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Anesthetic management for subtotal gastrectomy in a patient with paramyotonia congenita

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Abstract
We performed anesthesia for a subtotal gastrectomy in a 70-year-old female patient with paramyotonia congenita (PC). She had been diagnosed with PC at the age of 47 years by electromyogram analysis. Several points to consider have been revealed regarding the management of anesthesia in patients with PC. In this patient, anesthesia was safely maintained using sevoflurane and nitrous oxide together with concomitant epidural anesthesia using mepivacaine. Efforts should be made to prevent perioperative attacks of muscle weakness when planning anesthesia for patients with this kind of disorder. Specifically, refraining from the use of muscle relaxants, care with regard to the composition of infusion fluids during operations, and the maintenance of body temperature are required for anesthesia. In addition, postoperative pain management using a continuous epidural block proved to be a useful method.

Key words Paramyotonia congenita · Management of anesthesia

Introduction

Paramyotonia congenita (PC) is classified under the category of ion channel myotonias, which consists of channelopathies with myotonia. According to the classification of myotonia, it is thought that PC is a subgroup of myotonia congenita (voltage-gated chlorine channel) or hypokalemic periodic paralysis type I (voltage-gated calcium channel). PC is a disease associated with mutation in the α-subunit of the skeletal muscle voltage-gated sodium channel in the neuromuscular junction. This disease resembles hyperkalemic periodic paralysis and potassium-aggravated myotonia in its clinical features, because genetic analysis of patients with the three diseases (i.e., PC, hyperkalemic periodic paralysis, potassium aggravated myotonia) has shown that mutation of a gene at chromosome 17q is responsible for the symptoms. Mutant channels exhibit sustained sodium currents that lead to prolonged membrane depolarization, causing myotonia, followed by membrane desensitization (or inactivation), resulting in paralysis. Clinically, this disorder is characterized by transient stiffness (myotonia) precipitated by exposure to such factors as cold temperatures or physical exertion.

Several case reports of anesthesia employed for patients with PC have been published [1–3], and below we describe our experiences with anesthesia employed for a PC patient who underwent a subtotal gastrectomy for treatment of a gastric carcinoma.

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Case report

The patient was a woman aged 70 years with a body weight of 30.5 kg and height of 149.5 cm. Her chief complaint was a decrease in activity during daily life. The patient had experienced weight loss, and difficulty waking in the morning. An examination led to the diagnosis of gastric carcinoma, and a subtotal gastrectomy was scheduled. Her past medical history included hyperthyroidism, but the patient was in a euthyroid state. With regard to the clinical progress of PC, she had been unable to raise her head at 4–5 months after birth. At the age of 1 year, she began to experience stiffness of the limbs, which was aggravated by cold temperatures. At 47 years of age, bilateral digital myotonia of the hand had worsened and electromyograms revealed myotonic discharges. Provocation tests produced negative results for glucose and insulin, but myotonia was induced by the oral administration of 5 g potassium chloride. A whole-body cold-exposure test showed conspicuous provocation. A definite diagnosis of PC was then made, based on the results of these examinations and the patient's medical and family history. Preoperative exam-

Table 1. Preoperative examination values

WBC	$7380 \cdot \mu l^{-1}$	Na	$131.2\,mEq \cdot l^{-1}$	TP	$4.7\,g \cdot dl^{-1}$
RBC	$352 \times 10^4 \cdot \mu l^{-1}$	K	$4.5\,mEq \cdot l^{-1}$	Alb	$2.7\,g \cdot dl^{-1}$
Hb	9.4 g·dl	Cl	$98.4\,mEq \cdot l^{-1}$	CPK	$584\,IU \cdot l^{-1}$
Ht	30.8%	BUN	$14.5\,mg \cdot dl^{-1}$	GOT	$56\,IU \cdot l^{-1}$
PLT	$20.1 \times 10^4 \cdot \mu l^{-1}$	Cr	$0.21\,mg \cdot dl^{-1}$	GPT	$29\,IU \cdot l^{-1}$

BGA, pH 7.46; Pa_{CO_2} 37.9 mmHg; Pa_{O_2} 73.5 mmHg; HCO_3^- $26.4\,mmol \cdot l^{-1}$; BE $2.9\,mmol \cdot l^{-1}$
Chest radiograms, no abnormal findings
Electrocardiograms, normal sinus rhythm with no abnormal findings

BGA, blood gas analysis in artery

ination values are shown in Table 1. Echocardiography revealed an ejection fraction value of 74% and normal wall motion.

As premedication, ranitidine (150 mg) was administered orally. After the patient was brought into the operating theater, an epidural catheter was inserted at the intervertebral space between Th9 and Th10, approximately 9 cm cephalad from the skin puncture site. Anesthesia was induced by slow induction, using sevoflurane after the intravenous administration of fentanyl (0.1 mg); the trachea was intubated without muscle relaxant. Anesthesia was maintained with oxygen (2l), nitrous oxide (4l), and sevoflurane (1%–1.5%) in combination with epidural anesthesia. Eight ml of 1.5% mepivacaine was administered into the epidural space as an initial dose 10 min before the operation was commenced. Body temperature (BT) was monitored by measurements of the rectal temperature. After the induction of anesthesia, atropine sulfate (0.25 mg) was administered intravenously in order to rectify bradycardia (heart rate $48 \cdot min^{-1}$). During the operation, pressor agents (total, ephedrine 16 mg and phenylephrine 0.6 mg) were used as required to control blood pressure. The operation was completed in 71 min. The volume of surgical blood loss was 135 g, the urine output was 350 ml, and the volume of parenteral fluid given was 600 ml (potassium-free T1 infusion fluid).

Recovery from anesthesia was normal, and the endotracheal tube was removed upon confirmation of spontaneous respiration, movement of the extremities, and level of consciousness. The patient's condition was monitored for a while, and after a good arousal state was confirmed, the patient was returned to the ward. The duration of anesthesia was 125 min. Postoperative pain was controlled with a continuous epidural infusion, consisting of 200 ml of 0.2% ropivacaine, 0.6 mg (12 ml) of fentanyl, and 28 ml of physiological saline (240 ml in total at a rate of $5\,ml \cdot hr^{-1}$). This pain management was successfully performed, achieving adequate control of pain without any complications. This was an effective method of postoperative pain management in our patient with PC.

Discussion

PC is a very rare, highly pervious, autosomal dominant inherited disease that arises from mutation of the *SCN4A* gene on chromosome 17q. The first case of this disease was reported by Eulenburg in 1886, and in Japan, the first case report was published by Nogi et al. in 1948 [4]. There have been approximately ten positive family lines reported to date in Japan, but the present patient is an extremely rare case in that there have been seven phenotypically affected members in the four generations preceding and succeeding our patient, within a single pedigree (Fig. 1).

Clinical manifestations of PC can occur soon after birth and persist throughout life though not noticeably varying in the severity of symptoms. Attacks of muscular weakness occur but are neither severe nor fatal. Muscles related to respiration are not affected, and myopathy is rarely noted. It has been shown that cold, exercise, starvation, and the administration of potassium are factors that can induce myotonia [2]. It has been shown that high-carbohydrate diets and mild exercise are effective in the prevention of myotonia. Medicines including acetazolamide and mexiletine [5] have been recommended for the treatment of PC.

Several points to consider have been revealed regarding the management of anesthesia for patients with PC, and we planned the anesthesia in our patient taking into account the following points for intraoperative management.

Anesthesia was induced by slow induction, using sevoflurane to permit tracheal intubation without the use of muscle relaxants, and the trachea was intubated as soon a sufficient depth of anesthesia was achieved. In patients with PC, it is thought that the use of muscle relaxants should be avoided regardless of whether they are depolarizing or nondepolarizing agents. In particular, succinylcholine is contraindicated, as its use will result in increases in serum potassium concentrations and this can cause myotonic symptoms in these types of patients [6,7]. The use of anticholinesterase drugs should be avoided because they may induce myotonic reactions

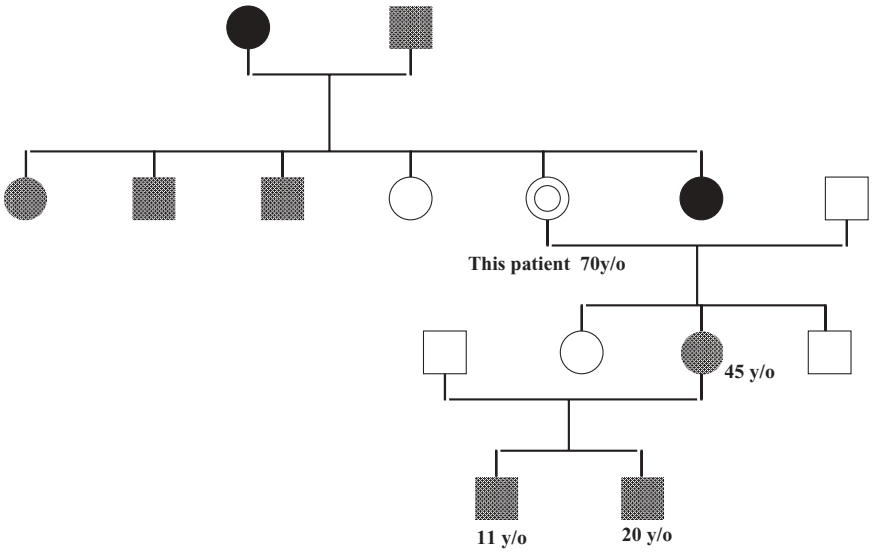

Fig. 1. Pedigree of the patient's family. *Hatched symbols*, patients with paramyotonia congenita. *black symbols*, deceased; *y/o*, years old

[8]. In any event, the use of muscle relaxants should be considered with care, and it is generally thought that their use should be avoided as a rule.

The maintenance of anesthesia was performed using sevoflurane (≤1.0 minimum alveolar concentration [MAC] approximately), nitrous oxide, and oxygen in combination with epidural anesthesia using 1.5% mepivacaine. With regard to the relationship between PC and anesthetics, volatile inhalation anesthetics may be related to this disorder as it is a channelopathy due to a sodium channel abnormality, in comparison with malignant hyperthermia, which is a channelopathy due to a calcium channel abnormality. Therefore, caution should be exercised when using volatile anesthetics in patients with PC. However, there have been no studies specifically dealing with the relationship between PC and inhalation anesthetics, or articles demonstrating a higher incidence of malignant hyperthermia among patients with PC as compared with healthy subjects [9], although myotonic symptoms appear in patients with this disorder. Anesthetics of this class can thus be used in the clinical setting for patients with PC, because underlying abnormal ion channels differ between malignant hyperthermia and PC. A report has described that the intravenous anesthetic propofol can be used without concern in inducing and maintaining anesthesia in patients with PC [10], whereas another report states that there is a delayed recovery in association with its use in PC patients because the sodium channel-mediated sodium influx current is blocked [11]. Local anesthetics are considered to be relatively safe for use in anesthesia in patients with PC. A study showed that a cesarean section did not involve any events of clinical concern in a patient with this disorder when performed under spinal anesthesia [12]. Another report has documented the use of epidural analgesia in a patient with this disorder who experienced pregnancy and parturition [13].

It is considered important to perform subtle BT management, including careful postoperative BT management in patients with PC. For maintenance of BT during the course of the operation in our patient, the extremities and body surface were actively kept warm to maintain adequate peripheral temperature, through the warming of fluid injections and the use of a machine to supply a warm breeze. As a result, the rectal temperature was maintained at 36.4°C to 35.8°C during the operation. As well as BT management, intraoperative fluid management is also important. We used the potassium-free intravenous solution Solita T1 (Ajinomoto Pharma, Tokyo, Japan) in order to avoid potassium administration. The preoperative potassium value was 4.5 $mEq \cdot l^{-1}$, and the potassium value after the operation was 4.1 $mEq \cdot l^{-1}$.

Finally, it is most important to design an anesthetic procedure that enables the prevention of perioperative attacks of muscle weakness in PC patients. From the experience with our patient, it is thought that an anesthetic procedure consisting of induction with sevoflurane and fentanyl, and maintenance with sevoflurane, nitrous oxide, fentanyl, and epidural anesthesia proved to be safe.

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