Laparoscopic Cholecystectomy for Cholelithiasis in a Patient With Myotonic Dystrophy

Haridimos Markogiannakis, MD, Maria Skalistira, MD, Lukas Georgiou, MD, Antonios Louizos, MD, Emmanuel Leandros, MD

ABSTRACT

Myotonic dystrophy (DM) is a rare autosomal dominant inherited neuromuscular disease involving several systems. The anesthetic method of choice remains uncertain. The risk of perioperative complications, particularly pulmonary and cardiac complications, in these patients is of major concern. We report on a 16-year-old female patient with DM type 1 undergoing laparoscopic cholecystectomy for symptomatic cholelithiasis, who had a smooth, uncomplicated recovery. Laparoscopic cholecystectomy is feasible and safe in patients with DM but requires individual multidisciplinary perioperative management.

Key Words: Myotonic dystrophy type 1, Cholelithiasis, Laparoscopic cholecystectomy.

INTRODUCTION

Myotonic dystrophy (DM) is an autosomal dominant RNA-mediated disease and is the most common inherited neuromuscular disease in adults. Two genetic loci have been associated with the DM phenotype: DM1 on chromosome 19, and DM2 on chromosome 3.1 The responsible mutation of DM type 1 is a (CTG) repeat expansion at the 3’ end of the dystrophic myotonic protein kinase gene (DMPK) on chromosome 19.2,3

The disease affects multiple systems (skeletal muscle, heart, eye, and the endocrine system), and the clinical picture includes myotonia (impaired muscle relaxation), progressive skeletal muscle wasting, arrhythmias, cataracts, insulin resistance, and altered CNS function.4 There are very few reports of DM patients undergoing laparoscopic cholecystectomy, so we present a 16-year-old female patient with DM type 1 who underwent laparoscopic cholecystectomy for symptomatic cholelithiasis with a smooth, uncomplicated recovery.

CASE REPORT

A 16-year-old female patient was admitted to our department for symptomatic cholelithiasis. She had been diagnosed with DM1 4 years earlier. Her body mass index (BMI) and nutritional status were normal; in particular, her BMI was 25kg/m². Abdominal ultrasonography revealed cholecystolithiasis. On admission, weakness of facial and anterior neck muscles and reduced muscular tone and reflexes in the lower extremities were found, but myotonic phenomena were not. Blood tests were within normal ranges. Chest x-ray, arterial blood gas analysis, pulmonary function tests, electrocardiogram, and echocardiogram were also normal.

A dose of cefazolin (500 mg) was administered intravenously (IV) one hour before the operation. In the operating room, midazolam (2.5mg) was initially administered IV. After insertion of an epidural catheter between the T₇–T₉ interspace, fentanyl (50 μg) was given epidurally; lornoxicam (8mg) was administered IV. After preoxygen-
ation, anesthesia was induced with bolus IV propofol (200 mg) and remifentanil (100 μg). The patient was intubated easily without administering muscle relaxants. Anesthesia was maintained with oxygen in the air and sevoflurane (1.6 minimum alveolar concentration).

A typical laparoscopic cholecystectomy, using 4 trocars, as previously described by our group, was performed. After the establishment of pneumoperitoneum and insertion of the laparoscope, a thorough inspection of the entire peritoneal cavity was performed and revealed no abnormality. The other 3 working ports were then inserted. Following ligation of the cystic duct and cystic artery, the gallbladder was mobilized and dissected. Meticulous hemostasis was achieved by using electrocautery. The resected specimen was introduced into a retrieval bag and removed. No technical difficulties were encountered during the whole procedure. The operating time from skin incision to skin closure was 53 minutes. The wounds were infiltrated with 0.5% Chirocaine hydrochloride.

The patient was extubated when she gained consciousness and had spontaneous respirations. The epidural catheter was removed; the patient was observed in the postoperative intensive care unit for 12 hours after the operation and then was transferred to the surgical ward. Lornoxicam (8 mg) was administered IV 12 hours after the operation for analgesia. The postoperative course was uneventful, and the patient was discharged the following day.

**DISCUSSION**

Various anesthetic techniques have been described, as there is no general agreement about the safest method. Hazards have been associated with the use of thiopentone, suxamethonium, neostigmine, and halothane, which may cause prolongation of recovery from anesthesia, apnea, and respiratory depression. However, in a Canadian study, halothane was used in 154 patients and suxamethonium in 113 patients without any episodes of myotonia or shivering. In a series of 13 DM patients undergoing oropharyngeal and head surgery, the authors used continuous propofol infusion with fentanyl and atracurium with no perioperative complications. Propofol was used for induction in the presented case, and although there is large variability in propofol sensitivity in DM patients, there was no adverse effect. Epidural anesthesia has been reported as a single method but was used in combination with general anesthesia in our patient. We also administered nonsteroidal inflammatory drugs, which are good analgesics and cause no respiratory depression. Local wound infiltration provided additional postoperative analgesia.

Patients with myotonic dystrophy have chronic restrictive respiratory insufficiency. Involvement of the upper airway and expiratory muscles occurs early, but inspiratory muscle weakness becomes prominent when proximal limb weakness is obvious. Expiratory muscle weakness may preclude effective cough and lead to atelectasis. The disability of inspiratory muscles decreases the inspiratory capacity and increases the risk of alveolar hypoventilation. The diaphragm has been found to be weak in such patients predisposing further to perioperative pulmonary complications. Central disturbances may lead to hypoventilation and reduced ventilatory response to carbon dioxide. The largest study of perioperative complications in 219 patients undergoing a variety of procedures found an 8.2% rate. Almost all complications (16 out of 18) were pulmonary complications. Eighteen patients underwent cholecystectomy, and 7 had respiratory complications (atelectasis, pneumonia, and ventilatory failure); death occurred in one of them. Multivariate analysis showed that the risk of complications was significantly higher after upper abdominal surgery (Odds Ratio: 24.4) and in patients with severe muscular disability (Odds Ratio: 14.1). Careful perioperative management in the described patient avoided any pulmonary complications.

Only 3 patients with DM who underwent laparoscopic cholecystectomy have been described. The authors reported no perioperative complications, and the patients were discharged after 3 and 4 days compared with the 1-day hospital stay of our patient. The fact that pulmonary function deteriorates significantly less in laparoscopic cholecystectomy compared with the open procedure contributes to the smooth postoperative course of these patients.

DM is also frequently associated with conduction disorders (atrioventricular block, bundle-branch block, or intraventricular conduction delays), severe arrhythmias (sinus bradycardia and tachyarrhythmias, including atrial flutter, atrial fibrillation, ventricular tachycardia, ventricular fibrillation) and dilated cardiomyopathy. The reported patient did not experience a perioperative cardiac event.

**CONCLUSION**

The presented case indicates that laparoscopic cholecystectomy can be performed safely in patients with myotonic dystrophy under careful multidisciplinary perioperative management. Because the severity of myotonic dystrophy varies among patients, the anesthetic strategy should be individually planned.
References:

1. Ranum L, Cooper T. RNA-mediated neuromuscular disorders. *Annu Rev Neurosci.* 2006;29:259–277.

2. Mahadevan M, Tsilfidis C, Sabourin L, et al. Myotonic dystrophy mutation: an unstable CTG repeat in the 3′ untranslated region of the gene. *Science.* 1992;255:1253–1255.

3. Brook J, McCurrach M, Harley H, et al. Molecular basis of myotonic dystrophy: expansion of a trinucleotide (CTG) repeat at the 3′ end of a transcript encoding a protein kinase family member. *Cell.* 1992;68:799–808.

4. Mathieu J, Allard P, Gobeil G, et al. Anesthetic and surgical complications in 219 cases of myotonic dystrophy. *Neurology.* 1997;49:1646–1650.

5. Konstadoulakis MM, Antonakis PT, Karatzikos G, Alexakis N, Leandros E. Intraoperative findings and postoperative complications in laparoscopic cholecystectomy: the Greek experience with 5539 patients in a single unit. *J Laparoend Adv Surg Tech.* 2004;14:31–36.

6. Bennun M, Goldstein B, Finkelstein Y, Jedeikin R. Continuous propofol anaesthesia for patients with myotonic dystrophy. *Br J Anaesth.* 2000;85:407–409.

7. White R, Bass S. Myotonic dystrophy and paediatric anaesthesia for patients with myotonic dystrophy. *Paed Anaesth.* 2003;13:94–102.

8. Begin P, Mathieu J, Almirall J, Grassino A. Relationship between chronic hypercapnia and inspiratory muscle weakness in myotonic dystrophy. *Am J Respir Crit Care Med.* 1997;156:133–139.

9. Bogaard J, van der Meche F, Hendriks I, Ververs C. Pulmonary function and resting breathing pattern in myotonic dystrophy. *Lung.* 1992;170:143–153.

10. Mercier M, Baghdadi H, Frosini C, Sielezneff I, Sastre B, Gouin F. Laparoscopic cholecystectomy in a patient with Steinert disease. *Ann Fr Anesth Reanim.* 1996;15:310–312.

11. Nardone A, Marradi C, Tiberio G, et al. Video laparoscopic cholecystectomy in a female patient with cholelithiasis in Steinert’s myotonic dystrophy. *Ann Ital Chir.* 2000;71:139–143.

12. Takhar AS, Thaper A, Byrne A, Lobo DN. Laparoscopic cholecystectomy in a patient with myotonic dystrophy. *J R Soc Med.* 2004;97:284–285.

13. Hendolin HI, Paakonen ME, Alhava EM, Tarvainen R, Kempinen T, Lahtinen P. Laparoscopic or open cholecystectomy: a prospective randomised trial to compare postoperative pain, pulmonary function, and stress response. *Eur J Surg.* 2000;166:394–399.

14. Bassez G, Lazarus A, Desguerre I, et al. Severe cardiac arrhythmias in young patients with myotonic dystrophy type 1. *Neurology.* 2004;63:1939–1941.