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Anesthetic Management of a patient With Charcot-Marie-Tooth Disease

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Abstract **Case Report**

Charcot-Marie-Tooth disease (CMTD) is a hereditary peripheral neuropathy and is characterized by progressive muscle atrophy and motor-senrory disorders in all 4 limbs. A 27-year- old man was scheduled to undergo endoscopic sinus surgery, septoplasty and turbinoplasty due to chronic paranasal sinusitis, deviated septal nose and both chronic hypertrophic rhinitis. He had been diagnosed with Charcot-Marie-Tooth disease in 2008. General anesthesia was induced with propofol 120 mg and the intubating condition was achieved with rocuronium 50 mg. Anesthesia was maintained with 1.5-2% sevoflurane and remifentanil. During surgery, the body temperature and end tidal concentration of CO2 were maintained within the normal range. Despite the continuous monitoring of the train-of-four (TOF) response, no more muscle relaxants were required during surgery and the patient recovered with suggamadex 200mg and the patient awaked without a delay. In the management of patients with Charcot-Marie-Tooth disease, it is desirable to evaluate the patient carefully, select the appropriate anesthetics and adjust the dosage of the drug according to the patients requirements.

Keywords: Charcot-Marie-Tooth disease, motor-senrory disorders, turbinoplasty, Anesthesia.

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INTRODUCTION

Charcot-Marie-Tooth disease is a rare hereditary disease, first reported by J. M. Charcot, P. Marie, and H. H. Tooth in 1886 [1]. This disease develops with occurrence of mutation in myelin sheaths surrounding an axon of a neuron. It is a typical hereditary peripheral neuropathy with prevalence of about 30 to 40 persons per 100,000 population. Atrophy of distal motor muscle and weakening of tendon reflexes occur in patients with this disease, and characteristic peroneus muscle atrophy ("stork-leg appearance") and foot deformities including club feet and talipes are exhibited [2]. In severe cases, the disease can progress to respiratory failure and patients may require mechanical ventilation. Increase in sensitivity to neuromuscular blocking agents, hyperkalemia [3] and delayed recovery due to prolonged effect of neuromuscular blocking agents may occur. Therefore, strict manangement of anesthesia should be performed under neuromuscular monitoring in patients with Charcot-Marie-Tooth disease. We report a case of a patient with Charcot-Marie-Tooth disease under a rhinologic surgery and experience in anesthesia with Train-of-Four monitoring and sugammadex.

CASE REPORT

A 27-year-old male patient diagnosed with chronic paranasal sinusitis, Deviated septal nose, and both chronic hypertrophic rhinitis, was scheduled to receive endoscopic sinus surgery, septoplasty and turbinoplasty under general anesthesia. He was diagnosed with Charcot-Marie-Tooth disease in 2008, and distal muscular weakness was progressing. The patient showed no signs of muscular atrophy or attenuated tendon reflex in extremities. Preoperative chest radiograph, electrocardiogram, and laboratory blood tests revealed no specific findings. No preoperative medication was administered, and upon entering the operation room, the patient was monitored with electrocardiogram, pulse oximeter, capnography, and nerve stimulator. Initial vital signs before induction were: systolic and diastolic blood pressure 120/70mmHg, pulse rate 80/min, oxygen saturation as measured by pulse oximetry of 100%. Nerve stimulator (Fisher & Paykel healthcare, New zealand) was attached to left wrist area, along the path of ulnar nerve, and contraction of adductor pollicis muscle in response to TOF (train of four, 2 Hz, 0.2msec) stimulation every 20 seconds was monitored. 5 minutes of preoxygenation was performed with 100% O_2, followed by induction of anesthesia by 120mg of propofol and 50mg of rocuronium. Anesthesia was maintained using

sevoflurane and remifentanil. Tracheal intubation was performed after complete loss response to TOF stimulation. Initial plan for additional rocuronium administration was to give 5mg of rocuronium intravenously every time when TOF response showed more than 2 counts. However, no additional doses of rocuronium were administered. The operation lasted 2 hours, and the end of the operation, response to TOF stimulation recovered to 2 counts after initial rocuronium administration. For reversal of neuromuscular blockade, 150mg of suggamadex, dose calculated with 2mg/kg of patient weight, was administered. Extubation was performed after confirming return of awareness, recovery of spontaneous ventilation, and response to TOF stimulation of 100%. Patient showed no specific post-anesthetic sequelae of complication in post-anesthetic care unit and in ward, and was discharged on post-operation day 1.

DISCUSSION

Charcot-Marie-Tooth disease can progress to respiratory failure and patients may require mechanical ventilation. Preoperative anesthetic concerns relating to this disease include: respiratory muscular weakness due to involvement of phrenic nerve [6], and possible requirement of mechanical ventilation due to respiratory failure [7], cardiac menifestations attributed to this neuropathy including conduction distarbance, atrial flutter, and cardiomyopathy. Propofol, which possess lesser risk of delayed recovery, was used instead of thiopental for induction of anesthesia. To prevent succinylcholine-induced hyperkalemia and related changes in electrocardiogram, rocuronium was used for muscle relaxation under TOF monitoring, without requirement for additional rocuronium administration after the induction dosing. Sugammadex was used for neuromuscular blockade reversal, and patient was extubated after confirming return of awareness, recovery of spontaneous ventilation, and response to TOF stimulation of 100%. Patient showed no further difficulty in spontaneous ventilation after the extubation.

Management of anesthesia in patients with Charcot-Marie-Tooth disease must be focused on reaction to neuromuscular blocking agents, and possibility of postoperative respiratory failure due to weakness of respiratory muscles. In case of succinylcholine, excessive release of potassium may occur in patients with neuromuscular disease, and must be used with caution. Malignant hyperthermia can be related with volatile anesthetics, with exception of N_2O, but with less possibility in mild form of Charcot-Marie-Tooth disease [8].

In conclusion, in patients with Charcot-Marie-Tooth disease, reaction to neuromuscular blocking agents, possibility of developing malignant hyperthermia and postoperative respiratory failure must be taken in to consideration when making choices for anesthetic medications, and appropriate measures for possible postoperative respiratory complications should also be prepared.

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