Case Report

Usefulness of sugammadex in a patient with Becker muscular dystrophy and dilated cardiomyopathy

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Abstract

A 54-year-old patient with Becker muscular dystrophy and dilated cardiomyopathy underwent laparoscopic cholecystectomy under total intravenous anesthesia. Muscle relaxation was induced by rocuronium (0.4 mg/kg body weight) under train-of-four (TOF) ratio monitoring. The TOF ratio was 0 at intubation, and 0.2 at the end of surgery. Residual muscle relaxant activity was successfully reversed by sugammadex (2 mg/kg body weight) without any hemodynamic adverse effects (TOF ratio 1.0 at extubation). The clinical and hemodynamic findings suggest that sugammadex can be safely used in patients with Becker muscular dystrophy and dilated cardiomyopathy.

1. Introduction

Becker muscular dystrophy is a form of myopathy caused by loss of dystrophin protein, characterized by weakness of proximal muscles, with the emergence of symptoms at 5–15 years of age, and is associated with a morbidity of 1/10,000. Patients usually develop abasia (inability to walk because of impaired muscle coordination) after their late 20s. The muscle dystrophy causes various intraoperative conditions such as aspiration pneumonia, prolonged artificial ventilation, malignant hyperthermia, rhabdomyolysis, hyperkalemia, cardiomyopathy, hypotension, and arrhythmia.

Patients with Becker muscular dystrophy are at higher risk for cardiomyopathy compared with those with Duchenne muscular dystrophy. Dilated cardiomyopathy is independent of age and severity of muscle weakness, and it is the most common cause of death in patients with Becker muscular dystrophy.

Here we describe a patient with Becker muscular dystrophy and dilated cardiomyopathy who underwent surgery under general anesthesia. Sugammadex was useful for the reversal of residual muscle relaxation induced by a nondepolarizing neuromuscular agent without any adverse cardiovascular effects. This is the first case report of using sugammadex with Becker muscular dystrophy in a patient with dilated cardiomyopathy.

2. Case Report

We received written permission from the patient to publish the report. A 54-year-old man (height 167 cm, weight 54 kg) was scheduled to undergo laparoscopic cholecystectomy for cholelithiasis. The patient received the diagnosis of Becker muscular dystrophy at the age of 31 years. He had difficulty walking because of muscle weakness. Dilated cardiomyopathy, with episodes of cardiac failure, had also been diagnosed in this patient.

Echocardiography (ECG) at admission showed left ventricular ejection fraction of 41%, all-round severe hypokinetic left ventricular wall motion, and moderate mitral regurgitation. The cardiac index was 2.7 L/min/m². The electrocardiogram demonstrated ventricular extrasystole with horizontal ST depression in leads II, III, and aVF. The spirogram was normal. Serum creatine kinase was normal. The American Society of Anesthesiologists Physical Status was 3.

General anesthesia was induced with fentanyl (3 μg/kg body weight) and midazolam (0.6 mg/kg body weight). Rocuronium was administered as a muscle relaxant. Neuromuscular blockade was monitored using train-of-four (TOF-Watch SX, Ireland Ltd, Dublin, Ireland) at 15-second intervals. We administered rocuronium 10 mg by 10 mg. The TOF ratio was 0 after administration of 20 mg...
Sugammadex in muscular dystrophy

3. Discussion

Becker muscular dystrophy is categorized as chloride channel myotonia. Anesthetic management of patients with Becker muscular dystrophy requires special attention to concomitant complications such as cardiomyopathy, hypotension, arrhythmia, and pulmonary hypertension, and also to the possible development of aspiration pneumonia, prolonged artificial ventilation, malignant hyperthermia, rhabdomyolysis, and hyperkalemia.

Becker muscular dystrophy is sensitive to depolarizing neuromuscular blocking agents and succinylcholine can produce prolonged contraction; thus, its use should be avoided. Although the risk of malignant hyperthermia is thought to be low in patients with Becker muscular dystrophy, succinylcholine increases the risk of rhabdomyolysis, which may cause cardiac arrest, probably mediated through hyperkalemia. Therefore, a non-depolarizing muscle relaxant should be used in such patients. However, the response to nondepolarizing muscle relaxants can be prolonged. It is often difficult to estimate the response and duration of nondepolarizing neuromuscular blocking agents in patients with Becker muscular dystrophy. Even though the ED95 of rocuronium is 0.3 mg/kg and we usually use rocuronium 0.6 mg/kg for patients with no complications, we administered rocuronium 10 mg by 10 mg under TOF ratio monitoring. Cisatracurium, a nondepolarizing muscle relaxant metabolized with no relation to renal or hepatic dysfunction, could be a good choice for such patients with muscular dystrophy, but it is not available in Japan.

As stated previously, patients with Becker muscular dystrophy are at risk of aspiration pneumonia because of low tidal volume, suppression of the swallowing reflex, laryngeal muscle weakness, and delayed excretion of gastric contents. Therefore, adequate and timely reversal of muscle relaxation is important. However, anticholinesterases, such as neostigmine, are difficult to use because they could cause either contraction or relaxation in patients with Becker muscular dystrophy; in other words, the reaction to anticholinesterases is unpredictable. Patients with myotonic dystrophy have been asked to avoid using anticholinesterase because of anticholinesterase-induced myotonia.

Sugammadex, a novel selective reversal binding agent, rapidly reverses muscle relaxation caused by steroidal neuromuscular blocking agents such as rocuronium. Sugammadex encapsulates steroidal neuromuscular blocking agents in the plasma and renders them unfavorable to neuromuscular junction receptors. Unlike other reversal agents, sugammadex has the ability to provide fast and thorough reversal of neuromuscular paralysis regardless of the level of blockade. Sugammadex is reported to have few adverse effects and no cardiovascular or autonomic effects commonly seen with conventional reversal using anticholinesterases plus anticholinergic agents. Recent studies reported the usefulness of sugammadex and rocuronium in patients with neuromuscular disease (including Becker muscular dystrophy). However, the role of sugammadex in surgical patients with neuromuscular disorders has not yet been fully established, and its role in muscular dystrophy patients who are concomitant with dilated cardiomyopathy is not known.

Based on conventional knowledge, the TOF ratio should be maintained at approximately 0.90 at the time of extubation in patients without neuromuscular disease. In our patient, the TOF ratio was 0.20 at the end of surgery, and accordingly, sugammadex was administered at 2 mg/kg body weight. Two minutes later, the TOF ratio increased to 1.0, and the patient was able to take a deep breath, grasp firmly, and open his eyes. Continuous TOF ratio monitoring of patients with muscular dystrophy after surgery is necessary. These patients are at risk of postoperative hypventilation because of prolonged action of neuromuscular blocking agents. However, in this patient, the use of sugammadex and rocuronium allowed safe extubation after surgery.

The use of volatile anesthetics in patients with muscular dystrophy also increases the risk of rhabdomyolysis, which could result in cardiac arrest and malignant hyperthermia. Accordingly, volatile anesthetic agents should be avoided and total intravenous anesthesia, with epidural anesthesia, is recommended.

Cardiac complications including cardiac arrest are also serious problems in the perioperative management of Becker muscular dystrophy. There are a few case reports using sugammadex for patients with Becker muscular dystrophy, but this report is the first to use sugammadex for patients with Becker muscular dystrophy and dilated cardiomyopathy. Myotonic disorders are sometimes associated with cardiomyopathy, which could enhance anesthesia-induced acute heart failure in patients with Duchenne muscular dystrophy. In patients with Becker muscular dystrophy, because motor dysfunction progresses more slowly than in those with Duchenne muscular dystrophy, prolonged workload on the morbid myocardium may lead to dilated cardiomyopathy. Cardiac involvement develops early in the right ventricle, with subsequent development of severe left ventricular dilatation with reduced ejection fraction, which could be complicated by life-threatening arrhythmias and the need for cardiac transplantation. Therefore, for the perioperative circulatory management of patients with dilated cardiomyopathy and Becker muscular dystrophy, the use of arrhythmogenic agents such as traditional reverse agents, combined with cholinesterase inhibitors and atropine sulfate, should be avoided. Sugammadex is less arrhythmogenic and induces the safe reversal of rocuronium-induced neuromuscular block in patients with heart failure and even with heart transplant recipients. This case also shows that sugammadex is useful even in a patient with dilated cardiomyopathy.

In this report, we described the successful reversal of rocuronium-induced muscle relaxation with sugammadex in a patient with Becker muscular dystrophy and dilated cardiomyopathy.
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References