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Anaesthetic Management of Patient with Limb Girdle Muscular Dystrophy Scheduled for Laparoscopic Cholecystectomy

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ABSTRACT

The multiple anaesthetic concerns in a patient with Limb-Girdle muscular dystrophy make the anaesthetic management of the patient difficult. It can be safely managed with adequate pre-operative assessment and careful choice of anaesthetic agents. Our case was managed with general anaesthesia using a short-acting opioid, total Intravenous anaesthesia with propofol and dexmedetomidine avoiding muscle relaxant.

Keywords: *Anesthesia, Intravenous; Dexmedetomidine; Muscular Dystrophies, Limb-Girdle*

INTRODUCTION

Limb-girdle muscular dystrophy (LGMD)is one of the several types of muscular dystrophy with variable age of onset. The anaesthetic concern in such cases includes risk of rhabdomyolysis, development of malignant hyperthermia, malignant arrhythmias during intraoperative period and muscle weakness and respiratory failure in post-operative period.

We present a successful management of a case of young female with limb girdle muscular dystrophy posted for laparoscopic cholecystectomy.

CASE HISTORY

A 17-year-old female, weighing 45 kg was scheduled for laparoscopic cholecystectomy. She was a known case of limb girdle muscular dystrophy diagnosed in the third year of life. Her symptoms predominantly affected

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the lower limbs. Her initial symptoms were bilateral lower limb weakness. Her symptoms progressed slowly with the inability to walk and getting bound to a wheelchair by 12 years of life. Her IQ level was low about her age. She underwent multiple examinations in past with persistent high creatinine kinase levels. There was no history of dyspnea in the past. Her second cousin (male) is also a known case of LGMD.

On examination, she couldn't communicate properly and the history was given by her parents. The lower limb examination revealed wasting of bilateral lower limbs and hypertrophy of bilateral calf muscles. The airway examination showed no findings predictive of the difficult airway. Her vitals were within normal limits. Her blood counts, blood chemistry and urinalysis. Chest X-rays were normal. ECG showed sinus rhythm. On echocardiography, all cardiac chamber/valve morphology was normal with a left ventricular ejection fraction of 65%.

The patient's parents were counselled for risk during the operation and high-risk consent was taken. The intensive care unit bed was booked before surgery.

In the operation room, a 20Gz IV cannula was opened over the dorsum of the right hand and the patient was monitored with ECG, noninvasive blood pressure, pulse oximeter and temperature probe. The patient was given IV dexmedetomidine 1mcg/kg over 10 minutes. She was induced with IV Fentanyl 50 mcg, IV Ketamine 40 mg, Inj.Propofol 60 mg. The laryngoscopy was performed, the patient did not struggle and the vocal cord was opened and the C-L grading was grade I. The trachea was intubated with a 7.0 mm cuffed tube. There was no coughing and no significant hemodynamic changes were noticed. The maintenance was done with an IV Propofol infusion of 75mcg/kg/min and a Dexmedetomidine infusion of 0.4mcg/kg/hr. There were no arrhythmias noted, no rise in temperature and no rise in end-tidal CO_2 level in the intraoperative period. The total duration of surgery was 1 hour 15 minutes. The IV Propofol and Dexmedetomidine were titrated to the hemodynamic parameters and movement of the patient. There was no significant hemodynamic fluctuation and the patient did not move during surgery. The same dose of infusion was continued till the end of surgery. Only after the suturing of all the skin site ports, the infusion of both drugs was stopped. The ventilation mode was changed to pressure support mode with pressure support of 12 cm of H_2O and a flow trigger of 1lt/min. She was extubated after twenty minutes when the patient had a tidal volume of more than 6ml/kg on spontaneous mode, acquired cough reflex and eye-opening on command.

She was managed post-operatively in a high-dependency unit for 72 hours. Her stay was uneventful. The pain was managed with IV ketorolac 30 mg as needed and IV paracetamol 1 gm three times a day. She was shifted to the ward on the fourth postoperative day and discharged on the fifth postoperative day.

DISCUSSION

Limb-Girdle muscular dystrophy is a rare type of neuromuscular disorder involving proximal upper and lower limb muscle weakness with varying degrees of pulmonary and cardiac manifestations. There is a sparing of facial and bulbar muscles in most cases. It is divided into two types, type 1 (Autosomal dominant) and type 2 (Autosomal recessive). [1]The symptom's onset is variable and progression is slower. Systemic involvement can include minor cardiac lesions significant cardiac lesions like dilated cardiomyopathy, and malignant arrhythmias. [2]The preoperative assessment may require an echocardiogram and electrophysiological studies based on cardiac signs and symptoms. Similarly, pulmonary involvement ranges from non to severe pulmonary restriction.[2]Depending on

the patient's history and physical examination, the degree of cardiac and pulmonary involvement needs to be stratified during the pre-operative anaesthetic assessment.

The anaesthetic management is quite challenging in these patients. Regional anaesthesia is preferred over general anaesthesia, whenever it is feasible. The risk of rhabdomyolysis, malignant hyperthermia due to exposure to anaesthetic agents especially inhalation agents, post-operative respiratory compromise due to opioid sensitivity and residual muscle paralysis is omitted in regional anaesthesia. There are case reports where regional anaesthesia has been used successfully, even for cholecystectomy.[3,4] Although there is concern about the myotoxic effect of local anaesthetic especially in the peripheral block, the neuraxial block has been used without any complication. Unfortunately, spinal anaesthesia couldn't be used, as the patient was mentally retarded and could not cooperate during the spinal procedure.

Our plan was general anaesthesia with the short-acting opioid Fentanyl, use of Propofol during induction as well as maintenance and total avoidance of muscle relaxants even during intubation.

The primary concern during the intraoperative period is the possibility of the development of rhabdomyolysis, and malignant hyperthermia due to exposure to initiating agents like succinylcholine and inhalation agents.[5] There are a series of reports of the use of inhalation agents in the case of various types of muscular dystrophy with a low incidence of such events. The authors have argued that even though there is no absolute contraindication to the use of volatile agents in muscular dystrophy, complications if occur, are life-threatening so avoidance of such controversial agents conforms 'principle of beneficence' to the patients.[6] Propofol doesn't trigger malignant hyperthermia and it is advisable to use such agents even for maintenance of anaesthesia so that the patient is not exposed to unnecessary risk. There is increased sensitivity to opioids so the use of a short-acting opioid is advocated.

The use of non-depolarizing muscle relaxants is also best avoided due to high sensitivity and prolonged duration of action leading to potential residual effects during recovery.[5] This possess a challenge during endotracheal intubation without the use of any muscle relaxant. The Dexmedetomidine 1mcg/kg over 10 minutes along with propofol and remifentanil have been used to intubate patients without the use of muscle relaxants and hemodynamic changes.[7] In our case, we use Ketamine, Propofol, Fentanyl and Dexmedetomidine for intubation. The Dexmedetomidine was continued as an infusion of 0.4mcg/kg/hr along with Propofol infusion during the maintenance period to avoid the use of muscle relaxants. The advantages of Dexmedetomidine also include less respiratory depression and analgesic effects.

The patient was also monitored intraoperatively for a sign of malignant hyperthermia using temperature monitoring and a capnogram. The patient's intraoperative as well as post-operative period was uneventful.

CONCLUSION

General anaesthesia for a patient with limb-girdle dystrophy possess a challenge for the anesthesiologist. It can be safely conducted using a short-acting opioid, total intravenous anaesthesia with Propofol and Dexmedetomidine without the use of muscle relaxant.

CONFLICT OF INTEREST

None

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None

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