

Sugammadex reversal of rocuronium-induced neuromuscular block in a patient with ataxia-telangiectasia

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A 17-year-old adolescent with ataxia-telangiectasia was scheduled to have laparoscopic colectomy for a resection of colon cancer. General anesthesia was performed, and rocuronium 30 mg was administered for muscle relaxation. Deep neuromuscular block (post-tetanic count: 0-8) was maintained for 95 minutes without additional rocuronium. On completion of surgery, sugammadex 80 mg was injected and train-of-four ratio was 0.93 at 210 seconds after administration. The tracheal tube was removed 5

min after the end of surgery. He recovered full spontaneous respiration and voluntary movements within 1 minute after extubation. The reversal of rocuronium induced neuromuscular block by sugammadex was fast, complete, and recovered to the initial preoperative level of neuromuscular function in this patient. (Rawal Med J 201;40:486-488).

Key Words: Ataxia-telangiectasia, rocuronium, sugammadex.

INTRODUCTION

Ataxia-telangiectasia is a progressive neurodegenerative disorder including cerebellar ataxia, immunodeficiency, and susceptibility to malignancies, recurrent sinopulmonary infections, and often respiratory failure.¹ We report the use of sugammadex for prevention of any respiratory complication in a patient with ataxia-telangiectasia after general anesthesia.

CASE PRESENTATION

A 17-year-old, 162 cm tall, and 40 kg weighing male was admitted for the management of suspicious colon cancer. He had ataxia-telangiectasia diagnosed at birth. He had dyspnea, generalized dystonia, dysmetria, ataxia, and telangiectasia on orbit. Colonoscopy showed a mass resulting in near obstruction at the transverse colon that was confirmed as adenocarcinoma. The preoperative laboratory findings were within the normal range. He was to have laparoscopic colectomy.

In operating room, electrocardiogram, pulse oximeter, capnogram, bispectral index, esophageal temperature, invasive measurement of arterial blood pressure, central venous pressure, and neuromuscular monitoring using post tetanic count (PTC) and train of four (TOF) once per minute were monitored. Neuromuscular monitoring was done

using a TOF-Watch SX device (Organon [Ireland] Ltd., Drynam Road, Swords, Co. Dublin, Ireland) and using the adductor pollicis muscle in response to the ulnar nerve. Propofol 80 mg, remifentanyl 0.2 µg/kg/min and desflurane 6.0 vol% by mask were administered for the induction of anesthesia. Rocuronium 30 mg was administered for muscle relaxation.

Following intubation, anesthesia was maintained with a 6-7% end-tidal concentration of desflurane in an oxygen-air mixture ($\text{FiO}_2=0.50$) and infusion of remifentanyl 0.1 µg/kg/min. Deep neuromuscular block (PTC: 0-8) was maintained for 95 minutes without additional rocuronium. After the skin closure, TOF count was 1. Subsequently, sugammadex 80 mg was given and TOF ratio was 0.93 at 210 seconds after administration. The endotracheal tube was removed 5 min after the end of surgery. He recovered to the preoperative level of spontaneous respiration and voluntary movements within 1 minute after extubation. The operation lasted for 70 min. Intended laparoscopic colectomy could not be completed because of multiple metastases, and the total period of anesthesia was 125 min. Estimated blood loss was about 100 ml and the total volume of infused crystalloid was 1400 ml. After the surgery, he was transferred to the intensive care unit and stayed for 1 day, and discharged 14

days after the surgery without any problem.

DISCUSSION

In this patient, rocuronium induced neuromuscular blockade was reversed within 5 minutes by sugammadex and no signs of residual curarization or recurarization occurred irrespective of the administration of the rocuronium. Ataxia-telangiectasia is a rare autosomal recessive genetic disorder due to a mutation in the ATM gene that encodes a serine/threonine protein kinase responsible for phosphorylating many target proteins.² This defect impinges on lymphocyte function in ataxia-telangiectasia³ commonly causing cellular immunodeficiency,⁴ as well as a predisposition to the development of lymphoid tumors at an early age.⁵

The neurological degeneration in ataxia-telangiectasia is progressive and typical ataxia-telangiectasia patients show neuromuscular features such as absent deep reflexes, atrophy, weakness and the presence of peripheral neuropathy.⁶ It can be accompanied by serious pulmonary problems such as interstitial lung disease. These are compounded from deficiencies in swallowing, aspiration,⁷ and respiratory muscle weakness. These respiratory problems are one of the major causes of increased mortality.⁸ In these patients, residual muscle relaxation or recurarization can lead to serious postoperative respiratory complications, as compared to other patients; and can furthermore aggravate underlying pulmonary disease.

There are reports on the use of sugammadex in the patients with neuromuscular disease or neuromuscular weakness. Hans et al.⁹ reported the clinical usefulness of sugammadex in patients with myasthenia gravis. Wefki et al.¹⁰ also found it useful in a patient with duchenne muscular dystrophy. However, there are a few studies that can confirm the safe use of sugammadex in patients with other minor neuromuscular disease. We found this effective and the safe in an adolescent patient with ataxia-telangiectasia, as described. The patients showed rapid emergence without any postoperative residual curarization.

In summary, anesthesiologists should pay attention

to the increased risk of the postoperative respiratory problem caused by ineffective cough, swallowing dysfunction, impaired airway clearance and immunodeficiency in ataxia-telangiectasia patients. The use of rocuronium should be considered in such patients because sugammadex can provide a rapid and safe reversal from rocuronium-induced neuromuscular block.

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Conflict of Interest: None declared

Rec. Date: June 15, 2015 Accept Date: July 21, 2015

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