

Case Report

Anesthesia management of deep brain stimulation insertion in a patient with generalized idiopathic dystonia: a case report

Junghee Choi¹, Jinsol Lee¹, Youngduck Shin^{1,2}, Soontaek Jeong^{2,3}, Jeongeun Moon¹, Sanghi Park¹

¹Department of Anesthesiology and Pain Medicine, Chungbuk National University Hospital, Cheongju, Korea;

²Department of Anesthesiology and Pain Medicine, Chungbuk National University College of Medicine, Cheongju, Korea; ³Charm Pain Clinic, Cheongju, Korea

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Abstract: Dystonia is a movement disorder in which excessive muscle contractions cause abnormal movement. It is classified according to its clinical manifestations (onset, distribution, temporal and associated features) and etiology (pathology and inheritance). Deep brain stimulation (DBS) is a surgical procedure used to treat medically intractable dystonia. In this study, we aim to share our experience with general anesthesia in systemic idiopathic dystonia that was not controlled by drugs, along with a literature review. A 21-year-old man with generalized idiopathic dystonia and developmental delay was scheduled to undergo deep brain stimulator implantation under general anesthesia. Intubation of the endotracheal tube and fixation of the stereotactic frame were performed in the intensive care unit (ICU) under sedation and neuromuscular blockade before arrival at the operating room. Total intravenous anesthesia was administered. After an uneventful surgery, the patient was discharged to the ICU with an endotracheal tube. As dystonia has a wide clinical spectrum and DBS requires special anesthetic considerations, anesthesiologists should adopt proper anesthetic depth and neuromuscular blockade according to each patient's condition.

Keywords: Idiopathic generalized dystonia, deep brain stimulation, general anesthesia

Introduction

Dystonia refers to dyskinesia that causes abnormal posture or movement due to continuous or intermittent muscle contraction [1]. It is a common form of movement disorder [2] and is classified according to clinical features based on age and site of onset, temporal distribution, accompanying symptoms, as well as according to the mechanisms based on pathology and heredity. Many types of dystonia exist with various treatments, including counseling, physical therapy, oral medications, botulinum toxin injection, and surgical intervention [1]. Deep brain electrode insertion can be performed for abnormal movement disorders that do not respond to drugs [3]. It has the peculiarities of stereotactic frame installation, magnetic resonance imaging, and arousal surgery, and requires careful airway management and anesthesia maintenance. For safe anesthesia and airway management in patients with dystonia,

anesthesiologists must select the appropriate anesthesia method for each patient after identifying the patient's movement, posture, and comorbidities in the pre-anesthetic evaluation. In this study, we aim to share our experience with general anesthesia in systemic idiopathic dystonia that was not controlled by drugs, along with a literature review.

Case

A 21-year-old man (height, 160 cm; weight, 30 kg) underwent deep brain stimulator implantation for uncontrolled systemic idiopathic dystonia. The patient was first diagnosed with a developmental disorder at the age of 2 years and continued to receive drug treatment at the Department of Pediatrics and Psychiatry to manage autism spectrum disorder and cerebral palsy. However, at the age of 20 years, the dystonia worsened, and the symptoms of bending the back and neck backward, and losing



Figure 1. Deep brain stimulation electrodes. Lateral X-ray of electrodes (white, upper right) in the brain.

strength of the whole body and falling forward were observed.

There were no abnormal findings in the preoperative blood test and electrocardiogram, and there were no specific findings except for mild scoliosis on a simple chest radiograph.

Since brain magnetic resonance imaging (MRI) failed despite sufficient sedation due to exacerbation of dystonia; on the morning of the surgery, after muscle relaxant administration in the intensive care unit, endotracheal intubation, head frame installation, brain MRI, and admission to the operating room were performed. Midazolam 3 mg, remifentanyl 0.2 mcg/mL, atracurium 6 mcg/mL, and succinylcholine 400 mg were intravenously administered in the MRI room 1.5 h before moving the patient to the operating room. The patient entered the operating room with ambu bagging of 5 L/min of oxygen. After arriving at the operating room, non-invasive blood pressure monitoring, electrocardiogram, pulse oxygen saturation, waveform variable index, and esophageal temperature were initiated. The following parameter values were noted: blood pressure, 169/112 mmHg; heart rate, 97/min; and oxy-

gen saturation, 99%. Glycopyrrolate 0.2 mg, lidocaine 40 mg, propofol at a target concentration of 1.0 $\mu\text{g}/\text{mL}$, and remifentanyl at a target concentration of 1.0 ng/mL were intravenously administered to induce anesthesia. Trans of Four (TOF) was confirmed to be 0%, and no additional administration of neuromuscular blockers was performed. Mechanical ventilation was initiated, the end-tidal carbon dioxide partial pressure was maintained at approximately 31 mmHg, and a ductus arteriosus was inserted into the right radial artery for continuous blood pressure monitoring. Thirty minutes after anesthesia induction, the patient's movement was detected before the operation began, and 20 mg of rocuronium was administered.

Subsequently, TOF was 0%. To maintain anesthesia, propofol was injected intravenously at a target concentration of 2.0 to 2.5 $\mu\text{g}/\text{mL}$ and remifentanyl at a target concentration of 0.1 to 1.0 ng/mL. The operation was completed with a stable systolic blood pressure of < 130 mmHg. It was confirmed that the patient's TOF was consistently 0%, and sugammadex 120 mg was injected intravenously to ensure recovery from the neuromuscular blockade. Since spontaneous breathing was insufficient, the patient was kept in the intensive care unit while maintaining intubation (**Figure 1**). After monitoring the patient for more than 1 h in the intensive care unit, he recovered stable spontaneous breathing and was extubated. The next day, he was transferred to a general ward, and he was discharged a week later as his condition improved to the point where he was able to use a wheelchair. Currently, he is able to live a sedentary life, and his dystonia in the trunk and neck has improved.

Discussion

Dystonia is generally defined as repetitive twisting movements or chronic or intermittent excessive muscle contractions [1]. In addition, it

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is classified according to the age of onset, site of involvement, temporal distribution, and clinical features of accompanying symptoms. There are many types of dystonia depending on the cause [4]. Various treatments are available, including counseling, physical therapy, oral medications, botulinum toxin injection, and surgical intervention. Deep brain stimulation (DBS) is a surgical intervention method for dystonia when the patient does not respond to conservative treatments, such as drug therapy [1, 5]. Stimulation electrodes were implanted under general anesthesia and positioned based on the direct anatomy of the posteroven-trobasal globus pallidus internus (GPi). GPi stimulation is an effective treatment for patients with dystonia with *DYT1* mutations [5].

To prepare for a stereotactic procedure, such as deep brain stimulator implantation, it is necessary to check for a blood coagulation disorder, including medication history. The most common type of anesthesia used for patients undergoing deep brain stimulator implantation is monitored anesthetic care or local anesthesia with mild sedation [6]. This is because this procedure requires intraoperative neurophysiological monitoring of awake patients and precise localization of the target nuclei [7]. Therefore, it is necessary to ensure adequate sedation and analgesia during surgery and thorough monitoring to prevent excessive respiratory depression or the disappearance of symptoms [8].

However, general anesthesia may be planned for patients who cannot cooperate because of accompanying developmental disabilities, such as in the present case [9]. Therefore, it is necessary to perform a brain MRI under sedation after fixing the stereotactic frame before surgery; however, the frame makes mask ventilation and endotracheal intubation difficult. The patient had dystonic movements, including intermittent retrocollis, and only mild scoliosis was observed on chest radiography. Although the frequency of difficult endotracheal intubation has been reported to be low in cervical torticollis, a representative of focal dystonia [10, 11], anesthesiologists should consider the possibility of difficult endotracheal intubation in advance and conduct a thorough interview and physical examination. In particular, our patient had a history of failed brain MRIs

despite sufficient sedation. Therefore, considering the possibility of respiratory depression, airway failure, and imaging failure, endotracheal intubation and head frame installation were performed after sedation and neuromuscular block in the intensive care unit, and a brain MRI was performed. Therefore, it is important to determine anesthesia and airway management methods in advance according to the patient's condition.

Unlike awakening surgery, there are no restrictions on anesthetic agents, because microelectrode recordings are impossible during general anesthesia. Since anesthetics do not uniformly affect brain regions, there have been insufficient studies on how anesthetics affect microelectrode recordings [12, 13]. We administered total intravenous anesthesia with propofol and remifentanyl. Continuous blood pressure monitoring was performed through the ductus arteriosus because increased blood pressure could cause intracerebral hemorrhage as a complication. After anesthesia induction, additional intravenous rocuronium was administered based on TOF monitoring and patient movement. This patient was sedated, and neuromuscular blocking agents were administered before the patient entered the operating room; however, TOF monitoring was not performed in the intensive care unit and MRI room. Therefore, it was difficult to predict the appropriate dose of neuromuscular blocking agents when inducing general anesthesia in the operating room. Accordingly, the patient was extubated after injecting sugammadex at a sufficient dose, and spontaneous breathing was monitored for more than 1 h in the intensive care unit.

Tsaroucha et al. reported an anesthetic method for Inguinal hernia repair surgery for patients with focal dystonia [11]. The patient had cervical dystonia called spasmodic torticollis (ST), which is the most common primary focal dystonia [14]. Recently, DBS has been known to be an effective first-line treatment for ST [15, 16], and patients underwent surgery while being treated with DBS. Bhojar et al. also reported the experience of DBS anesthesia in patients with Meige's syndrome [17]. Meige syndrome is a rare form of facial dystonia. However, in the case of refractory cases, pallidal DBS was reported to be effective [18, 19]. In this patient,

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Table 1. Comparison of other dystonia cases

Include the literature	Case	Athanasia [11]	Kalpesh [17]
Disease	Generalized idiopathic dystonia	Idiopathic focal dystonia	Meige's syndrome
Type of surgery	Deep brain stimulation insertion	Inguinal hernia repair	Deep brain stimulation insertion
Sedative agent	midazolam 3 mg propofol 1.0 mcg/mL by TIVA	propofol 2.5 mg/kg by TIVA	Dexmedetomidine 1 mcg/kg (60 mcg) continuous IV infusion @ 0.5 mcg/kg/h
Neuromuscular blockage agent	atracurium 6 mcg/cc, succinylcholine 400 mg, rocuronium 20 mg	rocuronium 70 mg	vecuronium 6 mg
Neuromuscular block reversal agent	sugammadex 120 mg	sugammadex 140 mg	neostigmine and glycopyrrolate
Complication	none	none	none

TIVA: total intravenous anesthesia.

dexmedetomidine was used for anesthesia, and hemodynamic stability was maintained. **Table 1** compares the generalized idiopathic dystonia anesthesia in the current study with the aforementioned anesthesia experiences in the literature. Dexmedetomidine, a relatively selective alpha2-adrenoceptor agonist, is suggested as an anesthetic for patients with dystonia by minimizing respiratory function deterioration and maintaining hemodynamic stability [20].

The use of local or general anesthesia during deep brain stimulator implantation is not contraindicated. With advances in medicine, the survival rate of patients with rare diseases, such as dystonia, will increase. As the indications for DBS increase, there is an increased risk of patients entering the operating room, or of operation in other departments for patients who already have undergone DBS. Therefore, standardized anesthesia protocols for patients undergoing DBS transplantation as well as orthopedic and gynecological surgeries, unrelated to neurosurgery, for patients of DBS transplantation are required.

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Disclosure of conflict of interest

None.

Address correspondence to: Dr. Sanghi Park, Department of Anesthesiology and Pain Medicine, Chungbuk National University Hospital, 1 Chungdae-ro, Seowon-gu, Cheongju 28644, Korea. Tel: 82-43-269-7492; Fax: 82-43-269-6238; E-mail: happycat19@hanmail.net

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