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Case Report

An Unusual Presentation of Motor Neuron Disease with Overlapping Features of both FTD-ALS and Madras Motor Neuron Disease

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ABSTRACT

Madras motor neuron disease (MMND) presents in the young in the first two decades. The majority of the cases reported are from South India. The hallmark of MMND is sensorineural deafness (hearing loss due to nerve defect), atrophy and weakness of the limb muscles, cranial nerve palsies involving cranial nerves VII, IX and XII, facial and bulbar muscles (kind of muscles that control the chewing, swallowing and speech). MMND variant has the additional features of optic atrophy and cerebellar signs. It is a slowly progressive disease. We report a case of motor neuron disease with sensorineural deafness like Madras motor neuron disease variety but with relatively late presentation and rapid progression and associated with frontotemporaldementia-like FTD-ALS variant. So it's a very rare case associated with some features of MMND and some features of FTD-ALS.

Keywords- MMND,FTD-ALS, South India, sensorineural deafness, optic atrophy

INTRODUCTION

Madras motor neuron disease is a sporadic disease occurring in young adults mostly in South India. It presents with slowly progressive asymmetric limb weakness with pyramidal signs, sensorineural hearing loss, facial and lower cranial nerve palsy. Optic atrophy and cerebellar signs may be present.

CASE REPORT

A 37 yr male presented with a behavioral abnormality in the form of increased anger, irritability, forgetfulness in the form of misplacing objects and money followed by apathy, decreased speech output, less interaction with people and executive dysfunction for last 3 yrs. Simultaneously he developed gradually progressive thinning of extremities and weak hand grip. He also developed b/l foot drop. The weakness gradually progressed over 2 yrs to involve the proximal muscles of the upper limb and lower limb. There was a flickering sensation over arms, thighs, and upper back. He had a history of hearing impairment in both ears for last 1 yr that was not associated with tinnitus. There were no sensory symptoms or bladder bowel involvement. There no h/o neck pain or trauma. There was no family h/o similar illness. There was no h/o DM,HTN,TB in the past. On examination his vitals were stable. His MMSE was 16.Lobar function test revealed the involvement of frontal and medial temporal lobe with the improvement of memory with cues. There were sensorineural hearing loss right side and b/l

lower motor neuron type facial weakness. Rests of the cranial nerves were normal. There was wasting of both distal and proximal muscles of extremities. Spasticity was present in the lower limb but hypotonia around the ankle joint. Muscle power in upper limb around 3/5 and in lower limb 4-/5. Bilateral foot drops present. All DTR were brisk. Plantar was equivocal bilaterally. System examination revealed no abnormality. There was the presence of visible fasciculations over arms and thighs.

Fig-1-showing wasting of proximal and distal muscles of the upper limb

Fig-2-showing wasting of proximal and distal muscles of the lower limb

The routine investigations like CBC, RBS, RFT, LFT, lipid profile was normal. HIV Ag was negative. Thyroid profile, it B12 was normal. EEG was normal. Pure tone audiometry revealed severe SNHL right ear and mild SNHL in left ear.MRI Brain and MRI Cervical spine were normal. Nerve conduction study showed motor axonal polyneuropathy. EMG was of neurogenic pattern with spontaneous activity in tongue, deltoid, vastus medialis, 1st dorsal interossei, and thoracic paraspinal muscles.

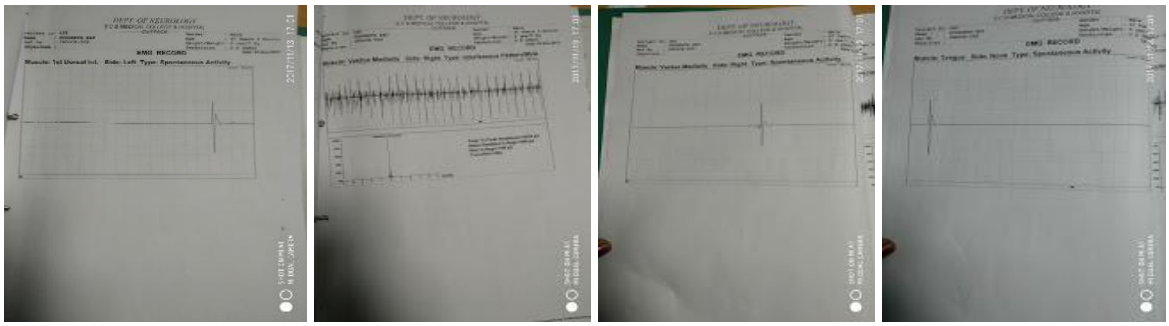

Fig -3- showing spontaneous activity in1st dorsal interossei, tongue, thoracic paraspinal muscles

DISCUSSION

So from the above findings it is clear that it is a case of anterior horn cell disease with 7th and 8th cranial nerve involvement and with frontotemporal dementia. In Madras motor neuron disease the age of presentation is a 1st or 2nd decade and the 8th cranial nerve is commonly involved but the disease is very slowly progressive. But in our case, the disease is rapidly progressive with features of Fronto temporal dementia. In FTD-ALS the age of presentation is late and 8th cranial nerve involvement is not commonly seen.

CONCLUSION

It is a very rare case of motor neuron disease with overlapping features of MMND and FTD-ALS with a relatively late presentation in comparison to MMND.

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