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Case Report / Olgu Sunumu

Anaesthesia Management and Use of Sugammadex in a Patient with Ullrich's Disease

Ullrich Hastalığı Olan Bir Hastada Anestezi İdamesi ve Sugammadeks Kullanımı

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Abstract / Öz

Here, we report anaesthesia management and the successful use of total intravenous anaesthesia and sugammadex in a patient with Ullrich's disease. Propofol and remifentanyl infusion was used for anaesthesia. After the end of the surgery, when the train-of-four value was 0%, 4 mg kg^{-1} sugammadex was administered, and the patient was successfully extubated after 36 s. No adverse effects or safety concerns were observed. In conclusion, we suggest that the use of propofol infusion to avoid the use of inhalation anaesthetics and the use of sugammadex for the reversal of the effects of rocuronium is safe in patients with Ullrich's disease.

Keywords: Sugammadex, muscular dystrophy, total intravenous anaesthesia

Bu olgu sunumuzda Ullrich hastalığı olan bir hastada başarılı total intravenöz anestezi ve sugammadeks kullanımını bildirmeyi amaçladık. Anestezi idamesi için propofol ve remifentanil infüzyonu kullanıldı. Cerrahi bitiminde train of four (TOF) değerleri %0 iken, 4 mg kg^{-1} sugammadeks verildi ve 36. saniyede hasta başarıyla ekstübe edildi. Herhangi bir yan etki ile karşılaşılmadı. Sonuç olarak, Ullrich hastalığı olan hastalarda inhalasyon anestetiklerinin kullanılmasından kaçınmak adına propofol infüzyonun kullanımının ve roküronyumun etkilerinin sonlandırılması için sugammadeks kullanımının güvenli olduğunu bildirmekteyiz.

Anahtar kelimeler: Sugammadeks, musküler distrofi, total intravenöz anestezi

Introduction

Scleroatonic muscular dystrophy also known as Ullrich's Disease is an autosomal recessive congenital muscular dystrophy that is characterized with generalized muscle weakness, multiple contractures in the axial joints and hyper-laxity in the distal joints with increased creatine phosphokinase levels (1). A defect in the collagen IV that is an extracellular matrix protein is held responsible (2). In muscular dystrophies, such as Ullrich's disease, perioperative respiratory and cardiac complications are common, and they have a higher risk for malignant hyperthermia and rhabdomyolysis (3). Hypersensitivity to non-depolarizing neuromuscular blockers can cause a prolonged requirement for ventilator support (4).

Sugammadex is a new agent used in reversing the effects of steroidal non-depolarizing neuromuscular agents such as vecuronium and rocuronium (5). By encapsulating the neuromuscular blocker, it decreases the free concentration of the drugs from the plasma causing a rapid recovery from the effects of the neuromuscular blocker. There are limited number of reports regarding the use of sugammadex in patients with muscular dystrophies, whereas there is no report regarding its use in Ullrich's disease.

In this case report, we report anaesthesia management and the successful use of total intravenous anaesthesia (TIVA) and sugammadex in a patient with Ullrich's disease.

Case Presentation

A 7-year-old male patient (weight 20 kg, height 120 cm) who had planned to undergo ureteroneocystostomy for bilateral vesicoureteral reflux had a history of Ullrich's disease that was diagnosed in the early 2012. He was not on medication. Clinical evaluation revealed hypotonia, proximal contractures and distal hyperlaxity in the upper extremities (Figure 1). He was unable to sit without help, to climb up stairs and hobbled while walking. His mother defined histories of aspiration during feeding.

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Preanaesthesia evaluation of blood samples revealed no abnormalities except the elevated creatine phosphokinase levels (480 UL^{-1}). His Mallampati score was II, and the movement of the neck and head was not limited. The physical evaluation revealed reversed v-shaped upper lips and concave temporal muscles resulting in an expressionless face that is a characteristic trait in patients with Ullrich's disease (Figure 2). He demonstrated normal central nervous system function and normal intelligence as expected.

After obtaining informed consent from the parents, the patient was taken to the operating room without any premedication, and intravenous line was inserted. Non-invasive arterial blood pressure, peripheral oxygen saturation (SpO_2), electrocardiography and nasopharyngeal temperature were monitored. Dantrolene was available if required. Anaesthesia was induced with 5 mg kg^{-1} propofol and 0.1 μg $kg^{-1}min^{-1}$ remifentanyl and 0.6 mg kg^{-1} rocuronium was administered. The neuromuscular function was monitored using train of four (TOF) stimulation of the ulnar nerve with TOF Watch® SX (Organon Ireland Ltd, Dublin, Ireland). TOF was applied with 20-s intervals to the ulnar nerve after the induction. When TOF ratio reached 0%, endotracheal intubation with a ID 5.5 tube was achieved in the first attempt. His Cormack–Lehane score was 2, and intubation was not difficult (6). For the management of anaesthesia, 200 μg $kg^{-1}min^{-1}$ propofol infusion for the first 10 min, 100 μg $kg^{-1}min^{-1}$ propofol infusion between 10^{th} and 120^{th} min and 80 μg $kg^{-1}min^{-1}$ propofol infusion from 120^{th} min to the end of the operation and 0.05–0.2 μg $kg^{-1}min^{-1}$ remifentanyl infusion was administered. The patient was ventilated (Datex Avence S5) in a pressure-controlled mode with 40% oxygen–air mixture to reach tidal volumes of 150–180 mL and maintain $EtCO_2$ levels between 32–36 mmHg with a respiratoy rate of 16–18 min^{-1}. Propofol and remifentanyl infusions were stopped 5 m before the end of the surgery. The first twitch of TOF recovered to 10% of control levels, 52 min after the induction of 0.06 mg kg^{-1} rocuronium was administered to obtain complete neuromuscular blockade, as observed by absent TOF responses. The duration of the surgery was 230 min, and three additional dose of rocuronium were required. At the end of the surgery, TOF ratio was still 0%; therefore, for the reversal of the effect of 4 mg kg^{-1} rocuronium, sugammadex was administered. When TOF ratio was 90% and the spontaneous ventilation was adequate, the patient was extubated. The time from the administration of sugammadex to extubation was 36 s. The patient was taken to the post-anaesthetic care unit. No sign of rhabdomyolysis was observed in the postoperative period, and creatine phosphokinase levels increased to 502 UL^{-1} from 480 UL^{-1} after the surgery.

Discussion

Scleroatonic muscular dystrophy also known as Ullrich's Disease carries the risk of postoperative respiratory failure because of repeated aspiration histories, prolonged block and residual curarization due to the use of non-depolarizing neu-

Figure 1. Proximal contractures in the upper extremities

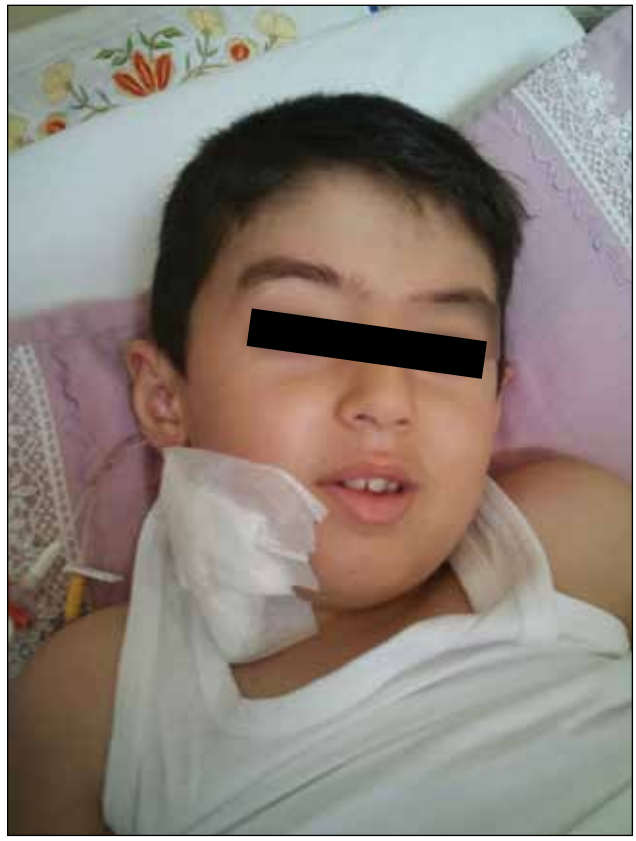

Figure 2. Reversed v-shaped upper lips, concave temporal muscles

romuscular blockers (7). In addition, many agents used in the induction and management of anaesthesia can cause malign hyperthermia in the patients with muscular dystrophies (8).

To avoid the agents that can trigger malignant hyperthermia, we planned on using total intravenous anaesthesia. Anaesthesia was induced with intravenous bolus propofol and remifentanyl infusion. Propofol is an agent safely used in patients with malignant hyperthermia risk, and it is believed that it does not trigger the disease (8) Anaesthesia management with TIVA and rocuronium was adequate for surgery.

Puangsuvan et al. (9) reported the successful anaesthesia management of a patient with Ullrich's disease who underwent appendectomy. They had intubated the patient only with propofol and remifentanyl infusion and avoided using neuromuscular blocker and inhalation anaesthetics.

Grosu et al. (6) reported using sevoflurane in a patient with Ullrich myopathy and concluded that halogenated agents can be safely used in patients with Ullrich's disease because the mutations of genes associated with malignant hyperthermia are not present in the disease. However, Li et al. (10) demonstrated that malignant hyperthermia is observed six times more in patients with musculoskeletal disorders on the basis of a large paediatric inpatient sample. We had chosen not to use sevoflurane and preferred TIVA as a safer method.

The patient was susceptible to postoperative respiratory failure because of the prolonged effects of neuromuscular blockers in patients with muscular dystrophy in addition to the preoperative hypotonia observed (4). The reversal of the effects of neuromuscular blockers is achieved with anti-cholinesterase agents; however, they carry the risk of increasing hypotonia in hypotonic patients. Furthermore, they may not be adequate in the reversal of the drug and carry the risk of residual block both in adults and children (11, 12). The requirement for the use of anticholinergic drugs with anti-cholinesterase agents also has its own side effects (12).

Sugammadex is a fast reversal agent of the steroidal neuromuscular blockers, such as rocuronium and vecuronium. Because sugammadex is different from anti-cholinesterase agents that are used for the reversal of most neuromuscular blocks, it can be used even when the patient is under profound blockage, and postoperative residual neuromuscular blockade can be significantly decreased (13).

Ustun et al. (4) reported the safe use of sugammadex in a patient with Becker muscular dystrophy. They reversed the effect of rocuronium with sugammadex and observed a fast and safe return of neuromuscular function.

De Boer et al. (14) used sugammadex in a paediatric patient with Duchenne muscular dystrophy with a dose of 4 mg kg^{-1} and suggested that the effects of high dose rocuronium can be fast and safely reversed with sugammadex in patients with Duchenne muscular dystrophy.

In our literature search, although there are reports on other muscular dystrophies, we were not able to find any patient with Ullrich's disease who was administered sugammadex for the reversal of the effects of rocuronium. Furthermore, we chose to use rocuronium as the neuromuscular blocker for intubation and during surgery. It is important to use neuromuscular function monitoring during the surgery and particularly during the reversal of the neuromuscular block to find the appropriate dose of sugammadex to reach a full recovery of the neuromuscular function (13). We used 4 mg kg^{-1} sugammadex in our patient at the end of the surgery according to a dose-ranging study (15). TOF value was 0% before the administration of sugammadex. When TOF values reached 90% with 4 mg kg^{-1} sugammadex and spontaneous ventilation was adequate, the patient was extubated. The time from the administration of sugammadex to extubation was 36 s. The patient's peripheral oxygen saturation remained 97% without any oxygen support. Haemodynamics was stable during the operation. We did not encounter any period of hypotension or hypertension. No sign of residual neuromuscular blockade or postoperative recurarization was immediately observed after extubation or in the post-anaesthetic care unit. The patient was observed in the post-anaesthetic care unit for 24 h and was then taken to the paediatric surgery ward the next day. He was released from the hospital on the 6th day following the operation without any complications.

Conclusion

We suggest that propofol infusion to avoid the use of inhalation anaesthetics and the use of sugammadex for the reversal of the effects of rocuronium are safe in patients with Ullrich's disease.

Informed Consent: Written informed consent was obtained from patients' parents who participated in this case.

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