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Motor Neuron Disease Presenting as Low Back Ache: A Case Report

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Abstract

Motor neuron disease (MND) is a progressive neurodegenerative disorder, manifested by upper and lower motor neuron signs and symptoms. It is characterized by progressive weakness, atrophy, spasticity, dysarthria, dysphagia, and respiratory compromise, ultimately resulting in death. The initial presentation of the disease may be diverse, cramps often being the earliest manifestation of the disease especially in the lower motor neuron component. We present a case of 69 year old female, who presented in the physiotherapy department with complains of low back ache, cramp like pain and weakness in both the legs for the last 6 months. Her neurological examination and investigations confirmed the diagnosis of MND.

Key Words: Motor neuron disease; Low back ache; Cramps.

Introduction

Motor neuron disease (MND), is a chronic neurodegenerative disorder of the motor system. It is characterized by the loss of motor neurons in the cortex, brainstem and spinal cord, manifested by upper and lower motor neuron signs and symptoms affecting limb, bulbar and respiratory muscles. It is a devastating illness which leads to progressive paralysis and eventual death. Death usually results from respiratory failure and follows on average 2 to 4 years after onset, but some may survive for a decade or more. The exact cause of the neuronal loss in MND is not understood. Motor neuron disease is however relatively uncommon with an annual incidence of 2 in 100,000 and prevalence of 5-7 per 100,000 worldwide.[1,2] There are four variants of motor neuron disease. Amyotropic lateral sclerosis is the most common form, with both upper motor neuron (such as spasticity, hyperreflexia, and Babinski responses) and lower motor neuron involvement (muscular atrophy and fasciculation). In progressive bulbar palsy, disease begins in the lower brainstem, causing dysphagia, dysarthria, and dysphonia. Progressive muscular atrophy is again a rare subtype of motor neuron disease (MND) which affects only the lower motor neurons.[1,2] Lastly, Primary lateral sclerosis which only affects upper motor neurons.We report a case of MND presenting as low back ache in the physiotherapy department of our hospital.

Case report

A 69 year old housewife came to the physiotherapy department of our hospital with low back ache since last 6 months. She also complained of cramp like pain and weakness in both the legs. There was history of frequent falls (3-4 times/month). Her symptoms were progressive in nature. The aggravating factors for her low back pain were prolonged standing and walking and the most relieving position for her was supine lying. On examination, her lumbo-saccral movements were restricted in the end range and there was diffuse tenderness in the lumbo-saccral area.The SLR on both the sides was 70^0, because of hamstrings tightness. The FABER's

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test was negative on both the sides. There was distal lower limb wasting seen bilaterally. On manual muscle testing, her lower limb strength was 4/5 proximally (hip & knee) and 3/5 distally (ankle) bilaterally. She had difficulty in heel standing and toe standing. The upper limbs examination revealed bilateral thenar wasting. The pinprick, touch, proprioception and vibration sensations were intact in the upper and lower limbs. The knee and ankle jerks were diminished in both the lower limbs, whereas biceps and triceps jerks were normally elicitable. There was no evidence of aphasia, dysartria, dysphagia, limitation of extraocular movements and visual field deficits. She was continent in bladder and bowel. In view of these findings, the patient was referred to a Neurologist and was subsequently advised MRI L-S spine and EMG/NCV of all 4 limbs. The MRI findings did not reveal any evidence of root compression, except degenerative disc disease at L4-L5 and L5-S1 levels. The nerve conduction was normal in tibial, ulnar and peroneal nerves. NCV could not be done for median nerve because of the atrophy of bilateral abductor pollicis brevis. Sensory potential were normal in sural, ulnar and median nerves. Facial muscles, dorsal paraspinal, C8/T1, T5-6-7, L3-S1 myotomes showed fibrillations and large motor units with reduced interference pattern. The electrophysiological findings were suggestive of motor neuron disease.

Discussion

Low back pain is one of the commonest conditions that we encounter in our everyday physiotherapy practice. Our assessment should focus on neuro-muscular-skeletal assessment in every patient and any unusual finding should not be overlooked. The management of patients should ensure a multidisciplinary approach if there is any doubt regarding the diagnosis. As in this case, the presence of atrophy was unrelated to the complain of low back ache. Therefore it required neurology consultation. The symptoms in MND are diverse and challenging including weakness, spasticity, limitation in mobility and activities of daily living, communication deficits and dysphagia, pain and psychosocial distress, fatigue and sleep disorders.[3] Cramps can often be the earliest manifestation of lower motor neuron component of MND.[2,4,5] Pain may be part of the sensory phenomena noted in few patients or it may arise from abnormal stresses on the musculoskeletal system imposed by weak musculature.[4]

Currently, there is no definite cure for motor neuron disease. Riluzole is the only drug identified to have a beneficial effect on survival. The effect is modest, with a prolongation of life for approximately 3-4 months on average. In MND the focus of management is on symptomatic rehabilitative and palliative therapy, delivered by a multidisciplinary team. Multidisciplinary approach improves care, reduces the frequency and length of inpatient stay, and improves survival.[1,2,3]

The role of physiotherapy in MND is to reduce secondary problems. A marked reduction in activity level secondary to MND can lead to cardiovascular deconditioning and disuse weakness, superimposed on the weakness caused by the disease itself. Reduced physical activity, particularly if prolonged, produces muscle atrophy, reduced strength of tendons and ligaments, osteoporosis, pain and contractures. Exercise programs might have positive physiological and psychological effect on people with MND, especially when implemented before significant muscular atrophy occurs.[4] Breathing exercises and incentive spirometry may improve the respiratory status of the patient. The greatest advancement in respiratory therapy has been the discovery of the beneficial effects of non-invasive ventilation, in which the patient uses a mask ventilator system overnight during sleep. Most patients will eventually have difficulty swallowing. Upright position while eating (upright, no distractions, chin tuck), and dietary modifications in the form of thickened fluids can be helpful. Current practice is to offer the option of enteral feeding to patients

either through percutaneous endoscopic gastrostomy or nasogastric tube when more than 10% of the promorbid weight has been lost.[1,2] Hence, supportive treatment in the form of physiotherapy, respiratory care, nutritional support etc remains the mainstay in the management of patients with motor neuron disease.

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