Leprotic involvement of peripheral nerves in the absence of skin lesions

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Sri Lanka Journal of Dermatology, 2007, **11**, 24-25

Abstract

In the absence of clinically apparent cutaneous lesions, purely neural involvement in leprosy is uncommon. These are recognized as pure neural leprosy and can be seen in 4 to 10% of leprosy patients. Primarily neural leprosy present clinically as peripheral neuropathy that most frequently affects motor nerves and occasionally sensory nerves as well. Since its involvement is patchy, most of the time histology is not supportive.

We present a 17-year old boy with glove and stocking hypoesthesia, intrinsic muscle wasting of hands and bilateral ulnar claw hand. There was no thickening of superficial nerves. Sural nerve biopsy was performed and histology revealed inflammatory infiltration and presence of *Mycobacterium leprae*.

We believe that in cases of peripheral neuropathy without a known aetiology, it is useful to perform a superficial nerve biopsy to exclude the diagnosis of pure neural leprosy mainly in developing countries where leprosy is prevalent.

This case is reported as pure neural leprosy is uncommon and the yield of positive histology with numerous *Mycobacterium leprae* is very rare.

Introduction

The main cause of morbidity in leprosy is the peripheral neuropathy that is responsible for the great bulk of disabilities and deformities displayed by many leprosy patients. It is important to consider leprosy as a cause of peripheral neuropathy, because it is readily treatable and produces crippling sequelae if it is not diagnosed and treated in its early stages.

Leprosy neuropathy may also present without skin lesions, this is known as the neuritic form of leprosy. The patients with this form of the disease displays only signs and symptoms of sensory impairment, parasthesia, nerve enlargement, nerve pain and muscle weakness without dermatological alterations. In the absence of detectable skin lesions the aetiologic diagnosis of peripheral neuropathy as a result of infection with Mycobacterium leprae is usually confirmed by means of histological examination of a biopsy specimen of a small peripheral nerve.

We report a case of pure neural leprosy that was histologically confirmed on sural nerve biopsy.

Case report

A 17-year old boy presented to a peripheral unit with a history of glove and stocking hypoesthesia, intrinsic muscle wasting of hands and bilateral ulnar claw hand for six months duration. His mother had multiple hypopigmented hypoesthetic patches on the body. Both of them were started on multibacillary leprosy treatment. After one month of treatment both of them developed fever, jaundice and scaling of the skin. Leprosy treatment was discontinued and they were transferred to National Hospital of Sri Lanka. He was diagnosed to have dapsone hypersensitivity syndrome and dapsone induced agranulocytosis.

As there were no cutaneous lesions suggestive of leprosy and no nerve thickening, he underwent investigations for peripheral neuropathy. His fasting blood sugar was normal. Nerve conduction studies showed sensory motor axonal polyneuropathy.

Sural nerve biopsy was performed and histology revealed inflammatory infiltration and presence of numerous *Mycobacterium leprae* bacilli.

He was started on ofloxacin and clofazimine. One month later he developed diabetes and was treated with insulin. He is currently undergoing follow up at our clinic.

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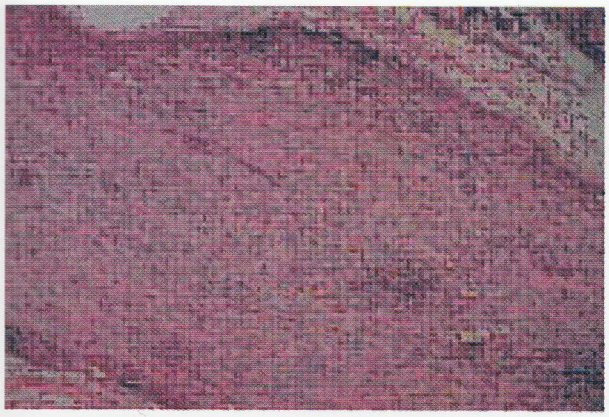

Figure 1. *Sural nerve biopsy – nerve tissue infiltrated with lymphocytes (H&E).*

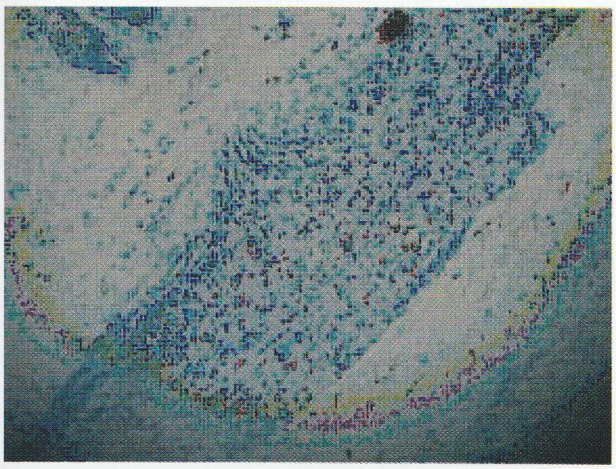

Figure 2. *Sural nerve biopsy showing numerous Mycobacterium leprae bacilli (Fite stain).*

Discussion

Leprosy is one of the most common peripheral nerve diseases in the world. It usually affects the skin and the nerves. It is caused by the *Mycobacterium leprae*, an acid fast bacillus. Although the prevalence of the disease is decreasing, leprosy continues to be a major cause of infective neuropathy in tropical and subtropical countries. Patients with leprosy may have only nerve involvement without obvious primary skin lesions. The pure neural leprosy is a well recognized clinical entity, comprising 4 to 10% of patients with leprosy[2]. In this pure neuritic form, the ulnar and the common peroneal nerves are the most frequently affected nerves.

It is important to be aware of the common neurological manifestations of peripheral neuropathy of leprosy such as local, asymmetric sensory impairment, the preferential involvement of intrinsic muscles of the hand and of the feet, nerve enlargement and tenderness to achieve an early diagnosis of this disabling and preventable deforming specific neuropathy.

In most cases of pure neural leprosy the diagnosis cannot be firmly established on clinical grounds alone. Only the nerve biopsy could establish the exact diagnosis. Superficial sensory nerve biopsy is a relatively simple procedure for the diagnosis of pure neural leprosy.

Limitations of this nerve biopsy technique are sample error, low sensitivity, and permanent nerve deficit. Recent improvements in sampling techniques, using fine needle aspiration cytology and specimen from nasal mucosa and dermatologically normal but hypoesthetic skin have resulted in significantly increased diagnostic yield and less side effects. The additional usage of antibodies specific to Mycobacterial antigens further enhances diagnostic accuracy.

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