

Posterior Fossa Subdural Hematoma Resulting in Locked-in Syndrome: Case Report

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BACKGROUND AND IMPORTANCE: Locked-in syndrome (LIS) is a well-known and devastating clinical entity, of which stroke is the most common cause; the distant second, trauma, usually results in LIS from basilar artery dissection. Our case report describes a posterior fossa subdural hematoma causing LIS, likely by direct compression of neural structures, which is a unique etiology and prognosis compared with other causes.

CLINICAL PRESENTATION: A 34-year-old female experienced a posterior fossa subdural hematoma. She was taken emergently for evacuation, and on postoperative examination was found to be locked-in. The symptom complex, while classic for LIS, resolved at an accelerated rate compared with previous reports. Emergent evacuation of clot and rehabilitation were performed.

CONCLUSION: Prompt treatment allowed this patient to experience an accelerated and more complete recovery compared with the ischemic causes of LIS.

KEY WORDS: Locked-in syndrome, Subdural hematoma, Trauma

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The “locked-in syndrome” (LIS) is a well-characterized syndrome, first described more than 200 years ago by Darolles, a French physician,¹ and given its current name in 1966 by Plum and Posner.² Its principal features include quadriplegia, anarthria, and lower cranial nerve palsies with preserved consciousness. LIS is caused by an insult to the brainstem; classically the ventral pons, although bilateral cerebral peduncle lesions have also been described.³ The etiology is generally infarction, with trauma, tumor, central pontine myelinolysis, drug toxicity, and vaccine reactions being less common causes.³ Trauma is generally considered to be the second leading cause of LIS, although very few cases are present in the literature.⁴ We report a case of LIS caused by a posterior fossa subdural hematoma.

CASE REPORT

The patient is a 34-year-old female who was admitted to our facility after a fall backward from

ABBREVIATIONS: LIS, locked-in syndrome; PTD, posttrauma day

standing, during which the back of her head hit a curb and she experienced loss of consciousness. She was taken to a local emergency department with a reported Glasgow Coma Scale of 15 (E4-M6-V5), ambulatory, and neurologically intact, but while in the emergency department, experienced a seizure and rapidly became unresponsive. She underwent rapid sequence intubation at the outside hospital and was transported to our facility as a trauma page. She was noted to have a Glasgow Coma Scale of 3 (E1-M1-V1 T); however, her pupils were reactive bilaterally. A computed tomography (CT) scan of the brain revealed a subdural hemorrhage in the posterior fossa under the tentorium and extending to the base of the cerebellum (Figure 1). The brainstem and cerebral aqueduct were compressed, resulting in obstructive hydrocephalus. A ventriculostomy was placed while the operating room was prepared, then the patient was taken to the operating room for evacuation of the subdural blood through a suboccipital craniectomy, shown in a postoperative CT image (Figure 2).

The patient’s neurological examination improved postoperatively; she opened her eyes to voice and moved them to command, but was unable to move her extremities, and on cranial nerve examination was noted to have bilaterally reactive and equal pupils, bilateral sixth nerve palsies, upgaze palsy, no corneal reflexes, facial symmetry but no movement of facial muscles to command, and a positive gag reflex and twelfth nerve palsy. Sensation in all 4 extremities was

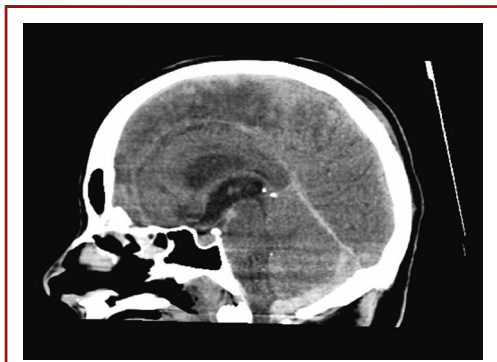


FIGURE 1. *Sagittal, noncontrasted computed tomography scan showing posterior fossa subdural hematoma with mass effect on the cerebellum and compressing the brainstem.*

intact. She demonstrated consistency of neurological examination, and no fluctuation of vigilance was noted, although she was pharmacologically sedated in the acute setting.

Because of her neurological examination, magnetic resonance imaging (MRI) of the brain was performed, which revealed abnormal signal intensity in the posterior pons, greater on the left, with extension into the left middle cerebellar peduncle (Figure 3). No diffuse axonal injury or other lesions were discovered at the cortical or subcortical level. There was no restriction on diffusion-weighted imaging. MRI of the cervical spine revealed no signs of acute trauma on short T1 inversion recovery or T2 sequences. Electroencephalogram, performed on posttrauma day (PTD) 7, revealed posterior rhythms of approximately 7 Hz, which were suppressed with eye opening (Figure 4). A diagnosis was made of posttraumatic LIS.

Attempts were made to wean the patient from the ventilator, but these were unsuccessful, and the patient underwent tracheostomy and gastrostomy tube placement on PTD 7. After removal of the endotracheal tube, the patient remained unable to mouth words, although she regained some movement of the tongue, thereafter communicating with tongue and horizontal eye movements to family and medical staff. By



FIGURE 2. *Sagittal, noncontrasted computed tomography scan showing suboccipital craniectomy and evacuation of subdural hematoma.*

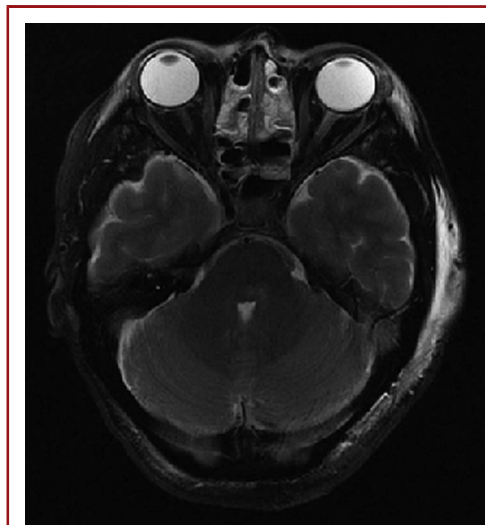


FIGURE 3. *Axial, nonenhanced T2-weighted magnetic resonance imaging demonstrating subtle abnormal signal intensity in the posterior pons, extending into the left middle cerebellar peduncle.*

approximately PTD 8, the patient had regained her corneal reflexes and was able to wrinkle her nose to command. She developed very trace withdrawal of the extremities to pain by PTD 10, and was able to move her toes to command by PTD 12, with extremity strength improving in a proximal to distal progression. Her ventilator and ventriculostomy were weaned, and she was able to be transferred from the intensive care unit. She was able to move bilateral fingers to command at PTD 18. She remained unable to vocalize despite downsizing and occlusion of the tracheostomy, although she could mouth simple, one-word answers to questions. She did not demonstrate pathological vocalizations such as cries or laughs. The patient gradually regained strength in her left upper and lower extremities, with continued weakness on the right. She was able to be discharged to a nursing home for continued rehabilitation and care on PTD 42. On 2-month follow-up, she was revealed to be alert and oriented, with full strength of the left upper and lower extremity and continued comparative weakness of the right side of 3/5. This weakness improved by 8 months postinjury.

DISCUSSION

Although trauma is considered the second most common etiology of LIS, it is a distant second after infarction with only 15 cases being present in the literature.⁴ These posttraumatic cases are generally due to traumatic basilar artery dissection and stroke, or hyperextension injury causing direct injury to the brainstem.⁵ Posttraumatic cases of LIS are generally thought to have a better outcome because of the younger age of the patient population (reflecting the generally improved outcomes of younger patients after traumatic brain injury) and possibly because of fewer medical complications.⁶ Cases of direct trauma to the brainstem may account for the reports of transient LIS, in which shearing or compressive injury allow more complete recovery than frank

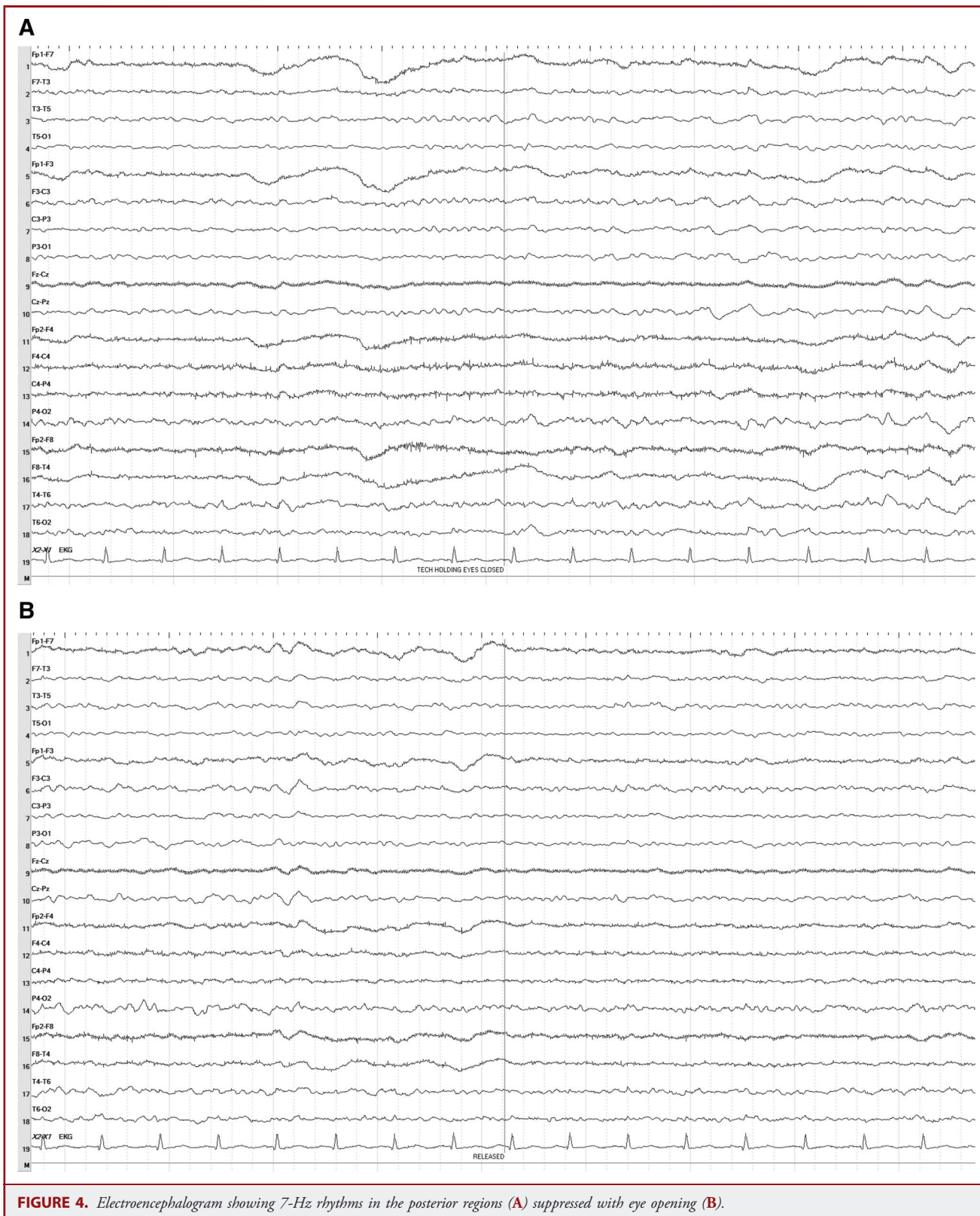


FIGURE 4. Electroencephalogram showing 7-Hz rhythms in the posterior regions (A) suppressed with eye opening (B).

ischemia. This is most often caused by hyperextension of the neck causing shearing trauma to the ventral pontomedullary junction in a high-energy trauma such as a motor vehicle accident.⁵

Both partial and complete recovery after LIS has been reported.^{5,7,8,11} Major return of neurological function generally occurs within 6 months of insult.⁴ According to Chia,⁹ return of horizontal pursuit eye movements is predictive of further improved outcome. In regard to improved motor function, Richard et al¹⁰ determined that improved strength occurred in a proximal to distal progression. Both of these phenomena were present in this case.

Two cases of transient LIS after trauma were reported: by Chang and Morariu⁵ in 1979 and Carrai et al⁸ in 2009. The first was a 25-year-old male after a motor vehicle accident, who was initially comatose and gradually increased in alertness more than 1 month later, with motor function improving gradually approximately 2.5 months after his accident.⁵ Like our patient, he first regained horizontal movement of his eyes, motor function improved in a proximal-to-distal progression and demonstrated asymmetry of strength, with his left side improving more quickly than his right.⁵ Unlike our patient, his level of consciousness remained poor for up to a month after his injury, and he demonstrated fluctuation of alertness.⁵ This patient's presentation was postulated to result from traumatic hyperextension of the head causing direct trauma to the pontomedullary junction, although this was able to be confirmed only by auditory evoked potential at the time of the case.⁵ More recently, Carrai et al⁸ reported on a 56-year-old male, also status post-motor vehicle accident, who was initially thought to be comatose, but who demonstrated electroencephalogram evidence of only a mild consciousness disorder on PTD 6, and demonstrated ability to follow commands by PTD 17. He began showing return of motor function by 4 weeks posttrauma and 1 year later was able to ambulate with a spastic gait.⁸ Unlike our case, this patient initially demonstrated a "total" LIS, in which eye and facial movements were absent and only electroencephalogram findings could differentiate his condition from a coma.⁸ This patient's first movements were in distal muscle groups; however, like our patient and the other previous case, he demonstrated asymmetry of his motor recovery.⁸ MRI in this patient showed diffuse axonal injury in the bilateral cerebral peduncles.

An additional notable difference between our patient and the 2 previously reported cases lies in the mechanism of injury and likely pathophysiology. Our patient was neurologically intact

after her fall, and only demonstrated an LIS after compression of the brainstem from accumulation of subdural blood. The expeditious removal of compression may account for her minimal MRI abnormalities as well as her rapid recovery.

We report what is, to our knowledge, the first case of traumatic LIS caused by a surgically treatable lesion: a posterior fossa subdural hematoma. Our patient demonstrated brainstem compression, both directly from the clot, and also from downward compression of obstructive hydrocephalus. She was taken to the operating room emergently. After evacuation of the hematoma, she demonstrated a clear case of LIS, with her improvement of neurological functioning markedly accelerated in comparison with previous cases in the literature. The prompt treatment of the surgical lesion, within an hour of presentation, may have limited neurological injury and had a positive impact on the outcome in this patient, allowing rapid improvement of her neurological function in comparison with patients in whom no treatment is possible after traumatic insult.

Disclosure

The authors have no personal financial or institutional interest in any of the drugs, materials, or devices described in this article.

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