Elmer Press

Case Report

J Med Cases. 2014;5(4):232-233

Using Sugammadex in a Patient With Friedreich Ataxia: A Case Report

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Abstract

Friedreich ataxia is a rare autosomal recessive disease characterized by demyelination of the nervous system, affecting both genders equally, starting in childhood with muscular coordination deficits that may also cause comorbidities such as cardiomyopathy and diabetes. The disease especially affects anesthesia management in patients planning to receive neuromuscular blockers. We aim to discuss the management of anesthesia technique in such a patient that underwent an appendectomy under general anesthesia.

Keywords: Friedreich ataxia; Anesthesia; Sugammadex

Introduction

Friedreich ataxia (FA) is a rare (1/50,000) upper and lower motor neuronal disease together with marked cerebellar atrophy with autosomal recessive inheritance that demonstrates degeneration in spinocerebellar and pyramidal pathways. It was first defined by German physician Nicholaus Friedreich in the 1860s. The disease is generally characterized by muscular coordination defects starting in childhood that progress over time. In addition, the disease causes comorbidities such as cardiomyopathy, diabetes and restrictive lung disease [1]. It has no effect on cognitive functions. The underlying pathology was demonstrated to be related to the mutation of a gene that codes for the mitochondrial protein frataxin. Decrease in frataxin production results in increased iron load, which causes mitochondrial disorder and cell death [1]. Neuromuscular dysfunction, in addition to cardiomyopathy and glucose intolerance seen in one-third of the patients, all affect the anesthesia to be applied in such patients [2].

Case Report

Appendectomy under general anesthesia was planned to be performed on a 17-year-old girl with a weight of 48 kg and a height of 160 cm. Upon physical examination of the patient, who was diagnosed as FA 7 years ago, body development and speech were assessed as normal with slight muscle weakness; she was noted to have an ataxic gait without help for short distances. The patient had a Mallampati score of 1 with no additional diseases, allergies or past history of any operations. ECG, chest X-ray, serum electrolytes, bleeding, liver and kidney function tests were normal. Her white blood cell count was 13,500/mm^3 and hemoglobin was 14.3 g/dL.

After 8 h of fasting, the patient was taken to the operating room after informed consent regarding her anesthesia was obtained. IV sedation with 1 mg midazolam was administered and a train-of-four (TOF) monitor for neuromuscular monitorization with 5 min intervals was placed on the ulnar nerve trace of her left hand. Induction was completed with 150 mg propofol and 20 µg of remifentanil was administered. Rocuronium bromide was added after airway was provided and TOF calibration was completed. TOF was observed from the moment of rocuronium administration until the time of recovery.

The patient was intubated with a 6.5 Fr endotracheal tube approximately 90 sec after the administration of neuromuscular blockade. Sevoflurane 2.5%, oxygen 50% and nitrogen protoxide 50% were used for maintenance anesthesia. Cefazolin 2 g for infection prophylaxis and tramadol 50 mg for preemptive analgesia were also administered. The patient needed no additional muscle relaxants during the operation. Her end-tidal carbon dioxide was maintained between 35 and 40 mmHg. Her hemodynamics were stable and no complications were encountered during the operation. Her operation was terminated after 1 h and 15 min. Spontaneous respirations returned after the first hour. Sugammadex (2

Manuscript accepted for publication March 3, 2014

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doi: http://dx.doi.org/10.14740/jmc1722w

mg/kg) was administered after the operation was completed. The patient was extubated uneventfully after she was able to draw sufficient tidal volumes. The patient did not desaturate on room air and was sent to the pediatric surgical ward 10 min after sugammadex administration.

Discussion

There is still no consensus on the type of general anesthesia that should be used in patients with FA. Although no complications after general anesthesia in the great majority of such patients are reported in the literature, which is comprised mainly of case reports, comprehensive studies on the management of general anesthesia in patients with FA are still needed. The main discussion for these patients is on the use of muscle relaxants [1]. There are a limited number of studies with controversial results on the use of non-depolarizing muscle relaxants in patients with FA. Due to the possibility of development of hyperkalemia after succinylcholine administration in denervated muscle diseases, it has been suggested to refrain from using depolarizing muscle relaxants [1]. Kume et al reported a hypersensitivity to tubocurarine [3]. On the other hand, there have been case reports on depolarizing muscle relaxant use with normal or near-normal results. Neuromuscular monitoring was performed in all those cases in which tubocurarine, atracurium, vecuronium and rocuronium were used with normal or near-normal results [4, 5]. Schmitt et al, on the other hand, used total intravenous anesthesia with propofol and sufentanil instead of volatile anesthetics for maintenance of anesthesia and reported no delay in recovery with rocuronium. After they used recuronium bromide uneventfully in the anesthesia of two patients diagnosed with FA, they reported that non-depolarizing muscle relaxants might be used safely in patients with FA [6]. Although no complications were reported in the majority of those studies, it is not possible to guarantee safe use of various drugs (such as muscle relaxants), since none of them have been studied with high power and randomization. Pancora et al reported that due to the hypersensitivity of FA patients to non-depolarizing muscle relaxants, muscle relaxants might potentially cause delays in recovery and discharge, and hence they facilitated the intubation of the patients with the use of propofol and sufentanil without the use of muscle relaxants. They used optimal drug infusion (remifentanil and propofol) during maintenance anesthesia by providing a bispectral index value of 45-60 [6].

Taking into account the insecurity of using muscle relaxants in the literature, the hypersensitivity of patients with FA to non-depolarizing muscle relaxants and delays in the postoperative recovery and discharge in patients with FA, we tried to demonstrate the safe use of non-depolarizing muscle relaxants in such muscle diseases by using non-depolarizing muscle relaxants for anesthesia induction, followed by reversal with sugammadex accompanied by necessary TOF monitoring in such a case.

Grant Support

No funding.

Conflict of Interest

None.

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