

Laparoscopic cholecystectomy in a patient with Steinert myotonic dystrophy. Case report

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SUMMARY: Laparoscopic cholecystectomy in a patient with Steinert myotonic dystrophy. Case report.

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Myotonic dystrophy (MD) is a serious multi-systemic autosomal dominant disease. The estimated incidence is 1 in every 8000 births, with an estimated prevalence of between 2.1 and 14.3 cases per 100,000 inhabitants. Signs and symptoms vary from a severe form of congenital myopathy, present from birth and often fatal, to a classic form and a delayed form, which generally presents after the age of 50 and in which the only sign is a cataract and life expectancy is completely normal.

We describe the clinical case of a 40-year-old woman with Steinert myotonic dystrophy who underwent laparoscopic cholecystectomy (under general anesthesia) for symptomatic gallbladder stones. The conduct of anesthesia in such patients must be carefully considered, as hypothermia, shivering, electrical and mechanical stimulation, and the drugs used can all trigger myotonia.

RIASSUNTO: Colectomia videolaparoscopica in paziente affetto da Distrofia Miotonica di Steinert. Caso clinico.

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La distrofia miotonica (DM) è un importante disordine autosomico dominante, multisistemico. L'incidenza è stimata di 1 caso ogni 8.000 nati con una stima di prevalenza da 2,1 a 14,3 casi ogni 100.000 abitanti. La sua espressione sintomatologica varia da una forma severa di miopatia congenita, già presente alla nascita e spesso fatale, ad una forma classica, fino ad arrivare ad una forma ad esordio tardivo, in genere dopo i 50 anni, in cui l'unica manifestazione è la comparsa di una cataratta, con aspettative di vita assolutamente normali.

Descriviamo il caso clinico di una paziente di 40 anni, affetta da distrofia miotonica di Steinert, sottoposta ad intervento chirurgico di colectomia videolaparoscopica per calcolosi sintomatica della colecisti (in anestesia generale). La condotta anestesiológica in pazienti affetti da distrofia miotonica di Steinert deve essere ben ponderata; l'ipoteremia, il brivido, le stimolazioni elettriche e meccaniche, i farmaci utilizzati possono scatenare crisi miotoniche.

KEY WORDS: Laparoscopy - Cholecystectomy - Myotonic dystrophy - Steinert disease.
Laparoscopia - Colectomia - Distrofia miotonica - Malattia di Steinert.

Introduction

Myotonic dystrophy (MD) is an autosomal dominant neuromuscular disease (1). The most common form in adults is type 1 myotonic dystrophy (MD1), or Steinert disease, characterized by important systemic signs and symptoms as well as wasting of the muscles (2).

The conduct of anesthesia in patients with MD must

be carefully evaluated, as hypothermia, shivering, electrical and mechanical stimulation, and the drugs used can all trigger myotonia. These patients are more sensitive to anesthetics and are at risk of heart and lung complications as well as malignant hypothermia (3).

We describe a case of an MD1 patient who underwent laparoscopic cholecystectomy for symptomatic gallbladder stones and discuss the conduct of general anesthesia.

Case report

A woman with Steinert disease, aged 40 years and weighing 85 kg, and scheduled for laparoscopic cholecystectomy for symptomatic gallbladder stones, underwent preoperative evaluation by the Anesthesiology and Intensive Care Service, Palermo University General Hospital in November 2010.

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The patient's medical history revealed hepatic steatosis, mild mitral stenosis and nodular thyroid disease with subclinical hypothyroidism. The patient had in the past undergone a tonsillectomy, five Cesarean sections and a voluntary abortion, all carried out under general anesthesia and characterized by breathing difficulties on reawakening.

Prior to the cholecystectomy, she underwent cardiological, neurological, and endocrinological evaluation, which confirmed her good clinical condition. Routine blood chemical tests, chest X-ray and ECG were all normal. A preoperative anesthesiological risk of ASA III was assigned. The patient underwent laparoscopic cholecystectomy through induction of pneumoperitoneum to a pressure of 10-12 mmHg with the open technique (subumbilical Hasson trocar). The French technique was used for the cholecystectomy, with the surgeon standing between the patient's legs and the use of 4 trocars. The operation was carried out under general anesthesia, with orotracheal intubation and use of an orogastric tube for aspiration, removed at the end of the procedure.

Anesthesia was induced with Atropine (0.5 mg), fentanyl 1 µg/kg (150 µg), Propofol 2 mg/kg (160 mg) and Cisatracurium 0.1 mg/kg (8 mg), and maintained with boluses of Fentanyl 0.03 mg/kg/min for analgesia and Sevoflurane 1% for hypnosis. Re-administration of cisatracurium was not needed, due to the short duration of the procedure. The patient was monitored throughout by pulse oximetry, non-invasive blood pressure measurement, ECG and capnography. At the end of the 50-minute procedure, after analgesic cover with acetaminophen 1 g i.v. and Ketorolac 30 mg i.v. the patient was extubated in the operating theater without any neuromuscular blocking drug antagonists. Vital signs were monitored continuously for around an hour in the recovery room. The patient was then

returned to the ward, as no intensive care was necessary.

Her post-operative course was regular and she was discharged on the second day, in line with all other patients undergoing laparoscopic cholecystectomy.

Discussion and conclusion

Laparoscopy is considered the gold standard for the treatment of symptomatic gall bladder stones and is the technique used in urgent cases of acute cholecystitis. In patients with muscular dystrophy, the choice of technique and drugs in the conduct of anesthesia can be problematic. The particular sensitivity of these patients to common depolarizing and non-depolarizing hypnotics and curare-like agents and their presumed sensitivity to neostigmine requires the anesthetist to carry out a careful evaluation.

Our approach was based on a decision not to use curare-like agents during the conduct of anesthesia, to avoid exposing the patient to any risk of cardiac or respiratory depression related to their use. Teamwork, and especially the cooperation between the surgeon and anesthesiologist, enabled the procedure to be concluded without complications, permitting the patient's early discharge and all the other benefits of laparoscopy.

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