Published online: 2019-08-06

Locked-In Syndrome after Trauma—A Rare Case in the Literature

Jennyfer Paulla Galdino Chaves[1] Pedro Henrique Mezadri[2] Thiago Suzuki[1] Carlos Alexandre Twardowsky[1] Tatiana Von Hertwig Fernandes Oliveira[3]

[1]Department of Neurology, Cajuru Hospital, Curitiba, Brasil
[2]Pontifícia Universidade Católica do Paraná (PUC-PR), Curitiba, Brasil
[3]Hospital Cajuru, Curitiba, Brasil

Address for correspondence Jennyfer Paulla Galdino Chaves, MD, Neurosurgery Resident, Department of Neurology, Cajuru Hospital, Curitiba, Brasil, Av. São José, 300 - Cristo Rei, Curitiba - PR, 80050-350, (e-mail: jennyfergaldino@hotmail.com).

Indian J Neurotrauma 2018;15:74–77

Abstract

The present report is about a 25-year-old woman who had a motorcycle accident with brain trauma injury. Although she was admitted at another institution disoriented but alert, she progressively evolved to an altered level of consciousness and required orotracheal intubation. She was then transferred to our hospital sedated, and the admission head computed tomography showed signs of posterior fossa ischemia. The angiography demonstrated a vertebral artery dissection with subsequent embolization to the basilar artery, which provoked its complete occlusion. After drug withdrawal, the patient recovered level of consciousness but was aphasic and did not exhibit purpose movements apart from eye blinking, as expected in patients with locked-in syndrome.

Keywords
- locked-in syndrome
- vertebral artery dissection
- trauma brain injury (TBI)

Introduction

The locked-in syndrome (LIS) was originally described in 1966 by Plum and Posner and consists of a complex neurological condition resulting from a brain stem lesion, more specifically the ventral pons, with complete injury of the corticospinal and corticobulbar tracts bilaterally.[1-5] As a result, this syndrome presents with quadriplegia and anarthria with preserved consciousness. Since the supratentorial structures are generally intact, the only possible movements are the verticalization of the eyeball and/or eye blinking, which become the only form of interaction with the external environment.[1-6]

Locked-in syndrome is divided into three different forms: the classical form is characterized by complete immobility except for ocular movement; the incomplete form by mobility of just a few muscles (hand fingers, for example); and the total or complete form by paralysis of all muscles.[1,3,6]

The most common and most described cause of LIS is a vascular or ischemic injury of the anterior region of the pons. In traumatic conditions (10%), LIS can occur due to direct brain stem lesion, secondary to injury or occlusion of the vertebrobasilar system, or by compression of the cerebral peduncle by tentorial herniation.[1,3,6]

In most cases, the diagnosis of LIS still depends on clinical and imaging criteria, but the information is not always concordant.[1,5] Furthermore, the correlation between lesion topography and clinical presentation is often challenging because the neurological evaluation may be compromised by sedation, predominantly in the acute phase.[1]

Case Report

A 25-year-old woman had a traumatic brain injury in a motorcycle versus car accident. She was initially sent to a primary level institution and was admitted alert but disoriented (Glasgow Coma Score 14), without any other alteration in her physical examination. During transportation to our hospital, she became comatose and had to be intubated and sedated. At her arrival, she was sedated and intubated, RASS-2, with miotic pupils but with ocular bobbing. The admission computed tomography demonstrated posterior fossa ischemia (►**Fig. 1**) and in the angiography, that was realized 1 day after admission, a vertebral artery recanalized (►**Fig. 2**) plus

Dr. Jennyfer Paulla Galdino Chaves's ORCID is https://orcid.org/0000-0003-2453-5205.

received
January 5, 2019
accepted after revision
March 26, 2019
published online
August 6, 2019

DOI https://doi.org/10.1055/s-0039-1694694
ISSN 0973-0508.

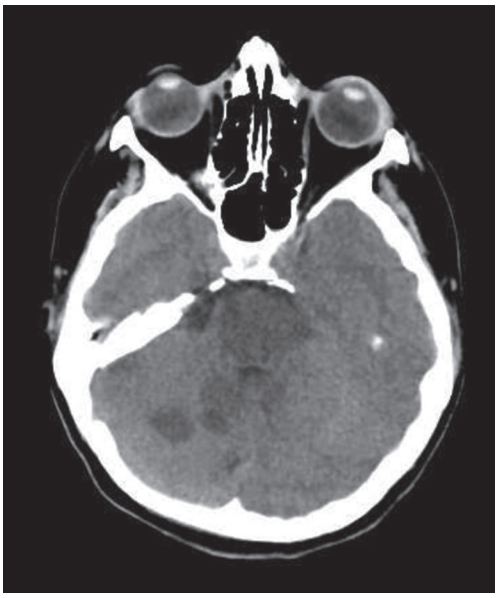

Fig. 1 Computed tomography showing posterior fossa ischemia.

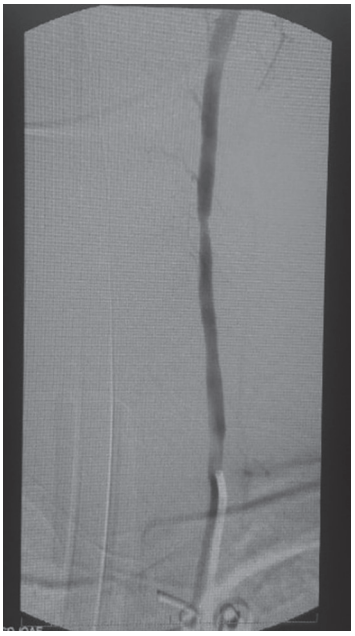

Fig. 2 Angiography showing vertebral artery dissection already recanalized.

a total occlusion of the basilar artery after the anterior inferior cerebellar artery (AICA) emergence (►**Fig. 3A**, **B**). The video electroencephalogram (EEG) was compatible with a waking state. She underwent treatment with dual antiplatelet therapy (clopidogrel plus aspirin). After sedation withdrawal—48 hours after admission—the patient recovered consciousness and was able to interact through vertical eye movements and blinking, but remained quadriplegic and aphasic.

Discussion

Locked-in syndrome is a neurological condition composed of tetraplegia and anarthria with preservation of consciousness, with the maintenance of vertical eye and eyelid movements, as a consequence from a bilateral interruption of corticospinal, corticobulbar, and inferior cranial nerves, sparing the supratentorial structures and the reticular system.[1-6] Mortality in the first 4 months reaches 87% in vascular cases, but may be reduced if the diagnosis and rehabilitation are initiated precociously.[1,7] After clinical stabilization, survival may reach 83% in 10 years and 40% in 20 years.[1,7]

The clinical picture should increase suspicion of LIS when the patient presents with quadriplegia and anarthria with preserved consciousness and cognition,[1-6] although ophthalmoplegia and reduced consciousness can be found during the acute phase of the injury.[1,3,5] Blurred vision, balance disturbance, dysarthria, bilateral paresthesia, or motor weakness may demonstrate the progression of posterior circulation symptoms that can lead to oclusion.[6] The most common etiology of LIS is ischemia of the ventral region of the pons, generally due to basilar artery thrombosis (75–80%) or other conditions such as central pontine myelinolysis, multiple sclerosis, trauma, encephalitis, tumors, pontine abscess, subarachnoid hemorrhage, basilar artery vasospasm, drug toxicity, severe or prolonged hypoglycemia, and iatrogenicity.[1,3]

Differential diagnosis with other conditions that may trigger complete paralysis with preservation of consciousness such as peripheral polyneuropathy (Guillain–Barré syndrome), amyotrophic lateral sclerosis in the ventral horn of the spinal cord, and cerebral cortex diseases should be made.[1,3] Furthermore, conditions that may mimic LIS but evolve with impaired consciousness should also be discriminated. These

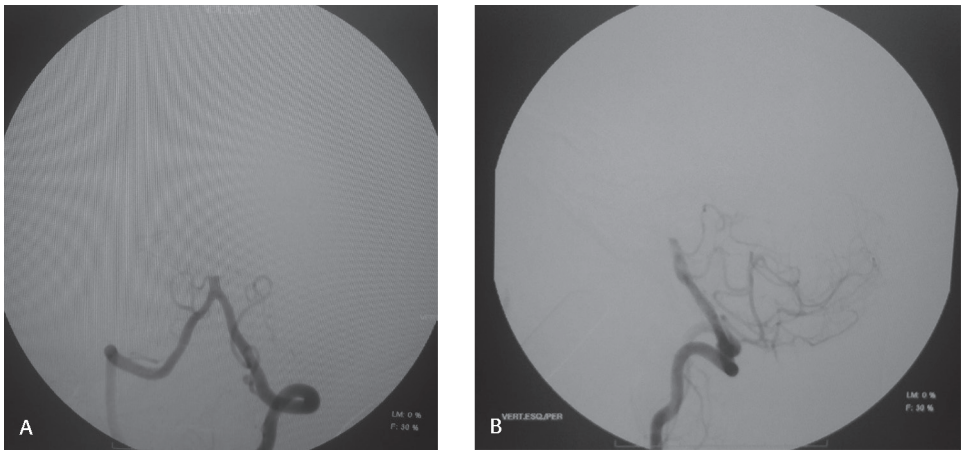

Fig. 3 (**A, B**) Angiography showing a total occlusion of the basilar artery after the AICA emergence.

include coma, vegetative states, and akinetic mutism, among others.[1,2,3,6] This differentiation, however, may not be straightforward as confounders such as altered cognition and ocular movements, which can be mistakenly interpreted as reflective in decerebrating patients, are usually found.[3] Studies show an average of 2 to 2.5 months between brain injury and correct diagnosis of LIS.[6,8] Patient characteristics such as age, previous cognitive status, previous neurological conditions, or other comorbidities, as well as the type and size of the ischemic lesion, are variables to consider in order to obtain an accurate diagnosis.[6]

The use of neuroimaging is fundamental to distinguish between cases with or without cognition impairment.[4,6] Its importance in LIS also lies in the fact that recent studies have shown cortical disturbance, which may have a cognitive and emotional impact on these patients.[4,9] This impairment is related to the sudden interruption of efferent corticospinal and corticobulbar tracts and the cortical and subcortical pathways reaction to the injury.[4]

Vertebral artery dissections (VADs) correspond to 20% of cases of cerebral infarction in patients under 45 years and LIS is the most feared complication, occurring in 10 to 15% of basilar artery occlusions (BAOs).[3,10,11] BAO is a serious condition that presents with 90% mortality in the first 4 months.[3,10] VADs are divided into spontaneous or traumatic, the latter being more common in young patients, younger than 45 years.[8,12] The incidence of traumatic lesion of the vertebral arteries varies on the literature, but may affect 0.5 to 2% of all polytraumatized patients and up to 20% of those with cervicocranial trauma.[8,11,12]

Recanalization of acute BAO significantly reduces mortality. Treatment can be made by intravenous or intraarterial thrombolysis, with similar outcomes in terms of survival and resolution of neurological symptoms, or by endovascular therapy.[13-15] Conservative management may be achieved as well with the association of anticoagulation with intravenous heparin, induced hypertension, and intravenous tissue plasminogen activator.[14,16]

Supportive care should also be provided as it is one of the mainstays of the treatment of these patients. Pulmonary rehabilitation should be optimized during the acute phase of the disease. Physical therapy for the control of secretions is critical for the disease, especially in the first 4 months of hospital stay, as well as pulmonary hygiene and respiratory pattern monitoring.[1,10,17] When respiratory problems are detected, early ventilatory support should be considered. In addition, consciousness is preserved, so it would be important to develop a definitive form of communication, an aim to avoid confusion and frustration for the patient, due to constant changes in the means of communication.[17]

Likewise, psychological support is required, as there is a significant increase in mood swings and depression among LIS patients. More than 25% of the patients might demonstrate suicidal ideation, a fact that negatively impacts quality of life.[14] Although 7% reported the desire of committing euthanasia, a study by the French Association for Locked-in Syndrome surprisingly showed that 72% of LIS patients considered themselves satisfied with their lives in a cohort of 65 patients despite the dramatic clinical picture.[13] This brings to light the importance of an adequate emotional and psychosocial support to improve the quality of life of these patients.

The communication adversities for LIS patients are still a challenge and various technologies have tried to overcome this condition. Brain–computer interfaces[18] bring the possibility of decoding human intentions from brain signals, creating a new communication channel for patients with severe motor impairments.[18] Maybe, with this technology, patients may have the chance to express themselves more appropriately and improve their quality of life.

Conclusion

Although rare, LIS should be considered a differential diagnosis when evaluating aphasic patients with quadriparesis that respond to stimulation with vertical ocular movements. The diagnosis is based primarily on clinical examination and radiologic findings of pons ischemia. Treatment can be made by intravenous or intra-arterial thrombolysis or by endovascular therapy in cases of BAOs. Besides that, pulmonary rehabilitation, physical therapy, and psychological support are essential for rehabilitation and to prevent complications.

Conflicts of Interest
None.

Funding
None.

Ethical Statement
The authors confirm that ethical approval and informed consent was secured during the conduct of this study.

References

1 Bruno M, Nizzi M, Laureys S, Gosseries O, Consciousness in the Locked-in Syndrome. In: Laureys S, Gosseries O, Tononi G, eds. The Neurology of Consciousness. 2nd ed. San Diego, CA: Elsevier; 2015;187–199
2 Kolić Z, Kukuljan M, Vukas D, Bonifačić D, Vrbanec K, Franić IK. Locked-in syndrome in a patient with acute obstructive hydrocephalus, caused by large unruptured aneurysm of the basilar artery (BA). Br J Neurosurg 2017;31(6):738–740
3 Kotchoubey B, Lotze M. Instrumental methods in the diagnostics of locked-in syndrome. Restor Neurol Neurosci 2013;31(1):25–40
4 Pistoia F, Carolei A. The role of neuroimaging in the diagnosis, prognosis and management of disorders of consciousness and locked-in syndrome. Open Neuroimaging J 2016;10:20–22
5 Desouza RM, Crocker MJ, Haliasos N, Rennie A, Saxena A. Blunt traumatic vertebral artery injury: a clinical review. Eur Spine J 2011;20(9):1405–1416
6 Surdyke L, Fernandez J, Foster H, Spigel P. Differential diagnosis and management of incomplete locked-in syndrome after traumatic brain injury. Case Rep Neurol Med 2017 Doi: 10.1155/2017/6167052 [epub ahead of print]
7 Rousseau MC, Baumstarck K, Alessandrini M, Blandin V, Billette de Villemeur T, Auquier P. Quality of life in patients with locked-in syndrome: evolution over a 6-year period. Orphanet J Rare Dis 2015;10(1):4-11 [Internet]

8 Thakral P, Jyotsna, Tandon P, Dureja S, Pant V, Sen I. Radiation dose to the occupational worker during the synthesis of ^{188} re-labeled radiopharmaceuticals in the nuclear medicine department. Indian J Nucl Med 2018;33(1):1–5
9 Lindsberg PJ, Mattle HP. Therapy of basilar artery occlusion: a systematic analysis comparing intra-arterial and intravenous thrombolysis. Stroke 2006;37(3):922–928
10 Al-Raweshidy YH, Sinha DM, Coward LJ, Guyler PC, O'Brien A. Locked in and out: a case of emerging basilar artery obstruction secondary to vertebral artery dissection thrombolysed with intravenous rt-PA. BMJ Case Rep 2011;2011:10–13
11 Idicula TT, Joseph LN. Neurological complications and aspects of basilar artery occlusive disease. Neurologist 2007;13(6):363–368 [Internet]
12 Patterson JR, Grabois M. Locked-in syndrome: a review of 139 cases. Stroke 1986;17(4):758–764
13 Bruno MA, Bernheim JL, Ledoux D, Pellas F, Demertzi A, Laureys S. A survey on self-assessed well-being in a cohort of chronic locked-in syndrome patients: happy majority, miserable minority. BMJ Open 2011;1(1):e000039
14 Pistoia F, Cornia R, Conson M, Gosseries O, Carolei A, Sacco S, et al. Disembodied mind: cortical changes following brainstem injury in patients with locked-in syndrome. Open Neuroimag J [Internet]. 2016;10(Suppl-1, M3):32–40
15 Johnson TM, Romero CS, Smith AT. Locked-in syndrome responding to thrombolytic therapy. Am. J Emerg Med 2018 ;36(10):1928.e5–1928.e7
16 Ojemann JG, Leuthardt EC, Miller KJ. Brain-machine interface: restoring neurological function through bioengineering clinical neurosurgery. Clin Neurosurg 2007;54:134–136
17 Acharya V, Chandrasekaran S, Nair S. An interesting case report of vertebral artery dissection following polytrauma. Int J Surg Case Rep 2016;28:196–199 [Internet]
18 Janjua N, Wartenberg KE. Meyers PM, Mayer SA. Reversal of locked-in syndrome with anticoagulation, induced hypertension, and intravenous t-PA. Neurocrit Care 2005;2(3):296–299