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Motor neuron disease-The Indian Scenario

Shankar Prasad Saha1 and Sinjan Ghosh2

1Institute of Neurosciences Kolkata, India

2NRS Medical College and Hospital, India

***Corresponding author:** Shankar Prasad Saha, Institute of Neurosciences Kolkata, India

Tel: 91 9433249923; E-mail: drspsaha@yahoo.co.in

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Introduction

MND is a progressive disease causing degeneration of the upper and/or lower motor neurons. The average age of onset is between 55 and 65 years according to the Western literature; however, the same has been found to be a decade earlier in the Indian population. There is a male preponderance (M: F-2:1). The frequency of ALS cases in India is 5 in 100,000.

Etiological Factors

ALS is mainly a polygenic disease (70%–90%). C9ORF72, TARDBP, FUS, and SOD1 genes have been implicated in familial ALS cases. Environmental factors such as intense physical activity, cigarette smoking and viral infections play a role in ALS.

Toxins

Dietary factor such as neurotoxin (BMAA) was associated with Amyotrophic Lateral Sclerosis/Parkinsonism Dementia Complex (ALS/PDC) in Guam). Lathyrism is a condition prevalent in India, presents with specific affection of the motor system which is attributed to toxin (BOAA) found in legume Lathyrus sativus.

Regional characteristics

The unique aspects of clinical presentation of Motor Neuron disease were reviewed and regional characteristics in the Indian subcontinent as noted in literature is being compiled. The first study from eastern India was performed by Saha et al (Published in 1997) with 110 cases from July 1993 to June 1995 (0.11% of all neuro cases in OPD).

Amyotropic lateral sclerosis (ALS) constituted 43.6%

Progressive muscular atrophy (PMA) 10.970

Post-polio progressive muscular atrophy (PPMA) l.8% Spinal muscular atrophy (SMA) 20%

Atypical form Madras pattern of MND (MMND) 0.9% Monomelic amyotrophy (MMA) 22.7% of cases

Predominantly younger age of onset and increased propensity in males.

Trauma > Electrocution were the most common antecedent event (ALS & MMA) Optic nerve involvement in 1 case.

A South Indian retrospective review of patients' medical records for clinical manifestations, electromyography, imaging, audiological and histopathology findings were performed in 116 patients over 36yrs (1971-2007). Madras motor neuron disease (MMND), MMND variant (MMNDV) and Familial MMND (FMMND) have a unique geographic distribution predominantly reported from Southern India. The salient features are as follows-

Young age (Mean age of onset was 15.8±7.9 years),progressive but benign course, persistent asymmetrical involvement, lower cranial nerve involvement, bilateral sensor neural deafness, sparing of cognitive and sensory system. Patients with MMND Variant in addition have optic atrophy.

A South Indian study (2008) by Nalini et al analyzed 1153 patients of classical sporadic ALS seen over 30 years for the clinical manifestations and survival pattern. When compared to studies among Caucasians the age of onset was one to two decades earlier and the male preponderance was more. The survival pattern is significantly longer compared to Caucasians who generally have a dismal prognosis. Thus, Indians appear to have a relatively younger age of onset and prolonged survival suggesting the relatively slow course of ALS among Indian patients.

Neurogenic atrophy is restricted to one limb is a heterogeneous disorder, involving one upper or lower limb. Gourie-Devi et al. (1984a, 1986) suggested the eponym “monomelic amyotrophy” (MMA) as a more appropriate term. Upper limb MMA -“brachial monomelic amyotrophy” Lower limb MMA - “crural monomelic amyotrophy” (Gourie-Devi and Nalini, 2003).

Salient features are- insidious onset of atrophy and weakness, onset 2nd or 3rd decade, male preponderance, sporadic occurrence, slow progression , stabilization within a few years, benign outcome, cranial nerves, pyramidal, sensory, cerebellar and extrapyramidal systems are not involved.

North Indian study by Prabhakar and Chopra et al discussed about Wasted Leg Syndrome. The illness was noticed incidentally with a strictly unilateral wasting of the whole lower limb (in 65% of cases)- a) all muscles below the knee (in 22.5%

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of cases), b) Quadriceps muscles only (in 12.5%of cases). 77.5% of the cases were heavy manual labourers. It may be presumed that occupational trauma may in some way be responsible for initiating or aggravating the disease process in majority.

A study performed with 32 native patients of western Himalayas were recruited over 1year (2013-2014) which highlights following findings: -

Male preponderance is less common in these patients as compared to earlier reports from India.

Bulbar onset is more common in elderly (age >60 years) females.

As per previous reports from India, compared to Western population present study reiterates the younger age of onset and longer duration of symptoms and slow course of disease in Indian patients.