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CLINICAL INFORMATION

Use of Sugammadex in Strumpell-Lorrain Disease: a Report of Two Cases

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Abstract
Content: Strumpell-Lorrain disease - or familial spastic paraplegia (FSP) - is a rare hereditary neurological disorder, mainly characterized by variable degrees of stiffness and weakening of the muscles, with cognitive impairment, deafness, and ataxia in the more severe cases. We describe two female siblings with FSP programmed for cholecystectomy and subtotal colectomy, respectively, and also how we dealt with the anesthetic management in both cases and review the literature on this disease in relation to anesthesia.

Introduction

Strumpell-Lorrain disease - also known as hereditary or familial spastic paraplegia (HSP or FSP) - comprises a group of rare neurological disorders affecting mainly the higher motor neurons, causing stiffness and weakness in the legs [1].

FSP is estimated to affect 7.4 in 100,000 individuals. It is a genetic, inherited or hereditary disease; that is, it is passed on from generation to generation. 'Autosomal dominant' (AD) FSP is the predominant form of the disorder, accounting for 70-80% of all cases [1,2].

Hereditary spastic paraplegia is mainly characterized by variable degrees of stiffness and weakening of the muscles of the legs, muscle spasms, and bladder control problems. A limited number of affected families may present some of the following more serious alterations: mental retardation, dementia, epilepsy, peripheral neuropathy, retinopathy, deafness, ataxia, dysarthria, and disorders of the extrapyramidal system [3].

The onset of the disease is usually gradual, slow and insidious, with symptoms that typically worsen progressively over time. The age at onset of the symptoms can be extremely variable among different families, as well as among affected members within the same family [1-3].

The diagnosis of hereditary spastic paraplegia is typically based on careful evaluation of the family and personal history of the patient, a complete physical examination, and

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assessment of the characteristic symptoms and findings. Diagnostic evaluation can also include different specialized tests (neurophysiological or genetic studies) [4].

The treatment of hereditary spastic paraplegia involves medical control of the symptoms and physiotherapy. No treatment is presently able to slow or modify the course of the illness, though baclofen may help reduce spasticity in some patients [1,4].

Patients with neurological diseases pose a challenge when planning anesthesia. In such situations careful selection of the drug type and dose is required. Anesthesia is especially rare as in two patients with this disease in a very short space of time.

We describe two female siblings with FSP respectively programmed for cholecystectomy and subtotal colectomy – the two operations being spaced two months apart. The main interest of our study is that both patients were subjected to general anesthesia, with the use of a specific reversal agent for non-depolarizing neuromuscular block.

Case Report 1

The first case was a 47 year old woman diagnosed with FSP at 23 years of age, with hypertension and dependent upon others for her basic activities. Treatment was being provided with hydrochlorothiazide and baclofen. The preoperative laboratory tests (complete blood count, biochemistry and coagulation) and chest X-rays yielded no relevant findings.

Upon arrival in the surgical area a venous catheter was inserted and 2 mg of intravenous midazolam was administered. In the operating room ECG, pulse oximetry, noninvasive blood pressure, bispectral index (BIS) and neuromuscular block (TOF and TOF ratio in the adductor muscle of thumb) were performed. After pre-oxygenating the patient, anesthetic induction was carried out with fentanyl 2 $\mu g.kg^{-1}$, propofol 1.5 $mg.kg^{-1}$ and rocuronium 0.6 $mg.kg^{-1}$; intratracheal intubation proved uneventful. Maintenance was carried out with sevoflurane for a minimum alveolar concentration (MAC) of 0.6 and remifentanil in continuous perfusion (0.1-0.2 $\mu g.kg^{-1}.min^{-1}$), adjusting the dosage to achieve BIS values between 35-50; relaxant use was not necessary during surgery.

There were no hemodynamic or respiratory incidents during surgery. Before the end of the operation, intravenous dexketoprofen 50 mg, ondansetron 4 mg and acetaminophen 1 g were administered. Extubation proved uneventful, following the administration of sugammadex (2 $mg.kg^{-1}$ body weight) due to the presence of moderate neuromuscular block, until a TOF ratio of over 0.9 was obtained. The patient was discharged from the awakening unit to the hospital ward within three hours, without adverse events.

Case Report 2

The second case corresponded to the younger (43 years of age) female sibling of the above patient, who had been diagnosed with FSP at 16 years of age. In this case, the clinical course was more severe, with severe deafness, cognitive impairment, important vision deficit, neurogenic bladder, repeated episodes of sigmoid volvulus and dolichomegacolon. Her usual treatment consisted of baclofen, tizanidine, clonazepam, and nicardipine. The patient was programmed for laparoscopic subtotal colectomy and ileostomy as treatment for the recurrent volvulus episodes. The preoperative laboratory tests (complete blood count, biochemistry and coagulation) and chest X-rays showed no alterations other than mild anemia (hemoglobin 10 $g.dL^{-1}$).

Upon arrival in the surgical area, a venous catheter was inserted and 1.5 mg of intravenous midazolam was administered. Exploration revealed no clinical sign that predicted difficult intubation. In the operating room ECG, pulse oximetry, noninvasive blood pressure, bispectral index (BIS) and neuromuscular block (TOF and TOF ratio in the adductor muscle of thumb) were performed. After pre-oxygenating the patient, anesthetic induction was carried out with fentanyl 1.5 $\mu g.kg^{-1}$, propofol 1.5 $mg.kg^{-1}$ and rocuronium 0.6 $mg.kg^{-1}$; intratracheal intubation proved uneventful. A central venous catheter was inserted through the right internal jugular vein, without incidents. Maintenance was carried out with a continuous infusion of propofol (4-8 $mg.kg^{-1}.h^{-1}$) until BIS values of 40-50 were obtained, and remifentanil in continuous perfusion (0.1-0.2 $\mu g.kg^{-1}.min^{-1}$); relaxant use was not necessary during surgery.

There were no hemodynamic or respiratory alterations during surgery, and no ventilatory problems were observed on performing the pneumoperitoneum at laparoscopy. Before the end of the operation, intravenous ondansetron 4 mg, paracetamol 1 g, dexketoprofen 50 mg and 150 µg fentanyl were administered. Following administration of a dose of 2 $mg.kg^{-1}$ of sugammadex, extubation proved uneventful, due to the presence of moderate neuromuscular block, to obtain a TOF ratio of over 0.9. The patient was moved to the resuscitation unit, where she remained for 48 hours with a favorable course. Serial laboratory tests and imaging studies were carried out and proved normal in all cases. The patient was discharged home after 8 days in the Department of Surgery.

Discussion

There are practically no studies in the literature on the safety of general anesthesia versus spinal anesthesia in Strumpell-Lorrain disease. A PubMed literature search with the keywords “Strumpell-Lorrain”, “hereditary spastic paraplegia” and “anesthesia” yielded a very limited number of articles, mainly in the field of obstetric anesthesia [5].

Three of the published articles used spinal anesthesia: McTiernan et al [5] described the use of epidural anesthesia for cesarean section, while Thomas et al. [6] and Deruddre et al. [7] used intradural anesthesia likewise for cesarean section. In turn, Mclve et al. [8], Kunisawa et al. [9] and Dallman et al. [10] decided to use general anesthesia for cesarean section, orthopedic surgery, and abdominal surgery, respectively.

The use of neuromuscular blockers is complicated in patients with familial spastic paraplegia. Succinylcholine is contraindicated since it may induce hyperkalemia, and there should be caution in the use of non-depolarizing muscle relaxants due to the risk of exaggerated muscle relaxation. The literature sources have not shown whether regional anesthesia exacerbate the neurological symptoms [11]. However, regional anesthesia is not always possible. Therefore general anesthesia with non-depolarizing neuromuscular blockers would represent a safe alternative – particularly considering that there are have drugs offering rapid and safe reversal of muscle block induced by rocuronium and vecuronium.

In our two patients we chose general anesthesia because of the duration and complexity of both operations.

Great care is required at extubation in patients with FSP, particularly if neuromuscular blockers were administered during the operation. If possible, long-acting neuromuscular blockers should be avoided, with routine monitoring of neuromuscular relaxation throughout the operation, using a standard peripheral nerve stimulator [12]. A TOF ratio of over 0.9 must be confirmed before awakening, accelerating patient recovery with neostigmine or drugs that selectively bind aminosteroid neuromuscular blockers, such as sugammadex.

Although the anesthetic management differed between the two operations (Total Intravenous anesthesia versus inhalatory anesthesia), agreement existed regarding the choice of muscle relaxant, i.e., rocuronium, due to the possibility of antagonizing its effects with sugammadex.

Functional deficiencies being similar to those prior to general anesthesia, subsequent follow-up of both patients revealed no significant worsening of neurological signs.

Conclusion

The main interest of our study is that both patients were subjected to general anesthesia, with the use of a specific reversal agent for non-depolarizing neuromuscular block, followed by complete recovery and no worsening of the existing neurological disease.

This article is an original piece that has not been presented at any congress, and which has not received economical support.

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