

Anestesia para Colectomia Videolaparoscópica em Paciente Portador de Doença de Steinert. Relato de Caso e Revisão de Literatura *

Anesthesia for Videolaparoscopic Cholecystectomy in a Patient with Steinert Disease. Case Report and Review of the Literature

Flora Margarida Barra Bisinotto, TSA ¹, Daniel Capucci Fabri ², Maida Silva Calçado ³, Paula Borela Perfeito ⁴, Lucas Vieira Tostes ⁵; Gabriela Denardi Sousa ⁵

RESUMO

Bisinotto FMB, Fabri DC, Calçado MS, Perfeito PB, Tostes LV, Sousa GD – Anestesia para Colectomia Videolaparoscópica em Paciente Portador de Doença de Steinert. Relato de Caso e Revisão de Literatura.

JUSTIFICATIVA E OBJETIVOS: As distrofias miotônicas são doenças neuromusculares de transmissão autossômica dominante. Dentre elas, a distrofia miotônica tipo 1 (DM1), ou doença de Steinert, é a mais comum no adulto e, além do envolvimento muscular, apresenta manifestações sistêmicas importantes. A DM1 representa um desafio para o anestesiológico. Os pacientes apresentam maior sensibilidade às drogas anestésicas e complicações, principalmente cardíacas e pulmonares. Além disso, há a possibilidade de apresentarem hipertermia maligna e crise miotônica. Descreveu-se o caso de um paciente que teve complicação pulmonar importante após ser submetido a anestesia geral.

RELATO DO CASO: Paciente de 39 anos, portador de DM1, foi submetido à anestesia geral para colectomia videolaparoscópica. A anestesia foi venosa total com propofol e remifentanil e rocurônio. O procedimento cirúrgico de 90 minutos não apresentou intercorrências, mas após a extubação o paciente apresentou insuficiência respiratória e crise miotônica, que tornou a intubação traqueal impossível. Utilizou-se a máscara laríngea, que possibilitou a oxigenação adequada, e a ventilação mecânica foi mantida até a recuperação total da atividade respiratória. Evolução ocorreu sem outras complicações.

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CONCLUSÕES: A DM1 é uma doença que apresenta várias peculiaridades para o anestesiológico. O conhecimento minucioso do seu envolvimento sistêmico, associado à ação diferenciada das drogas anestésicas nesses pacientes, proporcionará um ato anestésico-cirúrgico mais seguro.

Unitermos: DOENÇAS, Muscular: distrofia miotônica; CIRURGIA, Abdominal: colectomia; COMPLICAÇÕES, Pós-Operatórias: insuficiência respiratória

SUMMARY

Bisinotto FMB, Fabri DC, Calçado MS, Perfeito PB, Tostes LV, Sousa GD – Anesthesia for Videolaparoscopic Cholecystectomy in a Patient with Steinert Disease. Case Report and Review of the Literature.

BACKGROUND AND OBJECTIVES: Myotonic dystrophies are autosomal dominant neuromuscular diseases. Among them, myotonic dystrophy type 1 (MD1), or Steinert disease, is the most common in adults, and besides muscular involvement it also has important systemic manifestations. Myotonic dystrophy type 1 poses a challenge to the anesthesiologist. Those patients are more sensitive to anesthetics and prone to cardiac and pulmonary complications. Besides, the possibility of developing malignant hyperthermia and myotonic episodes is also present.

CASE REPORT: This is a 39-year old patient with DM1 who underwent general anesthesia for videolaparoscopic cholecystectomy. Total intravenous anesthesia with propofol, remifentanil, and rocuronium was the technique chosen. Intercurrences were not observed in the 90-minute surgical procedure, but after extubation, the patient developed respiratory failure and myotonia, which made tracheal intubation impossible. A laryngeal mask was used, allowing adequate oxygenation, and mechanical ventilation was maintained until full recovery of the respiratory function. The patient did not develop further complications.

CONCLUSIONS: Myotonic dystrophy type 1 presents several particularities to the anesthesiologist. Detailed knowledge of its systemic involvement along with the differentiated action of anesthetic drugs in those patients will provide safer anesthetic-surgical procedure.

Keywords: COMPLICATIONS, Postoperative: respiratory failure; DISEASES, Muscular: myotonic dystrophy; SURGERY, abdominal: cholecystectomy.

CASE REPORT

This is a 39 years old male weighing 68 kg, with the diagnosis of MD1 and cholelithiasis, who worked as a salesman, scheduled for videolaparoscopic cholecystectomy, and he was evaluated in the pre-anesthetic outpatient clinic. He had a history of muscular weakness since nine years of age, but only at age 34 when he presented significant dysphagia, gastroesophageal reflux, and cataract the MD was diagnosed. The patient did not have a history of anesthetic procedures, and he used *aloe vera*. The family history was significant for his mother who died at age of 50 years due to complications of MD and five siblings with the disease (three sisters and two brothers). He had five children, four healthy boys and a girl with MD. On physical exam, he had typical fascies (Figure 1) with lagophthalmos and frontal baldness, difficulty swallowing that affected speech (dysphonia), hypotrophy of the distal muscles of the upper and lower limbs (Figure 2 and 3), myotonia of the hands (difficulty to relax the hands after a contraction), and Mallampati IV. Laboratorial exams were normal except for CK-NAC of

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INTRODUCTION

Myotonic dystrophy (MD), the most common type of muscular dystrophy in adults, is a neuromuscular disorder also known as Steinert disease. It was first described in 1909 simultaneously by Batten and Gibb in England, and Steinert in Germany, who published several cases of patients of the same family with MD¹. Muscular dystrophy is caused by an abnormal expansion of the sequence of a trinucleotide (cytosine-thymine-guanine) in chromosome 19. It is transmitted genetically (autosomal dominant) and the phenotypical expression is highly variable².

It has an estimated incidence of one in 8,000 births, and the worldwide prevalence ranges from 2.1 to 4.3/100,000³. This disease is characterized by the presence of myotonia, i.e., persistent muscle contraction after voluntary contraction. It is a multisystem disease and it can be associated with severe complications. Anesthesiologists must know the clinical manifestations of this disease to provide appropriate and safe anesthesia for this group of patients when they undergo surgical procedures. The objective of the present report was to present the case of a patient with MD who underwent cholecystectomy under general anesthesia, discussing the anesthetic conduct. We then go on to review the literature on the subject.



Figure 1 – Facies of a patient with Steinert Disease



Figure 2 – Lower limb hypotrophy



Figure 3 – Upper limb hypertrophy

271.0 U.L⁻¹ (normal range for men from 24 to 204 U.L⁻¹). The electrocardiogram showed left branch block, isolated ventricular PVCs, left ventricular overload, and diffuse changes in ventricular repolarization. The chest X-ray and echocardiography were normal. The patient was evaluated by a cardiologist and cleared for the anesthetic-surgical procedure; cardiologic medications were not prescribed. His physical status was classified as ASA 3. The patient received ranitidine (150 mg) and metochlopramide (10 mg) 90 minutes before surgery, but not other pre-anesthetic medication. General anesthesia was induced with midazolam (5 mg), remifentanil (100 µg), and rocuronium (50 mg), being intubated with rapid sequence intubation and posterior introduction of an orogastric tube for aspiration. Anesthesia was maintained with O₂ and medicinal air at 50%, and continuous infusion of propofol (100 µg.kg⁻¹.min⁻¹) and remifentanil (0.15 µg.kg⁻¹.min⁻¹). Monitoring consisted of pulse oximetry, non-invasive blood pressure, electrocardiogram, capnography, monitoring of muscular relaxation using the train-of-four in the adductor pollicis muscle, and body temperature. The patient underwent videolaparoscopy with insufflation with carbon dioxide to induce pneumoperitoneum, but maintaining the intra-abdominal pressure below 15 mmHg. An electrical scalpel was used to facilitate hemostasia. Body temperature was maintained with an electrical blanket. Thirty minutes before the expected

end of the surgery, subcutaneous tramadol (100 mg) and intravenous dypirone (2 g) and ketoprofen (100 mg) were administered. The surgical procedure was completed in 90 minutes, without interurrences. At that time the patient did not show any response to the train-of-four. We waited for about 20 minutes until the development of at least one response and then the curarization was reversed with atropine (1.0 mg) and prostigmine (2.0 mg). He then showed four equal responses to the train-of-four and responded to verbal commands to open his eyes and mouth. However, his respiratory pattern was not satisfactory. When the patient was regaining consciousness, he reacted to the presence of the ET tube and he was extubated. Pulse oximetry was 98%, but after five minutes he showed progressive decrease in oxygen saturation, without improvement of the respiratory function. His extremities became cold and cyanotic, he developed muscular hypertonia, lost consciousness, and it was difficult to ventilate the patient with the face mask. Tracheal intubation was impossible due to generalized hypertonia. At that time, blood was drawn for ABGs and electrolytes. The laryngeal mask was used, allowing adequate ventilation and oxygenation. Arterial blood gases showed significant cyanosis (pH= 7.17, PaCO₂= 77.9 mmHg, PO₂= 125.3 mmHg, HCO₃ = 28.2 mEq.l⁻¹, and base excess = -2.3), but the level of electrolytes did not change. Myotonia was diagnosed and treated with midazolam (5 mg) and he remained on controlled ventilation with the laryngeal mask. The patient was transferred to the post-anesthetic recovery room because ICU beds were not available. He regained consciousness slowly and mechanical ventilation continued during this time. After five hours, the laryngeal mask was removed without interurrences. The patient was transferred to a regular room and discharged from the hospital after three days.

DISCUSSION AND REVIEW OF THE LITERATURE

Muscular dystrophies belong to a group of hereditary diseases characterized by progressive muscular weakness. There are several classification criteria based on genetic transmission, age of onset of symptoms, muscles involved, and speed of progression.

In the beginning of the 1990s, a mutation in chromosome 19 was identified for the first type of the disease identified genetically and classified as muscular dystrophy type 1 (MD1) or Steinert disease⁴. In 1994, a similar disease was identified, being called proximal myotonic myopathy or muscular dystrophy type 2 (MD2). Nowadays, MD1 refers to the types of the disease related to chromosome 19 and MD2 to new genetically distinct diseases. A repeated sequence of the nitrogenous bases cytosine (C), thymine (T), and guanine (G) of variable length, in which the size of the repeated sequence is correlated with the clinical symptoms of the disease, is the genetic alteration in MD1. It is an autosomal dominant disorder and in the majority of the cases the gene is inherited from the mother, but paternal transmission is seen in a small number

of cases. Although a neonatal type does exist most patients develop symptoms of MD1 in adult life.

The disease is characterized by muscular involvement and several associated systemic manifestations. Prognosis depends mainly on the cardiac and respiratory repercussions.

Muscular involvement: Myotonia can be seen in several neuromuscular disorders. This word describes a persistent muscle contraction observed after termination of voluntary contraction or stimulation. The patient is not capable of relaxing the muscle after using it. Initially, the muscular involvement manifests as myotonia and atrophy, predominantly in distal portions of the limbs, but it can affect the muscles of the face (facial and temporal muscles) and larynx, and the respiratory muscles. It occurs due to an intrinsic change in the muscle, and not in the peripheral nerve or neuromuscular junction. This can be demonstrated since myotonia is not abolished by peripheral nerve blocks or neuromuscular blockers. Its mechanisms have not been defined. Evidence suggests damage of chloride or sodium channels in the muscular membrane. It has been suggested that complete deactivation of sodium channels and an increase in the activity of calcium-dependent potassium channels are contributing factors to increase the excitability and susceptibility to myotonia in MD⁵. Alternatively, a dramatic reduction in transmembrane chloride conductance can also occur. The primary involvement of peripheral nerves is also controversial, and some authors have suggested that weakness and muscular dystrophy are consequences of a polyneuropathy⁶.

Cardiac involvement: It is represented mainly by deterioration of the conduction system of the heart, tachycardias, cardiomyopathy, and valve diseases⁷. First degree AV block with prolonged PR interval is the most common conduction defect in patients with MD1 and present in the ECG of more than 40% of the patients. The most common arrhythmia is atrial in origin, but mono or polymorphic ventricular tachycardia can also be seen. Delayed conduction in the His bundle can lead to the development of severe arrhythmias secondary to reentry. Severe rhythm disorders, such as fatal ventricular tachycardia, can develop early in patients with congenital or infantile MD as well as in asymptomatic teenagers with none or little disease manifestation. They can develop sudden cardiac arrest related mainly to physical exertion⁸. Heart failure is present in a significant proportion of patients, but it is not clinically apparent due to the limited physical activity. For this reason, echocardiography is an important exam to quantify the degree of disruption of the cardiac function. Cardiac lesions are progressive and they can evolve for more severe types faster than the evolution of the disease itself, and, in those cases, little correlation between the cardiac and muscular skeletal disease is observed. Therefore, those patients should be followed continuously, paying special attention for cardiovascular manifestations to make the best treatment decisions, and especially for the choice of anesthetic technique and drugs.

Pulmonary involvement: Pulmonary complications are multifactorial and frequent causes of morbidity in MD. Patients show weakness of the respiratory muscles, and altered central control of the respiration with changes in respiratory mechanisms, leading to global hypoventilation, microatelectasis, and reduction in pulmonary compliance. Ventilatory response to carbon dioxide is reduced, which might have a central origin or be attributed to a reduction in the contractile strength of the diaphragm⁹. This function is further diminished in upper abdominal surgeries. Several studies have suggested that central control mechanisms of respiration are normal in patients with MD^{9,10}. Chronic hypercapnia is a common finding and can be seen even in the presence of minimal signs of peripheral muscular weakness. Along with hypoventilation, patients have day time somnolence, which is also not related to the symptoms of muscular weakness, but it can be due to changes in the respiratory center or sleep disorders characterized mainly by the presence of central or obstructive sleep apnea. Muscular weakness can affect the diaphragm and other respiratory muscles causing difficulty to cough and reduction in functional residual capacity. Weakness of the abdominal muscles that despite showing increased work during respiration is not effective in improving respiratory parameters also contributes for the respiratory complications. Increased sensitivity to sedative and anesthetic drugs can be seen.

Gastrointestinal manifestations: Gastrointestinal manifestations are present in 80% of the cases, being considered important for the quality of life of MD patients. The most common symptoms include: nausea, vomiting, dysphagia, early satiety (due to a reduction in gastric emptying time), and gastroesophageal reflux¹¹. Both myopathic weakness and myotonia of the oropharyngeal muscles are highly important in the dysfunction during the oral and pharyngeal phases of swallowing in patients with MD¹². Cholelithiasis is a common finding due to the increase in vesicular biliary sphincter. For unknown reasons, liver function tests are abnormal¹³.

Endocrine involvement: It can include hypothyroidism, hypogonadism, altered secretion of the growth hormone, and abnormalities in glucose metabolism and insulin (frequently associated with diabetes mellitus)¹⁰. Asymptomatic female patients can also develop infertility.

Central nervous system: Regarding the personality, changes in behavior were observed in the first descriptions, but until the 1980s very few cases were reported, which are usually related to the severity of the muscular involvement. Obsessive-compulsive and passive-aggressive behaviors, as well as apathy and depression, are common in patients with MD. Mental retardation can be seen in 60% of the congenital types of MD, affecting severely all measures of general intelligence. Classical types of the disease, juvenile and adult, develop cognitive decline, characterizing dementia. Personality, motivation, and emotional changes can lead to social isolation¹⁴⁻¹⁶.

Anesthesia Planning and Perioperative Factors Associated with the Development of Myotonia

Several precautions should be taken not only during anesthesia, but also in the postoperative period in patients with Steinert disease to prevent known complications. The development of myotonia represents an important problem for anesthesia because, if laryngeal and respiratory muscles are involved, intubation can be difficult or even impossible¹⁷. Potential trigger factors include the surgery, hypothermia, tremors, electrical or mechanical stimulation during or after the surgery, drugs (clofibrate, propranolol, potassium), and anesthetic agents, such as succinylcholine, and anticholinesterase drugs¹⁸. Treatment is mainly preventive, avoiding all triggering factors. A protocol of safe anesthetics should be adopted. The use of electrical scalpel should be avoided. Body temperature should be monitored closely to minimize the risk of tremors.

The anesthetic technique of choice remains uncertain. Whenever possible, peripheral nerve block or neuroaxis block is the technique of choice¹⁹. When general anesthesia is indicated, extreme care should be taken during all phases of anesthesia. During anesthetic induction, thiopental is relatively contraindicated due to more prolonged respiratory depression. Propofol has been used both for induction and maintenance of anesthesia, but it can also be a problem. Changes in sensitivity, which may be increased or decreased, in patients with MD have been reported in the literature, and, consequently, it can be difficult to calculate the proper dose^{20,21}.

Although a direct relationship between MD and malignant hyperthermia has not been reported in the literature, depolarizing neuromuscular blockers represent a particular problem, since they can have unpredictable effects²². Succinylcholine seems to have a double effect in myotonic patients. It can cause a "normal" neuromuscular blockade, but it can affect the muscles directly causing muscular contraction and enough hyperkalemia to lead to cardiac arrest. Besides, it can initiate a generalized myotonic response, resulting in difficulty in tracheal intubation and ventilation²³. Therefore, the use of succinylcholine in patients with myotonic dystrophy should be avoided, although some cases of administration of this drug in emergency situations in patients with MD without complications have been reported in the literature²⁴. Since myotonia is caused by a primary defect in the musculature, the use of non-depolarizing agents does not abolish generalized contraction.

Non-depolarizing neuromuscular blockers have an unpredictably long action in patients with MD, and the cases reported in the literature suggest a reduction in the dose and use of drugs with intermediate duration, especially atracurium^{22,25}. Nish et al.²⁶ calculated the effective dose for 50% and 90% (ED₅₀ and ED₉₀ respectively) of the patients to achieve neuromuscular blockade of vecuronium for the orbicularis, adductor pollicis, and flexor hallucis muscles in a patient with MD undergoing general anesthesia with a laryngeal mask and maintenance with propofol, nitrous oxide, and fentanyl. The ED₅₀ for the orbicularis, adductor pollicis, and flexor hallucis muscles was 7.77 (3.10-16.8), 28.3 (20.7-43.3), and 29.5 (11.0-85.6) µg.kg⁻¹, respectively (p < 0.01), and ED₉₀ of 35.7 (14.8-66.5),

51.8 (29.3-145.0), and 50.6 (5.29-642.0) µg.kg⁻¹, respectively (p < 0.01), indicating a marked increase in susceptibility only for the orbicularis muscle. Other authors²⁷ had already shown an increase in muscular sensitivity to vecuronium, especially of the face muscles. Those drugs do not change the muscular tension triggered by the hyperexcitability of the muscular membrane in myotonia and metabolic exhaustion in malignant hyperthermia. It is not known whether the administration of anticholinesterase agents induces myotonia, although the unpredictability of its action has been reported²⁸. Those patients also show increased sensitivity to opioids and their dose should be decreased²⁹.

As for halogenated anesthetics, it has been questioned whether all myopathies are associated with an increased risk of malignant hyperthermia (MH). In 2004, it was recognized that approximately 50% of the patients identified as being susceptible to malignant hyperthermia by the contracture test had mutations of the ryanodine receptor, which is responsible for the development of this complication³⁰. Several studies have shown that 30-50% of the individuals susceptible to MH have myopathological changes³¹.

Takhar et al.¹⁸ reported a case of videolaparoscopic cholecystectomy using alfentanil, propofol and sevoflurane, and nitrous oxide without neuromuscular blockers, resulting in elevated abdominal insufflation pressures around 20 mmHg. Complications related with the anesthetic technique were not observed. The use of general anesthesia has been reported with diazepam, propofol, rocuronium, isoflurane, and morphine³². Due to the doubts, it is prudent to avoid drugs that could potentially trigger malignant hyperthermia in patients with myotonic dystrophy³³.

Large size surgeries in patients with Steinert disease have also been reported in the literature. Gelsomino et al.³⁴ reported a series of six patients with myotonic dystrophy who underwent cardiac surgery with hypothermic extracorporeal circulation. Anesthesia was maintained with midazolam, remifentanyl, propofol, and atracurium. Complications were not reported. Propofol, sufentanil, atracurium, and clonidine were used successfully in another patient undergoing cardiac surgery³⁵.

In a study³⁶ with 219 patients with MD undergoing anesthesia, the authors reported a rate of complication of 8.2%, mostly pulmonary and mainly in upper abdominal surgeries. Besides, the severity of the disease and age above 37 years also contributed to increase the incidence of respiratory complications. The risk of pulmonary complications was 14 times higher in patients with proximal muscular weakness when compared with those with less severe disease. Weakness of the pharyngeal musculature can lead to obstructive apnea and aspiration pneumonia. Weakness of the respiratory muscles can prevent effective coughing, leading to the formation of atelectasis. The inspiratory effort decreases inspiratory capacity and increases the risk of alveolar hypoventilation. Most complications in patients with MD are pulmonary in origin. Acute respiratory failure causing retention of carbon dioxide such as in the case presented here is a common complication in those patients. The majority of anesthetic drugs including

halogenated, hypnotic, and opioid drugs as well as neuromuscular blockers can induce hypoventilation and respiratory failure. Thus, ventilatory assistance is extremely important in the immediate postoperative period. Mechanical ventilation with tracheal intubation has been used for ventilatory support, although it can increase the risk of pulmonary infection, even for short periods. Besides, sedation is required to prevent the discomfort of the tracheal tube. This can prolong the recovery time of the respiratory function. The laryngeal mask minimized the need for sedation, since it is less traumatic for the airways and cardiovascular system. It is better tolerated by the patient, allowing calmer arousal, attenuating airways and hemodynamic reflexes. Potential complications of the laryngeal mask, such as hypopharyngeal trauma and aspiration, have been reported^{37,38}. We considered the possibility of aspiration of gastric contents during surgery, and maintaining the torso of the patient elevated in the recovery room was responsible for making the procedure safer regarding the risks of aspiration, since complications were not observed in the late postoperative period. Other ventilation modalities, such as positive pressure in the airways (BIPAP) with face mask, have been effective in improving oxygenation and reducing hypercapnia³⁹. Postoperative BIPAP has also been reported in conjunction with the laryngeal mask⁴⁰.

In view of what happened during anesthesia of this patient with MD, we conclude that deep knowledge of the disease, which is multisystemic and demands careful anesthetic planning by anticipating complications, is recommended.

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RESUMEN

Bisinotto FMB, Fabri DC, Calçado MS, Perfeito PB, Tostes LV, Sousa GD – Anestesia para Colecistectomía Videolaparoscópica en Paciente Portador de Enfermedad de Steinert. Relato de Caso y Revisión de la Literatura.

JUSTIFICATIVA Y OBJETIVOS: Las distrofias miotónicas son enfermedades neuromusculares de transmisión autosómica dominante. Entre ellas está la distrofia miotónica tipo 1 (DM1), o enfermedad de

Steinert, que es la más común en el adulto y además de la involucreción muscular, presenta manifestaciones sistémicas importantes. La DM1 representa un reto para el anestesiólogo. Los pacientes presentan una mayor sensibilidad a los fármacos anestésicos y complicaciones, principalmente cardíacas y pulmonares. Además de eso, existe la posibilidad de presentar hipertermia maligna y crisis miotónica. Se ha descrito el caso de un paciente que tuvo una complicación pulmonar importante después de haber sido sometido a la anestesia general.

RELATO DEL CASO: Paciente de 39 años, portador de DM1, sometido a la anestesia general para colecistectomía videolaparoscópica. La anestesia fue venosa total con propofol y remifentanil y rocuronio. El procedimiento quirúrgico de 90 minutos no presentó intercurencias, pero después de la extubación, el paciente presentó insuficiencia respiratoria y crisis miotónica, que hizo la intubación traqueal imposible. Se utilizó la máscara laríngea, que posibilitó la oxigenación adecuada, y la ventilación mecánica se mantuvo hasta la recuperación total de la actividad respiratoria. Evolucionó sin otras complicaciones.

CONCLUSIONES: La DM1 es una enfermedad que presenta varias peculiaridades para el anestesiólogo. El conocimiento minucioso de su involucreción sistémica, asociado a la acción diferenciada de los fármacos anestésicos en esos pacientes, proporcionará un acto anestésico-quirúrgico más seguro.