Case Report / *Olgu Sunumu*

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Anesthesia for a Patient with Myotonic Dystrophy

Miyotonik Distrofili Olguda Anestezi

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Abstract

Myotonic dystrophy is the most common myotonic syndrome causing abnormalities of the skeletal and smooth muscles as well as problems related to the cardiac, gastrointestinal and endocrine systems. In affected people, reduced functional residual capacity, vital capacity, and peak inspiratory pressure are observed within the respiratory system. As would be expected, anesthetic management of these patients is challenging for anesthesiologists. In addition, delayed recovery from anesthesia and cardiac and pulmonary complications may develop in the intraoperative and early postoperative periods due to sensitivity to sedatives, anesthetic agents, and neuromuscular blocking agents. Myotonic dystrophy can be performed with the use of appropriate anesthesia procedures as well as carefully communication between anesthesiologists and surgeons. In conclusion, myotonic dystrophy has variations, which makes it important to preoperatively determine specific surgical and anesthetic management strategies for each patient. In this article, we present a patient with myotonic dystrophy who underwent laparoscopic cholecystectomy surgery for symptomatic cholelithiasis and to discuss the relevant literature.

Keywords: Anesthesia, myotonic dystrophy, epidural

Öz

Miyotonik distrofi miyotonik sendromlar içinde en sık karşılaşılanıdır. İskelet ve düz kaslarda anormalliklerle birlikte kardiyak, gastrointestinal ve endokrin sistemle ilgili problemler içerir. Etkilenen kişilerde respiratuvar sistemde fonksiyonel reziduel kapasitede azalma, vital kapasite ve pik inspiratuvar basınçlarda azalma gözlenir. Bu hastaların anestezi yönetimi özelliklidir ve anestezistler için ciddi bir problemdir. Ayrıca sedatif, anestezik ve nöromusküler ajanlara hassasiyetten dolayı, intraoperatif ve erken postoperatif dönemde anesteziden derlenmede uzama, kardiyak ve pulmoner komplikasyonlar gelişebilir. Miyotonik distrofi farklı varyasyonlar şeklinde görülebildiğinden her hastaya özel cerrahi ve anestezi yönetiminin preoperatif olarak belirlenmesinin önemli olduğunu düşünüyoruz. Biz de bu yazıda semptomatik safra kesesi taşı nedeni ile laporoskopik kolesistektomi operasyonu olan bir hastayı sunarak literatür bilgileri ışığında tartışmayı amaçladık.

Anahtar Sözcükler: Anestezi, miyotonik distrofi, epidural

Introduction

Myotonic dystrophy (MD) was first identified by Steinhart in 1909 (1). The disease is transmitted in an autosomal dominant fashion and the mutated gene causing MD is located on the long arm of chromosome 19 (1). Symptoms, which usually arise between the second and fourth decades of life, coexist with cardiomyopathy, frontal baldness, testicular atrophy, cataracts, and intellectual and emotional abnormalities. Weakness and atrophy occur in the facial, sternocleidomastoid, distal, and pharyngeal muscles. Myotonia is the principle manifestation early in the disease, while atrophy becomes more prominent as the disease progresses (2). In affected people, reduced functional residual capacity, vital capacity, and peak inspiratory pressure are observed within the respiratory system. As would be expected, anesthetic management of these patients is challenging for anesthesiologists. Hypothermia, postoperative shivering, and electrical stimulation can induce myotonic contractions, therefore, careful observation is required in the intraoperative and postoperative periods. In addition, delayed recovery from anesthesia and cardiac and pulmonary complications may develop in the intraoperative and early postoperative

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periods due to sensitivity to sedatives, anesthetic agents, and neuromuscular blocking (NMB) agents. In this article, we present a patient who underwent laparoscopic cholecystectomy surgery for symptomatic cholelithiasis and to discuss the relevant literature.

Case

A 40-year-old male patient, weighing 75 kg and with American Society of Anesthesiologists physical status 2 was scheduled to undergo laparoscopic cholecystectomy surgery for symptomatic cholelithiasis. He had a medical history of MD diagnosed at the age of 15 and he had been smoking 1 pack per day for the past 10 years. Laboratory investigations were within normal limits and his physical examination findings of all systems were normal except for frontal baldness. After he provided informed consent, an 18-gauge (G) intravenous cannula was placed in the back of his left hand in the premedication room and a 0.9% sodium chloride infusion was initiated at 10 mL/kg-1. He was taken to the operating room without being given any premedication. Noninvasive arterial blood pressure, electrocardiography, and peripheral oxygen saturation were monitored with a Julian Plus (Drager, Germany) anesthesia device and the thoracic 9-10 interspace was determined with the patient in the sitting position. After the area was sterilized, 60 mg of lidocaine (Aritmal 2%, Osel Medicine, Turkey) was injected into the subcutaneous area. Using an 18-G (Portex, United Kingdom) epidural needle, we entered the epidural space using the loss of resistance technique, and placed the catheter. After negative aspiration was observed and a test dose was applied without a problem, 25 mg of 0.5% bupivacaine (Marcaine, Astra Zeneca, England) and 5 mL of 0.9% sodium chloride were injected through the epidural catheter. Anesthesia was induced with 200 mg of propofol (Propofol 1%, Fresenius Kabi AB, Sweden), 100 mg of aritmal, and 20 mg of atracurium (Neucurium 50 mg/5 mL, Vem Medicine, Turkey) and he was intubated with an 8.5-mm internal diameter endotracheal tube. Anesthesia was maintained with 3 L/min of (40% O_2+60% air) fresh air flow containing 1% sevoflurane to set the monitored anesthesia care at 0.5%. An intravenous infusion of remifentanil (Ultiva 2 mg/5 mL GlaxoSmithKline, Italy) at a rate of 0.1 µg/kg/minute was applied during the surgery. Intra-abdominal pressure was 12 mmHg intraoperatively. The surgery lasted one hour and no unusual circumstances or complications were encountered during the operative period.

For postoperative analgesia, 20 mg of 0.25% bupivacaine was injected through the epidural catheter. When the patient began to breathe spontaneously, he was extubated and taken to the postoperative intensive care unit. To avoid probable hypothermia, intravenous solutions were warmed with enFlow Controller Model 121 (GE Healthcare Company, New Jersey, and USA) during the operation. In the postoperative period, we prevented hypothermia and shivering, which might induce a myotonic crisis. Except for right shoulder pain, the patient did not complain of pain in the postoperative period. He was followed up for 24 hours and then discharged, as his control creatine kinase levels were within the normal limits and no additional problems were observed.

Discussion

MD, the most common myotonic syndrome, includes abnormalities of the skeletal and smooth muscles as well as, problems related to the cardiac, gastrointestinal and endocrine systems. All of these problems create serious difficulties for anesthesiologists in the perioperative and postoperative periods. In the respiratory system, in particular, intubation difficulty due to involvement of the laryngeal and pharyngeal muscles (3), reduced functional residual capacity and vital capacity and carbon dioxide retention caused by chronic alveolar hypoventilation are important problems.

Patients with MD are very sensitive to hypothermia, postoperative shivering, electrical and mechanical stimulations (4), and some anesthetic agents. As such, both surgeons and anesthesiologists should be very careful in the pre, intra and postoperative periods. Some inhalation agents, NMB agents and their antagonists and opioids are included among the agents that induce myotonic crisis (5). The safest anesthetic technique for MD is not yet clear. General anesthesia is often preferred, however to avoid the related risks, local anesthesia may be preferable (6). In the literature, there are cases of laparoscopic cholecystectomy using the thoracic epidural anesthesia technique (7). However, complications, such as shoulder pain, nausea, and vomiting can disrupt patient comfort in the intraoperative period (7). It was observed in the literature that, intra-abdominal pressure was kept high at 20 mmHg in a patient who was not administered any NMB agents. In our case, in addition to general anesthesia, we successfully used a thoracic epidural catheter for intra- and postoperative pain control. However, we did not use it alone due to probable shoulder pain and the deep intraoperative sedation requirement. Of the induction agents, thiopental causes prolonged apnea and respiratory depression (8). Alternatively, propofol can be used to induce or maintain anesthesia. In patients with diabetes mellitus, different responses to propofol have been observed (9). However, there are cases in which it was used without any problems (10). There are other cases in which etomidate was used safely in myotonic patients (11). We used propofol to induce anesthesia and encountered no complications. Although we do not prefer fentanyl for

anesthesia induction, we used an intravenous infusion of remifentanil during the surgery due to the short-acting nature of remifentanil. NMB agents are routinely used in laparoscopic cholecystectomy surgery. They are not used higher intra-abdominal pressure is required. Responses to NMB agents vary among patients as high sensitivity, normal responses or resistance to them (12,13). Of the NMB agents, succinylcholine, in particular, may induce myotonic crisis since it causes muscle contraction and hyperkalemia; as such, its use is not recommended in myotonic patients (14). In the literature, also there are cases in which succinylcholine has been used in urgent situations that did not result in any complications (15). In our case, we did not use succinylcholine because of its tendency to induce myotonic crisis. If NMB agents are going to be used, those with a short duration of action that do not require anticholinesterases should be preferred. Accordingly, atracurium seems to be the most suitable NMB (16). Neuromuscular monitoring is useful for evaluating muscle potentials in patients administered NMB. We used atracurium in this case and our patient did not require additional doses during the surgery. After sufficient tidal volume was confirmed, he was extubated at the end of the surgery. We could not provide train-of-four monitoring intraoperatively due to technical deficiencies, but we did not notice any desaturation during the postoperative follow-up.

In patients with MD, myotonia may be induced by perioperative shivering. As such, hypothermia is undesirable in these patients. A sympathetic blockade during local anesthesia causes heat loss precipitating shivering in these patients (17). Therefore, to avoid hypothermia, patients should be heated pre-and intraoperatively, and solutions should be warmed prior to administration. Opioids are often used to prevent shivering. However, epidural injection of opioids may lead to respiratory depression, thus, if opioids are to be used by epidural injection, breathing should be monitored more carefully (17).

Various complications can be observed during the postoperative follow-up of these patients. In a study of 219 patients with myotonia who underwent surgery for different procedures, it was observed that the rate of complications was 8.4% and that most were related to the respiratory system. These complications are more expected in patients undergoing upper abdominal surgery (18).

In light of the literature, laparoscopic cholecystectomy surgery in patients with MD can be performed with the use of appropriate anesthesia procedures as well as carefully communication between anesthesiologists and surgeons. Avoiding drugs that can induce myotonic crisis and carefully monitoring these patients is critical. MD has variations which makes it important to preoperatively determine specific surgical and anesthetic management strategies for each patient.

Ethics

Informed Consent: It was taken.

Peer-review: Internal peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: Dilek Kalaycı, Menşure Kaya. Concept: Dilek Kalaycı, Tuğba Aşkın. Design: Dilek Kalaycı, Özlem Şen, Süheyla Ünver. Data Collection or Processing: Dilek Kalaycı, Menşure Kaya, Fetiye Eylem Akkuş, Selda Muslu. Analysis or Interpretation: Dilek Kalaycı, Özlem Şen. Literature Search: Dilek Kalaycı, Özlem Şen. Writing: Dilek Kalaycı.

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References

1. Buxton J, Shelbourne P, Davies J, et al. Characterization of a YAC and cosmid contig containing markers tightly linked to the myotonic dystrophy locus on chromosome 19. Genomics 1992;13:526-31.
2. Ranum LP, Cooper TA. RNA-mediated neuromuscular disorders. Annu Rev Neurosci 2006;29:259-77.
3. Mahr A, Attof Y, Flamens C, et al. [Anaesthetic management of patients with Steinert myotonia: two case reports]. Ann Fr Anesth Reanim 2009;28:161-4.
4. Takhar AS, Thaper A, Byrne A, et al. Laparoscopic cholecystectomy in a patient with myotonic dystrophy. J R Soc Med 2004;97:284-5.
5. Russell SH, Hirsch NP. Anaesthesia and myotonia. Br J Anaesth 1994;72:210-6.
6. Araujo FS, Bessa Junior RC, Castro CH, et al. [Anesthesia in a patient with Steinert disease: case report.]. Rev Bras Anestesiol 2006;56:649-53.
7. El-Dawlatly A, Aldohayan A, Nawaz S, et al. Anesthetic management of a patient with myotonic dystrophy for laparoscopic cholecystectomy–a case report. Middle East J Anaesthesiol 2008;19:1135-40.
8. Dundee JW. Thiopentone in dystrophia myotonia. Curr Res Anesth Analg 1952;31:257-62.
9. White RJ, Bass SP. Myotonic dystrophy and paediatric anaesthesia. Paediatr Anaesth 2003;13:94-102.
10. Gandhi R, Jain AK, Sood J. Desflurane anaesthesia in myotonic dystrophy. Indian J Anaesth 2011;55:61-3.
11. Muller H, Punt-van Manen JA. Maxillo-facial deformities in patients with dystrophia myotonica and the anaesthetic implications. J Maxillofac Surg 1982;10:224-8.
12. Diefenbach C, Lynch J, Abel M, et al. Vecuronium for muscle relaxation in patients with dystrophia myotonica. Anesth Analg 1993;76:872-4.

13. Kodama K, Akata T, Sasaki T, et al. Unexpected resistance to pancuronium in a patient with myotonic dystrophy (myotonia dystrophica). J Anesth 2000;14:160-3.
14. Azar I. The response of patients with neuromuscular disorders to muscle relaxants: a review. Anesthesiology 1984;61:173-87.
15. Tomlinson S, Macartney I, Lam S. Dystrophica myotonia and suxamethonium. Anaesthesia 1999;54:1234.
16. Miller JD, Chingmuh L. Muscle disease. In: Katz J, Benumof JL, Kadis LB, editors. Anesthesia and Uncommon Disease. 3rd ed ed. Philadelphia: W.B. Saunders; 1990. p. 590-644.
17. Pauca AL, Savage RT, Simpson S, et al. Effect of pethidine, fentanyl and morphine on post-operative shivering in man. Acta Anaesthesiol Scand 1984;28:138-43.
18. Mathieu J, Allard P, Gobeil G, et al. Anesthetic and surgical complications in 219 cases of myotonic dystrophy. Neurology 1997;49:1646-50.