A CASE OF LOCKED-IN SYNDROME IN A PATIENT WITH BASILAR ARTERY THROMBOSIS AND PONTINE INFARCTION

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[Introduction]

The Locked-in syndrome (pseudo-coma) describes patients who are awake and conscious but no producing speech, limb or facial movements. Acute ventral pontine lesions are its most common cause. We describe a case of ischemic lesion in the basal pons, due to thrombosis of the basilar artery in the middle portion. The patient remained comatose for some days, needing artificial respiration; under anticoagulant therapy gradually wake up, remaining paralysed and voiceless.

To recognize this condition it is critical from a prognostic and diagnostic point of view, because superficially this patients recall a vegetative state or akinetic mutism.

Key words: Basilar artery thrombosis - pontine infarct - quadriplegia

The aetiology is multi-factorial, with the lesion in the ventral pons. The most frequent cause is the basilar artery occlusion or a pontine haemorrhage. Another relatively common cause is of post-traumatic nature (Fitzgerald et al., 1997; Golubovic et al., 2004).

Further, LIS has also been reported in sub-arachnoid haemorrhage, secondary to vascular spasm of the basilar artery (Landi et al., 1994); in brain stem tumours (Inci and Ozgen, 2003; Cherington et al., 1976); in central pontine myelinolysis (Lilje et al., 2002); in encephalitis (Acharya et al., 2001) and in pontine abscess (Murphy et al., 1979).

The cardinal signs of LIS include tetraplegia, lower cranial nerve palsy, and anarthria, with preserved consciousness.

We present a case of LIS, emerging after a period of coma, subsequent an acute stroke with underlying thrombosis of the basilar artery.
Case report

A 62 years old man was admitted in a unconscious state after an episodic dizziness and blurred speech with abrupt onset. Early he experimented also headache, nausea and vomiting. In the past history there was only diabetes and hypertension.

Neurological examination revealed: GCS 4/15; horizontal gaze palsy, without nystagmus or ocular bobbing; bilateral motor weakness with reduced reflexes; bilateral Babinski’s sign. Fundus oculi was normal, and no bruit was heard over the carotid. There were no signs of meningeal irritation.

An urgent computed tomography (CT) scan of the head was performed, showing a basilar artery with “hyper density” in the lower and middle sections. The control CT scan showed a low density area in the ventral pons, due to an infarction (fig. 1).

Discussion

LIS is categorized as classical, incomplete, or total (Bauer et al. 1978). According to this classification, our patient emerged from the coma, one week after stroke onset, into a “classical” LIS.

The main behavioural evidence of conscious awareness was the ability to execute vertical eye movements and blinking to command. One month after stroke onset, the patient’s condition evolved into incomplete LIS on the basis of his recovery of lateral head movements.

Brain structural imaging (CT) shows an isolated lesion (bilateral infarction) with extensive destruction of pontine basis. Permanent facial, oropharyngeal, and bilateral limb paralysis is explicated on the basis of descending cortical motor pathways involvement.

Consciousness was recovered because the reticular formation in the dorsal pons is unaffected by the lesion; ocular vertical movements are preserved because the lesion is caudal to the nucleus of the III cranial nerve.

EEG recordings show a predominance of “reactive” slow alpha (8-9 Hz) with mixed theta activity modestly present in the temporal anterior and frontal leads (fig. 2). This EEG findings are reported in literature (Patterson and Grabois, 1986).

We present an usual case of locked-in syndrome in a patient with vascular brain damage. The purpose for discussion and presentation of this case is to remind the diagnosis of LIS. Unless the physician is familiar with the signs and symptoms of the LIS, the diagnosis may be frequently missed and the patient may be erroneously considered in coma, vegetative state, or kinetic mutism. Of course is difficult to recognize unambiguous signs of conscious perception of the environment and of the self in severely brain-injured patients unable to speak. In view of the high margin of error for early misdiagnosis of LIS, serial neurological examinations combined with neuropsychological assessments adapted for use in nonverbal patients with quadriplegia are essential to accurately ascertain the level of consciousness and cognition (Laureys et al., 2005).

Indeed, the necessary substrate for LIS is bilateral paralysis despite preserved consciousness. It has been stated that long-term survival in LIS is rare: mortality is high in acute LIS (76% for vascular cases and 41% for nonvascular cases), with 87% of the death occurring in the first 4 months (Patterson and Grabois, 1986). Motor recovery of LIS of vascular origin is very limited (Doble et al., 2003).
References


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