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CASE REPORT

Intraoperative diagnosis of autonomic neuropathy in a case of Charcot–Marie–Tooth disease undergoing laparoscopic cholecystectomy: A case report

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INTRODUCTION

Anaesthetic management is often challenging to the attending anaesthesiologist when a rare disease is encountered. Charcot-Marie-Tooth (CMT) is one such hereditary disorder that affects the peripheral nervous system. CMT disorders are relatively uncommon, affecting about 1 in 2500 individuals.[1] It can manifest in various genetic mutations that alter the peripheral nerves. Symptoms typically involve progressive muscle weakness and atrophy with a distally pronounced sensory dysfunction. Although there may be an autonomic component, it is not always immediately apparent from a clinical perspective.

CASE DESCRIPTION AND MANAGEMENT

A 21-year-old woman weighing 83 kg was scheduled for laparoscopic cholecystectomy. She had walking difficulties since age six and underwent reconstructive surgery on her left ankle at age 12, but her condition deteriorated. By 18, she developed muscle weakness in her hands and became progressively bedbound. A neurological examination revealed intact sensory function in her limbs. However, the motor examination using the Bromage scale showed full motor power (5/5) in proximal muscles, while distal muscles had substantial reduced power (1/5) **(FIGURE 1)**. There were no signs of autonomic neuropathy during the preanesthetic visit .

LEARNING POINTS

1. Encountering a rare disease can make anaesthetic management challenging for anaesthesiologists.
2. Charcot-Marie-Tooth (CMT) disease is a hereditary disease that primarily affects the motor and sensory nerves but can also affect the autonomic nervous system.
3. It is crucial to assess the patient for preoperative autonomic neuropathy.
4. Patients with autonomic neuropathy may respond unpredictably to anaesthetic agents and vasopressors, necessitating careful titration.

The nerve conduction study of the patient showed unexcitable motor and sensory nerves, and an electromyogram revealed neurogenic motor unit action potentials in some upper limb muscles. Based on clinical presentation and electrophysiological studies, it is likely that the patient has peripheral neuropathy, specifically hereditary motor sensory neuropathy (HMSN-CMT) type. Whole exome sequencing (WES) wasn't possible due to unavailability and cost. Clinical presentation and radiological findings suggest that the patient may have HMSN-type-2, a variant of CMT.

The patient, with a baseline blood pressure of 130/80 mmHg, heart rate of 120 beats per minute, and SpO2 of 98%, was given 100 mg of propofol and 100 micrograms of fentanyl for anaesthesia induction, followed by atracurium for intubation. After the pneumoperitoneum was established, the blood pressure dropped sharply to 42/20 mmHg. The administration of 10 mg of

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ephedrine caused the blood pressure to spike to 168/128 mmHg, indicating significant instability. Throughout the procedure, the patient's blood pressure remained unstable, and she remained tachycardic.

Initially, anaesthesia was maintained with isoflurane in a 70% N2O and 30% O2 mix, but oxygen saturation levels dropped to 92-94%. The endotracheal tube position was checked, with no signs of bronchospasm or lung secretions. After turning off nitrous oxide, the SpO2 level returned to 100%. The air and oxygen mix was used for the rest of the procedure. Thirty minutes after the muscle relaxant was given, the patient could breathe spontaneously, and the recovery process was uneventful.

a b

FIGURE 1 (a) muscle wasting of upper limbs; (b) left foot deformity and muscle wasting of both lower limbs.

DISCUSSION

After administering anaesthesia, a slight drop in blood pressure is common due to the vasodilation caused by propofol. However, this patient experienced a severe and unexpected drop in blood pressure. While pneumoperitoneum can cause hypotension, it was ruled out by monitoring the pressure during the procedure. Usually, bradycardia and hypotension indicate high CO2 insufflation pressure, but this patient remained tachycardic throughout. The suspected cause was autonomic neuropathy, which affects the sympathetic and parasympathetic fibres of the autonomic nervous system.[2] A study of 17 adult patients with CMT disease found that six of them had signs of autonomic neuropathy, indicating this may be the underlying issue.[3] Response to anaesthetic agents, as well as vasopressors, is very unpredictable in patients with autonomic neuropathy, which is very obvious in our case.[4]

During laparoscopic surgery, hyperventilation helps eliminate CO2 from the patient's body. However, for healthy individuals, this may decrease blood pressure due to the impact of hypocapnia on peripheral vasculature. Patients with autonomic neuropathy may experience an even greater drop in blood pressure, as reflex vasoconstriction may not be effective in these cases.

Patients with CMT may have subclinical respiratory dysfunction resulting from axonal degeneration of intercostal and phrenic nerves.[5] In restrictive lung disease, a small airway collapses during tidal ventilation, resulting in shunting and hypoxaemia.[6] This issue is further exacerbated in the presence of pneumoperitoneum, necessitating the use of high oxygen concentrations.

Patients with autonomic neuropathy are at a higher risk during anaesthesia because anaesthetic drugs cause vasodilation. It is important to conduct a thorough evaluation of autonomic neuropathy before surgery. Patients with autonomic neuropathy may respond unpredictably to anaesthetic agents and vasopressors, so careful titration is required.

In conclusion, it is suggested to consider the possibility of subclinical autonomic neuropathy and associated abnormal vascular response in CMT disease before planning anaesthesia, to manage the situation proactively.

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Author contributions

Manuscript drafting and revising it critically: KMA, CSK. *Approval of the final version of the manuscript:* KMA, CSK, AKMA. *Guarantor of accuracy and integrity of the work:* AKMA.

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Conflict of interest

We do not have any conflict of interest.

Ethical approval

Ethical approval was not sought because this is a case report. However, informed written consent was obtained from the patient for preparation of this manuscript and publishing her pictures.

Data availability statement

The data that support the findings of this study are available on request from the corresponding author.

REFERENCES

1. Hines RL, Jones SB, editors. Stoeltings Anesthesia and Co-existing Disease E-book. 7th ed. Philadelphia, PA: Elsevier; 2021.
2. Stamboulis E, Katsaros N, Koutsis G, Iakovidou H, Giannakopoulou A, Simintzi I. Clinical and subclinical autonomic dysfunction in chronic inflammatory demyelinating polyradiculoneuropathy. Muscle Nerve. 2006 Jan;33(1):78-84. DOI: https://doi.org/10.1002/mus.20438.
3. Brooks AP. Abnormal vascular reflexes in Charcot-Marie-Tooth disease. J Neurol Neurosurg Psychiatry. 1980 Apr;43 (4):348-350. DOI: https://doi.org/10.1136/jnnp.43.4.348.
4. McGrane S, Atria NP, Barwise JA. Perioperative implications of the patient with autonomic dysfunction. Curr Opin Anaesthesiol. 2014 Jun;27(3):365-370. DOI: https://doi.org/10.1097/ACO.0000000000000072.
5. de Carvalho Alcantara M, Nogueira-Barbosa MH, Fernandes RM, da Silva GA, Lourenço CM, Sander HH, Marques Junior W. Respiratory dysfunction in Charcot-Marie-Tooth disease type 1A. J Neurol. 2015 May;262(5):1164-1171. DOI: https://doi.org/10.1007/s00415-015-7677-8.
6. Wang M, Argalious MY. Smith and Aitkenheads Textbook of Anaesthesia. 7th ed. Philadelphia, PA: Elsevier; 2020.