

Locked-in syndrome caused by the pressure exerted by the sound gun

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ABSTRACT

A 19-year-old male patient who wounded himself with a gun in the cranial region had a Glasgow coma scale of 3E. At posttraumatic day 7, locked-in syndrome was considered upon detection of vertical eye movements, meaningful winks, and quadriplegia. Apart from the classical view, computed tomography (CT) and postmortem examination of the brain showed an infarct area in the cerebellum. However, vertebrobasilar artery system was normal. In this case report, we would like to present that unlike cases with ischemia, specific CT findings may not be evident in posttraumatic cases and ischemia may occur in the cerebellum as a result of the pressure exerted by a sound gun.

Key words: *Locked-in syndrome, sound gun, trauma*

INTRODUCTION

Plum and Posner defined locked-in syndrome (LIS) for the first time in 1966 as a “combination of quadriplegia and anarthria observed as a result of damage to the corticospinal and corticobulbar pathways in pons.”^[1] The LIS is also known as a pseudocoma or de-efferented state in which patients can sense the environment because their sensory paths are intact however can only communicate with vertical eye movements and blinking. The most common cause of LIS is pontine infarction.^[2] In addition, it is also frequently observed in cases with trauma, tumor, multiple sclerosis, pontine abscess, brainstem encephalitis, and central pontine myelinolysis.^[3-7] The second leading cause of LIS is head trauma. However, since total LIS mimics coma, it is very difficult to diagnose posttraumatic LIS.^[3] Clinical situations that are often confused for LIS are coma, vegetative state, akinetic mutism, and a minimally conscious state.

Motor impairment is divided into three subgroups according to the degree of impairment.^[8] The first subgroup is the classical type in which quadriplegia and aphonia are present but cognitive functions, vertical eye movements, and blinking are preserved. The second subgroup is the incomplete impairment type which, along with vertical eye movements, some voluntary movements can be made. The third subgroup is the total impairment type in which only cognitive functions are preserved.

We are presenting the case of a 19-year-old male patient with a head injury that occurred as a result of sound gun trauma, where increased intracranial pressures lead to cerebellar damage and posttraumatic LIS.

CASE REPORT

A 19-year-old male patient who shot himself at the right temporal region with a gun was referred to our intensive care unit (ICU) on the day of the injury. The patient had a history of unknown recreational drug and alcohol use. He was in a very poor general condition, unconscious, intubated, and at a 3E Glasgow coma score. In addition, there was approximately a 1 cm skin incision matching the entrance wound and soft tissue defects in the right temporal region. Light and corneal reflexes were bilaterally negative. The patient did not display

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spontaneous breathing. His heart rate was 56 beats/min, blood pressure was 80/40 mmHg, body temperature was below 36°C and the central venous pressure was 1 cm H₂O. The initial brain computed tomography (CT) taken at the external center showed bone fragments in the right temporal region along with a wide hemorrhagic contusion area and edema in the right frontotemporal area, temporal lobe inferomedially and midline structures that resulted in a minimal left shift [Figure 1]. The patient was not breathing spontaneously and was intubated when admitted to the ICU and was treated with mechanical ventilation (synchronized intermittent mandatory ventilation, V_T: 540 ml, f: 12/min, I/E: 1:2, positive end-expiratory pressure: 5 cm H₂O). The patient had low blood pressure and central venous pressure and was administered a crystalloid fluid. Despite the adequate fluid resuscitation the desired increase in blood pressure was not obtained, thus dopamine infusion (0.3 mg/kg/h) was started and titrated according to the patient's hemodynamics. Mannitol therapy was started to reduce the cerebral edema and within days its dose was gradually reduced and finally cut off. In addition, the patient was also administered 20 mg furosemide (4 times/day), 10 mg metoclopramide HCl (2 times/day), 20 mg famotidine (2 times/day), and 300 mg acetylcysteine (3 times/day) until the end of the process. The patient's blood biochemistry values were normal (blood gas: FiO₂: 40%, pH: 7.35, pO₂: 62.7 mmHg, pCO₂: 35 mmHg, SaO₂: 93%, HCO₃: 24 mmol/L, BE: 0 mmol/L, glucose: 90 mg/dL, aspartate aminotransferase/alanine aminotransferase: 20/12 U/L, urine: 29 mg/dL, creatinin: 0.6 mg/dL, Na: 141 mmol/L, K: 3.7 meq/L, Cl: 109 mmol/L, Ca: 7.3 mg/dL, Mg: 1.6 mg/dL, albumin: 2.9 mg/dL, hemoglobin [Hb]: 10.7 g/dL, hematocrit [htc]: 32.6%, platelet: 146.000 K/mL, white blood cell: 15.050 K/uL, international normalized ratio: 1.03 mg/dL). Toxicological evaluation did not have any significance. A brain surgeon

assessed the patient's intracerebral hemorrhage due to the head trauma and an emergency operation was not planned.

On the 2nd day of the patient's ongoing follow-up and treatment in the ICU, 0.3 µg/kg/h noradrenaline infusion was added to the dopamine infusion and titrated according to hemodynamics. The patient's nutritional status was assessed and enteral feeding was started. At posttraumatic day 3, the patient started to intermittently open and close his eyes to the point where the blinking became more frequent and more pronounced. Light and corneal reflex was bilaterally positive. However, neither spontaneous limb movement nor responses to pain stimuli were present. A percutaneous tracheotomy was performed on the 6th posttraumatic day. At posttraumatic day 7, LIS was considered upon the detection of vertical eye movements that deviated to the right, meaningful blinking and quadriplegia. The patient's brain CT was repeated. Unlike previous CT scans, the new CT showed minimal pressure in the pons and mesencephalon due to brain edema, partial compression in the fourth ventricle and a hyperdense area of about 1 cm in diameter compatible with a contusion in the cerebellum of the left hemisphere [Figure 2]. A magnetic resonance imaging (MRI) was planned to better define the lesion. However, the MRI was delayed due to the increase in accompanying hemodynamic problems, fever, and infection. During this process, antibiotic treatment was started due to the proliferation of *Chryseobacterium indolegenes* in the endotracheal aspirates. During the patient's follow-up, pulmonary function tests and laboratory findings indicated pulmonary edema and acute respiratory distress syndrome symptoms. The ventilation modes were adjusted accordingly. At posttraumatic day 17, appropriate blood and blood product replacement and omeprazole were administered due to of the finding of melena and a concomitant decrease in Hb (6.2 g/dL) and htc (21.2%) values. The patient died at posttraumatic day 36 due to multiorgan failure. Postmortem macroscopic



Figure 1: The computed tomography of the patient's brain (coronal plane)

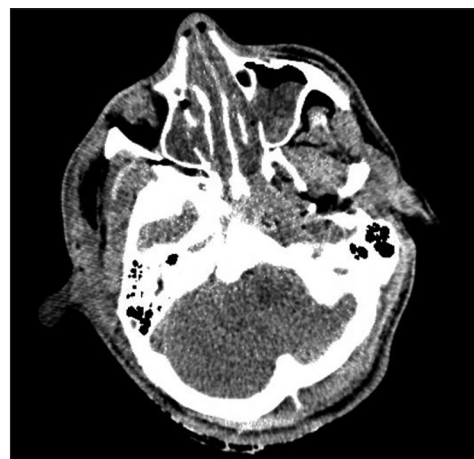


Figure 2: The computed tomography of the patient's brain (axial plane)

examination showed ischemia in the cerebellum; however, the vertebrobasilar artery system was found to be normal [Figure 3].

DISCUSSION

Locked-in syndrome is a condition that can be treated if diagnosed early by establishing communication with the patient and taking advantage of physical therapy techniques.

In LIS, family members are usually the ones who recognize that the patient is awake. León-Carrión *et al.* have reported that LIS is diagnosed on average in 78.8 days.^[2] In our case, the clinical LIS diagnosis was made on the posttraumatic day 7 by the medical team responsible for the treatment. It was pointed out in the literature that the diagnosis of LIS is delayed because it is difficult to detect the transition from a coma to LIS. In addition, there are a limited number of LIS case reports in the literature.^[2,9] In patients with suspected LIS, a CT or MRI can be used for diagnostic purposes. In some posttraumatic LIS cases, CT lesions may be nonspecific and may not always be located in the ventral pons.^[2,3,9] In addition, electroencephalogram and brainstem auditory evoked response may be helpful for diagnosis. In our case, the lesions were located in the cerebellum; further tests could not be done due to deterioration of the patient's general condition. However, clinical diagnosis of LIS was done in an earlier time frame than specified in the literature.

The most common cause of this syndrome is pontine infarction due to basilar artery occlusion. The frequency of improvement after posttraumatic damage is very rare, therefore difficult to diagnose. In the literature, there are isolated reports about the injuries caused by blunt^[10-16] and penetrating^[17] types of trauma, however in these cases vascular damage in the vertebrobasilar artery system and thrombotic occlusion are present. In the case report presented by Ahn and Aarabi,^[18] there was no damage in

the vertebrobasilar arterial system and it was indicated that LIS was caused by ventral pontomeduller contusion. In postmortem studies conducted by Britt *et al.* they indicated that posttraumatic LIS can occur as a result of damage in the pontomedullar junction without vertebrobasilar injury.^[3,19] In our patient, brain CT showed minimal pressure in the pons and mesencephalon as well as lesions in the cerebellum and left hemisphere compatible with a contusion. The postmortem macroscopic evaluation showed that the vertebrobasilar arterial system was normal and that there was widespread ischemia in the cerebellum. For this reason, we suggest that in our patient, LIS did not develop as a result of vertebrobasilar arterial system occlusion as a direct effect of trauma, but rather it developed as a result of ischemia in the cerebellum due to pressure exerted by the trauma.

When the cause of death of patients with LIS was analyzed, pneumonia, respiratory arrest, and respiratory failure with pulmonary embolism were among the most common causes.^[3,7,19] The other causes include respiratory failure, enlargement of the brain stem lesions, cardiac complications, sepsis, gastrointestinal hemorrhage, widely disseminated intravascular coagulation, and pontine abscess. In our case, the patient died at posttraumatic day 36 following pneumonia and gastrointestinal hemorrhage.

As a result, we suggest that careful clinical follow-up and neurological examination in patients in the ICU could allow for a faster diagnosis of LIS. In addition, in posttraumatic cases that are not like ischemic patients, specific CT findings may not be observed and the pressure produced by the sound gun may cause ischemia in the cerebellum, which may lead to the formation of LIS.

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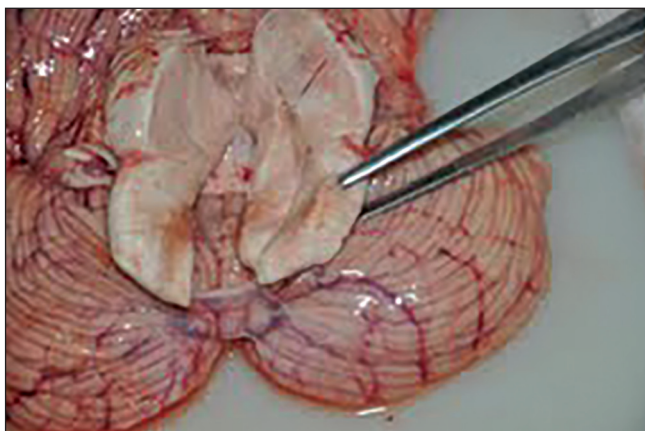


Figure 3: The postmortem view of the patient's cerebellum

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