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God locked you in the room, but left a window open: A case report of spinal cord stimulation in locked-in syndrome

Dear Editor:

Locked-in syndrome (LIS), caused by severe damage to the pons, is a serious neurological condition of movement deficiency, characterized by quadriplegia and aphonia. Spinal cord stimulation (SCS), the most common neuromodulation therapy, has recently been shown to restore walking in patients with spinal cord injury [1]. The possibility of SCS treatment in LIS is still unclear. We reported the first application of cervical SCS in a classic LIS patient with encouraging outcomes.

Case presentation

MAZ, a 37-year-old, right-handed Asian man, suffered from classical LIS for 6 months. The LIS was caused by occlusion of the basilar artery. He had a prosthetic valve implanted for rheumatic heart disease 10 years prior, and had not taken anticoagulant drugs regularly in recent years. He was sent to a local hospital for intensive treatment, but the patient still exhibited complete quadriplegia and aphonia. Thus, he was transferred to our hospital for further treatment.

Upon admission, in addition to quadriplegia and inability to speak, the limbs muscular tension was severely elevated, accompanied by involuntary spasm. He also showed hyperhidrosis on the torso and head. He was able to communicate with others by blinking, indicated a classic LIS. His long-term medication consisted of dabigatran etexilate 110 mg bid and eperisone 50 mg tid. Computed tomography (CT) and angiography (CTA) before surgery were shown in Fig. 1 A and C.

The patient agreed and his parents gave written informed consent to confirm all the procedures for this study, which had prior ethics approval from the research ethics committee of the Third Affiliated Hospital of Sun Yat-sen University. Under general anaesthesia, a 16-contact electrode array (Lead Set 39286; Medtronic, USA) was inserted into the dorsal epidural surface at the mid-line of the C2–4 level (Fig. 1 B). The spinal electrode was connected to the permanent stimulator implanted subcutaneously below the right clavicle. A frequencies of 40Hz, an amplitude of 1.0V and a pulse duration of 180 μs were chosen as the main stimulation parameters, with subtle adjustments based on the changes of symptoms.

The patient was followed-up by our team and his relatives, and any changes in his clinical condition was recorded. The hyperhidrosis significantly improved 3 days after the SCS implantation. The patient showed continuous improvement of muscular tension and spasm. After approximately 6 months of electrical stimulation and rehabilitation training, he was able to pronounce vaguely and the limbs muscle strength recovered to 2–3/5. In the recent follow-up (26 months after surgery), his right upper limb muscle strength was approximately 4/5 (Supplementary video S1). Along with the improvement of clinical symptoms, CTA also showed partial recanalization of the basilar artery (Fig. 1 D).

Supplementary video related to this article can be found at https://doi.org/10.1016/j.brs.2019.08.006.

Discussion

To our knowledge, this is the first case report of the application of SCS in LIS. Although SCS has been used successfully to improve motor function after spinal cord injury [1–3], it remains unclear whether SCS has beneficial effects in central nervous system injury at higher levels, such as in the brainstem. Our case exhibited severe motor dysfunction, and his baseline motor function did not

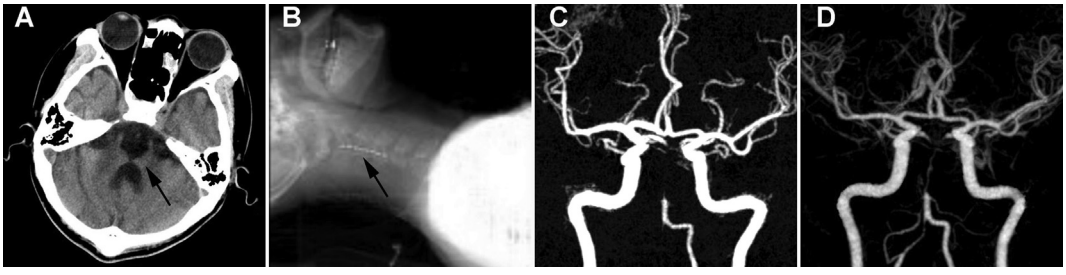

Fig. 1. (A) Computed tomography (CT) showed liquefaction necrosis in the ventral pons (arrow). (B) X-ray showing the implantation location of the SCS electrode (arrow). (C) CT angiography (CTA) before SCS implantation. (D) CTA after 6 months of SCS implantation.

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improve over 3 months prior to SCS implantation. By contrast, after SCS implantation and treatment for 2 years, the patient showed improved hyperhidrosis, decreased muscle tone, partial recovery of locomotor function. By imaging, we also found evidence of partial recanalization of the basilar artery. These results were remarkable based on previous findings of high mortality and limited recovery in LIS [4].

Previous animal studies reported that SCS can induce a robust and consistent rhythmic stepping-like activity and partially restore locomotion in complete spinal cord injury rats [5–7]. In a subsequent study of patients with severe spinal cord injury, SCS in lumbosacral segments produced step-like movements and restored independent standing after complete paraplegia [3,8]. Further, implantation of SCS at the injury level in patients with cervical spinal cord injury improved volitional control of the hands [2]. The improvement in motor function was mostly stimulus-dependent in the above human studies, which is hypothetically mediated via activation of interneuronal networks in the spinal cord. It is worth noting that our findings differed from previous studies. The improvement of motor function in present patient was gradual and not stimulus-dependent. In addition, the SCS system was implanted at the C2-4 level below the injury (brain stem), instead of in the cervical or lumbosacral enlargement or just near the injury site.

In present case, the basilar artery is completely occluded for 6 months. We performed dynamic imaging tracing of the occluded basilar artery. The basilar artery and its branches were partly shown on CTA after 6 months of SCS treatment. Though cervical SCS has been shown to improve cerebral blood flow both in animals and humans (review [9]), this is the first clinical clue that cervical SCS may has positive impact on recanalization of large artery.

Four possible mechanisms might explain how cervical SCS improved LIS syndrome in present patient: (1) the physiological state of the interneuronal networks in the brainstem can be modulated by SCS, though implanted below the injury level; (2) there may be a numerous anatomically-connected, but non-functional, conduction pathways crossing the lesion that can be activated by SCS; (3) SCS may enhance the function of residual pathways, and/or promote regeneration of axonal pathways, as reported in animals [6]; (4) artery recanalization and improvement of cerebral blood flow may be helpful to restore locomotor function.

Our present clinical observations have cautiously suggested that cervical SCS may be a safe and effective treatment option for LIS. More cases or a multicenter controlled study are required to confirm the efficacy of cervical SCS in LIS treatment in the future.

We thank the patient for taking part in this study and agreeing to share his treatment.

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Declarations of interest

None.

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Zhenchao Huang[1]
Department of Neurosurgery, The Third Affiliated Hospital of Sun Yat-sen University-Lingnan Hospital, Guangzhou, 510530, PR China
E-mail address: raymond25x@aliyun.com.

Xiaofeng Xu[1]
Department of Neurology, The Third Affiliated Hospital of Sun Yat-Sen University, Guangzhou, 510630, PR China
E-mail address: xuxf5@mail.sysu.edu.cn.

Qing Dong
Department of Neurology, The Third Affiliated Hospital of Sun Yat-Sen University, Guangzhou, 510630, PR China
E-mail address: dongqing2009tracy@163.com.

Lei Wei
Department of Neurology, The Third Affiliated Hospital of Sun Yat-Sen University, Guangzhou, 510630, PR China
E-mail address: weilei068@163.com.

Yanjun Lin
Department of Neurology, The Seventh Affiliated Hospital of Sun Yat-Sen University, Shenzhen, 518107, PR China
E-mail address: linyanjun00@163.com.

Ju Jiao
Department of Nuclear Medicine, The Third Affiliated Hospital of Sun Yat-Sen University, Guangzhou, 510630, PR China
E-mail address: jiaoju2@mail.sysu.edu.cn.

Zhengqi Lu
Department of Neurology, The Third Affiliated Hospital of Sun Yat-Sen University, Guangzhou, 510630, PR China
E-mail address: lzq1828@aliyun.com.

Feng Qin*
Department of Neurosurgery, The Third Affiliated Hospital of Sun Yat-sen University-Lingnan Hospital, Guangzhou, 510530, PR China

* Corresponding author.
E-mail address: qinfeng2@mail.sysu.edu.cn (F. Qin).

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[1] These authors contributed equally to this work.