



Anesthetic Management of A Patient with Charcot-Marie-Tooth Disease for 2-stage Revision of Total Knee Replacement

Eleftheria Soulioti , Foteini Kavezou , Georgia Efstathiou , Chrysanthi Batistaki , Agathi Karakosta ,
Georgia Kostopanagiotou 

Department of Anesthesiology, Attikon University Hospital, National and Kapodistrian University of Athens, Athens, Greece

Cite this article as: Soulioti E, Kavezou F, Efstathiou G, Batistaki C, Karakosta A. Anesthetic Management of A Patient with Charcot-Marie-Tooth Disease for 2-stage Revision of Total Knee Replacement. Turk J Anaesthesiol Reanim 2021; 49(2): 178-80.

Abstract

Charcot-Marie-Tooth disease is characterised by hereditary motor and sensory neuropathy. Its anaesthetic management is challenging owing to the unpredictable response observed in patients, especially to non-depolarising neuromuscular blocking drugs, and the risk of malignant hyperthermia and cardiorespiratory complications. A 66-year-old woman underwent anaesthesia for 2 different surgical procedures, a 2-stage revision of total knee replacement over a 4-month period. She presented with severe anatomic disorders, accompanied by severe motor and sensory impairment. An anaesthetic plan without neuromuscular blocking drugs or volatile anaesthetics, using a clean ventilator, with dantrolene available, was successfully used both times. There were no complications during the administration of general anaesthesia or postoperatively at the post-anaesthesia care unit, and the patient did not complain of pain at any time. General anaesthesia with a careful selection of anaesthetic drugs proved to be a safe option for the management of a patient with Charcot-Marie-Tooth disease.

Keywords: Anaesthesia, Charcot-Marie-Tooth, neuromuscular blockade, total knee replacement

Introduction

Charcot-Marie-Tooth disease was first described in 1886 and refers to a clinically and genetically heterogeneous group of disorders characterised by hereditary motor and sensory neuropathy and progressive loss of muscle tissue, with a prevalence of 1 in 2500. The disease progresses into muscular atrophy of the lower extremities (which can affect the upper extremities as well), slowly aggravating weakness and sensory dysfunction (1).

Case Presentation

We present the case of a 66-year-old woman with Charcot-Marie-Tooth disease who underwent anaesthesia for a 2-stage revision of total knee replacement over a 4-month period.

During preoperative evaluation, the patient presented with severe motor and sensory demyelinating polyneuropathy. Numbness and muscle weakness were present in both hands and feet, with foot drop at both sides. Muscle weakness was more severe at the lower limbs, whereas sensory impairment was more evident in the feet below the ankles. The patient had severe anatomic disorders (Figure 1a, 1b) and absence of lower extremity reflexes was observed. Genetic control was requested to determine the severity of the disease. The patient was under antihypertensive therapy and had undergone a gastric bypass surgery owing to obesity 10 years ago; at the time of the pre-anaesthetic evaluation, her body mass index was still elevated (31.1 kg/m²). Surgery was considered important for the patient at the time, as she complained of pain during movement and poor quality of life. We decided to proceed with the operation, both times, under general anaesthesia. Written informed consent was obtained the day before surgery.

Total intravenous anaesthesia was selected and administered for both procedures (2). Midazolam, propofol and fentanyl were used for inducing anaesthesia, and infusions of propofol and remifentanyl for maintenance. Neuromuscular blockade was not administered. Ventilation of the lungs, both times, was achieved using a clean ventilator, free of volatile anaesthetics, with disposable circuits. Dantrolene was available, given the risk of malignant hyperthermia (3). Monitoring included electrocardiogram, pulse oximetry, invasive blood pressure, end-tidal CO₂ and bispectral index monitoring. Paracetamol and patient-controlled analgesia with intravenous morphine were administered for postoperative analgesia.

The first operation (removal of implants of previous total knee replacement and placement of cement spacer) lasted 3.5 hours, whereas the second stage lasted 5.5 hours. At the end of the procedures, the patient was extubated, without any complications. After both surgical operations, for closer observation, the patient was transferred to the post-anaesthesia care unit (PACU) for 3 hours, without any desaturations or other adverse events. At the time of discharge from the PACU, she had no complaints of pain and declared that she was satisfied with the anaesthetic management.

Discussion

Patients with Charcot-Marie-Tooth disease represent a special patient group. The anaesthetic management is of crucial importance and must be based on patients' individual profile and type of surgery. To the best of our knowledge, the literature lacks case reports regarding this rare disease, and this is the first one regarding a revision of total knee arthroplasty. This type of surgery is challenging for the anaesthesiologist, as it is a more complex intervention than primary total knee arthroplasty and is associated with increased complications and mortality rates (4). In addition, the use

of tourniquet inflation may be associated with nerve injury and ischaemia, especially when tourniquet release occurs after prolonged inflation time. Reperfusion can only modestly decrease the nerve damage (5) Major considerations for these patients include the variable, prolonged, unpredictable response to non-depolarising neuromuscular blocking drugs (6) and the risk of malignant hyperthermia. Avoidance of medication-induced exacerbation of neuropathy is also of great importance.



Figure 1. a, b. The appearance of the patient's feet.

Main Points:

- Patients with Charcot-Marie-Tooth disease may present with a variable and unpredictable response to non-depolarising neuromuscular blocking drugs and volatile anaesthetics.
- The anaesthetic management is challenging because these patients are at a risk of developing malignant hyperthermia and cardiorespiratory complications.
- Appropriate monitoring for the patient and avoidance of medication-induced exacerbation of neuropathy are of crucial importance for major surgical procedures such as revision of total knee replacement.
- General anaesthesia with the avoidance of neuromuscular blockade and volatiles, using a clean ventilator with disposable circuits, led to successful anaesthetic management.

Conclusion

To conclude, the anaesthetic management of a patient with Charcot-Marie-Tooth disease is challenging owing to progressive neuropathy that may lead to an abnormal response to anaesthetic agents, especially volatiles and muscle relaxants, and the risk of cardiorespiratory complications. In our case, general anaesthesia with the avoidance of neuromuscular blockade and volatiles, using a clean ventilator, free of volatile anaesthetics with disposable circuits, led to successful anaesthetic management in both surgical procedures, with the patient consequently expressing satisfaction on account of absence of complications and postoperative pain.

Informed Consent: Written informed consent was obtained from the participant.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - E.S., F.K., G.E., C.B., A.K., G.K.; Design - E.S., F.K., G.E., C.B., A.K., G.K.; Supervision - C.B., G.K.; Literature Search - E.S., F.K., G.E., C.B., A.K., G.K.; Writing - E.S., F.K., G.E., C.B., A.K., G.K.; Critical Reviews - E.S., F.K., G.E., C.B., A.K., G.K.

Conflict of Interest: The authors have no conflicts of interest to declare.

Financial Disclosure: The author declared that this study has received no financial support.

References

1. Barreto LC, Oliveira FS, Nunes PS, et al. Epidemiologic study of charcot-marie-tooth disease: A systematic review. *Neuroepidemiology* 2016; 46: 157-65. [\[Crossref\]](#)
2. Gratarola A, Mameli MC, Pelosi G. Total intravenous anaesthesia in Charcot-Marie-Tooth disease. Case report. *Minerva Anesthesiol* 1998; 64: 357-60.
3. Ducart A, Adnet P, Renaud B, Riou B, Krivosic-Horber R. Malignant hyperthermia during sevoflurane administration. *Anesth Analg* 1995; 80: 609-11. [\[Crossref\]](#)
4. Japer LL, Jones CA, Mollins J, Pohar SL, Beaupre LA. Risk factors for revision of total knee arthroplasty: A scoping review. *BMC Musculoskelet Disord* 2016; 17: 182 [\[Crossref\]](#)
5. Horlocker TT, Hebl JR, Gali B, et al. Anesthetic, patient, and surgical risk factors for neurologic complications after prolonged total tourniquet time during total knee arthroplasty. *Anesth Analg* 2006; 102: 950-5. [\[Crossref\]](#)
6. Schmitt HJ, Munster T. Mivacurium-induced neuromuscular block in adult patients suffering from Charcot-Marie-Tooth disease. *Can J Anaesth* 2006; 53: 984-8. [\[Crossref\]](#)