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Case Report

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A CASE OF MADRAS PATTERN OF MOTOR NEURON DISEASE - CASE REPORT

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ABSTRACT

A case of Madras pattern of Motor Neuron Disease (MMND) is a unique geographically distribution preponderantly reported from Southern part of India that is presented and other forms of Motor Neuron Disease are discussed. This disease affect males and females in an equal rate. Disease condition happened to have wasting confined to distal part of limbs known to be Lower Motor Neuron (LMD)disease and Upper Motor Neuron (UMD) together pyramidal dysfunction. We present a18 years old female presented with atrophy and weakness of left and right legs and is unable to walk for about 3 years who is early diagnosed with MMND. There is no wasting of proximal and trunk muscles and also have normal tone regardless of the presence of extensor plantar response, deep tendon reflexes are elicited. The involved group of muscles have power about 4/5.The patient also had multiple lower cranial nerve palsies and sensorineural hearing loss. With supportive treatment, the patient feels better. Continuing physiotherapy, regular monitoring of sensory and motor system with the disease condition makes a profound effect of patients healthy life.

KEYWORDS: Madras Motor Neuron Disease, Lower Motor Neuron Disease, Upper Motor Neuron Disease.

INTRODUCTION

A sub group of rare motor neuron disease that is originated in 1970 from the Southern part of India and was first described by Meenakshisundaram et al.,[1] Further reports of this case were reported from south Indian city of Bangalore[2,3,4] and also from other Indian cities.[5] Reports of isolated cases from other countries are also available.[6,7,8,9] It is classified into Familial Madras Motor Neuron Disease (FMMND) and Variant Madras Motor Neuron Disease (MMNDV).This disease has a rare younger age onset progressive neuromuscular disease taking relatively benign course. Multipronged approach is required to evaluate various causal factors such as genetic, environmental and also some ethnic factors as well. Here we report a case of MMND with wasting limited more to lower limbs with normal tone and DTR also present with extensor plantar response. This appears to be Juvenile Motor Neuron Disease.

CASE REPORT

A 18 years old female presented with the complaints of atrophy and weakness of left and right legs which is insidious onset and progressive in nature. She is not able to walk herself for about 3 years. On hospitalization the patient had the complaints of cough and cold, headache, fever. No past history of trauma or family history of this condition reported. There is no wasting of proximal and trunk muscles and also have normal tone regardless of the presence of extensor plantar response, deep tendon reflexes are elicited. The involved group of muscles have power about 4/5. The patient also had multiple lower cranial nerve palsies and sensorineural hearing loss. DTR is also present.Extensive work up including blood biochemistry, serology, vasculitic profile, magnetic resonance imaging of brain and spine were normal. Nerve conduction study showed abnormal motor as well as sensory conduction. Needle electromyography (EMG) showed acute and chronic denervation in genioglossus, distal and proximal muscles of all four limbs, cervical and thoracic paraspinal muscles and reinnervation in distal muscles of left upper limb, suggestive of preganglionic lesion, anterior horn cell involvement. Pure tone audiometry showed bilateral mild sensory neural hearing loss. Brain stem auditory evoked response was abnormal and visual evoked response was normal. On arrival at hospital, the patient was given Inj. Ceftrixone 1g, Syrup.Ascoril 50ml, T.Paracetamol 650mg and also with Neb. Duolin twice a day.

DISCUSSION

Motor neurons are the type of cells in the nervous system that conveys impulses which directly or indirectly carry out muscle movements. Motor neurone disease (MND) is the name given to a group of related diseases affecting the motor neurones in the brain and spinal cord. Degeneration to these may leads to weakness and

wasting of muscles, causing increasing loss of mobility in the limbs, and difficulties with speech, swallowing and breathing.[1,2]

Advancement is persistant and generally prompt, with a life expectancy of between two and five years from the onset of symptoms. Approximately 20% of patients can survive for 5-10 years but the rate of progression varies greatly from one person to another. Death usually occurs due to respiratory failure. Observations on postmortem spinal cord reveal an extreme loss of anterior horn cells, demyelination and sclerosis of the ventrolateral columns. Bulbar motor nuclei and cochlear nucleus show neuronal depletion. Inflammatory etiology in MMND is suggested. MMND closely resembles several other disease conditions, including relatively common ALS (amyotrohic lateral sclerosis) and are considered in the differential diagnosis. There are disputes if MMND and Brown-Vialetto-Van Laere syndrome (BVVL) described from Western Europe are similar clinical entities. However, there are several finer clinical features that differentiate MMND from BVVL. There is no specific treatment for MMND is available yet. Intravenous immunoglobulin administered in one case showed moderate improvement in certain symptoms. Yet, immunoglobulin treatment is based on the assumption of inflammatory etiology of MMND, but not a proven treatment. Rarity of MNND in itself is an impediment for evolving a possible treatment. Symptomatic treatment and supportive care, such as offering hearing aids, are part of MMND management that might help maintain patients' routine activities.

CONCLUSION

The positive aspect of this disease is that many patients continue to live for several decades after the onset of the disease. In our case LMN signs are present without CN involvement. Distal part of lower limb is severely involved & regardless of extensor plantar response, universally tone was normal & DTR were just present. Continuing physiotherapy, regular monitoring of sensory and motor system with the disease condition makes a profound effect of patients healthy life. The purpose of this presentation is to feature one more presentation of MND.

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