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Man in a Barrel Syndrome

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This is a rare variant of amyotrophic lateral sclerosis which presents only with motor weakness of bilateral upper limbs

Introduction:

Man in barrel syndrome (MIBS) also known as the flial arm variant of amyotrophic lateral sclerosis is characterized by brachial diplegia with motor function preserved in legs and facial musculature giving to a patient the clinical appearance of seeming constrained in a barrel. Clinically a patient appears as though they are "stuck in a barrel" with impaired bilateral arm movement and normal facial, cervical, and lower extremity strength.

MIBS can result from bilateral symmetric injury to the brain affecting motor fibres that control arm movement and can also occur following injury to the brainstem, cervical spinal cord, bilateral brachial plexus, or peripheral nerves. It was initially described by JP Mohr in 1969 as a brachial diplegia with normal motor function in the legs and face, giving the appearance of being constrained in a barrel.

History: A male patient of age 75 years, a farmer by occupation presented with complaints of difficulty in using the bilateral upper limb which was flial for more than one year without any weakness in the lower limbs there is no history of difficulty in speech, memory and other higher mental functions, sensory disturbances, autonomic involvement. He also had a history of muscle wasting, fasciculations and postural tremors without the involvement of the bowel and bladder. There was no history suggestive of cranial nerve involvement.

Evaluation:

Systemic examination revealed wasting of both shoulders with power less than 3/5 in the bilateral upper limb, with intact sensory and cranial nerves, and cortical and cerebellar functions. Upper limb reflexes are brisk compared to the lower limb. Polyminimyoclonus was noticed.

Figure: 1

Figure: 2

The complete hemogram, liver and renal parameters, serum electrolytes, viral markers, and thyroid profile were normal.

Imaging such as CT brain and MRI brain-normal; looked for compressive myelopathy, spinal cord ischemia, and degenerative change in the brain, but none of them was seen.

Other imaging modalities, to rule out paraneoplastic syndrome considering the age were done and turned out to be inconclusive

A nerve conduction study showed normal distal latency with reduced CMAP and conduction velocity in bilateral ulnar, radial musculocutaneous and axillary nerves suggestive of motor axonal neuropathy with normal conduction in lower limbs.

EMG showed large amplitude polyphasic action potentials with positive waves and reduced interference suggestive of the neuropathic pattern.

MRI cervical spine with whole spine screening showed significant bony canal narrowing and severe degenerative changes from C4 to C6 with a degenerative fusion of C4 and C5 (acquired block vertebra)

HISTORY: A 41 years old male, farmer by occupation presented with complaints of chronic progressive bilateral upper limb weakness for a period of 6 months without any weakness in lower limbs or sensory involvement. There was no history suggestive of any cranial nerve, bowel or bladder involvement.

Figure: 3

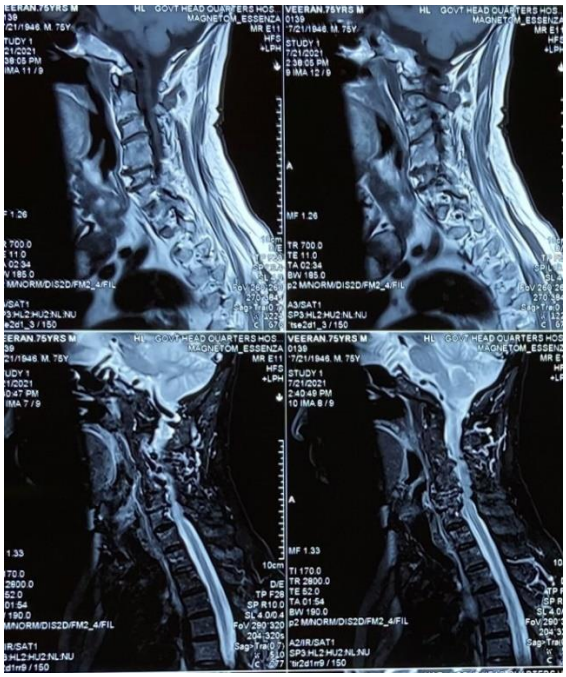

Figure: 4

Evaluation:

Systemic examination showed thinned, wasted upper limbs with a power of 2/5. Hypotonia of the upper limb with brisk triceps reflex was noticed. Sensory, autonomic, cortical, cranial nerves and cerebellar functions were intact.

Figure: 5

Figure: 6

The complete hemogram, liver and renal parameters, serum electrolytes, viral markers, HIV serology, and thyroid profile were normal.

Imaging such as CT brain and MRI brain-normal; looked for compressive myelopathy, spinal cord ischemia, and degenerative change in the brain, but none of them was seen.

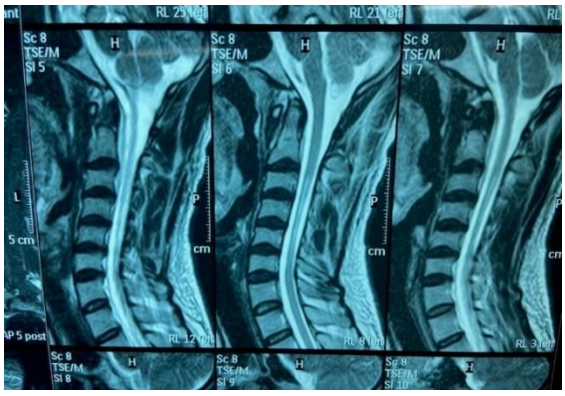

Figure: 7

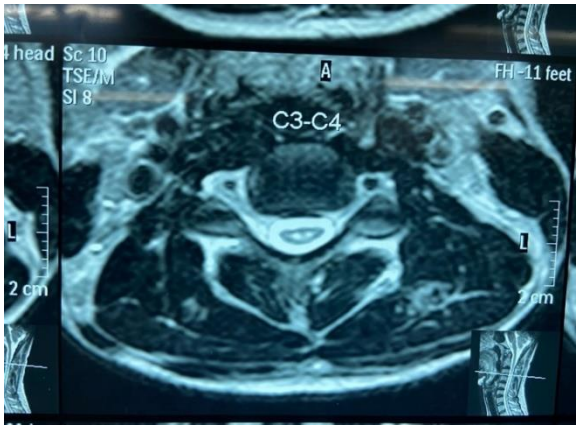

Figure: 8

Other imaging to rule out paraneoplastic syndrome was done and turned out to be inconclusive.

A nerve conduction study showed normal distal latency with reduced CMAP and conduction velocity in bilateral ulnar, radial musculocutaneous and axillary nerves suggestive of motor axonal neuropathy with normal conduction in lower limbs.

EMG showed large amplitude polyphasic action potentials with positive waves and reduced interference suggestive of the neuropathic pattern.

MRI cervical spine with whole spine screening showed significant bony canal narrowing and severe degenerative changes from C4 to C6 vertebra, with **owl eyes sign** in the C3-C4 vertebra.

Management:

Both patients were provided supportive measures and Physiotherapy. The nature of the disease was clearly explained to both patients and advised follow-up.

Discussion:

This is a rare variant of amyotrophic lateral sclerosis, it has male predominance the ratio of male to female being 4-10:1. Damage to the anterior horn cells, such as in HTLV-1 or amyotrophic lateralizing sclerosis, can also present with bilateral upper extremity weakness.

It is characterized by a relatively symmetric involvement of proximal muscles of both arms without muscle weakness of leg or bulbar sites and sparing sensory, bowel and bladder involvement.

Diagnosis is chiefly by clinical examination and supported by radiological investigation, there are criteria for diagnosing ALS which involve AWAJI, EL ESCORIL, GOLD COAST CRCITERIA

Management chiefly involves the symptomatic treatment, physical rehabilitation, speech and communication management nutritional care and home care. The prognosis of the flial arm variant of ALS is significantly better than that of other variants of ALS, in terms of median survival rate.

The differential diagnosis for man in a barrel syndrome includes the following lesions along the neuroaxis:

- Bilateral cerebral upper extremity motor fibers (watershed ischemic stroke, hemorrhagic, traumatic injury, inflammatory, metastatic disease)
- Cervical spine (external compressive lesion, ischemia, inflammatory or infectious process)
- Bilateral brachial plexus (mechanical injury, inflammatory)
- Peripheral neuropathic process (toxic or metabolic neuropathy, inflammatory, auto-immune such as multifocal motor neuropathy or myasthenia gravis)

The prognosis for man in a barrel syndrome depends on the type and location of the lesions. The prognosis for recovery from bilateral watershed strokes is variable depending on the extent of ischemic damage. In comatose patients with MIBS following extensive watershed strokes, survival is less than 10%.

Complications from bilateral intracerebral injury can include cognitive deficits, sensory loss, language dysfunction, weakness, and spasticity. Cervical spinal cord injury complications include weakness, sensory loss, spasticity and bowel, and bladder dysfunction. Bilateral brachial plexus injury complications include motor and sensory deficits in the bilateral upper extremities.

Conclusion:

This is a rare variant of ALS and patients of varying ages were presented with similar signs and symptoms. This is a diagnosis of exclusion, although early diagnostic evaluation is necessary.

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