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Locked In Syndrome, a Rare Clinical Presentation: A Case Report

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Abstract: Locked in syndrome is a rare neurological entity which is usually misdiagnosed as a spinal pathology. Here we report 70 years-old hypertensive and diabetic gentleman presented with quadriplegia without upsetting his conscious level.

Key Words: Locked in, Rare presentation, Stroke, Quadriplegia

Locked in syndrome is a rare neurological entity characterized by complete paralysis of voluntary muscles expect ocular muscles that control eye movement.

2. Case Description:

A 70-year-old long standing hypertensive and diabetic gentleman presented with sudden onset of vomiting for multiple episodes, dizziness, slurred speech and generalized weakness of body, which was more on the right side for one day. He was under regular antihypertensive and oral hypoglycemic medications since last 12 years. On arrival to emergency, his blood pressure was 180/100 mmHg, random blood sugar was 142mg/dl and spo2 98 with high flow oxygen with normal body temperature. He was awake and was obeying commands. His both pupils were reactive and normal in size. There was weakness of power 2/5 in right upper and lower limbs. Emergency CT head revealed no significant intracranial abnormality. He was admitted in intensive care unit with provisional diagnosis of autoimmune disease. He developed shortness of breath, fall in oxygen saturation despite high flow oxygen the following day and was intubated and kept in mechanical ventilator. Though there were no changes in his conscious level and was following commands with normal eye movement in both horizontal and vertical directions. He later developed weakness of all four limbs of power 1/5. CSF analysis was normal. Internal medicine team was also involved and his initial investigations revealed dyslipidemia, leucocytosis, uncontrolled diabetes, renal impairment and enteric fever. In house MRI was nonfunctioning hence he was sent to other hospital for MRI brain stroke protocol. Diffusion restriction was prominent in brainstem involving median Pons and medulla confirming acute ischemic infarction. On the basis of his clinical presentation and neuroimaging, a diagnosis of locked in syndrome was put forward.

3. Discussion

Locked in syndrome is a rare neurological disorder characterized by complete paralysis of voluntary muscles except those muscles controlling the eye movement. This syndrome was 1st describe by Fred

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Plum and Jerome B Posner coined the termed in 1966.[1] Patients are conscious and can think and reason but are unable to speak or move. Communication can be achieved by vertical eye movement and blinking.[2] It has relatively equal frequency among male and female. This phenomenon is most frequently seen in 41-52 years age group. Among them 60% of the cases are due to basilar artery occlusion.[3]. The 2nd largest cause of locked in syndrome after vascular insult is traumatic brain injury. It is either due to thrombotic occlusion of vertebrobasilar tree or shearing injury to the brain stem. [4]

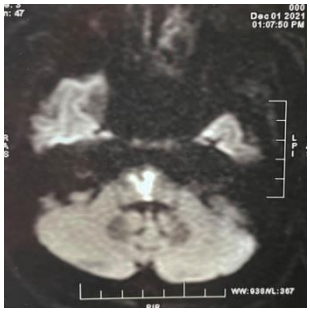

Figure 1: Diffusion restriction image demonstrated acute ischemic infraction in brainstem involving median Pons and medulla

Patient usually presents with quadriplegia and inability to speak in otherwise cognitively intact. They are conscious, aware and can communicate via blinking and moving their eyes due to intact integrity of cerebrum. They may lack coordination between breathing and voice which prevents them from producing voluntary sounds, though vocal cords themselves may not be paralyzed. 3rd nerve nucleus lies ventromedial in the midbrain near the cerebral aqueduct and tracts travel interiorly from the nucleus to innervate eye muscles. Cranial nerves 4 and 6 control downward eye movement via the superior oblique and lateral rectus muscles of the eye affect the ventral pontine lesion. Corticobulbar tracts passing through the midbrain and pons contain upper motor neurons for 5, 7 and 9-12 cranial nerves.[5] Pontine respiratory centers are also affected leading to apnea and respiratory distress. RAS responsible for consciousness lies dorsally and does not affect by ventral pontine lesion. Corticospinal and spinothalamic tracts responsible for limb movement and sensation travel through the crus cerebri of the midbrain and ventral pons hence affected in locked in syndrome.[6]

There are three main types of locked in syndrome.[1] In classic type, patient is unable to move either limb but could perform vertical eye movements, blink and maintains normal consciousness. In incomplete type, classic type with some motor function is preserved. Complete type defined as total immobility including eye movement and only EEG form of cortical function is preserved. Most commonly locked in syndrome is caused by ventral pontine stroke. (**Figure 1**)

This is mainly due to basilar artery or its perforating artery occlusion. Other causes might be bleed, tumors, and infection, trauma and hypotensive events in the brain stem areas. Other caused could be central pontine myelinosis[7], motor neuron disease or demyelination.[8] Other than rapid correction of serum sodium (>12mEq/day), central pontine myelinosis has been reported due to hyperosmolar hyperglycemic condition.[9] Locked in syndrome has also been reported after SARS-coV-2 infection and cervical manipulation by a chiropractor. [4, 10]

Locked in syndrome carries significant morbidity and mortality. They need intensive care, full range of supportive therapy with extensive physiotherapy and rehabilitation for functional improvement. Some papers in the English literature have been reported regarding recovery from this particular phenomenon due to resolution of the edema in pontine infarction and probably due to neuro-plasticity in brain stem.[11]

Conclusion:

Locked in syndrome is a rarely encountered neurological disease with variable underlying causes. This syndrome is usually missed in clinical practice or misdiagnosed as spinal pathology. Hence MRI brain is recommended to confirm the diagnosis.

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