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A Variant of Motor Neuron Disease "Madras Motor Neuron Disease" (MMND)

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INTRODUCTION

A 9 yrs old male child born out of non-consang-uinous parentage presented to our department with complaints of hearing impairment, facial weakness, weakness and clawing of both hands with involuntary movements and speech disturbances, weakness was progressive involving the lower limb also but with benign course & with breathlessness. Electromyography shows chronic partial denervation. Mini polymyoclonus, atrophy and fasciculation of tongue, weakness and atrophy of upper extremities with sensorineural deafness were the important clues to the clinical diagnosis. A sub group of motor neuron disease in the younger age group was described from Madras, Tamilnadu State, India by Meenakshi sundaram et al in 1970.

HISTORY

Presenting complaints are
Deafness since - 3yrs
Facial weakness & deviation of angle of mouth to left - 2yrs
Weakness and clawing of both hands– since 5-6 months
Palpitations & Breathlessness - since 4 months.
Bilateral involuntary movements of hands - since 1 month
Speech disturbances since 1 month

History of Presenting Complaint:

1. Child was apparently well till 6 yrs of age then his class teachers and mother noticed that he responds slowly and inability to hear voices, then his mother noticed facial weakness & deviation of angle of mouth to left, for which they admitted in hospital and deafness doesn't improved and weakness relieved temporarily,No history of ear pain, discharge, trauma. Sensations over face are well preserved.
2. From the last 6 months his mother noticed weakness and clawing of both hands. He is unable to hold objects, mix food, buttoning his shirt which was gradual in onset and progressive, without any sensory loss. No history of Difficulty in combing hair No history of Slippage of chappals while walking ,No history of Tripping and falling, Buckling and falling
3. History of breathlessness since 4 months which is gradual in onset. History of

breathlessness aggravated on lying down, on climbing steps & during daily routine activity also, for which the child had wakening in nights and sits up to relieve it. No history of chest pain, cough, wheeze, vomiting, recurrent respiratory tract infections. No history of pedal edema, abdominal distension, facial puffiness.

4. History of involuntary movements observed in both the hands since 1 month which is present even during rest and aggravated on attempted extension of wrist and fingers.
5. History of difficulty in standing from squatting position since 1 month.
6. History of slurring of speech since 1 month

NO HISTORY OF

Decreased perception of smell, Blurring of vision, decreased vision and ocular movements.

Difficulty in chewing, Nasal regurgitation, difficulty in swallowing, drooling of saliva.

Gait disturbances,Seizures, syncopal attacks, Fever, headache, vomitings, dizziness, neck pain, Loss of appetite, myalgias, rashes over skin.Bladder and Bowel disturbances

OTHER HISTORY

- No similar complaints in past. No history of TB contact, Bronchial asthma, seizures, Trauma, recurrent respiratory infections.
- Two siblings. No similar complaints in family members, no history of consanguinity or early neonatal deaths.
- Uneventful, Delivered spontaneously, no history of birth asphyxia, seizures. Attained milestones as per age.
- Routine immunization followed as per the IAP schedule. BCG scar was found on the left arm.

EXAMINATION

GENERAL EXAMINATION – child conscious, coherent, moderately built & nourished. No signs of Pallor, cyanosis, lymphadenopathy, oedema, clubbing.

SYSTEMIC EXAMINATION OF CENTRAL NERVOUS SYSTEM (CNS):

Higher Mental Functions – well preserved

Cranial Nerves: All cranial nerves are normal except bilateral 7^{th},8^{th}, 12^{th} nerve i.e., facial diplegia, bilateral sensorineural deafness, Presence of fasciculations and atrophy of tongue.

Motor System

There was marked distal weakness of upper limbs in form of atrophy of intrinsic muscles of hand and weakness associated with Minipolymyoclonus and proximal weakness of lower limbs. There was weakness of truncal muscles and diaphragmatic weakness. Muscles of upper limb except hands and lower limb were of normal bulk. Absent biceps, triceps, supinator jerks, Knee & Ankle Tendon reflexes were brisk, with plantars extensors. The sensory examination was normal. No ataxia, nystagmus, coordination deficits. No signs of meningeal irritation. Autonomic Nervous System was Normal. Skull and cranium were Normal

OTHER SYSTEMS

- **CARDIOVASCULAR SYSTEM**

Apex beat: left 5^{th} inter-costal space, ½ inch med.to the mid-clavicular line.

S1, S2 heard.

- **RESPIRATORY SYSTEM**

Bilaterally airway entry present. Bilateral normal vesicular breath sounds present. Tachypnea present

- **GASTRO INTESTINAL SYTEM**

Per abdomen – Soft, No organomegaly, tenderness, Bowel sounds heard

- **INVESTIGATIONS**

✓ Complete blood count – Shows Microcytic Hypochromic Anemia

✓ Chest X-ray – Elevation of Left Hemidiaphragm

✓ MRI Brain, Cervical spine – Normal

- ✓ ENMG Studies – Nerve concuction studies – Neuropathic potentials
  Needle EMG - CMAPs: Small amplitude, SNAPs: Normal
- ✓ Serum fasting Citrate level is low, moderately elevated serum fasting Pyruvate levels
- ✓ An altered Citrate Pyruvate ratio is considered diagnositic.
- ✓ BAER – Reduced.

CONCLUSION

A 9yr old male child with chronic progressive bilateral sensorineural deafness with LMN involvement in upper limb with minipolymyoc-lonus, weakness distally and fasciculation of tongue and UMN involvement in lower limb with exaggerated tendon jerks and weakness Proximally, involvement of multiple lower cranial nerve nuclei (7^{th}, 8^{th}, 12^{th} nerve), evolving bulbar signs like dysphonia & dysarthria with Diaphragmatic weakness, Sporadic Occurrence of the disease. Above features makes a clinical diagnosis of a variant of MOTOR NEURON DISEASE i.e., MADRAS MOTOR NEURON DISEASE.

CASE DISCUSSION

MADRAS MOTOR NEURON DISEASE

Sporadic; Occasional recessive inheritance
Epidemiology- Common in South India, Family history of similar disorder: 2%, Male = Female
Genetics - A8302G mutation in mt- tRNA
Onset Age: Juvenile; Range 2nd & 3rd decade; Mean 16 years
Hearing loss: 53%, Arm weakness: 38%, Facial weakness: Diplegia, Deafness: Sensorineural (30% to 80%), Bulbar involvement (40%), Dysph-agia, Dysarthria Tongue: Weakness; Fascicula-tions, Lower motor neuron signs (85%) Weakness in Limbs: Arms > Legs, Asymmetric, Diffuse: Distal > Proximal, Pyramidal signs (80%) Wasting, Fasciculations, Tendon reflexes: May be brisk in legs, Plantar: May be extensor (50%) Sensory: Normal. Other: Minipolymyoclonus related to fasciculations. Course is Slowly progressive Most remain ambulatory

DIFFERENTIAL DIAGNOSIS

1) Fazio-londe disease
2) Kennedy's disease
3) Juvenile amyotrophic lateral sclerosis.

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