Anesthesia for laparoscopic surgery in a patient with Myotonic Dystrophy (Steinert's disease): beneficial use of sugammadex, but incorrect use of pethidine: A case report

P. MAVRIDOU, V. DIMITRIOU, A. MARGARITIS and A. MANATAKI

Abstract: Patients with Myotonic Dystrophy show an unpredictable response to several anesthetic drugs including opioids, neuromuscular blocking agents and especially reversal agents like neostigmine. We describe the case of a 40 year old patient with myotonic dystrophy who underwent laparoscopic cholecystectomy and ovarian cyst removal under general anesthesia. The authors suggest the use of the new reversal agent sugammadex, for reversing neuromuscular blockade caused by rocuronium, in patients suffering from neuromuscular disease and especially from Myotonic Dystrophy, because it rapidly and completely reverses any residual neuromuscular blockade, but also underline the increased susceptibility of these patients to opioids.

Key words: Myotonic Dystrophy; congenital; sugammadex; opioids.

INTRODUCTION

Myotonic Dystrophy (MD) is the most common disorder of the dystonias (prevalence 1:8000/1:20000), the others being myotonia congenita and paramyotonia. It is an autosomal dominant disorder, usually localized to chromosome 19 and diagnosis is confirmed by genetic tests. MD is typically presented in the second to fourth decade of life with myotonia (persistent contraction and slowing of relaxation after muscle contraction in response to electrical or percussive stimuli) being the principal manifestation early in the disease. Factors that may trigger myotonic reaction are hypothermia, shivering, and mechanical or electrical stimulation. As the disease progresses, muscle weakness and atrophy become more prominent, usually affecting cranial muscles (orbicularis oculi and oris, masseter and sternocleidomastoid) and result in the typical facial appearance. As opposed to most myopathies, distal muscles are more involved than proximal muscles. Multiple organ systems are involved in the disease as evidenced by presenile cataracts, premature frontal baldness, hyperinsomnia with sleep apnea and endocrine dysfunction leading to pancreatic, adrenal, thyroid, and gonadal insufficiency. In addition these patients are predisposed to pulmonary aspiration due to gastric hypomotility with a diminished cough reflex. Respiratory involvement leads to decreased vital capacity while pulmonary or central nervous system dysfunction lead to alveolar hypoventilation. Cardiac manifestations are often present before other clinical symptoms appear. Degeneration of the cardiac conduction system causes dysrhythmia, mostly atrial, varying degrees of heart block and less frequently, depression of ventricular function. Mitral valve prolapse occurs in 20% of patients.

The anesthetic management of patients with myotonic dystrophy can be challenging. Cardiorespiratory complications, pulmonary aspiration and myotonia aggravation are the main anesthetic considerations. An unpredictable response to several anesthetic drugs including neuromuscular blocking agents and especially reversal agents is responsible for most of the anesthetic problems (1). Sugammadex is a relatively new, unique reversal agent that encapsulates and deactivates rocuronium and vecuronium. It reverses completely and effectively any residual neuromuscular blockade, thus it could be extremely useful in patients with MD. As far as we know, until today only two cases have been reported considering patients with MD receiving sugammadex.

Paraskevi MAVRIDOU, Staff Anesthesiologist, M.D.; Varvara DIMITRIOU, Resident Anesthesiologist, M.D.; Athanasios MARGARITIS, Resident Anesthesiologist, M.D.; Adamantia MANATAKI, Consultant Anesthesiologist, M.D.

Anesthesiology Department, Hatzikosta General Hospital, Ioannina, Greece.

Correspondence address: Dr. Paraskevi Mavridou, Hatzikosta General Hospital, Anesthesiology Department, Makriyianni Avenue, 45001, Ioannina, Greece. Tel.: +302651080537. Fax: 00302651080538. E-mail: voulam@otenet.gr

© Acta Anaesthesiologica Belgica, 2011, 62, n° 2
CASE REPORT

A 40 year old female patient (body weight: 74 kg, height: 160 cm) was scheduled for laparoscopic cholecystectomy and right ovarian cystectomy. The first symptoms of the disease developed at the age of 17, with lower limb numbness and cramps. At the age of 35 she began to present symptoms of lower limb fatigue and weakness and the disease was confirmed at the age of 38 with genetic tests. Family history was free.

Previous spinal anesthesia in our hospital (2 years ago) for loop electrosurgical excision procedure to treat high grade cervical dysplasia was uneventful.

Preoperative assessment showed that the patient had the typical facial appearance of the disease due to weakness and atrophy of cranial muscles and incipient frontal baldness. Muscle strength of distal muscles in both upper and lower limb was slightly affected. Minor walking impairment and easy fatigue were observed.

Electrocardiograph (ECG), chest X-ray and echocardiography were normal. Cardiologic medications were not prescribed. Other laboratory and biochemical test results were within normal ranges. Pulmonary physical examination by specialist and spirometry, revealed mild mixed pulmonary disorder and the patient received salbutamol for three days preoperatively. Auscultation did not reveal any abnormal breath sounds. SpO₂ was 97%.

The operation theatre was adequately heated before the patient’s admission and warm intravenous fluids were administered during the procedure. Monitoring consisted of electrocardiogram and non-invasive blood pressure measurement, pulse oxymetry, capnography and body temperature. For monitoring of muscular relaxation the train-of-four (TOF) (Watch SX, Schering-Plough Ireland, Dublin, Ireland) was used at the adductor pollicis muscle. Repetitive TOF stimulation was applied at the ulnar nerve until the end of anesthesia. Ondansetron and omeprazole were administered to the patient 30 min before anesthesia induction.

Anesthesia was induced with propofol at a dose of 2 mg/kg. For complete muscle relaxation a rocuronium dose of 30 mg was required (titrated by TOF indication) followed by intubation of the trachea. A gastric tube was inserted. Anesthesia was maintained with O₂ and air mixture at 50% and continuous infusion of propofol (4 mg/kg/h) and remifentanil (0.2 µg/kg/min). No additional dose of rocuronium was required. The patient underwent videolaparoscopy with insufflation with carbon dioxide to induce pneumoperitoneum. The intra-abdominal pressure was maintained under 12 mmHg. The gall bladder and a large ovarian cyst were laparoscopically removed. Thirty minutes before the end of the procedure pethidine (50 mg iv) was administered for postoperative analgesia. The duration of the procedure was 90 min without any undesirable events.

After the end of the procedure at reappearance of T2, sugammadex 2 mg/kg was administered to the patient. In 2 minutes time, TOF was 1 but the patient was in respiratory depression and she did not respond to simple verbal commands. The patient remained under mechanical ventilation and 15 min later there was no clinical improvement, we decided to administer 0.4 mg naloxone iv, presuming that the patients’ sedation and respiratory depression was due to pethidine.

After receiving naloxone the patient gradually started regaining adequate spontaneous ventilation. She was extubated approximately 10 minutes after naloxone administration when we confirmed that she was able to respond to simple commands, to open her eyes, lift her head and squeeze the anesthesiologists’ hand.

The patient was transferred to the Postanesthesia Care Unit (PCU) where she received 100% O₂ and salbutamol through ventilation mask. During spontaneous breathing with constant stimulation SpO₂ was approximately 90-92%. When the patient started complaining about pain she received tramadol (50 mg, iv) and lornoxicam (8 mg, iv). After remaining in the PCU for 3 hours, we decided that she could be transferred to a surgical clinic ward with thorough postoperative instructions.

There were no postoperative complications and the patient was discharged from the hospital the third postoperative day in good condition and fully satisfied.

DISCUSSION

Patients with MD have altered responses to a number of anesthetic medicines. Anesthetic complications in MD patients occur in approximately 8.2% and are mainly pulmonary, especially in patients undergoing upper abdominal surgery and in those who have more severe muscular weakness (2). In another study the complications rate is up to 52% (3). One death has been reported (after 391 days in the ICU) of a patient with severe, but undiagnosed MD (4).

Laparoscopic surgery has the advantages of fast postoperative recovery, minimizing pulmonary
Complications, which is very helpful when dealing with MD patients. Laparoscopic surgery has been widely used with (5, 6), or without neuromuscular blockers (7) or even under epidural analgesia (8).

Regional anesthesia is preferable for patients suffering from MD. However, when general anesthesia is required, we ought to take under consideration some necessary precautions as far as the drugs we use are concerned. A number of anesthetic agents have been used for induction (thiopental, inhalation agents, propofol). All drugs should be administered cautiously, with a careful dose titration. Propofol has been used in many cases safely, for induction and maintenance of anesthesia (9, 10). While propofol is considered a safe choice, there are reports of myoclonic movements occurring after propofol administration (11). Inhalation agents have been associated with postoperative shivering, especially combined with low operation room temperature, which can trigger myotonic contractions. However, the safe use of sevoflurane has been described (12). Although propofol has not been proven to be superior to inhalation agents in MD, we chose to administer propofol, motivated by the fear of postoperative shivering.

Suxamethonium (succinylcholine) should be avoided because it produces prolonged muscle contraction and rigid jaw, which may make intubation, ventilation and surgery difficult (13, 14). In patients with MD it is preferable to use short or medium acting non depolarizing neuromuscular agents. Atracurium (15), cis-atracurium (16), mivacurium, vecuronium (17) and rocuronium have been safely used. The reversal agents for these drugs should be avoided, because they can cause unpredictable reactions to patients with MD. However, neostigmine has been used without any complications (14). We chose to use rocuronium since a special reversal agent, sugammadex, is now available offering fast and effective muscle blockage reversal, ideal for patients suffering from MD. We believe that sugammadex is a useful aid for reversing muscle blockade and since it has been used in few patients with MD, further studies are required in order to come to some safe conclusions about its use. To the best of the authors’ knowledge two more cases of successful sugammadex administration have been reported until now (18, 19).

An increased susceptibility to opioids is a common feature of the disease, thus their administration should be done carefully. Opioids can cause respiratory depression and prolonged sedation. Continuous infusion of remifentanil (the drug of our choice) has been used intraoperatively with good results (16), mainly due to its short acting properties and its metabolic pattern. Fentanyl (9) and alfentanil have also been used successfully. The epidural use of opioids has been reported. However, epidural administration of morphine has caused serious respiratory depression (20).

In our case we believe that the administration of pethidine was responsible for the sedation and respiratory depression observed in our patient. The pethidine dose we used in this patient, following our protocol for postoperative analgesia, proved to be excessively high. Pethidine’s action was successfully reversed by administering naloxone. Analgesia is best provided by other drugs such as nonsteroidal anti-inflammatory drugs (NSAIDs), in order to avoid the systemic depressant effects of opioids.

In conclusion we would like to highlight two main points: first, the effective and safe use of sugammadex in this patient with MD which provides fast and adequate recovery from rocuronium induced neuromuscular blockade, TOF being normal within two minutes of sugammadex administration and second, the need for careful administration of opioids for postoperative analgesia. Long acting and powerful morphinomimetics should be avoided and other drugs, which don’t cause respiratory depression, should be preferred. Above all a good knowledge of the special features of the disease is required in order to plan a proper anesthetic strategy, individually modified for each patient with MD.

References


© Acta Anaesthesiologica Belgica, 2011, 62, n° 2