Clinical Case Report

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Opioid-free general anesthesia in patient with Steinert syndrome (myotonic dystrophy)

Case report

Tomasz Gaszynski, MD, PhD*

Abstract

Introduction: We report on the anesthetic management using opioid-free method of a patient with Steinert syndrome (myotonic dystrophy, MD), autosomal dominant dystrophy which is characterized by consistent contracture of muscle following stimulation. A myotonic crisis can be induced by numerous factors including hypothermia, shivering, and mechanical or electrical stimulation. In patients with MD, hypersensitivity to anesthetic drugs, especially muscle relaxants and opioids, may complicate postoperative management. If opioids are employed (systemic or neuraxial), then ICU care and continuous pulse oximetry must be considered given the high risk for respiratory depression and aspiration. Patients with MD present high sensitivity to the usual anesthetics such as volatile and muscle relaxants (both depolarizing and nondepolarizing). Opioids may induce muscle rigidity in this type of MD. Therefore, omitting opioids is recommended. Due to hypersensitivity to opioids and increased susceptibility to intra- and postoperative complications, it is recommended to introduce opioid-free anesthesia (OFA), for example, with use of dexmedetomidine (DEX). This is a new method of conducting general anesthesia without opioids and is based on concept of multimodal approach to pain management.

Methods: A 31-year-old male patient (183 cm, 69 kg) was scheduled for laparoscopic operation of cholecystectomy. The patient received intravenously (IV): propofol in a dose of 250 mg followed by continuous infusion, rocuronium in a dose of 20 mg, and DEX in a loading dose of 0.6 μg/kg over 10 minutes followed by continuous infusion of dose of 0.2 μg/kg/hour.

Results: The course of anesthesia and postoperative period were uneventful. The patient exited the operating theatre in a good medical state, with vitals within normal limits and fully regained consciousness.

Conclusion: DEX is effective and safe for moderately painful procedures in patients with the elevated risk of respiratory and cardiovascular failure. This substance provides adequate analgesia level during surgeries of patients suffering from MD.

Abbreviations: CPAP = continuous positive airway pressure, DEX = dexmedetomidine, MD = myotonic dystrophy, NSAIDs = Non-steroid anti-inflammatory drugs, OFA = opioid-free anesthesia.

Keywords: dexmedetomidine, general anesthesia, myotonic dystrophy, opioid-free anesthesia

1. Introduction

We report on the anesthetic management using opioid-free method of a patient with Steinert syndrome (myotonic dystrophy, MD), autosomal dominant dystrophy which is characterized by consistent contracture of muscle following stimulation. An abnormal nucleotide sequence on chromosome-19 causes prolonged stimulation of the actin–myosin complex due to a larger sodium current, causing delayed relaxation of contracted muscle. It may manifest in early childhood and is a multisystem disease. The estimated incidence is 1 in every 8000 births, with an estimated prevalence of between 2.1 and 14.3 cases per 100,000 inhabitants.

Providing a successful anesthesia to patients suffering from Steinert syndrome (MD) constitutes a serious challenge. These difficulties are an implication of multiple disorders which may be related to respiratory, cardiac, and central nervous systems. A myotonic crisis can be induced by numerous factors including hypothermia, shivering, and mechanical or electrical stimulation. In patients with MD, hypersensitivity to anesthetic drugs, especially muscle relaxants and opioids, may complicate postoperative management. Patients may present poor outcomes related to the following complications: loss of airway secondary to medication-induced respiratory depression, aspiration of stomach contents, and sudden death that is usually secondary to cardiac conduction delays and dysrhythmias.

Myotonia is described as muscle contraction (voluntary or otherwise) with abnormal, prolonged relaxation.[1] Triggers for myotonia include certain medications, potassium, hypothermia, shivering, or any mechanical or electrical stimulus.[1–3] Patients also exhibit profound skeletal muscle weakness secondary to muscle degeneration. Dystrophia myotonica (DM, myotonic dystrophy) patients are exquisitely sensitive to the respiratory depressant effects of anesthetic medications.[1] Postoperative pain control should be managed with non-steroid anti-inflammatory

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Department of Anaesthesiology and Intensive Therapy, Medical University of Lodz, Lodz, Poland.

* Correspondence: Tomasz Gaszynski, Department of Anaesthesiology and Intensive Therapy, Medical University of Lodz, 90-419 Lodz, Poland (e-mail: tomasz.gaszynski@umed.lodz.pl).

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drugs (NSAIDs), regional techniques using local anesthetics, and acetaminophen when possible. If opioids are employed (systemic or neuraxial), then ICU care and continuous pulse oximetry must be considered given the high risk for respiratory depression and aspiration.

Rapid sequence induction with cricoid pressure is recommended. Weakness of the pharyngeal muscles and a delayed gastric emptying time predispose DM patients to aspiration.[1,4] Also, succinylcholine effects are unpredictable in DM patients. Succinylcholine should be omitted.

Respiratory muscle weakness predisposes MD patients to restrictive lung disease with concurrent dyspnea and ineffective cough.[1] Ventilatory weakness contributes to the complex sleep disorders of DM. DM subjects have heightened central nervous system sensitivity to sedatives, anxiolytics, and analgesics, further impeding ventilatory drive and airway protection. It is not uncommon for DM patients to have a history of hypoxia, dyspnea, sleep apnea requiring continuous positive airway pressure (CPAP), or marked ventilatory muscle weakness necessitating ventilation support, DM patients can be exquisitely sensitive to the respiratory depressant effects of commonly used premedications (e.g., opioids and benzodiazepines).

Patients with MD present high sensitivity to the usual anesthetics such as volatile and muscle relaxants (both depolarizing and nondepolarizing). Opioids may induce muscle rigidity in this type of MD. Therefore, omitting opioids is recommended. Therefore, a choice of adequate drug with minimal risk of complication remains a serious problem. The exquisite sensitivity of DM patients to the respiratory depressant effects of opioids (systemic or neuraxial) can equate to fatal outcomes in the postoperative period. The most common route of opioid administration that places DM patients at high risk for respiratory depression is intravenous, yet there is a case report that details respiratory depression following a small dose of epidural morphine as well.[5] Due to hypersensitivity to opioids and increased susceptibility to intra- and postoperative complications, it is recommended to introduce opioid-free anesthesia (OFA), for example, with use of dexmedetomidine (DEX). This is a new method of conducting general anesthesia without opioids and is based on concept of multimodal approach to pain management. The drugs used instead of opioids for intraoperative analgesia include clonidine, DEX, ketamine, and continuous intravenous infusion of lidocaine. This method was supposed to maintain stability of vital parameters, and prevent undesired events with irreversible effects.

Hypothermia and shivering can induce a myotonic contracture.[2] Therefore, keep the operating room and table warm so that the patient will be better able to maintain a normal body temperature. Using the warmed IV fluids as well as forced-air blankets during surgery is strongly recommended. If a muscle relaxant is needed, then a nondepolarizing agent with a short recovery index should be chosen (e.g., rocuronium, cis-atracurium).[6] DM patients are no more susceptible to the development of malignant hyperthermia than the rest of the general population.[7,8] Neostigmine has been purported to induce myotonia.[9] Sugammadex can be a good option in these patients.

2. Case description

A 31-year-old male patient (183 cm, 69 kg) was scheduled for laparoscopic operation of cholecystectomy. The rapid sequence intubation guidelines were followed during the induction of general anesthesia. The patient received intravenously: propofol in a dose of 250 mg followed by continuous infusion, rocuronium in a dose of 20 mg, and DEX in a loading dose of 0.6 μg/kg over 10 minutes followed by continuous infusion of dose of 0.2 μg/kg/hour. During the intubation, cricoid pressure was maintained. Except hemodynamic parameters, patient's body temperature was monitored carefully and corrected if needed. No opioids were administrated. The anesthesia lasted for 1 hour. All the time patient was warmed by air-blanket. At the end of surgery, 200 mg of sugammadex was administrated to reverse muscle relaxation. The course of anesthesia and postoperative period were uneventful. The patient exited the operating theatre in a good medical state, with vitals within normal limits and fully regained consciousness. Patient's consent was obtained for publication. Ethical approval for this case was not necessary because one of the indications for OFA is hypersensitivity to opioids.

This is the first case of opioid-free general anesthesia using DEX in patient with Steinert syndrome described in the medical literature.

The successful use of DEX as adjunct to regional anesthesia was described in a 53-year-old woman with MD for a total abdominal hysterectomy by Yoshino et al.[10] Combined spinal and epidural block was used in this patient and DEX was used for sedation during surgery. Airway obstruction was observed after the initial administration of DEX at 2 μg/kg, therefore authors concluded that DEX was proved to be useful in this case; however, use of the drug should be carefully started at a low initial dose in patients with MD. We used dose of 0.6 μg/kg and did not observed airway complications.

There are several reports on safe use of DEX for anesthesia in other muscular diseases.[11] Rozmiarek et al reported successful use of a combination of DEX and ketamine to provide sedation and analgesia in a 21-year-old patient with Duchenne muscular dystrophy (DMD) undergoing bone marrow aspiration and biopsy.[11] Kako et al used 2 different doses of DEX for sedation during muscle biopsy in patients with DMD: 1.0 or 0.5 μg/kg was administered as a loading dose over 3 minutes followed by a continuous infusion of 1.0 or 0.5 μg/kg/hour. Ketamine 1 mg/kg was administered along with the DEX loading dose.[12]

The regional anesthesia in DM patients is preferred if possible[13,14] (thoracic epidural even for cholecystectomy[13]); however, in most described cases of general anesthesia of patients with MD, general anesthesia was performed using continuous infusion of propofol.[3,15–17] For analgesia during general anesthesia, remifentanil was used in described cases.[3,15–17] However, remifentanil has been reported to induce hyperalgesia causing difficulties in postoperative pain management, especially after intra-abdominal surgery.[18] We decided to perform OFA, in which we have great experience because of bariatric surgery. In myotonic diseases, use of rocuronium and sugammadex was described.[19,20]

3. Conclusion

DEX is effective and safe for moderately painful procedures in patients with elevated risk of respiratory and cardiovascular failure. This substance provides adequate analgesia level during surgeries of patients suffering from MD.

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