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Case Report

Total intravenous anesthesia without muscle relaxant for pulmonary wedge resection in a patient with amyotrophic lateral sclerosis: a case report

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Abstract: Muscle relaxants may exacerbate the symptoms of amyotrophic lateral sclerosis (ALS). Furthermore, ALS patients often experience respiratory muscle weakness. Herein, we report the case of a 63-year-old man with ALS who underwent pulmonary wedge resection using total intravenous anesthesia without muscle relaxant and single lumen endotracheal tube. After an unremarkable surgical procedure, the patient was transferred to the intensive care unit after extubation. The patient did not experience any worsening of ALS symptoms over the one-year follow-up period. Our experience shows that total intravenous anesthesia without muscle relaxants can be used as an anesthetic method for lung surgery in ALS patients. We report this case along with a brief literature review.

Keywords: Amyotrophic lateral sclerosis, neurodegenerative diseases, general anesthesia, neuromuscular blocking agents, video assisted thoracic surgery

Introduction

Amyotrophic lateral sclerosis (ALS) is a degenerative neurological disorder of the motor ganglia in the anterior horn of the spinal cord and pyramidal tracts that affects both the upper and lower motor neurons [1], with an incidence of 1-2 cases per 100,000 patient-years [2]. ALS was initially known as Charcot's sclerosis but is now more familiarly known as Lou Gehrig's disease. As ALS progresses, patients develop symptoms that may include bulbar and respiratory muscle weakness, spasticity, hyperreflexia, muscular atrophy, fasciculations, and cramps [3].

Atrophy and weakness of the respiratory and bulbar muscles can affect the management of anesthesia. In individuals with ALS, an abnormal response to muscle relaxants can lead to ventilatory depression. Furthermore, neuroaxial blocks, such as spinal and epidural anesthesia, are also relative contraindications in patients with motor neuron diseases due to the possibility of exacerbating the disease [4].

General anesthesia without muscle relaxants in ALS patients is rare but has been reported. However, the administration of total intravenous anesthesia without muscle relaxants in ALS patients undergoing lung surgery has rarely been reported. Herein, we report the case of an ALS patient who received total intravenous anesthesia without muscle relaxants for pulmonary wedge resection.

Case report

This case report was approved by the Chungbuk National University Hospital Clinical Research Review Committee (approval number 2022-01-020-001). The patient provided informed consent for its publication.

History

A 63-year-old man (weight, 65 kg; height, 164 cm) visited a neurology outpatient department for dysarthria and dysphagia that had presented five months before admission. He had been taking antihypertensive agents for ten years and choline alphoscerate 400 mg for mild cog-

Table 1. Clinical characteristics of the patient

Characteristic	Value
Age (years)	63
Sex	Male
BMI (kg/m^2)	24.2
ASA	3
Symptoms	
Sensory	-
Motor	-
Dysarthria	+
Dysphagia	+
Physical examination	
Hoffman sign	+
Tromner sign	+
Deep tendon reflex	Brisk
EMG	The electrophysical findings are in keeping with a widespread denervating process.
PFT	
FEV1 (L)	2.10
FVC (L)	2.10
FEV1/FVC (%)	100

Note: BMI = Body mass index; ASA = Anesthesiologists physical status score; EMG = Electromyography; PFT = Pulmonary function test; FEV1 = Forced expiratory volume in 1 second; FVC = Forced vital capacity.

nitive impairment for three months. The patient had no sensory complaints and no definite weakness of the limbs, besides the dysarthria and dysphagia. ALS was diagnosed by active denervation observed on the patient's electromyography, a positive Hoffman and Tromner sign, and brisk deep tendon reflex findings. At that time, the patient was prescribed 50 mg of riluzole per day.

Diagnosis

Three months after the diagnosis of ALS, a ground glass nodule, approximately 17 × 12 mm, was discovered in the left upper lobe of the lung by chest computed tomography. Positron emission tomography-computed tomography revealed a mild hypermetabolic semi-solid ground glass nodule in the left upper lobe. As malignancy could not be excluded, a wedge resection was planned for biopsy and surgical resection.

His laboratory findings were within the normal limits, except for a decreased forced expiratory volume in 1 s (FEV1) of 2.10 L (52% of predicted value) and forced vital capacity (FVC) of 2.10 L (70% of predicted value); the FEV1/FVC was 100%. The patient's blood pressure remained within the normal range, and his American Society of Anesthesiologists physical status score was 3 (**Table 1**). Although the patient desired the surgery, he wanted to avoid the use of muscle relaxants as he was concerned regarding their effect on his ALS. Therefore, after consulting with the surgeon, the patient and his family, we chose total intravenous anesthesia without muscle relaxants via a single lumen tube.

Procedure

Before anesthesia induction, the patient's blood pressure, heart rate, respiratory rate, oxygen saturation, and bispectral index value were 140/80 mmHg, 77 beats*min^{-1}, 11 breaths*min^{-1}, 100%, and 95, respectively. Preoxygenation was performed with 100% oxygen at 5 L/min with the patient in the supine position. After an injection of 0.2 mg of glycopyrrolate and 40 mg of lidocaine, we initiated a continuous anesthetic infusion with an effect-site drug concentration of 3.0 ng*mL^{-1} for remifentanil and 4.5 μg*mL^{-1} for propofol. After the patient lost consciousness, manual mask ventilation was performed with 100% O_2 for 2 min.

At the time of laryngoscopy, the patient was immobile, and his vocal cords were fixed.

Figure 1. Patient's train-of-four ratio. The train-of-four ratio was maintained at 100% from the start of anesthesia to the end of anesthesia.

Intubation was performed without difficulty, using a plain 7.5-cuffed tube. The patient was mechanically ventilated at tidal volumes of 8 $mL*kg^{-1}$ and 14-16 $breaths*min^{-1}$ with a 50:50 mixture of air/oxygen, and the end-tidal carbon dioxide was maintained between 29 and 31 mmHg.

After the Allen test, an arterial line was placed in the patient's right radial artery. Arterial blood gas analyses revealed a partial pressure of carbon dioxide (PCO_2) of 42.4 mmHg and an oxygen saturation of 96.4% with a 50% fraction of inspired oxygen. The patient's blood pressure, heart rate, and oxygen saturation were 116/84 mm Hg, 85 $beats*min^{-1}$, and 100%, respectively. The bispectral index value was 45, and the train-of-four ratio was 1.0 (**Figure 1**).

After the patient was placed in the right decubitus position and disinfected, a video-assisted thoracic surgery port was inserted into the fourth and sixth intercostal space. A yellow and gray color round mass of approximately 1.2 cm was observed.

The surgeon asked for three breathing stops: one to secure the visual field and two for wedge resections. We initiated manual ventilation approximately for one minute at tidal volumes of 8 $mL*kg^{-1}$ and 24-28 $breaths*min^{-1}$ with a 50:50 mixture of air/oxygen, and the end-tidal carbon dioxide was maintained between 25 and 28 mmHg. While the ventilation was stopped, the patient's lungs remained motionless and deflated (**Figure 2**). After the two wedge resections were performed, a 16-Fr chest tube was inserted. Upon completion of the surgery, the continuous infusion of propofol and remifentanil was terminated.

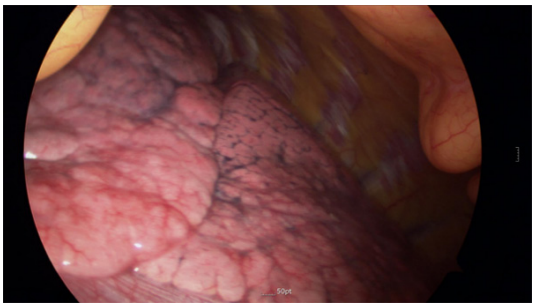

Figure 2. Image of the patient's lungs viewed using video-assisted thoracoscopy. After hyperventilation, the patient's lungs remained motionless and deflated.

Approximately three minutes later, voluntary respiration was restored with tidal volumes of 6-8 $mL*kg^{-1}$, 18-20 $breaths*min^{-1}$, and an oxygen saturation of 100%. Once the patient regained consciousness and motor power (hand grip, head elevation), oral secretion suction and tracheal extubation was performed. The total anesthesia time was 50 minutes.

The patient was moved to the intensive care unit. Ten hours post surgically, arterial blood gas analysis revealed a PCO_2 of 34.4 mmHg and an oxygen saturation of 96.5% with a 30% fraction of inspired oxygen. No worsening of neurological signs or symptoms was observed. On postoperative day 2, the patient was discharged. He has since undergone outpatient follow-up without further complications for a period of one year.

Discussion

This case describes the successful application of total intravenous anesthesia without muscle relaxants in an ALS patient undergoing pulmonary wedge resection.

In ALS patients, involvement of the upper motor neurons manifests as weakness, spasticity, hyperreflexia, and Babinski signs, and involvement of lower motor neurons causes weakness, muscular atrophy, fasciculations, and cramps. In general, muscle weakness usually begins in the extremities but eventually involves the bulbar and respiratory muscles [3]. Bulbar muscle weakness causes slurring of speech

Table 2. Anesthesia use in amyotrophic lateral sclerosis cases as reported in the literature

First author (publication year)	Type of anesthesia			Agent		Surgical procedure		Side effects
	Spinal	Epidural	General	Rocuronium	Sugammadex	Name	Site	
Hara K (1996) [1]		o				Inguinal herniorrhaphy	Abdomen	x
Panchamia JK (2020) [7]	o					Total hip arthroplasty revision	Lower extremity	x
Kim B-R (2018) [10]			o	o	x	Laparoscopic low anterior resection with ileostomy, total abdominal hysterectomy with bilateral salpingo-oophorectomy	Abdomen	x
Chun HR (2020) [11]			o	o	o	Laparoscopic nephroureterectomy	Abdomen	Incomplete neuro-muscular recovery, aspiration pneumonia
Xiao W (2017) [12]			o	x	x	Cesarean section	Abdomen	x
Lee D (2008) [13]			o	x	x	Open reduction & internal fixation, tibia	Lower extremity	x
This case			o	x	x	Video assisted lung wedge resection	Thorax	x

and leads to aphasia, while atrophy and weakness of the respiratory muscles eventually leads to respiratory failure and death.

In ALS patients, death usually occurs within a few years of symptom onset and is due to worsening respiratory failure or complications, such as dysphagia or immobility [3]. The average life expectancy after symptom onset is three years; however, at least 14% of patients live more than five years [5]. Currently, riluzole and edaravone are the only treatments approved by the United States Food and Drug Administration for ALS with extremely limited survival benefits [6]. Therefore, the management of these patients largely depends on symptom improvement and the prevention of side effects.

Anesthesiologists face many challenges in choosing the safest anesthetic method for ALS patients as must consider the nature and progression of the disease. Moreover, these patients require special care during the preoperative period for the following reasons. First, the neurons of ALS patients are considered vulnerable to muscle relaxants; therefore, they may exhibit an altered response (either depolarized or non-depolarized) to muscle relaxants. Second, spinal anesthesia has rarely been reported for ALS patients [7]. However, neuroaxial blockades are relatively contraindicated [4] because the demyelination in ALS makes the spinal cord more vulnerable to the potential neurotoxic effects of local anesthetics [8]. Further, neuroaxial blockade may exacerbate ALS due to needle injury and/or drug toxicity [9].

General anesthesia with muscle relaxants in ALS patients has often been reported. Kim et al reported the application of general anesthesia using low-dose rocuronium for laparoscopic low anterior resection in ALS patients [10]. Chun et al reported delayed neuromuscular recovery after the use of sugammadex in an ALS patient [11]. Total intravenous anesthesia without the use of muscle relaxants in ALS patients has also been occasionally reported. Xiao et al reported total intravenous anesthesia without muscle relaxant for a cesarean section in an ALS patient [12]. Lee et al reported total intravenous anesthesia without muscle relaxant for open reduction and internal fixation of the right tibia in an ALS patient [13].

Similarly, we also selected total intravenous anesthesia without muscle relaxant; however, in our case the surgical site was the lung (**Table 2**), which is responsible for ventilation necessary for breathing during the operation of the patient. We chose total intravenous anesthesia because of the required pauses in ventilation during the lung wedge resection. If an inhaled anesthetic was used, there was a possibility that the patient would regain consciousness during the time when the ventilation was stopped.

There were several reasons for not using muscle relaxants in this procedure. First, the patient did not want to use them. Second, ALS patients are considered vulnerable to muscle relaxants. Third, sugammadex reverses the rocuronium-induced blockade by encapsulating and inactivating neuromuscular blockers [14].

However, there was a report that neuromuscular recovery was delayed after using sugammadex [11].

In our case, ventilation was stopped three times during the surgery by appropriate sedation and hyperventilation. Propofol inhibits the respiratory drive and diaphragm effort to an extent that depends on the depth of sedation [15]. As more CO_2 is released during hyperventilation, intense forced breathing temporarily terminates the breathing due to the absence of a physiological CO_2 stimulus to respiration [16].

In summary, we successfully administered total intravenous anesthesia without the use of muscle relaxants for pulmonary wedge resection in an ALS patient. However, further research is needed to determine whether our anesthesia method can be generalized for surgery in ALS patients.

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Disclosure of conflict of interest

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