CASE REPORT

Anesthesia for deep brain stimulation in a patient with X-linked dystonia-parkinsonism/Lubag disease

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Abstract : Lubag disease is a genetic X-linked dystonia-parkinsonism syndrome afflicting Filipino men. This disease is characterized by dystonia dominating the first 10-15 years of the disorder, which is associated with or replaced by parkinsonian features in later years of life. A 49-year-old man with Lubag disease underwent general anesthesia for deep brain stimulation (DBS) surgery. Anesthesia was maintained mainly with propofol, remifentanil, rocuronium bromide, and sevoflurane. During magnetic resonance imaging, the patient was anesthetized with midazolam, fentanyl, and rocuronium bromide. The surgery was completed safely using these anesthetic agents. After DBS, some symptoms including involuntary movement improved within 10 days. J. Med. Invest. 60 : 146-148, February, 2013

Keywords **:** *lubag disease, dystonia, parkinsonism*

INTRODUCTION

Lubag disease, X-linked dystonia-parkinsonism (XDP), is an X-linked recessive progressive neurodegenerative dystonia-parkinsonism that is endemic to the island of Panay in the Philippines (1, 2). The onset of XDP occurs in adulthood and by the end of the third or early fourth decade manifests predominantly as dystonia. Dystonia usually manifests focally and becomes generalized or multifocal after 5 years in most patients. The disease plateaus until the 10th year of the illness and then its severity reduces or is replaced by Parkinsonian symptoms by the 15th year of illness (1, 2). Evidente *et al.* reported the first case of XDP that demonstrated a good response to bilateral pallidal stimulation under local anesthesia (3). Here, we report a case of XDP who presented to us for deep brain stimulation (DBS) under general anesthesia.

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CASE PRESENTATION

The patient was a 49-year old Filipino male (height, 162 cm ; weight, 45 kg) having maternal roots in the Philippine Island of Panay. He first presented with involuntary movements of bilateral halluces at the age of 41 years. The condition began with involuntary movements, his ambulatory ability worsened progressively and he developed generalized dystonia. On presentation, the patient had dystonic symptoms mainly on the left side of his body with a retracted head, twisted trunk, and involuntary movements of his upper limbs. He also had difficulty

walking and dysarthria because of a twisted trunk ; however, no communication difficulties were observed.

Magnetic resonance imaging (MRI) studies revealed atrophy of the caudate and putamen. The patient had a medical history of hypertension without medication and a family history of dystonia. His mother was unaffected carrier and his elder brother had dystonia.

No premedication was administered. An electrocardiography was conducted in the patient. His oxygen saturation, heart rate, body temperature, and urine output volume were monitored. Anesthesia was induced with propofol (1.3 mg/kg), remifentanil (0.3 μg/kg/min), and rocuronium bromide (0.6 mg/kg). A stereotactic frame was fixed to the patient's skull in the operating room after tracheal intubation. The patient was transferred to the MRI suite to identify target nuclei and determine surgical planning. Anesthesia was maintained during MRI with intravenous fentanyl (100 μg), midazolam (10 mg), and rocuronium bromide (30 mg), and the patient was returned to the operating room 1 h after MRI. A burr hole was made in the cranium to insert a DBS electrode that was passed down to the target area using microelectrode recording (MER) to identify brain structures and conduct clinical testing. Specific brain structures were identified based on the unique MER neuronal firing patterns, and the electrode was inserted at the exact target location. Anesthesia was maintained with infusions of propofol (4.0 mg/kg/h), remifentanil (0.3 μg/kg/min), sevoflurane (0.8%), and rocuronium bromide (3.7 μg/kg/min). A pulse generator was subsequently implanted in the subclavicular subcutaneous area and connected to the electrode extension cable. Anesthesia was maintained during this procedure with sevoflurane (1.0-1.5%), remifentanil (0.15-0.3 μg/kg/min), and rocuronium bromide (3.7 μg/kg/min). This patient did not present with Parkinsonian symptoms at the time of surgery. After the surgery, sugammadex (4 mg/kg) was injected and the patient regained consciousness. He was alert and oriented and therefore extubated. Total surgical time was 434 min, and anesthesia time was 591 min. Total infusion volume was 2,900 ml, bleeding was 80 ml, and urine output volume was 2,025 ml.

The stimulation was started 1 day after the surgery, and involuntary movements of the patent's upper limbs, difficulty walking, and dysarthria improved markedly within 10 days.

DISCUSSION

DBS is used for treating patients with neurogenic disorders such as Parkinson's disease, essential tremors, dystonia, and certain psychiatric conditions (4). DBS is a procedure that includes placing electrodes into deep brain structures. The DBS electrode is connected to an implantable pulse generator (5).

Anesthetic techniques vary depending on the traditions and requirements of each institution performing these procedures (5, 6). In our hospital, a stereotactic frame is usually placed on the patient's skull followed by MRI for targeting using three-dimensional optical marker-based technology under local anesthesia. After MRI, general anesthesia is initiated to insert the electrode into the target area for stimulation and implantation of the pulse generator in the subclavicular subcutaneous area.

The awake technique has an obvious advantage during the mapping phase because it accurately detects cellular activity and movement-related responses to neurostimulation (5). In addition, if the patient is conscious and cooperative, intraoperative electrical stimulation and clinical improvement can be observed (7).

However, general anesthesia may be necessary in children and adults who cannot tolerate the awake technique. Total intravenous and inhalation anesthesia have been used in patients who are considered unsuitable for the awake technique (5, 8, 9). In this case, the patient presented with severe dystonia and was restless ; therefore, general anesthesia was selected.

Intravenous anesthesia is preferred over volatile anesthesia for DBS anesthetic management in many cases (8, 9). It is unclear which general anesthesia agents are the most effective and suitable for this procedure. Trombetta *et al.* reported delayed awakening after propofol anesthesia during DBS for dystonia (10) ; therefore, we used low dose sevoflurane to minimize propofol dose. Propofol administration was stopped when the pulse generators were implanted in the subclavicular subcutaneous area.

Propofol causes a significant decrease in target nuclei neuronal activity (7, 8). Steigerwald *et al.* reported that overall neuronal firing rates in patients with dystonia decrease (11), differences in globus pallidus internus neuronal firing rates occur between patients with dystonia and Parkinson's disease, and the effect of anesthesia may be more pronounced in patients with Parkinson's disease (5). These

changes may interfere with localization of MER-based DBS electrodes (6) ; therefore, administering propofol before electrophysiological mapping begins is not recommended (10). In addition, Pralong *et al.* reported that low doses of propofol and sevoflurane influence pallidal activity in a similar way (12) ; therefore, studies to prove the anesthetic agents that are the most effective and suitable for this surgery must be conducted.

We anesthetized a patient with XDP disease who underwent DBS. XDP is a rare disease and its mechanism is unclear ; however, the operation was completed safely by the same anesthesia method as the one for other dystonia patients.

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