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CASE REPORT

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Successful use of spinal anesthesia for incarcerated inguinal hernia repair in a patient with Hunter syndrome

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Abstract
Mucopolysaccharidosis type II (MPS II, Hunter syndrome) is a rare X-linked lysosomal storage disorder caused by a deficiency of the enzyme iduronate-2-sulphatase, leading to the accumulation of glycosaminoglycan (GAG). GAG accumulation causes difficult airway due to changes in the upper airway. In this report we present spinal anesthesia that was administered to a patient with Hunter syndrome to avoid complications related to anticipated difficult airway.

Keywords: Spinal anesthesia, Hunter syndrome inguinal hernia

Introduction

MPS II is a rare X-linked disease resulted from the deficiency of IDS lysosomal enzyme that causes GAG accumulation in various tissues and organs [1,2]. Excessive accumulation of these mucopolysaccharides in the skeletal system and in the connective tissues causes facial dysmorphism, hepatomegaly, splenomegaly, skeletal and articular deformities, valvular heart disease, and hearing problems [1]. Obstructive airway disease is the main clinical symptom of Hunter syndrome [3]. Recurrent upper airway infections are frequently reported, and these infections cause nasal congestion and thick nasal secretions [3].

Case Report

An 18-year-old male patient, weighing 37 kg and a height of 128 cm, diagnosed with MPS II was scheduled for emergent incarcerated inguinal hernia repair. In the preanesthetic evaluation, the patient had typical features of Hunter syndrome with macrocephaly, a wide forehead and rough facial lines, shortness of the body height, neck and extremities, thick brows and lips, wide tonsils, and history of snoring. The patient had Mallampati score of III, a 5 cm thyromental distance, and 10 cm sternomental distance (Figure 1). The patient was being followed for Grade 1 aortic and mitral regurgitation and was receiving enalapril therapy. The patient's family was informed about the anticipated difficult airway and anesthesia risks, and the expected respiratory problems in the postoperative period.

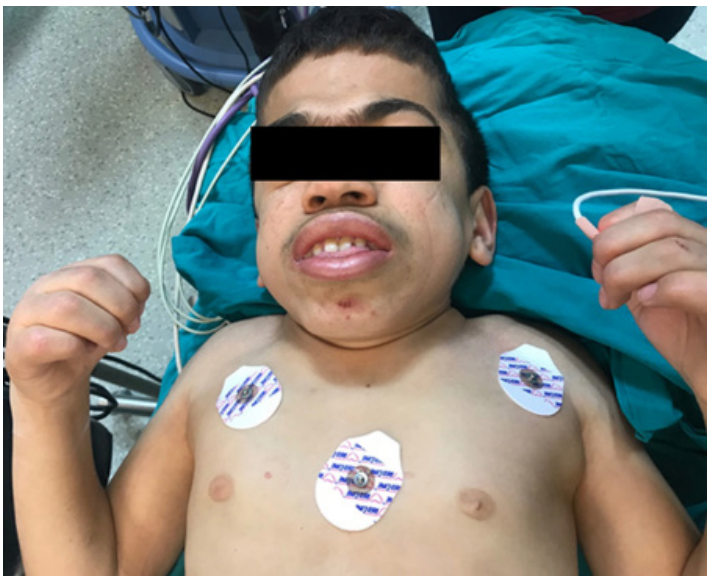

Figure 1. 18 year old male patient with Hunter syndrome

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The patient was allowed the anesthesiologist to submit his own case report and photograph without any identification. Considering the anticipated difficult airway and the safety of regional anesthesia, subarachnoid block under midazolam sedation was planned for this patient. Difficult airway equipment were made available including different size airways, smaller size endotracheal tubes, various size LMAs, C_MAC video laryngoscope and fiberoptic bronchoscopy. Two experienced anesthetists for to manage the anticipated difficult airway with fiberoptic bronchoscope were present in the operating room. After the routine anesthesia monitoring (ECG, pulse oximeter and non-invasive arterial blood pressure), 1 mg midazolam IV bolus was administered. The subarachnoid block was performed while the patient was in the sitting position. Following local infiltration, 10 mg fentanyl and 7.5 mg heavy bupivacaine were injected in L3 / L4 intervertebral space with 25G Quincke needle. The level of anesthesia was controlled after the subarachnoid block. The patient received supplemental oxygen through facemask during surgery. The surgery lasted 90 minutes, and an additional 1 mg midazolam was administered intraoperatively. Hemodynamics of the patient were stable during the operation. During the postanesthesia care unit for 2 hours, no respiratory or cardiac complication was observed. He was referred to the ward and discharged to home on the third day.

Discussion

The main concern of Hunter syndrome is the difficulty in ensuring the safety of the airway in anesthesia management. Adenotonsillar hypertrophy, a large tongue, thickened vocal cords, and narrowed tracheobronchial tract cause challenges related to management of the airway [4]. In a study investigating anesthetic techniques and airway complications in patients with Hunter syndrome, the rate of difficult mask ventilation, difficult intubation, and failed intubation were reported. In that study, difficult mask ventilation was observed in 26.7%, and difficult intubation in 35% of the patients [5]. In a case planned for umbilical hernia surgery Ozer et al. [6] reported that the patient could not intubated after induction of general anesthesia, and although emergency tracheotomy was performed the patient died from postoperative respiratory and cardiac complications. Since airway anatomy change by increasing age, we made difficult airway materials ready as we predicted the difficult airway in our patient. Preference of spinal block should be decided depending on status of the patient. When choosing a subarachnoid block for obtaining anesthesia, the duration of the operation, the patient's communication and the willingness and experience of the surgeon should be considered. Due to the accumulation of mucopolysaccharide with toxic effects on neurons, regional techniques may fail [7]. The safety and success of the subarachnoid block and the airway related problems in this patient led us to administer anesthesia with this block [8]. In this case, difficult airway materials were kept ready and two experienced anesthesiologists were present for patient safety with alternative plans for the management of anesthesia.

Conclusion

We conclude that spinal anesthesia may be considered as a safe and predictable option for abdominal surgery in children with Hunter syndrome in terms of anticipated difficult airway and respiratory tract related complications. However, further randomized studies with wide populations are needed to obtain a definitive conclusion.

Competing interests
The author confirms that this article content has no conflict of interest.

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