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SHORT REPORT

Locked-in syndrome in a patient with acute obstructive hydrocephalus, caused by large unruptured aneurysm of the basilar artery (BA)

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ABSTRACT

We describe a case of acute obstructive hydrocephalus as a consequence of compression of the brainstem by a large aneurysm of the basilar artery (BA) in a 62-year-old male. After the insertion of the ventriculoperitoneal shunt (VPS), we encountered the “locked-in syndrome” clinical condition. “Locked-in syndrome” is a clinical state characterized by quadriplegia and anarthria with preserved consciousness, most commonly caused by ischemia in the ventral part of pons.

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Hydrocephalus; aneurysm; locked-in syndrome; ventriculoperitoneal shunt

Introduction

The term “locked-in syndrome” is used to describe a clinical state characterized by quadriplegia and the paresis of lower cranial nerves. In addition, a patient suffering from the “locked-in syndrome” is conscious but is unable to speak. Its first appearance in the medical literature was recorded in 1966.^{1,2}

Since 1986, this syndrome is defined as quadriplegia and anarthria with preserved consciousness. The patients suffering from the syndrome communicate via the vertical movement of their eyes and through the movement of their eyelids. The most common cause of the “locked-in syndrome” is the vascular occlusion in the basilar artery (BA) area with ischemia of the ventral part of pons.¹⁻³

Before its appearance in medical literature, the “locked-in syndrome” was described by the French novelists Alexandre Dumas and Emile Zola.

In 1845, in Chapter 58 of his novel “The Count of Monte Cristo”, Dumas described one of the characters, Monsieur Noirtier de Villefort, in the following way: after suffering a stroke, he remains paralyzed and is able to communicate with his son, his grandchild, and the servant by moving his eyelids.²

In 1868, in the novel “Therese Raquin”, Emile Zola introduced the character of Mrs. Raquin who, although completely paralyzed after an argument between her daughter-in-law and her lover, realizes the truth about the death of her son.²

The insight into the state of a patient suffering from the “locked-in syndrome” comes from Jean-Dominique Bauby, the editor of the French fashion magazine “Elle”. In 1995, at the age of 42, he suffered a stroke. Upon waking up from coma, he could only move the upper eyelid of his left eye. With the help of a speech therapist, he learned how to communicate by blinking

and with the help of his associate, Anne Consigny, he wrote the best-seller, “The Diving Bell and the Butterfly”.²

Case report

A 62-year-old patient arrived in a comatose state GCS 03 (E-1, VR-1, MR-1). His pupils were narrow and areflexic. The following is the anamnesis taken from the patient’s family: during the previous month, he has had trouble walking and has experienced bouts of vertigo. He suffers from high blood pressure that is controlled nephrologically and he regularly takes antihypertensive drugs. The week before hospitalization, the patient felt weakness, had trouble getting out of bed, was generally drowsy, and yet had no trouble communicating with people around him. On the day when he was hospitalized, he ceased communicating, had trouble breathing, and was subsequently taken to the hospital.

After a radiological exam, the brain multi-slice computed tomography (MSCT) showed the presence of the acute obstructive hydrocephalus caused by a ventral compression of the brainstem with a giant aneurysm of BA (Figure 1). An emergency drainage cerebrospinal fluid (CSF) with ventriculoperitoneal shunt (VPS) was recommended.

Following the procedure, the patient was taken off the respirator, he was able to breathe on his own, and his pupils were not dilated. Furthermore, he was able to open his eyes at will and move them vertically. He was otherwise not responsive to physical stimulation. The drainage proceeded without any problems.

The MSCT cerebral angiography showed multiple aneurysmic changes in the patient’s cerebral circulation. In addition, it also showed the correct positioning of the VP drainage (Figure 2).

The MRI of the brain showed the ventral compression of pons (Figure 1).

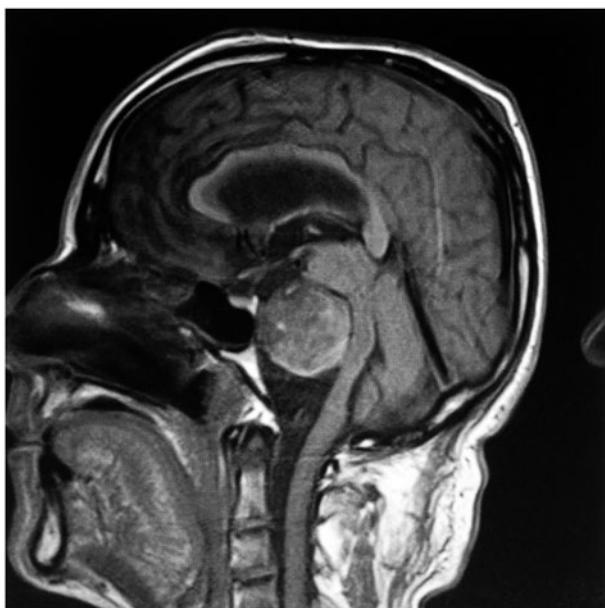


Figure 1. Sagittal T1-weighted MRI shows a giant aneurysm of the basilar artery with the compression of pons and the IV ventricle.

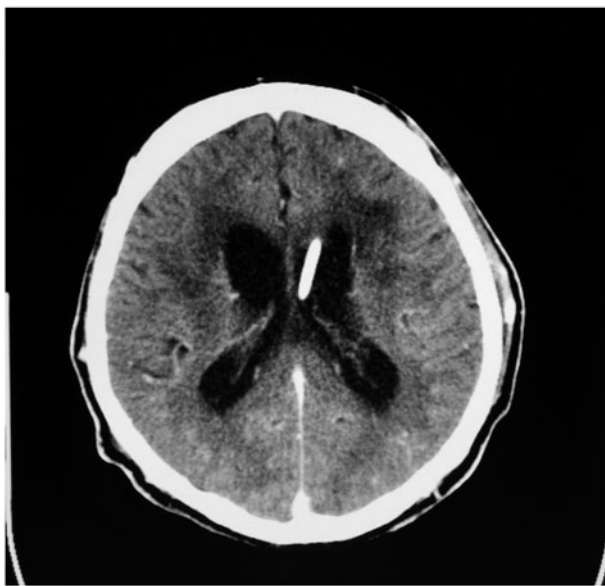


Figure 2. Postoperative MSCT shows correct position of ventricular drain.

Three weeks after the operative therapy of hydrocephalus, the patient was released and sent to the rehabilitation center with the diagnosis of the “locked-in syndrome”. During this period, he communicated with his family by blinking. His neurological condition remained without change. However, he experienced respiratory difficulties which were accompanied by high fever and the signs of respiratory infection. Four months after the diagnosis of the “locked-in syndrome”, the patient died because of respiratory complications.

Discussion

The “locked-in syndrome” was for the first time described in medical literature in 1966 as quadriplegia of a conscious patient

who is unable to speak. The patient’s only way of communicating was through the vertical movement of his eyes and eyelids. This definition was changed in 1986. Since then the “locked-in syndrome” is defined as quadriplegia and anarthria with preserved consciousness.^{1–3}

“Locked-in syndrome” is most commonly caused by ischemia in the ventral part of pons. The most common cause of ischemia of pons is the thrombosis of the BA (75–80%). Less frequent causes of ischemia are trauma, bleeding, infection, multiple sclerosis, primary, and secondary tumors of posterior cranial cavity, prolonged vertebrobasilar ischemia, central pontine myelinolysis, abscess of pons and iatrogenic cause.^{1–3}

It is important to clinically differentiate a case of the “locked-in syndrome” from coma and akinetic mutism.

Coma is a state of complete unconsciousness in which the reticular activation system in the brainstem and diencephalons is destroyed. The comatose patients are unconscious and their eyes are mostly closed.

Akinetic mutism is a clinical state in which a patient although seems awake does not respond to stimulation. The patient is not paralyzed, yet does not react to command and does not speak, and is incapable of any movement. Communication is not possible despite the fact that the patient seems awake, or mildly absent.

The “locked-in syndrome” is a state in which the reticular activation system in tegmentum of pons and mesencephalon is preserved. This is necessary for the maintenance of consciousness. The vertical movement of the eyes and the blinking of the eyelids are made possible through the preservation of the corticobulbar tract in the tegmentum of pons and the possible innervation of the oculomotor nerve nuclei (N III). The horizontal eye movement is disabled because of the destruction of the abducens nerve nuclei (N VI) and roots which pass through the ventral pons. A patient suffering from the “locked-in syndrome” is not aphatic, but is aphonic because of the paralysis of the inferior cranial nerves.^{1,2}

In addition to the importance of the clinical differentiation of these three syndromes, it is essential to distinguish them for ethical and medical-legal reasons: euthanasia, establishing of the cerebral death, medical staff’s conduct toward the patients ...

The medical prognosis for patients suffering from the “locked-in syndrome” is grim even though partial recovery is possible, but rare. The mortality rate is considerable (20–60% in the first year), and the survival depends on the quality of the rehabilitation. The mortality rate peaks in the first four months after the onset of the locked-in syndrome, and it is the highest in patients who suffer a stroke. The most common causes of death are pulmonary complications (pneumonia, atelectasis, etc.).^{2,3}

In our case, the “locked-in syndrome” was caused by the long-term compression of the ventral pons which in turn disrupted the nerve fibers and cut the pontomesencephalic connection. This was confirmed by the fact that neither the initial CT scan nor the subsequent MRI revealed any signs of the ischemic lesion in pons following the vascular occlusion.

The clinical state of our patient developed over the course of one month. Before the diagnosis of the mentioned disorders, the acute hydrocephalus had already advanced. After the surgical treatment of the acute hydrocephalus, the “locked-in syndrome” was diagnosed.

Disclosure statement

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