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Case Report

Laparoscopic Cholecystectomy under Total Intravenous Anaesthesia in a Patient with Myotonic Dystrophy Type 1 (Steinert's disease) – a Case Report

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Abstract

Myotonic dystrophy type 1 or Steinert's disease is an autosomal dominant multisystem disease which is characterized by consistent contracture of muscle following stimulation (myotonia). Hypothermia, shivering, mechanical or electric stimulation during surgery can precipitate episodes of myotonia which may complicate the course of anaesthesia. The present case report focuses on successful strategies for providing general anaesthesia for laparoscopic cholecystectomy in a patient affected by this genetic disorder, at a hospital which does not have the facility for postoperative ventilation.

Keywords

myotonia, neuromuscular genetic disease, rapid sequence intubation, short acting opioid

INTRODUCTION

Myotonic dystrophy (MD) type 1 or Steinert's disease is an 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 (myotonia).[1] The neuromuscular symptoms, myotonia, progressive weakness, and waisting of facial, respiratory, laryngeal, axial, and distal limb muscles, start to appear between the second and third decades of life. The neuromuscular symptoms can be associated with sleep apnea, endocrine disorders, and cardiac, gastroenteric or cognitive disorders.[1-3]

Patients with myotonic dystrophy present many potential problems in terms of management of anaesthesia due to the multisystemic effects of the disease and extreme sensitivity of these patients to sedatives, anaesthetic agents, opioids, and neuromuscular blocking agents.[4-8] Prolonged recovery from anaesthesia[9-12] or development of perioperative cardiovascular and respiratory complications[5,6,13] were described in previous case reports. Other factors such as hypothermia, shivering, mechanical or electric stimulation during surgery can precipitate episodes of myotonia which may in turn complicate the course of anaesthesia.[6,8]

The present case report focuses on successful strategies for providing general anaesthesia for laparoscopic cholecystectomy in a patient affected by this genetic disorder at a hospital which does not have the facility for postoperative ventilation. Perioperative concerns regarding total intravenous anaesthesia (TIVA) in patients with MD are presented, and previous reports regarding anaesthetic management in similar clinical scenarios are reviewed.

CASE REPORT

A 44-year-old male patient (175 cm, 86 kg, BMI 28.1 kg/m²), with a previous diagnosis of MD was scheduled for elective laparoscopic cholecystectomy surgery because of repeated attacks of acute cholecystitis. The patient complained of peripheral muscle weakness which started at the age of 15 and gradually progressed to mild walking impairment and some difficulty in relaxing his handgrip. He reported easy fatigue but denied shortness of breath or other cardiovascular symptoms. Respiratory or swallowing difficulties were not reported. There was no history of systemic disease or anaesthetic procedure and he was not taking medication. Family history was free. At the preoperative evaluation, the patient was alert and oriented. He was able to walk unassisted and attend to his own bodily needs without assistance. Facial atony and symmetrically depressed reflexes of the distal extremities were noticed. The cardiology examination revealed decreased left ventricular function with an ejection fraction of 50%, without any conduction disorders. The pulmonary function test and other physiologic examinations and laboratory analyses were normal. His general condition was classified as ASA (American Society of Anaesthesiologists) physical status 2. The patient was found to be in grade 3 in the muscular impairment rating scale (MIRS) and the perioperative risk was considered as intermediate. The preoperative airway assessment (Mallampati score III, limited ability to open the mouth and short neck) indicated the risk of a difficult endotracheal intubation. Patient's consent was obtained for the procedure, anaesthesia, and publication.

A laparoscopic procedure was decided on. A general anaesthetic technique (TIVA) using propofol, fentanyl, remifentanil, and rocuronium was planned. No premedication other than IV ondansetron (8 mg) and omeprazole (40 mg) was given. Prior to induction of anaesthesia, a three-lead electrograph, a pulse oximeter, a non-invasive haemodynamometer, a nerve stimulator, and a bispectral index (BIS) were placed. Preoperative vital signs included blood pressure of 130/75 mmHg, a heart rate of 90 beats/min, oxygen saturation rate of 98%, and body temperature of 36.7°C. After preoxygenation, a rapid sequence induction (RSI) was accomplished by midazolam 1 mg, fentanyl 100 μg, propofol 2 mg kg^{-1}, followed by rocuronium 0.6 mg kg^{-1}, intravenously. Intubation of the trachea with an 8.0 cuffed tube was easily performed with a C-MAC3 video laryngoscope. Mechanical ventilation was initiated in the intermittent positive pressure ventilation mode. The end-tidal carbon dioxide levels was monitored and maintained at the range of 30-35 mmHg, and the airway pressure was noted to be within normal limits. The stomach decompression was achieved by careful suctioning using a catheter. Anaesthesia was maintained with oxygen (1.5 L/min), medical air (1.5 L/min), and continuous infusion of 120-150 mcg $kg^{-1}min^{-1}$ of propofol and 0.2-0.3 mcg $kg^{-1}min^{-1}$ of remifentanil using an infusion pump. The BIS was maintained in the range of 40-60. Monitoring of the train-of-four ratio (TOF) confirmed that muscle relaxation was adequate. A further dose of rocuronium 10 mg was required 75 min after the first dose when the TOF showed four twitch responses. All the time, warm intravenous fluids and a forced air warmer were used to maintain normothermia.

At surgery, a standard 4 port technique was used, and an adequate pneumoperitoneum was maintained at an insufflation pressure of 15 mmHg. At commencement of the surgery, the skin and the underlying tissues were infiltrated with ropivacaine 0.75% and near closure, the suture line was again infiltrated with ropivacaine 0.75% (total volume administered was 20 ml) in order to reduce opioid requirement. About mid-way through the operation, IV lornoxicam 8 mg and paracetamol 1.0 g were given. Throughout the procedure, which lasted about 90 min, the patient remained stable, and the operation was completed uneventfully. The infusion of propofol/remifentanil was discontinued and after the detection of a TOF ratio of 0% with the neuromuscular monitor, the neuromuscular block was reversed with IV sugammadex 2 mg kg^{-1}. At that time, BIS was 78. Two minutes later, four equal TOF responses appeared, the patient was breathing spontaneously, but the tidal volume achieved was deemed insufficient. We decided that it was safer to keep the endotracheal tube in place until he demonstrated adequate respiratory function. The trachea was extubated ten minutes later, after making sure that the patient regained consciousness (BIS 93), breathing spontaneously and following commands with adequate muscle strength. He was kept under strict surveillance in the recovery room for about two hours during which time he was doing well without any signs of muscular fatigue or respiratory distress. No further opioids were given, and he did not complain of pain. He was transferred then to the surgical ward in good medical state, with vitals within normal limits (blood pressure 135/70 mmHg, heart rate 87 beats/min, oxygen saturation rate 98%, respiratory rate 14 bpm), fully awake. A regimen of IV dexketoprofen trometamol (50 mg 12 hourly) and paracetamol (1 gr 6 hourly), was prescribed for postoperative pain relief in the first 48 h. The postoperative course was unremarkable, and the patient was discharged home on the third postoperative day on oral dexketoprofen trometamol (25 mg 12 hourly) and paracetamol (1 gr 6 hourly), as needed for pain control.

DISCUSSION

Patients with myotonic dystrophy type 1 present many potential problems in terms of management of anaesthesia due to the multisystemic effects of the disease and extreme sensitivity of these patients to sedatives, anaesthetic agents[4,5], opioids[5,6], and neuromuscular blocking agents[7,8]. Such unpredictable sensitivity may cause prolonged recovery from anaesthesia as described in previous case reports[9-12], or development of perioperative cardiovascular and respiratory complications[5,6,13]. Furthermore, other factors such as hypothermia, shivering, mechanical or electric stimulation during surgery can precipitate episodes of myotonia which may in turn complicate the course of anaesthesia.[6,8] The anesthesia team

considered the full clinical picture and decided to proceed with a laparoscopic procedure at our hospital, which does not have the facility for postoperative ventilation. Our anaesthetic plan consisted of total intravenous anaesthesia (TIVA) with propofol/remifentanil drip and rocuronium boluses, wound infiltration prior to incision with local anaesthetic, administration of anti-inflammatory drugs (lornoxicam and paracetamol) intraoperatively, fast reversal of the neuromuscular block with sugammadex, avoidance of trigger factors that could induce myotonia (hypothermia, shivering, surgical manipulation, diathermy) and postoperative analgesia based on anti-inflammatory drugs during the first 48 h. An appropriate postoperative monitoring was assured in the first 2 hours in a high dependency unit (recovery room). A literature search on this subject confirmed successful cases of total intravenous anaesthesia using propofol and remifentanil for adult patients with MD.[14-18] Although there are reports of exaggerated physiological responses[11] and a myotonic state after propofol administration[8,19], and hyperalgesia induced by remifentanil causing difficulties in postoperative pain management[20], we also used those agents in our patient, and no adverse effects were observed. Sugammadex has also been successfully used for the reversal of rocuronium in a few cases.[21,22] In light of the literature, a general anaesthetic technique (TIVA) was often preferred for laparoscopic cholecystectomy in patients with MD.[18,23]

CONCLUSIONS

In the present patient, who only had minor symptoms of the disease, our anaesthetic plan was successful; the intraoperative period was stable, and the recovery uneventful without postoperative complications.

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Лапароскопическая холецистэктомия под тотальной внутривенной анестезией у пациента с миотонической дистрофией 1 типа (болезнь Штейнерта) – клинический случай

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Резюме

Миотоническая дистрофия 1 типа, или болезнь Штейнерта, представляет собой аутосомно-доминантное мультисистемное заболевание, характеризующееся стойкими контрактурами мышц после стимуляции (миотония). Гипотермия, озноб, механическая или электрическая стимуляция во время операции могут спровоцировать приступы миотонии, которые могут осложнить курс анестезии. Настоящий клинический случай фокусируется на успешных стратегиях обеспечения общей анестезии при лапароскопической холецистэктомии у пациента, страдающего этим генетическим заболеванием, в больнице, не имеющей возможности для послеоперационной вентиляции.

Ключевые слова

миотония, нервно-мышечное генетическое заболевание, быстрая последовательная интубация, опиоид короткого действия