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Case Report

Management of Patient With Limb Girdle Muscular Dystrophy for ERCP and Regional Anaesthesia for Laparoscopic Cholecystectomy: Case Report

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Abstract

Limb-Girdle muscular dystrophies (LGMDs) are a clinically and genetically heterogeneous group of disorders characterized in general by predominantly limb-girdle weakness. It manifests as proximal muscle weakness involving the pelvic girdle and scapular girdle. We report the anaesthetic management of a patient who is a known case of limb-girdle muscular dystrophy, presented with cholecystitis to our institution. Patient underwent endoscopic retrograde cholangio pancreatography (ERCP) under total intravenous anaesthesia followed by laparoscopic cholecystectomy under spinal anaesthesia which were managed successfully.

Keywords: Muscular dystrophies, Limb-Girdle, Malignant hyperthermia, Anaesthesia

Introduction

Limb-Girdle muscular dystrophies (LGMDs) are a clinically and genetically heterogeneous group of disorders characterized in general by predominantly limb-girdle weakness [1]. It manifests as proximal muscle weakness involving the pelvic girdle and scapular girdle, elevation of the creatine kinase levels and has a widely variable onset as it may present from infancy to adulthood [2]. The estimated incidence of this disease is 1 to 6.5 in 100,000 [3]. Increased risk of rhabdomyolysis and malignant hyperthermia (MH) when exposed to volatile anaesthetics and succinylcholine are life-threatening complications [4]. We report a successful management of a patient known case of limb-girdle muscular dystrophy for endoscopic retrograde cholangio pancreatography (ERCP) and laparoscopic cholecystectomy.

Case Report

A 62-years old male known case of hypertension, type2 diabetes mellitus and bronchial asthma presented to our hospital with complaints of pain in the right upper abdomen and vomiting for 7 days. The pain was acute in onset, colicky, aggravated on eating food and relieved on taking analgesics. He was diagnosed to have autosomal recessive limb girdle muscular dystrophy, 35 years ago. Patient had a history of tibia plating under spinal anaesthesia 6 months back.

On general examination, patient was found to be afebrile with pulse rate of 68/min, blood pressure of 130/90 mm Hg and respiratory rate of 16/min. Cardiorespiratory system examination was normal. On neurological evaluation patient had significant proximal muscle weakness in lower limbs greater than upper limbs. He had left sided foot drop and walked with support (walker). The power in the bilateral extremities was shoulder-4/5, elbow-5/5, wrist-4/5, hip-2/5, knee-2/5 and ankle-2/5. Deep tendon reflexes were absent and superficial reflexes were normal. Neuro physician's fitness was taken. Airway assessment showed Mallampati Class-I.

His electrocardiogram (ECG), chest X-ray and pulmonary function test were normal. 2D echocardiography revealed good left ventricular function with no evidence of RWMA. His complete blood count, blood chemistry and urinalysis were within normal range except liver function test. His total bilirubin was 3.2 mg/dl with direct of 1.7 mg/dl and

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AST/ALT/ GGT were 140 IU/L, 234 IU/L and 238 IU/L respectively. Gastro-enterology reference was given and ERCP was planned. Written informed consent was taken explaining the risk associated with general anaesthesia.

Patient was kept nil by mouth for 6 hours. An intravenous line was secured by 20G cannula in the left hand in the endoscopy suite. Oxygen was administered through nasal prongs at 3L/min. Patient was premedicated with Inj. Midazolam 1mg and inj. Fentanyl 50 micrograms. For induction Inj. Propofol 100 mg and inj. Ketamine 20 mg was administered. Anaesthesia was maintained with continuous propofol infusion 6-10 mg/kg/hour without volatile anaesthetics and patient was maintained on spontaneous ventilation throughout the procedure. Procedure lasted for 45 minutes. Patient was then shifted to post anaesthesia care unit with stable haemodynamics.

After 2 days patient was posted for laparoscopic cholecystectomy. Spinal anaesthesia was planned after discussing with the patient and his relatives about high risk of rhabdomyolysis and malignant hyperthermia associated with general anaesthesia and muscle relaxant in muscular dystrophies. Written informed consent was taken for the same.

Patient was kept nil by mouth for 6 hours. Standard hemodynamic monitors pulse oximeter, electrocardiogram (ECG), non-invasive blood pressure (NIBP)monitor was attached in the operating room. His fasting blood sugar was 135 mg/dl so, intravenous fluid of ringer's lactate was started through 20 G intravenous line. Nasogastric tube no 14F was inserted to prevent the gastric distension. Under all aseptic precautions subarachnoid block was performed with 3.5 ml of 0.5% hyperbaric bupivacaine at the L3/4 level with a midline approach in sitting position using a 25G Quincke's spinal needle after free flow of cerebrospinal fluid. Oxygen was administered with nasal prongs at 3 litres/minute.

The level of sensory (pinprick) block was assessed by the pinprick test using a 24-gauge hypodermic needle, while the motor block level was assessed by the modified Bromage scale and recorded 10 min after placement in the supine position. T4 level of sensory block was achieved after 15 mins and was considered adequate and surgery was commenced.

After port placement, patient was given reverse Trendelenburg position and right sided upward tilt. Gradual pneumoperitoneum insufflation was done with intra-abdominal pressure of 10-12 mm hg and flow of 6 L/min. Intra-abdominal pressure was kept below 10 mmHg, and insufflation flow was kept low at 2 L/min.

To reduce patient's anxiety intravenous Midazolam 1 mg was administered. The patient complained of diaphragmatic discomfort and right shoulder pain intermittently, which was managed using inj. fentanyl 100 µg given in 20 µg aliquots and with also the option of spraying the hepatic surface of diaphragm with 20 ml of 0.25% bupivacaine. IV Ephedrine 6 mg bolus was used to treat hypotension.

Gall bladder dissection was difficult due to inflammatory changes. The cystic duct and artery were ligated, by which time after about 60 minutes in to the procedure the patient started to regain tone of abdominal muscles. Further dissection of the gall bladder from the hepatic fossa became difficult due to patient's active breathing, which prevented maintenance of pneumoperitoneum and therefore loss of visualisation. Propofol infusion was started at 8 mg/kg/hour and surgery was completed uneventfully.

Total 1 litre Ringer's lactate was administered intravenously. At the end of surgery nasogastric tube was removed. Postoperative pain was managed with 1gram intravenous paracetamol and port site infiltration with 20 ml of 0.25% bupivacaine. Postoperatively the patient was satisfied with the anaesthesia.

Discussion

LGMD belongs to the group of progressive muscular dystrophies, characterised by mutation of transmembrane proteins (dystroglycans, sarcoglycans) with dystrophin. Based on its genetic origin, it is classified into limb-girdle MD1 and MD2. Type 1 dystrophy exhibits cardiac manifestations such as malignant arrhythmias that require the implantation of cardiac defibrillators or pacemakers, and type 2, in which 50% of the cases develop dilated cardiomyopathy [5].

The current reports about anaesthetic management of LGMD patients are limited. The risk of rhabdomyolysis leading to hyperkalaemia and cardiac arrest, malignant arrhythmias, increased muscle weakness, airway management problems, and exacerbation of the respiratory failure are associated with LGMD. These patients are also susceptible to Malignant Hyperthermia and prolonged recovery from non-depolarizing muscle relaxants. Patients with muscular dystrophy have an absolute contraindication to receive depolarizing muscle relaxants such as succinylcholine [6].

ERCP was done under total intravenous anaesthesia (TIVA) to avoid volatile anaesthetics and muscle relaxants. Non depolarising muscle relaxants when administered show fast onset, but long duration of effect with high risk of residual paralysis. In the absence of sugammadex, neostigmine for reversal can trigger acute myotony, rhabdomyolysis, malignant arrhythmias and heart failure. Low dose ketamine, midazolam, propofol, etomidate and opioids have been reported as safe options in these patients [7].

Laparoscopic cholecystectomy is generally performed under general anaesthesia with endotracheal intubation and

requires positive pressure ventilation. There was a concern about the ability to wean this patient from the ventilator postoperatively. So, it was planned under spinal anaesthesia. Laparoscopic cholecystectomy under spinal anaesthesia has been described. Sinha et al. published a retrospective review of 3,492 laparoscopic cholecystectomy procedures under spinal Anaesthesia [8]. Findings included a lower incidence of nausea and vomiting and decreased intravenous analgesic requirements. Shoulder and neck pain occurred in 12.3% of patients. Conversion to general anaesthesia was required in 0.5% of cases. Gerbershagen MU et al. suggested regional anaesthetic techniques should be chosen whenever possible in myopathic patients [9]. So far there was no study about myotoxicity after neuraxial anaesthesia. It's preferred to use neuraxial anaesthesia in LGMD patients whenever general anaesthesia had to be avoided or carried an increased risk of complication. avoided or carried an increased risk of complication.

Complications associated with spinal anaesthesia in laparoscopic surgeries are vasovagal attack, persistent hypotension due to increased intra-abdominal pressure, spontaneous ventilation with pneumoperitoneum, hypoventilation and hypercarbia, shoulder-tip pain due to subdiaphragmatic irritation of the peritoneum by the carbon dioxide pneumoperitoneum, and anxiety.

The insufflation pressure used for laparoscopic cholecystectomy (12–15 mmHg) has significant effects on both cardiovascular and respiratory function and may be dangerous in American Society for Anaesthesiologists physical status classification III and IV patients with severe restrictive disease. The use of low-pressure pneumoperitoneum at 6–8 mmHg has been shown to reduce adverse haemodynamic effects [10]. In our case we also kept IAP less than 10 mm Hg.

Post-operative pain management in this case is also challenging. Non-steroidal anti-inflammatory drugs should be used with care as these may also trigger crises of rhabdomyolysis. The use of opioids and sedatives during the postoperative period should also be avoided [11]. Shoulder pain mostly reduces with NSAIDS and spraying the inferior surface of the diaphragm with local anaesthetics. Superficial cervical plexus or brachial plexus block with low dose of local anaesthetics can be done in order to control the pain if not getting reduced by above mentioned modalities. In our case, we used the combination of port site infiltration with 0.5% bupivacaine and intravenous acetaminophen to minimise post-operative pain.

Conclusion:

Neuroaxial anaesthesia or general anaesthesia with TIVA both can be administered. Safe anaesthesia for surgery is the goal, any strategy has to be well planned for the perioperative period.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the Journal. The patient understands that his name and initials will not be published, and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil **Source of support:** None

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