



CASE REPORT

Medicine Science 2017;6(1):109-10

**Anesthetic management in a patient with Friedreich's Ataxia:
A different approach**

Sedat Akbas, Erol Toy, Ahmet Selim Ozkan

Department of Anesthesiology and Reanimation, Inonu University, Faculty of Medicine, Malatya, Turkey

Received 27 May 2016; Accepted 22 June 2016

Available online 29.06.2016 with doi: 10.5455/medscience.2016.05.8497

Abstract

Friedreich's Ataxia (FA) is a rare autosomal recessive disease. FA is clinically characterized by progressive ataxia, especially loss of reflex and power in the lower extremities, spasticity and loss of proprioceptive sensory. There is an increased sensitivity to non-depolarizing muscle relaxants in neurodegenerative diseases. Care must be taken of anesthesia management for the use of volatile anesthetics and muscle relaxants, and precautions should be taken for difficult airway management which is due to secondary to kyphoscoliosis and comorbidities. In this case, anesthesia management of emergency operation for septal hematoma by TIVA without using of muscle relaxants and minimized hemodynamic response depending on the intubation by videolaryngoscope were presented.

Keywords: Friedreich's Ataxia, anesthesia management, neuromuscular blockade, videolaryngoscope

Introduction

Friedreich's Ataxia (FA) is a rare autosomal recessive disease and the most common cause of inherited ataxias by the prevalence of 1/30000-1/50000 [1]. FA comprises the features of neuropathological as the degeneration of dorsal columns of the spinal cord, spinocerebellar and pyramidal tracts [2]. FA is clinically characterized by progressive ataxia, especially loss of reflex and power in the lower extremities, spasticity and loss of proprioceptive sensory. Furthermore, it shows togetherness with diabetes mellitus, cardiomyopathy, pes cavus, kyphoscoliosis, restrictive lung disease and congestive heart failure [3]. The most common cardiac anomaly in FA is hypertrophic cardiomyopathy in proportion as 63% [2]. It leads to early death which cause arrhythmias at the advanced stages and the average life expectancy of up to 30-40 years [2]. There is an increased sensitivity to non-depolarizing muscle relaxants as in neurodegenerative diseases. predisposition to hyperkalemia may occur after succinylcholine administration. In this patient group, care must be taken of anesthesia management for the use of volatile anesthetics and muscle relaxants, and precautions should be taken for difficult airway management which is due to secondary to kyphoscoliosis and comorbidities [4]. In this case, anesthesia management of emergency operation for septal hematoma were presented.

Case Report

Twenty years old, 55 kg, 165 cm, male patient was admitted to the emergency room after a fall-related injury. An emergency decompression surgery was planned because of septal hematoma considered to develop due to the trauma. In the patient's history, it was learned that diagnosed for FA 6 years and diyabetis mellitus type II 14 years ago. On the other hand, patient has become wheelchair-dependent in years because of muscle weakness and scoliosis. In the preoperative evaluation; dysarthric speech, complete loss of muscle strength in the lower extremities (5/5), partial muscle strength in the upper extremities (2/5) and sensory loss were present. Mallampati score was evaluated as 2. There was no additional pathology in cardiological assessment. Biochemical tests were normal. The process was explained to patient and his relatives and written consent form was obtained. The patient was taken to the operating room without premedication. Electrocardiogram (ECG), oxygen saturation (SpO₂), noninvasive blood pressure (NIBP) and bispectral index (BIS) monitoring were performed. ECG was sinus rhythm, NIBP: 120/84 mmHg, heart rate: 96 min⁻¹, SpO₂ 97% and respiratory rate: 14 min⁻¹. Following the preoxygenation with face mask by 4 L min⁻¹ for 5 min, anesthesia induction was performed with propofol 2 mg kg⁻¹ IV, remifentanyl 1.5 mcg kg⁻¹ IV without using muscle relaxants. Patient was intubated by videolaryngoscope (McGRATH® MAC) after sufficient depth of anesthesia. Anesthesia was maintained by total intravenous anesthesia (TIVA) as infusions of propofol 100-200 mcg kg⁻¹ min⁻¹ IV ve remifentanyl 0,1 mcg kg⁻¹ min⁻¹ IV and BIS value was adjusted to 40-60. Mechanical ventilation was maintained by 50% oxygen-air mixture without the use of volatile anesthetics.

*Corresponding Author: Sedat Akbas, Department of Anesthesiology and Reanimation, Inonu University, Faculty of Medicine, Malatya, Turkey.
E-mail: drsedatkb@ gmail.com
Fax: +904223410728
Tel: +904223410660-3106

Aneesthesia was terminated and patient was extubated after started breathing spontaneously and returned protective reflexes. Patient with Modified Aldrete score 9 at recovery room was transferred to the relevant service without the complication.

Discussion

Detailed examinations of neurological, cardiac, pulmonary and skeletal systems are very important in the preoperative evaluation of patients with FA. Difficult airway management, neuromuscular monitoring because of increased sensitivity to muscle relaxants, neglecting use of volatile agents, following hemodynamic parameters carefully in anesthesia induction and maintenance are all fundamental points of anesthesia management [5].

Schmitt et al. [4] reported that anesthesia was maintained to patients with FA by propofol and sufentanil as TIVA instead of volatile anesthetics, there was no delay by rocuronium at recovery room and could be used non-depolarizing muscle relaxants. For the patients with denervated muscle disorder, non-depolarizing muscle relaxants are preferred when required the use of muscle relaxants because of using of succinylcholine lead to hyperkalemia. It has been observed that volatile anesthetics were used in many cases. There are very few studies that demonstrating the use of non-depolarizing muscle relaxants in patients with FA [4]. Doğu et al. [5] reported that patient with FA intubated by rocuronium with neuromuscular monitoring, aneesthesia was terminated without the complication by using TIVA for maintenance.

There are many case reports for patients with FA performed anesthesia induction and maintenance without using the neuromuscular blocker. Levent et al. [6] reported that they used for patients with FA propofol and alfentanil for the induction of anesthesia, intubated without the neuromuscular blocker, maintained the anesthesia by TIVA without the neuromuscular blocker and recovery was faster and no complication. Akçaboy et

al. [3] reported that they performed to patient with FA laryngeal mask airway by propofol and alfentanil for the induction of anesthesia without the neuromuscular blocker, maintained the anesthesia by sevoflurane and nitrous oxide-oxygen mixture and terminated surgery without no complication. In this case that planned emergency operation for septal hematoma, videolaryngoscope was performed by using propofol and remifentanyl without neuromuscular blocker to minimize the hemodynamic responses due to intubation and the operation was completed successfully.

Conclusion

TIVA should be preferred for maintenance of anesthesia and avoiding muscle relaxants as much as possible, although there are many cases showing that used safely non-depolarizing muscle relaxants and inhalation anesthetics in patients with FA. hemodynamic stabilization must be provided carefully for the induction of anesthesia, intubation and maintenance for hypertrophic cardiomyopathy with a high incidence in this group of patients.

References

1. Kurul SH, Yiş U, Güzel Aİ, Kasap H, Başak N, Dirik E. Evaluation Of The Cases With Friedreich Ataxia. *Gulhane Med J.* 2013;55(2):123-7
2. Elinx-Benizri S, Glik A, Merkel D. et al. Clinical Experience With Deferiprone Treatment for Friedreich Ataxia. *J Child Neurol.* 2016;31(8):1036-40.
3. Akçaboy ZN, Akçaboy EY, Özek ÖC, Yılmaz AN, Göğüş N. Friedreich ataksisi olan bir hastada anestezi yaklaşım. *Türk Anest Rean Der Dergisi.* 2011;39(1):50-3.
4. Schmitt HJ, Wick S, Münster T. Rocuronium for muscle relaxation in two children with Friedreich's ataxia. *Br J Anaesth.* 2004;92(4):592-6.
5. Doğu T, Şahin H, Şimşek T, Altınışık U, Toman H, Altınışık H.B, Erbaş, M. Friedreich ataksili hastada anestezi yönetimi. *Abant Med J.* 2015;4(3):266-8.
6. Levent K, Yavuz G, Kamil T. Anaesthesia for Friedreich's ataxia. Case report. *Minerva Anesthesiol.* 2000;66(9):657-60.