CASE REPORT

Clinical and histopathological profile of a patient with localized lepromatous leprosy

JACQUILINE JOSEa, LORETTA DASa, P.A. SINGHb & RUBY MARSHALAA
aThe Leprosy Mission Community Hospital, Naini, Prayagraj, Uttar Pradesh, India
bAnoop Pathology, Prayagraj, Uttar Pradesh, India

Accepted for publication 15 July 2019

Summary Lepromatous leprosy is a polar form of the disease characterized by very low to absent cell mediated immunity, resulting in unrestricted multiplication of the leprosy bacilli. This leads to infiltration and papulo-nodular lesions, which initially favor the cooler areas of the body and later become generalized. However, lepromatous leprosy may rarely be localized to a single or limited sites. Routine skin smears are negative but a skin smear and histopathology from the affected sites are confirmatory. These cases may be misdiagnosed as paucibacillary leprosy with subsequent under-treatment of the patient. Clinicians should be aware of this entity as a simple skin smear from the site can confirm the diagnosis. A 57-year old man presented with an erythematous plaque surrounded by a few satellite papules on the right hand, right ulnar paralysis and partial anaesthesia of the right hand and both feet, for one year. Slit skin smears from routine sites were negative for acid fast bacilli. However, a skin smear from the satellite lesions revealed a bacillary index of 4+. Histopathology of the skin was consistent with lepromatous leprosy. He was initiated on the WHO multibacillary multidrug therapy (MB-MDT).

Keywords: localized lepromatous leprosy

Introduction

One of the oldest infectious diseases known to mankind, the first reports of leprosy date back to 1400 BC. However, many aspects of its pathogenesis are still undetermined. One of the rare presentations of leprosy is localized lepromatous leprosy, where there is a discrepancy between the clinical findings and histopathology.
Case report

A 57 year old gentleman from Uttar Pradesh, India presented to our outpatient department with a slowly progressive, reddish, anaesthetic, raised lesion on the right wrist and hand present for one year. He complained of weakness, clawing and loss of sensation involving the right 4th and 5th fingers for one year, as well as partial anaesthesia of both soles. He did not have any systemic symptoms. There was no history of contact with leprosy.

General examination was unremarkable, with no signs of madarosis, ear lobe thickening or other infiltration. Cutaneous examination revealed a single, ill-defined 10 × 5 cm
erythematous anaesthetic plaque extending from the right wrist to the right palm, surrounded by a few reddish papules proximally (Figures 1 and 2). Peripheral nerve examination was significant with enlargement of both ulnar nerves, both common peroneal nerves and the left posterior tibial nerve. Voluntary muscle testing showed a right ulnar nerve paralysis (Medical Research Council grade 0) with clawing of the right 4th and 5th fingers. Sensory testing demonstrated anaesthesia along the ulnar distribution on the right hand and partial anaesthesia on both soles. There were no trophic ulcers or other deformities.

Based on the history and clinical findings of a single anaesthetic plaque with asymmetrical peripheral nerve involvement, a provisional diagnosis of Borderline Tuberculoid Hansen’s disease with right ulnar paralysis was made. A slit skin smear was performed from 3 routine sites as per institutional policy and also from the lesion on the right hand. The bacillary index (BI) from the plaque was 4+ and negative from all other sites. Histopathology was performed from the papules on the wrist and showed normal epidermis

Figure 2. Absence of madarosis, ear lobe thickening and diffuse infiltration of the face.
with an underlying clear grenz zone (Figure 3); the dermis exhibited massive infiltration by foamy macrophages containing numerous acid fast bacilli (AFB) as demonstrated by Fite-Faraco stain and a relative paucity of lymphocytes (Figure 4). Thus, this patient was diagnosed bacteriologically and histopathologically as lepromatous leprosy. He was initiated on the World Health Organization multibacillary multidrug therapy (MB-MDT) and physiotherapy, and subsequent reconstructive surgery and family screening was planned.

Discussion

Leprosy or Hansen’s disease is an important public health concern, with new case reports from India constituting 59.8% of the global burden in 2017.1 It is a chronic mycobacterial infection of low virulence that primarily affects the peripheral nerves and skin. The aetiological agent, *Mycobacterium leprae*, is an obligate intracellular acid fast bacillus with an affinity for the cooler areas of the body such as the respiratory tract, skin, anterior chamber of the eye, testes and the reticulo-endothelial system.

There is an inverse relationship between cell mediated immunity (CMI) and bacillary load which determines the spectrum of disease in the affected individual. In lepromatous leprosy, diminished CMI permits unrestricted bacillary multiplication and widely disseminated multi-organ disease. Diffuse dermal infiltration by the bacilli manifests as enlargement of the ear lobes, widening of the nasal root, fusiform swelling of the fingers, prominent skin folds and loss of cutaneous appendages. Other skin findings include numerous generalized macules, papules, painless nodules or confluent plaques. This may be accompanied by a symmetrical enlargement of peripheral nerves in advanced disease. Sensory loss may be present in a characteristic glove and stocking pattern. Mucosal
involvement may manifest as nasal stuffiness or epistaxis. Skin smears are highly positive and the lepromin test is negative. Histopathological examination of the skin shows a normal or atrophic epidermis with abundant AFB within foamy histiocytes, a clear grenz zone and enlarged dermal nerves with few inflammatory cells.

In contrast, tuberculoid leprosy is characterized by a near-normal CMI, which confines the mycobacteria to localized areas. There is phagocytosis and digestion of AFB by macrophages, which mature into epithelioid cells. Tuberculoid spectrum disease presents as a single or few asymmetrically distributed anaesthetic, anhidrotic patches or plaques. Peripheral nerves may be enlarged asymmetrically early in the course of the disease. Mucosae are typically spared. Skin smears are usually negative and the lepromin test is strongly positive. Histopathology reveals epithelioid granulomas with very few to absent AFB and abundant lymphocytes. Dermal nerves are destroyed and a grenz zone is usually absent.

Localized lepromatous leprosy is a rarely reported entity where clinical findings are suggestive of tuberculoid spectrum disease but there are localized areas teeming with AFB as confirmed by skin smears and histopathology.\textsuperscript{2–9} It is generally believed that lepromatous leprosy in India originates from the borderline spectrum.\textsuperscript{2} So these unusual presentations may be considered as borderline lesions in the early stages of evolution into polar lepromatous leprosy. Another possibility is that these lesions are inoculation lepromas wherein susceptible hosts acquire the infection from viable bacilli present in the environment through minor trauma.\textsuperscript{3,9,10} This little known variant of leprosy emphasizes that there are as yet unknown factors which determine the host response to the leprosy bacilli.\textsuperscript{4,5} This case report also highlights the importance of performing slit skin smears and histopathology on unusual skin lesions, thus ensuring accurate diagnosis and an adequate duration of treatment.

\textbf{Figure 4.} Plenty of acid fast bacilli (BI 4+), modified Fite stain, 1000X.
Localized lepromatous leprosy

References