Case Report

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Anesthetic management in a patient with steinert syndrome undergoing laparoscopic cholecystectomy: A case report

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Abstract

Introduction: Steinert Syndrome (SS) is a progressive, hereditary, multisystem muscular dystrophy. We aim to describe the anesthetic management of a patient with SS undergoing laparoscopic cholecystectomy (LC).
Case report: Male, 47 years old with SS underwent LC with total intravenous anesthesia. Induction was performed with 100mg propofol, 200µg fentanyl, and 10mg rocuronium, followed by maintenance with 2% propofol (target between 1.5 and 3.0µg/ml keeping the BIS between 40 and 50) and 0.1 µg/kg/min remifentanil. The procedure lasted 50 minutes, with no intercurrences. In the end, dipyrone 1g, parecoxib 40mg, and ondansetron 8mg were administered. When TOF reached >2%, 100µg of sugamadex was administered. The patient was extubated when the TOF index exceeded 90%. After 24 hours in postsurgical ICU, followed by 12 hours in the ward, the patient was discharged uneventfully.
Conclusions: With correct planning of anesthetic technique, and knowledge of associated risks, perioperative management can be done safely.
Keywords: Case report, steinert syndrome, general anesthesia, laparoscopic cholecystectomy.

Introduction

Steinert syndrome (SS) is a progressive, hereditary, multisystem muscular dystrophy, characterized by musculoskeletal, gastrointestinal, neurological, respiratory, endocrine, and cardiovascular features [1]. It considered the most common type of muscular dystrophy in adults [2, 3].

The higher prevalence of cholelithiasis justifies studying the response of this group to surgical-anesthetic stress. We aim to describe the anesthetic management of a patient with SS undergoing laparoscopic cholecystectomy (LC).

Case Report

A male patient, 47 years old, 67 kg, 1.70 m, with SS for 30 years, on preanesthetic evaluation to perform LC. He had muscular atrophy, myotonia, history of asthma and cataract, and had been submitted to tracheostomy and gastrostomy three years ago.Domperidone (20 mg), amitriptyline (25 mg), lactulone (13 g), muvinlax (14 g), dipyrone (500 mg), montelukast (10 mg), and loratadine (10 mg) were used. He had a brother and mother with SS.In electroneuromyography of the limbs, during mild exertion, he presented motor unit potentials of decreased durations and amplitudes in all muscles. During rest, fibrillation potentials and positive spikes wore observed. The patient was classified as physical status ASA III. Monitoring included continuous electrocardiogram, pulse oximetry, noninvasive blood pressure (NIBP), esophageal thermometer, capnography, bispectral index (BIS), train-of-four (TOF), and posttetanic count (PTC). Intraoperatively, he maintained a BP of 120x50mmHg, 60-70 bpm, an SatO2 100%, an EtCO2 30-24 mmHg, a BIS between 45 and 60, and normothermia.Total intravenous anesthesia (TIVA) was used. Induction was performed with 100mg propofol, 200µg fentanyl, and 10mg rocuronium, followed by maintenance with 2% propofol (target between 1.5 and 3.0µg/ml keeping the BIS between 40 and 50) and 0.1 µg/kg/min remifentanil.

The procedure lasted 50 minutes. In the end, dipyrone 1g, parecoxib 40mg, and ondansetron 8mg were administered. When TOF reached >2%, 100µg of sugamadex was administered, and extubated when reached >90%. After 24 hours in

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the postsurgical ICU, and 12 hours in the ward, the patient was discharged uneventfully.

Discussion

Several factors can influence the surgical-anesthetic act, such as facial dystrophy, respiratory muscle dysfunction, arrhythmias, and altered sensitivity to drugs in general, which can cause apnea, even in small doses [1].Complications in the postoperative period usually result in cardiopulmonary dysfunction and weakness of the pharyngeal muscles, with a higher incidence of hypoxia, atelectasis, aspiration, and respiratory failure, especially in the laparoscopic approach [1]. These patients are more susceptible to ventilatory depression, either because of greater sensitivity to depressant drugs, weakness of the respiratory muscles [4], or abnormal central ventilatory control mechanisms, leading to reduced sensitivity of the respiratory center and central sleep apnea [2].
Shivering and hypothermia can induce and prolong myotonic contractures, so normothermia should be maintained throughout the perioperative period using active warming devices, as well as considering increasing the temperature in the operating room [2].

Suxamethonium can precipitate exaggerated contraction, masseter spasm, and laryngospasm, and should be avoided. Nondepolarizing NMB and anticholinesterasics, if needed, should be used with caution. Reports suggest that it may be safe to use sugamadex. In patients not already tracheostomized, rapid sequence induction should be considered, given the increased risk of aspiration due to delayed gastric emptying and pharyngeal muscle weakness [4].Five case reports of patients with SS who underwent anesthetic procedures for LC were found [1,5-8]. Three studies used the TIVA [5,6,8], whereas only one [1] used spinal anesthesia and the last one used a “combined spinal-epidural” technique associated with TIVA [7].

There is evidence of an increased risk of malignant hyperthermia in these patients; therefore, triggering agents, such as depolarizing NMB, and halogenated agents should be avoided [9]. We chose TIVA with propofol, remifentanil, and rocuronium, which was effective for the surgery, causing minimal intraoperative hemodynamic compromise and no complications in postoperative periods, aligning with reports of SS undergoing LC [1,5-8].

Myocardial involvement is rarely identified, but mild abnormalities of systolic function can be seen on echocardiography. Only two cases reported preoperative changes. One case had an electrocardiogram showing left bundle branch block, isolated ventricular extrasystoles, left ventricular overload, and diffuse repolarization changes [5]. In the other echocardiogram, changes were found, with reduced left ventricular capacity, ejection fraction of 40%, and pulmonary test results with a restrictive pattern [7]. In this report, the patient presented only isolated supraventricular arrhythmia, with no changes in cardiac morphology or function, showing only minimal pericardial effusion on echocardiography.

Myotonia can affect any muscle group and is caused by several factors and drugs used in general anesthesia, such as hypnotics, sedatives, and opioids [10]. Propofol is the most commonly used hypnotic, despite reports of triggering myotonia and prolonged recovery [5,10]. It was the hypnotic of choice in this case, which was planned to maintain the BIS between 40 and 50 with a target dose of 1.5 to 3.0 μg/ml. We used a small dose of fentanyl at induction (200 μg) and remifentanil (0.1 μg/kg/min) at maintenance.

Prolonged use of neuromuscular blockers (NMBs) should be done with caution. Depolarizing agents should be avoided, as they may trigger a myotonic crisis and make ventilation and intubation more difficult [1]. The effects of succinylcholine are unpredictable and may precipitate a myotonic crisis. Nondepolarizing drugs usually elicit a normal response, but if there is a loss of muscle mass, prolongation of the response may occur [4,10].

As a NMB, we used rocuronium with monitoring of the neuromuscular junction, and the effect was reversed with sugammadex, which has also already been successfully used in other cases of general anesthesia for patients with SS [5,9].
In this report, there were no intraoperative complications, highlighting the reports of myotonic crisis [5] and delayed reversal of neuromuscular blockade [7], which represent possibilities to be considered in the anesthetic planning.

In other studies [1,5–10], postoperative pain management was performed with intraoperative administration of iv dipyrone (2g) alone or combined with tramadol SC (100 mg) + ketoprofen (100 mg) IV, fentanyl (80 mcg) + acetaminophen (1g) + parecoxib (40 mg) with intraoperative spinal perfusion with ropivacaine 0.1% (2 ml/h), maintained for 24h or more; NSAIDs with a bolus of 10 mL of bupivacaine 0.125% were administered by an epidural catheter. We chose to use dipyrone 1g, parecoxib 40mg and local infiltration of the portals with ropivacaine 0.2%, and the patient had a good evolution and

was discharged after 24 hours of observation in a semi-intensive therapy unit and 12 hours in the ward.sics can precipitate myotonia and should also be used with caution [5]. Reports suggest that it may be safe to use sugamadex in patients with myotonic dystrophy [5]. In patients not already tracheostomized, rapid sequence induction should be considered if the patient requires general anesthesia given the increased risk of aspiration due to delayed gastric emptying and pharyngeal muscle weakness, keeping in mind the risks associated with nondepolarizing neuromuscular blockade [5].

In the literature review, 12 case reports of patients diagnosed with Steinert syndrome who underwent surgical and anesthetic procedures were found[3,4,6-15].Of these, five studies addressed cases of LC, allowing direct comparison to the anesthetic procedures adopted[3,7,11,13,15]. Three studies used the total intravenous technique[7,13,15], whereas only one[3] used spinal anesthesia (fentanyl 20 mcg + normal saline 1 ml, followed by hyperbaric bupivacaine 5 mg + normal saline 2 ml), and the last one used a "combined spinal-epidural" technique associated with the total intravenous technique [11].

Although it is a controversial topic, there is evidence in the literature of an increased risk of malignant hyperthermia in these patients; therefore, triggering agents, such as depolarizing NMB, and halogenated agents should be avoided [6].
In the present report, according to what has already been exposed, we chose total intravenous general anesthesia with propofol, remifentanil, and rocuronium, which was effective for the surgery, causing minimal intraoperative hemodynamic compromise and no complications in the immediate and late postoperative periods, aligning with reports of SS undergoing LC [3,7,11,13,15].

Myocardial involvement is rarely identified in myotonic dystrophy, but mild abnormalities of systolic function can be seen on echocardiography. Only two cases reported preoperative changes. One case had an electrocardiogram showing left bundle branch block, isolated ventricular extrasystoles, left ventricular overload, and diffuse repolarization changes [15]. In the other echocardiogram, changes were found, with reduced left ventricular capacity, ejection fraction of 40%, and pulmonary test results with a restrictive pattern [11]. In this report, the patient presented only isolated supraventricular arrhythmia, with no changes in cardiac morphology or function, showing only minimal pericardial effusion on echocardiography.

In some studies, [3,4,6-15] intraoperative monitoring was performed with pulse oximetry, noninvasive blood pressure, electrocardiogram, capnography, muscle relaxation monitoring using the adductor pollicis muscle with a train-of-four sequence and body temperature. In other studies, as in this report, the bispectral index was added [12]. In the report where only spinal anesthesia was chosen, only ECG, pulse oximetry, noninvasive blood pressure, spirometry, and arterial blood gas analysis were used [3].

Myotonia can affect any muscle group and is caused by several factors and drugs used in general anesthesia, such as hypnotics, sedatives, and opioids [4]. Propofol is the most commonly used hypnotic, despite reports of triggering myotonia and prolonged recovery [4,15]. It was the hypnotic of choice in this case, which was planned to maintain the BIS between 40 and 50 with a target dose of 1.5 to 3.0 µg/ml. We used a small dose of fentanyl at induction (200 µg) and remifentanil (0.1 µg/kg/min) at maintenance, as cited in other reports [8,10].
Prolonged use of neuromuscular blockers (NMBs) in SS patients should be done with caution. Depolarizing agents should be avoided, as they may trigger a myotonic crisis and make ventilation and intubation more difficult[3]. In addition, the effects of succinylcholine in these patients are unpredictable and may precipitate a myotonic crisis. Nondepolarizing drugs usually elicit a normal response, but if there is a loss of muscle mass, prolongation of the response may occur [4, 5].
As a neuromuscular blocker, we used rocuronium with monitoring of the neuromuscular junction, and the effect was reversed with sugammadex, which has also already been successfully used in other cases of general anesthesia for patients with SS [6,15].

In this report, there were no intraoperative complications, highlighting the reports of myotonic crisis [15] and delayed reversal of neuromuscular blockade [11], which represent possibilities to be considered in the anesthetic planning of patients with SS.

Postoperative analgesia can be performed with NSAIDs and opioids and prevention of symptoms, such as nausea and vomiting, with ondansetron or droperidol.

In these studies[3,4,6-15], postoperative pain management was performed with intraoperative administration of iv dipyrone (2g) alone or combined with tramadol SC (100mg) + ketoprofen (100mg) IV, fentanyl (80mcg) + acetaminophen (1g) + parecoxib (40mg) with intraoperative spinal perfusion with ropivacaine 0.1% (2 ml/h), maintained for 24 h or more; NSAIDs with a bolus of 10 mL of bupivacaine 0.125% were administered by an epidural catheter. In the present report, we chose to use dipyrone 1 g, parecoxib 40mg and local infiltration of the portals with ropivacaine 0.2%, and the patient had a good evolution and was discharged after 24 hours of observation in a semi-intensive therapy unit and 12 hours in the ward. It is also necessary to highlight the importance of multidisciplinary perioperative planning for adequate management of the patient to reduce risks and allow early identification of

complications related to the syndrome.

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