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CASE REPORT

Deep Brain Stimulation of the Globus Pallidus in a Microcephaly Patient

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Pallidal stimulation for medically intractable dystonia is known to be effective. However, the optimal target of globus pallidus interna (GPi), so called sweet spot is not clear when the anatomy of deep brain structures are not in ordinary fashion. A 29-year-old woman who presented with severe general dystonia was admitted for deep brain stimulation in our center. The patient was diagnosed with microcephaly at birth. Preoperative magnetic resonance image (MRI) revealed that her GPi was located 17mm lateral from the midplane far medial than the classical coordinate, which is usually 21mm lateral from the midplane. Directional electrodes (St. Jude, Little Canada, MN, USA) were implanted to avoid possible side effects from a narrow effective volume. After optimizing the stimulation direction and amplitudes, her BFMDRS movement and disability scores decreased from 30 to 9 and from 11 to 1, respectively 6 months after surgery. Presentation with a distorted or undeveloped brain, in such case are in need of direct targeting and directional lead stimulation can help increase target precision.

KEY WORDS: Deep brain stimulation · Dystonia · Microcephaly.

INTRODUCTION

Medical treatment is the first-line treatment modality for dystonia followed by botulinum toxin injection.[9)10)] Deep brain stimulation (DBS) for cervical dystonia was first introduced by Mundiner in 1977,[11)] and DBS for GPi was approved by the United States Food and Drug Administration (FDA) in 2003. Since then, DBS has been a convincing treatment modality for medically intractable dystonia.[11)] However, the anatomical location of the target, so called sweet spot, is not clear especially when the patient's deep brain structures are not in ordinary fashion. In such case, direct visualized targeting with magnetic resonance imaging (MRI) and directional stimulation electrodes can be used for precise stimulation. Herein, we present a rare case of a distorted or undeveloped brain due to microcephaly in a patient with a severe form of general dystonia, who was then admitted for DBS in our center.

CASE REPORT

A 29-year-old woman who presented with a severe form of general dystonia was admitted for DBS in our center after a neurologist conducted a series of evaluations. The patient was diagnosed with microcephaly at birth, and she was also born preterm needing incubation care for an extended period of time. Then, she presented with a mild intellectual disability early in her youth. Her physical appearance was atypical. That is, she had a short stature (136cm) and a head circumference of 48.3cm, corresponding to an adult head circumference of 3 percentile.[3)] She complained of torticollis in 15 degrees and anterocollis in 60 degrees as well as palpable right sternocleidomastoid muscle contraction. After the patient was diagnosed with cervical dystonia, she received multiple cycles of botulinum injection and medication adjustment for 1 year. However, the symptoms eventually aggravated, and the patient was referred to our neurosurgical department for consideration of DBS. The severity of dystonia was evaluated before and 6 months after surgery with stimulation using the movement and disability subscores of the Burke-Fahn-Marsden dystonia rating scale (BFMDRS). Written informed consent was obtained from the patient, and she underwent bilateral DBS in the GPi. The patient was under monitored anesthesia care for microelectrode recording, and the x, y, and z coordinates for the bilateral GPi were set based on 3T MR images obtained prior to surgery. Her GPi was located 17mm lateral from the midplane, 2mm anterior to the midcommisur-

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ral point, and 3mm below the intercommissural line, which was far medial than the classical coordinate (usually located 21mm lateral from the midplane, 2mm anterior to the midcommisurral point, and 5mm below the intercommissural line).[2)] Neurophysiological target verification was conducted during the procedure via simultaneous microelectrode recording and was later on reflected to the lead stimulation settings. Directional leads (St. Jude, Little Canada, MN, USA) were implanted bilaterally to avoid possible side effects and better stimulation effect for the patient with microcephaly who may have narrow effective volume for stimulation. The markers that indicate the directions of the directional lead was set toward the anterior direction to allow stimulation, considering the direction of the implanted leads (Fig. 1). Subsequently, an internal pulse generator (Infinity, Little Canada, MN, USA) capable of bilateral stimulation was inserted in the left subclavicular pocket and was connected to the DBS leads via subcutaneous tunneling. Postoperative computed tomography (CT) and preoperative MRI were merged to confirm the target region (Fig. 2). The device was turned on after 30 post operative days. With the optimization of stimulating direction and amplitudes, her BFM-DRS movement and disability scores decreased from 30 to 9 and from 11 to 1, respectively, during follow-up 6 months after surgery. On the right side, lead 3A (anterior lead of the proximal third lead) was used as the cathode, and the electrical setting included an amplitude of 1.50mA, a frequency of 160Hz, and a pulse width of 140μs. On the left side, lead 2A (anterior lead of the second proximal lead) was used as the cathode, and the electrical setting included an amplitude of 1.40mA, a frequency of 160Hz, and a pulse width of 140μs. During optimal setting, the patient complained of aphasia, lip smacking, and facial twitching. As we gradually decreased the amplitude and altered the setting from a bipolar setting to monopolar set-

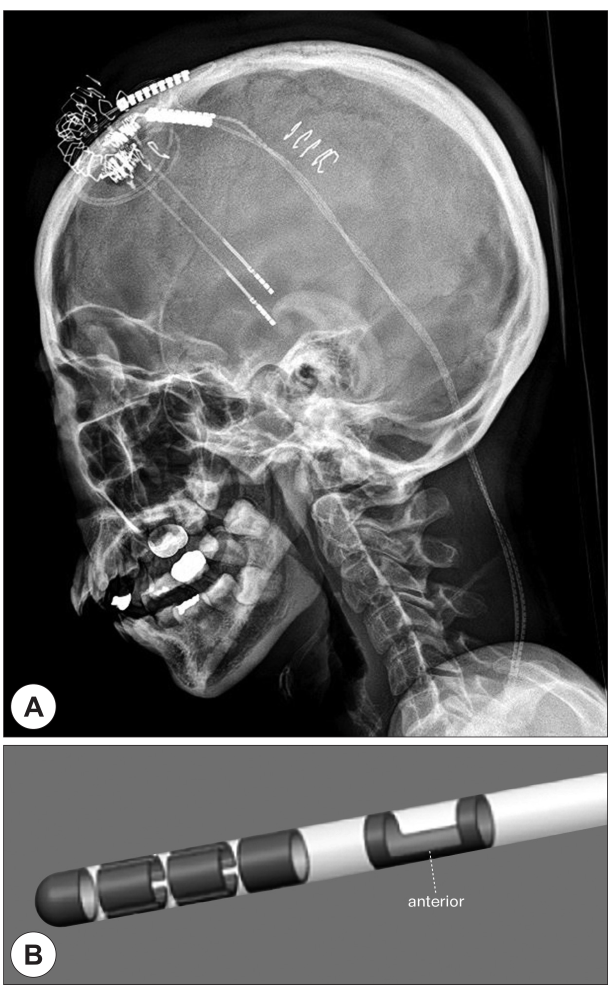

Fig. 1. Shape of directional deep brain stimulating electrode. A : Postoperative skull X-ray lateral view image of a patient with microcephaly showing directional indicators toward the anterior. B : Schematic visualization of the directional lead showing the anterior direction (St. Jude Medical Infinity), image courtesy of the official website (https://www.sjm.com).

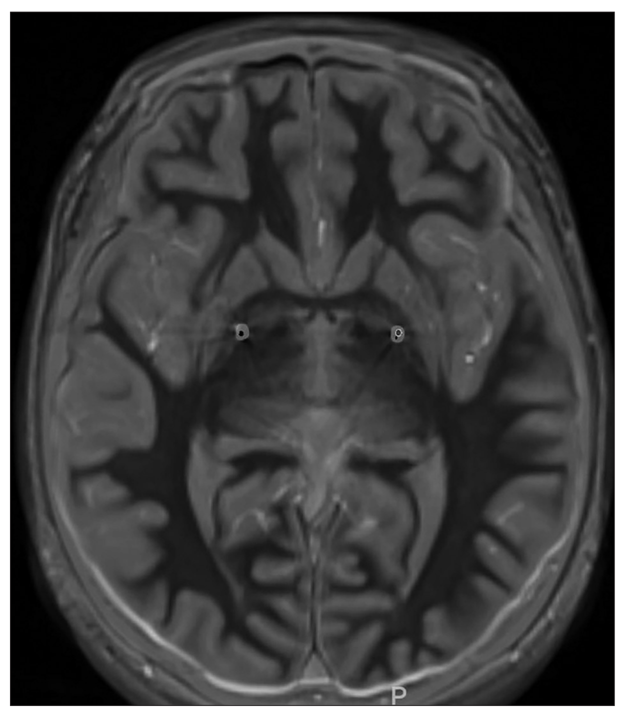

Fig. 2. Merged postoperative computed tomography and preoperative magnetic resonance image of a patient with microcephaly showing appropriate placement of the deep brain stimulation electrodes in the bilateral globus pallidus pars interna. The dots indicate the left and right of the preoperative magnetic resonance image target, respectively, and the surrounding high density indicating the electrode lead tip in the postoperative computed tomography image.

ting, the symptoms eventually diminished. No complications were observed, and the patient did not have any complaints after the final setting was confirmed.

DISCUSSION

DBS is an effective surgical treatment for patients with refractory dystonia, and it has been approved by the FDA. A successful DBS mainly depends on optimal target selection and precise implantation of electrodes to the target region. Several studies have shown that GPi should be targeted to improve motor dysfunction in individuals with primary dystonia[1)4)6)7)] and that the optimum therapeutic target was the posterolateroventral region of the GPi.[8)12)] However, targeting the accurate region remains a challenge for all neurosurgeons, particularly in patients with anatomical variations. In patients with distorted and atypical brain anatomy, direct visualization and targeting with MRI are crucial. DBS using directional leads theoretically provides a more precise control of stimulation leading to effective DBS and lesser side effects.[5)] Directional leads differ from the conventional DBS leads that use radially segmented electrodes that allow the field to move the plane in a perpendicular manner or to use anodes and cathodes allowing stimulation in the direction of each patient's individual anatomical variety.[5)] In our case, the patient was diagnosed with microcephaly, and she had an obvious anatomical variation. Smaller anatomical structures result in a narrow therapeutic window, which may lead to an unsuccessful lead positioning. The classical coordinates for the GPi were placed 21mm lateral from the midplane, 2mm anterior to the midcommisurral point, and 5mm below the intercommissural line.[2)] However, based on preoperative MRI, the target coordinates of our patient were located 17mm lateral from the midcommisural line, 2mm anterior to the midcommisurral point, and 5mm below the intercommissural line, which were far medial and caudal. MRI-guided stereotactic targeting resulted in a successful targeting of the GPi in such patient.

By inserting bilateral directional leads, the stimulation field that is perpendicular to the lead can be altered, which precisely controls the volume of tissue activated in a narrow therapeutic window. In addition, stimulation toward the functional target and away from the side-effect structure results in a higher side-effect threshold.

CONCLUSION

Most patients with movement disorder have a normal brain anatomy. However, some have a distorted or undeveloped brain anatomy. Direct targeting is mandatory for these patients, and directional lead stimulation improves DBS stimulation precision and lowers potential side-effects in such case.

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