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Review

Neurological manifestations of Hansen's disease and their management

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Abstract

Hansen's disease is almost eliminated from developed countries but in developing countries of Africa, Asia and Latin America leprosy is still considered to be a public health problem. *Mycobacterium leprae* have the affinity for peripheral nerves and neuropathy is a cardinal manifestation of the disease. The nerve damage affects sensory, motor, and autonomic fibers resulting in the physical impairments and limitation of physical activities and social participation. Leprosy is a curable disease and treatment provided in the early stages will avert the disabilities. Approach to the neuritic leprosy depends on its clinical characteristics, nerve biopsy, and histological appearance of dermatological and neurological lesions. In this article we review the literature and discuss the pathology, clinical features, diagnosis and management of neurological manifestations of leprosy.

Keywords: Hansen's disease; Leprosy; Neuritic leprosy; Lepra reactions

Contents

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1. Introduction

Leprosy was recognized in the ancient civilizations of China, Egypt and India. The earliest report of Leprosy dates back to 600 BC. In 1864 G. Armour Hansen reported his observations on tissue from a Norwegian patient and became the first to link a bacterium to human disease. This organism, which later came to be known as *Mycobacterium leprae*, is one of the important causes of treatable neuropathy worldwide. Failure in early detection often leads to severe disability in spite of eradication of mycobacteria at a later date. The most likely mode of *transmission* is through nasal secretions and skin contact. The disease is thought to be of low infectivity. In most populations, over 95% of individuals are naturally immune [1]. In spite of this the disease accounts for approximately 10 million affected people world wide, mostly in developing countries of Africa, Asia and Latin America where leprosy is still considered a public health problem.

2. Pathogenesis

M. leprae multiplies very slowly and the incubation period is about five years. Symptoms can take as long as 20 years to appear. Nerves, which are generally resistant to bacterial infections are consistently invaded by *M. leprae*. Another extraordinary feature is that the critical temperature required for multiplication of *M. leprae.* It fails to multiply at core body temperature of 37 °C and optimal growth occurs at 27–30 °C which is responsible for the occurrence of leprosy in superficial and cooler areas such as skin, nerves, testis and upper respiratory tract. Peripheral neuropathy is the main cause of morbidity in leprosy and responsible for most of the disabilities and deformities displayed by many leprosy patients [2,3]. The nerve damage affects sensory, motor, and autonomic fibers. These nerve lesions are characterized by a chronic or subacute inflammatory infiltrate containing epithelioid cells or *M. leprae*-glutted macrophages. This infiltrate will occupy the endoneurium, perineurium and epineurium [4]. This will lead to progressive impairment of unmyelinated and myelinated neural fibers followed by a replacement of the peripheral nerve parenchyma with fibrous tissue [5]. Necrotic caseation in tuberculoid granulomas can lead to abscess formation and complete destruction of the nerves [6].

Pure neuritic form of neuropathy may present without skin lesions. Pure neuritic form has a varied incidence among the total number of cases in an endemic leprosy population comprising 4–10% of patients. Males are significantly more affected than females [7,8]. In this neuropathy, the small nerve fibers conducting pain and temperature sensations are affected significantly before the large myelinated fibers that conduct vibration sense, position sense, and motor impulses. This selective sequential involvement of the nerve fibers impairs the detection of leprosy neuropathy at the initial stages of the disease by neurophysiological evaluation since routine nerve conduction studies only record potentials originating from fibers wider than 7 μm in diameter. Histologic preparations in these patients usually show changes compatible with borderline or tuberculoid leprosy [2,9].

3. Clinical features

3.1. Symptoms

Leprosy mainly affects the skin and nerves and resembles many dermatologic and neurologic conditions. Untreated the disease is progressive and results in permanent damage to the skin, nerves, limbs and eyes. Leprosy can be classified according to the number of skin lesions present and the number