CHARCOT-MARIE-TOOTH:
GENERAL ANESTHESIA GUIDELINES

by

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Abstract
Charcot-Marie-Tooth disease (CMT) is the most common inherited neuromuscular disorder. This case report will detail the events of a 31 year-old female undergoing a uterine ablation under general anesthesia. CMT is a genetically inherited disease with a prevalence of 1 in 2,500 people.\textsuperscript{1} CMT affects both the motor and sensory nerves in affected patients. CMT has a typical onset within the first two decades of life and shows a slow, continual progression for the remainder of the patient’s life.\textsuperscript{2} These patients often require multiple orthopedic surgeries in the course of their life, necessitating the need for anesthesia professionals to be aware of the anesthetic risks associated with CMT.
Case Report

A 31 year-old, 72kg female presented for uterine ablation secondary to persistent abdominal pain and vaginal bleeding. The patient’s only current medication was drospirenone. Her vital signs and all laboratory values were within normal limits. She did not have any known drug allergies. A pre-operative interview found that the patient suffered from occasional acid reflux, but that she did not take any medications for it. She was not feeling any reflux at the time. An assessment of her neuromuscular function was conducted and she was found to have complete motor and sensory capabilities in all four extremities with only mild weakness noted in both lower extremities. The patient denied any shortness of breath or difficulty breathing. The patient was assigned a physical status classification of II using the American Society of Anesthesiologists (ASA) guidelines. Risks and benefits of anesthesia were explained and anesthesia consent was obtained. The patient was given midazolam 2mg (IV) in pre-operative holding and then transferred into the operating room. Once on the operating table, oxygen was administered to the patient via facemask with flows of 10 L/min. Standard ASA monitors were applied. The patient was allowed to pre-oxygenate for a period of 3-5 minutes before anesthesia was induced with the administration of propofol 140 mg in the patient’s IV catheter. Once the patient was asleep, a laryngeal mask airway (LMA) was placed without complication. Placement was verified by noting the presence of end-tidal carbon dioxide (CO₂) and by auscultation of bilateral breath sounds. Oxygen was administered through the LMA via the anesthesia circuit at a rate of 2 L/min and anesthesia was maintained by the administration of sevoflurane 2%. A skin temperature probe was then placed in the patient’s right nare. All pressure points were padded and the case began. The patient’s vital signs were stable throughout the case and spontaneous ventilation was maintained. The patient received a total of fentanyl 150 mcg for pain control. She
was also administered dexamethasone 4mg and ondansetron 4 mg to minimize nausea. At the end of the case the sevoflurane was discontinued and 100% oxygen was administered via the LMA at 10L/min. The patient opened her eyes on command and obeyed all verbal orders prior to removal of the LMA. Once the LMA was out, the patient was transferred to the PACU where she denied pain or nausea. Another neuromuscular assessment was performed after she was fully awake and the same results were found as in the pre-surgery assessment.

Discussion

Signs and symptoms of CMT include: hammer toes, foot drop, motor and sensory loss of the distal portion of both lower and upper extremities, and a reduced or absent deep tendon reflex.\(^2\) Symptoms begin in the lower extremities, then progress to the upper extremities. Rarely does the disease affect the muscles of respiration, however, involvement of the respiratory system can be difficult to assess. “Patients may have few or no symptoms despite considerable abnormalities in pulmonary function. The presence of proximal muscle weakness of the arms may be a predictor for respiratory muscle weakness.”\(^3\) Volume flow loops of a patient with CMT display a restrictive lung pattern.\(^3\) A patient displaying respiratory weakness before surgery may require ventilation post-operative period.\(^4\) Best practice technique is to adequately assess for the presence of muscle weakness in the arms and to ensure a thorough pre-oxygenation prior to induction.

As a result of the muscle weakness caused by CMT, patients should be considered at risk for muscle wasting and the associated hyperkalemia. Therefore, the administration of succinylcholine in patients suffering from CMT is contraindicated.\(^3\) Special care must be given
when giving non-depolarizing muscle relaxants. The response to non-depolarizing muscle relaxants can be unpredictable, exacerbated or attenuated, varying from patient to patient.\textsuperscript{3,4,5} It is of the utmost importance that if the use of non-depolarizing muscle relaxants is necessary that the dosage be decreased and that a thorough assessment of relaxation be completed via peripheral nerve stimulation. Patients with CMT must have their neuromuscular blockade completely antagonized and the anesthesia professional must ensure that no muscle weakness persists prior to extubation.

The use of nitrous oxide on patients with CMT can predispose them to neurotoxicity through inhibition of methionine synthesis, but only through prolonged exposure.\textsuperscript{3,5} Nitrous oxide is classified as posing a “moderate to significant” risk to patients with CMT.\textsuperscript{5} That being said, there are several reports of nitrous oxide being safely administered to patients with CMT with no worsening neuropathy.\textsuperscript{5} Anesthesia professionals must use sound judgment when deciding whether or not to use nitrous oxide and determine if the potential risks outweigh the potential benefits of its use.

A prominent risk of malignant hyperthermia (MH) in the CMT patient population has long been speculated, but never truly established.\textsuperscript{6} The safest approach for anesthesia professionals is to assess if there is a personal or familial history of MH with the patient.\textsuperscript{3,6} The patient in this case study had previously undergone anesthesia without any complications of MH and had no familial history of MH. For this reason sevoflurane was administered to the patient. Special attention was given to monitor her body temperature and carbon dioxide (CO\textsubscript{2}) levels for signs and symptoms of MH.
Another anesthetic consideration is the proper and careful positioning of CMT patients to protect against nerve compression that could result in the exacerbation of neuropathy. Proper body alignment and the use of padding on areas of boney prominences will decrease the risk of worsening a patient’s neuropathy.

In this case study the patient displayed very few of the classic signs and symptoms of CMT, indicating that she was in the early stages of the disease. A thorough assessment of her neuromuscular function provided a good basis for the state of the disease progression. In addition it provided a baseline assessment for the anesthesia practitioner that allowed for a post-operative assessment to determine if any exacerbation of symptoms had occurred. No exacerbations were found. The decision was made to avoid both depolarizing and non-depolarizing paralytics and use an LMA rather than an endotracheal tube. This decision was based upon shortness of the surgical case and the fear of prolonged muscle relaxation. The patient had no physical contraindication to using an LMA. Spontaneous ventilation was maintained throughout the case with tidal volumes between 6-8 ml/kg, indicating that the patient was not at risk for respiratory weakness in the post-operative period. Nitrous oxide was not used because the potential risks did not outweigh the benefits of its use, even considering the brevity of the case.

In the future similar cases could be managed differently by using non-depolarizing muscle relaxants at a decreased dosage and intubating the patient with an endotracheal tube if the need for a more secure airway is evident. Nitrous oxide could also be used in order to decrease the minimum alveolar concentration (MAC) of the volatile anesthetic agent.
References


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