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EXAGGERATED PHYSIOLOGICAL RESPONSES TO PROPOFOL IN MYOTONIC DYSTROPHY

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SUMMARY

A patient with marked manifestations of myotonic dystrophy presented for surgical correction of cataracts. Propofol was used as part of the anaesthetic technique. The patient demonstrated marked sensitivity to its central depressant effects.

KEY WORDS

Anaesthetics i.v : propofol Myotonia dystrophica.

Myotonic dystrophy is a familial disease with autosomal dominant transmission and variable penetrance. Typically, the disease process becomes manifest in the second and third decades of life. Prevalence is equal between the sexes, but pathological changes may be more obvious in the male. Myotonia may be an early symptom encountered most often in the muscles of hand grip and face. Voluntary muscles may become progressively weaker with associated increase in fatigue and decrease in exercise tolerance. Typically, patients exhibit a lethargic demeanour with apathetic expression and ptosis of the eyelids. There may be frontal balding caused by premature recession of the hairline. Muscle wasting may be marked and is most obvious in the face, neck and distal limbs. Early presentation to the medical services occurs with infertility, or loss of visual acuity because of cataract formation. The latter may require surgical correction and this usually involves some type of anaesthesia. Life expectancy is reduced and death results usually from cardiopulmonary failure during the fifth decade of life.

Myotonic dystrophy is a multi-system degenerative disease. Striated muscle is affected by a variable degree of myotonia induced by voluntary or reflex contraction, mechanical stimulation (including surgical), and aggravated by cold. Muscular atrophy is progressive and leads to loss of mobility and ability to work. It also decreases ventilatory capacity and ability to clear chest secretions by coughing. Involuntary muscle atrophy may cause gastrointestinal paresis and loss of oesophageal competence (increasing the risk of regurgitation); loss of laryngeal competence (increasing the risk of aspiration); and loss of pharyngeal tone (increasing the risk of obstructive apnoea). The myocardium may be affected, with arrhythmias and pump failure. Degeneration of endocrine glands may lead to diabetes mellitus, hypoadrenalism, myxoedema and infertility. Degeneration in the brain may lead to apathy, somnolence, disorders of affect and pre-senile dementia. Degenerative change in the ocular lens leads to cataract formation [1].

The first case report of anaesthesia in this disease was published in 1915 [2]. The patient underwent muscle biopsy under ether general anaesthesia. The operation was complicated by muscle spasm induced by the surgical incision. While the patient was on the operating table, ventilation became grossly inadequate and cyanosis occurred. After operation the patient remained cyanotic and unconscious for 24 h. Many other reports have appeared subsequently in the literature [3–7]. These case reports attest to the ease with which apnoea can be induced with a variety of agents and techniques and the frequency of postoperative respiratory failure and delay in recovery. More recent publications have described the use of newer drugs [8–10]. However, there is still no universally recommended technique; outcome is unpredictable, and disaster awaits the unwary or unprepared [11].

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CASE HISTORY

The patient was a 31-yr-old, 65-kg, unmarried male with a father affected severely by myotonic dystrophy. In the past 10 years he suffered from progressive limitation of exercise tolerance and gradual deterioration in eyesight. On presentation to the department of ophthalmic surgery his visual acuity was almost zero in one eye and only 6/12 in the other. Exercise tolerance was limited to one flight of stairs. In the preceding year he had been affected also by somnolence.

He gave no other medical history and had had no previous surgery or anaesthesia. He took no regular medications and had no known allergies. On examination he had a typical deadpan expression with obvious bilateral ptosis and muscle wasting in upper and lower limbs. He exhibited generalized weakness of all muscle groups and myotonia in response to handgrip and percussion of clenched masseteric muscles. There was no evidence of dyspnoea, hypoxia or hypercapnia at rest. Haematological and biochemical values were normal, as were chest x-rays and ECG. Bedside respiratory function testing revealed a reduced FVC (2.5 litre) but normal FEV_1:FVC ratio.

Arrangements were made for postoperative recovery and care in the intensive care unit. The patient was anxious and was premedicated with temazepam 20 mg orally, 90 min before surgery. On arrival in the anaesthetic room the patient was fully conscious but relaxed, and exhibited no difficulty with head-lift, swallowing or breathing. ECG and arterial pressure were monitored before and during induction.

Propofol 25 mg was given into a free flowing, warmed, i.v. infusion of normal saline while the patient was observed closely. After 3 min the patient was still conscious and no obvious change in ventilation or circulation was seen. A further 25 mg of propofol was given. One minute later the patient became unconscious and apnoeic. Arterial pressure decreased from 130/80 to 101/65 mmHg. The lungs were ventilated easily with 50% nitrous oxide in oxygen. Laryngoscopy was performed without difficulty and a 9-mm oral, cuffed tracheal tube was inserted without gagging or coughing or change in heart rate and arterial pressure. The patient was transferred to theatre and placed on a heated water-blanket, and the lungs were ventilated with 50% nitrous oxide and 0.5% isoflurane in oxygen. Ventilation was adjusted to achieve normocapnia.

On preparing the patient's eye for surgical incision it was noticed that the eyeball was held in extreme downward gaze and some weak coughing movements were apparent. The inspired isoflurane concentration was increased to 1%. This abolished the coughing movements and decreased the ocular muscle spasm within 1 min. Surgery was commenced and over the next 10 min arterial pressure decreased progressively to 65/35 mmHg. A rapid infusion of normal saline 1000 ml had no effect, so the inspired isoflurane concentration was decreased to 0.5%. Arterial pressure increased steadily over the next 10 min to 110/70 mmHg. The operation was completed uneventfully in a further 25 min. At the end of surgery, isoflurane and nitrous oxide were discontinued and the lungs ventilated with 100% oxygen.

After 25 min no sign of waking was apparent, the patient remained apnoeic and no airway reflexes could be elicited, although end-expired carbon dioxide was increased to 7.0 kPa. Doxapram 100 mg i.v. had no effect. The circulation remained stable with an arterial pressure of 120/80 mmHg and heart rate of 80 beat min^{-1}. After a further 80 min the patient started gagging on the tracheal tube. On discontinuing controlled ventilation, the patient took occasional spontaneous breaths. During the next 10 min ventilatory rate increased to 15 b.p.m., but he would not respond to command and his eyelids remained closed. Extubation was achieved easily, but the patient could not maintain an adequate airway. Insertion of an oropharyngeal airway allowed comfortable, regular ventilation, but he remained unconscious. He was then escorted to ITU breathing spontaneously via an oxygen mask.

He regained consciousness after a further 2 h when he was able to maintain his own airway. The remainder of his recovery was uneventful. On questioning, he had no recall of any event between induction and removal of the oropharyngeal airway.

DISCUSSION

The potential risks associated with general anaesthesia in this patient were appreciated, but it was felt that surgical success might be severely compromised by myotonia in the ocular muscles and by movement if it were undertaken in a conscious, nervous patient.

The use of propofol in this condition has not

been described before. The drug was administered cautiously and titrated to effect. It was clear that a profound depth of anaesthesia was achieved with a dose of drug that would be considered sub-anaesthetic in a healthy patient. The use of suxamethonium to facilitate intubation was avoided because of the risk of inducing generalized myotonia [12, 13] and intubation conditions were ideal after propofol alone. Because of the apparent ease with which deep anaesthesia could be induced and maintained it was decided to avoid the use of a neuromuscular blocking agent. This also ensured that neostigmine would not be required. As anticipated, surgical manipulation provoked myotonia of the ocular muscles and the use of isoflurane successfully overcame this problem. Reasonable precautions were taken to prevent hypothermia, as postoperative shivering can induce generalized myotonia. It was hoped that the small dose of isoflurane used, for only a short period of time, would not induce the shivering-like muscle contractions seen sometimes during recovery from volatile anaesthesia. As the operation is not associated with much pain, either during or after surgery, the use of opioids was avoided.

The small doses of propofol and isoflurane used not only induced surgical anaesthesia but also caused hypotension. This may have been caused by depression of the central nervous system, or a direct effect on the circulation, or both. In spite of the very small total doses of anaesthetic agents used, the patient was deeply unconscious and apnoeic for nearly 2 h after operation and remained unconscious and unable to maintain a clear airway for a further 2 h. No problems were experienced with regurgitation of gastric content and aspiration, as the patient was not acutely ill and had been well starved.

Less than 1 mg kg^{-1} of propofol was adequate to induce general anaesthesia deep enough to permit tracheal intubation in this patient. Anaesthesia was maintained at a depth adequate to facilitate controlled ventilation and surgery, by nitrous oxide and a small concentration of isoflurane. Small doses of this short acting i.v. anaesthetic agent combined with the inspiration of a rapidly eliminated anaesthetic may be associated with an exaggerated and prolonged anaesthetic effect in this disease.

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