



## Case Report

# Madras Motor Neuron Disease –A Rare Tropical Variant

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## ABSTRACT

*Madras Motor Neuron Disease is an anterior horn cell disorder seen in young individuals with prominent cranial nerve involvement and hearing loss. It is sporadic, slowly progressive juvenile onset anterior horn cell disease. In contradiction to classical ALS the clinical course of this disorder is benign and long term survival over decades common. Here we presented a patient with complaints of distal muscle weakness, wasting of upper limbs and sensorineural hearing loss in his medical history. Madras motor neuron disease was diagnosed with neurological examination and EMG findings*

## Introduction

Motor neuron diseases (MND) are a heterogeneous group of disorders affecting the upper motor neuron, lower motor neuron or both. Amyotrophic lateral sclerosis is the most common type of MND, Tropical variants like madras motor neuron disease (MMNDs) are relatively rare.

Madras motor neuron disease can be considered as unique variant of ALS with atypical features like benign course of disease, lower cranial nerve involvement, sensorineural hearing loss

## Case Report

46 year old gentleman from coastal margin of kerala came with history of wasting and weakness of his hands for 30 years and slurring of speech for one year. Wasting was more than weakness and it was symmetrical affecting both hands. The

wasting was first noticed over the dorsal aspect of right and left hand at the age of 16 years. The wasting was gradually progressive and reached a static phase within five year. Weakness affected his palms and there was history of difficulty for buttoning and unbuttoning shirts, making food bolus. The wasting and weakness confined to the palms for several years. There was history of hearing impairment. After thirty years of wasting and weakness of palms patient developed slurring of speech for past one year. He gives history of Pure motor weakness without sensory involvement or bladder involvement.

Nervous system examination revealed dysarthria with predominant affection of linguals and labials. Eighth cranial nerve was affected with positive rinne. Weber was localised to left ear and absolute bone conduction was defective. Tongue

showed extensive fasciculations and wasting with near normal power. Tongue was flaccid on palpation. There was symmetrical wasting of his hands with split hand sign (thenar wasted more than hypothenar), dorsal interossei and lumbricals were severely wasted with poor hand grip bilaterally. Wrist flexors, extensors were weak. Lumbricals, dorsal & palmar interossei were weak, Ext & flexor pollicis longus were weak. No lower limb weakness. Tone was normal. Fasciculations were noted over right deltoid. Superficial and deep tendon reflexes were normal. Reflexes were preserved. Sensory system examination was normal. Gait was normal. Skull and spine normal

including full movement of spine. Other system examination does not reveal any abnormality. Investigations revealed normal routine blood results. BRE, RFT, LFT, RBS, FBS, PPBS, HBA1C, VDRL and viral markers was normal. Peripheral smear-normal TFT, PTH, S. Electrophoresis, Ach and Musk Antibody, S.calcium, S.phosphorous - normal. Serum immunoglobulin -normal Creatine kinase (CK) level was normal ANA including ANA profile -negative MRI spine with brain screening was normal Nerve conduction studies (NCV) showed low amplitude median and ulnar compound muscle action potentials (CMAP).

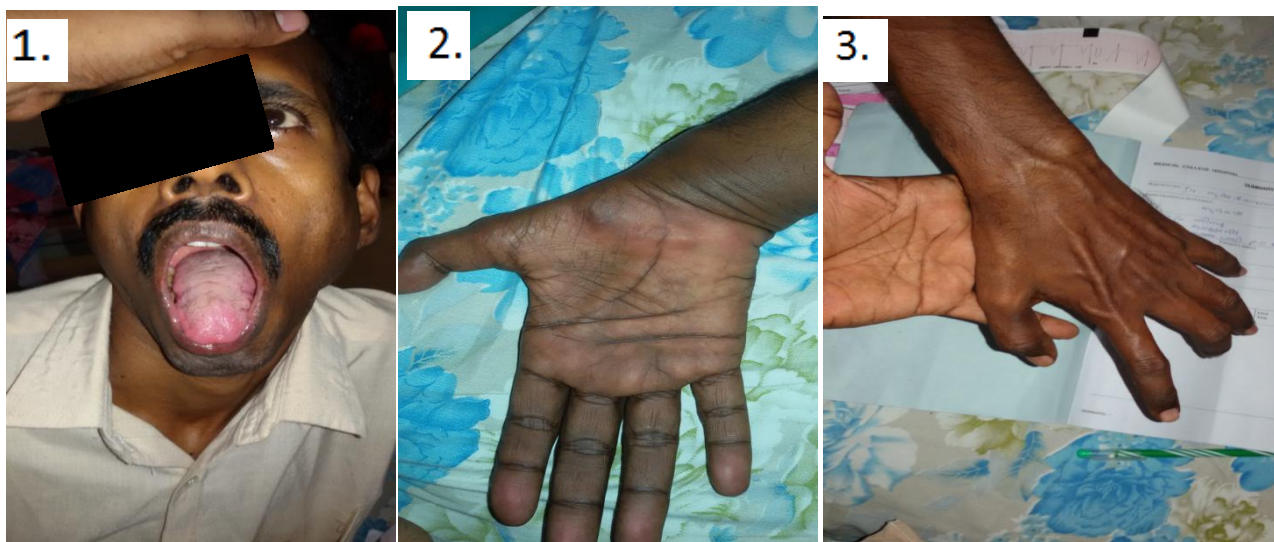


Fig 1,2,3 – Wasting of tongue and hands

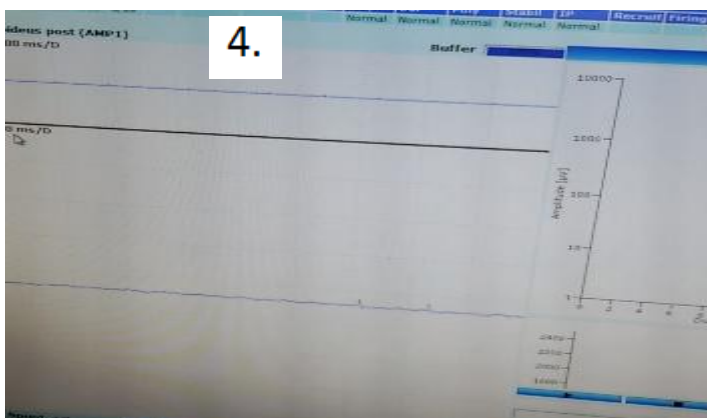


Fig 4 – Fibrillatory waves of right posterior deltoid in EMG showing spontaneous activity

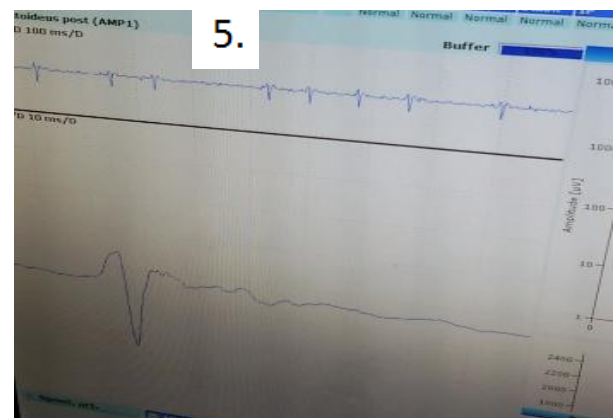


Fig 5 – Positive sharp wave of left deltoid in EMG showing spontaneous activity

There was no electrodiagnostic evidence of conduction block or demyelination.

Median, ulnar and radial sensory nerve action potentials (SNAP) were normal.

EMG showed Positive sharp waves and fibrillation

Lower limb electro physiological study was normal

Pure tone audiometry showed moderate bilateral sensorineural hearing loss

In view of pure motor weakness, wasting of muscles of hands, sensorineural hearing loss, wasting and fasciculations of tongue without any sensory involvement and electrophysiological evidence of motor neuron disease, we arrived at a diagnosis of Tropical variant of motor neuron disease madras motor neuron disease(MMND).

### Discussion

Madras motor neuron disease (MMND) was first described by Meenakshi sundaram et al in 1970. It is an anterior horn cell disease seen in young individuals with prominent cranial nerve involvement and hearing loss. Mean age of onset is 16 years. Males and females affected equally. Initial manifestation include weakness and wasting of distal limb muscles and upper motor neuron signs, a slender body habitus is noted in majority. Audiological hearing impairment is seen in all patients. Bifacial weakness, palatal weakness and tongue fasciculations were comm.-only noted. Deep tendon reflexes are usually brisk but normal or sluggish in some patients. One fourth of patients have optic atrophy in addition which has been called MMND variant syndrome. Cerebellar signs have also been reported in few cases. The syndrome has relatively benign course with over all mean survival of 27 years

### Conclusions

Over a period of fifteen years, the original case series reported by Meenakshi sundaram et al had been expanded to forty typical cases. 70% of this forty patients exhibited clinical signs of lower cranial nerve dysfunction involving 7 th to 12 th

nerves in varying proportions. The presence of UMN signs occur in 65% of cases thus making the resemblance to classical ALS very close. Therefore in most discussions of MND, madras motor neuron disease (MMND) usually finds a place. The absence of reported identical cases from other parts of world, the lack of established neuro pathological correlates of the clinical features, and its rarity has shrouded a cloak of mystery over this elusive entity.