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General anesthesia for progressive external ophthalmoplegia syndrome

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Introduction

The progressive external ophthalmoplegia syndrome (ocular myopathy) is a slowly advancing disorder which is associated with increased weakness of the extraocular muscles and paresis of ocular movement [1,2]. Recently, abnormalities of muscle mitochondria have been reported in patients with progressive external ophthalmoplegia syndrome [3–5]. Although there is only one report of anesthesia in a patient with mitochondrial myopathy [6], no report has been made on the anesthesia for progressive external ophthalmoplegia syndrome. We present the anesthesia for a patient with progressive external ophthalmoplegia syndrome.

Case report

A 55-year-old woman, weighing 46 kg and 157 cm in height, was admitted to our hospital because of bilateral blepharoptosis of unknown origin. The patient first experienced bilateral blepharoptosis at the age of 41. The blepharoptosis worsened over the next 3 years. There were no other neurological symptoms such as muscle weakness. Hematological studies, renal and liver function, and arterial blood gas analysis were within the normal range. A tensilon test was negative. Anti-acetylcholine-receptor antibody was not detected. An electromyogram (EMG) test did not demonstrate muscle and neuromuscular junction abnormalities. A preoperative chest roentgenogram and brain computed tomography (CT) scan were normal. The retrenchment of the bilateral palpebralis was planed.

A neuromuscular blockade monitor (Datax relaxograph, Helsinki, Finland) was set before the induction of anesthesia. Anesthesia was induced with thiopental and vecuronium. The train-of-four (TOF) ratio reduced to 30% 5 min after the intravenous administration of 1 mg of vecuronium. The TOF ratio reduced to 0% after the further addition of 2 mg of vecuronium. The trachea was intubated and anesthesia was maintained with nitrous oxide (60%) and isoflurane (1.0%–1.5%) in oxygen. No neuromuscular blocking agent was added after the induction of anesthesia. Blood pressure, heart rate and body temperature were stable during the surgery. Antibiotics were not administrated pre- or intra-operatively. The TOF ratio was 50% 1 h after initial administration of vecuronium and about 80% after 3 h of surgery. The TOF ratio returned to 100% following administration of neostigmine 1 mg and the tracheal tube was extubated.

The patient underwent retrenchment of the right palpebralis 2 months after the first surgery. Induction and maintenance of anesthesia were performed in a similar way using thiopental, vecuronium, nitrous oxide, and isoflurane. The prolonged neuromuscular blocking effect of vecuronium was again observed and neostigmine was administrated (Fig. 1). The tracheal tube was extubated and the postoperative course of the patient was uneventful.

Discussion

Since Von Graefe first described the disorder of palpebralis as progressive external ophthalmoplegia syndrome in 1868 [7], this syndrome has been elucidated by several investigators [1,8,9]. In 1972, Olsen et al. showed muscle fiber abnormalities including excessive accumulation of mitochondria in seven patients with progressive external ophthalmoplegia [4].

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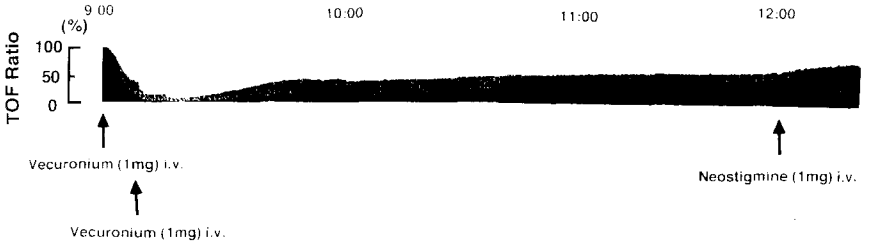

Fig. 1. The effect of vecuronium in a patient with the progressive external ophthalmoplegia syndrome. The effect of 2 mg of vecuronium on train-of-four (*TOF*) ratio in a patient with progressive external ophthalmoplegia syndrome in the re-trenchment of right palpebralis. The datum was recorded by stimulating the left ulnar nerve with a neuromuscular blockade monitor. The duration of the action of vecuronium was longer than that in normal patients

Recently, Maraes et al. suggested that deletions of muscle mitochondrial DNA are associated with progressive external ophthalmoplegia [5].

Special anesthetic problems in the patients with progressive external ophthalmoplegia syndrome seem to be: (1) possible susceptibility to malignant hyperthermia, (2) hypersensitivity to nondepolarizing neuromuscular blockades, and (3) postoperative respiratory dysfunction.

It is well known that there is a relation between depolarizing neuromuscular blockades and malignant hyperthermia in muscular disorders [10]. Previously, the uneventful use of vecuronium in a patient with Kearnes-Sysre syndrome, a subtype of mitochondrial myopathy, was reported [6]. We used a nondepolarizing muscle relaxant, vecuronium, to facilitate tracheal intubation.

In normal human adults, 0.1 $mg \cdot kg^{-1}$ body weight is necessary for complete muscle relaxation and 100% recovery time of TOF is about 1 h [11]. A smaller dose of vecuronium (0.044 $mg \cdot kg^{-1}$) was sufficient to reach 0% TOF ratio indicating that this patient was more sensitive to vecuronium than normal adults (Fig. 1). Although vecuronium is a nondepolarizing neuromuscular blockade with short duration of action, about 1 h in normal patient, its effect lasted for over 3 h in this patient. Neostigmine was effective to reverse vecuronium. The neuromuscular blockade monitor was useful to continuously assess the extent of neuromuscular junction blockage.

Postoperative respiratory depression was another consideration in this patient because of the possibility that effects of inhalation anesthetic and vecuronium might persist postoperatively. The postoperative course of this patient was uneventful.

In summary, we described a case of general anesthesia for a patient with progressive external ophthalmoplegia syndrome. The patient was hypersensitive to a nondepolarizing neuromuscular blocking agent. A smaller than usual dose of vecuronium was sufficient.

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