

LOCKED-IN SYNDROME — A REPORT OF THREE CASES

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SYNOPSIS

Three cases of 'locked-in' syndrome are reported. The patients are mute and quadriplegic with paralysis of the lower cranial nerves. They present a bilateral horizontal gaze palsy with ocular 'bobbing'. They are able to communicate by preserved eye movements and blinking. Jaw movements are also preserved. All three patients are still alive, the longest surviving being four months after onset. The main clinical features of this syndrome and their anatomical correlates are discussed.

INTRODUCTION

The term 'locked-in' syndrome was coined by Plum and Posner (1966) to describe a clinical picture in which the patient is quadriplegic and mute, yet conscious and able to communicate with others by preserved eye movements and blinking. Synonyms are 'ventral pons syndrome', 'de-afferented state' or 'cerebromedullospinal disconnection'. To our knowledge no such case has been reported locally before.

We report three cases of 'locked-in' syndrome to illustrate their clinical features and discuss their anatomical correlates.

CASE REPORTS

Case 1 (see photos in Fig. 1, 2 and 3).

B.B.I. a 56 year old Indian Muslim with established diabetes and hypertension for 3 and 4 years respectively, woke up on the 18th January 1978 with severe giddiness, headache, nausea and vomiting. The next morning, his wife found him in a state of unconsciousness with the body held straight and breathing heavily. On admission to hospital, he was comatose, responding only to pain. He was in decerebrate rigidity. The neck was supple, the pupils equal and reactive to light; there was no papilloedema and the oculo-cephalic reflex was abolished. There was spastic quadriplegia. All the deep tendon reflexes were very brisk and the plantar response was extensor bilaterally. Blood

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Fig. 1 Patient in Case 1. B.B.I., 56-year-old male

pressure was 140/80 mmHg. The pulse was regular. No carotid bruit was heard. His heart was clinically not enlarged. Breathing was ataxic and remained so for the initial two weeks during which time he remained deeply comatose, sometimes responding only to painful stimuli. The cerebrospinal fluid was normal except for a raised protein level of 110 mg/dl. The EEG revealed diffuse low amplitude theta activity. Two weeks after admission, he was noticed to open and close his eyes on command. The eyes started to move vertically but not horizontally. However, reflex horizontal ocular movement occurred on vestibular stimulation. Ocular 'bobbing' (rapid downward movement of the eyes followed by a slower return to the initial position) was subsequently observed. The corneal reflex was present. There was no facial muscle response to supra-orbital pressure. He was not able to swallow, necessitating tube feeding. The gag reflex was absent and cough reflex very poor. He was and still is, unable to communicate verbally. Conjugate lateral movements returned partially. He is now capable of masticating movements of the jaw.



Fig. 2 B.B.I. A close-up to show patient looking downwards



Fig. 3 B.B.I. Patient looking to the left

Case 2

K.K.Y. a 60 years old Chinese female was admitted on 1.5.78 to a surgical ward for suspected perforated peptic ulcer. At laparotomy, chronic duodenal ulcer and toxic ileus were found and decompression of the gastrointestinal tract was carried out. The next day, she went into sudden cardio-respiratory collapse and was resuscitated and put on a respirator. She remained deeply comatose and developed flaccid quadriplegia with bilateral facial palsy. The deep tendon reflexes were absent and plantar response extensor bi-laterally. She went into episodes of decerebrate rigidity. The CSF was normal. Four days later, she began to open her eyes and to blink. 'Ocular bobbing' was noted. The fundi were normal. Subsequently masticating movements of the jaw and partial horizontal eye movements were noted. She remained mute and quadriplegic.

Case 3

C.C.C. a 47 year old Chinese male had been suffering from episodic 'black-outs' for one month before he

was admitted to hospital on 2.4.78, when he had 3 consecutive episodes of giddiness, the last of which was associated with stiffening of the limbs, mild twitching of the arms, uprolled eyes, cyanosis and loss of consciousness. There was no previous history of hypertension or diabetes. On admission, he was drowsy but responded to commands by vertical eye movements. The oculo-cephalic reflex was absent. Both pupils were about 1 mm in size. The fundi were normal. He was in decerebrate rigidity with spastic quadriplegia. The deep tendon reflexes were brisk and the plantar responses were extensor. At the time of writing, he remained quadriplegic and mute, but able to blink his eyes and has incomplete horizontal ocular movements with occasional eye bobbing.

COMMENTS

The main features in our patients are spastic quadriplegia and lower cranial nerve paralysis. They are conscious but anarthric, communicating with the outside world by blinking and vertical eye movements. These features conform to the 'locked-in' syndrome described by Plum and Posner (1966). In contrast, classical akinetic mutism is a different entity in which the patient's higher intellectual functions and awareness are totally or severely impaired. The akinetic mute has the ability to move his extremities but usually does not do so.

The quadriplegia and lower cranial nerve palsies are explained by interruption of the corticospinal and cortico nuclear pathways either in the basis pontis or cerebral peduncles, or both in continuity.

The presence of vertical eye movements would seem to indicate that the mesencephalic tegmentum is spared as the oculomotor and trochlear nuclei are located in this region. 'Ocular bobbing' was first reported by Fisher (1959) in patients with basilar artery thrombosis or pontine haemorrhage. All three of our patients demonstrated this phenomenon. The selective loss of voluntary lateral gaze with sparing of lateral vestibular-ocular reactions (shown in Case 1) indicates a ventral pontine lesion. Hasley et al (1967) reported such a case in a conscious patient who was also mute and quadriplegic after a ventral pontine infarction secondary to basilar artery thrombosis. Their case supported the concept of a route for the corticobulbar gaze pathways that was separated ventrally from the MLF as has been shown in animals by Cohen et al (1968). With more dorsal pontine damage, the MLF is also involved so that all lateral eye movements may be abolished, as occurred in the three cases of Nelson and Johnston (1970).

The fact that our three patients are conscious would suggest that a large part of the tegmentum of the pons and midbrain is spared since it is generally accepted that preservation of this area is vital for retention of the conscious state. In the patients studied by Chase et al (1968) the largest lesion permitting full consciousness involved 25% of the tegmentum on one side. The initial unconsciousness of our three patients is probably due to oedema affecting the tegmentum.

The presence of masticating movements would imply that the trigeminal motor nuclei are spared.

Taking all the clinical findings into account, it would appear that the lesion involves the lower pons bilaterally starting from the basis pontis and spreading dorsally into parts of the tegmentum.

In most reported cases, the cause is usually a basilar artery thrombosis producing an infarction of the ventral rostral pons extending dorsally in various degrees into the paramedian tegmentum and sometimes rostrally into the cerebral peduncles. Kamp and Hurtig in 1974 reported a case with bilateral infarction of the lateral two-thirds of the cerebral peduncles with only scattered microinfarcts dorsal to the basis pontis, but in this case, there was no lateral gaze palsy as expected. On this evidence, they rightly emphasised that the term 'ventral pons syndrome' previously used synonymously with the 'locked-in' syndrome should give way to a broader term like 'ventral brain stem syndrome'. Another case reported by Kumar et al, (1978) was attributed to trauma as the patient developed the syndrome following a road traffic accident although no pathological evidence was presented. Ocular movement in all directions was preserved.

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