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Case Report:

A case report: Leprosy Polyneuritis from Bangladesh

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Abstract

A 40 year old man presented with progressive weakness, wasting and sensory disturbance of all 4 limbs for 1 and half months. He was diagnosed as a case of Leprosy Polyneuritis. Relevant studies were reviewed. The Mycobacterium survives better at a temperature close to 30°C rather than 37°C. Menifestations of leprosy can be divided into five clinical forms based upon clinical and immunological diffrences, they are tuberculoid leprosy, borderline tuberculoid, borderline, borderline lepromatous and lepromatous leprosy[1]. The diagnosis of leprosy requires high clinical suspicion, confirmation done by bacteriological and histological analyses, which reveals caseating granulomas. Nerve biopsy may also show evidence of the lepra bacilli residing in endothelial, perineurial, Schwann cells and also in macrophages[3]. An useful skin test, Lepromin test, performed by intradermal injection of a standardized extract of the inactivated "leprosy bacillus"; usually positive in tuberculoid form but can be negative in lepromatous leprosy. Differentials include SLE, tertiary syphilis, sarcoidosis, peripheral nerve pathology, lymphoma etc.

Keywords: Leprosy Polyneuritis.

Introduction

Leprosy or Hansen's disease, is a chronic systemic granulomatous disease caused by infectious acid fast bacteria *Mycobacterium leprae*. In the peripheral nervous system it primarily infects schwann cells, can also affect histiocytes in the dermis, mucosa of upper respiratory tracts and other tissues such bones[1]. Mode of spread of *M. laprae* is thought to be by airborne droplets and has an incubation period ranging from 3 months to 40 years (average 7 years)[2]. The Mycobacterium survives better at a temperature close to 30°C rather than 37°C. Menifestations of leprosy can be divided into five clinical forms based upon clinical and immunological diffrences, they are tuberculoid leprosy, borderline tuberculoid, borderline, borderline

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Case Report:

A 40-year-old male, normotensive, non-diabetic, right handed service holder working abroad visited at the outpatient department of neurology, Bangabandhu Sheikh Mujib Medical University. He presented with weakness and wasting of all four limbs for one and half months. He states that, he was reasonably well one and half months ago, then suddenly developed weakness of all four limbs starting from both lower limbs and within a week involving both upper limbs, which was associated with muscular pain of all four limbs. He also complained of diminished sensation of his hands which persisted for about a week at the onset of his weakness. For this he experienced mild thermal injury. He had no history of difficulty in deglutition, speech or respiratory distress. He denied of any bowel or bladder dysfunction, neck or back pain, any joint pain or stiffness, skin ulceration or discoloration, abdominal pain or reddish discoloration of urine. As his weakness became more severe, he became bedbound within two weeks of illness. He was initially admitted in a hospital abroad, where at first he received symptomatic treatment along with analgesics but as his condition deteriorated. Then he was further evaluated with investigations including serology, nerve conduction study and cerebrospinal fluid examination. This revealed neutrophilic leucocytosis, hyponatraemia, hyperkalaemia, normal CSF study along with nerve conduction studies suggestive of motor polyneuropathy with predominantly axonal involvement. Considering these along with generalized areflexia, he was labelled as a case of GBS (Guillain-Barre syndrome) and transferred into intensive care unit and was prescribed intravenous immunoglobulin therapy for five days and antibiotics (meropenem, vancomycin, metronidazole). But after the completion of IVIG therapy his condition improved only minimally. With these complaints he visited at BSMMU. He is a known case of asthma for last two years. He is on salbutamol and budesonide inhaler for two years with occasional exacerbation which required hospitalization for several times. He has no parental consanguinity and none of his family members suffered similar illness.

On general examination he had normal vital parameters, breath sound was vesicular with prolonged expiration with no skin lesion or thyromegaly or lymphadenopathy. Nervous system examination revealed higher cerebral function including speech was normal. Cranial nerves were intact including fundoscopy. There was wasting of all four limbs which was more marked distally with no fasciculation. There was hypotonia, muscle power MRC grade 02 on right upper limb & left lower limb, MRC grade 03 on left upper limb and right lower limb both proximally and distally, areflexia and non responsive planter response. The sensory examination revealed patchy pattern of sensory deficit not following any dermatome or nerve

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pattern. There was no cerebellar or autonomic disturbance, no abnormal spinal curvature or thickened peripheral nerves.

Investigations revealed leucocytosis (25,500/cumm) with marked eosinophilia (41 percent), raised C reactive protein (80.75mg/L), positive p-ANCA (18.5 U/L), positive RA test (59.5 IU/ml), repeat NCS showed severe sensory motor polyneuropathy (both axonal and demyelinating) affecting all four limbs. All other investigations including serum electrolytes, creatinine, muscle enzymes, thyroid function, chest radiology were normal. Because of the asymmetrical pattern of weakness, asthma, marked eosinophilia, p-ANCA & RA positivity, the patient was suspected to be a case of Eosinophilic Granulomatosis with Polyangiitis, and prescribed pulse methylprednisolone (1gram daily for five days) followed by oral prednisolone (1mg/kg/day). Following pulse therapy patient's muscle power improved partially and became MRC 03.But his weakness did not improve any more. So he was reviewed again for any possible alternative diagnosis and sural nerve biopsy was done. It revealed multiple granulomas copmosed of epitheloid cells surrounded by cuffing of lymphocytes in perineurium and blood vessels features of lymphocytic vasculitis. Fitefaraco stain reveals gobli of lepra bacilli in the nerve bundles.

After confirming diagnosis now he was put on anti leprosy treatment which resulted in marked improvements in his conditions. Now his muscle power is MRC grade 04 proximally and MRC 04 distally in all four limbs.

Discussion

The neurological manifestations of leprosy is rare and restricted to the peripheral nervous system, that is peripheral and cranial nerves. The distribution of neural damage depends on the bacterial load within the nerve and on the immunological response mounted by the host against the infected nerves. Involvement of the peripheral nerves can result in several clinical patterns of peripheral neuropathy.

The most common neurological presentation of leprosy is Mononeuritis, and the nerves that are more involved are the ulnar, median, posterior auricular and superficial radial of upper limbs and in case of lower limbs, common fibular, superficial fibular and posterior tibial are affected[4]. Some patients may present with distal neuropathy where there is loss of temperature and pain, muscle weakness due to a mononeuritis multiplex[4, 5]. Leprosy is known to mainly involve only the exteroceptive sensations that is light touch, pain and temperature. Proprioceptive sensory loss that is joint position, vibration has been uncommonly described in leprosy. Cases of symmetrical neuropathies outnumbered cases having mononeuritis multiplex (12:7) in a series of 19 patients in Mumbai, India, presenting with proprioceptive loss in leprous neuropathy[6].

Purely neurological Leprosy is very rare, can occur in up to 4-8% patients, with no associated dermatological involvement [7]. These cases are very challenging to diagnose and often requires histological confirmation by nerve biopsy. Jardim et al. assessed 49 patients with pure neuritic leprosy. According to his observations the most common presentations were paraesthesias(55%), motor dysfunction (24%), neural tenderness (12%), and sensory loss (8%) [8]. Similar clinical picture was seen in this patient, where signs were confined to the peripheral nervous system, that is patient suffered from pain in all limbs followed by loss of sensation, weakness and wasting. There was no clinical feature suggesting spinal cord injury.

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Electrodiagnostic approaces is helpful in evaluating the characteristics of neural damage, to asses wheather they are motor or sensory and nerve conduction studies is considered superior in diagnosing neuropathy than the clinical evaluation. Many authors regarded NCS highly sensitive in these patients[9]. NCS is useful for not only reaching diagnosis but also for detecting and evaluating the extension of leprosy neuropathy .The pattern of nerve conduction abnormalities are variable in Leprosy neuropathy, it can be both axonal and demyelinating. Demyelinating features are observed in earlier part of disease and at the sites of nerve entrapment, such condition is that of the ulnar nerve at the elbow[10]. In later part of disease as there is more nerve damage, axonal pathology is noted[11]. In our patient the nerve conduction study in earlier phase of the disease revealed motor polyneuropathy with predominantly axonal involvement, no significant feature of demyelination and normal CSF study. Later nerve conduction study showed severe sensory motor polyneuropathy (both axonal and demyelinating) affecting all four limbs. This findings were similar to pattern described by different authers over time [12, 13].

Diagnosis of Leprosy requires both clinical and laboratory evidence. According to experts, there are three diagnostic signs of leprosy: skin hypopigmentation with loss of sensation, thickening of peripheral nerves and skin-smear positive for the acid-fast bacilli and it is important to remember that all leprosy lesions are not always devoid of sensation and peripheral nerve thickness is also found in other diseases[14]. The diagnosis of leprosy is confirmed by histopathological studies of patients. Slit-skin smears and skin biopsies demonstrating acid-fast bacteria often provide histopathological diagnostic confirmation. In patients with pure neuritic Leprosy, a nerve biopsy is required to confirm the diagnosis. The histopathological features can determine the subtype of the neuropathy. Findings in patients with robust cell-mediated immunity include well-defined tuberculoid granulomas and nonspecific lymphocytic inflammatory infiltrates with scarce or no bacilli, in patients with poor cell-mediated immunity, granulomas mostly contain vacuolated foamy cells and bacilli and during reactions, intraneural caseous necrosis and micro-abscesses can be seen[15]. Because of asymmetry of presentation and lack of response to steroid, sural nerve biopsy was done in our patient. It revealed multiple granulomas composed of epitheloid cells surrounded by cuffing of lymphocytes in perineurium and blood vessels and features of lymphocytic vasculitis. Fite-faraco stain revealed gobli of lepra bacilli in the nerve bundles.

After receiving standard treatment patient`s condition has improved and his muscle power is now MRC grade 4/5. He is on regular follow up.

Conclusion

Leprosy neuropathy is challenging to evaluate and control as it can develop at any stage of disease without dermatological presentations. The neurological manifestations of leprosy are restricted to the peripheral nervous system. Nerve involvement commonly manifests with a sensory abnormality, which progresses to painful neuritis and finally results in severe sensory motor deficits. Pure neural leprosy requires histological confirmation by nerve biopsy. Patient must undergo full neurological evaluation of the peripheral nerves at different stages so ongoing clinical and neurophysiological patterns of neuropathy can be understood. Recovery from Leprosy neuropathy depends on a few conditions such as point in time of recognition of

disease and treatment of neuritis, number and extent of reactional episodes, and the clinical form of the disease. A failure to detect leprosy early is common and specially in purely neuritic form, which leads to the development of permanent disability from nerve damage.

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The authors report no conflicts of interest related to this study.

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