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CASE REPORTS

Anesthesia for bariatric surgery in patient with mitochondrial myopathy – case report

Carlos Eduardo Coimbra Melonio[a], **Ciro Bezerra Vieira**[a], **Plínio C. Leal**[a], **Caio Marcio B. de Oliveira**[a], **Elizabeth T.N. Servín**[b], **Lyvia Maria Rodrigues de Sousa Gomes**[b], **Ed Carlos R. Moura**[a,b,*]

[a] *Universidade Federal do Maranhão, São Luis, Maranhão, Brazil*
[b] *São Domingos Hospital, São Luis, Maranhão, Brazil*

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Abstract Mitochondrial Myopathy is a rare pathology caused by a defect in the mitochondrial DNA metabolism, leading to defects in the formation of adenosine triphosphate, in the Krebs citric acid cycle, fatty acid oxidation and oxidative phosphorylation. It is manifested by exercise intolerance, muscle fatigue on small efforts, muscle weakness, tachycardia, and difficulty breathing. There are few case reports on the operative management of adult patients suffering from mitochondrial myopathy. With this report, we intend to describe the anesthetic management of a patient with mitochondrial myopathy who underwent laparoscopic gastroplasty and outline some anesthetic considerations about this pathology.

Introduction

Mitochondrial myopathies comprise a large heterogeneous group of neuromuscular disorders resulting from primary mitochondrial respiratory chain dysfunction, causing damage to energy metabolism.[1,2]

Although many different anesthetic techniques have been successfully used in these patients, there are reports of serious complications occurring during and after exposure to anesthesia. Tissues that require high energy are exclusively dependent on the energy released by mitochondria and, therefore, have the lowest threshold to exhibit symptoms of mitochondrial disease.[1]

We describe the perioperative anesthetic management of a patient with mitochondrial metabolic myopathy undergoing videolaparoscopic gastroplasty and discuss important anesthetic considerations regarding this rare condition.

* Corresponding author.
E-mail: edcrmoura@yahoo.com.br (E.C. Moura).

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Case report

KRMN, female, 48 years old, caucasian, suffering from mitochondrial myopathy, weighing 112 kilos, 1.65 meters tall, Body Mass Index (BMI = 41). The patient reported onset of myopathic symptoms after performing general intravenous anesthesia with propofol for cholecystectomy 8 years ago. Weeks after the surgery, she noticed difficulty in flexing her arm, fatigue after great efforts, muscle pain, headache, tachycardia accompanied by sweating and diarrhea. Six years ago, she underwent a muscle biopsy showing: Sparse muscle necrosis with macrophagy and subsarcolemal mitochondrial proliferation, and intrasarcoplasmic lipid accumulation.

The patient had the following comorbidities: obesity, depression, Hashimoto's thyroiditis, superficial purpura, hypercholesterolemia, systemic arterial hypertension (SAH), and tendonitis in the lower limbs. Former smoker quit 25 years ago. Regarding the treatment of myopathy, the patient took L-Carnitine (2 $g.day^{-1}$), Vitamin E (800 $mg.day^{-1}$), Vitamin C (1 $g.day^{-1}$), Coenzyme Q10 (100 $mg.day^{-1}$), Risoflavin (50 $mg.day^{-1}$), Pregabalin (150 $mg.day^{-1}$), Cyclobenzaprine (10 mg), Citoneurin 5000 (thiamine nitrate 100 mg + pyridoxine hydrochloride 100 mg + cyanocobalamin 5,000 $mcg.day^{-1}$). Regarding the treatment of SAH, the patient took ramipril (5 $mg.day^{-1}$).

She has been undergoing epidural block anesthesia for 20 years due to cesarean section and spinal anesthesia for 16 years for second cesarean section and tubal ligation. She also underwent general anesthesia for cholecystectomy. She reports an episode of seizure after the childbirth of his first son and was reversed with intravenous Diazepam 10 mg. During the second cesarean section, she reported having severe pain during the procedure, even under spinal anesthesia.

Recently, the patient underwent laparoscopic gastroplasty in order to reduce obesity. The patient underwent a cardiopulmonary evaluation, where no electrocardiographic, echocardiographic, or physical examination changes were found. The patient has good preoperative functional capacity. Regarding the classification of the clinical scenario by the American Society of Anesthesiologists, the patient was classified as ASA III. Preoperative laboratory tests showed: leukocytes (8,400 mm^3), neutrophils (6,669.6 mm^3), hemoglobin (14.5 $g.dL^{-1}$), hematocrit (42.4%), platelets (68,000 mm^3), Reactive Protein C (RCP) (3.54 $mg.dL^{-1}$) and potassium 4.2 $mmoL.L^{-1}$.

On admission to the operating room, she was eupneic, normal-colored mucosa, acyanotic, anicteric, and hemodynamically stable. Physical examination showed good mouth opening, Mallampati 2, with no predictors of difficult airway. In the operating room, she was monitored with pulse oximeter, cardioscope, noninvasive blood pressure, Train-Of-Four (TOF) monitoring, and Bispectral Index (BIS) brain function monitoring. General anesthesia was induced with 0.05 $mg.kg^{-1}$ of midazolam, 4 $mcg.kg^{-1}$ of fentanyl, 0.5 $mcg.kg^{-1}$ of ketamine, 0.2 $mg.kg^{-1}$ of etomidate, and 1 $mg.kg^{-1}$ of rocuronium. Orotracheal intubation was performed without complications with a 7.5 mm tube and connected to the gas analyzer. Mechanical ventilation was adjusted with the following ventilatory parameters: Tidal Volume = 600 mL; Respiratory Frequence = 12 irpm; Intrathoracic Pressure of 22 cmH_2O. Dexmedetomidine was also used in 10 min – only induction (0.6 $mcg.kg^{-1}$) – and maintenance with sevoflurane 1%–2%.

During surgery, the patient had 99% of oxygen saturation, expired carbon dioxide ranging from mmH_2O to 35 mmH_2O, with a body temperature of 36.5 °C, adequate TOF and BIS between 35–55.There were no great blood pressure or heart rate oscillations, being the patient maintained in the anesthetic plane only with the above mentioned agents. There were no intraoperative complications, being administered ondansetron 8 mg, dipyrone 2 g, parecoxib 40 mg and dexamethasone 4 mg at the end of surgery. The surgical procedure lasted 90 minutes and the anesthetic procedure 110 minutes, being the patient extubated immediately after the use of sugammadex 100 mcg, with 100% TOF and 95 BIS. Five minutes after extubation the patient was awake, conscious, lucid, time and space oriented, without pain, respiratory or motor deficit complaints. She was referred to the Intensive Care Unit (ICU).

The postoperative period was uneventful, the patient was hemodynamically stable, afebrile, and breathing eupneic. In the immediate postoperative period, laboratory tests were requested: leukocytes (10,600 mm^3), neutrophils (9,222 mm^3), hemoglobin (4.2 $g.dL^{-1}$), hematocrit (38.8%), platelets (162,000 mm^3), Prothrombin time (14.6 seconds), Prothrombin Activity (79%), International Standardized Ratio (1.16), Activated Partial Promtrobin Time (25.8 seconds). Throughout the hospitalization period, the parameters remained within normal range.

After 72 hours in the ICU she was discharged to the ward and on the 7th postoperative day, discharged from hospital without reports of paresthesias or motor deficits. Until 4 weeks after the surgical procedure had not presented new symptoms of his disease.

Discussion

Mitochondrial myopathy is a rare condition that is particularly important in the anesthetic- surgical context because it causes varied organ dysfunctions. Patients with mitochondrial diseases often have metabolic dysfunction and heart problems such as reduced heart function, hypertrophic cardiomyopathy, and conduction disorders. Anesthetic effect may cause respiratory depression, lactic acidosis and muscle fatigue.[1]

It is important to avoid circumstances that require metabolic demand such as prolonged fasting, hypoglycemia, postoperative nausea and vomiting, hypothermia, acidosis, and hypovolemia. Due to the difficulty of these patients in lactate metabolism, 0.9% saline or lactate-free Ringer's should be administered.[1,2] As per the patient's preoperative cardiopulmonary evaluation, the presence of diseases was not found; therefore, we chose not to carry out invasive arterial monitoring as it is more comfortable for the patient and to avoid possible complications, especially infection of the puncture site. However, immediate intraoperative and postoperative gasometric analyzes were performed and hydration by PlasmaLyte was chosen.

Anesthetic induction and surgical stress generate an increase in abnormal metabolites that may exacerbate myopathic symptoms. Some studies indicate that patients

suffering from malignant hyperthermia-related myopathies require anesthesia without triggering agents, which means strictly avoiding succinylcholine and volatile substances. However, there is no genetic link established between the phenotype and a genotype associated with malignant hyperthermia, which makes its association a controversial point.[3]

Essentially, all general anesthetics depress mitochondrial function. Volatile anesthetics decrease oxidative phosphorylation and depress breathing. Each class, however, depresses breathing to varying degrees. Isoflurane and desflurane depress the ventilatory response to CO_2 more than sevoflurane. Sevoflurane and desflurane cause more direct muscle relaxation than isoflurane. From the ventilation point of view, sevoflurane appears to be advantageous in patients with mitochondrial defects. Propofol infusion syndrome is a rare but lethal complication in patients undergoing long-term, high-dose propofol infusion. Propofol acts on mitochondria by inhibiting multiple electron transport chain complexes and fatty acid transport and it seems likely that some patients suffering from mitochondrial defects may be susceptible to adverse drug reactions. However, it is not known whether the patients suffering from mitochondrial disease are more likely to have the syndrome.[1]

Triggerless agents such as propofol, barbiturates, etomidate, benzodiazepines, opioids, nitrous oxide, xenon, non-depolarizing muscle relaxants can be safely used. Ketamine is also often very useful in these situations. Although several techniques have already been successfully used, there is no consensus on the ideal anesthetic technique for these patients.[4]

In our patient, we performed previous oxygenation for one minute with spontaneous ventilation under mask and opted for rapid sequence intubation with rocuronium to replace succinylcholine. However, a mistake was made when calculating the rocuronium and a sub-dose of the medication was administered, but fortunately, this did not pose a risk to our management and the patient. The use of rocuronium has one major advantage: any remaining muscle paralysis, can be safely and effectively antagonized by sugammadex.[2]

It has been observed that in some types of myopathy, propofol can trigger major adverse effects. In addition, our patient has an unfavorable history of using propofol in previous cesarean surgery. Therefore, the team opted to use etomidate as it is regarded less harmful. Advantages have been observed in patients with mitochondrial diseases with the use of sevoflurane, as it offers safety in muscle relaxation and depression of the respiratory pattern.[2] Therefore, we chose to maintain sevoflurane anesthesia.

The complications of bariatric surgery are varied. From the anesthetic point of view, pulmonary complications, thromboembolism and hemorrhage are the most common.[5] We observed in our case that the patient evolved without complications, being discharged in the 7th postoperative day. Up to 4 weeks after the surgical procedure, the patient had no new symptoms of the disease.

Mitochondrial myopathy is a rare disease with potential postoperative motor dysfunction. Anesthetic procedures are decisive and will result in potential complications. A thorough preoperative evaluation, a multidisciplinary approach and careful consideration of the function of the organs involved are essential. Although historically the result is challenging, we should not allow the scarcity of studies to influence the negative prognosis linked to this disease.

Conflicts of interest

The authors declare no conflicts of interest.

References

1. Niezgoda J, Morgan PG. Anesthetic considerations in patients with mitochondrial defects. Paediatr Anaesth. 2013;23:785–93.
2. Schieren M, Defosse J, Böhmer A, et al. Anaesthetic management of patients with myopathies. Eur J Anaesthesiol. 2017;34:641–9.
3. Litman RS, Griggs SM, Dowling JJ, et al. MH susceptibility and related diseases. Anesthesiology. 2017;30:1–9.
4. Lerman J. Perioperative management of the paediatric patient with coexisting neuromuscular disease. Br J Anaesth. 2011;107:79–89.
5. Bagatini A, Trindade RD, Gomes CR, et al. Anestesia para Cirurgia Bariátrica. Avaliação Retrospectiva e Revisão da Literatura. Rev Bras Anestesiol. 2006;56:205–22.