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Case report
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Neuraxial anaesthesia in a patient with Charcot-Marie-Tooth disease for Caesarean delivery: a case report

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Abstract

Charcot-Marie-Tooth (CMT) disease is a hereditary peripheral neuropathy characterised by slowly progressive, distal-to-proximal muscle weakness. The anaesthetic concerns are the associated respiratory and diaphragmatic muscle weakness, phrenic nerve palsy, and increased sensitivity to anaesthetic agents. Our case report described a patient with underlying CMT disease who had undergone elective Caesarean delivery (CD) under combined spinal-epidural (CSE) anaesthesia. She had underlying bronchial asthma with recent exacerbation secondary to pneumonia at 25 weeks' gestation. Her lung function test showed a moderately restrictive pattern. We decided to perform CSE using an intrathecal dose of heavy bupivacaine 8.5 mg and fentanyl 15 mcg under pre-procedural ultrasound guidance. The surgery was uneventful, and postoperative analgesia was maintained with an epidural infusion of ropivacaine 0.1% combined with fentanyl 2 mcg/mL running at 5 mL/hour. The patient's neurological status returned to baseline on postoperative day one. Our case showed the successful use of CSE for elective CD in the patient with CMT disease without worsening neurological symptoms. With a thorough preoperative assessment of neurological status, utilisation of ultrasound guidance and minimum effective local anaesthetic dosage, neuraxial anaesthesia can be safely performed in patients with CMT disease.

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Introduction

Charcot-Marie-Tooth (CMT) disease is a group of inherited neurological disorders comprising motor and sensory neuropathy that can be classified into distinct types based on the associated gene mutations. Those gene mutations can either result in nerve demyelination (occurring in *CMT1*, *CMT3*, and *CMT4* type) or direct axonal damage (as in *CMT2* type). The symptoms consist of tingling, numbness, distal muscle weakness, atrophy and foot deformities that can worsen progressively over time.[1] The anaesthetic management can be challenging due to its association with phrenic nerve palsy, reduced lung capacity due to diaphragmatic weakness and increased sensitivity to anaesthetic agents and muscle relaxants.[2] Neuraxial anaesthesia can mitigate the risk of pulmonary complications, but there are not many case reports describing its use in patients with CMT disease.[3,4]

Due to the low prevalence of this disease, we share our anaesthetic management in this case report describing a patient with underlying CMT disease who underwent elective Caesarean delivery (CD) under combined spinal-epidural anaesthesia (CSE).

Case presentation

A 34-year-old woman of gravida two para one with underlying CMT disease and bronchial asthma was scheduled for CD at 37 weeks' gestation. She was obese, with a body mass index of 37 (height 150 cm, weight 84 kg). She had been diagnosed with CMT disease at the age of 33 years, despite having developed lower limb weakness since the age of four years. She had an uneventful spontaneous vaginal delivery for her first pregnancy. During her current pregnancy, she was admitted to the intensive care unit with pneumonia at 25 weeks' gestation. Her echocardiography results were normal, but lung function test showed a restrictive pattern (Table 1), evidenced by her forced vital capacity (FVC) being only 45% of the predicted value. In view of her reduced respiratory reserve due to her medical condition, CD was planned for her to reduce the physiological demands of labour.

Upon assessment in the ward, she was well under room air. The muscle power was four-fifths in her lower limbs, whereas her upper limbs strength was full.

Table 1. Spirometry result of the patient

Parameters	Predicted	Best	% Predicted
FEV_1 (litre)	2.35	1.07	46
FVC (litre)	2.70	1.22	45
FEV_1/FVC (%)	87.04	88.12	101
PEF (litre/second)	6.12	3.15	52
MMEF (litre/second)	2.93	1.49	51

FEV_1: forced expiratory volume in one second, FVC: forced vital capacity, PEF: peak expiratory flow, MMEF: maximal mid-expiratory flow

Fig. 1. (Left) Hand and *(right)* feet deformities of the patient.

Sensation was otherwise intact. There were deformities on both her feet and hands (Fig. 1). Other systemic examinations were unremarkable. No obvious spine deformities were detected during the examination.

To reduce the risk of postoperative pulmonary complications related to general anaesthesia (GA), CSE was planned for her CD. She had received CSE when undergoing plating for an uneventful left femur fracture in 2023. A pre-procedural

ultrasound scan of the lumbar spine was performed on the day of surgery. Based on the spinal sonography technique described by Karmakar,[5] a low-frequency, curved-array probe was used to visualise the paramedian sagittal oblique view of the vertebral laminae and interlaminar spaces at the lower back starting from the sacrum. By sliding the transducer cranially and counting interlaminar spaces upwards, the L3/L4 interspace was located. A transverse midline view was then obtained to mark the spinous process for the midline approach insertion. Visualisation was achieved without much difficulty, and CSE was performed in a single attempt with intrathecal administration of bupivacaine 8.5 mg and fentanyl 15 mcg. The sensory block was achieved up to T6 level within a comparable time of onset to the usual obstetric population, without any patchy block. The operation was uneventful, and no epidural top-up was required. The postoperative recovery room assessment demonstrated a Bromage score of 1 before the patient was discharged to the ward with an epidural infusion of 0.1% ropivacaine combined with fentanyl 2 mcg/ml at 5 ml/hour.

During the follow-up by our acute pain service team, the patient was well, with motor and sensory function of her lower limbs returning to baseline on postoperative day one while still receiving epidural analgesia. In view of the patient's consistently low pain score, we decided to discontinue the epidural infusion and maintain analgesia with oral paracetamol 1 g QID and diclofenac 50 mg TDS.

Discussion

CMT disease is uncommonly encountered in anaesthetic practice. Its prevalence is heterogenous worldwide, affecting 1 in 125,000 individuals in the United States, whilst Japan reports 10.8 cases per 100,000 individuals. European countries have an even higher prevalence, with 28.2 cases per 100,000 in the Spanish population. CMT disease is diagnosed clinically and confirmed with investigations such as electromyography, nerve conduction studies and/or genetic testing.[1] In our case, the patient's nerve conduction study revealed severe bilateral sensorimotor axonal polyneuropathy, but genetic testing was not done as the patient did not consent.

The mode of delivery for pregnant women with CMT disease does not differ significantly from the normal population.[6] The decision should be individualised, with multidisciplinary planning based on the patient's neurological status, cardiorespiratory reserve, and obstetric indications. In our case, the patient had a moderately restrictive pattern in her lung function test and had history of asthma exacerbation secondary to pneumonia at 25 weeks' gestation. Despite having an uneventful spontaneous vaginal delivery for her previous pregnancy, it was decided that CD would be safer instead of subjecting her to the risk of hypoxaemia from the

increased physiological demands of labour.

When considering anaesthesia in patients with CMT disease, both GA and regional anaesthesia (RA) present distinct concerns. Generally, neuraxial anaesthesia is always the preferred anaesthetic technique for CD due to its excellent postoperative pain control allowing early mobilisation. Often, there are concerns whether RA can potentially worsen the nerve injuries in patients with CMT disease due to the "double crush phenomenon".[6] Nevertheless, numerous case reports and cohort studies support the safety of RA in this group of patients, be it peripheral nerve block or neuraxial anaesthesia.[2,4,6] One case report demonstrated the successful use of CSE for CD in a patient with CMT disease,[3] while another highlighted effective epidural analgesia during labour.[7] A retrospective study in the United Kingdom involving 87 patients with CMT disease who received spinal or epidural anaesthesia reported complications in only 5% of the cases, including prolonged anaesthetic effect and technical difficulty in a scoliotic patient.[8] Our clinical case further supports the safety of neuraxial anaesthesia patients with CMT disease without worsening neurological deficit or prolonging the anaesthetic effect.

Administering spinal or epidural anaesthesia can be particularly challenging due to the presence of scoliosis, which increases the technical difficulty.[8] In order to minimise the risk of nerve injuries, it is crucial to employ strategies such as establishing baseline neurological status during the preoperative assessment, risk disclosure to the patients during counselling, avoidance of high-dose local anaesthetics, and utilisation of ultrasound guidance for neuraxial anaesthesia.[5] In our case, although the patient had no spinal deformities, ultrasound guidance was used to identify the L3/L4 interspace, thereby minimising the risk of traumatic attempts. Besides, the bupivacaine dose administered was lower than the advocated effective doses in 95% of obstetric patients, yet slightly higher than the effective dose in 50% for CD,[9] with the hope of minimising the "double-crush phenomenon" risk, particularly given the patient's prior exposure to neuraxial anaesthesia.

On the other hand, there are undesirable implications of GA for such patients. The upregulation of acetylcholine receptors in chronically immobile patients can cause increased sensitivity and potentially prolonged muscle relaxant effect. In addition, there are also cardiorespiratory concerns, namely, postoperative pulmonary complications due to respiratory muscle weakness, autonomic dysfunction, and cardiac dysrhythmias, such as prolonged QT interval.[2,6] A case report has also described GA-induced delayed recovery of spontaneous breathing in the patient affected with axonal CMT type two.[10] These specific concerns, together with other GA-related risks in CD (namely aspiration, difficult intubation, postoperative nausea, and vomiting) render GA a considerably less favourable anaesthetic technique in this context.

Conclusion

Our case has demonstrated the successful use of CSE for CD in the patient with CMT disease without worsening neurological symptoms. With a thorough preoperative assessment of neurological status, utilisation of ultrasound guidance, and the minimum effective local anaesthetic dosage, neuraxial anaesthesia can be safely performed in patients with CMT disease. After all, it is still the more favourable anaesthetic technique as its benefits outweigh the potential risks associated with GA in this setting.

Declarations

Informed consent for publication
The patient provided written informed consent for the use of the clinical data and images contained in this case report.

Competing interests
None to declare.

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References

1. Divakara K. Charcot-Marie-Tooth disease [Internet]. [place unknown]: Medscape; [updated 2023 February 6; cited 2025 Aug 4]. Available from: https://emedicine.medscape.com/article/1232386-overview
2. Zanjani AP, Ghorbani A, Eslami B, Mirzashahi B. Epidural Anesthesia Combined with Light General Anesthesia for a Juvenile with Charcot-Marie-Tooth Disease Undergoing Corrective Spine Surgery: A Case Report. Anesth Pain Med. 2017;7:e14189. https://doi.org/10.5812/aapm.14189
3. Brock M, Guinn C, Jones M. Anesthetic management of an obstetric patient with Charcot-Marie-Tooth disease: a case study. AANA J. 2009; 77: 335-337.
4. Rodríguez-Ortiz E, Martínez E, Martín J, Maiza L, Medina J. Spinal anesthesia in a patient with Charcot-Marie-Tooth disease undergoing orthopedic surgery: case report. Colomb J Anesthesiol. 2019;47:180-183. https://doi.org/10.1097/CJ9.0000000000000115

5. New York School of Regional Anesthesia [Internet]. Spinal sonography and application of ultrasound for central neuraxial blocks; cited [2026 Apr 9]. Available from https://www.nysora.com/techniques/neuraxial-and-perineuraxial-techniques/spinal-sonography-and-applications-of-ultrasound-for-central-neuraxial-blocks/
6. McClain RL, Rubin DI, Bais KS, Navarro AM, Robards CB, Porter SB. Regional anesthesia in patients with Charcot-Marie-Tooth disease: a historical cohort study of 53 patients. Anesthésie régionale chez les patients atteints de la maladie de Charcot-Marie-Tooth : une étude de cohorte historique de 53 patients. Can J Anaesth. 2022;69:880-884. https://doi.org/10.1007/s12630-022-02258-5
7. Roriz D, Goncalves D, Brandao J, et al. Charcot-Marie-Tooth And Analgesia for Labor- Case Report. Reg Anesth & Pain Med. 2019; 44(Suppl 1)A143. https://doi.org/10.1136/rapm-2019-ESRAABS2019.203
8. Skorupinska M, Ramdharry G, Byrne B, Laurá M, Reilly MM. Pregnancy and delivery in patients with Charcot-Marie-Tooth disease and related disorders. Obstet Med. 2023;16:83-87. https://doi.org/10.1177/1753495X221107328
9. Eiko O, Mamoru M, Keiji H, Miho K. Optimal intrathecal hyperbaric bupivacaine dose with opioids for cesarean delivery: a prospective double-blinded randomized trial. IJOA. 2017;31:P68-73. https://doi.org/10.1016/j.ijoa.2017.04.001
10. Vinci P, Lapi G. Anesthetic management in Charcot-Marie-Tooth disease type 2 due to a mutation in the mitofusin-2 gene. J Anaesthesiol Clin Pharmacol. 2011;27:286-287. https://doi.org/10.4103/0970-9185.81845