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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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PALAVRAS-CHAVE
Período de recuperação da anestesia;
Despertar prolongado da anestesia;
Dexmedetomidina;
Cateter para troca de tubo traqueal;
Distrofia miotônica;
Teofilina

Anestesia geral sem opioide e recuperação induzida da anestesia em paciente com distrofia miotônica tipo-1: relato de caso

Resumo: A Distrofia Miotônica (DM) tipo-1 (Doença de Steinert) é uma doença multissistêmica progressiva autossômica dominante em que a crise miotônica pode ser desencadeada por vários fatores incluindo dor, estresse emocional, hipotermia, tremores e estímulo mecânico ou elétrico. O presente relato descreve anestesia geral realizada com dexmedetomidina em combinação com peridural torácica para colecistectomia laparoscópica em paciente com Doença de Steinert. Para evitar laringoscopia, a intubação traqueal foi realizada utilizando cateter de intubação Aintree guiado por broncofibroscopia óptica. Os efeitos anestésicos prolongados do propofol foram revertidos e a recuperação anestésica foi acelerada pelo uso de infusão intravenosa de teofilina.

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 $\mu g.kg^{-1}$ 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 (nº 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 stimulus 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.

Table 1 Perioperative vital parameters of the patient.

	Heart rate (bpm)	Blood pressure (mmHg)	SpO_2 (%)	$EtCO_2$	BIS	TOF (%)	
	Preoperative	109	155/102	93	–	97	–
Anesthesia induction	Induction	100	136/90	100	–	40	0
	Intubation	112	146/93	100	–	45	0
	Incision	93	132/87	98	46	50	0
	Trocar insertion	99	138/88	95	40	42	0
Anesthesia maintenance	10 min	83	105/74	96	36	45	0
	20 min	78	110/78	95	35	50	0
	30 min	80	113/81	95	36	48	0
	40 min	81	108/80	94	38	51	0
	50 min	76	106/72	94	37	43	0
End of anesthesia	0 min; sugammadex	74	111/81	95	37	40	84
	10 min	79	109/85	94	36	38	105
	20 min	65	121/92	93	37	40	100
	30 min; theophylline	83	115/78	97	38	41	100
	40 min; extubation	88	132/80	93	–	93	104

BIS, Bispecteral Index; TOF, Train-Of-Four stimulation.

Discussion

MD type-1 (Steinert Disease) is associated with more frequent perioperative complications and more severe multisystem involvement compared with MD type-2.[1,2] Additionally, anesthetic management of Steinert disease remains controversial because the response of patients to anesthetic drugs is so variable. Thus, the use of reduced anesthetic doses is recommended. Although propofol has been used uneventfully in some cases, it was reported to lead to a prolonged anesthetic effect even with low doses as low as 1 $mg.kg^{-1}$.[3–5] Although patients with MD are considered to be sensitive to opioids and susceptible to opioid-related adverse effects, fentanyl and remifentanil have been used without any anesthesia-related complications.[4,6]

However, the possibility of hyperalgesia related to remifentanil infusion should not be overlooked. Sugammadex has been successfully used for the reversal of rocuronium in a few cases.[7,8] Because of the response of MD patients to muscle relaxants is unpredictable, neuromuscular block monitoring is mandatory. The use of regional anesthesia in patients with MD is recommended, if possible.

Dexmedetomidine is an α-receptor agonist that can be used as a non-opioid adjuvant in general and locoregional anesthesia, due to its sedative, analgesic, and sympatholytic effects.[9] In this report, dexmedetomidine infusion, in combination with a thoracic epidural, was used for intraoperative analgesia. Reduced doses of propofol (less than 1 $mg.kg^{-1}$) and rocuronium (less than 0.5 $mg.kg^{-1}$), followed by dexmedetomidine infusion and sevoflurane, enabled sufficient conditions for intubation and surgery. In this case, to avoid laryngoscopy, the patient was intubated through an Aintree intubation catheter with the aid of a fiberoptic bronchoscope.

In the present patient, although the muscle relaxant was reversed successfully with sugammadex (TOF 100%), and end-tidal sevoflurane concentration was zero, BIS values remained at low values (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 acceleromyography seems to underestimate neuromuscular blockade in DM1 patients, especially at submaximal levels of neuromuscular block.[10] Furthermore, the electrical TOF stimulus could induce myotonia and be misinterpreted as an indication that neuromuscular blockade has been fully reversed.[11] 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 hypersomnia, 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.[12] Therefore, the BIS values in some of the MD patients can indicate an incorrect hypnotic state and should, therefore, be interpreted with caution. BIS, entropy, or NeuroSENSE monitoring could be useful to evaluate the hypnotic component of general anesthesia provided care is taken to measure a baseline control value in the awake patient before inducing anesthesia.[1] 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 MD patients.[13] Additionally, eight newborn babies with congenital myotonic dystrophy were reported to be weaned off the mechanical ventilator successfully with aminophylline.[14] 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.[15] 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

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 Aintree 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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