CASE REPORTS

Opioid-free general anesthesia and induced recovery from anesthesia in a patient with myotonic dystrophy type-1: a case report

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Abstract Myotonic dystrophy type-1 (Steinert disease) is an autosomal dominant, progressive multisystem disease in which myotonic crisis can be triggered by several factors including pain, emotional stress, hypothermia, shivering, and mechanical or electrical stimulation. In this report, dexmedetomidine-based general anesthesia, in combination with a thoracic epidural for laparoscopic cholecystectomy in a patient with Steinert disease, is presented. An Aintree intubation catheter with the guidance of a fiberoptic bronchoscope was used for intubation to avoid laryngoscopy. Prolonged anesthetic effects of propofol were reversed, and recovery from anesthesia was accelerated using an intravenous infusion of theophylline.

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Introduction

Myotonic Dystrophy (MD) type-1, or Steinert disease, is an autosomal dominant, progressive multisystem disease characterized by consistent contracture of muscle following stimulation. A myotonic crisis can be triggered by several factors, including pain, emotional stress, hypothermia, shivering, and mechanical or electrical stimulation. Most affected patients become severely disabled in the fifth or sixth decades due to muscle weakness and recurrent pulmonary infections. These patients are reported to be more susceptible to the depressant effects of general anesthetic agents.1

In this report, opioid-free anesthesia management in a disabled patient with Steinert disease is presented. Written consent was obtained from the patient for the publication of this case report.

Case description

A 48-year-old female patient (160 cm, 85 kg) who was diagnosed as having MD type-1 four years ago, was scheduled for elective laparoscopic cholecystectomy surgery. The family history included four siblings who had previously been diagnosed as having MD. Additionally, the obstetric history was gravida 3, para 3, none of which survived. The patient was wheelchair dependent and unable to walk unassisted. She had a moderately severe disability, and she was unable to attend her own bodily needs without assistance. Moreover, the patient had breathing difficulty while lying flat. A preoperative cardiology examination revealed decreased left ventricular function with an ejection fraction of 40%, without any conduction disorders. The pulmonary function test showed a restrictive pattern with a lowered vital capacity (68% of the predicted value). Preoperative airway assessment revealed a Mallampati score III and short neck. Her other preoperative physiologic examinations and laboratory analyses were normal.

No premedication was administered preoperatively. The patient was quite anxious, and she refused to stay awake during the operation; thus, dexmedetomidine-based opioid-free general anesthesia was reserved for the patient. In the operating room, routine monitoring of noninvasive blood pressure, electrocardiogram, oxygen saturation, Bispectral Index (BIS), and acceleromyograph (by using Train-Of-Four [TOF] ratio at the adductor pollicis brevis) were established. A warming blanket and warm intravenous fluids were used to prevent hypothermia. With the patient in the sitting position, an epidural catheter was inserted from the T7/8 interspace. Following a test dose, a local anesthetic (15 mL of 0.375% bupivacaine) was injected from the epidural catheter until the sensory level of T4 until pinprick was reached.

After the epidural anesthesia, general anesthesia induction was performed with dexmedetomidine in a loading dose of 0.6 μg.kg⁻¹ over 10 minutes, followed by an injection of propofol 60 mg and rocuronium 30 mg. There was no difficulty in mask ventilation, and the patient was intubated using a fiberoptic bronchoscope in order to avoid laryngoscopy. An Aintree intubation catheter was passed through I-gel laryngeal mask under fiberoptic bronchoscopy guidance. Following the removal of the laryngeal mask, an endotracheal tube (ø 7) was inserted through the Aintree intubation catheter. An arterial line was inserted in the left radial artery for intra- and postoperative monitoring immediately after securing the airway, to avoid further painful stimuli while the patient was awake. General anesthesia was maintained with a 1% sevoflurane, air, and oxygen mixture, and continuous infusion of dexmedetomidine 0.4 mg.kg⁻¹.hour⁻¹ (BIS: 45 ± 5) (Table 1). The surgery was ended after 60 minutes uneventfully. After the detection of a TOF ratio of 0% with the neuromuscular monitor, sugammadex 2 mg.kg⁻¹ was administered as a reversal agent. However, the patient was still unconscious (BIS: 40), and there was no spontaneous breathing in the following 25 minutes, although her TOF ratio was 100%. Following 200 mg theophylline administration over 10 minutes, the patient gained consciousness (BIS: 90) and was extubated (Table 1). She was then transferred to the intensive care unit for close follow-up. Postoperative pain management included non-steroidal anti-inflammatory drugs and local anesthetic boluses (10 mL of 0.125% bupivacaine) through the epidural catheter. On the 3rd postoperative day, the patient was discharged from the hospital.
Additionally, remifentanil is a sensitive opioid for the induction of anesthesia. Although previous reports have demonstrated its effectiveness for the induction of anesthesia, it is often recommended to be used in low doses (less than 1 mg/kg) due to its short duration of action and the risk of respiratory depression.

In the present patient, although the muscle relaxant was reversed successfully with sugammadex, ToF values remained at low levels (BIS: 40) 25 minutes following the end of the anesthesia. One of the possible reasons that can explain the lack of spontaneous breathing despite the sufficient TOF values may be our false conclusions about the exact level of neuromuscular block. It was shown that accelerometry seems to underestimate neuromuscular blockade in DM1 patients, especially at submaximal levels of neuromuscular block. Furthermore, the electrical TOF stimulus could induce myotonia and be interpreted as an indication that neuromuscular blockade has been fully reversed. However, this does not explain why BIS values remained at low levels, even 25 minutes after anesthesia in the present patient. MD patients commonly demonstrate some degree of hyperactivity, excessive daytime somnolence, or cognitive impairment. BIS values, observed in the awake state of these patients, can be significantly lower compared to those of normal controls. Therefore, the BIS values in some of the MD patients can indicate an incorrect hypoxic and should, therefore, be interpreted with caution. BIS, entropy, or NeuroSENSE monitoring could be useful to evaluate the hypoxic component of general anesthesia provided care is taken to measure a baseline control value in the awake patient before inducing anesthesia.

In our patient, the initial BIS value was measured as 97 before applying any sedative or hypnotic drugs (Table 1).

Methylxanthine derivatives, including aminophylline and theophylline, work by stimulating the central nervous system, respiratory drive, and respiratory muscles. Aminophylline has been used previously to enhance recovery from anesthesia in MD1 patients. Additionally, eight newborn babies with congenital myotonic dystrophy were reported to be weaned off the mechanical ventilator successfully with aminophylline. It was demonstrated in a previous animal study that theophylline accelerates recovery from general anesthesia via elevation of intracellular cAMP levels and blockade of adenosine A2 receptors. In the present case, the patient regained consciousness immediately after administration of theophylline, and this was clearly monitored with BIS. Based on the previous reports of methylxanthine derivatives, it may be helpful to use aminophylline or theophylline in MD patients to accelerate the recovery process.
recovery from anesthesia; additionally, respiratory stimulant effects of these drugs may be advantageous in patients with decreased vital capacity. However, one should be aware of their arrhythmogenic potential.

Conclusions

In summary, a successful dexmedetomidine-based opioid-free general anesthesia in combination with thoracic epidural anesthesia was conducted for laparoscopic cholecystectomy in a patient with advanced MD type-1 (Steinert Disease). If endotracheal intubation is essential, an Ain-tree intubation catheter inserted under the guidance of a fiberoptic bronchoscope can be used to avoid laryngoscopy. Although propofol has been used uneventfully in some patients with MD, it can prolong recovery from anesthesia, as in the present case. Theophylline was used successfully for the acceleration of recovery from anesthesia. Given the variable clinical presentations, a standard anesthesia protocol for MD cannot be recommended.

Conflicts of interest

The authors declare no have conflicts of interest.

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