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Case Report

Total Intravenous Anesthesia with Propofol and Remifentanil for Adult Patient with Charcot-Marie-Tooth Disease

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Abstract

Charcot-Marie-Tooth disease (CMTd) is the most common form of peripheral neuropathy characterized by progressive distal muscle weakness and atrophy. Although life expectancy is normal, quality of life is lessened due to deformity, immobility or chronic pain. Limited safety data for the kind of anesthesia to be administered in such patients has caused an argumentative concern for the anesthesia provider. The purpose of this case report is to emphasize the prospective edge of total intravenous anesthesia (TIVA) using propofol and remifentanil in CMTd patient without getting into litigious or controversial methods of administering drugs, gases or regional techniques.

ABBREVIATIONS

CMTd: Charcot-Marie-Tooth Disease; TIVA: Total Intravenous Anesthesia

INTRODUCTION

Charcot-Marie-Tooth disease (CMTd) is considered as the most common type of inherited peripheral neuropathy, occurring as 1 in 2500 people in United States to 1 in 10,000 people worldwide [1,2]. Early manifestation is unusual but it may prevail between the first and third decade [2]. CMTd is slowly progressive, with spells of alleviation and exaggerations of clinical symptoms. No sexual or racial predisposition is seen in CMTd. Life expectancy is normal. However, quality of life suffers depending upon the degree of disability (muscle power in lower limbs), ageing and the disease stage [3].

Patient with CMTd is presented with peroneal muscle atrophy, distal limb muscle weakness, skeletal deformities including pes cavus and decreased or absence of tendon reflexes are the most common clinical manifestations [3]. The lower limb is exemplary portrayed as having "inverted champagne bottle" or "stork leg" appearance [2]. Due to foot drops, patients repeatedly lapse over objects and have a predilection to sprain their ankles.

Inheritance patterns and genetic studies forms the basis of categorization of CMTd [3,4]. PMP22 or MPZ genes encodes for peripheral myelin protein, which are essential for the normal function of peripheral nerves. Mutations in these encoding genes lead to demyelination. Type 1 affects the myelin sheath (PMP 22 gene) in 60-80% of cases, however type 2 which alter the axons (MPZ gene) occur in only 5-15% of cases. Inheritance can be autosomal dominant, autosomal recessive or X-linked [3,4].

A major debate appeared in the literature about the safest anesthesia technique for patients with CMTd [5-12]. Concerns have been raised about using muscle relaxants [5,6,10,11], inhalational drugs [12], and regional techniques due to the existence of neurological defects. Therefore, in this case total intravenous anesthesia has been used.

Written patient informed consent and Institutional ethical committee approval obtained.

CASE PRESENTATION

A 42-year-old male weighing 71 kg, height 165 cm, with Charcot-Marie-Tooth disease (CMTd) of unknown type visited our anesthesia clinic scheduled for an open reduction internal fixation of the left distal radial fracture. Patient was diagnosed with this condition during 2nd decade of his life when he started having weakness in the lower limb muscles followed by atrophy years later (Figure 1). Patient was wheelchair dependent as he cannot walk because of lower limbs deformity and muscle wasting. There was no inheritance pattern associated and genetic test was undocumented. His neurological examination revealed bilateral facial weakness, small muscle wasting of both hands and bilateral foot drops in association with mild inversion and high arched foot along with marked wasting of lower limb muscles. The details of his last anesthetic history was unavailable. He was a non-smoker and rest of his antecedent medical history was insignificant.

On systemic preoperative assessment, spirometry revealed a moderate restrictive pattern with FVC of about 63.3% of the predicted value (Figure 2).

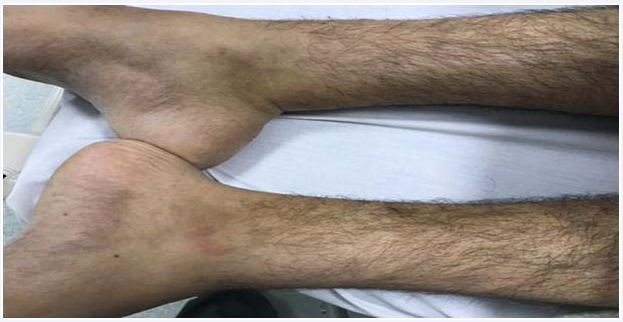
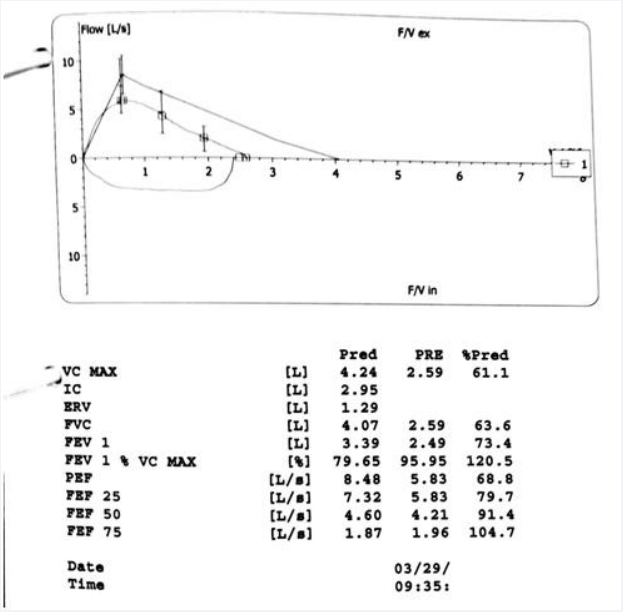


Figure 1 Bilateral foot drops, high arch foots and wasting of lower limb muscles.



		Pred	PRE	%Pred
VC MAX	[L]	4.24	2.59	61.1
IC	[L]	2.95		
ERV	[L]	1.29		
FVC	[L]	4.07	2.59	63.6
FEV 1	[L]	3.39	2.49	73.4
FEV 1 % VC MAX	[%]	79.65	95.95	120.5
PEF	[L/s]	8.48	5.83	68.8
FEF 25	[L/s]	7.32	5.83	79.7
FEF 50	[L/s]	4.60	4.21	91.4
FEF 75	[L/s]	1.87	1.96	104.7
Date			03/29/	
Time			09:35:	

Figure 2 Respiratory functions tests shows mild restrictive pattern.

Considering cardiac involvement with age in CMTd, echocardiography and Holter monitoring were performed. Echocardiography indicated no structural and functional involvement of myocardium, however ambulatory analysis displayed frequent supraventricular ectopic activities.

Airway examination was optimum in this patient. In view of the nature of the disease patient opted for total intravenous anesthesia (TIVA). Midazolam 2 mg IV was administered for anxiolysis as premedication in receiving area. Patient shifted inside the operating room and standard monitoring was attached. Anesthesia was induced by fentanyl 50 mcg, ketamine 25 mg and propofol 100 mg. To suppress the laryngoscopic response, a supplemental dose of 100 mcg remifentanil was administered and oropharynx was sprayed with 8% lidocaine prior to intubation. After intubation and fixation of a cuffed endotracheal tube sized 7.0 mm, volume control mode of ventilation was started and anesthesia was maintained with propofol infusion at the rate of 50-100 mcg/kg/h and remifentanil infusion at the rate of 0.6-1.0 mcg/kg/h with 100% oxygen. Bispectral index (BIS) was maintained throughout the procedure between 40-60. One-gram paracetamol was infused before the end of the procedure. Neuromuscular blockers were not used for this surgery. The surgery continued for 2 hours and the patient was extubated and observed in post anesthesia care unit for an hour. Post-operative pain control was achieved with the help of paracetamol, tramadol and fentanyl *pro re nata*. Patient was discharged on post-operative day 4 and was contented with the procedure.

DISCUSSION

There is lack of adequate data pertaining to determine the most appropriate kind of anesthesia for patients with Charcot-Marie-Tooth disease (CMTd). The main concerns to anesthesiologists are unforeseeable effect to muscle relaxants, inhalational agents, nitrous oxide, cardiac arrhythmias, postoperative respiratory complications and exaggeration of neurological manifestations. In spite of the conjectural and circumscribed evidence, issues about anesthetizing patients with CMTd essentially reflect on the use of muscle relaxants. While some have suggested a hypothetical risk of developing hyperkalemia and rhabdomylysis with use of succinylcholine in patients with the neuro-myopathic conditions [5,6], in contrast a retrograde review of 86 cases by Antognini, succinylcholine was utilized for 41 patients without adverse side effects [7]. The author specified that yet no clinical consequence was illustrated, serum potassium levels were not quantified and hence the safety cannot be substantiated. Similarly, no adverse outcome has been reported by Greenberg and Parker following the use of succinylcholine [8]. A case series using non-depolarizing muscle relaxant mivacurium had similar outcome in CMTd patients when compared to cases without neuromuscular diseases [9]. However, both normal and prolonged recovery has been noted by Baraka and Pogson et al., with the use of vecuronium [10,11]. Regardless of the types of muscle relaxant administered, monitoring of neuromuscular function using train-of-four stimulation has been advised. Considering the surgical procedure involving extremity in our case, neuromuscular blockade was not required.

There are meagre data to illustrate the advantage of total intravenous anesthesia (TIVA) including propofol over volatile anesthetics based methods. In spite one report raised concern for malignant hyperthermia with the use of inhalational agents [12], however Antognini in a retrospective study of 86 CMTd cases used malignant hyperthermia triggering agents (halothane) in 77 patients without detrimental outcome[7]. Maintaining anesthesia with nitrous oxide is another safety issue. Due to its interaction with vitamin B_{12} and the hypothetical effect on neurological function, the administration of nitrous oxide has been cautioned during anesthesia of CMTd [13].

In the setting of functional denervation resulting in muscular atrophy and weakness, the use of regional anesthesia is relatively controversial. Due to preexisting invariable neurological involvement, there are issues of aggravation of neurologic deficits or new onset neurologic complications with the use of regional techniques. However, studies support the safety of peripheral and neuraxial methods of anesthesia in CMTd without exacerbating the medical condition [14,15]. Dhir et al., illustrated the peripheral regional blocks with the guidance of ultrasound imaging in 3 cases of CMTd without any adverse complications [14]. Likewise, Bosenberg et al reported the usage of a single

shot sciatic nerve block as well as the continuous infusion with a catheter placed with the assistance of a nerve stimulating device for postoperative analgesia with no side effects [3]. A successful administration of central neuraxial blockade for labor and cesarean section in patients with CMTd was also demonstrated by Brock et al., [15]. In our case patient did not consent for any regional techniques.

CONCLUSION

With comparatively inadequate and controversial evidence considering the safety and efficiency of modes of anesthetic techniques, issues with general, neuraxial and regional anesthesia continued to pose challenges to anesthesia provider for Charcot-Marie-Tooth disease (CMTd). In this case authors considered the use of total intravenous anesthesia (TIVA) as a safe and effective method for administering anesthesia in a patient with CMTd. This method allowed authors to remain away from relatively questionable practice of using neuromuscular blockers, inhalational agents, nitrous oxide as well as regional techniques in patient with CMTd.

REFERENCES

1. Emery AE. Population Frequencies of Inherited Neuromuscular Diseases-A World Survey. Neuromuscul Disord. 1991; 1: 19-29.
2. Bösenberg A, Larkin K. Anaesthesia and Charcot-Marie-Tooth Disease. Southern South Afr J Anaesth Analg. 2006; 12: 131-133.
3. Pareyson D, Marchesi C. Diagnosis, natural history and management of Charcot-Marie-Tooth Disease. Lanct Neurol. 2009; 8: 654-667.
4. Saporta AS, Sottile SL, Miller LJ, Feely SM, Siskind CE, Shy ME. Charcot-Marie-Tooth disease subtypes and genetic testing strategies. Ann Neurol. 2011; 69: 22-33.
5. Martyn JA, Richtsfeld M. Succinylcholine-induced hyperkalemia in acquired pathologic states: etiologic factors and molecular mechanisms. Anesthesiology. 2006; 104: 158-169.
6. Mills P. Charcot-Marie-Tooth disease: suxamethonium and malignant hyperthermia triggering agents. Anaesthesia. 1998; 53: 1134.
7. Antognini JF. Anaesthesia for Charcot-Marie-Tooth disease: A review of 86 cases. Can J Anaesth. 1992; 39: 398-400.
8. Greenberg RS, Parker SD. Anesthetic management for the child with Charcot-Marie-Tooth disease. Anesth Analg. 1992; 74: 305-307.
9. Schmitt HJ, Wick S, Münster T. Onset and Duration of Mivacurium-Induced Neuromuscular Blockade in Children with Charcot-Marie-Tooth Disease. A Case Series with Five Children. Pediatr Anaesth. 2006; 16: 182-187.
10. Baraka AS. Vecuronium Neuromuscular Block in a Patient with Charcot-Marie-Tooth Syndrome. Anesth Analg. 1997; 84: 927-928.
11. Pogson D, Telfer J, Wimbush S. Prolonged Vecuronium Neuromuscular Blockade associated with Charcot Marie Tooth Neuropathy. Br J Anaesth. 2000; 85: 914-917.
12. Ducart A, Adnet P, Renaud B, Riou B, Krivosic-Horber R. Malignant hyperthermia during sevoflurane administration. Anesth Analg. 1995; 80: 609–611.
13. Weimer LH, Podwall D. Medication-induced exacerbation of neuropathy in Charcot Marie Tooth disease. J Neurol Sci. 2006; 242: 47-54.
14. Dhir S, Balasubramanian S, Ross D. Ultrasound-guided peripheral regional blockade in patients with Charcot-Marie-Tooth disease: a review of three cases. Can J Anaesth. 2008; 55: 515–520.
15. 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.

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