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Target-controlled infusion of propofol for a patient with myotonic dystrophy

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Abstract
We present a patient with myotonic dystrophy (MD) who was anesthetized with propofol using a target-controlled technique for electrophysiologic examination and cardiac catheter ablation. The patient became apneic unexpectedly at the same time when he fell asleep, with effect-site propofol concentration of 1.6 μg ml^{-1}. We had to insert a laryngeal mask airway (LMA), and mechanical ventilation was performed. The patient opened his eyes on verbal command at an effect-site concentration of 1.2 μg ml^{-1} after the procedure. This concentration (1.2 μg ml^{-1}) was slightly lower than our institutional average for adult male patients (1.5 ± 0.2 μg ml^{-1}). However, the time from the end of anesthesia to the patient's awakening was about 10 min. We considered that emergence from anesthesia was not delayed in this case. Careful titration of propofol by target-controlled infusion (TCI) enabled to evaluate the patient's sensitivity to propofol. We conclude that TCI of propofol was a useful anesthetic technique in the MD patient. Respiratory depression might occur in MD patients at low propofol concentrations. Precise control and titration over target propofol concentration is important in anesthetic management for MD patients.

Key words Myotonic dystrophy · Target-controlled infusion · Propofol

Introduction

Myotonic dystrophy (MD) is an autosomal dominant disorder and the most common of the myotonic syndromes [1]. MD is characterized by myotonia, marked wasting of the muscles of neck, pharynx, and distal limbs, frontal baldness, and multiple system involvement. Extramuscular features include cataracts, cardiomyopathy, conduction abnormalities, sleep apnea syndrome, and dysphagia.

Anesthetic management of these patients may pose a serious problem to the anesthesiologist. Sensitivity to anesthetics and neuromuscular blocking agents may result in perioperative cardiovascular and respiratory complications, as well as prolonged recovery from anesthesia [1]. Propofol has been used in this disorder with variable responses, including prolonged recovery [2], altered dose response, and precipitation of the myotonia [3]. We present a patient with MD who was anesthetized with propofol using a target-controlled technique for electrophysiologic examination and cardiac catheter ablation.

Case report

The patient was a 20-year-old man, 175 cm in height, and weighing 60 kg. Arrhythmia was indicated 5 years ago, and his electrocardiogram showed atrial flutter. When he was 17 years old, he began to experience grip myotonia and dysphagia. Atrial flutter and myotonia had been followed in an outpatient department with no specific treatment. However, the symptoms of myotonia had been aggravated, and further examination of myotonia and atrial flutter was scheduled.

Examination revealed myotonia in the upper limb muscle. Serum creatine phosphokinase was 334 IU/l (normal range, 76–251). An electromyogram obtained from the right biceps showed myotonic discharge at rest. From these findings, he was diagnosed as having myotonic dystrophy. Cardiac electrophysiologic examination and radiofrequency catheter ablation was scheduled under local anesthesia and propofol sedation. Before the procedure, arterial blood gas analysis at

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$F_{I_{O_2}}$ = 0.21 showed arterial oxygen tension (Pa_{O_2}) of 85 mmHg and carbon dioxide tension (Pa_{CO_2}) of 47 mmHg.

No premedication was given. Propofol infusion was begun to obtain target blood concentration of 1 μg ml^{-1} using a Diprifuser (TE-371; Terumo, Tokyo, Japan). When the effect-site concentration reached 1 μg ml^{-1}, the patient was still awake. Then the target concentration was carefully increased in steps of 0.5 to 2 μg ml^{-1}. At an effect-site concentration of 1.6 μg m^{-1}, he became unconscious and apneic. To support respiration, a laryngeal mask airway (LMA) was inserted and mechanical ventilation was performed. Anesthesia was maintained with 50% oxygen and propofol. When cardiac electrophysiologic examination was begun, target propofol concentration was set at 3 μg ml^{-1}. The target concentration was then gradually reduced. Spontaneous respiration began at an effect-site propofol concentration of 1.8 μg ml^{-1}. End-tidal CO_2 concentration was 62 mmHg and respiratory rate was 18 min^{-1}. The circulatory state was stable at this level of propofol concentration. However, when catheter ablation began, his arm moved suddenly. The target propofol concentration was increased to 2 μg ml^{-1} to deepen the level of sedation. Spontaneous respiration and arm movement disappeared at this level of propofol concentration. The procedure time was 5 h and 30 min, and the total consumption of propofol was 1290 mg. When the procedure was finished, the target propofol concentration was 1.6 μg ml^{-1} and the infusion of propofol was then stopped. The patient opened his eyes on verbal command about 10 min after the end of the procedure at an effect-site concentration of 1.2 μg ml^{-1} with sufficient spontaneous respiration. Thereafter, the LMA was removed. An arterial blood gas analysis at $F_{I_{O_2}}$ = 0.21 showed Pa_{O_2} of 80 mmHg and Pa_{CO_2} of 54 mmHg. The postanesthetic course was uneventful, and no exacerbation of myotonia was observed. When the patient was interviewed on the first postoperative day, he did not recall any events during anesthesia.

Discussion

MD is a rare, but serious, inherited disorder that may pose substantial problems for anesthetic management. The patient with MD has increased sensitivity to drugs used in anesthesia, such as induction agents, neuromuscular blocking agents, and opioids [1]. The use of inhalational anesthetics may produce shivering that can precipitate myotonia in the early postoperative period. The relationship between MD and malignant hyperthermia still awaits clarification.

We had planned the anesthesia for the MD patient with target-controlled infusion (TCI) of propofol for electrophysiologic examination and cardiac catheter ablation with spontaneous respiration. This technique is a common practice in our institite for prolonged procedures in normal patients. Unexpectedly, the patient became apneic at the same time as he lost consciousness. Although mask ventilation was easy, we had to insert an LMA and mechanical ventilation was performed for safe anesthetic management.

The use of propofol as an induction and maintenance agent in MD is controversial. In some reports, propofol had been successfully used [4,5]. However, other reports suggested prolonged recovery [2], exaggerated physiologic responses [3], and myotonia [6]. Tzabar et al. [2] used target-controlled propofol infusion in a patient with MD. They found prolonged recovery from anesthesia. In their case, spontaneous respiration appeared at a target propofol concentration of less than 2 μg ml^{-1}, and the patient showed signs of awakening at a blood propofol concentration of less than 1 μg ml^{-1} (precise concentration not described). Aquilona and Groves [7] also reported a case of TCI sedation in a patient with MD. Awareness occurred at a target propofol concentration of 1 μg ml^{-1} and over sedation with loss of airway control occurred at 2 μg ml^{-1}. In our case, the effect-site concentration at awakening was 1.2 μg ml^{-1} and spontaneous respiration began at 1.8 μg ml^{-1}. These concentrations were similar to those previously reported.

The effect-site propofol concentration at which the patient awoke (1.2 μg ml^{-1}) was slightly lower than our institutional average for adult male patients (1.5 ± 0.2 μg ml^{-1}) [8]. This result indicates that the sensitivity to propofol might be high in MD patients. However, the time from the end of anesthesia to the patient's awakening was about 10 min. In this case, emergence from propofol anesthesia was not delayed. Tzabar et al. [2] reported prolonged recovery time from propofol anesthesia; however, a high target concentration (6 μg ml^{-1}) during surgery might be the reason for delayed recovery. Other reports [4,5] showed rapid emergence from anesthesia from continuous propofol infusion.

Iwama et al. [9] assessed an anesthetic technique for achieving spontaneous breathing through the LMA during combined epidural block and propofol anesthesia for normal patients having elective lower extremity surgery. An induction dose of 1.5–2.0 mg kg^{-1} and infusion at a rate of 5 mg kg^{-1} h^{-1} was suggested as an appropriate dose to preserve spontaneous breathing with LMA. The estimated blood propofol concentration at this dose was about 2.5 μg ml^{-1}. In our case, respiratory depression occurred at effect-site propofol concentration of 1.6 μg ml^{-1}, which is lower than the concentration reported by Iwama et al. Although we had planned to perform the procedure in our patient using propofol sedation with spontaneous respiration, we had to insert

an LMA and to maintain anesthesia with mechanical ventilation. It is still not clear whether respiratory depression can occur more easily with propofol in MD patients. Further experience would be needed to confirm the respiratory depressive effect of propofol in MD patients.

TCI of propofol had advantages over conventional infusion for our patient. Iwakiri et al. [10] reported that the effect-site concentration of propofol at loss of consciousness was similar to that at awakening. This finding suggests that the effect-site concentration at loss of consciousness can be used as a guide to anesthetic management. We induced anesthesia with an incremental increase in target propofol concentration. The patient had not lost consciousness at an effect-site concentration of $1.0\,\mu g\,ml^{-1}$, so the concentration was then increased. The effect-site concentration of propofol at loss of consciousness was $1.6\,\mu g\,ml^{-1}$. Therefore, we could expect that the patient might wake up at an effect-site propofol concentration around $1.6\,\mu g\,ml^{-1}$ at the emergence period. Careful titration of propofol by TCI enabled evaluation of the patient's sensitivity to propofol.

We did not measure the actual blood concentration of propofol in this case. The accuracy of the TCI system for MD patients has not been evaluated. We should consider that the different body composition in patients with MD, including a decrease in muscle volume and abnormal muscle characteristics, might influence the pharmacokinetics of propofol.

We conclude that TCI of propofol is a useful anesthetic technique in MD patients. Respiratory depression might occur in MD patients at relatively low blood propofol concentrations. Precise control and titration of target propofol concentration is important in the anesthetic management of MD patients.

References

1. Russell SH, Hirsch NP (1994) Anaesthesia and myotonia. Br J Anaesth 72:210–216
2. Tzabar Y, Marshall R (1995) Myotonia dystrophy and target-controlled propofol infusions. Br J Anaesth 74:108–109
3. Speedy H (1990) Exaggerated physiological responses to propofol in myotonic dystrophy. Br J Anaesth 64:110–112
4. Bennun M, Goldstein B, Finkelstein Y, Jedeikin R (2000) Continuous propofol anaesthesia for patients with myotonic dystrophy. Br J Anaesth 85:407–409
5. White DA, Smyth DG (1989) Continuous infusion of propofol in dystrohia myotonia. Can J Anaesth 36:200–203
6. Kinney MA, Harrison BA (1996) Propofol-induced myotonia in myotonic dystrophy. Anesth Analg 83:665–666
7. Aquilona A, Groves J (2000) A combined technique utilising regional anaesthesia and target-controlled sedation in a patient with myotonic dystrophy. Anaesthesia 57:385–386
8. Morimoto Y, Matsumoto A, Koizumi Y, Ishida K, Tamura T, Sakabe T (2003) Effect of body fat percentage on estimated propofol concentrations at awakening from anesthesia using target controlled infusion (in Japanese with English abstract). Masui (Jpn J Anesthesiol) 52:967–971
9. Iwama H, Nakane M, Ohmori S, Kato M, Kaneko T, Iseki K (2000) Propofol dosage achieving spontaneous breathing during balanced regional anesthesia with the laryngeal mask airway. J Clin Anesth 12:189–195
10. Iwakiri H, Nishihara N, Nagata O, et al. (2005) Individual effect-site concentrations of propofol are similar at loss of consciousness and at awakening. Anesth Analg 100:107–110