

SPINAL ANESTHESIA IN A PATIENT WITH CHARCOT-MARIE TOOTH DISEASE, WHO UNDERWENT OSTEOSYNTHESIS WITH TIBIA NAIL PLATE

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ABSTRACT

Introduction: Charcot-Marie Tooth disease is a hereditary peripheral neuropathy characterized by muscular atrophy and sensitive, progressive neuropathy. The authors describe the clinical case of a patient with the disease, who underwent subarachnoid spinal block for orthopedic surgery following a fracture of the left tibia. The patient also had limited neck movement, small mouth rim, reduced bite space (approximately 3cm), and Mallampati Score: Class III. Although there is not much literature on anesthesia for this disease intervention, there is some concern about the use of muscle relaxant drugs due to a risk of prolonged action as in the case of vecuronium, and hypercalcemia as in local anesthetics.

Case presentation: The subarachnoid spinal block was performed through an injection of 4ml Ropivacaine 0.5% in level L4-L5 of the subarachnoid space, using a Whitaker type 25G needle, without bleeding or immediate complications. Although there is scarce literature on the subject, this case confirms the prior evidence that this type of local anesthesia is acceptable in surgery for this disease. In addition, this form of local-regional anesthesia proved useful to reduce the risk of upper respiratory problems due to abnormal anatomy.

Results: The operation proceeded without any problems, and there were no immediate or long-term complications attributable to the subarachnoid anesthesia observed in the postoperative period.

Key words: Charcot-Marie Tooth disease, subarachnoid anesthesia, general anesthesia, hereditary peripheral neuropathy, orthopedics.

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Introduction

Charcot-Marie Tooth disease (CMT) is a hereditary peripheral neuropathy characterized by muscular atrophy and sensitive, progressive neuropathy, which strikes the ends of the limbs and is often associated with hollow foot (pes cavus). The prevalence is estimated to be 1/2,500 in the U.S.A., and 1/10,000 worldwide⁽¹⁾, and there is no sexual or racial predilection. It is a form of hereditary sensory-motor neuropathy caused by defects in the myelin (CMT1, CMT3, CMT4), or axonal damage (CMT2)⁽²⁾. Lower limbs are most commonly affected, resulting in weakness and muscle wasting below the knees and in the feet, causing significant foot deformities. It is characterized by an early loss of deep tendon reflexes.

The hands may be affected, but this usually occurs later in the disease progression. Due to their deformities on their legs, patients frequently trip over objects and have a tendency to sprain their ankles. Limb deformities are often accompanied by a sensory component, resulting in a loss of sensation (heat, cold and pain) in the glove and stocking distribution on the hands and feet. Autonomic dysfunction is also impaired in some forms of the disease. When recognized and treated appropriately patients can lead a relatively normal life. However, diagnosis and treatment are often delayed, which can cause serious bone deformity and a need for surgical correction of the bones and pathological contracture of the soft tissue.

Given the rarity of this disease, our workgroup decided to share this clinical case with the

worldwide scientific community, with the intent of sharing our own experience in the anaesthesiologic management of patients affected by this syndrome.

Case presentation

The patient was a female, 48 years old, weight 65kg, height 160cm, presenting with a fracture of the left tibia following a fall, which required a metal nail plate. The patient history revealed arterial hypertension and Type 1A Charcot-Marie Tooth disease diagnosed at approximately age 20 according to documentation. She had never undergone surgery before, nor anesthesia. On examination the patient exhibited reduced mobility of the upper and lower limbs, and bone-joint deformity consistent with CMT. Examination of the upper respiratory tract revealed hypo-mobility of the neck, small mouth rim, reduced bite space (approximately 3 cm), and Mallampati Score: Class III, which was not modified by phonation. Routine clinical and laboratory examinations were normal, so subarachnoid anesthesia was planned. After positioning the patient on the operating table and monitoring blood pressure, ECG, and oximetry, we injected 4ml Ropivacaine 0.5% in the subarachnoid space at level L3-L4 through a Whitaker type 25G needle, without bleeding or immediate complications. We then sedated the patient using Midazolam i.v.

The hemodynamic parameters remained stable for the duration of the operation which proceeded without problems. The operation ended after two hours, and the patient was transferred to the resuscitation room where she was monitored until she fully regained consciousness. Postoperative exams occurred at 2, 4, and 6 hours after the end of the operation, and the patient gradually regained mobility until full recovery evaluated at the lower right limb. At 1-month and 3-month follow-up the patient reported no neurological disturbances attributable to the local-regional anesthesia.

Discussion

There is not much literature on anesthesia for CMT patients. A review of 161 operations on 86 CMT patients found no increased risk associated with the use of succinylcholine⁽³⁾. There are few and inconsistent studies on non-depolarizing fasciculation. One study on a limited number of cases

showed that mivacurium has an analogical effect on CMT patients compared to children without neuromuscular disease⁽⁴⁾, while other studies indicate a prolonged duration of action of vecuronium in CMT^(5,6). In contrast, it has been shown that there is an increased risk of hypercalcemia with neuromuscular blocking drugs. The difficulty in controlling the airways in patients who develop respiratory insufficiency related to muscle relaxants also increases the perioperative risks for CMT patients under general anesthesia. There is also some concern about the use of local anesthetics in regional or spinal anesthesia in patients with pre-existing neurological disease, which is a contra-indication for local-regional anesthesia. One study on CMT women in labor showed good results with the use of epidural anesthesia during natural and Caesarian delivery⁽⁷⁾.

In addition, another study reported three cases of CMT1 patients who successfully underwent local-regional anesthesia⁽⁸⁾. Although further studies are needed, we believe that local-regional anesthesia is safe and effective for CMT patients. The clinical case presented here confirms the prior evidence in literature cited, in which local anesthesia is not contra-indicated for this disease. The use of local-regional anesthesia in our case reduced the risks involved in upper airway management, which indicated concrete difficulty on the objective examination, as well as reduced duration of post-operative recovery and a better safety profile compared to general anesthesia.

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