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Radiofrequency Ventro-oral Thalamotomy for Post-stroke Focal Dystonia in a Pediatric Patient

Noriko HIRAO,[1] Takashi MORISHITA,[1] Kazuya SAITA,[1] Tomohiro TAKAGI,[1] Shinsuke FUJIOKA,[2] and Tooru INOUE[1]

[1]*Department of Neurosurgery, Fukuoka University, Fukuoka, Fukuoka, Japan*
[2]*Department of Neurology, Fukuoka University, Fukuoka, Fukuoka, Japan*

Abstract

Dystonia is a movement disorder that has various treatment options. For primary dystonia, stereotactic procedures such as deep brain stimulation (DBS) have demonstrated favorable outcomes. For secondary dystonia, however, the treatment outcomes remain inconclusive, and the heterogeneous etiological background is considered to contribute to the poor outcomes of the disease. Here, we report a rare pediatric case of post-stroke focal dystonia treated with conventional radiofrequency ventro-oral (Vo) thalamotomy. The patient was an 11-year-old girl with secondary focal dystonia in her right hand. The dystonia was considered to result from a stroke lesion in the putamen due to vasculitis following varicella-zoster virus infection. We hypothesized that the infarction of the putamen resulted in hyperactivity in the thalamus, and, thus, performed a radiofrequency Vo thalamotomy. Markedly decreased muscle tone in her right hand was noted immediately after surgery. However, the improvement was temporary, as her symptoms returned to baseline level by the 6-month follow-up. Although the observed improvement was temporary in this case, our findings may elucidate the possible mechanisms of secondary focal dystonia. Further studies are needed to establish an effective surgical treatment for secondary focal dystonia.

Keywords: focal dystonia, secondary dystonia, stroke, thalamotomy, ventralis oralis nucleus

Introduction

Dystonia is a movement disorder characterized by abnormal muscle tone resulting in muscle spasm and abnormal posture with treatment options that include medical therapies and surgical procedures, such as intrathecal baclofen therapy and stereotactic surgeries. Favorable outcomes of stereotactic procedures, such as deep brain stimulation (DBS) have been reported for primary dystonia; however, the outcomes of such procedures for secondary dystonia have demonstrated mixed results.[1)] The heterogeneous etiological background of secondary dystonia is considered to contribute to the poor outcomes. Here, we report a pediatric case of post-stroke focal dystonia due to vasculitis, which was treated with conventional radiofrequency ventro-oral (Vo) thalamotomy and discuss the possible etiology based on our findings.

Case Report

The patient was an 11-year-old girl with secondary focal dystonia in her right hand. When she was 1-year old, she was infected with the varicella-zoster virus and subsequently had cerebral vasculitis, which resulted in the occlusion of the middle cerebral artery (Fig. 1). She had a cerebral infarction in the left basal ganglia region, following which she developed motor impairment in the right upper extremity (UE). She was then diagnosed with right upper limb dystonia due to the left basal ganglia infarction at 8 years of age. This symptom affected her daily activities and was refractory to medical therapies and rehabilitation.

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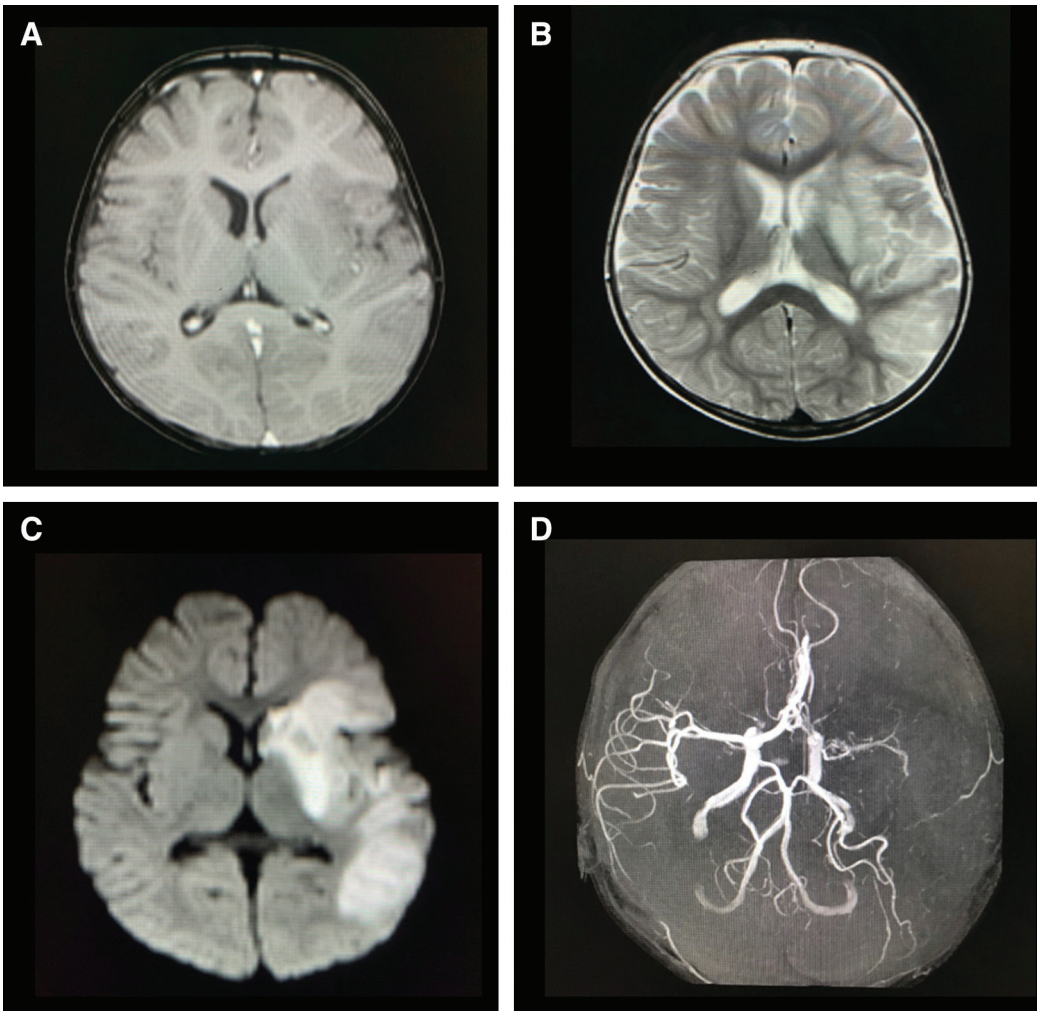

Fig. 1 Magnetic resonance images at stroke onset. (A) T1-weighted image with contrast. (B) T2-weighted image. (C) Diffusion-weighted image. (D) Magnetic resonance angiography.

Increased forearm muscle tone made voluntary hand opening difficult; therefore, once she held an object with her right hand, she could not release it. Since the symptoms were localized in the right hand, we hypothesized that a stereotactic surgical procedure might alleviate her focal dystonia similar to other types of focal dystonia, such as writer's cramp and musician's cramp.[2–4] Following discussion with the patient and her family concerning the risks and benefits of lesion therapy and DBS, the patient and her family decided on her undergoing radiofrequency Vo thalamotomy. We considered that the patient was unable to tolerate the awake procedure, so we proposed to treat with minimum lesion for safety under general anesthesia.

During preoperative stereotactic planning, the dentatorubrothalamic tract was determined to identify the ventrolateral nucleus, as described in our previous report,[5] and the trajectory was planned so that the radiofrequency electrode would pass the Vo anterior/posterior border. The preoperative stereotactic targeting (tip of the electrode) coordinates relative to the mid-commissural point and trajectory angles were as follows: 13.5 mm to the left, 0.5 posterior, 2.5 superior, anterior commissure-posterior commissure angle = 69.0, and centerline angle = 28.5 to the left. The target coordinate may have been unusual compared to conventional target due to ex-vacuo change.

On the day of surgery, a Leksell G frame (Elekta, Stockholm, Sweden) was attached under general anesthesia, and a computed tomography scan was performed to translate the stereotactic planning based on the Cartesian coordinate system into the coordinate system of the Leksell G frame. The minimum dose of muscle relaxant was administered for

intubation but the drug was reversed so that the muscle response to the stimulation could be observed. Following a 3-cm straight skin incision, a burr hole was made along the trajectory. A single-pass microelectrode recording (MER) was performed to confirm the preoperative plan. Background activities were relatively quiet due to general anesthesia; however, kinesthetic responses to passive movements of lower and UEs and jaw were observed from lateral to medial along the MER pass but no sensory responses were obtained (Supplementary Figure 1; Supplementary material is available online). We, therefore, considered that the electrode was positioned in the ventrolateral nucleus but not passed sensory thalamic nucleus as expected. Then, a radiofrequency electrode (2 mm in diameter, 4 mm length) was inserted so that the tip of the electrode was positioned 2 mm above the planned target along the trajectory to avoid lesioning the subthalamic area and to prevent complications such as choreo-ballistic movements.[6] Macrostimulation was performed at 100 microseconds and 133 Hz, and the stimulation intensity was gradually increased to the threshold level of the motor response (muscle twitch) in her right hand, which was observed at 7.5 mA. Radiofrequency coagulation was performed for 60 seconds with a temperature of 70°C. The lesion shown on the postoperative MRI images was considered to be located along the preoperative planning as expected, and the perilesional edema had been subsided by the 3-month follow-up (Fig. 2).

Functional improvement was observed in the voluntary opening and closing movements of the right hand, and muscle tone and grip strength decreased. Upper limb motor function was evaluated using the Fugl-Meyer assessment-UE (FMA-UE; range 0–66) and the action research arm test (ARAT; range 0–57). The FMA and ARAT scores at baseline were 36 and 21, respectively. A maximal improvement was observed at the 1-month follow-up, with FMA and ARAT scores of 43 and 24, respectively; however, the scores returned to the baseline level (FMA-UE score 37; ARAT score 23) by the 6-month follow-up. We discussed further treatment options including additional lesioning and DBS with the patient and her family, but they were not willing for her to undergo additional surgery.

Discussion

In this case, we performed a radiofrequency lesioning under general anesthesia; however, there may be an argument over our procedure and the treatment modalities. To maximize the safety, all of the possible electrophysiological data were gathered as reported by the electrophysiological studies under general anesthesia,[7,8] and the lesion size was minimized in our case. Even though no adverse events were observed in the present case, some clinicians may advocate that DBS may be a better option for a pediatric patient who require general anesthesia. Clinicians should be cautious in procedure section and explain about the potential risks and benefits of each procedure.

Hyperactivity of the thalamus may have contributed to the pathophysiology of focal hand dystonia in the present case. In the cortico-striato-thalamo-cortical loop, the thalamus receives inhibitory signals from the striatum.[9] For this case report, we hypothesize that an infarction of the left putamen resulted in hyperactivity of the motor area in the thalamus (Fig. 3). The observed improvement following Vo thalamotomy, although temporary, supports our hypothesis.

For this case report, other potential stereotactic targets for radiofrequency lesioning could have been considered, such as the globus pallidus interna and other thalamic nuclei. However, based on the phenomenology of focal hand dystonia, we selected the thalamic Vo nucleus as a target on the assumption that a similar outcome as with primary focal dystonia could be expected.[2–4]

The effectiveness of stereotactic neurosurgery for primary dystonia has been well established. In contrast, the clinical outcomes of stereotactic surgery, especially radiofrequency thalamotomy, have been reported with mixed results in cases of secondary dystonia (Table 1).[10–14] In this case report, our patient underwent temporary improvements, and this result was consistent with previous reports.[10–14] As seen in the magnetic resonance images, the peri-lesional edema subsided by the 3-month follow-up (Fig. 2). Postoperative imaging showed the optimal lesion location along the preoperative stereotactic planning (Fig. 2), the degree of improvement seemed to be associated with the degree of the edema. In this context, wider area in the thalamus may have been required to maintain the therapeutic effect similar to Holmes tremor,[15] or DBS may have been a better treatment option to obtain a sustainable effect as DBS therapy is adjustable; however, its results remain unconvincing to date.

Conclusion

We report a rare pediatric case of stroke-induced secondary focal dystonia in which Vo thalamotomy resulted in a temporary improvement. Even though we treated the patient with a conventional method,

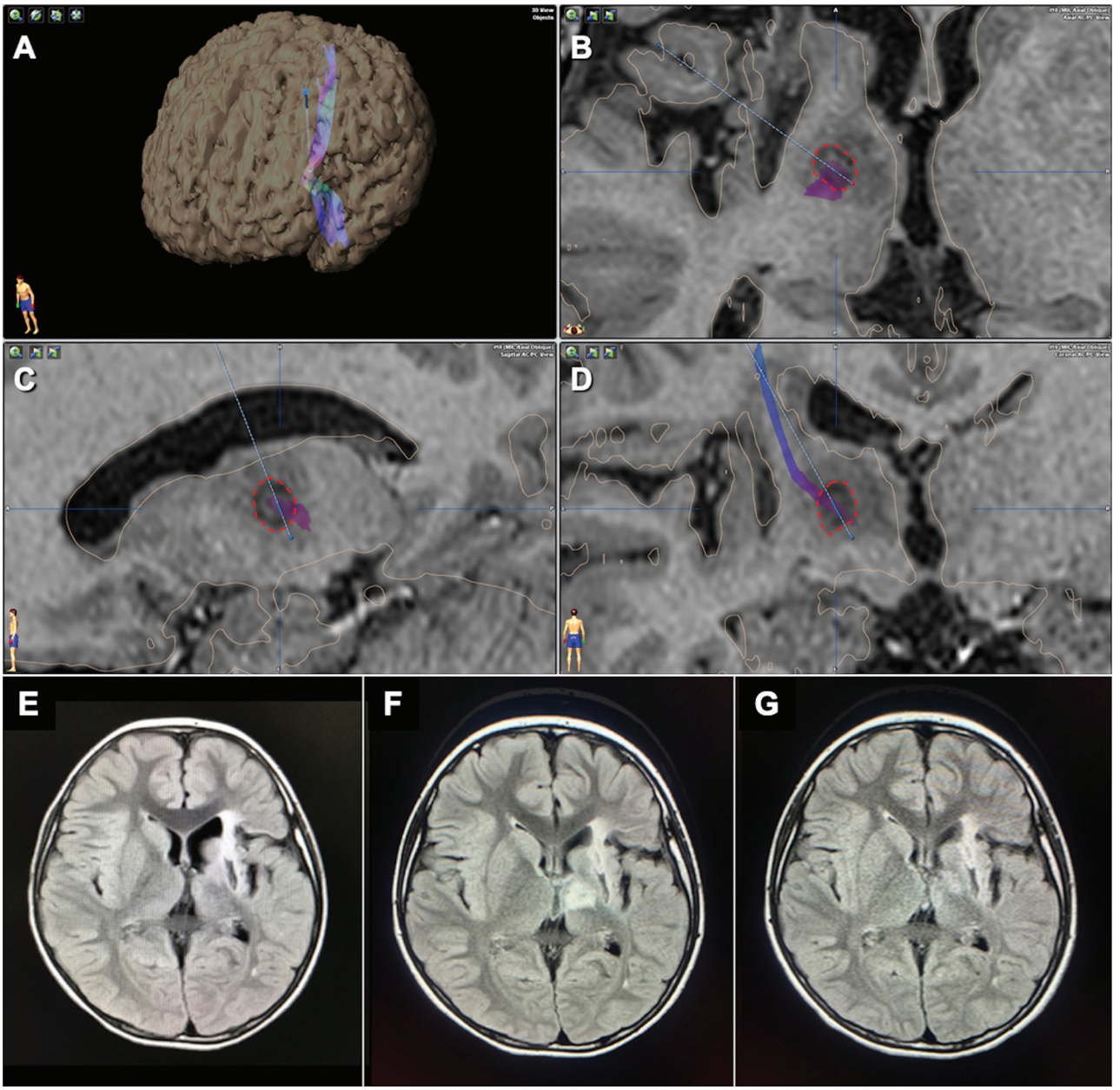

Fig. 2 Postoperative volumetric T1-weighted images with dentatorubrothalamic tract on stereotactic planning software (A–D) and preoperative and postoperative magnetic resonance images (E–G). (A) Three-dimensional brain image. (B) Axial image. (C) Sagittal image. (D) Coronal Image. Red dotted lines: demarcation of the radiofrequency lesion. Blue lines: preoperatively planned trajectory. A preoperative fluid-attenuated inversion recovery image (E) showing an old infarction lesion in the left putamen. A postoperative fluid-attenuated inversion recovery image performed 1 week (F) and 3 months (G) after surgery. The perilesional edema subsided by the 3-month follow-up.

our findings and strategy based on the basal ganglia circuit theory may elucidate the mechanisms of secondary dystonia. Even though our case did not experience any adverse events, treatment modality should be cautiously selected concerning the risks and benefits. Further studies with increased number of patients and standardized evaluations are needed to establish an effective surgical treatment for secondary focal dystonia.

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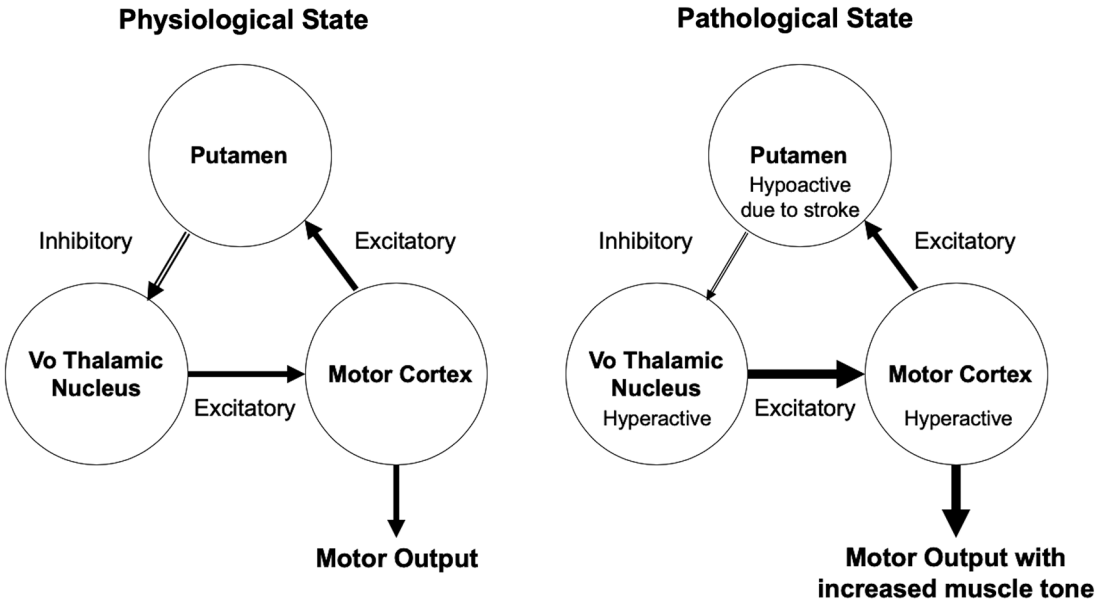

Fig. 3 Cortico-striato-thalamo-cortical loops explaining the pathological state in the present case. The stroke lesion in the putamen impaired the inhibitory signal, which resulted in a hyperactive state of the thalamus. This resulted in a hyperactive state of the motor cortex and subsequent abnormal motor output with increased muscle tone. Vo: ventro-oral.

Table 1 Reported clinical outcomes of thalamotomy alone for secondary dystonia study

	Etiology	Target	Follow-up	Outcome
Krauss and Jankovic, 1997	Stroke due to gun shot	VL	5 years	Mild improvement
Cordoso et al., 1995	Stroke (6 cases) Perinatal injury (2 cases) Prolonged seizures (1 case) Trauma (1 case)	Ventro-oral	41 months (mean)	5/10 cases achieved greater than moderate improvements
Loher and Krauss, 2009	Head trauma	1st surgery: Zi and medial pulvinar 2nd surgery: Vop	14 years	Torticollis was improved. Tremor and hemidystonia improved to a lesser degree
Yen et al., 2012*	Venous angioma	Vim	72 months	Symptom-free
Alvarez et al., 2014	Post-thalamic stroke	Vim	1 year	Improvement

*Gamma knife was applied. Vim: ventral intermediate, VL: ventrolateral, Vop: ventro-oral posterior, Zi: zona incerta.

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Conflicts of Interest Disclosure

The authors declare no conflict of interest related to this work. All authors who are members of the Japanese Neurosurgical Society (JNS) have registered online Self-reported COI Disclosure Statement Forms through the website for JNS members.

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Corresponding author: Takashi Morishita, MD, PhD
Department of Neurosurgery, Fukuoka University Faculty of Medicine, 7-45-1 Nanakuma, Jonan-ku, Fukuoka, Fukuoka 814-0180, Japan.
e-mail: tmorishita@fukuoka-u.ac.jp