ANESTHESIA FOR CHARCOT-MARIE-TOOTH DISEASE: CASE REPORT

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Charcot-Marie-Tooth disease comprises a group of disorders characterized by progressive muscle weakness and wasting. Reviewing the anaesthetic literature produced conflicting reports about the best anaesthetic options for patients with CMTD; as they are at increased risk of prolonged response to muscle relaxants, malignant hyperthermia and risks of regional anaesthesia. We present a case of the successful use of total intravenous anaesthesia with dexmedetomidine and propofol combined with caudal block using bupivacaine mixed with dexmedetomidine without any complications, for a 17 year old male patient with Charcot Marie-Tooth disease who underwent a lower limb orthopedic surgery.

Introduction

Charcot-Marie-Tooth disease (CMTD) is a spectrum of various subtypes of inherited peripheral neuropathies. The disease affects approximately 1 in 2500 people and is caused by mutations in more than 30 genes which are necessary for myelin structure, maintenance, and formation. Most patients have a “classical” CMTD phenotype characterized by onset in the first two decades of life, distal weakness, sensory loss, foot deformities, and absent ankle reflexes. Life expectancy is not shortened and the treatment of the disease is mainly supportive. Particularly, orthopedic procedures are often required to treat foot deformities by the time of adolescence.

Due to nerve-damaging nature of CMTD, anesthesiologists fear possible exacerbation of the disease by different classes of drugs that act on these nerves in regional anesthesia. Moreover, some concerns have been expressed regarding the use of muscle relaxants in these patients. Although the issue of increased risk of malignant hyperthermia (MH) has been raised in these patients, most patients received MH triggering agents without untoward effects. However, available information does not exclude a potential link.

One way of minimizing the risks of anesthesia in these patients is by using total intravenous anaesthesia (TIVA). This can be done by using dexmedetomidine, a potent highly selective and specific α2-adrenoceptor agonist that has both sedative and analgesic effects which has been shown to consistently reduce the requirements of other anesthetic drugs.

We present a case report of a 17 year old CMTD patient who underwent an uneventful general anesthesia for a lower limb orthopedic surgery with total intravenous anesthesia using propofol and dexmedetomidine combined with caudal block using bupivacaine mixed with dexmedetomidine without any complications.

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Case presentation

A 17-year old male patient (height = 135 cm weight = 19kg) was admitted for an orthopedic surgery. He is a known case of CMTD for 13 years. The diagnosis was made clinically, and confirmed by nerve conduction study and genetic studies. His physical examination reveals high arched left foot, distal muscle wasting, knee and ankle jerks were absent and sensation was decreased just below the knees. In the last month he developed difficulty in walking and he could not walk without assistance. He was planned for a posterior tibial tendon transfer, planter fascia release, gastrocnemius release and first ray dorsiflexion osteotomy. Preoperatively, the patient was fasting for eight hours, his American Society of Anesthesiologists (ASA) classification was 2, and his cardio-respiratory assessment was normal with good air entry bilaterally.

On the day of the operation, Soda lime, filters and circuit were changed, the anesthetic machine was washed with Oxygen 10 liter/minute for 15 minutes to eliminate any residual gas. Anesthesia plan was TIVA and Caudal block.

The patient was admitted to the operating room, and his blood pressure: 92/50 mm Hg, Pulse rate of 98 beats/minute, O2 saturation of 95% on room air. Intravenous (IV) access was secured and pre-oxygenation was done. Intravenous dexmedetomidine (Precedex®; Hospira, Lake Forest, Illinois, USA) of 20 mcg was administered over 15 minutes then propofol (60 mg) was given over two minutes. Ventilation was assessed, and then cisatracurium 0.1 mg/kg was given. Intubation was successful with oral endotracheal tube (size 5.5) from the first attempt without difficulties and the patient was mechanically ventilated. Anesthesia was maintained with propofol 5-10 mg/kg/hr and dexmedetomidine 0.2-0.5 mcg/kg/hr. Patient was placed in left lateral position and caudal block was done with 16 ml bupivacaine 0.25% mixed with preservative-free dexmedetomidine (20 mcg). Surgery lasted 140 minutes, through which there was no hypotension or bradycardia, and no muscle relaxant or opioids boluses were needed intraoperatively. At the end of the operation, patient was extubated and transferred to the recovery room and observed for two hours with uneventful course. Patient required first analgesia 12 hour after the operation. During a follow-up inquiry three months after surgery, the patient did not report any worsening of the underlying disease.

Discussion

Sporadic reports on the anesthetic management of CMTD have appeared in the literature, yet the main problem is the lack of controlled studies in these patients to evaluate potential risks of anesthesia. Up to this day, there is still a debate about the proper anesthetic care for surgical procedures for CMTD patients. In general, anesthetic care was adjusted to fit the individual needs of each patient.6-10

Therefore, due to the lack of enough evidence to exclude the risks of anesthetic drugs, the best way to deal with these patients is by minimizing the use of anesthetic drugs that have potential risks in these patients even though some reviews state that their use was safe and without untoward effects.4

CMTD patients have a chronic denervation, often in all extremities.11 Since denervation is a potent predisposing factor for potassium release after exposure to succinylcholine (SCH)12, it is cautioned to use SCH in these patients. Moreover, the response to non-depolarizing neuromuscular blocking drugs is variable in patients with CMTD and their effects may be prolonged.13-15 In this context, we used a single dose of cisatracurium (0.1 mg/kg) to facilitate the intubation.

Association between MH and the use of inhalational anesthesia in CMTD remains unclear. Despite the fact that sevoflurane is able to trigger malignant hyperthermia in susceptible patients,16 the review of 86 cases indicates that most patients received MH triggering agents without untoward effects. Moreover, according to The Charcot-Marie-Tooth Disease North American database, nitrous oxide is considered in the moderate to significant risk group of drugs for CMTD patients.17 For that, no inhalational agent was used and the anesthesia machine was flushed with oxygen for 15 minutes before the surgery in order to eliminate any gas residues.

Patients with an underlying peripheral neurological disorder may be more susceptible to
nerve injury with the use of regional techniques for anesthesia\textsuperscript{18}, and the presence of chronic underlying neural compromise, may theoretically, place these patients at increased risk of further neurological injury\textsuperscript{19}. In order to alleviate the effects of regional anesthetic drugs in our case, we combined dexmedetomidine (20 mcg) with bupivacaine (40 mg) in the caudal block. This combination was found to prolong the duration of analgesia and decrease the amount of anesthetic drugs needed\textsuperscript{20,21}.

Dexmedetomidine is known for its remarkable pharmacological properties including sedation, anxiolysis, and analgesia with the unique characteristic to cause no respiratory depression. In addition, it possesses sympatholytic and antinociceptive effects that allow hemodynamic stability during surgical stimulation. It has shown to consistently reduce opioids, propofol, and benzodiazepines requirements\textsuperscript{5,22}.

CMTD has been reported to cause cardiac dysrhythmias and conduction disturbances, and is associated with increased incidence of mitral valve prolapse\textsuperscript{23,24}. Although bradycardia and hypotension are considered to be the most prominent adverse effects of \(\alpha_2\)-adrenoreceptor agonists, these side effects appear to be less pronounced when dexmedetomidine is given slowly over 15 minutes.

CMTD patients may have an increased sensitivity to thiopental, and the dose required is strongly related to the severity of the motor and sensory disturbance\textsuperscript{25}. However, propofol, as part of the total intravenous technique, is thought to be safe and effective\textsuperscript{26,27}. Therefore, we combined dexmedetomidine and propofol in TIVA to achieve the safest results in our case.

**Conclusion**

In conclusion, this case demonstrates the safety and effectiveness of the combined use of caudal dexmedetomidine and bupivacaine with intravenous dexmedetomidine and propofol for the anesthetic management of a patient with CMTD.
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**REFERENCES**
1. BRIDION Summary of Product Characteristics (SPC)

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