INCOMPLETE LOCKED-IN SYNDROME

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ABSTRACT

In this paper, we present a case of 62 year-old male with quadriplegia, dysarthria and preserved consciousness. Our case belongs to the incomplete variety of locked-in syndrome due to pontine infarction, because beside the vertical eye movements and eye lid movements, the patient had horizontal eye movements and finger movements on the right side. In general, the basilar artery occlusion is associated with poor outcome, however the urgent thrombolytic therapy may increase the chances to survive. The prognosis of our patient was disquieting and he died after 6 weeks from the onset.

Key words: locked-in syndrome, basilar artery thrombosis

BACKGROUND

The term "locked-in syndrome" (LIS) was initially used by Plum and Posner in 1966 (1), for patients whom maintains consciousness, but loses all voluntary movements, except for vertical eye movements and movements of eyelids.

This syndrome is caused, most commonly, by an infarct of the ventral pontine, due to occlusion or thrombosis of the basilar artery, which interrupts the corticospinal and corticobulbar fibers to the lower cranial nerves, sparing the reticular activating system. Other causes have been cited in the literature, such as tumors or encephalitis of the brainstem, traumatic lesions, central pontine myelinolysis, multiple sclerosis, drug intoxication (2,3). Three categories of LIS have been described by Bauer (4): classical, incomplete, total. Classical form is with quadriplegia, anarthria and preserved consciousness and vertical eye movements. In incomplete variety, patient has residual movements beside eyelid and vertical eye movements. The total variety is composed by patients who are totally immobile and unable to communicate.

CASE PRESENTATION

A 62 year-old right-handed Caucasian male with a history of hypertension, diabetes mellitus, thyroid goiter operation and traumatic brain injury 5 years ago was admitted to our hospital with quadriplegia and dysarthria. The relatives revealed that the patient have been presented headache, nausea, vomiting, dizziness, and abrupt onset of left hemiparesis, when he was admitted to the local hospital. The brain computed tomography (CT) scan was normal. During the hospitalization the patient's status was stationary, but 25 days later suddenly became tetraplegic, dysarthric, dysphagic and that was the reason of his transfer to our clinic. On admission the patient was obese, afebrile, normotensive, in sinus rhythm, with 75/min heart rate, with wet rales at the lung auscultation. He was somnolent, anarthric but conscious, responding to the verbal command with eye movements (vertical and horizontal), eye lid blinking and right hand fingers movements. The neck was supple, the pupils were equals and reactive to light, with left central facial palsy, bilaterally horizontal nystagmus and severe

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dysphagia for fluids and solids. The deep tendon reflexes were diminished and Babinski sign was present bilaterally. The sensory examination was difficult to perform. The laboratory findings, in exception of triglyceride (221 mg %), were unremarkable. The cranial MRI scan showed post traumatic left frontal laceration and an extended lesion in T2 and T2 flair hypersignals at the level of the pons (Figure 1). The magnetic resonance angiography (MRA) demonstrated no blood flow in the vertebral arteries and in the basilar artery and the posterior cerebral arteries were filling from posterior communicating arteries (Figure 2). The carotid artery Doppler ultrasonography one day later revealed mild atherosclerotic plaque bilaterally, with 20% stenosis of the right common carotid artery. The Doppler ultrasonography of V1, V2 segments of vertebral arteries revealed blood flow bilaterally, with 33,7/10 cm/sec maximal velocity at the right side and 31,3/9, 5 cm/sec at the left side. *These hemodynamic findings* were not suggestive for basilar artery occlusion (Figure 3). The transcranial Doppler examination of the distal vertebral arteries and basilar artery was not performed *because* of *technical difficulties due to the local anatomy (short and thick neck) and severe state of the patient.*

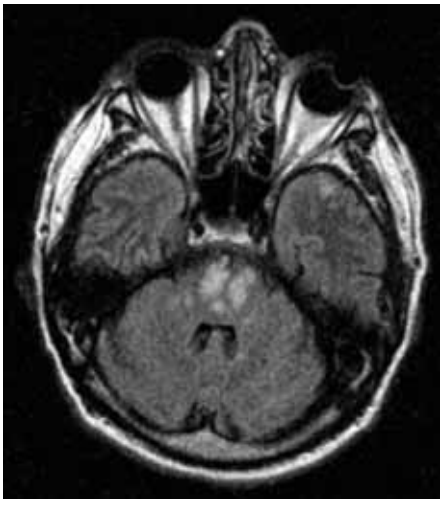

Figure 1. *MRI show acute and severe pontine infarction involving more than two thirds of the pons.*

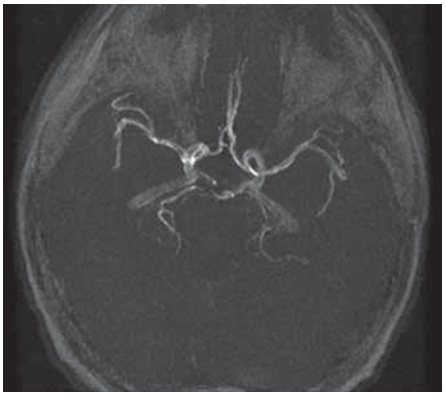

Figure 2. *Brain MR angiography showing no signal in vertebral and basilar arteries.*

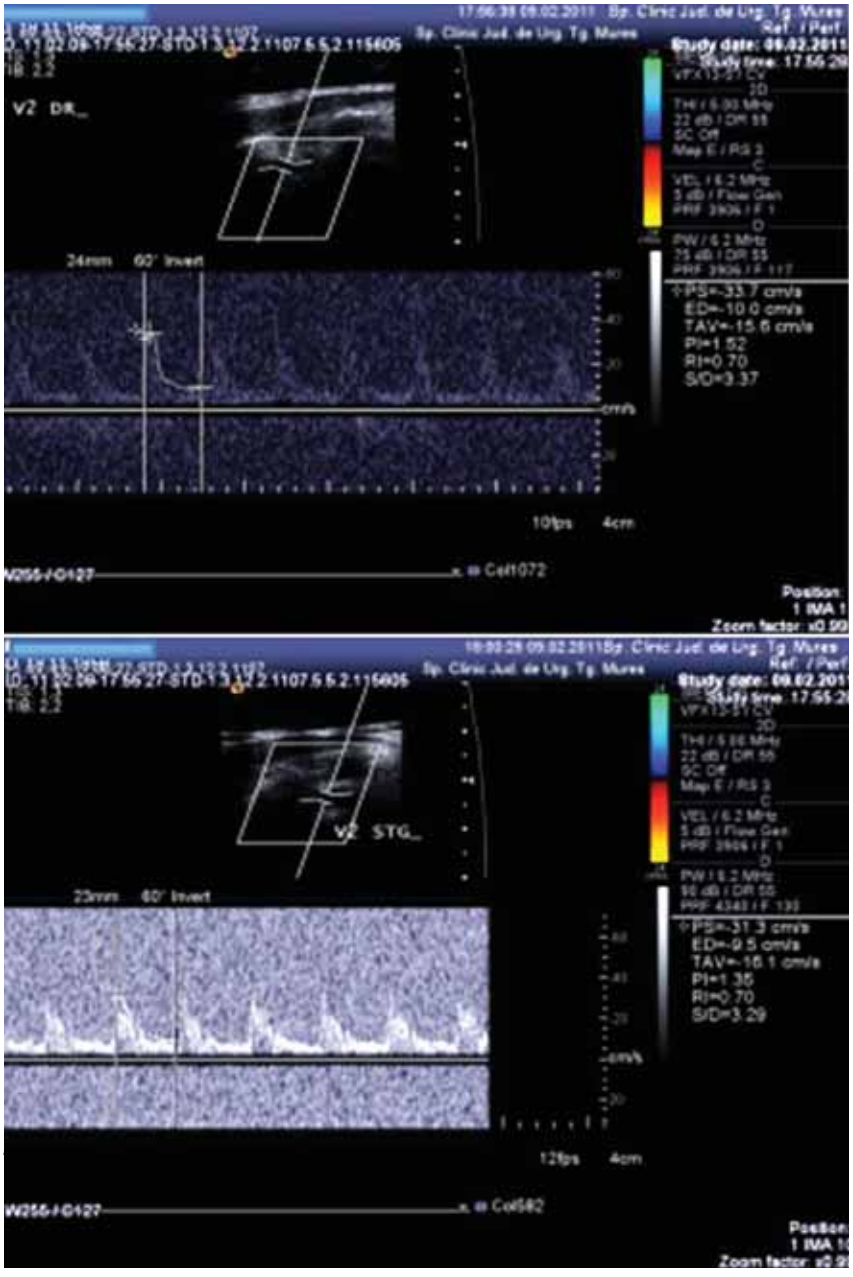

Figure 3. *Duplex ultrasonography. V2 segment of the vertebral arteries. The blood flow is present in the vertebral arteries with slightly increased pulsatility index.*

At the course of hospitalization, the patient became febrile, dyspneic, with retained tracheobronchial secretion and progressive aggravation of neurological status. He was feed by nasogastric tube and the secretion was aspirated. He received treatment with intravenous fluids, antiplatelet drug, low molecular weight anticoagulant, antipyretic, antibiotics, antihypertensive, hypolipemiant, thyroid hormone substitution. The patient died after 6 weeks from onset.

DISCUSSION

The causes of locked-in syndrome is varied: aortic dissection, vertebral artery dissection, basilar artery ectasia, stellate ganglion block, abnormal orientation of vertebral artery, tumors, encephalitis of the brainstem, traumatic lesions, central pontine myelinolysis, multiple sclerosis, air embolism, drug intoxication (2, 3, 9, 10, 11, 12). The neuroimaging has an important role in diagnosis and differential diagnosis. MRI can easily show the brainstem infarction and MRA can assess accurately both extra and intracranial vertebral arteries. A pitfall with MRA can be the over reporting of occlusion in cases of stenosis. The explanation is that time of flight MRA relies on flow related enhancement to show the arteries and a low flow can suggest an occluded artery, therefore conflicting results between ultrasound and MRA are not rare (13).

The particularity of our case is the two phase course of the diseases and the long period between the two events. Firstly the patient had become hemiparetic without speech impairment, just after 25 days he became quadriplegic and disarthric and he remained in this state for more than two weeks, when he died.

The etiological diagnosis of our case was probably basilar artery thrombosis, which has poor prognosis if early recanalization is not achieved in a specialized stroke unite (7). Although improved therapy forms, intravenous thrombolysis, intra-arterial thrombolysis, and endovascular mechanical thrombectomy, are necessary for basilar artery thrombosis (5, 6), hospitals are not equipped for these interventions. Our patient firstly was admitted in a community hospital, without a specialized stroke unite.

The prognosis for survival and recovery was found to be better in the group of patients whose syndrome was nonvascular in origin than those with a vascular etiology (3), but it has been described three cases with favorable outcome after extensive pontine infarction (8). Recognition of locked-in syndrome in these cases is important because patients are assumed to be in come, and they may be severely traumatized by inappropriate conversation around the bedside.

Intensive nursing care and early rehabilitation programs, physiotherapy, respiratory system management, effective system of communication can improve the functional recovery and reduce the mortality rate (14). Aspiration of saliva due to dysphagia and impaired cough reflex, leads to complications, including atelectasis and pneumonia, immobility predisposes to pulmonary embolus (15).

In a recent largest study which asses the quality of life in chronic locked-in syndrome, the majority of patients have reported that they have adapted to their situation and are generally happy with their lives, contrary to what outsiders may assume (16).

CONCLUSIONS

This case of bilateral pontine infarction with incomplete locked-in syndrome required special intensive care and a peculiarity was the slow deterioration of the neurological status for weeks.

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