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CASE REPORT

Anesthesia Management in a Patient with Patau Syndrome Who Underwent Bilateral Vitrectomy

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ABSTRACT

Patau syndrome was the first described by Patau et al. and associated with trisomy 13. The syndrome is characterized by microphthalmia, polydactyly, and cleft lip-palate. In this case report, we discuss the anesthesiology approach and management in a 2-year-old girl with Patau syndrome undergoing bilateral vitrectomy surgery.

A 2-year-old girl weighing 12.5 kg was undergoing bilateral vitrectomy surgery. There was no additional pathology related to the cardiovascular, central nervous, and respiratory systems. Following anesthesia induction, the patient was intubated using a # 4.5 uncuffed endotracheal tube. At the end of the operation, 3 mg/kg sugammadex was given to the patient and she was transferred to the remission unit without any problems. During her follow-up in the unit, nurses called for help after her oxygen saturation level dropped to 90%. Mask ventilation support was supplied with a chin lift maneuver. As re-curarization was thought to be associated with the situation, an additional sugammadex dose was given to the patient. After 20 minutes of follow up, she was transferred to the pediatric ward in a steady state.

The doses of anesthetic and analgesic drugs should be titrated carefully because intraoperative opioid and neuromuscular blocker use can lead to postoperative respiratory problems. Therefore, close follow-up of patients is essential in the postoperative period, as in the intraoperative period.

Keywords: Anesthesia management, Patau syndrome, re-curarization, sugammadex

INTRODUCTION

Patau syndrome was first described by Patau et al. and associated with trisomy 13 [1]. The syndrome is characterized by microphthalmia, polydactyly, and cleft lip-palate. In addition to a limited lifespan, cardiac, vascular, and central nervous system malformations give importance to the syndrome [2]. It is the third most commonly seen autosomal trisomy after trisomy 21 and 18, and among them, it is the most severe [3]. The death rate in a study on 172 patients with trisomy 13 was found as 28% in a week, 44% in a month, and 86% in a year [4]. In this case report, we discuss the anesthesiology approach and management in a 2-year-old girl with Patau syndrome undergoing bilateral vitrectomy surgery.

CASE PRESENTATION

The characteristic features of Patau syndrome such as low nasal bridge, low-set ears, sloping forehead, prominent glabella, sunken eyes, microphthalmia in the right eye, alopecia, and high arched palate were present in a 2-year-old girl weighing 12.5

kg who underwent bilateral vitrectomy surgery. In the preoperative evaluation, there was no additional problem or pathology related with the cardiovascular, central nervous, and respiratory systems, and she was evaluated as American Society of Anesthesiologists (ASA) Class II. After premedication with midazolam, the patient was taken into the operating room and monitored with standard ASA monitorization. A video laryngoscope and fiberoptic bronchoscope were prepared because of the risk for difficult intubation. Following anesthesia induction with 3 mg/kg propofol, 1 mcg/kg fentanyl, and 0.6 mg/kg rocuronium, the patient was intubated with a No. 4.5 uncuffed endotracheal tube using a Miller No. 1 blade without any difficulties. For maintenance of anesthesia, total intravenous anesthesia (TIVA) was preferred with propofol and remifentanil infusions in dosages of 10 mg/kg/hour and 0.05 mcg/kg/min, respectively. The propofol dosage was reduced to 6 mg/kg/hour over time. During the procedure, which took approximately 2 hours, the vital signs of the patient remained stable. At the end of the operation, 3 mg/kg sugammadex was administered to the patient for reversal of rocuronium. After the patient reached full awareness and was breathing spontaneously, she was successfully extubated and transferred to the remission unit without any problems. During her follow-up in the remission unit, nurses called for help after her oxygen saturation level dropped to 90%. The patient's respiration was superficial and her body muscles appeared flaccid. Balloon-mask ventilation support was supplied with the chin lift maneuver. As re-curarization was thought to be associated with the situation, an additional sugammadex dose was administered to the patient. After 20 minutes, the oxygen saturation of the patient was maintained over 97%, then the patient was transferred to the pediatric ward in a steady state.

DISCUSSION

Cardiac-craniofacial malformations and growth retardation are significant features in the anesthesia management of trisomy 13 [2]. There is a risk of difficult intubation and vascular access, postoperative apnea, and hemodynamic problems [5]. Short neck, micrognathia, and small mouth are features of trisomy 13, which make endotracheal intubation difficult [6]. In a case of a patient with trisomy 13 in the emergency department, after 2 unsuccessful intubation attempts with direct laryngoscopy, although ear-nose-throat and anesthesia doctors performed fiberoptic bronchoscopy and used a cricothyroid retrograde wire, the intubation process was unsuccessful [7]. Our patient had a low nasal bridge, low-set ears, sloping forehead, prominent glabella, sunken eyes, microphthalmia in the right eye, alopecia, and high arched palate, but no micrognathia, small mouth, and short neck. However, video laryngoscopy and fiberoptic bronchoscopy were prepared because of the risk for a difficult intubation scenario, but we faced no difficulties or complications during the endotracheal intubation.

Sugammadex is a modified γ-cyclodextrin molecule prepared for a reversal of the myorelaxant effect of rocuronium. It has a more rapid onset than the neostigmine+atropine combination and it irreversibly encapsulates rocuronium molecules. Recurarization after sugammadex reversal of rocuronium is theoretically unexpected but there are some examples of re-curarization following the reversal of rocuronium with sugammadex in the literature [8-11]. Kotake et al. reported that the use of sugammadex did not eliminate the risk of residual paralysis when the decision of extubation was not based on objective neuromuscular monitoring [12]. Recurrence of neuromuscular blockade after sugammadex administration can be explained by two processes: the first is a redistribution of rocuronium from peripheral compartments to the central compartment, and the second is the insufficient, unbound sugammadex molecules in the plasma [13]. We did not use neuromuscular monitorization but the symptoms of the patient and the results after additional sugammadex administration make us think that re-curarization had occurred.

In conclusion, the doses of the anesthetic and analgesic drugs should be titrated carefully because intraoperative opioid and neuromuscular blocker use can lead to postoperative respiratory problems. Although re-curarization after reversal of rocuronium with sugammadex is seen very rarely, it is not impossible. Therefore, it should be kept in mind that close follow-up of patients is essential in the postoperative period, as in the intraoperative period.

CONFLICT of INTEREST STATEMENT

The author has no conflicts of interest to declare.

REFERENCES

[1] Patau K, Smith DW, Therman E, et al. Multiple congenital anomaly caused by an extra autosome. Lancet 1960; 1: 790-3.

[2] Duarte AC, Menezes AIC, Devens ES, et al. Patau syndrome with long survival. A case report. Genetics and Molecular Research 2004; 3(2): 288-92.

[3] Fogu G, Maserati E, Cambosu F, et al. Patau syndrome with long survival in a case of unusual mosaic trisomy 13. Eur J Med Genet. 2008; 51(4): 303-14.

[4] Magenis RE, Hecht F, Milham S Jr. Trisomy 13 (D1) syndrome: Studies on parental age, sex ratio, and survival. J Pediatr. 1968; 73(2): 222-8.

[5] Demir HF, Erakgün A, Çertuğ A. Patau sendromu ön tanılı pediyatrik olguda anestezik yönetim: Olgu sunumu ve literatüre kısa bir bakış. Ege Tıp Dergisi / Ege Journal of Medicine 2013; 52(4): 205-207.

[6] Baum VC, O'Flaherty JE. Anesthesia for genetic, metabolic and dysmorphic syndromes of childhood. Lippincott Williams and Wilkins 2007; 374.

[7] Roberts R, Verghese ST, Bauman N. Failure to intubate a child with Patau (Trisomy 13) Syndrome with fiberoptic via the LMA and retrograde wire insertion through the cricothyroid with the ENT surgeon in the ER. CR39 2010 SPA/AAP Pediatric Anesthesiology. Winter Meeting; April 15-18, 2010.

[8] Bellod Jr A, March X, Hernandez C. Et al. Delayed recurarisation after sugammadex reversal. Eur J Anaesthesiol 2014; 31: 708–721

[9] Corre FL, Nejmeddine S, Fatahine C, et al. Recurarization after sugammadex reversal in an obese patient. Can J Anesth 2011; 58: 944–947

[10] Carollo DS, White WM. Postoperative Recurarization in a Pediatric Patient After Sugammadex Reversal of Rocuronium-Induced Neuromuscular Blockade: A Case Report. A&A Practice. 2019 Apr 11 (Publish Ahead of Print).

[11] Murata T, Kubodera T, Ohbayashi M, et al. Recurarization after sugammadex following a prolonged rocuronium infusion for induced hypothermia. Can J Anesth 2013; 60: 508–509.

[12] Kotake Y, Ochiai R, Suzuki T, et al. Reversal with sugammadex in the absence of monitoring did not preclude residual neuromuscular block. Anesth Analg. 2013; 117: 345–51.

[13] Iwasaki H, Renew JR, Kunisawa T, et al. Preparing for the unexpected: special considerations and complications after sugammadex administration. BMC Anesthesiology 2017; 17: 140.