

CASE REPORT / OLGU SUNUMU

THE USE OF SUGAMMADEX IN A PEDIATRIC PATIENT WITH TAY SACHS SYNDROME

TAY SACHS SENDROMLU ÇOCUK HASTADA SUGAMMADEKS KULLANIMI

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ABSTRACT

Tay Sachs is an infrequently seen neurodegenerative, lysosomal sphingolipid storage disease. The disease has infantile, juvenile and adult forms. In the juvenile form, the patient may be normal until the age of 2 years, motor skills are lost around the age of 4 years. Sugammadex (Bridion; MSD, Oss, The Netherlands), is a cyclodextrin ring agent used in recent years for the recovery of neuromuscular block provided by rocuronium and vecuronium and to accelerate recovery. In this case report we aimed to present the use of sugammadex in the anesthesia approach to a patient diagnosed with Tay Sachs undergoing rigid bronchoscopy.

KEYWORDS: Tay Sachs, Sugammadex, Pediatric

ÖZ

Tay Sachs nadir görülen bir nörodejeneratif, lizozomal sfingolipid depolama hastalığıdır. Hastalığın infantil, juvenil ve erişkin formları vardır. Juvenil formunda hasta 2 yaşına kadar normal olabilirken, motor becerileri 4 yaş civarında kaybolur. Sugammadex (Bridion; MSD, Oss, Hollanda), son yıllarda rokuronyum ve vekuronyum tarafından sağlanan nöromüsküler bloğun geri dönmesini sağlamak ve hızlandırmak için kullanılan siklodekstrin halkalı bir ajandır. Bu olguda rijid bronkoskopi yapılan Tay Sachs sendromlu hastanın anestezi yönetiminde sugammadex kullanımını sunmayı amaçladık.

ANAHTAR KELİMELE: Tay Sachs, Sugammadex, Çocuk

INTRODUCTION

Tay Sachs is an infrequently seen neurodegenerative, lysosomal sphingolipid storage disease, which develops as a result of hexosaminidase A enzyme deficiency. Abnormal accumulation of GM2 gangliosides in the brain tissue leads to neuronal cell loss and degeneration in the white matter.

Tay Sachs disease is seen in the three forms of infantile, juvenile and adult. The infantile form starts at 3-6 months and the patient is lost towards the age of 2 years. In the juvenile form, the patient may be normal from birth then lose speech and other motor skills at approximately 2 years, dysphagia, ataxia and spasticity develop and from around the age of 4 years, life continues in a bedridden, vegetative state and mortality occurs between the ages of 5-15 years (1).

Sugammadex (Bridion; MSD, Oss, The Netherlands) is a cyclodextrin ring agent used in recent years for the recovery of neuromuscular block provided by rocuronium and vecuronium and to accelerate recovery. A complex is formed by binding to steroid structure muscle relaxants in the circulation and neuromuscular junctions and it is eliminated via the renal route without metabolism (2).

The case is here presented of the use of Sugammadex in the anesthesia approach to a patient diagnosed with Tay Sachs undergoing rigid bronchoscopy.

CASE

A male child aged 4 years 5 months, weighing 12 kg was referred to the Pediatric Surgical Clinic for rigid bronchoscopy because of persistent pneumonia and possible thick mucous or suspected foreign body.

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In the preoperative evaluation the general condition of the patient was poor, he was tachypneic, and respiratory sounds were uneven. The lung volume had decreased and the chest cage was observed to have narrowed, there was kyphoscoliosis in the right lateral position and difficult breathing in the supine position.

After obtaining written, informed consent from the parents, the patient was admitted to the operating room. Monitorization was applied of non-invasive blood pressure (BP), ECG, heart rate and peripheral oxygen saturation (SpO₂). The oxygen saturation values on admission to the operating room were 84%, BP:80/50 mm hg⁻¹ and pulse:122 min⁻¹. With the patient in a lateral position, a vascular route was opened with a 22 gauge cannula under inhalation of 4% sevoflurane and 50/50% oxygen/air mixture. For monitorization of neuromuscular conduction, a TOF Watch®-S (Organon, Dublin, Ireland) device was used. For ulnar stimulation, 2 surface electrodes were placed over the course of the ulnar nerve in the right wrist. The hand was fixed leaving the thumb free. The acceleration transducer was placed on the wide surface of the thumb to evaluate the adduction response of the thumb with ulnar nerve stimulation.

Anesthesia induction was applied with 2 mg kg⁻¹ propofol and 1µg kg⁻¹ remifentanyl intravenously (iv). After checking the eyelash reflex, the patient was ventilated. The TOF Watch®-S device was calibrated. The supramaximal stimulation was defined automatically by the neuromuscular monitorisation system and 0.6 mg kg⁻¹ iv rocuronium was administered. The supramaximal stimulation was applied with the 'train-of-four' (TOF) stimulation. After obtaining a TOF 1 value at approximately 50 secs, rigid bronchoscopy was then applied by the pediatric surgeon. No foreign body was observed and thick consistency mucopurulent secretions were cleared by aspiration. Throughout the bronchoscopy procedure, remifentanyl infusion was continued at the rate of 0.5 µg kg⁻¹ min⁻¹. The bronchoscopy procedure lasted 15 mins. Remifentanyl was stopped and ventilation of the patient was continued with a mask. Sugammadex at a dose of 2 mg kg⁻¹ was administered to the patient who had 50% oxygen with 92% saturation. After 210 secs when TOF was > 90%, the patient was woken.

The patient was followed up at room air without oxygen and when saturation was 89%, was transferred to the pediatric intensive care unit (ICU) with 4 lt min⁻¹ oxygen.

DISCUSSION

No case could be found in literature related to the use of Sugammadex and general anesthesia in a patient

with Tay Sachs syndrome. The only related case was of the anesthesia method used for anal sphincter surgery on a 14-month old infant under spinal anesthesia with ketamine and nitrogen sedation (3).

Most patients with Type A worst prognosis are lost before the age of 4 years. Various difficulties can be experienced in the anesthesia method used in patients with Tay Sachs. There could be restrictive problems in the lungs and problems such as acid or hepatosplenomegaly which push the diaphragm upwards could cause difficulty in ventilation (1). Preparations must be made for difficult intubation as there is insufficient information in literature related the anesthesia method for these patients.

In a report by Bujok et al. the anesthesia method used in a case of Niemann Pick syndrome, which is a lysosomal sphingolipid storage disease similar to Tay Sachs, it was stated that the posture of the patient caused difficulties in ventilation and intubation (4). In the current case, there was kyphoscoliosis and although difficulties in ventilation and intubation were feared because of the impaired posture, no difficulties were encountered. No problems were experienced during the rigid bronchoscopy procedure.

In interventions with anticipated difficult airway and neuromuscular block, for antagonising rocuronium, sugammadex can be safely used without concern of a postoperative residual effect (5). In the current case, using rocuronium under TOF guidance ensured a smaller amount used and through the use together with Sugammadex on wakening, a rapid and safe recovery was provided.

There are few cases in literature related to the use of Sugammadex with rigid bronchoscopy where ventilation difficulty and bronchospasm have developed (6). In the current case, after loss of consciousness with remifentanyl, the conscious state was monitored and low-dose Sugammadex was used. No bronchospasm, ventilation difficulty or other complications were observed.

In Tay Sachs patients who develop hepatomegaly, care is necessary in respect of anesthetic agents used which could cause liver injury because of acid. Propofol and remifentanyl are suitable for use because of the short-term effects and that they do not cause postoperative respiratory failure or prolong sedation and their metabolism is independent of the liver and kidneys (7). In the current case, short-term effect agents such as propofol and remifentanyl were used.

As withdrawal from the mechanical ventilator could have been difficult, the patient was woken with Sugammadex and at spontaneous respiration of 4 lt min⁻¹

oxygen at saturation <89% without intubation, was transferred to the Pediatric ICU.

Tay Sachs is seen to have its own specific technical difficulties in the anesthesia approach and there may be pulmonary problems in end-stage patients which create problems in extubation just as in intubation.

In conclusion, an end-stage Tay Sachs patient with respiratory muscle weakness with a serious pulmonary infection was successfully awakened with the use of Sugammadex without intubation after rigid bronchoscopy.

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