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Spinal anesthesia in a patient with Charcot–Marie–Tooth disease undergoing orthopedic surgery: case report

Anestesia raquídea en un paciente con enfermedad de Charcot–Marie–Tooth sometido a cirugía ortopédica: reporte de caso

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Keywords: Anesthesia, Spinal, Charcot-Marie-Tooth Disease, Hip Fractures, Postoperative Period, Neuromuscular Manifestations

Palabras clave: Anestesia Raquidea, Enfermedad de Charcot-Marie-Tooth, Fracturas de Cadera, Período Posoperatorio, Manifestaciones Neuromusculares

Abstract

Introduction: It is uncommon to come across patients with neuromuscular diseases in the daily practice of anesthesia, given the low prevalence of those conditions. Charcot–Marie–Tooth (CMT) disease is the most frequently, caused by an inherited abnormal myelin structure pattern. In view of the low prevalence of this condition (1:25,000), there is little information, derived mostly from case reports, about the use of neuroaxial anesthesia in these patients.

Case presentation: Description of a patient with underlying CMT disease compromising lower limb mobility, who comes to the emergency service due to lower limb pain. After being diagnosed with an acetabular fracture, the patient underwent orthopedic surgery under spinal anesthesia, selected based on patient comorbidities, and the immediate postoperative follow-up.

Results: The anesthetic and surgical procedures proceeded uneventfully and no neuropathic worsening was observed during the next 24 hours.

Conclusion: Uneventful neuroaxial anesthesia is reported in a patient with neuromuscular disease. The case contributes to show the benefits and safety of this form of anesthesia when compared with other options.

Resumen

Introducción: En la práctica anestésica diaria es raro enfrentarse a pacientes con patologías neuromusculares, dada la poca prevalencia de dichas patologías. La más frecuente de ellas es la enfermedad de Charcot–Marie–Tooth, en la cual se hereda un patrón alterado en la estructura de la mielina. Debido a la baja prevalencia de esta patología (1:25000), el uso de anestesia neuroaxial en dichos pacientes no cuenta con mucha información, y mucha de ella proviene de reportes de casos.

Presentación del caso: Se describe el caso de un paciente con enfermedad de Charcot–Marie–Tooth, de base, con compromiso de la movilidad en miembros inferiores, y quien asiste a urgencias por dolor en miembro inferior. Tras ser diagnosticado con fractura de acetábulo, fue sometido a cirugía ortopédica bajo anestesia

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raquídea, indicada a la luz de sus comorbilidades, y el posterior seguimiento inmediato.

Resultados: Se realiza el procedimiento anestésico y quirúrgico sin complicaciones, y no se presenta empeoramiento de la neuropatía en las 24 horas posteriores.

Conclusiones: Se reporta un caso de anestesia neuroaxial en paciente con enfermedad neuromuscular sin incidencias, que ayuda así a ir mostrando los beneficios de la mencionada anestesia y su seguridad frente a otras opciones.

Introduction

Charcot–Marie–Tooth (CMT) disease is the most frequent inherited peripheral neuropathy, with an incidence of 1:25,000. It is both a motor as well as a sensory disease and involves striated muscle weakness and wasting. The most common presentation is peroneal nerve atrophy with anterior and lateral compartment weakness, although there are countless variants. Sensory deficit is usually milder than the motor compromise. It is not one disease but a whole spectrum of disorders caused by myelin gene or axonal mutations.[1]

Motor compromise varies according to the type of inherited pattern. In most cases, it starts with muscle weakness followed by wasting; it is a distal progressive disorder that impairs mobility. In some individuals, axial muscle involvement has been described, presenting as respiratory failure secondary to diaphragmatic weakness. The sensory disorder consists of the presence of neuropathic pain and sensory loss, predominantly distal.

Experience with the anesthetic management of these patients is limited given the small number of case reports, and there is some controversy regarding the use of general anesthesia versus a locoregional technique, and their impact on the underlying disease. However, both techniques have been used successfully in these patients.

The case report is of a patient diagnosed with CMT seen at the traumatology service and diagnosed with left hip fracture. The decision made following patient assessment was to take him to surgical intervention under spinal anesthesia.

Case report

A 63-year-old male Caucasian patient, born and living in Spain, with a history of dyslipidemia, diabetes mellitus type II, mild chronic obstructive pulmonary disease with sequelae from pulmonary tuberculosis infection, and CMT (confirmed by means of genetic testing, as the only affected individual in the family). The patient presented to the traumatology service complaining of left lower limb pain (LLLP), predominantly of the knee and the upper anterior surface of the thigh, with no evidence of trauma. Clinical assessment showed evidence of slight external rotation and leg length discrepancy of the involved limb compared with the contralateral leg. Magnetic resonance imaging ruled out knee joint involvement, and a left acetabular fracture was diagnosed. The patient was scheduled for surgical intervention and was admitted to the traumatology suite.

As a result of CMT disease, the patient exhibited distal weakness of the lower limbs, club feet, hammer toes, abolished Achilles reflexes, and abnormal heel to toe gait.

The initial assessment upon admission revealed stable respiratory status without recent coughing episodes, absence of wheezing or dyspnea, and no edema, hemodynamic or cardiac disorders were found. The patient was scheduled for open reduction and internal fixation of the acetabular fracture.

However, before the surgery the patient was found to have dyspnea, cough, little expectoration, poor decubitus tolerance, oxygen saturation 90% to 91% with oxygen delivered at 4 L/minute through nasal prongs. On auscultation, lower breath sounds were found in both lung fields, together with increased expiration time, with no evidence of wheezing or crepitation. Blood gases showed pO_2/FiO_2 171 mm Hg, pCO_2 33.8 mm Hg, pO_2 61.4 mm Hg; the chest X-ray did not show changes compared with the previous film, and computed tomography did not show pulmonary thromboembolism or changes compared with previous computed tomography scans.

Internal medicine assessment ruled out overinfection or exacerbation, but it was decided to avoid general anesthesia given the patient's respiratory status. After explaining the risks associated with spinal anesthesia and the growing evidence about the safety of this form of anesthesia in relation to the underlying condition, the patient accepted the technique and signed the informed consent.

Following assessment by our anesthesia service, locoregional anesthesia was selected for the procedure. Given the nature of the fracture, it was proposed to carry out the procedure under subarachnoidal or epidural anesthesia, the former being selected in the end given that there was no need for postoperative pain control, while, on the other hand, early ambulation was required in order to start rehabilitation. The procedure was carried out under subarachnoidal anesthesia using a single, atraumatic medial puncture in the L2 to L3 space with a 25G needle (Braun Whitacre; Braun España, carretera de terrasa, 121, 08191 Rubí, Barcelona, España) and 13 mg of 5% bupivacaine injection. The patient remained hemodynamically stable, with good tolerance of the decubitus positioning, 90% to 91% oxygen saturation with 50% Ventimask®, and no episodes of bronchospasm or coughing spells.

Postoperatively, no sensory or motor abnormalities additional to the underlying disease were documented. The patient and the treating physician confirmed that there were no changes from baseline in the motor and sensory status of the right lower limb. As for the LLLP, changes associated with intervention were documented.

The case is reported with the stated informed consent from the patient.

Discussion

Diseases of this type are rarely found during the routine practice of anesthesia; consequently, it is difficult to find relevant information in the literature regarding regional anesthesia in patients with CMT.

Some papers do not make a distinction between the different types of peripheral neuropathies. In fact, J. Neil and J. Ratel, in 2007, suggest that "chronic peripheral nerve involvement, secondary to a mechanical, ischemic, toxic or metabolic disease condition, create a high risk for these patients,"[2] referring to potential complications of regional anesthesia.

The book entitled *Tratado de anestesia regional y manejo agudo del dolor (Treaty of regional anesthesia and acute pain management)*[3] contains a short section on anesthesia in CMT patients, and concludes that drugs that may trigger malignant hyperthermia should be avoided. It goes on to state that respiratory function must be assessed, reviewing prior pulmonary function tests and blood gases, and also assessing the risk of respiratory complications. There is no evidence in the literature in terms of prolonged response to muscle relaxants in CMT patients.

As pertains to regional anesthesia, despite certain relative advantages, its usefulness is open to question. Actually, avoiding interscalene blocks in patients with severe pulmonary dysfunction (because of 30% lung volume reduction) is suggested, as well as avoiding lower limb blocks in cases of severe muscle wasting or atrophy.[3]

Respiratory compromise associated with CMT disease has been reported. Diaphragmatic weakness secondary to phrenic nerve compromise is the finding in patients with progressive dyspnea and orthopnea with no changes in resting blood gases or spirometry[4]; consequently, baseline respiratory status is important, and worsening is not exclusively secondary to the use or not of muscle relaxants or potent inhaled anesthetics, as was concluded from a retrospective review of 161 surgical procedures in 86 patients, although they were related to weakness within the first few hours into the postoperative period.[5]

Different types of regional anesthesia are reported in the obstetrics arena, ranging from a case of epidural management for vaginal delivery in a patient with no respiratory symptoms and no major lower limb involvement[6]; there is at least 1 report of spinal anesthesia for cesarean section, with intradural catheter placement for bolus anesthetic administration in a patient with underlying respiratory failure and rib cage deformity, scheduled for cesarean section because of worsening of the respiratory condition[7]; cases with no reported episodes of dyspnea or worsening of dyspnea or of the underlying condition; and 1 report of spinal anesthesia with the development of an episode of dyspnea and hypotension, difficult to manage, secondary to a sudden rise in the level of anesthesia together with anaphylactic reaction to intravenous (IV) medication given afterwards.[8]

Perhaps the area with the largest number of reports is orthopedic surgery, including patients managed with uncomplicated subarachnoidal anesthesia, except for those attributable to added IV medication[9–11]; epidural anesthesia with evidence of longer block duration than expected for that technique but no other late complications[9]; spinal anesthesia with ultrasound-guided peripheral block, uneventful following the procedure and during the immediate postoperative period[12]; brachial plexus blockade techniques, with no reports of worsening dyspnea or respiratory compromise immediately after the procedure or during follow-up, and no changes from baseline, although with reported motor response abnormalities to neurostimulation – hence the recommendation to perform these techniques under ultrasound guidance to ensure safety in patients with muscle atrophy[13]; peripheral blockade combinations with reports of the same limitation[14]; and combination of general anesthesia with peripheral block, with no reported incidentes[14] and caudal block with evidence of prolonged blockade effect.[15]

Given the growing number of case reports regarding the use of different types of regional or locoregional anesthesia in patients with this form of neuropathy, we believe that individual patient assessment is needed to determine the safety of the technique, based on the clinical findings and the resources available in the workplace.

In the case presented here, given the complications and the patient's clinical condition and test results, subarachnoidal anesthesia was selected with no impact on the patient's respiratory status or worsening of the underlying disease in the immediate postoperative period or later, showing the benefits and safety of this type of anesthesia over other options.

Ethical responsibilities

Human and animal protection. The authors declare that the procedures used in the study followed the ethical standards of Responsible Human Experimentation Committee and were in accordance with the World Medical Association and the Declaration of Helsinki.

Data confidentiality. The authors declare having followed the protocols of their centre of work regarding patient data disclosure.

Right to privacy and informed consent. The authors obtained the informed consent of the patients and/or subjects reported in the article; the form is kept by the corresponding author.

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Conflict of interest

The authors declare having no conflict of interest.

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