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CASE REPORT

PYELOLITHOTOMY IN PATIENT WITH MUSCULAR DYSTROPHY: MUSCLE RELAXANT - FREE ANESTHESIA

Panovska Petrusheva A[1], Gavrilovska Brzanov A[1], Chavkoska M[2]

[1]University Clinic for Traumatology, Orthopedics, Anesthesiology, Resuscitation, Intensive Care and Emergency Center - Skopje, Department of Anesthesiology, Resuscitation and Intensive Care Medicine, University "Ss. Cyril and Methodius" - Skopje, Faculty of Medicine
[2]Department of Anesthesiology and Intensive Care Medicine, General Hospital of Ohrid

Abstract

Group of genetic conditions that cause the muscles to weaken, are known as Muscular dystrophies (MD). Over time, these inherited and hereditary diseases lead to an increased level of disability. MD is caused by interframe deletion/ duplication, missense or nonsense mutations in the genes responsible for the structure and functioning of a muscle. The increased disability, over time interfere with patient's everyday life and may affect patinet's vital functions. There are many kinds of muscular dystrophy, each of them caused by a different mutation, but the most frequent of them in variety, have something in common, such as begging in childhood, mostly in boys. On the other hand, there are types that don't manifest until adulthood. There is no well-known and accepted cure for monotherapy for muscular dystrophy, but medications and multimodal treatment can help in facilitating symptoms and can slow the course of the disease. The main sign of muscular dystrophy is progressive muscle weakness. Specific signs and symptoms begin at different ages and in different muscle groups, depending on the type of muscular dystrophy. Signs and symptoms usually include frequent falls, difficulty rising from a lying or sitting position, trouble running and jumping, waddling gait, walking on the toes, large calf muscles, muscle pain and stiffness, learning disabilities and delay in growth and development.
This case report is about 43-years-old male patient, diagnosed with a type of muscular dystrophy and right kidney nephrolithiasis, admitted for a classic pyelolithotomy.

Key Words*: muscular dystrophy, muscle relaxant, pyelolithotomy.*

Introduction

Muscular dystrophies are a group of genetic diseases which cause progressive degeneration of skeletal muscle along with weakness (1-3). Different types of muscle diseases are associated with different mutations which interfere with the function of genes that are necessary for muscle contraction and function. Duchenne's and Becker's muscular dystrophies are caused by a

mutation in the DMD gene, which codes the dystrophin, a protein included in myocyte's protection during movement. These types of changes may be an interframe deletion/ duplication, missense or nonsense mutations, but, commonly, all of them are inherited from the parents. Much less often, they may occur spontaneously, named as de-novo mutations (1). Even though epidemiology of MD says that they have prevalence rates between 1 and 10 per 100,000 population, we have to mention that this prevalence is rising and can be present in patients for anesthesia. This group of patients has to be evaluated by the anesthesiologist, especially for complications that may happen because of the muscle disorders. These people may experience weakness of the musculature with reduced strength of the respiratory muscles and the possibility of increased sensitivity to drugs with a neuromuscular mechanism of action, which predisposes them to complications, both during the procedure itself and the recovery period. Complications can be related to the anesthesiologic treatment itself (drugs which are commonly used during anesthesia), and the most often they affect cardiovascular and respiratory system. Recovery period may be followed by difficulty in movement and walking, which leads to increased risk of traumatic events, such as falling and injuries (1). Introduction and conduction of anesthesia have to be precisely administered in the patients with muscular dystrophy, especially considering the fact that the muscle relaxants, directly affect neuromuscular junction and muscle contraction. Preoperative, a detailed neurological assessment is required in order to better insight the risk during surgery and anesthesia. It is necessary to confirm the diagnosis if possible, as well as to determine the degree of disease progression in each patient (2). The preoperative management also requires respiratory and cardiovascular evaluation due to the possibility of concern of both. The assessment of respiratory function should include accurate medical documentation, physical examination, lung radiography, evaluation of sleep-disordered breathing, as well as measurement of respiratory function and cough efficiency (3). In all patients, it is necessary to have an electrocardiogram and an echocardiogram performed before anesthesia or sedation if this has not been done in the previous 12 months (4).

Case Presentation

Our patient was 43-years-old male, 70kg by weight, height of 165cm, smoker, with diagnosed muscular dystrophy, according to molecular genetic analysis, which has shown 60% deletions and duplications to the Dystrophin gene.
He was admitted to our Urology department, one week ago, with pain in his right lumbar quadrant, and he was diagnosed with nephrolithiasis. The patient was posted for right nephrolithotomy. According to the already mentioned history of muscular dystrophy, the patient was induced and maintained with TIVA. Previously, we had an insight into his respiratory function, which was unaffected at that moment, without absolute cardiologic contraindication for surgery. The patient did not have any allergies to food and medicaments, using no chronic therapy, laboratory well arranged. Before induction, his vital parameters were monitored, his blood pressure was 130/70mmHg, heart rate 60/min and saturation 98% without oxygen mask.

After preoxygenation for 3 minutes with 8L oxygen, the patient was premedicated with 2mg Midazolam i.v., 0.1mcg Fentanyl and inducted to general anesthesia with 150mg Propofol i.v. bolus. TIVA was maintained with continuously use of Remifentanil 0.05mg/kg and Propofol 0.05mg/kg/h. The patient was intubated without use of muscle relaxant, with video laryngoscope and there was placed ETT number 8. After induction and intubation, we had stable parameters, with a slight decrease in blood pressure, which was 100/65mmHg, no change in heart rate and 100% saturation. The stability of parameters was maintained throughout the operation, and parameters were in rank 100-130mmHg for systolic pressure and 60-80 mmHg for diastolic. Heart rate was about 57-60 beats per minute and blood oxygenation 99-100%, with monitored capnography which showed etCO2 29. Intraoperatively, we administered ampoule Dexason 8mg i.v., with adequate gastroprotective therapy with Famotidine 20mg and Metoclopramide 10mg, non-opioid analgesia was provided with Acetaminophen 1g i.v. Fluid replacement was with 1500ml NaCl 0.9%. Three hours later, surgery was done, the patient awakened and was transported to the recovery room, all well and pain free. Postoperative pain management was with NSAIL, antibiotic, gastroprotective and anticoagulant therapy.

Discussion

Difficult intubation, prolonged duration of neuromuscular block, impossibility of extubating due to muscle weakness and postoperative ventilation, potential rhabdomyolysis and arrhythmias when exposed to halogenated volatile anesthetics and depolarizing muscle relaxants, are all anesthetic worries and possible events (4). In the case of motor neuron and peripheral nerve diseases, the use of volatile anesthetics is possible, while the use of succinylcholine is absolutely contraindicated (12, 25). In patients with a disorder of the neuromuscular junction general anesthesia maintained with volatile anesthetics is considered safe, until MAC is less than 1. Using inhaled anesthetics and succinylcholine can be highly risky for malignant hyperthermia or acute rhabdomyolysis in all previously mentioned patients (5). Avoiding the use of succinylcholine and halogenated anesthetics in these patients is general recommendation. Nondepolarizing neuromuscular relaxants may show a prolongation of neuromuscular block, no matter of their short activity. There are several reports that recommend avoiding these relaxants whenever possible (6). If their use is necessary, then it is preferable to reduce the doses and use TOF (train-of-four) monitoring (7). Reversion of neuromuscular block, if used, is also challenge, according to the fact that anticholinesterase drugs are not recommended in patients with MD, due to possibility to predicate hyperkalemia, the use of sugammadex as an antidote to rocuronium may be beneficial, according to reduced risk of postoperative residual muscle paralysis. The combination of rocuronium and sugammadex can replace the use of succinylcholine in rapid sequence intubation of patients with neuromuscular dystrophies (8).

Conclusion

Due to the rarity and peculiarities associated with MD, there is no established superior anesthetic technique. The approach will always be guided by the type of surgery and clinical status of the patient, with the primary target being greater patient's safety. Total intravenous anesthesia with propofol and remifentanil administered by continuous infusion without neuromuscular blockers is a safe and effective option for MD patients.

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