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Case Report Open Access

Anesthetic Management of an Obese Child with Charcot-Marie-Tooth Disease: A Case Study

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Abstract

Charcot-Marie-Tooth (CMT) disease is an inherited motor and sensory neuropathy predominantly affecting the feet and legs, with an estimated prevalence of one per 2500 [1]. Some of anesthetic medications present varying degrees of potential risk for worsening CMT neuropathy. This is a case study of a 14-year-old, obese (body mass index= 36.5), male child known to have CMT disease and listed for urgent laparoscopic appendectomy. Obesity as a co-morbidity was an additional burden affecting the anesthetic management along with calculating the doses of anesthetic drugs.

Keywords: Charcot-Marie-Tooth disease; Neuro muscular disorders; Obesity

Introduction

Charcot-Marie-Tooth (CMT) disease is an inherited motor and sensory neuropathy that takes different forms. It is predominantly affecting the feet and legs but the hands and arms can be affected in the advanced stages of disease. Presently incurable, this disease is one of the most common inherited neurological disorders, with an estimated prevalence of one per 2500 [1]. Subtypes of CMT may be differentiated by medical and family history, clinical examination, nerve biopsy, nerve conduction velocity studies (NCV), electromyogram (EMG) and genetic studies [2]. Clinically distal predominant wasting, weakness, and sensory loss, with foot deformities are present. Reilly et al. [3] classified CMT on bases of genetic cause and upper limb motor nerve conduction velocity (MNCV) [3]. Demyelinating (CMT1), is considered if MNCV < 38 m/s and axonal (CMT2) if MNCV > 38 m/s [4]. Physiotherapy, orthopedic devices or surgery can help the affected individuals cope with the disabling symptoms of the disease. The degree of disability is unpredictable between and within families [5]. Some of anesthetic medications present varying degrees of potential risk for worsening CMT neuropathy. Sensitivity to thiopentone was reported in 20 patients with CMT compared to a control group [6].

Nitrous oxide, by inhibiting the cobalamin-dependent enzyme, methionine-synthase, may be considered neurotoxic in prolonged cases [5]. The possibility of succinylcholine induced hyperkalaemia can be prevented by a small "defasciculating" dose of a non-depolarizing muscle relaxant that may lessen the potassium release from diseased muscle [7]. Very few patients can develop pulmonary complications due to respiratory muscle affection, resulting in a restrictive lung pattern. Patients may have few or no symptoms despite considerable abnormalities in pulmonary function. The presence of proximal muscle weakness of the arms may be a predictor for respiratory muscle weakness. Theoretically, muscle weakness related to loss of motor units might sensitize a patient to non-depolarizing muscle relaxants [8].

Case Report

A 14-year-old male child (1.58m, 91kg) was admitted to the emergency room of our hospital suffering from acute abdominal pain and listed for urgent laparoscopic appendectomy. He has no history of medical treatment, no allergy to known medications, but he was diagnosed to have Charcot-Marie-Tooth disease (CMT) in the form of difficulty in running and a tendency to fall. His mother started to notice weakness of his lower extremities few months ago. Weakness of both lower extremities was noted during flexion and extension. Physical findings of the cardiovascular and respiratory systems were unremarkable, and laboratory values were all within normal limits. The mother stated that the diagnosis of CMT had been initially based on clinical symptoms and the positive family history of CMT on his mother's side; and confirmed by EMG as CMT1A and diagnosis was confirmed as demylinating CMT1 with CMTNS Charcot-Marie-Tooth neuropathy score of (14) which correlates with moderate form of the disease [9]. The mother was very knowledgeable about the diagnosis and associated co-morbidities. History of malignant hyperthermia was excluded as the mother had been subjected to several operations before and the child himself did a minor ear surgery in the early childhood period. The previous anesthetic management for their surgeries was reported to be uneventful.

An intravenous line was inserted for good hydration with warm normal saline that was started at a rate of 125ml/hour. The patient was transferred to the operating room, received 2 mg midazolam IV; he was wrapped with warm sheets to prevent hypothermia during the procedure.

Oxygen saturation, non-invasive blood pressure, ECG, capnography, peripheral nerve stimulator (Innervator 272, Fisher & Paykel Healthcare Ltd, New Zealand) and temperature monitoring were in place. Baseline vital signs were recorded, and showed normal ranges.

As the child was considered obese with a body mass index of 36.5 Kgm^{-2}, drug doses were given according to his calculated ideal body

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weight that was approximately 48 Kg for his age, height and sex. Anesthesia was induced by fentanyl 2μgm/Kg, propofol 2 mg/Kg, and a reduced dose of cis-atracurium 4mg to facilitate endotracheal intubation. After loss of eye lash reflex, the depth of anesthesia was increased by inhalation of sevoflurane 3 vol. % in 100% O_2 for 3 minutes. After assuming adequate muscle relaxation as evident by the nerve stimulator, endotracheal intubation with the suitable size tube was performed, and anesthesia was maintained by sevoflurane 1.5±0.5 vol.% in 40% oxygen in air, 1.5 mg cis-atracurium was added 35 minutes after induction (upon 25% recovery of response to T1 % as detected by the nerve stimulator). Intravenous Paracetamol 1gm was given immediately after induction of anesthesia. The operation proceeded uneventfully. At the end of surgery, residual muscle paralysis was monitored by nerve stimulator and muscle relaxant was reversed by neostigmine 0.05 mg/Kg and atropine 0.02 mg/Kg. After reaching acceptable tidal volume and Oxygen saturation, patient was extubated. Enoxaparin 40 mg was given subcutaneously as a thromboprophylaxis at the end of surgery. Recoveries were smooth and prompt along with uneventful postoperative period. Child was contacted 24 hours and one week postoperatively, and there were no complications.

Discussion

Anesthetic management of patients with CMT is a challenge for anesthetist due to the common concern of drug-induced exacerbation of such neuropathy and the unpredictable patients' response to anesthetics such as thiopental, both depolarizing or non- depolarizing muscle relaxants and nitrous oxide. In addition to the importance of good hydration and proper preservation of body temperature from possible hypothermia. Also the incidence of malignant hyperthermia should be taken into consideration during anesthesia.

The Unpredictable response to various medications in CMT is seemed to be due to altered sensitivity to some medications in such neuropathy.

Kotani N et al. [6] have been found a strong relationship between the severity of both motor and sensory disturbances and the minimal induction dose of thiopenton where as this dose was not related to age, gender, inherited type, body weight, and cardiac output. Hence they recommended reduction of thiopental dose according to the individual patient's response [6].

Another important medication is the neuromuscular blocking agents (NMA), as many factors may influence either response to these drugs or performance of neuromuscular monitoring in patients with CMT [10].

On the bases of these factors, it is easily concluded that the response to the same drug especially muscle relaxants may be either normal or prolonged in different CMT patients. Hence there are 2 reports that showed different response to mivacurium in CMT. Schmitt H J et al. [10], found a normal response to a standard dose of mivacurium in five children with CMT1 [10] Whereas, Pogson D et al. [11] noticed a prolonged vecuronium neuromuscular blockade in adult patient with CMT [11].

In the present case report, the patient was fasting so the induction of anesthesia was carried out by non- depolarizing muscle relaxant in a reduced dose according to the calculated ideal body weight (IBW): *IBW = [Body mass index at the fiftieth percentile for the child's age]X [Height (m)]²* [12]. This was different than anesthetic management of Schmitt HJ et al. [10], who anesthetized 5 children with CMT undergoing orthopedic surgeries with propofol based anesthesia without inhalational anesthetic agent and used muscle relaxant as maintenance not for intubation [10].

Succinylcholine was avoided due to the documented risks. As the nerves which are used clinically to monitor neuromuscular function may be affected by (CMT), including the posterior tibial, ulnar and facial nerves, the use of nerve stimulator may be helpful in such situation to exclude the presence of residual neuromuscular blockade and to overcome the possibility of increased risk for postoperative pulmonary complications, which increases further if a residual neuromuscular blockade is present. So the ulnar nerve was supra maximally stimulated at wrist every 5 minutes using train of four principles to ensure adequate muscle relaxation and to avoid higher doses of muscle relaxant.

The negative family history of malignant hyperthermia, along with uneventful minor surgery done during patient's early childhood; makes it safe to use an inhalation agent to maintain of anesthesia. Good I.V. hydration with warm normal saline and wrapping the patient with blanket was essential as complaints of cold intolerance are common in CMT patients but their peripheral responses to cold have not been documented [13].

Although obesity in our patient is most probably not a part of CMT syndrome, yet it represented an additional anesthetic challenge due to the respiratory, cardiovascular, pharmacokinetic, and neurological abnormalities associated with obesity [14]. To our knowledge, there are some reports in literature about successful anesthetic management of children with Charcot Marie tooth syndrome [5,10]. But, there are no reports of any procedures done for obese children having this syndrome. As the incidence of obesity in children increased, co-morbidity with higher incidence of respiratory complications accordingly increased compared with non obese children. So awareness of risk factors for the perioperative complications will help in optimizing the anesthetic management of these children [15]. Especially if the incidence to face respiratory problem may be doubled in CMT –obese child. Dematties M et al. [16], in a family study on 14 family member with CMT have been suggested that there is high correlation between the severity of neuropathy and sleep apnea, that correlated with Age and body mass index and it was higher in male CMT[16].

Another dilemma that will face the anesthetists is the titration of medication dosing for this population [17], due to the effect of obesity on the pharmacokinetics and pharmacodynamics of drugs. As the fat mass which presents 75% of excess weight will affect the distribution of lipophilic drugs and the remainder lean mass alter drug clearance, so calculating the optimum therapeutic dose of a drug depends on understanding the pharmacology of various drugs [18].

Unfortunately there is lack of data on the impact of obesity upon the pharmacokinetics and pharmacodynamics for the majority of medications [17].

To overcome this problem, we calculated drug doses on the expected ideal body weight of the child. However, titrating the dose of non-depolarizing muscle relaxant in this patient according to the calculated IBW will result in suitable surgical muscle relaxation but prompt recovery without residual neuromuscular blockade was another aim. So, the use of nerve stimulator was very important to clarify the actual state of muscle relaxation and the need of relaxant increments. But in our opinion, the sensitivity for muscle relaxants in this neuropathy case is not the same as the normal case, adding to this the problem of obesity. So the primary dose and need to supplement of muscle relaxants for the reported case was titrated by using the neuromuscular monitor. In this case the calculated dose of cis-atracurium according to the ideal

body weight was 7.5 mg, but we reduced it to 4 mg and depended on the nerve stimulator to ensure adequate muscle relaxation. This was different than the anesthetic management of Bösenberg A and Larkin K [5],who anesthetized 2 non obese boys aged 14&19 yrs old with CMT for foot surgery , they avoided muscle relaxant, but combined Propofol and Sevofluraneinduction, with maintenance on sevoflurane in 35% oxygen, and nerve block [5].

Thromboprophylaxis is particularly essential in this patient due to the expected difficult ambulation postoperatively as a result of both obesity and his clinical condition.

Conclusion

In summary, ensuring a sufficient NPO, good perioperative hydration, prevention of hypothermia, thromboprophylaxis, and calculating the dose of non-depolarizing muscle relaxants on the basis of ideal body weight, with dose reduction titrated by using the neuromuscular monitor, all provided a safe and uncomplicated anesthetic management of an obese child with CMT disease. However, further studies should be carried on to reach the optimum management of such cases.

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