

Case Report

Preserved motor-evoked potentials but without good motor recovery in a patient with decerebrate rigidity[☆]

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Abstract

The corticospinal tract is not incriminated in decerebrate rigidity (DR). However, this has not yet been proven in humans. We applied transcranial magnetic stimulation (TMS) in a decerebrate patient to support the hypothesis. A patient suffering from pontine hemorrhage with the fourth ventricular extension was admitted unconscious and in a decerebrate posture. Five days later, she regained consciousness but remained in a decerebrate posture. Motor-evoked potentials (MEPs) to TMS were measured 1 week after she had regained consciousness, and this provoked muscle responses in her hands and feet bilaterally. During the follow-up, the patient's muscle tone became persistently flaccid, although her strength increased to varying degrees in different body and limb muscles. She remained bedridden for 3 years after the stroke and could neither turn on the bed by herself nor perform skilled movements using her hands. The findings of TMS confirmed the animal studies in that the mechanism of decerebrate rigidity did not come through a damage of the corticospinal pathway. This also implies that a preserved corticospinal tract function cannot guarantee a good motor recovery in a stroke patient.

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1. Introduction

The technique of transcranial magnetic stimulation (TMS) has been widely applied in the clinical investigation,¹ especially in patients with motor disabilities as a result of cerebrovascular disorders. TMS is also of prognostic utility, and a preserved TMS response usually indicates a better outcome.²

The term “decerebrate rigidity” (DR) was first used in 1898 by Sherrington³ in describing posture after a pre-pontine transection. He also concluded that the corticospinal tract could not be incriminated in either causing or reversing decerebrate posture after a series of animal studies. To our knowledge, TMS has not been applied in the study of corticospinal tracts in DR patients. Here, we report a patient with DR because of a brainstem hemorrhage. She failed to execute voluntary movement of her limbs, although her consciousness was clear and the function of the corticospinal tract was preserved after the acute stage of DR had passed. We reviewed the mechanism of DR and the phenomenon of poor motor control even with preserved function of the corticospinal tract.

[☆] The authors declare no conflicts of interest.

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2. Case report

A 48-year-old woman suffering from pontine hemorrhage with the fourth ventricular extension was admitted because of a sudden change of consciousness. The Glasgow Coma Scale on admission was 4, the best motor response was 2 (decerebrate posture), the best verbal response was 1 (no response), and the eye-opening response was 1 (no response). Neurological examination revealed a normal light reflex, an absence of right-eye corneal reflex, and oculocephalic reflex. A mechanical ventilator was used to support her respiration. DR, which developed extension of her four limbs with internal rotation of bilateral arms and plantar flexion, clenching of jaws and opisthotonos, had been noted since admission, and these findings were enhanced by pain or pressure stimulations on her limbs. She regained consciousness 5 days after the stroke onset and was able to obey simple orders (such as opening and closing her eyes). Limb movement was limited, and she could only move the right upper limb slightly. Motor-evoked potentials (MEPs) to TMS 1 week after she had regained consciousness showed preserved corticospinal tract responses, although the onset latencies were delayed and the peak amplitudes were decreased (Fig. 1).

Three months later, there were residual neurological deficits, with right abducens palsy, right facial and crossed left hemibody sensory impairment, poor hand dexterities, and truncal unsteadiness. The patient was able to follow commands to execute simple tasks, such as moving her shoulders and flexing and extending her wrists or elbows. She could not perform some tasks, such as using her hands to write or to eat with chopsticks, a fork, or a spoon. She could not maintain a steady sitting position without support, not to mention an upright posture with support. The muscle response to TMS became larger along with her muscle strength improvement, although her muscle tone decreased as hypotonia. She remained in flaccid weakness and did not have

a significant improvement in motor function after 3 years of follow-up.

3. Discussion

Series of animal studies have revealed that the integrity of reticular, cerebellar, vestibular, and cortical descending neural systems is responsible for the development of DR.⁴ Our TMS findings confirmed these animal studies in that the DR mechanism did not come through a damage of the corticospinal pathway. The DR mechanism of our patient was mainly because of a lesion involving the rubrospinal pathway and an indirect disinhibition of vestibulospinal and reticulospinal effects.⁵

Preserved MEPs to TMS usually indicate a good motor recovery of stroke patients. However, our patient has remained bedridden for 3 years after the stroke. In a general sense, the control of posture and locomotion is based on a spinal central pattern generator influenced by supraspinal structures and peripheral afferents. From the original concepts of Kuypers,⁶ the supraspinal control is divided between two systems: a fine control of locomotion (the lateral system, including the corticospinal and rubrospinal tracts) and a provision of postural support for the fine control (the medial system, including the reticulospinal and vestibulospinal tracts). During the control of goal-directed locomotion, selection of a motor program is performed in the basal ganglia and then reaches the command centers in the diencephalic locomotion region and the mesencephalic locomotion region, which can modulate the spinal central pattern generator for locomotion via the reticulospinal neurons.⁷ In our patient, preserved MEP might represent certain integrity of the corticospinal tracts. The poor control of posture and fine voluntary movement might have come through a locomotion dysfunction by disruption of the rubrospinal or in part reticulospinal tracts involving the locomotion command center at the brainstem level. However, locomotion failure has been used to describe gait disorders in humans, and it may not account for the loss of hand dexterity in our patient.

The concept of incomplete lesion was first proposed by Dimitrijevic et al,⁸ describing a preserved electrophysiological function of the corticospinal tract but clinically exhibiting complete paralysis in spinal man. He supposed that a substantial number of nerve fibers can survive the trauma but insufficient to elicit effective contraction of muscles. A similar concept can be applied to our case, where the lesion involved the brainstem instead of the spinal cord. The TMS study of our patient showed residual connectivity of the corticospinal tract, but she did not have any voluntary movement of the hands and had poor motor recovery later. This may in part share a similar “incomplete” mechanism of spinal man.

Loss of dexterity and prolonged flaccid weakness were noted in our patient. Brainstem lesions may produce cerebellar diaschisis,^{9,10} which may last 20 years after stroke.¹¹ Although single photon emission computed tomography or positron emission tomography was not done in our patient, we infer that she had cerebellar dysfunction because of diaschisis. Cerebellar dysfunction may decrease human muscle tone and

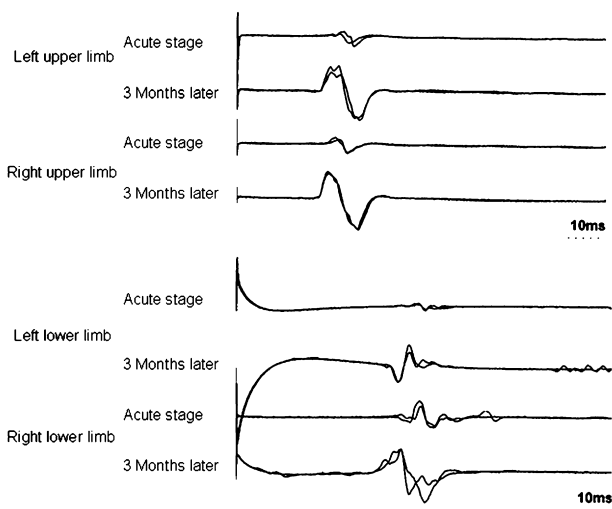


Fig. 1. The motor-evoked potentials of a 48-year-old patient, who presented with decerebrate rigidity because of a pontine hemorrhage during the acute stage and 3 months after stroke onset. Recordings: abductor pollicis brevis and tibialis anterior.

disturb human motor control. Hence, persistent cerebellar dysfunction because of diaschisis might in part account for her hypotonia and clumsiness.

In conclusion, DR may occur following a massive, bilateral cerebral trauma, anoxic damage, or midbrain destructive lesion, such as the hemorrhagic stroke in our case. Although it is rare, preserved MEP can occur in humans with DR. Our case also emphasizes that preserved MEP does not always indicate a good voluntary movement and motor recovery.

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