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AMYOTROPHIC LATERAL SCLEROSIS PLUS SYNDROME (COIMBATORE MOTOR NEURON DISEASE)

Neurology

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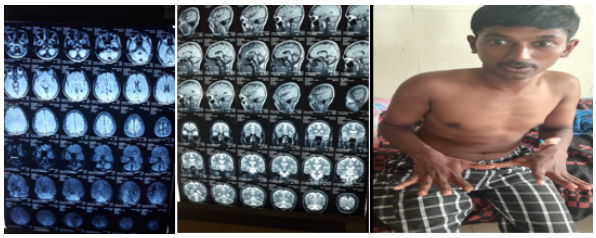
KEYWORDS

CASE REPORT:

26 Year old Male admitted with history of unsteadiness with swaying while walking for the past two years, which is insidious in onset and progressive in nature. It is associated with gradual diminution of vision for past 1year and gradual decrease in speech for two months .General examinations showed fasciculations in shoulder and thigh region associated with wasting of dorsal muscles in hand and foot. Vitals were normal.CNS examination showed Mini Mental Score of 27/30. The visual acuity of both eyes showed perception of fingers and light only, with color blindness. Visual field examination showed field constriction on both sides. Saccades were slow, horizontal and vertical gaze were impaired, oculomotor apraxia was present and fundus were pale with macular dystrophy. Gag reflex were impaired on both sides with uvula in midline with nasal tone of voice. Other cranial nerve examinations were normal. Motor system examination showed bulk was normal, tone increased in all 4 limbs with power 4/5 in all 4 limbs, left side sustained clonus and right side ill sustained clonus present. Plantar bilateral flexor. Sensory, extra pyramidal system is normal. Patient had bilateral axial and appendicular cerebellar signs in the form of truncal ataxia, finger nose in coordination, dysmetria, dysdiadochokinesia, tandem walking impaired with broad based gait. Other systems were normal. Routine lab investigations were normal. MRI brain showed cerebral and cerebellar atrophy. NCS in upper and lower limbs were normal. VEP showed poorly formed wave pattern with both axonal and demyelinating changes. EMG was neurogenic pattern. This patient had features of amyotrophic lateral sclerosis with pancerebellar degeneration and ocular cranial nerves and optic nerves degeneration. We are reporting this rare entity as Coimbatore motor neuron disease, since it has been reported from this part of land.



DISCUSSION:

Degeneration is defined as change of tissue to a lower or less functionally active form Neurodegeneration is the progressive loss of structure and function of neurons including death of neurons. Many neurodegenerative disorders including amyotrophic lateral sclerosis, Parkinsons disease, Alzheimers disease and Huntingtons disease occur as a result of neurodegenerative process.

Amyotrophic lateral sclerosis is a disease in which motor neurons are selectively targeted for degeneration.

Cerebellar degeneration is a condition in which cerebellar neurons become damaged and there will be progressive loss of cerebellar functions.

Optic nerve degeneration results in progressive loss of vision

Ocular cranial nerve degeneration results in progressive loss of ocular muscle function resulting in impaired gaze Amyotrophic lateral sclerosis, also known as motor neuron disease or Lou Gehrig's disease is specific disease causing death of the motor neurons controlling voluntary muscle activity. It is characterized by stiff muscles, muscle fasciculation, gradually worsening weakness of muscles of arms neck and difficulty in speaking or swallowing. Motor neuron disease includes Amyotrophic lateral sclerosis, primary lateral sclerosis, progressive muscular atrophy, bulbar palsy, pseudobulbar palsy and monomelic amyotrophy.

ALS plus syndrome is a set of non motor features with ALS such as extrapyramidal features, ocular motility abnormalities, apraxia, cerebellar features, autonomic dysfunction and cognitive disease. ALS plus syndrome is a multi system disorder. A clinical and pathological feature of ALS plus syndrome extends beyond motor system.

This patient had features of ALS, ataxia, optic neuropathy and occulomotor apraxia. Amyotrophic lateral sclerosis signs include spasticity of all four limbs, exaggerated reflexes, and ankle clonus with bulbar palsy. Ataxia features include truncal incoordination, finger-nose incoordination , finger-finger-nose incoordination, dysmetria, disdiadokokinesia, and intention tremor. Occulomotor apraxia signs include slow saccades, impaired horizontal and vertical gaze. Optic neuropathy signs are pale disc, with hypo attenuated retinal vessels with macular dystrophy.(1,2,3)

In this case pyramidal, cerebellar and cranial nerve degeneration was present without any family history.

Since rarity of these features we are naming this case report as ALS plus syndrome – Coimbatore Motor Neuron Disease. Coimbatore Motor Neuron Disease is characterized by Amyotrophic lateral sclerosis, ataxia, optic atrophy and occulomotor apraxia.

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