

Locked-In Syndrome: A Case Report

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Key Words

Electroencephalography
Locked-in Syndrome
Magnetic Resonance Imaging

INTRODUCTION

The term "locked-in" was introduced by Plum and Posner¹ to denote the state of quadriplegia and mutism with preserved consciousness, usually demonstrated by communication via intact vertical eye movements. The locked-in syndrome (LIS) is usually caused by an infarct of the ventral pons² or bilateral infarct of the midbrain.³

We report a case of LIS with ventral pontine infarction shown by Magnetic Resonance Imaging (MRI). The EEG and brainstem auditory evoked potentials (BAEPs) and somatosensory evoked potentials (SEPs) of the patient were all normal.

CASE REPORT

A 46-year-old man with hypertension developed right hemiparesis and dysarthria. After 2 days, he developed rapidly progressive quadriplegia and inability to speak and swallow. Consciousness was not altered. The patient understood spoken and written commands. On admission, his blood pressure was 160/90 mmHg, pulse rate was 86/min and respirations were 27/min. He was mute and quadriplegic, but could communicate voluntarily through eye movements and blinking. Pupils were equal and reacted to light. Corneal reflexes were present, fundoscopic examination was normal. The palate was immobile and the pharyngeal reflex was absent. He was never able to move his jaw, face, or tongue. Tendon reflexes were brisk and Babinski signs were present bilaterally. Pain sensation was perceived on the face and in all limbs.

Lumbar puncture revealed normal CSF findings. Computerized Tomography (CT) scans demonstrated bilateral median and paramedian pontine hypodense areas. MRI showed low

signal intensities (T₁ - weighted TR/TE 500/70 msec) high signal intensities (T₂ - weighted TR/TE 2500/90 msec) within the ventral pons, more marked on the right side (Figure 1 A, B).

EEG showed a normal background of 8 to 9 cps reactive alpha activity. There were no focal, lateralizing or paroxysmal patterns (Figure 2).

Somatosensory evoked potentials (SEPs) from median nerve and brainstem auditory evoked potentials (BAEPs) were recorded during the acute and chronic phases of the clinical course. Both SEPs and BAEPs showed no abnormality. (Figures 3, 4).

Left-sided strength improved in the subsequent 4 weeks. The patient's speech was dysarthric, but functional. He had full strength on the left, and had slowly improving strength on the right side 3 months later.

DISCUSSION

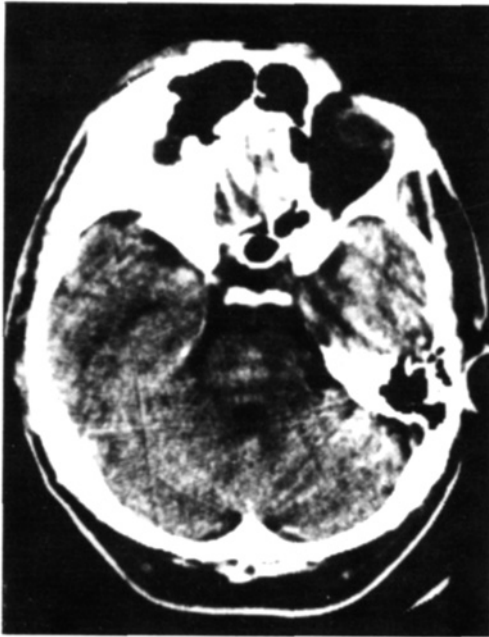
Most cases of LIS are caused by basilar artery occlusion resulting in brainstem infarction in the ventral pons.^{2,4} While there may be no prior history of hypertension or vertebralbasilar insufficiency, there are often premonitory symptoms or evidence of evolving vertebralbasilar ischemia.⁵

Since the corticospinal and corticobulbar tracts traverse the ventral pons, infarction in this region disrupts the voluntary motor supply to the limbs and medullary motor nuclei, resulting in quadriplegia, often with loss of articulation, swallowing, and facial movement. In our patient, MRI demonstrated an infarction in a small hypointense area within the ventral pons.

The EEG of the patient was normal and reactive of altering stimuli. BAEPs and SEPs recorded several times were also normal. Some authors reported abnormal background

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A.



B.

Figure 1.
CT scan and MRI showing bilateral infarcts in ventral pons
A) CT.
B) MRI (T₂ - weighted, TR/TE 2500/90 msec)

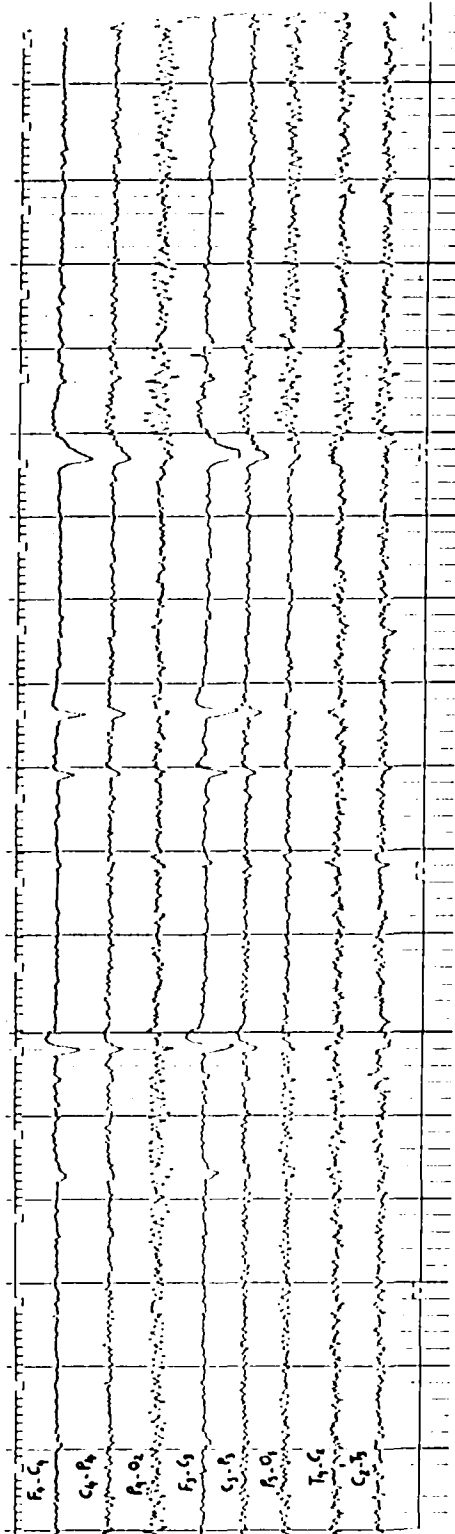


Figure 2.
EEG showing reactive background alpha activity.

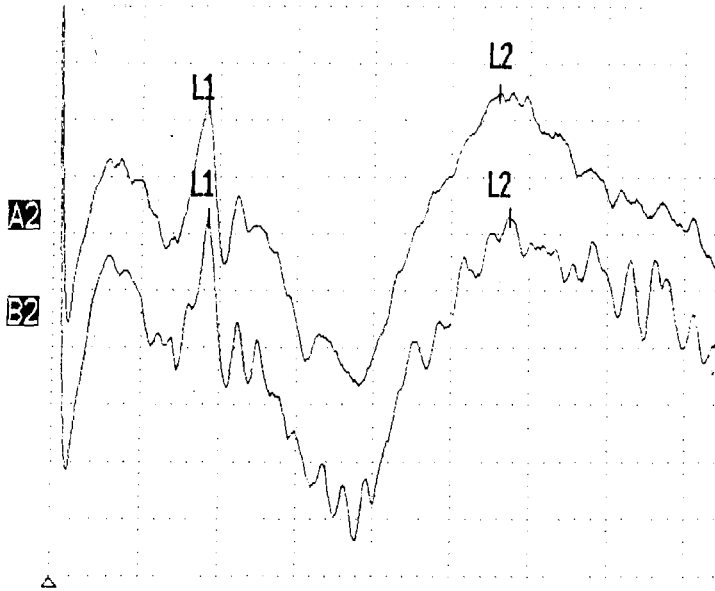
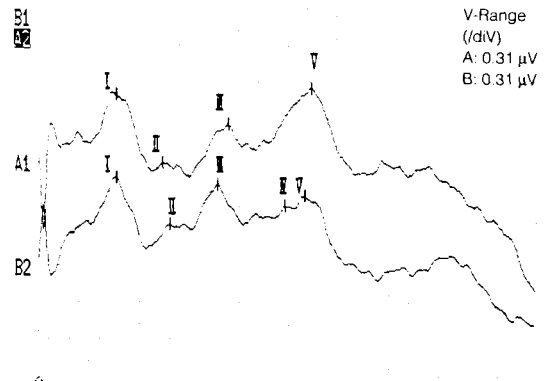
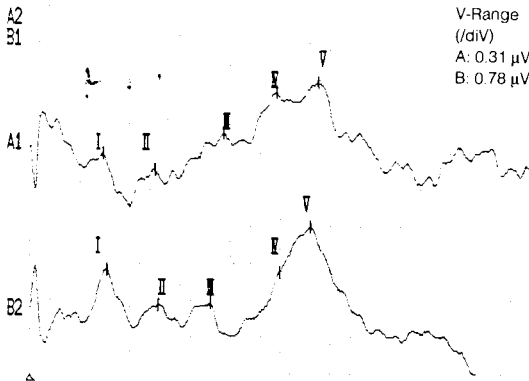


Figure 3. Median Nerve Somatosensory Evoked Potentials recorded from C3 - A1 and C4 - A2. Latency and amplitude values are within normal limits. (18.8 msec, 9.2µV)

Analysis 10 ms
 Electrode (-) - (+) 1: C3-A1, 2: C4-A2, Stim A: 17.2 mA, B: 17.2 mA
 L1 L2 L2-L1
 A2 - 18.4, 56.0 ms; 0.26 µV
 B2 - 18.4, 57.4 ms; 0.03 µV



Analysis 10 ms
 Electrode (-) - (+) 1: A1-Cz, 2: A2-Cz,
 Stim A: L, 95 dB, B: R 95 dB

Analysis 10 ms
 Electrode (-) - (+) 1: A1-Cz, 2: A2-Cz,
 Stim A: L, 95 dB, B: R 95 dB

	I	II	III	IV	V
A.	A1-1.46,	2.50,	3.86,	4.92,	5.76 ms
	B2-1.54,	2.56,	3.60,	4.98,	5.60 ms
	Interval		Amplitude		
	I-III	I-V	III-V	III-I	III-IV
	A1-2.40,	4.30,	1.90 ms;	0.18,	0.34 µV
	B2-2.06,	4.06,	2.00 ms;	0.28,	0.26 µV

	I	II	III	IV	V
B.	A1-1.54	2.46,	3.78,		5.46 ms
	B2-1.54,	2.60	3.56,	4.92,	5.32 ms
	Interval		Amplitude		
	I-III	I-V	III-V	III-I	III-IV
	A1-2.24,	3.92,	1.68 ms;	0.25,	µV
	B2-2.02,	3.78,	1.76 ms;	0.06,	0.18 µV

Figure 4. Brainstem Auditory Evoked Potentials recorded from Cz - A1 and A2 with 95 dB rarefaction click stimulation. Latency and amplitude values are within normal limits.

activity in EEG in LIS. The most frequent abnormality reported is the presence of slowing over the temporal or frontal areas, or more diffuse slowing.^{4,6} Thus, the presence of a relatively normal rhythm that is reactive to some form of altering stimuli (i.e., EEG of waking state) in a patient who appears to be comatose could indicate the possibility of a "locked-in state."

SEPs from median nerve were normal in our patient. Cutaneous sensation is frequently preserved, because the spinothalamic pathway, which lies dorsolaterally, is spared.⁷ BAEPs were also normal. Brainstem auditory pathways traverse the pontine tegmen laterally. Gilroy et al reported a slightly delayed wave V in the brainstem auditory evoked potentials of two patients with LIS.⁸ In one LIS patient with a brainstem lesion medial to the lateral lemnis-

cus and rostral to the trapezoid body and superior olives, the auditory brainstem potential latencies were normal.⁹ Neurophysiological studies reveal that the lesion may have broader functional implications than may be gauged from the neuroradiological evidence.

MRI offers a clear anatomic delineation of the posterior fossa and brainstem due to absence of artifact arising from the petrous bone. It may show well-delineated ischemic lesions some time after the event.

SUMMARY

A 46-year-old man developed a "locked-in" syndrome due to ventral pontine infarction. MRI showed abnormal signal intensity regions in the ventral pons. The EEG, BAEPs and SEPs were all within normal limits.

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