Case report

Myotonic Dystrophy and Volvulus: Anesthetic Considerations For an Urgent Situation And the Role of Sugammadex; A Case Report

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Abstract

Myotonic dystrophy (MD), although an uncommon disorder, is challenging for anesthesiologists as it restricts medication choices and involves organs other than muscles. Here we report a known case of an MD who underwent laparotomy for bowel obstruction. Anesthesia was induced rapidly, and muscle relaxation was achieved by using a high dose of rocuronium. Reversal muscle relaxation was successfully done by using sugammadex with no residual relaxation. Rocuronium could be used as a safe means of rapid muscle relaxation in MD, and the complete reversal could be achieved with sugammadex.

Keywords: Myotonic dystrophy, Steinert disease, Sugammadex, Rocuronium

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Introduction

Myotonic dystrophy (MD) is an autosomal dominant hereditary disorder that involves not only skeletal but also cardiac and smooth muscles (1), gonads, and eyes (2, 3). Provided that muscles responsible for vocal cords coordination get involved, it may result in aspiration, respiratory failure, and eventually grave morbidities (2). The most common form is type 1 MD, due to CTG trinucleotide repeat in protein kinase gene on chromosome 19q13.3 that usually leads to symptoms in 2nd and 3rd decades of life span. General anesthesia may have harmful effects on these patients since it may require muscle relaxants, which they are sensitive to (2). It could result in myotonia crisis, need for prolonged ventilation secondary to hypoventilation, respiratory failure, prolonged recovery time dysrhythmia, and even death (4-6).

Most reports about MD and anesthesia explain an elective procedure for a prepared patient, while here we report an urgent case of an MD who suffered volvulus.

Case Report

A 25 years old man who was a known case of myotonic dystrophy (MD), with abdominal pain, nausea, vomiting, and abdominal distention, was admitted to our hospital. An X-ray study confirmed volvulus and bowel obstruction, so the patient was urgently transferred to the operation room (OR). Physical examination showed frontal balding (Figure 1), decreased distal muscular tone (Figure 2) of extremities, slurred speech, muffled sound, and mental retardation.

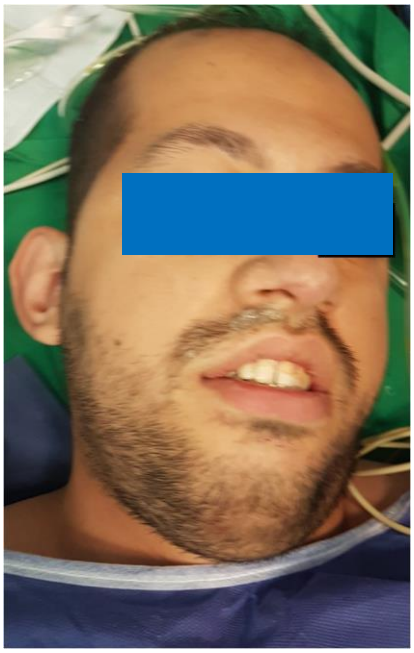

Figure 1. Frontal balding.

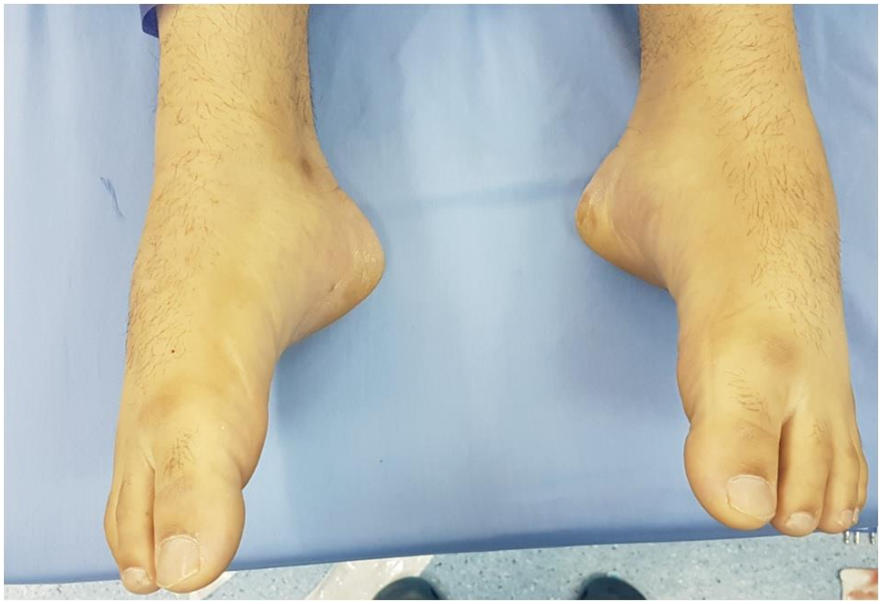

Figure 2. Decreased distal muscular tone.

His 28-year-old elder sister also suffered from MD; his mother suffered from myasthenia gravis and passed away four years ago. In OR, vital signs and hemodynamic indices were in the normal range. An electrocardiogram (ECG) showed a right bundle branch block (RBBB). He received intravenous (IV) fentanyl 1.5 µg/Kg as premedication. We had planned rapid sequence induction of anesthesia, but MD mandates omitting succinylcholine. Therefore, the anesthesia was induced by propofol 2 mg/Kg/IV and rocuronium 1.2 mg/Kg/IV. Intubation, anesthesia maintenance, and surgery, which lasted two hours, were uneventful. At the end of the surgery, sugammadex, 4 mg/Kg/IV, was injected, and Train of Four (TOF) showed 0.9 after 3 minutes. The tube was removed from the trachea, and the patient was transferred to the post-anesthesia care unit. Then discharged to ICU to be monitored closely for possible

adverse respiratory events while fully awake with competent spontaneous breathing.

Discussion

Although rare, myotonic dystrophy could challenge anesthesiologists since it interferes with neuromuscular blocking medications. Succinylcholine must be avoided in these patients. Rocuronium and sugammadex are wise choices to provide the desired level of muscle relaxation and reversal.

Myotonia means maintained musculature tone after voluntary contraction with delayed muscle relaxation (2, 3), which may happen at rest or after exercise (7). Myotonic dystrophy (MD) is an autosomal dominant inherited disorder with two subtypes. MD1 or Steinert disease secondary to trinucleotide (CTG) repeat in dystrophic myotonia protein kinase (DMPK) gene on chromosome 19q.13.3 that involves distal muscle. Usually, symptoms and signs appear in adult-affected patients (2, 3). MD2 is due to mutations in cellular nucleic acid-binding protein (CNBP) on chromosome 3q.21 that mainly affect proximal muscles in children (3). Ninety-eight percent of patients are type 1 (3), and it is estimated that 1 in 8000 births carry the mutation and 2 to 14 in 100000 population show symptoms and signs (8). MD1 patients usually suffer muscle weakness and loss, which may include cranial muscles and lead to the involvement of vocal cords (2). Patients may also have frontal balding, testicular atrophy, insulin resistance, mitral valve prolapsed and cataracts, (2, 3) cardiac conduction abnormalities, and cardiac muscle involvement (1, 2). For a long time, it was mistakenly considered to be linked to malignant hyperthermia, but recently it was realized that there is no direct link between them (9).

Although the disease name refers to muscular involvement, some authorities believe that neuromuscular junction (NMJ) may also be a part of the problem (10). The explanation for NMJ involvement is that the affected muscle responds to succinylcholine, neostigmine, and acetylcholine (Ach) surge in NMJ. Secondary to motor nerve damage (axonal neuropathy) and reinnervation, multiple endplates may form on muscle fibers; therefore, after succinylcholine administration or Ach surge in NMJ, these multiple endplates make action potentials on the muscle fiber surface and lead to accommodation and muscle contraction which does not lead to muscle relaxation but prolonged muscle contraction. Depolarizing muscle relaxants are hazardous to MD patients since they may elicit muscle spasms and hyperkalemia. In contrast, non-depolarizing muscle relaxants (NDMR) show normal responses (3). Therefore, a vital anesthetic consideration for MD patients is avoidance of succinylcholine and judicious use of NDMRs. Opioids are another source of controversy as they might provoke muscle rigidity; some authorities recommend omitting this class of medication from the list to diminish respiratory complications (2, 8), while others have reported its use uneventfully (3). We only administered fentanyl as premedication, and no muscle rigidity was seen.

Most of the reports spotlighted omitting muscle relaxants even for tracheal intubation before elective procedures (3) or using different volatile anesthetics to achieve a rapid and safe emergence (2). Recently, reports about using sugammadex as the reversal agent of choice for NDMRs have been published, but most of them emphasize elective procedures (3) since cholinesterase inhibitors may exacerbate myotonia.

The challenge we encountered was an urgent case with a high risk for gastrointestinal content regurgitation and pulmonary aspiration due to volvulus resulting in bowel obstruction. Awake tracheal intubation under topical anesthesia of the glottis and airway was not a wise choice since it blocks the protective reflexes of the airway and increases the danger of aspiration. The usual approach is rapid sequence induction using succinylcholine to achieve an immediate relaxation of muscles and ease of laryngoscopy and tracheal intubation, but our patient's situation disqualified this option. The only wise choice that remained was a high dose of rocuronium to induce rapid muscle relaxation and using sugammadex to reverse the remaining effects of rocuronium as its high dose will make a long-lasting muscle relaxation and MD mandates omitting neostigmine.

Fortunately, the post-operative period was uneventful, and the patient was discharged from ICU to the general ward and then home. We believe this report shows the value of the combination of rocuronium and sugammadex, which enabled us to

avoid succinylcholine and cholinesterase inhibitors.

Conclusion

Although rare, MD is a serious challenge to anesthesiologists, so we should be familiar with the disease and the pros and cons of the medications we use. We believe that our report shows the importance of the availability of sugammadex in the operating theatre for the safe management of MD.

Acknowledgment

None.

Conflicts of Interest

The authors declare that there are no conflicts of interest.

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