

Anesthetical Management of a Patient with Hereditary Muscle Sensory Neuropathy Type 2: Case of a 17-Year Old with Sacral Dermoid and a Short Overview of the Anesthesiological Considerations

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Abstract

We hereby present a short overview of the anaesthesiological considerations regarding the patient with Charcot-Marie-Tooth disease also known as hereditary muscle and sensory neuropathy, which affects peripheral nerves and muscles. Due to pathophysiology of the disease certain anaesthesiological complications associated with HMSN can be related. A case report describing protocol of the total venous anesthesia in the 17-year old patient operated on sacral dermoid with fistulae is presented. The patient recovered without any further complications. In the conclusion we would like to bring the importance of awareness to prepare the HMSN patient for a surgical procedure as well from anesthesiological as from surgical point of view to avoid possible unwanted event such as malignant hyperthermia, hyperkalemia, seizures, prolonged effect of muscle relaxants and worsening of the disease. As an important alternative to general anesthesia regional anesthesia should be considered.

Keywords

Hereditary Muscle Sensory Neuropathy Type 2, Anesthesia, Complications

1. Introduction

Muscle sensory neuropathy type 2, also known as Charcot-Marie-Tooth Syndrome, is the most common inherited neuropathy and it is more common in

women than in men [1]. It affects about one in every 2500 people. HMNS is genetic disease that is associated with degeneration of select populations of motor and sensory neurons with accompanying atrophy and degeneration of their axons. Mostly myelinated axons are affected. Fibre degeneration and fibre loss progresses from distally to proximally. The clinical onset is often in first or second decade of life [2] and it includes peroneal and distal leg muscle wasting and weakness, distal sensory loss and areflexia [3]. Signs of sensory system dysfunction are common and include loss of vibration and joint position sense followed by decrease pain and temperature sensation in stocking and glove distribution. Neuropathic bone deformities develop including pes cavus (high-arched feet) and hammer toe. With further progression the hands become weak [2]. Hand muscles may be severely affected in the autosomal and X-linked dominant forms [3]. Following further progression the severity of the disease worsens, and respiratory dysfunction may develop and become restrictive in nature [4]. Currently treatment of the disease consists only from supportive and surgical treatment because there is no effective drug treatment for the disease itself [5].

Regarding the anesthesia of such patients, possible malignant hyperthermia, hyperkalemia, seizures, prolonged effect of muscle relaxants and worsening of the disease should be taken in consideration.

The present case report describes anesthesia of a patient with HMNS type 2, who underwent the TIVA (Total Intravenous Anesthesia), for a short surgical procedure and the anaesthesiological considerations considering anesthesia of such patients.

2. Case Report

A 17-year old patient with hereditary muscle sensory neuropathy was brought to hospital for operation of sacral dermoid. He was diagnosed with HMSN 15 years ago. No follow ups to reevaluate the progression of disease at the neurologist were made. He already had the typical changes of the feet, which affected his gait but he was still able to walk. Also the fine movement of the arms was affected (troubled signing the consent to anesthesia). From the anaesthesiologic status there were no particularities. According to American society of Anesthesiology (ASA) his physical status was rated class 2. The patient could open his mouth wide, his upper airway was classified with Mallampati 2, neck was not limited in movement. Spirometry showed lightly reduced vital capacity of the lung with no signs of obstruction.

For the anesthesia TIVA with 1% Propofol and Fentanyl was chosen. Propofol was infused with TCI Schneider protocol that was started with 6 mcg/ml. During the anesthesia it was reduced to 3.5 mcg/ml. The sufficient analgesia was achieved with bolus of fentanyl 0.15 mg. The depth of anesthesia was monitored with BIS and it was held between 35 and 50. The medial arterial pressure was held at about 70 mmHg. Profenid was infused as antiphlogistic therapy at the beginning of the procedure. Additional antiemetic therapy was given in the form of On-

dansetron and Dexamethason. During the procedure, which lasted about an hour, he received 500 ml infusion of Elomel Isoton.

Open airway was achieved with Proseal supraglottal device. The mechanical ventilation was performed in pressure control mode with tidal volumes between 300 - 400 ml, frequency 14/min and maximal inspiration pressure of 8 mm H₂O. Saturation was 97% at 50% inhaled Oxygen during the procedure.

After the operation spontaneously breathing patient was brought to recovery room. At admission Catapressan 150 mcg was given as a treatment for the onset of tachycardia. The blood pressure was normal all the time. During the recovery course patient denied pains.

As the patient was fully recovered in one hour and was returned to ward.

The patient was followed up in the surgical outpatient clinic. There weren't any complications due to anesthesia reported.

Informed consent for publication was obtained.

3. Discussion

In discussion I would like to bring the topics that we considered important during anaesthetical management of such particular patient. In the foreground were taken most expected complications such as malignant hyperthermia, hyperkalemia, seizures, prolonged effect of muscle relaxants and worsening of the disease. In our opinion all the possible measures were taken in considerations to avoid the trigger factors that either can cause the acute complications or long term worsening of the disease.

First of all is the type of anaesthesia. The most commonly used inhalation anaesthesia was taken in discussion as a possible predisposing factor for the development of malignant hyperthermia. A clear trigger for malignant hyperthermia has not been defined. After reviewing described cases we have concluded that malignant hyperthermia could be triggered at any HMSN patient. Joseph F. Antognini reviewed anaesthesia of 86 patients with HMNS 2. He concludes that 90% of the patients received either Sevofluran and/or Succinylcholine as triggering factor. Complications other than malignant hyperthermia were present among 22% of the patients. Patients susceptible to hyperkalemia following Succinylcholine developed arrhythmias—from subtle to malignant. Also hemodynamic changes, muscle weakness and pulmonary complications were described [6]. Individuals with certain neuromuscular disorders have long been suspected to have MHS (Malignant Hyperthermia Susceptibility). Due to small sample size of the patients with these disorders, who have developed one or more clinical symptoms of MH-like reactions during or after anesthesia, the anesthesiologists are confronted with the problem whether there is a MH or MH-like reaction in patients with known neuromuscular disorders [7].

Therefore inhalation agents should be used with caution when dealing with patients with HMSN 2, while using TIVA may be a wiser choice.

Additionally there are also possible cardiac manifestations of the HMNS, mostly

described by children with CMTD that underwent anesthesia and may include prolonged QT Syndrome, paroxysmal atrial flutter, cardiomyopathy, premature ventricular complexes (PVC) and a complete atrioventricular block [8] [9]. One of authors describes frequent PVC during an anesthesia with Thioenthal, Atracurium, nitrous oxide/oxygen and Halothan, at a child with plantar release procedure [9]. Also the inhalation anesthetic Sevoflurane has been reported to cause prolongation of the QT interval, and thus, it should be used with caution in patients with QT prolongation. To confirm increased incidence of these cardiac manifestations more research is required [5].

The second problem addressed is a choice of a muscle relaxant. It was not needed in our case, but should be chosen for possible use if the intubation should be performed. There are three possible known complications concerning use of the muscle relaxants. Firstly, the longer effect of the muscle relaxants that can be attributed to the loss of motor units. The latter shows itself in atrophy and muscle weakness. Simultaneous up-regulation of acetylcholine receptors in neuromuscular junction and increased sensitivity to these receptors occur [10]. Pulmonary impairment and the weakness of intercostal muscles and diaphragm may also be present [11]. When such findings are present, the prolongation of neuromuscular blocking agent should be expected. At comorbidities showing phrenic nerve and vocal cord dysfunction, the aspiration risk would be increased [5]. According to Joseph F. Antognini paralyzing dose of non-depolarizing agent was used in 45% of the cases. Most of the patients affected complained about “weakness” postoperatively, two patients developed pneumonia, of which one died due to it [6].

On the other hand the same effect, hence up-regulation of acetylcholine receptor will cause normal or even moderate resistance to the non-depolarizing muscle relaxants in patients in a state of chronic demyelination disease [5]. There is a significant positive correlation between the increase of acetylcholine receptors and the dose of non-depolarizing muscle relaxant [12]. Also should be taken in an aspect, that when using inhalation anesthetics, the neuromuscular blocking effect is prolonged in patients with HMSN, because this condition increases the neuromuscular blocking effect of the inhalational anesthetic itself [18]. Also aminoglycoside antibiotics, hypercarbia, acidosis, hypothermia, hypocalcemia and hypercalcemia may cause the prolongation of a neuromuscular blockade [5].

The neuromuscular monitoring is recommended, but instead of monitoring ulnar nerve, facial nerves are more appropriate due to the fact, that disease affects peripheral nerves [13]. The relaxation of the eye muscle corresponds well with relaxation of diaphragm [14].

The third problem that should be addressed at patients that are in chronic denervation state, is possible hyperkalemia as a consequence of malignant hyperthermia [15], which was discussed earlier, or at usage of defasciculating muscle relaxant. The latter occurs due to overexpressed isoform acetylcholine receptors,

neuronal (nicotinic) alpha 7AChR, that are usually not active and preliminary evidences suggest, that they are up-regulated with either denervated states, injuries, at disuse atrophy and infection [16]. Despite these facts many authors do not take this complication as highly possible or problematic [6].

As a reversal of a muscle relaxation Sugammadex, a modified gamma – cyclo-dextrin, selective antagonist for steroidal neuromuscular blocking agents, such as vecuronium and rocuronium, was proven as an efficient drug for reversing moderate and deep neuromuscular block, also at high dosage of Rocuronium [17]. We should be also aware of possible side effects, such as rash, erythema, hypotension, wheezing and periorbital edema [19] [20]. There have been noticed possible QT prolongations and one case of atrioventricular block, nevertheless several larger studies have proven safety of the drug [21] [22] [23]. Otherwise medication has been successfully used for reversing neuromuscular blockade at the other myopathies (myasthenia gravis, dermatomyositis, Duchenne muscular dystrophie, spinal muscular atrophy, amyotrophic lateral sclerosis) [24]

In the option of anaesthetical management of our patient was also taken the spinal anaesthesia. There are few case reports describing use of spinal anaesthesia at such patients [25] [26]. The 5% Bupivacain injection was delivered with atraumatic 25 G needle. The patient remained haemodynamically stable with oxygen saturation 91% on 50% Ventimask. No special complications that would suggest possible worsening of the patient's neurological condition were described [25]. With a thought, that the infection could be spread around the infected site, we decided to avoid spinal anaesthesia and general anaesthesia was used instead.

4. Conclusion

To conclude, we would like to emphasize the importance of proper preparation of the HMSN patient for a surgical procedure as well from anaesthesiological as from surgical point of view. Important is preoperative status mostly to determine existing neurological deficits. In consideration the proper anaesthesiological technic has to be planned. If it is possible, the regional anaesthesia is a good choice. The right dosage of local anaesthetic has to be chosen and under ultrasound guidance delivered at the right structure [27]. General anaesthesia is also an option. TIVA would probably be in advantage over inhalation anaesthesia due to possible complications that could arise during procedure even though many authors do not find them controversial [6]. Keeping in mind that at these patients postoperatively the prolonged neuromuscular blockade could be expected, there are not many doubts of using the Sugammadex as a reversal drug [21] [22] [23] [24].

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Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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