Anaesthetic implications in patient with Charcot-Marie-Tooth disease

Sir,

Charcot-Marie-Tooth disease (CMT) is a hereditary sensory and motor neuropathy with an incidence of 1 in 2,500.[1] It is the most common inherited neuropathy starting in the lower limbs and subsequently involving hands and forearms.[2] Patients often have pes cavus, hammer toes, clawed hands, loss of deep tendon reflexes, and later on scoliosis and involvement of respiratory muscles. 'A 24-year-old male, a known case of CMT since 14 years presented for renal calyceal stone removal. Nerve conduction studies had revealed generalised axonal sensorimotor peripheral neuropathy. Currently, he had wasting and weakness of small muscles of the hands with bilateral claw hands. Deep tendon reflexes were absent. His lower limb motor power was 0/5 in both limbs and he was bed ridden. There were no complaints of dysphagia or breathing difficulty.Chest X-ray showed significant scoliosis. He had no other comorbidities and his blood investigations were within normal limits. Electrocardiogram and 2D echocardiography were normal. The pulmonary function test reported a normal Forced expiratory volume in 1 second (FEV1)/Forced vital capacity(FVC) ratio excluding any significant obstructive defect. However, FVC was reduced (55% of predicted value) suggesting moderate restrictive defect. Currently, he was not on any steroids or analgesics, only regular physiotherapy.

In the operation theatre, standard monitors including non-invasive blood pressure, arterial oxygen saturation, and five lead electrocardiogram were attached. Bispectral index (BIS) was monitored throughout and was maintained in the range of 40-60.

Anaesthetic agents used for induction and maintenance were midazolam, fentanyl, and propofol in graded aliquots and dexmedetomidine infusion was started at 0.5 μg/kg/h. Atracurium was used for muscle relaxation followed by intubation. As peripheral nerve stimulator for train-of-four monitoring would be ineffective in this patient, visual notching of capnograph was used as an indicator for repeating dose of neuromuscular blocking agent. Intraoperatively, the vital parameters were stable and serum potassium level was checked and reported as 4.12 mmol/L. At the end of the surgery, on return of spontaneous respiration, the neuromuscular blockade was reversed with neostigmine. The patient

demonstrated all clinical signs of adequate reversal and had an uneventful recovery.

Patients with CMT undergoing surgery are usually prediagnosed by a neurologist and under regular follow-up. Analgesics for neuropathic pain and pain related to skeletal deformities and steroids for immunosuppression form the mainstay of drug therapy. Increased incidence of mitral valve prolapse, prolonged QT interval, AV block, paroxysmal atrial flutter, and other cardiac dysrhythmias have been noted in these patients.[2] During anaesthesia, drugs enhancing conduction disturbances are avoided. Chronic denervation is a predisposing factor for hyperkalemia, however this has not been frequently observed probably because the denervation process is slow.[3] Preoperatively, potassium levels should be checked. Use of succinylcholine is controversial because of expected rise in potassium levels hence we did not use it. Similarly, use of inhalational agents is also unclear because of their role in triggering malignant hyperthermia.[1] Since this disease is a peripheral neuropathy and not myopathy, the fear of malignant hyperthermia seems to be unfounded. Inhalational agents are also known to prolong muscle relaxation. Atrophy of muscles can lead to upregulation of acetylcholine receptors at neuromuscular junction which can lead to prolonged effect of neuromuscular blocking agents. Proximal muscle weakness of the arms has been used to predict respiratory muscle weakness.[2] Weakness of intercostal muscles and diaphragm can further prolong the effect of neuromuscular blocking agents. Similar to other reports, prolongation in the duration of neuromuscular blockade was not seen.[2] We continuously monitored depth of anaesthesia using BIS to avoid awareness during anaesthesia.[4] Dexmedetomidine is used for intensive care unit (ICU) sedation, procedural sedation, and has an anaesthetic sparing effect.[5] It is known to reduce the requirement of concomitantly administered intravenous or inhalational anaesthetics but cannot replace them. We used dexmedetomidine infusion with fentanyl and propofol boluses and maintained BIS below 60. Neuromuscular monitoring using facial nerve has been suggested in these patients. Theoretically, nitrous oxide can produce neurotoxicity, hence is avoided in these patients. Regional anaesthesia in patients with neurological disorders is controversial. Ultrasound-guided nerve blocks are gaining popularity as they have lower risk of neurological complications.

Proper assessment of neurological deficits and titration of anesthesia drugs is essential for smooth conduct of anesthesia in patients of CMT.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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Submitted: 13-Feb-2021
Revised: 13-Mar-2021
Accepted: 25-Mar-2021
Published: 23-Jul-2021

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Access this article online	
Quick response code	Website: www.ijaweb.org
	DOI: 10.4103/ija.IJA_135_21

How to cite this article: Kapoor H, Shah M, Desai S. Anaesthetic implications in patient with Charcot-Marie-Tooth disease. Indian J Anaesth 2021;65:561-2.