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CASE REPORT

Epidural anesthesia for open gastrostomy in a patient with amyotrophic lateral sclerosis

Anestesia epidural para realizar gastrostomía abierta en paciente con esclerosis lateral amiotrófica

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Abstract

Amyotrophic lateral sclerosis is characterized by the progressive degeneration of motor neurons, causing a constellation of symptoms that include muscle weakness, atrophy, fasciculations, spasticity, and hyperreflexia. Currently, Rulizol is the only treatment that has been shown to delay its progression, though to a very small extent. Disease prognosis is grim, with death caused mainly by respiratory failure secondary to muscle weakness, making anesthetic management of these patients a true challenge. The use of muscle relaxants must be avoided as much as possible because of the high risk of ventilatory depression, considering that these patients have an abnormal unpredictable response as a result of heightened sensitivity related to the lower number of acetylcholine receptors. If muscle relaxants are required during the surgery, rocuronium, among nondepolarizing muscle relaxants, is the drug of choice because of its short half-life, while depolarizing relaxants such as succinylcholine are contraindicated because of the risk of lethal hyperkalemia. In terms of intraoperative hypnotics and analgesics, propofol and remifentanil are ideal because of their short half-life. Regarding neuroaxial anesthesia, despite widespread reluctancy to use it, its benefits and rather uncommon adverse effects lead many anesthetists to consider it as an important alternative when it comes to deciding between general or neuroaxial anesthesia, because it reduces airway manipulation significantly, thus reducing respiratory complications as described in this clinical case.

Resumen

La esclerosis lateral amiotrófica se caracteriza por la degeneración progresiva de las neuronas motoras provocando una constelación de síntomas que incluyen debilidad muscular, atrofia, fasciculaciones, espasticidad e hiperreflexia. Actualmente, el único tratamiento que ha demostrado retrasar mínimamente su progresión ha sido el Rulizol. Su pronóstico es infausto, falleciendo mayoritariamente por insuficiencia respiratoria secundaria a la debilidad de su musculatura, siendo el manejo anestésico de estos pacientes, un importante desafío. El uso de relajantes neuromusculares deberá evitarse en la medida de lo posible por el riesgo elevado de depresión ventilatoria, ya que

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estos pacientes tienen una respuesta anormal e impredecible a ellos al presentar una sensibilidad aumentada por el menor número de receptores de acetilcolina; Si la cirugía requiere de ellos, es de elección dentro de los relajantes neuromusculares no despolarizantes, el rocuronio por su vida media corta, contraindicándose el uso de los despolarizantes como la succinilcolina, por el riesgo de hiperkaliemia letal. En cuanto a los hipnóticos y analgésicos que deberemos de usar intraoperatoriamente, el propofol y remifentanilo serían los más ideales por su vida media corta. En cuanto a la anestesia neuroaxial, a pesar de la reticencia extendida a su uso, su beneficio y sus no tan frecuentes efectos adversos, hacen a muchos anestesistas, considerarla como una alternativa de peso a la hora de decidir entre anestesia general o neuroaxial, pues reduce considerablemente la manipulación de la vía aérea, disminuyendo por tanto complicaciones respiratorias posteriores, como referimos en el caso clínico que describiremos a continuación.

Introduction

Amyotrophic lateral sclerosis (ALS) is one of the most frequent types of motor neuron disease in adults, which includes, besides ALS, progressive bulbar paralysis, progressive muscular atrophy, and primary lateral sclerosis. In all these disease types, both the upper and the lower motor neurons are affected.

The etiology is unknown and multiple factors have been proposed (aging, virus, metal poisoning, paraneoplastic syndrome), but there is no strong evidence of their relevant role in the etiology of the disease. Family forms of dominant or recessive autosomal transmission account for close to 10% of the cases, the main cause being a mutation in the superoxide dismutase gene.[1]

The disease is characterized by weakness and progressive muscle atrophy. In its advanced stages, it affects respiratory muscles, leading to progressive muscle weakness, dysarthria, dysphagia, and cramps. The clinical picture is characterized by extensive though asymmetrical amyotrophy, fasciculations, weakness, reactivity of myotatic reflexes, and a combination of bulbar and pseudobulbar syndromes, with preservation of both sensation and mentation, as well as ocular motility and sphincter control, although the latter is impaired in advanced stages. It is a progressive disease lasting 3 years in average. The only pharmacological treatment currently available is Rulizol 100 mg/day.[1,2]

Electromyography is the main diagnostic test, as it confirms neurogenic effects on the muscles manifesting clinical compromise and allows to discover subclinical involvement of other muscles. These alterations include the loss of motor units, a significant increase in the territory of the motor unit with polyphasic potentials, and spontaneous denervation activity (positive waves, fibrillations, and fasciculations). Nerve conduction velocity is normal and there are no cerebrospinal fluid (CSF) abnormalities.

Anesthetic considerations in these patients include preoperative respiratory function tests in order to determine the degree of respiratory weakness and decide on the most appropriate anesthetic technique; forced vital capacity under 50% points to a high probability of complications during emergence and the postoperative period.[3] A thorough neurological examination to document the existing deficit is also important,[4] and the presence of bulbar symptoms (dysphagia, dysarthria) must raise awareness of a high risk of aspiration and respiratory failure.[3] Because of all these reasons, these patients should not be pre-medicated.[5]

Intraoperatively, the use of short half-life drugs is recommended as well as hypnotics such as propofol, desflurane, and sevoflurane because of their low liposolubility. The combination of propofol with inhaled hypnotics is also a possibility during maintenance, as they allow faster awakening because of their different clearance pathways.[6] Opioid analgesics such as remifentanil, with an ultra-short half-life, are advisable in order to allow emergence without the risk of secondary respiratory depression associated with other agents with a more prolonged half-life such as fentanyl or morphine chloride.[7] As for neuromuscular relaxants, they should be avoided whenever possible due to the high risk of respiratory depression, considering that these patients have an abnormal unpredictable response to those agents because of their heightened sensitivity resulting from the lower number of acetylcholine receptors. The use of succinylcholine is contraindicated because of the risk of lethal hyperkalemia, and in terms of nondepolarizing agents, rocuronium is the drug of choice, provided smaller doses than usual are administered and muscle relaxation is monitored.[8] Sugammadex has been used for rocuronium reversal with good results.[2,6,9] The use of propofol and remifentanil for induction allows for correct orotracheal intubation without the need for neuromuscular relaxation.[7]

As for neuroaxial anesthesia, there are reports that caution against its use for fear of accelerating the progression of the disease and causing exacerbations either due to spinal cord trauma caused by the needle or catheter, technical issues, pharmacological toxicity (lidocaine), secondary neural ischemia (vasopressor use), although the actual mechanism is unknown.[10,11] Even though it is important to bear in mind that the lack of a protective cord lining may render the spinal cord more susceptible to potential neurotoxic effects of local anesthetics, these exacerbations of the disease may also be due to surgical trauma, patient positioning during the intervention, tourniquet use, or even drug interactions between medications used for treating the disease and the drugs used intraoperatively and later for analgesia.[4]

As increasingly reflected in the literature, neuroaxial anesthesia may be an important alternative that would benefit patients with significant respiratory muscle

weakness because it reduces and could even eliminate airway manipulation. It can also be a very effective adjunct to general anesthesia, as it allows for regional nerve blockade, ensuring correct anesthesia, immobilization, and analgesia without the need for neuromuscular relaxants, and reducing the need for opioids.[12] Cases of ALS have been described in which combined epidural and general anesthesia with the use of a laryngeal mask (provided there is no bulbar or trunk dysfunction, ie, full stomach)[13] have been used in order to avoid the use of neuromuscular relaxation and reduce the need to manipulate the airway as much as possible; it can even allow spontaneous ventilation if the surgery permits it. Therefore, neuroaxial anesthesia is the safest modality for the respiratory system, with the adverse effects previously believed to exist being infrequent.[5,11,14] Epidural anesthesia is the choice because local anesthetic concentrations are smaller in the white matter of the spinal cord following its administration, as compared to the subarachnoid space; the lack of a protective sheath around the nerve may render the spinal cord more prone to potential neurotoxic effects of local anesthetics following intrathecal administration.[11] It is worth noting that neuroaxial anesthesia may have effects on lung function if the sensory level is at T5 or above, mainly affecting vital capacity, which has been found to drop temporarily by 13% when the sensory level is T5; therefore, it is important to know baseline lung function in order to determine the sensory level that cannot be surpassed.[15]

Clinical case

We present the case of 56-year-old female patient diagnosed with ALS, presenting with tetraparesis and severe dysfunction, scheduled for open feeding gastrostomy.

The patient has type 1 diabetes, depression, and memory impairment. Her usual pharmacological treatment includes Oliclinomel, Insuline lantus, Fluoxetine, Zolpidem, Rulizol, Adiro. She has had multiple surgeries, including open cholecystectomy, splenectomy, abdominal hysterectomy with double adnexectomy, subtotal gastrostomy, caudal pancreatectomy, and left superficial parotidectomy.

Two percutaneous gastrostomies were attempted because of her dysphagia but were unsuccessful perhaps due to an adhesion syndrome associated with multiple prior surgical interventions. Open gastrostomy was proposed following an attempt at placing a nasogastric tube, which was not tolerated due to pharyngeal pain, and assessment by our service was requested.

On pre-anesthetic assessment, the patient was found in poor general condition, with a baseline SpO_2 of 91%, systolic blood pressure (SBP) 130 mm Hg and diastolic blood pressure (DBP) 80 mm Hg, impaired speech due to bulbar involvement, chest X-ray showing slight density increases in the right lung base perhaps related with microaspirations, impossibility to assess lung function after 8 failed attempts at spirometry, and marked respiratory muscle weakness. The patient was classified as American Society of Anesthesiology (ASA) IV/V and fit for surgery.

In the operating room, the patient was monitored in accordance with the standards of the Spanish Society of Anaesthesiology and Resuscitation, using Ventimask with an FiO_2 of 0.5 and maintaining a mean saturation of 97% throughout the procedure.

Epidural anesthesia was chosen followed by sedation with propofol. General anesthesia was discarded in order to avoid respiratory complications related to potential difficult emergence. The T8–9 epidural space was localized and an initial bolus of 0.5% levobupivacaine 40 mg and fentanyl 100 μg was administered. Next, sedation with 1% propofol was initiated at around 2 mg/kg/h. Adequate blockade was achieved at 15 minutes, reaching T5 sensory level. The surgical procedure was started with correct analgesia of the area. Pain was evidenced 40 minutes into the procedure, requiring epidural boluses up to a total dose of 80 mg. Good analgesic quality was maintained during the rest of the procedure, although assessment of motor blockade was not possible due to the underlying disease. Ephedrine boluses were required in order to maintain adequate blood pressure values following epidural anesthesia, and good ventilation dynamics were maintained with no sign of discomfort or respiratory distress at any time.

Upon completion of the surgical procedure, the patient was transferred to the PACU, where she remained until cessation of epidural motor blockade, with no incidents during her stay.

Conclusion

Despite widespread reluctance to use neuroaxial anesthesia in patients with ALS, its benefit in this type of patient makes it the modality of choice for many professionals when it comes to deciding between general or neuroaxial anesthesia, given that it allows for less airway manipulation, reducing potential respiratory complications.[10,11] In our case, it allowed us to preserve spontaneous ventilation and airway protective reflexes. We decided to use epidural anesthesia because of the possibility to titrate the dose, maintain an adequate sensory depth, and avoid, as much as possible, direct contact between local anesthetics and the spinal cord with its heightened susceptibility. Regarding the type of local anesthetic, levobupivacaine was used because, besides being less neurotoxic and cardiotoxic, it produces less motor blockade. Mepivacaine was also used as an adjunct to levobupivacaine, in our case because of its fast onset and short action, avoiding lidocaine that has been found to produce spinal cord toxicity.[16] It is worth highlighting that neuroaxial anesthesia, despite all its

benefits, is not always successful and it is important to keep in mind the possibility of general anesthesia in these cases. A good option could be the use of the laryngeal mask because of the ability to maintain spontaneous ventilation in the patient, provided the surgical procedure allows it and the patient does not have contraindications for its use, as was the case in our patient (dysphagia with risk of aspiration, full stomach). If not, there will be a need to intubate the patient but avoiding muscle relaxants whenever possible is something to remember, with the use of depolarizing relaxants, in particular succinylcholine, being contraindicated because of the risk of lethal hyperkalemia. Among the nondepolarizing neuromuscular relaxants, rocuronium is the agent of choice due to its short half-life, but always under neuromuscular relaxation monitoring. In addition, noncumulative, rapid-clearance hypnotics, and analgesics should be used, in order to avoid postoperative respiratory complications or problems during patient education, propofol, and remifentanil being the drugs of choice for this purpose.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have fol-lowed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors declare that no patient data appear in this article.

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Conflicts of interest

The authors declare having no conflict of interest.

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