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CASE REPORT

A CASE REPORT ON INCOMPLETE LOCKED-IN SYNDROME

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ABSTRACT

"Locked-in syndrome" is most often caused by a lesion of the ventral pons as a result of occlusion of the basilar artery which essentially interrupts the corticobulbar and corticospinal pathways but spares both the somatosensory pathways, and the ascending neuronal systems. LIS is categorized into three: pure form, incomplete form and total form. There is no cure for LIS, nor is there a standard course of treatment. A 58 year old female with a known case of hypertension for 3 years was brought to our hospital with a sudden onset of weakness of all the four limbs and difficulty in swallowing and the case was diagnosed as Pontine infarction and she eventually developed Incomplete Locked In Syndrome.

INTRODUCTION

"Locked-in syndrome" (LIS) or the deefferented state is most often caused by a lesion of the ventral pons (basis pontis) as a result of occlusion of the basilar artery which essentially interrupts the corticobulbar and corticospinal pathways but spares both the somatosensory pathways, and the ascending neuronal systems. [1] Patients with this syndrome have complete paralysis of all voluntary muscles but the cognitive function is not impaired. They are fully alert and aware about their surroundings, can see, hear and have preserved sleep-wake cycles, and can communicate through purposeful movements of their eyes or blinking or both. But they cannot consciously chew, swallow, breathe, speak, or produce any movements other than those involving the eyes or eyelids. Some affected individuals can move their eyes vertically but not horizontally.[2] Bauer classified the LIS based on the amount of motor response, which are preserved in patients. It may be categorized in three different clinical forms: a) Pure form - the patient loses the control of all body movements with the exception of blinking and vertical eye movements, b) incomplete form - when some voluntary movements and eye movements are preserved. c) total form - when motor functions are completely lost. The final form is the most complicated state since the patients completely fail to interact and express their needs and thoughts.[3,4] There is no cure for LIS, nor is there a standard course of treatment.[5] Given the severity of the neurologic impairment, clinicians are frequently challenged with questions relating long-term morbidity and mortality.[6] In this case report, we are discussing a case of 58 year old female diagnosed as Pontine infarction who eventually developed Incomplete Locked In Syndrome.

CASE REPORT

A 58 year old female with a known case of hypertension for 3 years was brought to our hospital with a sudden onset of weakness of all the four limbs and difficulty in swallowing.

On clinical examination GCS – E4 V1 M6, Muscle Power was 0/5 on all the 4 limbs and multiple cranial nerve palsy was present. She was able to move her eye balls vertically and protrate her tongue slightly outside the mouth. MRI brain report shows Pontine Infarct. In view of severe respiratory distress and signs of aspiration, patient was intubated and connected to a ventilator. She was treated with Anti-platelets, Anti-hypertensives, Statins, Antibiotics and other supportive care.

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Early elective tracheostomy was done. Later the patient was slowly weaned off from the ventilator and connected to BiPAP. Patient was discharged with home BiPAP support. Her prognosis was clearly explained to her attenders.

DISCUSSION

Incomplete variety of LIS resembles the pure variety, except that patient has residual movements beside eyelid and vertical eye movements. Despite the complaints of quadriplegia, dyphasia and preserved consciousness, our patient was able to move her eye balls vertically and able to protrate the tounge slightly outside the mouth indicated that the patient belongs to Incomplete variety.

Infarction of the basis pontis disrupts the corticospinal tracts (producing quadriplegia), Paramedian Pontine Reticular Formation and abducens nuclei (eliminating conjugate horizontal eye movements), and the facial nerves in their course around the abducens nuclei (cause complete facial paralysis). Because the Midbrain Reticular Formation is spared, however, the patient is awake and alert; and because the sensory pathways have migrated to a posterior position in the pons, both the auditory and somatosensory systems are functional. The conjugate vertical eye movements is the only voluntary motor act of which the patient is capable.[7]

Most common etiology is ventral pontine stroke is caused by vertibrobasilar artery thrombosis or occlusion.[8] The other reasons of LIS were varied like also occur due to tumors or encephalitis of the brainstem, traumatic lesions, demyelinating diseases, air embolism, drug intoxication.[1] Our patient has the history of hypertension developed ponitine infarction which ultimately leads to present condition. Jong S. Kim *et al* study revealed that hypertension was the single most common and important risk factor, and the pathogenetic mechanisms of ischemia were likely to be small arterial (lacunar) occlusion or basilar atheromatous branch occlusion in most of the patients.[9] The acute treatment of patients with LIS will similar to patients with other acute brain stem insults. An important aspect is involvement of respiratory system in our patients, is same as in Dr.Murthy TVSP case.[5] Our initial emphasis was on maintaining an airway and adequate oxygenation.

More intensive nursing care and early rehabilitation programs, physiotherapy, respiratory system management, effective system of communication can improve the functional recovery and reduce the mortality rate.[10]

All LIS patients should have to be rehabilitated either in a national or regional specialist centre that has specific multidisciplinary rehabilitation experience . Although the cognitive ability of these patients should not be overestimated, survivor's views regarding the focus of acute treatment, rehabilitation goals, and life choices should be formally sought.[11]

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