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Letter to the Editor

Anesthesia management in charcot-marie-tooth disease

Merve Yusifov, Resul Yilmaz, Aysegul Bilge , Ercan Kaya, Ahmet Topal

Necmettin Erbakan University, Meram Medical Faculty, Department of Anesthesiology and Reanimation, Konya, Turkey

Dear Editor,

Charcot-Marie-Tooth (CMT) disease is a genetically inherited polyneuropathy defined by JM Charcot, P Marie and HH Tooth in 1886 (1). Its frequency is 1/2500-400 and there is no relation with race or gender (2). The syndrome is manifested by loss of strength of the lower extremity distal muscles, and a progressive muscular atrophy from distal to proximal is observed (3).

Neuropathy in patients with CMT, may lead to serious problems in general anesthetic practice and make neuromuscular blockade an important problem. In this case report, we aimed to share our experience in a patient with CMT and present the problems that may develop during the management of anesthesia of these patients.

A 42-year-old, 80 kg, female patient was scheduled for laparoscopic cholecystectomy. Preoperative evaluation revealed that she had hypertensive disease, and she was taking antihypertensive medication, she had underwent general anesthesia during a cesarean operation, and that it took a long time for her to recover. According to the patient's history, when she was 12 years old she went to the neurology clinic with complaints of numbness in her feet, not being able to stand for a long time and falling down. After some time, similar symptoms were revealed in her sibling and as a result of examinations and tests performed because of the appearance of similar symptoms, she was was diagnosed with Charcot-Marie-Tooth disease. It was discovered that over time numbness and deformity occurred in her hands and toes, followed by development of drop foot. During the preoperative examination, numbness and deformity were present in hands and feet (Figure 1). Upper extremity muscle strength was 6/6, lower extremity was 4/6, while upper extremity deep tendon reflexes were normal and lower extremity deep tendon reflexes were weak. Blood count and biochemical parameters were normal. The respiratory function test was in the expected range according to age, weight and height measurements.

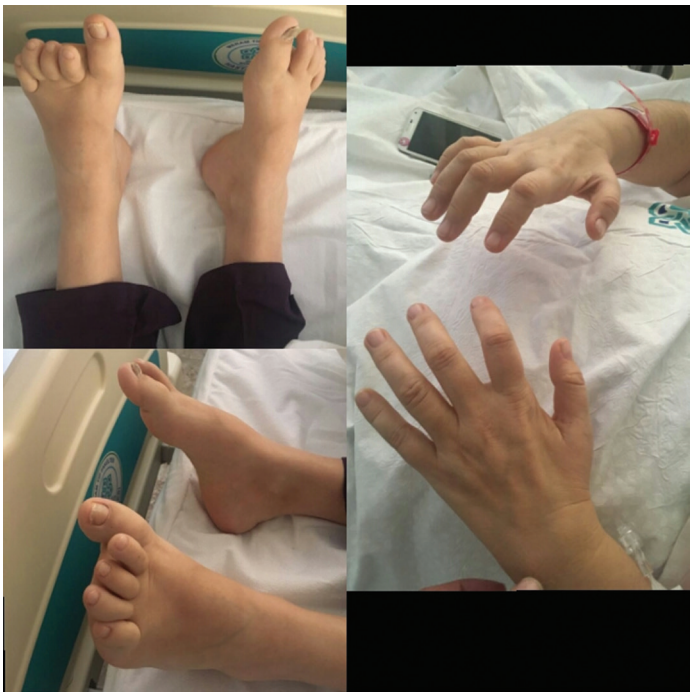

Figure 1. The appearance of the patient's hands and feet

The Electrocardiogram (ECG), pulse oximetry (SpO2), noninvasive blood pressure (NIBP), temperature and neuromuscular transport (NMT) were monitored in the operation room. Our BIS device is defective, for that reason BIS monitorization could not be done. The baseline hemodynamic values were recorded as: pulse 85 beat/min, SpO2 96%, NIBP 145/88 mmHg. The patient was given 1 mg midazolam intravenously for anxiety. Induction of anesthesia was continued with propofol 2 mg/kg and after NMT supramaximal value was taken, 0.5 mg/kg rocuronium was used for intubation and the train of four (TOF) was started. When optimal intubation conditions were reached, the patient was intubated and the operation started. Total intravenous anesthesia (TIVA) was used for anesthesia maintenance and, propofol (6 mg/kg/min) and remifentanil (0.25 mg/kg/min) was administered. The

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Corresponding Author: Resul Yilmaz, Necmettin Erbakan University, Meram Medical Faculty, Department of Anesthesiology and Reanimation, Konya, Turkey. E-mail: dr.r.yilmaz@gmail.com

operation lasted approximately 30 minutes and during that time hemodynamic parameters were stable. 150 mg of tramadol was administered intravenously for analgesia. Because of the continuation of the neuromuscular deep block, the patient was given sugammadex (2 mg/kg) intravenously. After spontaneous breathing started, she was extubated and oxygen support was provided by mask ventilation. After 20 minutes, the patient was able to communicate comfortably and resume breathing without desaturation in room air. For close observation, the patient was kept in the postoperative care unit for about 1 hour and no problems developed during this period. The patient was discharged on the third postoperative day. The main features of Charcot-Marie-Tooth disease are muscle atrophy and motor-sensory conduction disturbance. It is a peripheral neuropathy that starts in childhood and is clinically manifests when the patient is middle aged(1). It has been reported that some patients who manifest symptoms at an earlier age have been followed for a long time with the diagnosis of poliomyelitis (4). Nerve degeneration and clinical manifestations are varied (1). Our patient had muscle weakness and anatomic disorders in the hands and lower extremity, but she was able to perform her daily activities. The main causes for problems that may be encountered in anesthesia practices are genetic disorders in the myelin sheaths or axons and can result in motor, sensory, and autonomic dysfunctions (5). Loss of strength and sensory loss in the extremities, anatomic disorders, as well as cardiac and respiratory problems may accompany the disease. Postoperative respiratory distress, pneumonia, long-term need for ventilator support were reported in the literature (5). Inadequate respiratory effort or deteriorated thoracic structure can be the cause of the supine position. A wide variety of cardiac rhythm disturbances can be seen from premature beats to a-v complete blocks (6). The use of thiopental for general anesthesia is of interest. Kotani N et al. (7) reported in their study, that the needed dose of the drug was inversely related to neuropathy involvement in patients with CMT, and that the use of thiopental should be reduced as the severity of the disease increases. Alzaben KR et al. (8) reported successful propofol and dexmethothymidine administiration for intravenous anesthesia. We prefered propofol as the hypnotic agent and combined remifentanil with propofol for the duration of the surgery and completed the anesthesia management without any problems. Another issue to consider for patients with CMT is malignant hyperthermia. Therefore, the volatile anesthetic agents and succinylcholine are not preferred options (5). We preferred TIVA and dantrolen was kept ready for the risk of malignant hyperthermia. Heat follow up of the patient was conducted through the case.

When the literature is examined, it is seen that different muscle relaxants are used without problems, but denervation in the muscles makes the use of these agents and follow-up of the patients difficult (9). No prolonged effects have been reported in patients treated with succinylcholine. However, the possibility of hyperkalemia and possible triggering of malignant hyperthermia in polyneuropathies and patients with nerve trauma should be considered in the use of succinylcholine (10). We also considered malignant hyperthermia in our patient, and we chose to use rocuronium that we have more experience with. In addition, neuromuscular monitoring was performed, and we evaluated the patient response to rocuronium and the return of the block objectively. At the end of the case we used sugammadex to restore effective muscle strength. In neurological diseases based on neuropathy, regional anesthesia techniques are believed to worsen the disease and are generally not preferred. However, there are case reports of regional anesthesia in patients with CMT. Patients with CMT are a special patient group and the involvement areas of the disease should be evaluated carefully preoperatively. Anesthesia management should be conducted in a way that is most appropriate for the patient's specific clinical features.

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