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Case Report

Use of remifentanil and propofol without muscle relaxant with Duchenne muscular dystrophy - A case report -

Heung Soo Kim, Seung Youp Baek, Dong Ho Park, and Keon Hee Ryu

Department of Anesthesiology and Pain Medicine, Eulji University Hospital, Daejeon, Korea

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Corresponding author
Keon Hee Ryu, M.D., Ph.D.
Department of Anesthesiology and Pain Medicine, Eulji University Hospital, 95 Dunsanseo-ro, Seo-gu, Daejeon 35233, Korea
Tel: 82-42-611-3883
Fax: 82-42-611-3882
E-mail: ryu4912@naver.com
ORCID
http://orcid.org/0000-0001-5781-6658

Duchenne muscular dystrophy (DMD) is a relatively rare muscle disease with severe symptoms. Owing to the commonly limited mouth opening, cervical spine immobility, and deformation, DMD patients often present with a difficult airway. Patients with DMD are sensitive to sedation, anesthesia, and neuromuscular blockade. This risk increases as the disease progresses with age. The anesthetic management of these patients can cause various issues, presenting a challenge to anesthesiologists. We administered anesthesia for an orchiectomy in a patient with testicular cancer using total intravenous anesthesia with propofol and remifentanil without muscle relaxants. Although the patient was Mallampati grade IV due to neck stiffness, tracheal intubation was successfully performed with a portable videolaryngoscope. The intraoperative course was uneventful and recovery was rapid without postoperative complications. In conclusion, anesthesia without a muscle relaxant was successful and the patient recovered rapidly, even with a difficult tracheal intubation.

Key Words: Airway management, Duchenne muscular dystrophy, Intravenous anesthesia, Neuromuscular blocking agents.

Duchenne muscular dystrophy (DMD) is a recessive inherited genetic disorder caused by a mutation in the X chromosome and occurs in approximately 1 in 3,500 males. Generally, degeneration and atrophy of the respiratory muscles and myocardium lead to death before the age of 30 years. During anesthesia, rhabdomyolysis, malignant hyperthermia, and cardiac arrest are known risks caused by an inhalation anesthetic or succinylcholine; therefore, the selection of inhalation anesthetics or muscle relaxants requires caution. According to a study by Muenster et al. [1], 4% of DMD patients had difficult intubations and the frequency was especially high in older patients [2]. Thus, difficulty in securing the airway can be anticipated and sufficient muscle relaxation is required. The risks are known for depolarizing muscle relaxants. For non-depolarizing muscle relaxants, it is generally known that sensitivity increases, but the response and stability have not yet been established [2,3]. Therefore, preoperatively securing the airway and recovering respiratory function postoperatively can be considered the most challenging aspects in anesthesia.

Thus, together with a review of the relevant literature, we report a case of general anesthesia management without muscle relaxation in a patient with DMD undergoing surgery for testicular cancer.

CASE REPORT

A 29-year-old male patient with a height of 160 cm and a weight of 40 kg was diagnosed with testicular cancer and admitted for a radical resection. He was diagnosed with DMD

through electromyography and biopsy at 7 years of age. As his respiratory muscles had been damaged by DMD over the past 5 years, he was able to maintain his breath for only 20 minutes without a support. Thus, the outpatient Department of Rehabilitation Medicine prescribed a supportive ventilation device from, and a portable ventilator (Trilogy®, Phillips, USA) was used in spontaneous/timed pressure support mode (respiratory rate, 12 breaths/min; 19 cmH_2O support during inhalation, inspiratory trigger sensitivity 5 L/min, inspiratory time 1.2 seconds) with a mask. Before the operation, the muscle strength in his legs became weaker than that of his upper body and activities of daily living were nearly impossible. There were no abnormal results from his preoperative blood tests or the additional ultrasound examination he underwent owing to the observation of intermittent premature ventricular contraction on electrocardiography. Cardiac function tests showed no abnormal findings, including a left ventricular ejection fraction of 62%. Premedication was not administered before anesthesia. Physical examinations conducted during the preoperative interview revealed a decrease in strength and joint contracture. The patient was classified as a Mallampati IV owing to contracture of the cervical vertebrae. Nevertheless, tracheal intubation was favored over using a supraglottic airway device since the patient had a jaw joint deformity [4] (Fig. 1).

The patient showed symptoms of general dehydration, such as anuria, since he had been administered only 500 ml of normal saline during 13 hours of preoperative fasting. Vital signs measured after entering the operating room included blood pressure of 110/60 mmHg, pulse rate of 102 beats/min, respiratory rate of 16 breaths/min, and body temperature of 36.2°C. Electrocardiography showed a normal sinus rhythm. Because of the risk of malignant hyperthermia, total intravenous anesthesia using a target-controlled infusion system was planned. After sufficient denitrogenation with 6 L/min of oxygen, anesthesia was induced using effect-site target concentrations of 4 μg/ml of propofol and 3 ng/ml of remifentanil. After manual ventilation with 6 L/min of oxygen and a partial pressure of inhaled oxygen of 100%, intubation was attempted via a laryngoscope using an inner diameter (ID) 7.0 mm single-lumen endotracheal tube (Mallinckrodt™ TaperGuard tracheal tube, Covidien, Ireland) without muscle relaxants. However, since the intubation was difficult, a ID 6.5 mm single-lumen endotracheal tube was used under the guidance of a portable video laryngoscope (GlideScope®, Verathon, USA). To reduce the risk of dyspnea due to laryngeal edema after intubation, 5 mg of dexamethasone was administered. Anesthesia was maintained at an FiO_2 of 0.5 with a mixture of oxygen and air, 3 L/min, and 3 μg/ml of propofol and 2.5 ng/ml of remifentanil. Bispectral index and intra-arterial blood pressure were monitored via the left radial artery. Mechanical ventilation was used during the operation in continuous mandatory ventilation mode with a tidal volume of 400 ml, peak inspiratory pressure of 20 mmHg, and respiratory rate of 12 breaths/min. After 10 minutes of anesthetic induction, the patient's systolic blood pressure decreased to 80 mmHg and his heart rate increased to 120 beats/min, indicating tachycardia. Hydration with Hartmann's solution returned him to a stable condition within 20 minutes of anesthetic induction. Throughout the operation, his vital signs were generally stable. The total operation time was 60 minutes, and no specific issues developed during the operation. A total of 600 ml of intraoperative fluids were administered: 400 ml of Hartmann's solution and 200 ml of 6% volume expanders (Volulyte®, Fresenius Kabi, Germany). Postoperatively, propofol and remifentanil administration was ceased and the patient was transferred to the intensive care unit for respiratory monitoring, received manual ventilation with a self-inflated resuscitator bag during transfer. The patient showed a spontaneous respiratory response within 15 minutes of the end of the infusion and recovered consciousness 25 minutes

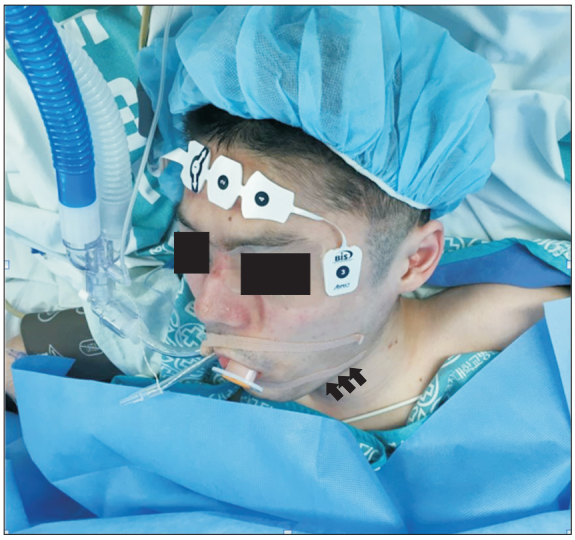

Fig. 1. The patient's jaw movement and rotation, were limited due to cervical fixation or contraction.

after the end of the infusion. However, because the patient was not breathing adequately, the ventilator was used in synchronized intermittent mandatory ventilation mode with a tidal volume of 400 ml and respiratory rate of 8 breaths/min.

Eight hours after the end of surgery, arterial blood gas analysis showed a pH of 7.41, PO_2 of 99 mmHg, and PCO_2 of 47 mmHg at an FiO_2 of 0.4. After confirming that spontaneous respiration had been recovered, the tube was removed. After extubation, the portable ventilator that the patient had used previously was used in spontaneous/timed pressure support mode. On postoperative day 2, the patient showed stable respiration and arterial blood gas analysis showed a pH of 7.33, PO_2 of 78 mmHg, and a PCO_2 of 46 mmHg in room air. The patient was transferred to the general ward, and subsequently recovered and was discharged without any abnormal findings.

DISCUSSION

DMD is the most common form of muscular dystrophy, resulting from a dystrophin gene mutation on the X chromosome. According to Boland et al. [5], DMD is diagnosed at an average age of 4.6 years. By 10 years of age, patients with DMD rely on wheelchairs. As the disease progresses, it affects the respiratory muscles and myocardium, leading to death from cardiopulmonary complications, with an average life expectancy of 17 years. As age increases, the incidence of difficult intubation becomes significantly higher and the morbidity rate of the extremities and myocardium also increase. Particularly, as respiratory muscle function worsens, the pulmonary reserve and vital capacity decrease, and anesthesia becomes more challenging [2].

In a study by Segura et al. [6], 90% of the patients who were over 30 years of age had cardiac involvement. However, even though the patient in the present report was 29 years old, no particular cardiac abnormalities were observed.

Since the first attempt at intubation was highly difficult, a video laryngoscope was used for intubation. In addition, even though there were no problems with cardiac function during the preoperative examination, difficulties were anticipated in postoperative respiratory care owing to the weakness of the respiratory muscles. According to a study by Muenster et al. [1], intubation is difficult in patients with DMD owing to their large tongue and limited mouth opening; therefore, it is not advisable to administer pretreatments such as sedatives in such cases. We did not use any pretreatment in the present case.

Our patient had limitations in the range of motion of his temporomandibular joint and cervical spine. According to Muenster et al. [1], 4% of DMD patients experience difficult intubation, while the rate increases to 7.5% among elderly patients. Furthermore, Ramachandran et al. [4] argued that the risk of supraglottic airway failure is increased 3.34-fold in patients with limited cervical spinal range of motion and 1.25-fold in patients with limited mouth opening compared with the normal group, although this was statistically insignificant. Therefore, we determined that intubation would be more appropriate.

Regarding the choice of methods for anesthesia, rhabdomyolysis and life-threatening hyperkalemia may occur due to inhalation anesthesia or succinylcholine, leading to an increased risk of anesthesia. Intravenous anesthesia using propofol, ketamine, or dexmedetomidine is known to be relatively safe [7,8]. In the present study, propofol and remifentanil were selected based on reports that described their safe use for anesthesia [1,6]. Anesthesia was administered using the target effect-site concentration method.

The patient was in a dehydrated state because he had received only 500 ml of normal saline during the 13 hours of fasting on the day before surgery. Thus, a large amount of fluid relative to the patient's weight was administered during the operation.

Although recovery in a patient with DMD is known to be limited due to an increased sensitivity to nondepolarizing muscle relaxants [2,8], there has not been enough research on why these changes in sensitivity occur. However, currently, it is thought that a decrease in the concentration of acetylcholinesterase in the endplate of the neuromuscular junction, caused by decreased secretions of choline acetyltransferase and acetylcholinesterase, prolongs the effects of muscle relaxants [8]. As a result, recovery is delayed and close observation of the respiratory function during recovery is required postoperatively. Moreover, it has been reported that reversal by anticholinesterase drugs in patients with advanced DMD may cause rhabdomyolysis [9].

The abnormal response of patients with DMD to muscle relaxants results from the structural changes in the neuromuscular junctions of these patients due to DMD [10]. The

prediction of abnormal responses in older patients with advanced disease is expected to be even more difficult [7]. Considering the reports that recovery time increased three to six times even when muscle relaxation was provided using atracurium or vecuronium, it was thought that, rather than using muscle relaxants, it would be better for the patient's recovery time if intubation were attempted only after maintaining a sufficient depth of anesthesia. As a result, in this case, anesthesia was performed safely without any specific incidents during the anesthesia and recovery process. This is consistent with previous reports from Capozzoli et al. [11] and Cossu and Caboni [12] on successful use of anesthesia without muscle relaxants.

In previous studies of anesthesia in patients with DMD [3,5], the average age of the patients was 9 to 14 years. The authors have not collected any cases in patients aged 30 years or older, and case reports of DMD patients aged 30 years or older are especially rare. Given the risk of muscle relaxant use in older patients due to the progression of respiratory complications, this case is significant because anesthesia was safely completed without muscle relaxants in a patient with advanced DMD, despite the difficult intubation.

However, since this was an operation in which muscle relaxation was not essential, there are limitations to applying this method to other operations that require muscle relaxation. Further research is needed to evaluate the results of not using muscle relaxants during thoracic or abdominal surgeries, which require muscle relaxation. For example, 2 mg/kg of sugammadex can provide a rapid and safe reversal of neuromuscular relaxation in DMD patients [13,14]. Thus, it is recommended to administer sugammadex to reverse the effects of a muscle relaxant after its use in thoracic and abdominal surgery when muscle relaxation is essential.

In conclusion, anesthesia without muscle relaxants using the target-controlled infusion total intravenous technique with propofol and remifentanil could be safely performed in a 29-year-old patient with DMD.

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