Anaesthesia for abdominal hysterectomy in Charcot-Marie-Tooth disease

A case report

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Summary

A 44-year-old white woman with Charcot-Marie-Tooth disease underwent an abdominal hysterectomy. The pre-operative preparation and anaesthetic management are presented, and the specific problems discussed.

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Charcot-Marie-Tooth disease is hereditary and belongs to a broad spectrum of degenerative diseases. Classically there is chronic degeneration of peripheral nerves and roots, resulting in distal muscle atrophy that begins in the feet and legs and later develops in the hands. The anaesthetic management of a patient with established Charcot-Marie-Tooth disease is described.

Case report

A 44-year-old white woman with Charcot-Marie-Tooth disease was admitted to Tygerberg Hospital, Parowvallei, CP, for an abdominal hysterectomy. She had had a sick sinus syndrome (SSS), and a permanent pacemaker had been inserted. The patient had been on nomifensine 50 mg twice a day, lorazepam 2.5 mg nightly, carbamazepine 200 mg nightly, and amiloride 5 mg in the morning.

Pre-operatively the patient's pacemaker was functioning normally, and other than the muscle weakness she was fit for surgery and anaesthesia. Pre-operative serum electrolyte values were normal. She received lorazepam 2.5 mg the night before the operation. Premedication consisted of diazepam 10 mg orally 2 hours before the operation. The ECG pulse rate and blood pressure were monitored continuously and temperature was also monitored using a nasopharyngeal probe. Before induction, the blood pressure was 110/70 mmHg and pulse rate 87/min.

The patient was pre-oxygenated for 5 minutes with 100% oxygen. A peripheral intravenous line was inserted and an infusion of Hidrolyte begun. Anaesthesia was induced with propofol 200 mg and suxamethonium 20-40 µg/kg/min. Pancuronium 6 mg (0.1 mg/kg) was used for muscle relaxation. The patient was ventilated with 60% nitrous oxide and 40% oxygen. Normocapnia was maintained throughout the procedure. A peripheral nerve stimulator was used for the evaluation of neuromuscular function. No inhalational anaesthetic agent was used during the procedure, and no intubation problems were experienced. The patient remained haemodynamically stable throughout the operation. The pancuronium action was reversed with glycopyrrolate 0.2 mg and neostigmine 3 mg. The patient began to breathe spontaneously postoperatively and no ventilation problems were experienced.

Discussion

A pragmatic anaesthetist can manage the vast majority of routine anaesthetic cases, but occasionally may be challenged with the anaesthetic management of a rare condition. Management of these cases must be based on sound physiological and pharmacological principles. This patient presented with two problems of importance to the anaesthetist: (i) Charcot-Marie-Tooth disease; and (ii) SSS, for which the patient had received a permanent pacemaker.

Charcot-Marie-Tooth disease

Meryon1 first described so-called 'idiopathic disease of muscle' in 1864. In the same year Griesinger2 reported the first muscle biopsy performed under chloroform anaesthesia — a biopsy specimen was taken from the deltoid muscle of a patient with muscular dystrophy. Since then anaesthetists have become more aware of the anaesthetic complications in patients with this condition.

Sporadic reports on the anaesthetic management of this group of diseases have appeared in the literature.3 The possibility of respiratory insufficiency should always be borne in mind and since these patients may also have myocardial damage dysrhythmias commonly appear during anaesthesia. Although malignant hyperthermia is not thought to be a problem, inhalational anaesthetic agents must be administered with great caution. These patients may be sensitive to the non-depolarizing group of muscle relaxants and so a peripheral nerve stimulator is mandatory. Avoidance of suxamethonium may prevent the occurrence of rhabdomyolysis and hyperkalaemia.4

In 1886 Charcot and Marie5 in Paris and Tooth6 independently in Cambridge described a syndrome in which wasting and weakness started in the lower limbs (affecting predominantly the peroneal and anterior tibial muscles and the small muscles of the feet) and slowly spread proximally. The wasting stopped abruptly in a transverse line across the lower part of the thigh, giving an 'inverted champagne bottle' appearance. Several years later weakness and wasting would occur in the hands and forearms. The disease usually starts in adolescence or early childhood and is often hereditary. It may become progressive and severe or arrest may occur spontaneously.
Charcot-Marie-Tooth disease is a chronic muscular dystrophy and does not influence life expectancy. This disease is usually of academic interest only to anaesthetists because muscles of respiration and deglutition are usually unaffected; the myocardium is also rarely concomitantly involved. In one variety of Charcot-Marie-Tooth disease — hereditary sensory neuropathy — there is sensory disturbance only; this begins distally in the legs with selective loss of pain and temperature sensation and sometimes leads to chronic ulceration and destruction of the bones in the feet. During anaesthesia in such a case attention must therefore be given to the pressure points.

Our patient received the normal dose of the non-depolarizing muscle relaxant pancuronium, neuromuscular relaxation being monitored by a peripheral nerve stimulator. No problems were experienced with the reversal of pancuronium. It is advisable to avoid a depolarizing muscle relaxant in these cases. No inhalational anaesthetic agent was used, although malignant hyperthermia has not as yet been reported in Charcot-Marie-Tooth disease.

Sick-sinus syndrome
Anaesthetic management of our patient was complicated by the presence of SSS for which the patient had a permanent pacemaker. SSS, first described by Ferrer in 1968, is now well recognized, and treatment with permanent pacemakers is an established procedure. Although special circumstances predominate whenever a patient with a pacemaker receives anaesthesia, problems are relatively few in comparison with those in patients with SSS but without a pacemaker. The bradycardia in SSS may be due to disordered impulse generation in the sino-atrial node or impaired conduction of an impulse from the node in the atrium. Predisposing factors for SSS are coronary atherosclerosis, hypertensive heart disease, cardiomyopathies, pericarditis, progressive muscular dystrophy, amyloidosis and rheumatic heart disease. Cases of familial disease of the sino-atrial node have also been described, and others have been associated with diabetes mellitus and thyroid disease. No association of SSS and Charcot-Marie-Tooth disease could be found in the literature.

In patients with SSS and a permanent pacemaker thorough pre-operative evaluation is necessary, for the patient with a permanent pacemaker usually has underlying heart disease. Ventricular dysrhythmias are also said to be more common in patients with pacemakers, and the elderly patient probably has concomitant ischaemic heart disease, hypertension and respiratory disease. These patients may also be taking a variety of drugs, such as antihypertensive agents, digoxin, diuretics or β-blockers. It is important to be sure of the type of pacemaker (fixed-rate or demand) and that it is in working condition. Assessment of proper pacemaker function also involves careful review of the pre-operative ECG. To determine whether a pacemaker is functioning properly, the patient’s pulse rate is measured and the ECG examined; each beat paced must coincide with the pulse beat. During anaesthesia, pacemaker function may be influenced by electromagnetic interference, direct electromagnetic interference being the most common type found in the operating-theatre.

To minimize the possible adverse affects of electrocauterization on the pacemaker patient, the University of Michigan Medical Centre has recommended a number of preventive guidelines. Careful observation of the patient during anaesthesia is very important, and the electrical-mechanical function of the heart must also be monitored, by the ECG and by feeling the pulse, using a pulse monitor or via an oesophageal stethoscope. A ventricular complex followed by a pulse wave shows that electrical activity of the heart is followed by ventricular contraction and ejection. An irregular or fast peripheral pulse can indicate interference with the normal pacemaker function. The pacemaker can fail during surgery because of hypokalaemia, hyperkalaemia, myocardial infarction, the use of diathermy, and the administration of a depolarizing muscle relaxant. Precurcarization can attenuate the problem associated with suxamethonium but the effectiveness of this has not yet been confirmed. Pacemaker function can also be influenced by shivering. Diazepam can be used. Various intravenous induction agents (etomidate, thiopentone and ketamine) can be used with safety for a patient with a permanent pacemaker. The non-depolarizing muscle relaxant pancuronium can also be used, and so can inhalational anaesthetic agents provided there are no other contraindications. It must be remembered, however, that no single protocol is suitable for all patients with pacemakers. The choice of anaesthetic agents depends entirely on the extent of organ involvement and the experience and preferences of the anaesthetist. It is based on the type of surgery, medical status of the patient, contraindications to the use of certain drugs, and surgical risks. Constant monitoring is the key to optimal management. Normally the pacemaker is not the problem: the real problem is the underlying disease.

REFERENCES