

Anesthetic management for deep brain stimulation in a patient with pantothenate kinase-associated neurodegeneration

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Key points

Pantothenatekinase-associated neurodegeneration is a rare progressive disorder characterized by dystonia, rigidity, choreoathetosis and mental deterioration. Deep brain stimulation of different targets in the basal ganglia circuitry can be a choice in patients who do not give response to medical treatment. Patients requiring general anesthesia with this syndrome may have many anesthesia-relevant symptoms that influence the preanesthetic management, the induction of anesthesia and the postoperative care.

Abstract

Pantothenatekinase-associated neurodegeneration is a rare progressive disorder characterized by dystonia, rigidity, choreoathetosis and mental deterioration. Patients requiring general anesthesia with this syndrome may have many anesthesia-relevant symptoms that influence the preanesthetic management, the induction of anesthesia and the postoperative care. In this case report, we present the anesthetic management of a 10-year-old male scheduled for bilateral Globus pallidus internus stimulation for pantothenatekinase-associated neurodegeneration under general anesthesia. The patient had to be sedated before the operation because of the extreme dystonia. Anesthesia was induced uneventfully by the use of modern anesthetic agents and a multidisciplinary approach.

Keywords: Pantothenate kinase-associated neurodegeneration, Hallervorden Spatz disease; deep brain stimula-

tion; children; airway management

Background

Pantothenate kinase-associated neurodegeneration (PKAN) was first described by two German fellows: the neuropathologist Julius Hallervorden and the neurologist Hugo Spatz in 1922 (1). The active involvement of Hallervorden in euthanasia in Germany during World War II and the discovery of the defective gene for this disease removed the name “Hallervorden Spatz disease” to PKAN (2,3). The disease is characterized by progressive generalized dystonia involving the oromandibular muscles, rigidity and choreoathetoid movement (4). Medical treatments can be incapable of reversing dystonic symptoms and deep brain stimulation (DBS) of different targets in the basal ganglia circuitry can be a choice. We present a case with PKAN undergoing DBS surgery. A legal representative of the patient consented to publication.

Case report

A 10-year-old, 20-kg male was scheduled for bilateral Globus pallidus internus (GPI) stimulation for PKAN under general anesthesia. The disease was diagnosed at age 6 years when he exhibited developmental delays in motor skills. In the years following diagnosis, treatment with Baclofen, Levadopa, Pramipexole and Trihexyphenidyl hydrochloride produced minimal improvement. Magnetic resonance imaging (MRI) of the brain revealed diagnostic “eye of the tiger” lesions within the globus pallidus (Fig. 1).

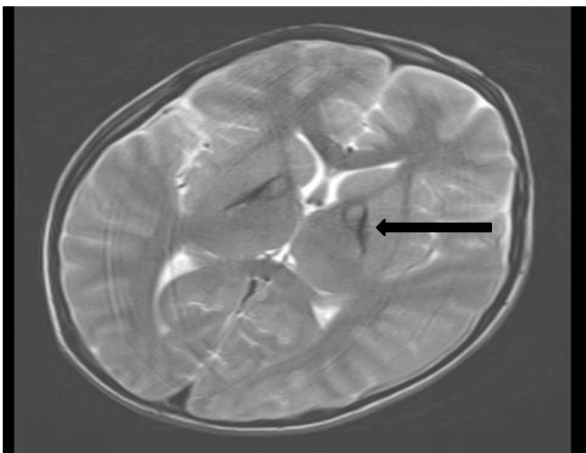


Figure 1. "Eye of the tiger" lesion on MRI

By age 10 years, he had swallowing difficulties and a percutaneous endoscopic gastrostomy tube was planned for nutrition. But he was admitted to the emergency service because of dystonic storm; extreme dystonia causing self-inflicted injuries. The patient was accepted to the pediatric intensive care unit (ICU) to control severe spasms. Physical examination revealed a skinny patient (BMI: 15.1 kg/m²) with normal vital signs. He had limited mouth opening because of trismus, frequent choreoathetoid movement in all limbs, extreme rotation of head and neck and severe dystonia. His feet were deformed bilaterally. He was unable to sit or walk (Fig 2). Midazolam infusion had to be begun and bilateral DBS of the GPI was planned. Serum biochemical and hematological profiles were normal. The entire surgical pro-

cedure was planned to perform under general anesthesia. In the operative room, standard monitoring including electrocardiography, pulse oximetry, capnography, non-invasive blood pressure and body temperature measurements were established.



Figure 2. Dystonic storm of the patient

Radial artery and urinary bladder catheters were placed. Anesthesia was induced with propofol (2 mg/kg) and remifentanyl (0.3 µg/kg). After controlling ease of face-mask ventilation, neuromuscular block with vecuronium (0.1 mg/kg) was achieved and the trachea was intubated with a 5.5 mm cuffed endotracheal tube with no difficulty. Anesthesia was maintained with remifentanyl infusion (0.1 µg/kg/min) and sevoflurane 1-2% inspired in O₂ and air. After making scalp block (bilaterally blockage of supraorbital, auriculotemporal and greater occipital nerves) with bupivacaine 0.25% 2 ml at each site, a stereotactic Leksell frame was applied to the head of the patient and the patient was transferred to the intraoperative MRI suite to obtain localizing images. In MRI suite, anesthesia was maintained with sevoflurane 2% inspired in O₂ and air by a MRI compatible anesthesia machine. After 30 minutes, the patient was transferred to the operating room. A bicoronal linear scalp incision was made and bilateral burr holes were opened. Low dose sevoflurane (1%) and remifentanyl (0.05-0.1 µg/kg/min) infusion with no additional neuromuscular blockers

were preferred as an anesthetic technique so as to monitorize the response of electrode stimulation. Using computer-assisted stereotactic techniques based on MRI data, a microelectrode was inserted at the exact target location. As the patient was anesthetized, the immediate clinical effect of the lesioning and possible side effects could not be determined. A pulse generator was then implanted in the subclavicular subcutaneous area and connected to the electrode extension cable. The stimulation was started 1 day after the surgery. Anesthesia time was 525 min, total surgical time was 450 min, urine output volume was 200 ml and total infusion volume was 800 ml. After the procedure, the patient was transferred to the ICU, mechanical ventilation was continued and sedation was provided over night with remifentanyl and midazolam infusions to allow slow, gradual emergence. Next day the sedation was stopped and endotracheal tube was removed with no accident when the patient awoke, with stable hemodynamics (HR: 90 bpm, BP: 90/55 mmHg, RR: 16/min) and arterial blood gases (pH: 7.40, PaO₂: 110 mm Hg, PCO₂: 42.0 mmHg). As dystonia came back after 3 hours, sedation with dexmedetomidine (0.3 µg/kg/hr) was began. Endotracheal intubation was not needed. His dystonia had considerably diminished as a result of the surgery and the patient was discharged from the ICU 10 days after surgery.

Conclusions

Patients with PKAN requiring surgery under general anesthesia are usually in a status of uncontrolled dystonia and rigidity. They may require intensive care because of this life-threatening state. Dystonia may involve oromandibular muscles leading swallowing impairment, malnourishment and pulmonary aspiration (5). Dynamic upper airway obstruction and breathing difficulty can be seen as a result of involvement of pharyngeal muscles (6). Contractures, jaw and cervical spine stiffness can cause difficult airway. Detailed preanesthetic evaluation is not always easy because of associated mental retardation. Endotracheal intubation may be difficult; awake intubation techniques are not suitable as noxious stimu-

lation can intensify the dystonia and involuntary movements. Physicians should be aware that emergency tracheostomy can be needed. We made a limited examination as our patient had to be sedated preoperatively because of his severe dystonia. Airway anatomy seemed to be unaffected by his chronic disease state. Induction of anesthesia was uneventful as our patient was also under sedation. Limited case reports also show most dystonia get relieved after intravenous anesthetics (6,7). There was no difficulty in endotracheal intubation.

DBS surgery may be considered in PKAN patients with severe dystonia who have not responded to medical therapy (8). Awake or slightly sedated patients are preferred so as to monitorize the response of electrode stimulation. But our patient was not a suitable candidate for an awake procedure because of his gross pathologic movements. Total intravenous or low dose inhalation anesthesia can be used in DBS surgery. In many cases intravenous anesthesia is preferred over volatile anesthesia but it was also shown that propofol can decrease target nuclei neuronal activity (9,10). In our institution we make a scalp block with bupivacaine before placing the stereotactic frame. This provides us to use low dose sevoflurane and remifentanyl without muscle relaxants. Additional propofol boluses are used if it is needed by avoiding to administer just before the electrophysiological mapping.

For most case reports with PKAN patients, as dystonia came back postoperatively after the anesthetics wore off, a delayed tracheal extubation was suggested (6,7). Patients should be monitored longer than usual in the recovery room or admitted to the intensive care unit. We preferred to send our patient intubated in ICU. Endotracheal tube was removed next day without any difficulty under the activation of DBS. Dexmedetomidine infusion was used effectively without any adverse effects in the period of modification of stimulation parameters.

In conclusion, PKAN is a rare disease. DBS is an effective treatment for intractable generalized dystonia of these patients. Anesthetic management begins preopera-

tively by sedation to control severe dystonia. Detailed pre anesthetic evaluation and postoperative care are critical. Modern anesthetics and techniques can be used without problems.

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