification places leprosy patients into a spectrum with polar tuberculoid (TT) and lepromatous (LL) and middle types of borderline tuberculoid (BT), mid-borderline (BB) and borderline lepromatous (BL) leprosy.¹ These have been categorised, for treatment purposes, into paucibacillary (PB) and multibacillary (MB) depending on lesion counts and the results of skin smear examinations.²

Lepromatous leprosy (LL) presents at onset usually with symmetrically distributed nodules, plaques or hypopigmented macules on the trunk. If LL progresses, skin begins to thicken predominantly in the forehead, earlobes, eyebrows and cheeks, which eventually leads to the classic leonine facies.²

In some cases leprosy can manifest with unusual clinical forms that may mimic different cutaneous diseases; early recognition of these diverse clinical features is the key for early treatment and prevention of disabilities.³

Case report

A 58 year-old man, a farmer, presented with a 6-month history of asymptomatic erythematous skin lesions on his chest and abdomen. A few small nodules initially appeared on his chest, and then over a few weeks they increased in number and size and spread to the abdomen. This progression led him to seek medical advice. The patient was not immunosuppressed,

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and he was not taking any medication. A general examination was unremarkable. There was no family history of leprosy or contact with a known leprosy case. The patient had not observed any other skin condition affecting the same sites prior to the present episode.

Skin examination disclosed multiple, asymptomatic infiltrated, shiny, erythematous nodules of different sizes, some of which showed confluence into plaques, on his right chest and abdomen with an asymmetric, segmental, distribution (Figure 1).

A plaque on the right nipple had altered its normal morphology. Sensitivity to light touch was preserved in the lesions and no other skin lesions were observed on the rest of the body surface. There was no abnormal thickening or tenderness of peripheral nerves, and routine nerve function assessment revealed no deficiency. Ophthalmology consultation was unremarkable.

The progressive clinical course and presentation suggested a differential diagnosis of cutaneous lymphoma, cutaneous malignant metastasis, Paget's disease, dermatofibrosarcoma protuberans and leprosy with atypical presentation. Histopathological examination of a nodule led to a diagnosis of LL leprosy on the basis of: an atrophic epidermis with an underlying narrow clear Grenz zone in the papillary dermis and a diffuse infiltrate of foamy macrophages in the reticular dermis. Wade-Fite stain revealed abundant bacilli in clumps and globi (Figure 2).

Slit skin smears taken from the ear lobes showed a bacterial index (BI) 6+ and a morphological index (MI) of 50%. The small nodules on the chest showed a BI 5+ with a MI of 50%. Apparently normal skin showed a BI 4+ and MI 5%.

The World Health Organization recommended multidrug therapy (WHO-MDT) regimen for MB leprosy was started. All household contacts were examined and none was found with suspect lesions.

At the follow-up after a year, infiltrated small nodules were still present on the chest and abdomen, demonstrating only little clinical improvement. The BI was still 5+ with MI 1%. In accordance with the Guide to Leprosy Control, Ministry of Health of Brazil (2002) that recommends a 24-month therapy for multibacillary leprosy patients with little or no clinical improvement one year after WHO-MDT regimen,³ treatment was extended for another 12 doses.



Figure 1. Infiltrated shiny erythematous small nodules, small nodules and plaques on the right nipple and right side of the abdomen.

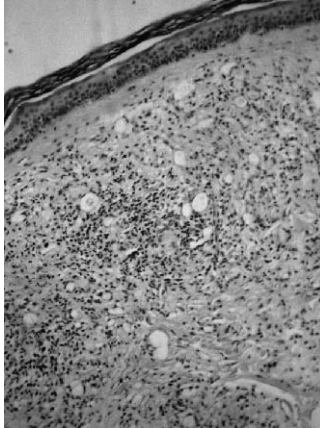


Figure 2. Histopathology of lepromatous form: dermal infiltrate of Virchow cells and the Unna band or Grenz zone separating the epidermis from the infiltrate.

Follow-up examination after 2 years' treatment revealed that the skin lesions were either less infiltrated or had healed with post-inflammatory hyper-pigmentation (Figure 3).

No reactions or adverse effects of MDT had been observed. Clinical improvement was confirmed by a reduced BI of 3 + with MI 0% repeated at the same site as at the time of the diagnosis.

Discussion

Leprosy still remains endemic in many tropical and subtropical countries despite dramatic reduction of the number of registered cases as a result of MDT.⁴ Leprosy presents a broad clinical and histopathological spectrum that is better correlated with the immunological response of the patient than with bacillary invasion.⁵

Definitive classification of leprosy can only be made with the clinical-bacteriological and pathological correlation. Unfortunately, only approximately 70% of leprosy patients can be correctly classified on clinical evidence alone. The remaining 30% of cases of leprosy present with atypical skin lesions and other unusual signs that may mimic a broad spectrum of dermatological conditions. In such patients, leprosy diagnosis might be a challenge, resulting in therapy delay and consequent possible neural sequelae.^{6,7}



Figure 3. After two years of continuous treatment, skin lesions were either less infiltrated or healed with postinflammatory hyperpigmentation.

Reports in the literature of unusual leprosy presentations include those of a single nodule on the face or a single plaque on the leg being described as the first manifestation of multibacillary leprosy.^{7,8} Clinical presentations with the rare histoid leprosy type and erythema nodosum leprosum (even necrotic) have also been reported.^{9–11} Recently, Da Costa Nery *et al.* described different, uncommon types of LL manifested as erythema multiforme-like lesions, lymphadenopathy masquerading lymphoma and long standing leg ulcers.³ Diffuse infiltration of the skin as in Lucio's leprosy, diffuse oedema of fingers, hands and feet, and palmo-plantar cyanosis are also other uncommon presentations of leprosy.³ Moreover, signs and symptoms resembling rheumatic diseases can also accompany the onset of leprosy as reported by Ribeiro *et al.*¹² These authors described two women with multiple, purpuric, ulcerated and necrotic skin lesions, livedo reticularis, joint pain, oligoarthritis, myalgia, and leg edema resembling systemic lupus erythematosus or polyarteritis nodosa.¹²

To our knowledge, there are no reports in the English literature of zoster-like or segmental presentation of LL. Leprosy is endemic in Amazonas (the detection rate of new cases in 2008 was 22.78 per 100,000) but LL diagnosis in our patient was a clinical challenge, as both morphology and distribution of the lesions were not typical for LL and simulated different conditions. The diagnosis was only established using skin smears and histopathological investigations.

Leprosy referral facilities with experienced clinical staff and easy access to skin smear services are of utmost importance for supporting peripheral dermatological services in diagnosis, and management of such unusual cases. In endemic areas, skin smears should be always performed first whenever leprosy is suspected, even if the clinical presentation is unusual as in the case presented here. Delayed diagnosis and treatment of leprosy may result in irreversible sequelae.^{13,14}

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