ANESTHETIC MANAGEMENT OF A PATIENT WITH MYOTONIC DYSTROPHY FOR LAPAROSCOPIC CHOLECYSTECTOMY

- A Case Report -

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Abstract

Myotonic dystrophy (MD) is rare disease that offers challenges to anesthesiologists. We report a case of adult patient with myotonic dystrophy who underwent laparoscopic cholecystectomy.

A 48-year-old male patient, known case of MD, was presented for laparoscopic cholecystectomy. Physical examination revealed, young man, calm, quite, cooperative, not in pain or distress with frontal baldness, temporal bone recession, elongated face, mild degree of ptosis and swan neck. Under complete aseptic conditions, thoracic epidural T6-7 with catheter insertion was performed while the patient was placed on left lateral side. Plain bupivacaine 0.5% 7cc was injected through the catheter. Level of analgesia tested with ice reached up to T4. Intravenous sedation was achieved with midazolam 2 mg and ketamine 50 mg. The patient was comfortably lying supine on warm heated mattress, except of bilateral shoulder pains which was relieved with midazolam and ketamine.

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In conclusion, regional anesthesia, spinal or epidural, is preferable in MD patients. Shoulder pains is the main intraoperative problem encountered in our patient. Therefore, studies are needed for treating shoulder pain. To the best of our knowledge, this is the first case report in a patient with MD who underwent laparoscopic cholecystectomy under thoracic epidural analgesia.

Myotonic dystrophy (M) is rare disease and represents challenges to anesthesiologists. We report a case of adult patient with myotonic dystrophy who underwent laparoscopic cholecystectomy.

Case Report

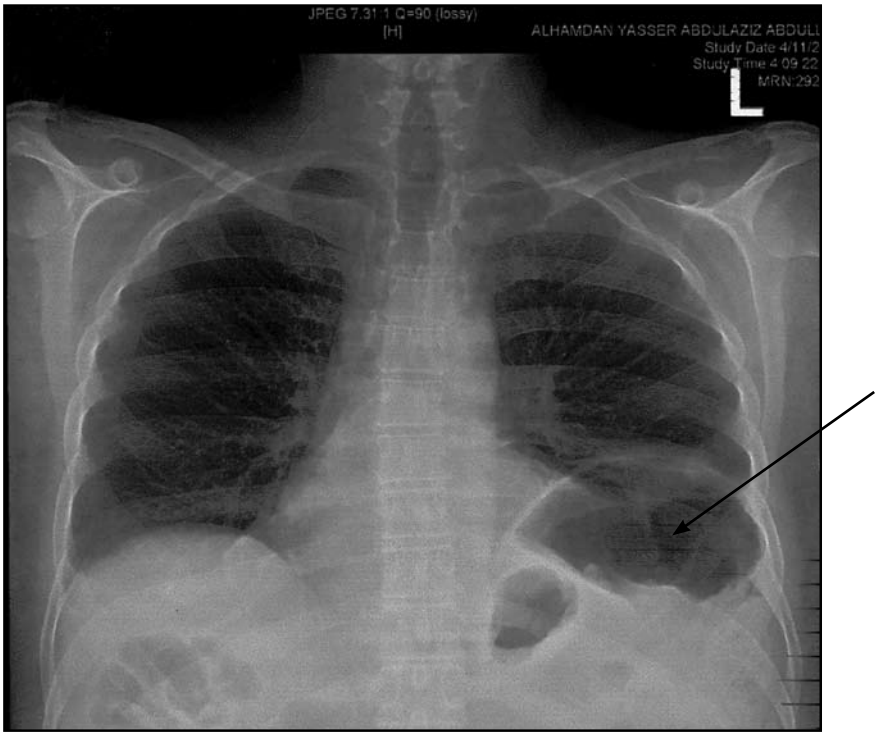
A 48-years-old male patient, height 156 cm and body weight 56 kg, known case of MD, was presented for laparoscopic cholecystectomy.

The patient gave a history of previous admission in our hospital complaining of difficulty in releasing his fist after grasping, and which had started in the left then moved to the right hands. At that time EMG was done and revealed myotonic nature, when during the insertion of the needle, myotonic discharges were heard. The findings in EMG besides the clinical diagnosis were suggestive of MD. In addition, he gave positive family history that had involved his mother and sister who died recently from the disease.

Physical examination revealed, young man, calm, quite, cooperative, not in pain or distress with frontal baldness, temporal bone recession, elongated face, mild degree of ptosis and swan neck. He gave history of bilateral cataract operation and primary infertility. Central nervous system examination showed: normal intact higher cerebral functions, normal cranial nerves, normal sensations, gait and distal muscular atrophy with weakness and myotonia and diminished reflexes in both limbs. Chest, heart and abdomen were normal. ECG showed first degree heart block. Echocardiography was normal. Other laboratory and biochemical investigations were within normal ranges. Chest-x-ray showed large air bubble in the stomach which was constantly present in the previous chest-x-rays taken during his admission in the hospital (Fig. 1).

Fig. 1

Chest-x-ray shows the large air bubble in the stomach. (black arrow)



Premedication consisted of oral diazepam 10 mg and ranitidine 150 mg 2 hr preoperatively. In the operation theatre, routine monitoring were connected.

Under complete aseptic conditions, thoracic epidural T6-7 with catheter insertion was performed in the left lateral position. Plain bupivacaine 0.5% 7cc was injected through the catheter. Level of analgesia tested with ice reached up to T4. Intravenous sedation was achieved with midazolam 2 mg and ketamine 50 mg. The patient was breathing spontaneously throughout the procedure via face mask. CO_2 insufflation was performed with intraperitoneal pressure of 12 mmHg. The patient was comfortably lying supine on warm heated mattress, except of bilateral shoulder pains which was relieved with midazolam and ketamine. The theatre temperature was kept warm. The procedure took 2 hr. Surgery was uneventful.

The patient was then transferred to surgical intensive care for further follow up. He made uneventful recovery and was transferred 24 hr later to the surgical floor.

Discussion

Myotonic dystrophy is a post-junctional autosomal-dominant myopathy disorder. It is characterized by incomplete muscle relaxation[1]. The disease is associated with frontal baldness, cataracts and testicular atrophy. It may also associated with cardiomyopathy and conduction abnormalities, restrictive lung disease, central and obstructive sleep apnea[2]. In the case presented the patient was having all clinical signs in addition to cardiac conduction abnormalities in the form of 1st degree heart block, but no sleep apnea disorder. Myotonia may be precipitated by hypothermia, shivering and mechanical or electrical stimulation. Sensitivity to sedatives and anesthetics has been described with MD patients. Succinylcholine can induce myotonic contracture and is not recommended for patients with MD[3]. Non-depolarizing muscle relaxants can be used safely, however use of anticholinesterase drugs as reversal agents can induce myotonic response secondary to increased sensitivity to the stimulatory effect of acetylcholine[2].

In one case report of a patient with MD[4] who underwent cardiac catheter ablation, it was concluded that target controlled infusion of propofol was a useful anesthetic technique in MD patient. In another case report, persistent apnea was described in an obese patient with MD following ovarian resection under general anesthesia and vecuronium relaxant. The conclusion was, anesthesia for patients with MD requires careful attention for perioperative respiratory management[5]. In another case report[6] it was shown that MD patients may have clinically significant increased sensitivity to vecuronium. In still further report[7] induction and maintenance of general anesthesia was achieved with propofol and airway was maintained with laryngeal mask in a patient with MD who underwent distal gastrectomy without respiratory complications such as respiratory depression or atelectasis occurring after surgery.

Regional anesthesia, including spinal or epidural, has been recommended as preferred anesthetic technique for patients with MD[8]. In pediatrics, combined general and epidural anesthesia has been recommended for patients with MD because it provides good conditions to the surgeon and adequate analgesia perioperatively. It also avoided the need for reversal of the muscle relaxant, postoperative ventilation or intravenous opioid use[9,10].

In our case, general anesthesia and muscle relaxants were avoided. Thoracic epidural analgesia was used without major intraoperative problems. The only problem encountered was bilateral shoulder pains. Shoulder pain is a common complaint following laparoscopic surgery.

A number of studies have looked at methods to reduce the severity of shoulder pain following laparoscopic surgery with conflicting results[11,12]. We have used intravenous ketamine to abort intraoperative shoulder pains.

In conclusion, myotonic dystrophy patients present challenges to the anesthesiologists. Regional anesthesia either spinal or epidural is preferable in MD patients. Shoulder pain is the main intraoperative problem encountered in our patient. Therefore, studies are needed for treating shoulder pain. To the best of our knowledge, this is the first case report of a patient with MD who underwent laparoscopic cholecystectomy under thoracic epidural analgesia.

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