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Anesthetic Management of a Patient with Amyotrophic Lateral Sclerosis undergoing Percutaneous Endoscopic Gastrostomy

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This is a case of a 62-year-old female with amyotrophic lateral sclerosis (ALS) and bulbar involvement. The patient was scheduled for percutaneous endoscopic gastrostomy under general anesthesia. The TOF stimulation was used to monitor neuromuscular blockade from the beginning of the induction until extubating process. General anesthesia was induced by intravenous administration of propofol and was intubated with rocuronium. General anesthesia was maintained by sevoflurane. Pain control with local infiltration of 0.5% bupivacaine at the surgical site without an intravenous opioid. The patient emerged from general anesthesia smoothly and was extubated after an administration of sugammadex without any complications. This case report showed that rocuronium and sugammadex could be used safely in these patients; however, neuromuscular monitoring and clinical correlation should be performed to determine the muscle power to ensure the extubating process, and further research is needed to determine the appropriate dose of rocuronium and sugammadex for ALS patients.

Keywords: Amyotrophic lateral sclerosis; Bulbar symptom; Rocuronium; Sugammadex

Amyotrophic lateral sclerosis (ALS) is a rare disease. The incidence was 2.16 per 100,000 person-years in the general European population[1] and 3.9 per 100,000 person-years in the general U.S. population.[2] The progressive neurodegenerative disease, which involves both upper and lower motor neurons, causes symptoms, including muscular weakness, atrophy, fasciculation, spasticity, and hyperreflexia. Most patients died from respiratory failure within 3 to 5 years. ALS typically begins in extremities; however, thirty percent of the cases have bulbar involvement. The symptoms include difficulty in chewing, speaking, swallowing, and showing facial expression.[3] The etiology of the disease remains unknown, but ten percent of the cases appear

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to be familial, and 90% are sporadic.[4] There is no curative treatment for ALS, and all current therapies are focused on palliative treatment.[3,5,6] ALS patients commonly require surgical procedures such as percutaneous endoscopic gastrostomy, tube replacement, and tracheostomy.[5] An improvement in the understanding of anesthetic considerations in ALS patients is necessary to increase awareness and promote patient safety. The anesthetic management of ALS patients undergoing general anesthesia is reported.

Case Report

A 62-year-old female underwent an elective percutaneous endoscopic gastrostomy under general anesthesia. She was previously diagnosed with ALS for nine months and had progressive bulbar symptoms, including hoarseness, difficulty swallowing, sialorrhea, and slurred speech. She had undergone general anesthesia for colectomy without complications eight years ago. Medications for symptom control are riluzole, mecobalamin, and folic acid. On examination, the patient had a positive gag reflex, positive tongue atrophy, hoarseness of voice, and grade V motor power of all extremities without sensory losses. Preoperative laboratory findings showed that the chest radiography and electrocardiography were normal. In addition, her pulmonary function test was preserved. ALS Functional Rating Scale-Revised (ALS-FRS-R) was 42.

The patient received preoperative aspiration prophylaxis with omeprazole and metoclopramide. With standard ASA monitoring and peripheral nerve monitoring (train-of-four, TOF) at right ulnar nerve, general anesthesia was induced with propofol 2 mg/kg after the patient fell asleep. The TOF ratio was measured for baseline. A total dose of 0.6 mg/kg of rocuronium was administered via a 20-gauge IV cannula by titrating in doses of 0.3 mg/kg with continuous TOF monitoring every 15 seconds. Then, the airway was secured with a No.7 endotracheal tube when the TOF reached 2 responses, and anesthesia was maintained with 0.4% mixture of air-oxygen (FiO2) and 2% sevoflurane. Pain control with local infiltration of 0.5% bupivacaine by a surgeon at the surgical site without intravenous opioid was used. The operation was successful in 25 minutes without surgical complications. TOF count was 2, and 2 mg/kg of sugammadex was administered. Four minutes after sugammadex administration, the TOF ratio was 0.97. The patient had adequate spontaneous breathing (5.5 mL/kg) and was responded to verbal commands. The patient was later extubated. She had an uneventful recovery in the post-operative period.

Discussion

Even though ALS is a rare disease, it is vital to understand its clinical manifestation and peri-operative management. ALS is a disease that primarily affects motor neurons, so it raises a concern toward anesthetic techniques. Dysfunction of motor neurons causes bulbar dysfunction, such as speech, salivation, swallowing. This increases the risk of aspiration, peripheral nerve weakness, muscle weakness, and muscular atrophy of limbs and respiratory muscles. The degree of functional impairment was assessed with pulmonary function test and ALS Functional Rating Scale-Revised (ALS-

FRS-R). The ALS-FRS-R[7] includes 12 questions, which assess the functional impairment in four functional domains: bulbar function, fine motor, gross motor, and respiratory function, with the overall scores ranging from 0 to 48. A preoperative pulmonary function test can predict the risk of ventilatory support postoperatively. Pulmonary function tests should include vital capacity, inspiratory capacity, and negative inspiratory force. The American Academy of Neurology recommended that ALS patients with FVC less than 50% receive non-invasive respiratory ventilation.[8]

Because ALS patients with advanced bulbar symptoms had an increased risk of aspiration, an intraoperative drug should be a short-acting analgesic. The amnestic agents, such as propofol and remifentanil infusion, are an ideal drug of choice. Inhalation agents are preferred to be either desflurane or sevoflurane rather than isoflurane because of low lipid solubility and faster recovery time. Moreover, extubation should be performed while the patients are fully awake.[5]

ALS is a neuromuscular disorder. The drug, which depolarizes the neuromuscular blocker such as succinylcholine, is contraindicated. This type of drug can cause severe elevation of serum potassium and contribute to cardiac arrest.[9] Nondepolarizing neuromuscular blockers are preferred and should be used with caution as these agents can also prolong the drug duration in ALS patients. Some reports showed that sugammadex could be used to reverse safely in ALS patients with an intravenous dose of 2 mg/kg.[10] However, a previous case report demonstrated residual paralysis after rocuronium reversal with a standard dose of sugammadex. The patient required a higher dose of sugammadex to reach the adequate TOF ratio and tidal volume for safe extubation.[11]

In this case report, the patient had only bulbar symptoms without limb weakness and respiratory problems. For the intubation process, at first, we decided to apply 0.3 mg/kg of rocuronium[10] but after 60 seconds, the TOF ratio was 0.93 then we added another 0.3 mg/kg of rocuronium to provide the TOF ratio appropriate for tracheal intubation and the 2 mg/kg of sugammadex was used in the reversal process. This showed that we could not predict the dose of rocuronium and sugammadex.

Although there are case series that percutaneous endoscopic gastrostomy in ALS patients was safely done under monitored anesthesia care with sedation[12,13], we decided to choose general anesthesia with muscle relaxant with endotracheal intubation for this patient because of the patient's aspiration history to make certain that the airway was secured.

Eventually, it is necessary to evaluate the degree of functional impairment. Although the patient has mild symptoms, it is critical to meticulously select the neuromuscular blocker, closely monitor neuromuscular outcomes, and assess clinical correlation such as vital capacity.

Conclusion

This case report showed that rocuronium and sugammadex could be used safely in these patients; however, neuromuscular monitoring and clinical correlation should be performed to determine the muscle power to ensure the extubating process, and further research is needed to determine the appropriate dose of rocuronium and sugammadex for ALS patients.

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