



High Dose Sugammadex Administration in a Case of Huntington Chorea

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Abstract

Huntington Chorea, a central nervous system disease that is inherited autosomal dominantly which causes miscellaneous difficulties and has features by the aspect of anesthesia management. It was aimed to present the first administration of such high dose sugammadex, in a Huntington Chorea case. Additionally, our case report is the second one in literature.

Keywords: General anesthesia, sugammadex, Huntington Chorea

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Introduction

Huntington Chorea, a rare central nervous system disease that is inherited autosomal dominantly. Symptoms include personality disorder, choreic movements and progressive cognitive functional failure [1]. The worst worrisome symptom is dysphagia which is caused by dysfunction of pharyngeal muscles. Dysfunction of pharyngeal muscles also causes tracheal aspiration. Death commonly occurs due to complications of tracheal aspiration [2]. There are limited numbers of case reports on management of anesthesia in Huntington Chorea patients [3-5]. Sensitivity to effects of midazolam, prolongation in action of sodium tiopental and increased risk for pulmonary aspiration were reported in these patients [4,5]. Although there are case reports about administration of miscellaneous anesthetics and potential beneficial effects of these agents, report on these patients with sugammadex is limited only one case report [4-6].

In this case report it was aimed to present administration of both repeated and high dose sugammadex and naloxane HCL, on the case who was planned for emergency bronchoscopy because of tracheal foreign body aspiration.

Case Report

Forty-eight years old male patient (body weight 60 kg, height 180 cm) was planned for emergency bronchoscopy due to tracheal foreign body aspiration. The patient who has choreic movements was cachectic. He was conscious but probably due to pharyngeal muscle dysfunction his speech was blurred. It was reported that he has had Huntington Chorea for 18 years and had no additional disease and his father has died because of same disease by his relatives. He has been treated by interested clinics since his disease has been diagnosed, but he withdrawn his medication for 3 months by himself and he has dysphagia for 15 days. Infiltrative areas were seen in pulmonary radiograph in the lower zone of right lung. There was no significant laboratory evidence except that aspartate aminotransferase: 147 U/L and alanine transaminase: 130 U/L. Patient was monitored with pulse oximeter, electrocardiograph, non-invasive blood pressure and additional neuromuscular monitoring by TOF-Watch® -S (Organon, Dublin, Ireland). Propofol 2 mg/kg and remifentanyl 1 mcg/kg administered for anesthesia induction. Before neuromuscular blocker agent has administered, TOF-Watch® -S was calibrated and then rocuronium was administered 0.5 mg/kg. TOF was measured and

bronchoscopy was started after measurement was % 0 of first. Maintenance of anesthesia was provided by infusion of propofol (0,1 mg/kg/min) and remifentanyl (0,1 mcg/kg/min), and infusions were stopped at the end of bronchoscopy. Bronchoscopy procedure took 20 minutes. TOF measurement was % 84 and after administration of 200 mg sugammadex there was not enough recovery from rocuronium. Because of this reason 200 mg additional sugammadex was administered and TOF measurement was above % 95. However respiration effort and respiration capacity of patient were enough, he was not conscious and his pupils were miotic after 30 minutes passed remifentanyl infusion stopped, so 0,08 mg naloxane was administered. After 10 minutes he was conscious and he was transferred to postoperative care unit. After enough time passed in postoperative care unit he was transferred to Department of Thoracic Surgery without any complication.

Discussion

Although there were case reports and case series about anesthetic management of Huntington Chorea patients, there is only one case report was detected in the literature about the administration of sugammadex in those patients [3-7]. Sugammadex is a gamma- cyclodextrin structured agent that was used to reverse steroidal neuromuscular blocker agents such as rocuronium and vecuronium. Sugammadex forms complexes by binding steroidal neuromuscular blocker agents both in the systemic circulation and in neuromuscular junction and then these complexes are excreted to urine without any metabolism [8]. Sugammadex was preferred in this case because sugammadex reverses neuromuscular block rapidly and neostigmine was avoided because of its choreiform movement increasing side effect [9,10]. Although enough dosage was used, TOF measurement was not sufficient enough so additional dosage was used. As recommended in many publication, total intravenous anesthesia administered by infusion of remifentanyl and propofol [11,12]. Although it was emphasized that propofol may be used safely in these publications, in a case report, increase in choreiform movements during magnetic resonance imaging with propofol sedation was reported [13].

In literature, management of anesthesia in cases with Huntington Chorea included bispectral index monitoring but in our case it was not feasible because the operation was performed in emergency state.

Further more no publication was detected about the prolonged opioid action on patients with Huntington Chorea. The half life of remifentanyl is 8-10 minutes and it is characterised without no cumulation with repeated doses or long periods of infusions [14]. Although this features, naloxane was administered because the patient was not conscious and his pupils were miotic, even though 30 minutes passed after infusion withdrawal.

As a result, although as pointed in many publications that total intravenous anesthesia with propofol and remifentanyl infusion is safe, reversal agents such as sugammadex and naloxane may be needed because the risk of prolongation of remifentanyl action and neuromuscular blocker agents.

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