Anaesthetic Management of a Child with Limb-Girdle Muscular Dystrophy

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Limb-girdle muscular dystrophies are a group of disorders with wide genetic and clinical heterogeneity. These disorders may lead to an increase in life-threatening complications related to surgery and anaesthesia. In this case, the anaesthetic management of a child with limb-girdle muscular dystrophy is presented.

Key Words: Child, muscular dystrophies, general anaesthesia

Introduction

Limb-girdle muscular dystrophies (LGMD) are a heterogeneous group of diseases showing autosomal dominant or autosomal recessive inheritance and are characterized by progressive weakness and atrophy in the pelvic and shoulder girdle muscle groups. Although clinical symptoms and pathological findings are frequently seen in the musculo-skeletal, respiratory and cardiovascular systems, it may occur as a systemic disease by central nervous system and gastrointestinal system involvement (1).

Respiratory insufficiency due to muscle weakness; difficult intubation and airway problems; increased risk for pulmonary aspiration, dysphagia and reflux; cardiomyopathy, conduction disturbances and arrhythmias; increased rhabdomyolysis risk in dystrophic myopathies; epilepsy and psychomotor retardation are symptom and signs that may increase anaesthesia related risks (2).

Although it is a rare condition, we aimed to present the clinical experience related to sugammadex and intravenous anaesthesia agent use in a child patient with LGMD, who have special anaesthesia needs.

Case Presentation

An 8-year old male patient, 20 kg in weight was admitted to the hospital. In the assessments performed due to difficulties in walking, running and climbing stairs at 5 years, aspartate aminotransferase (AST), alanine aminotransferase (ALT) and creatine kinase (CK) levels were found to be high. Muscle biopsy revealed deficiencies in alpha and beta sub-units and dystrophin and the patient was diagnosed as having LGMD. CK, AST and ALT levels remained high, although there was improvement in the movements of the patient, who was under steroid treatment for three years. The patient, who was admitted with acute abdominal pain, was taken to surgery for appendectomy by the consent of his parents. The patient was using steroids before the intervention and the preoperative laboratory values were as follows; AST=265 U L⁻¹, ALT=261 U L⁻¹, LDH=1178 U L⁻¹, CK=11361 U L⁻¹, CK-MB=230.3 U L⁻¹, leukocyte count=19.94 10³ uL⁻¹. Before the patient was transferred to the operating room, the anaesthesia machine was prepared with a disposable patient circuit, and the soda lime was flushed with a fresh gas flow rate of 15 L min⁻¹. Dantrolene was brought into the operating room. The patient was premedicated with intravenous 1 mg midazolam given from the venous line immediately after he was taken into the operating room, non-invasive blood pressure, electrocardiogram and pulse oximetry monitoring was started and a temperature measurement probe was placed on the skin. Anaesthesia was induced using 3 mg kg⁻¹ propofol, 0.5 g kg⁻¹ sufentanil and 0.6 mg kg⁻¹ rocuronium. After intubation was performed with a 5.5 F endotracheal tube with a cuff, a 22 gauge (diameter) cannula was placed in

the left radial artery and invasive arterial pressure monitoring was started. A nasopharyngeal temperature probe and a bladder catheter were placed. Total intravenous anaesthesia with propofol infusion (total dose=100.6 mg) and bolus doses of sufentanil (total dose=10 mcg) was used for the maintenance of anaesthesia. Before induction and during the intervention, a total of 10 mg methylprednisolone was intravenously infused. End tidal carbon dioxide levels were monitored. During the surgery, heart beat rate was recorded as 85-118 beats min^{-1} , mean blood pressure as 69-90 mmHg, skin temperature as 33.9-34.4°C and nasopharyngeal temperature as 36.2-36.4°C. After the surgical intervention that took 16 minutes, the patient was given intravenous sugammadex 2 mg kg^{-1} (40 mg) and extubated. The patient who showed a rapid recovery approximately within 2 minutes, was applied 2 mg kg^{-1} of tramadol for postoperative analgesia, and was transferred to the intensive care unit. The laboratory tests at postoperative 2 hours were as follows; LDH=685 U L^{-1} , AST=198 U L^{-1} , ALT=205 U L^{-1} , CK-MB=106.2 U L^{-1} , and myoglobin=242.54 ng mL^{-1} . The patient, who was followed up in the intensive care unit for approximately 4 hours, was taken to the ward and discharged from the hospital at postoperative 2 days without any problem.

Discussion

The susceptibility of patients with muscular dystrophy to sedative, anaesthetic and neuromuscular blocking agents may lead to intraoperative and early postoperative cardiovascular and respiratory complications as well as prolonged recovery. Although risk of malignant hyperthermia is not increased in these patients in comparison to the general population, muscular dystrophy patients who receive inhalation anaesthetics may rarely develop a malignant hyperthermia-like syndrome with acute rhabdomyolysis. As in any other muscular diseases, succinylcholine use may lead to life threatening hyperkalaemia, and therefore should not be used (3).

Safe anaesthesia techniques have been defined for these patients. Anaesthesia protocols that do not involve neuromuscular and inhalation agents but involves short-acting agents like propofol, fentanyl and remifentanyl are included among these (4, 5). Among anaesthetic agents, propofol does not activate ryanodine receptor Ca^{+2} release channels in patients who are susceptible to malignant hyperthermia and does not cause contractures during in vitro testing; therefore it is accepted that it can be used safely in place of inhalation agents (6). In LGMD, which is a progressive disease, nocturnal hypoventilation may develop before the development of significant respiratory failure. Therefore, propofol providing a rapid recovery, with no persistent effect on the respiratory system and more easily titrated depth of anaesthesia, is used in patients with LGMD (4). Although propofol and remifentanyl like short-acting agents are recommended to be used as they do not lead to postoperative respiratory failure and prolonged sedation, have their own metabolism independent of liver

and kidney functions, suppress hemodynamic responses during laryngoscopy and surgery and provide decreased awareness; sufentanil was preferred to be used in this patient, as its analgesic effect continues in the postoperative period, and provides a better suppression of cardiovascular responses to laryngoscopy and intubation in comparison to fentanyl (5, 7). A rapid recovery was achieved with a stable hemodynamic course during the surgery without any residual effect and with contribution to postoperative analgesia.

Sugammadex is a modified cyclodextrin that encapsulates and inactivates steroidal neuromuscular blockers, introduced into clinical use in the recent years. Its use in patients with muscular dystrophy is gradually increasing (8). It is possible to encounter difficult intubation and airway problems in these patients (2). As the patient would undergo abdominal surgery, rocuronium was used to facilitate the intubation and the intervention. A rapid recovery was provided with sugammadex in our patient. In interventions where neuromuscular blockade is mandatory, considering the difficulties in airway management, rocuronium antagonized by sugammadex can reliably be used without being concerned about postoperative residual effects.

Although dantrolene was provided for the possibility of malignant hyperthermia, the role of dantrolene in rhabdomyolysis caused by anaesthetic agents is not known. The mechanism of action of dantrolene in malignant hyperthermia is inhibiting the excess Ca^{+2} release from the sarcoplasmic reticulum by binding to the ryanodine receptor isoform 1. However, as it involves mechanisms like damage to the muscle cell membrane and efflux of cell contents, it may not be effective in rhabdomyolysis (9).

Conclusion

In the present case, a painless, pleasant, safe and rapid recovery was provided in a short time period by using propofol, sufentanil and sugammadex. The patient's being clinically diagnosed, allowed us to make anaesthesia arrangements more consciously. However, in patients without a diagnosis, a careful preoperative evaluation may help to choose the most suitable anaesthesia for the individual patient. Additionally, the use of the possible highest standards in perioperative monitoring may make the anaesthesia applications safer.

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