Introduction

The existence of myotonic dystrophy in a patient can go undetected because of its variable expressivity and its uncommon occurrence in anaesthetic practice. Nevertheless, failure to detect the disease preoperatively may result in significant morbidity and mortality.

Case history

A 45-year-old female, who was 1.63 m tall, and who weighed 55 kg, presented to our preanaesthetic clinic for an elective laparoscopic cholecystectomy. The patient did not provide a history of any previous medical illnesses, drug allergies, or surgeries under anaesthesia. Her vital signs were normal and the general examination was unremarkable, except for the presence of an elongated facies and bilateral ptosis. Examination of the airway revealed an interincisor gap of 3 cm, the presence of retrognathia and a high, arched palate, Mallampati class IV, medially protruding canines and premolars, a thyromental distance of > 6.5 cm, and normal submandibular compliance. Her neck movements were adequate. She was unable to perform the upper-lip bite test.

The full blood count, urine investigations, liver function tests, chest X-ray and electrocardiogram were within acceptable limits. Written informed consent was obtained from her for her to undergo surgery and anaesthesia. Prior to surgery, she was advised to fast for eight hours, and was given oral alprazolam 0.25 mg at night and on the morning of surgery.

On the morning of surgery, she was moved to the operation theatre. An electrocardiogram, non-invasive blood pressure monitor, pulse oximeter, capnograph, temperature probe and oxygen analyser were utilised as monitors, and warm lactated Ringer’s solution infusion initiated. She was premedicated with an intravenous injection of morphine 6 mg and induced with intravenous propofol 120 mg and 1.2% isoflurane with N₂O (66%) in O₂. After confirmation of bag and mask ventilation, 5 mg of intravenous vecuronium bromide was administered to facilitate endotracheal intubation. Initially, a Proseal® laryngeal mask airway (LMA) number 4 was used to attempt to secure the airway as it could not be negotiated due to protruding canines and premolars, following which a Proseal® LMA number 3 was inserted digitally over a 14G Ryle’s tube. The patient was maintained on isoflurane 0.6% and N₂O (66%) in O₂. No relaxants were required throughout the intraoperative period which lasted one-and-a-half hours.

On completion of the surgery, all inhalational anaesthetics were turned off and 100% O₂ administered. The patient's
residual motor block was reversed with intravenous 3 mg neostigmine and 0.6 mg glycopyrrolate. The Proseal® LMA was removed after confirmation of swallowing reflex, gag reflex, eye opening and mouth opening on command. Soon after the removal of the Proseal® LMA, the patient failed to generate adequate tidal volume. Her ventilation was assisted with a bag and mask to achieve adequate oxygenation and normocarbia. In view of the inadequate reversal, another dose of intravenous neostigmine 1.2 mg and glycopyrrolate 0.1 mg was administered. However, there was not much improvement. The possibility of sedation due to morphine was considered, and the patient was given a bolus dose of intravenous naloxone 0.4 mg, followed by infusion at 1 µg/kg/minute. The patient continued to show no improvement. Her arterial blood gas revealed respiratory acidosis (pH 7.20, PaCO₂ 100 mmHg). Other causes of inadequate reversal viz. hypothermia and hypoglycaemia were ruled out (temperature 36.5°C and random blood glucose 148 mg %). In case there was a possibility of hypocalcaemia, 30 ml of 10% calcium gluconate was administered intravenously. When no improvement was observed, the patient was intubated nasotracheally under 30 mg of propofol and transferred to the intensive care unit (ICU) for elective ventilation.

In the ICU, the patient was conscious, with a Ramsay sedation scale score of 2. She was put on a continuous positive-pressure ventilation (CPAP) trial which she tolerated well, maintaining adequate oxygenation and normocarbia. Investigations to rule out other causes of inadequate reversal (e.g. serum electrolytes and a thyroid function profile) were requested. No muscle relaxants, sedatives or opioids were given to the patient. For analgesia, administration of 30 mg of intramuscular ketorolac was advised.

Investigations revealed normal serum electrolytes and thyroid functions. A diagnosis of a neuromuscular disorder or myasthenia gravis was then contemplated. As the patient had not shown any improvement with repeated doses of neostigmine, a diagnosis of myasthenia gravis was relegated to the background. The literature was reviewed for a neuromuscular disorder associated with ptosis, a high arched palate, dental abnormalities, cholecystitis, and increased sensitivity to sedatives. All these features were found to be associated with myotonia dystrophica. A neurological opinion was sought regarding evaluation and management. Based upon advice, a muscle biopsy from the vastus muscle was obtained under local anaesthesia, and a blood sample was sent for genetic study. An electromyogram was deferred for a later date after stabilisation of the patient’s acute condition. Over the next 12 hours, the patient was weaned off CPAP followed by a T-piece trial and finally extubated. The rest of the postoperative period remained uneventful.

The muscle biopsy revealed many muscle fibres with central nuclei and selective atrophy of histochemical type I fibres. The DNA analysis of the blood showed numerous repeats of CTG codon on chromosome 19 at 19q13 locus, both suggestive of myotonia dystrophica. An electromyogram could not be performed as the patient was unwilling to undergo any further tests. She was discharged and advised to follow-up in a neurology clinic. Written informed consent was obtained from her for publication of her case report.

Discussion

Myotonic dystrophy, also known as Steinert’s syndrome, is the most common adult muscular dystrophy (prevalence of 2.5-5.5:100 000). Its mode of inheritance is autosomal dominant, with a male:female ratio of 1:1. It presents in the second to fourth decade, classically as a triad of cataract, frontal baldness and mental retardation. However, all these three features were absent in our patient. This can be explained on the basis of incomplete penetrance or variable expressivity that are commonly seen with autosomal dominant disorders. There is also progressive weakness and muscle wasting of distal extremities, sternocleidomastoid, cardiac and respiratory muscles. Progressive wasting of the facial muscles, especially temporalis and masseter, results in “hatchet-shaped facies”, as seen in our patient. Weakness of the levator muscles result in ptosis. Weakness of smooth muscle can lead to decreased gastrointestinal motility and cholelithiasis. The presence of ptosis and cholelithiasis in our patient also pointed towards myotonic dystrophy. The presence of endocrinopathy is manifested by insulin resistance and gonadal atrophy. Although our patient never had elevated blood sugar levels, her marital history revealed that she had been divorced, with no children. As the patient had not previously undergone any tests to identify the cause of her infertility, and because she denied any further tests for the same, we could not establish the presence of gonadal atrophy in our patient.

The diagnosis of myotonic dystrophy is established by muscle biopsy, DNA analysis and electromyography. Serum creatine kinase levels may not be helpful, as they can remain normal or slightly elevated. The “myotonia” in myotonic dystrophy rarely warrants treatment. Phenyoitin is the preferred agent in patients who occasionally require treatment. Phenyoitin decreases Na⁺ influx into muscle cells, and thus decreases muscle excitability. A cardiac pacemaker should be inserted in patients with unexplained syncope or advanced conduction defects. As our patient did not have any previous history of syncope or muscle weakness in the past, and because she...
had subsequently regained her muscle power completely, no treatment, except for regular follow-up in the neurological outpatient department, was advised for her.

The anaesthetic management\textsuperscript{1,4,5,7} of patients with myotonic dystrophy warrants caution with the use of premedicants, opiates, induction agents and muscle relaxants, as they can be exquisitely sensitive to these agents.\textsuperscript{1,7} There have been reports of a generalised myotonic state associated with the use of propofol in such patients.\textsuperscript{8} However, many workers have used propofol without any apparent adverse effects.\textsuperscript{7,8,10} Inhalational agents may abolish myotonic contractions. However, the postoperative occurrence of “halothane shakes” can precipitate a myotonic crisis.\textsuperscript{4,7} Halothane can also cause cardiac depression. Patients with myotonic dystrophy are prone to malignant hyperthermia.\textsuperscript{1,4,5} Inhalational agents and succinyl choline should be avoided. Muscle fasciculations that occur with succinyl choline can also precipitate a myotonic crisis.\textsuperscript{1,7} The theoretical concern of prolonged muscle contractions by facilitating depolarisation with the use neostigmine has not been completely validated.\textsuperscript{1} The prevention of postoperative shivering is imperative due to the possibility of prolonged myotonic contractions.\textsuperscript{4,6,7} Although pethidine can reduce postoperative shivering, such patients can be extremely sensitive to opioids. Myotonic contractions can also occur during surgical manipulations and use of electrocautery, and their use should be minimised.\textsuperscript{1,7} Patients with myotonic dystrophy are prone to sudden cardiac death due to cardiomyopathy, cardiac dysrythmias, and mitral valve prolapse. Any history of syncope, or evidence of advanced conduction block, warrants detailed cardiac evaluation and insertion of a cardiac pacemaker prior to surgery.\textsuperscript{7} A postoperative pulmonary complication in the form of aspiration pneumonia, can occur due to weakness in the pharyngeal muscles. Atelectasis and alveolar hypoventilation can also take place due to weakness in the diaphragm and accessory respiratory muscles, a low central ventilatory drive and sensitivity to opiates. A high anaesthetic risk, with the possibility of postoperative ventilatory support, should be explained to such patients prior to anaesthesia and surgery.

In conclusion, it must be emphasised that the preoperative detection of myotonic dystrophy is crucial to avoid morbidity and mortality. Hence, indicative signs of any muscular weakness, for example ptosis and distal muscle weakness, as well as a history of unexplained syncope, should always be sought in regular anaesthetic practice.

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