ANAESTHESIA FOR MAJOR ABDOMINAL SURGERY IN A PATIENT WITH MYOTONIA DYSTROPHICA

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Myotonia dystrophica is a hereditary disease which may present serious problems for the anaesthetist. It is characterized mainly by myotonia and muscular wasting (Miller and Lee, 1981). Myotonia may be provoked by suxamethonium, neostigmine, hypothermia, shivering or hyperkalaemia, and may render ventilation difficult and tracheal intubation impossible.

Patients are sensitive to barbiturates, opioids, benzodiazepines and volatile agents, all of which may cause apnoea when administered in small doses. These and other anaesthetic problems have been reviewed by Aldridge (1985).

We present a patient suffering from all of the typical signs and symptoms of the disease, who was successfully managed while undergoing an urgent partial gastrectomy.

CASE REPORT

The patient was a 50-yr-old woman. Her mother had died 10 years earlier following the administration of suxamethonium for an orthopaedic operation; her aunt had suffered from myotonia, but her father and brother are fit and well.

At the age of 10 yr the patient underwent an uneventful tonsillectomy. The first symptoms of myotonia dystrophica appeared at the age of 20 yr, with dysarthria, atrophy of the extremities and myotonia in both hands. The patient also complained of difficulties with mastication and in lifting her head while supine. She later developed frontal baldness, inexpressive facies, mental retardation and inability to walk more than a short distance (grade III myotonia) (Gillam et al., 1964).

During the 15 years before admission she had suffered from recurrent biliary colic, but always refused surgery. However, a recently diagnosed gastric carcinoma in situ made surgery imperative.

Preoperative assessment

Laboratory findings were normal. A chest radiograph showed a normal sized heart, but atelectasis of the lower lobe of the left lung. Pulmonary function tests indicated moderate airway restriction. The ECG showed some characteristics typical of the disease: P–R 200 ms, Q waves, and an incomplete right bundle branch block.

Bearing in mind the tragic death of her mother, the patient refused a general anaesthetic initially. She even asked for acupuncture and other alternative techniques. However, following good psychological preparation, she accepted general anaesthesia.

SUMMARY

A case report is presented of a woman with known myotonia dystrophica, who required partial gastric resection and cholecystectomy. Neuromuscular blockade was obtained using a single dose of vecuronium, and neuromuscular function was determined by the “train-of-four count” technique. The intraoperative course was uneventful, and it was not necessary to use neostigmine to antagonize neuromuscular blockade at the end of the operation. There were no postoperative complications and the patient was discharged from hospital 10 days later.

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Intra- and postoperative periods

The patient was not premedicated. ECG, arterial pressure, heart rate, CVP, oesophageal temperature and urinary output were monitored during the operation. A “sleep” dose of thiopentone (240 mg) was administered, followed by vecuronium 6 mg. After 2 min of manual ventilation of the lungs with 100% oxygen, the trachea was intubated easily. Anaesthesia was maintained with 70% nitrous oxide in oxygen, supplemented with 0.5% halothane. Ventilation was controlled mechanically (Engstrom 300 ventilator). \(P_aO_2\), \(pH\), \(P_cO_2\) and electrolyte concentrations were maintained within normal limits.

Neuromuscular function was monitored using a “Myotex” peripheral nerve stimulator, delivering a train of four stimuli at 2 Hz every 12 s to the ulnar nerve via cutaneous electrodes. Unfortunately, a twitch analyser was not available, and the extent of neuromuscular blockade was estimated by counting the number of palpable responses following each series of train-of-four stimuli (d’Hollander et al., 1986). Twenty minutes after the administration of vecuronium, the observed number of twitches had recovered to four. However, as operating conditions remained satisfactory, even during peritoneal closure, no further doses of neuromuscular blocking drug were administered during the remainder of the operation (which lasted 3 h).

No other drugs were administered, except fentanyl 0.1 mg given just before the surgical incision. Normovolaemia was maintained by administering concentrated red cells 2 units, physiological saline 1500 ml and 5% dextrose-in-water 500 ml, all of which had previously been warmed to a temperature of 36 °C.

At the end of the operation halothane was discontinued and 100% oxygen administered. After several minutes the patient opened her eyes and regained spontaneous breathing. Since there were no signs of residual neuromuscular blockade, the tracheal tube was removed without the prior administration of neostigmine. The patient remained pain-free for several hours after extubation.

With no postoperative complications, she was discharged from hospital 10 days later.

DISCUSSION

This patient presented considerable anaesthetic difficulties because her myotonia dystrophica had reached an advanced stage and she required a major abdominal operation.

Since exaggerated responses to thiopentone, opioids, benzodiazepines and volatile agents have been reported, regional anaesthesia may be preferable when clinically feasible (Wheeler and James, 1979). However, this approach was not considered appropriate for upper abdominal surgery. The chosen technique aimed to avoid those stimuli (chemical, mechanical, electrical, thermal) or drugs which could provoke sustained contractures.

Suxamethonium and neostigmine were contraindicated. Although Azar (1984) has warned that responses to non-depolarizing neuromuscular blockers may be exaggerated, they appeared to be normal in this case.

Similar cases have been reported. Two were managed using atracurium (Boehnimer, Harris and Ward, 1985; Nightingale, Healy and McGuinness, 1985), and another with dantrolene (Phillips et al., 1984). However, vecuronium is particularly free from adverse effects and was considered to be the neuromuscular blocker of choice.

Neuromuscular function monitoring was delayed until tracheal intubation had been accomplished, in order to avoid any possible sustained electrical contraction during incomplete paralysis. The maintenance of good abdominal relaxation for 3 h with a single dose of vecuronium may be related to:

(a) Muscular weakness attributable to the disease itself;
(b) A prolonged effect of the neuromuscular blocker. Although prolongation of effect was not detected with the train-of-four count, this remains a possibility because of the imprecision of that method. It would have been much more satisfactory to have measured the train-of-four ratio.
(c) An exaggerated response to barbiturates, halothane and fentanyl. This possibility appears unlikely in view of the normal rate of recovery from anaesthesia.

In conclusion, a patient with grade III myotonia dystrophica was managed successfully during major surgery by the use of balanced anaesthesia in the form of a small dose of thiopentone, a single dose of vecuronium, plus halothane and fentanyl.
REFERENCES


