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CASE REPORT

Perioperative hazards in myotonic dystrophy

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Summary

A 32-year-old man with myotonic dystrophy underwent a thoracotomy for removal of a mediastinal thymoma. Pre-operative examination revealed features of myotonic dystrophy, the only other abnormality was mild restrictive pulmonary disease. Anaesthesia was induced with thiopentone and maintained with enflurane, nitrous oxide, oxygen and curare. Following surgery, the patient was mechanically ventilated for several hours and remained intubated for 9 hours. The anaesthesia and surgery were tolerated well; however, postoperative complications included refractory dysrhythmias, hypoxia, pneumoccocal pneumonia and pulmonary emboli. More intensive preoperative pulmonary evaluation and physiotherapy coupled with more aggressive postoperative pulmonary care might have resulted in a smoother recovery phase.

Key words

Complications; myotonia dystrophica.

Myotonic dystrophy is a multisystem disease requiring the skill and knowledge of the anaesthetist in dealing with organ dysfunction and unique drug responses. Intraoperative problems have received considerable attention in the literature; however, the postoperative period is also hazardous and the care during this time has been under-emphasised. An illustrative case is presented to demonstrate the need for the involvement of the anaesthetist during the pre- and postoperative periods.

Case history

The patient, a 32-year-old white male, presented for evaluation of a mediastinal mass. He had myotonic dystrophy from the age of 17 and at admission he presented with the classical features (Fig. 1): frontal baldness, ptosis, lenticular opacities, expressionless face, wasted sternocleidomastoids, peripheral muscle weakness, areflexia of the limbs, testicular atrophy and dysphagia. His father and one brother also have myotonic dystrophy. He had smoked 20 cigarettes a day for 15 years but denied frequent respiratory infections or chronic bronchitis. The patient had an uneventful tonsillectomy under general anaesthesia in childhood, and recently had viral pericarditis complicated by a pericardial effusion and dysrhythmias.

On examination his weight was 66 kg, blood pressure 134/70 mmHg, pulse 68 and regular.

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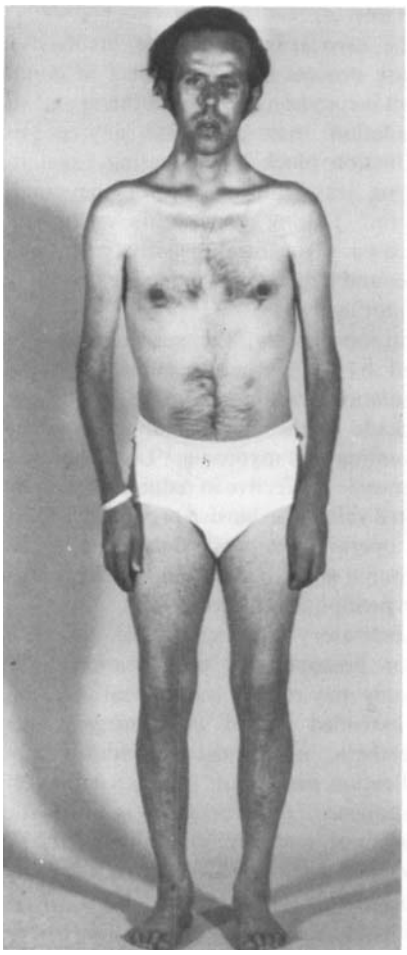

FIG. 1. Physical appearance of patient.

Haematological and serological studies were normal except for an unexplained prothrombin time 70% of normal. Pulmonary function studies showed a maximal voluntary ventilation of 85% of the predicted normal value and a forced expiratory volume of 51% of the predicted volume, with arterial blood gases: pH 7·41, PaO_2 75 mmHg, and $PaCO_2$ 46 mmHg. Electrocardiogram showed non-specific widening of the T waves and the chest X-ray revealed the mediastinal mass as the only abnormality.

On the day of his thoracotomy the patient came to a warmed operating room without premedication and was preoxygenated for 10 min. An induction dose of 100 mg of thiopentone rendered the patient apnoeic, though able to breathe on command. An additional 200 mg of thiopentone were given intravenously and the patient was ventilated by mask with oxygen and enflurane for 15 min. The trachea was then intubated without the use of any muscle relaxant. He was maintained on nitrous oxide, oxygen and enflurane. Movement necessitated incremental doses of curare totalling 25 mg during the 3½ hour procedure. Except for runs of premature ventricular contractions as the tumour was stripped from adherent pericardium, a thymoma was removed uneventfully from the mediastinum. (The subsequent neostigmine test ruled out myasthenia gravis as a diagnosis.) At the end of the procedure the patient was alert, making weak respiratory efforts, and indicating pain. Fentanyl 0·05 mg was injected intravenously and the patient ceased to make voluntary ventilatory efforts. He was transferred to the intensive care unit and placed on mechanical ventilation.

Early in the postoperative period the patient developed frequent premature ventricular contractions but maintained an acceptable blood pressure. Nine hours postoperatively he extubated himself. Despite 100% supplemental oxygen he had a hypoxic period (PaO_2 47 mmHg) which improved over the next 8 hours. His cough was weak and ineffective and suction produced thick yellow secretions. He was not reintubated.

On the third postoperative day the patient had several episodes of ventricular tachycardia requiring lignocaine, quinidine and procaine amide for their control. Five days postoperatively he became febrile with a deterioration of the arterial blood gases and was found to have pneumococcal pneumonia. On the tenth day the patient complained of severe chest pain and shortness of breath. A scan showed pulmonary emboli to both right and left lower lobes. Thirty days post surgery the patient was discharged still requiring quinidine and procaine amide for control of dysrhythmias. The majority of his postoperative period was spent in the intensive care unit.

Several months after discharge the patient received facial trauma. Examination at that time revealed an asymptomatic maxillary tumour thought to be an odontoma. He was admitted for surgical removal under general anaesthesia. Regional blockade was not considered a possibility because of the location of the tumour and the patient's inability to cooperate.

Pulmonary function studies showed a deterioration from the previous admission and were compatible with restrictive lung disease: vital capacity 68% of predicted normal, forced vital capacity 66%, forced expiratory volume 1 second 63%, and maximal voluntary ventilation 47%. Arterial blood gases were: pH 7.36, PaO_2 74, $PaCO_2$ 46. Holter monitoring* showed sinus rhythm interrupted by frequent ventricular unifocal premature ventricular depolarisations occurring in a pattern of ventricular bigeminy. After discussions involving the surgeon, patient, and anaesthetist, it was decided that the patient represented too high a risk for elective surgery and that the tumour would be followed clinically.

Discussion

This case typifies many of those reported in the anaesthetic literature and exemplifies the numerous problems, particularly in the postoperative period, when a patient with myotonic dystrophy undergoes general anaesthesia.[1–4]

The two major intra-anaesthetic problems are maintenance of adequate ventilation and control of cardiac dysrhythmias. Some patients respond to the depolarising muscle relaxant succinylcholine in an abnormal fashion.[3,5,6] Instead of producing flaccid paralysis, the drug may cause persistent contracture involving the muscles of the chest wall making intubation or ventilation impossible.[3] Fortunately the duration of the contracture is approximately that of the action of succinylcholine, about 3–4 min, and is not life-threatening provided that the patient has been preoxygenated.[3] The response to the non-depolarising muscle relaxants is normal; however, reversal of these drugs by neostigmine may precipitate myotonia.[6,7]

Many of these patients demonstrate sensitivity to the respiratory depression of barbiturates, narcotics, and diazepam.[6,7] Since patients with myotonic dystrophy may have impairment of ventilation secondary to atrophy or myotonia of the diaphragm and the accessory muscles of respiration, drug-induced central depression may cause severe hypoventilation or apnoea. These drugs must be used with caution as premedicants or in the postoperative period when mechanical ventilation is not readily available.

The myocardium may be involved in the disease process and some form of conduction defect is common.[2,7,8] Anaesthesia and surgical stimulation may aggravate any pre-existing conduction block by increasing vagal tone or causing transient hypoxia of the conduction system.[9] During anaesthesia, there may be increased myocardial sensitivity to catecholamines and hypercarbia requiring careful cardiac monitoring.

Intraoperatively, the surgeon may be hindered by a myotonic response to mechanical stimulation. Neither regional anaesthesia nor blockade of the motor endplate is effective in preventing this myotonia.[7] Local infiltration of the muscle is effective in reducing spasm but is of limited value in extensive procedures.[7] Warming the operating room reduces the severity of myotonia and the incidence of shivering which may precipitate myotonia.[7]

Respiratory insufficiency or failure is the major postoperative complication and these patients may require mechanical ventilation for an extended period after surgery. Residual anaesthetic, neuromuscular blockade, or pain medication may result in hypoventilation. As a consequence of weakness in the thoracic, pharyngeal, and laryngeal muscles, the patients may have diminished cough reflexes rendering them unable to raise secretions or prevent aspiration, leaving them vulnerable to bronchopneumonia and atelectasis.[1,10,11] In patients given a general anaesthetic, the tracheal tube should remain in place until all effects of the anaesthetic have dissipated, the patient has the muscular strength to cough, and large doses of narcotics are no longer required for pain. Postanaesthetic regional blockade may obviate the need for narcotics. After extubation, physiotherapy should be instituted early and at the first indication of secretion difficulties or hypoventilation, the patient should be reintubated. Infections must be recognised early and treated vigorously.

Preoperatively these patients should undergo cardiac evaluation, including Holter monitoring as any electrocardiogram abnormalities may be intermittent.[8] Current pulmonary infection must be identified and treated with antibiotics, pulmonary therapy, and instruction in breathing exercises. Arterial blood gases and pulmonary

* Holter monitoring is a continuous electrocardiographic recording for a period of 24 hours in an ambulatory patient.

function studies should be obtained and a particular request made for the maximal expiratory force as this is the index most frequently reduced.[10]

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