

A case of Motor Neuron Disease – Case Report

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Abstract- A case of Motor Neuron Disease (MND) with wasting confined to distal part of one upper limb and signs of Upper Motor Neuron (UMN) disease but for normal tone in all four limbs and preserved Deep tendon reflex (DTR).

Index Terms- motor neuron disease, upper motor neuron, deep tendon reflex

I. INTRODUCTION

MND is due to various causes, it is also dividable into various types .the most common one is amyotrophic lateral sclerosis, having both UMN&LMN signs. Here we report a case of MND with wasting limited to predominately to right hand with normal tone and DTR also just present with extensor plantar response. This appear to be a variant of MND

II. CASE REPORT

A 55 yrs old female presented with wasting and weakness of right hand predominately, fasciculations also observed on right side. There was no wasting of proximal muscles and trunk muscles. Power is 4/5 in the involved group of muscles. Tone was normal in four limbs despite the presence of extensor plantar response. Cranial nerves, sensory system & bladder & bowels were not involved. DTR were also just present. All investigations were normal, including complete blood picture, vit-B12 levels, Thyroid function tests, Serum electrolytes, computerized tomography (CT Scan)-Brain, Magnetic Resonance Imaging (MRI)-Cervical spine, Nerve Conduction Velocities (NCV) study-normal.

III. DISCUSSION

MND is a neurodegenerative disease and is arguably the most devastating of the neurodegenerative disorders. Amyotrophic lateral sclerosis (ALS) is the most common form of progressive motor neuron disease. It is a prime example of a neurodegenerative disease. Combined upper & Lower MN features present in Amyotrophic lateral sclerosis. Lower Motor Neuron Disorders are X-Linked Spinobulbar Muscular Atrophy (Kennedy's Disease), Adult Tay-Sach's Disease, Spinal Muscular Atrophy, Multifocal Motor Neuropathy with Conduction Block. Upper Motor Neuron Disorders are Primary Lateral Sclerosis, Familial Spastic Paraplegia. Purely UMN in Brainstem –

Pseudobulbar palsy, Purely LMN in Brainstem - Progressive bulbar palsy. Various newer types of MND have been reported; there were no conduction block in these cases. They are several varieties like MADRAS MND seen in 10% of MND in south india with gradual asymmetry in 50% of cases, gradually progressive involvement of four limbs and progress to ALS, sensorineural deafness, weakness of facial and bulbar muscles. Dr.GOWRI DEVI MND is monomelic type of amyotrophy ; LMN signs in one upper or lower limb; Dr.CHOPRA's MND is wasted leg syndrome; Dr.HIRAMA's MND is upper limb involvement but Brachioradialis muscle is spared.

In our case UMN & LMN signs are present without CN involvement. Distal part of one upper limb is severely involved & inspite of extensor plantar response, universally tone was normal & DTR were just present. The purpose of this presentation is to highlight one more presentation of MND.

REFERENCES

- [1] CMDT -2010
- [2] Davidson's Text book of Medicine 21st edition
- [3] Harrison's principles of Internal Medicine 17th edition
- [4] Brown RH et al: *Amyotrophic Lateral Sclerosis*, 2nd ed. London, Informa Healthcare, 2006
- [5] Pasinelli P, Brown RH: Molecular biology of amyotrophic lateral sclerosis: Insights from genetics. *Nat Rev Neurosci* 7:710, 2006 [PMID: 16924260]
- [6] Kevin Talbot and Rachael Marsden's text book of MND
- [7] Dr Stuart Neilson and Dr Frank Clifford Rose " text book of MND
- [8] Raffaele L. Mancini" text book of MND
- [9] P. N. Leigh"text book of MND
- [10] Christopher G goetz"text book of clinical Neurology 3rd edition.

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