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GENERAL ANESTHESIA FOR PACEMAKER IMPLANTATION IN A PATIENT WITH AMIOTROPHIC LATERAL SCLEROSIS: CASE REPORT

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ABSTRACT
Amyotrophic lateral sclerosis (ALS) is the most common form of upper and lower motor neuron disease. Due to rapidly progressive neurodegeneration, surgical interventions are necessary to improve and prolong the life of patients. With the advancement of therapeutic options, anesthesiologists have increased contact with ALS patients, and it is essential to master the pre, intra and postoperative management of this population, which requires special anesthetic care. The objective of this article is to describe the case report of a patient with ALS who underwent total intravenous general anesthesia for surgical implantation of a multisite pacemaker.

Keywords: Amyotrophic lateral sclerosis, Artificial Pacemaker, General anesthesia.

INTRODUCTION

Amyotrophic Lateral Sclerosis (ALS) or Lou Gehrig's Disease is a relatively rare, fatal, rapidly progressing neurodegenerative disease that affects 1-2.6 per 100,000 people annually. Its incidence increases with age, peaking between 60 and 79 years. ALS is the most common form of upper and lower motor neuron disease. Ninety percent of ALS cases are sporadic, while 10% are familial. Clinical manifestations include muscle weakness and atrophy, lack of coordination, spasticity, hyperreflexia, Babinski sign, fasciculations, and cramps. Motor impairment and functional deterioration are measured using the Revised ALS Functional Rating Scale (ALSFRS-R), a tool for assessing the functionality and disease-specific severity. The scale evaluates 12 items (Speech, Salivation, Swallowing, Handwriting, Cutting Food, Dressing and Hygiene, Transfers, Walking, Climbing Stairs, Dyspnea, Orthopnea, and Respiratory Insufficiency), with a score of zero to four for each item, and a total score ranging from 0 to 48 points, where 48 indicates normal functionality and 0 indicates complete disability. The treatment is palliative and includes symptomatic relief and supportive care. As the disease progresses, surgical interventions may be necessary to improve and prolong the lives of patients. With advances in therapeutic options,

the anesthesiologist's contact with ALS patients has increased. Pre-, intra-, and postoperative management of this population requires special care, as they are more susceptible to anesthesia-associated complications.[1-6]

CASE REPORT

Male patient, 67 years old, 70 kg, with recently diagnosed ALS (less than a year ago). The patient is on home oxygen therapy (nasal cannula oxygen support). Admitted to the Surgical Center on room air, presenting with hypoxemia (70%), which improved after oxygen administration with an oxygen mask. The patient had no motor deficits, and no impairments in speech, gait, or swallowing. An echocardiogram showed moderate dilation of the left heart chambers, left ventricle with moderate to severe global diastolic dysfunction, and moderate diastolic dysfunction. The mitral and tricuspid valves were structurally normal with mild insufficiency, and mild to moderate pulmonary hypertension (pulmonary artery pressure of 50 mmHg). A Holter exam revealed a sinus rhythm with atrial flutter, ventricular arrhythmias, polymorphic extrasystoles, ventricular bigeminy, runs of ventricular extrasystoles, and non-sustained ventricular tachycardia. A chest CT scan showed signs of interstitial edema, changes in the left lower lobe suggestive of bronchopneumonia and inflammatory bronchopathy, small right-sided pleural effusion and laminar effusion on the left side, with adjacent atelectasis, and an enlarged heart.

Surgical proposal for multi-site pacemaker implantation and anesthetic planning for total intravenous general anesthesia. Monitoring included cardioscope, pulse oximeter, temperature, invasive blood pressure in the right radial artery, capnography, Conox, TOF (Train-of-four), and urine output. Pre-oxygenation was performed using a facial oxygen mask. Anesthetic induction was carried out with Sufentanil 15 mcg, Propofol TCI (target-controlled infusion), Rocuronium 50 mg, and Dobutamine 0.15 mg/kg/h. Periglottic block with Ropivacaine 1% 5 ml was performed. Orotracheal intubation was performed using direct laryngoscopy with an 8.0 tube. Intravenous anesthetic maintenance was achieved with Remifentanil 0.15 mcg/kg/min and Propofol TCI. The patient remained hemodynamically stable, with Dobutamine 0.15 mg/kg/h throughout the procedure. No additional doses of neuromuscular blockers were required after anesthetic induction. For symptomatic control and prophylaxis, the following were administered: Dipyrone 2 g, Ondansetron 8 mg, Lidocaine 80 mg, Omeprazole 40 mg, Dexamethasone 10 mg, and Cephalothin 2 g. The procedure was completed without complications, lasting approximately three hours. The patient was extubated in the operating room after the administration of Sugammadex 200 mg, guided by TOF, with total reversal of the neuromuscular block. The patient was transferred to the ICU, hemodynamically stable, without the use of any drugs. The patient was discharged.

DISCUSSION

ALS manifests through a combination of upper and lower motor neuron dysfunction, affecting the bulbar, cervical, thoracic, and lumbar segments. The complete molecular basis of its pathophysiology is not yet fully understood. Pathophysiological processes are divided into four main categories: impaired RNA metabolism, altered autophagy, cytoskeletal defects, and mitochondrial dysfunction. It is believed that there are three variants of ALS: the classic

sporadic type, familial ALS, and the Western Pacific type, which is often associated with dementia. Approximately 10% of ALS cases are familial and caused by genetic mutations, typically inherited in an autosomal dominant Mendelian pattern. Besides the genetic component, environmental exposure appears to influence disease susceptibility. Suspected associated risk factors include smoking, athletic predisposition or activity, military service, β-N-methylamino-L-alanine, head trauma, electromagnetic fields, agricultural chemicals, and exposure to lead and other heavy metals.[1, 4]

The pattern of neurodegeneration follows a heterogeneous course, progressively affecting an increasing number of muscle groups until the condition presents a symmetrical distribution. Initially, there is progressive weakness of the voluntary skeletal muscles involved in limb movement, evolving asymmetrically and spreading contralaterally, rostrally, and caudally, most often in an anatomically contiguous manner. The disease then progresses to the bulbar muscles, leading to impaired swallowing (dysphagia) and speech (dysarthria). Involvement of the respiratory muscles results in respiratory insufficiency with hypercapnia, an inability to clear secretions, which in turn increases the risk of aspiration and respiratory disorders. This constitutes the main cause of death in ALS, which occurs on average two to three years after the onset of symptoms. Sphincter and extraocular muscles are usually spared.[3,7]

Despite the predominance of motor dysfunction in the manifestations of the disease, cognitive and behavioral changes can occur early in the disease course in 35% to 50% of cases. Individuals with ALS may present with language and executive function impairments, apathy, compulsive behavior, loss of empathy, irritability, disregard for hygiene, changes in eating habits, emotional lability, depression, anxiety, and sleep disturbances.[3]

Given the progressive nature of the disease and advances in palliative treatments to ensure the quality of life for patients, surgical procedures have increasingly been performed in this population, such as percutaneous enteral gastrostomy, long-term catheter insertion, and tracheostomy. Consequently, the anesthetic management of this group of patients, which requires special care, has increased.[8]

Anesthetic techniques, whether regional or general, pose different risks for patients with ALS. Risks include gastric aspiration, postoperative ventilatory support, autonomic instability, and increased and unpredictable sensitivity to opioids, sedatives/hypnotics, and non-depolarizing neuromuscular blocking agents. The anesthetist should begin care for the ALS patient in the preoperative phase. It is important to conduct pulmonary function tests to assess the possibility of respiratory complications. The forced vital capacity (FVC) is a parameter used to evaluate the success of extubation: if < 50%, patients should receive non-invasive positive pressure ventilation. Advanced bulbar symptoms increase the risk of aspiration and respiratory inadequacy.[2, 4]

In the intraoperative phase, fast-acting and reversible analgesic and amnesic agents should be used. The infusion of remifentanil and propofol for induction is an alternative used due to its ultrashort action. Regarding inhalational anesthetics, attention should be paid to the potential for respiratory depression in the postoperative period. Sevoflurane has low lipid solubility, making it suitable for its rapid reversal, as does desflurane. Desflurane is the least soluble of the inhalational anesthetics, ensuring the early recovery of airway functions. Additionally, when used above a minimal alveolar concentration, it

promotes muscle relaxation in a dose-dependent manner.[2, 7]

Neuromuscular blockers should be used in low doses for patients with ALS. Depolarizing neuromuscular blockers should be avoided due to the potential risk of hyperkalemia. Non-depolarizing blockers act as competitive antagonists of the postsynaptic receptor, preventing acetylcholine from binding to its receptor, leading to flaccid paralysis and prolonged weakness. The reversal of the blockade with Sugamadex at a dose of 2 mg/kg accelerates the reversal of paralysis.[4]

In the postoperative period, monitoring the respiratory pattern is crucial, as this group of patients has impaired respiratory function and limited functional reserve, along with an inability to clear secretions and chronic carbon dioxide retention. Non-invasive ventilatory devices can be helpful in ensuring the success of extubation. Additionally, changes in the level of consciousness and mental confusion may indicate hypercarbia. Supplemental oxygen therapy should also be limited, as respiratory drive and control are dependent on oxygen saturation during sleep.[4]

Local and regional anesthetic techniques, such as peripheral nerve blocks, have increasingly been indicated for patients with ALS, with fewer complications reported compared to general anesthesia. These techniques can be utilized both intraoperatively and for postoperative pain management. Traditionally, they were avoided due to concerns about worsening pre-existing neurological symptoms, justified by the "double crush" theory. This theory posits that a nerve with a prior compressive injury is more likely to be injured again, suggesting that patients with motor neuron diseases like ALS, who already have neurological disturbances as the "first crush" could experience a "second crush" when subjected to mechanical, ischemic, or toxic insults associated with regional anesthesia. However, reported worsening of neurological symptoms has been associated with factors such as surgical stress, positioning, body habitus, and local anesthetic concentration. There has been an increase in case reports documenting successful use of regional anesthetic techniques without exacerbation of neurological symptoms, as seen in this case report.[2, 4]

In summary, the anesthetic management of patients with ALS should begin with pre-anesthetic care, assessing the functional impairment of the disease and its systemic repercussions to tailor the approach. Motor impairment and functional deterioration, measured by the ALSFRS-R, are critical for evaluating the disease's severity, which was not conducted in the current case report. Nonetheless, intraoperative care with monitoring using a cardiograph, pulse oximeter, temperature, invasive blood pressure in the right radial artery, capnography, Conox, TOF, and diuresis was essential for guided and successful anesthetic management. The choice of short-acting medications like Propofol and Remifentanil for maintenance anesthesia also aligns with recommendations found in the literature. The success of extubation was ensured by using Rocuronium in low doses, with monitoring of neuromuscular function via TOF guiding reversal with Sugammadex. Postoperative care in the ICU for ventilatory support was also crucial for the case's outcome.

CONCLUSION

The anesthesiologist must pay close attention to the specific characteristics of patients with ALS, conducting a thorough evaluation starting from the preoperative phase. The adoption of appropriate medications and adequate support during the anesthetic procedure and postoperative care can ensure, as demonstrated in this case, that the surgery proceeds without

complications, allowing the individual to be extubated in a short time without compromising their motor and cognitive functions.

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