Aspiration pneumonia and coma - an unusual presentation of dystrophica myotonia

Vincent M. Hannon MB B CH FFARCSI,
Anthony J. Cunningham MB B CH FFARCSI DABA FRCPC,
Michael Hutchinson B SC MB MRCP,
Walter McNicholas MD MRCPI FRCPC

A 30-year-old female patient presented in a comatose state with clinical and radiographic signs of aspiration pneumonia 16 hours following elective surgery. Subsequent clinical assessment and investigations revealed the characteristic facies, proximal muscle weakness, lenticular opacities, pulmonary function defects, arterial desaturation and abnormal breathing during rapid eye movement (REM) sleep often associated with myotonia dystrophica. Although these characteristic features were evident on clinical examination postoperatively they were not noted in the preoperative assessment. The aspiration pneumonia and coma were unusual presenting features of this disease.

Unsuspected myotonia dystrophica should be considered in the differential diagnosis of unexplained respiratory depression, aspiration or comatose state following surgery. Recognition of the disorder during the preoperative assessment is the key to avoiding complications during the perioperative management of such patients.

Key words
COMPLICATIONS: aspiration pneumonitis,
MUSCULOSKELETAL: myotonic dystrophy.

From the Departments of Anaesthesia, Neurology and Respiratory Medicine, St. Vincent's Hospital, Elm Park, Dublin, Ireland.

Address correspondence to: A. J. Cunningham, Professor of Anaesthesia, Royal College of Surgeons in Ireland, Dublin 2, Ireland.

CAN ANAESTH SOC J 1986 / 33: 6 / pp 803–6

Myotonia dystrophica is a degenerative disease involving smooth, skeletal and cardiac muscle. The problems associated with general anaesthesia in these patients are well described.[1] With prior knowledge of the disease many of these problems can be avoided.[2–4] However, the disorder may present as unexplained hypoventilation in a previously undiagnosed patient following anaesthesia. The multisystem nature of this disease must be considered when administering anaesthesia to known or suspected cases.

Case report

A 30-year-old female patient weighing 56 kg presented for elective left ovarian cystectomy and uterine suspension in a gynaecological hospital. Her only previous surgery was an uneventful laparoscopy three months previously for investigation of infertility. Her past medical history included rheumatic fever at age four years and tuberculosis at age nine years. She was not taking any medications. She gave a history of having developed a rash after taking penicillin ten years earlier. There was no family history of problems during anaesthesia and four siblings were alive and well. No abnormal findings were detected on general systems review or on physical examination. Routine haematological and biochemical tests were also within normal limits.

Meperidine 50 mg and promethazine 25 mg were administered intramuscularly one hour prior to surgery. Anaesthesia was induced with thiopentone 250 mg and fentanyl 100 μg intravenously. Vecuronium 8 mg was administered to facilitate endotracheal intubation. Anaesthesia was maintained with 70 per cent nitrous oxide/30 per cent oxygen. The duration of surgery was one hour and fifteen minutes at the end of which neostigmine 2.5 mg and atropine 1.2 mg were administered to reverse the

residual neuromuscular blockade. The patient was extubated and transferred to the recovery room. During the 35-minute observation period in the recovery room she was noted to be awake, alert and able to sustain head lift for five seconds.

She received papaveretum 20 mg intramuscularly on three occasions at four hourly intervals after surgery. Sixteen hours following surgery the patient was noted to be comatose and cyanosed, blood pressure was 80 mmHg systolic and pulse rate 130 beats·min^{-1}.

Initial arterial blood gases revealed profound hypoxaemia (PO_2 6.19 kPa) and severe metabolic and respiratory acidosis (pH 6.91, PCO_2 9.26 kPa, HCO_3 9.18 mmol, base excess (BE) – 17.3 mmol·L^{-1}). Naloxone 0.8 mg in divided doses, dexamethasone 16 mg and sodium bicarbonate were given, without improvement in clinical condition. The patient was intubated and intermittent positive pressure ventilation (IPPV) with positive end expiratory pressure (PEEP) commenced. Six hours later the patient's respiratory and metabolic status had improved. However, she remained comatose with decorticate posturing, hyper-reflexia of both lower limbs and bilateral extensor plantar responses. On chest auscultation there was decreased air entry in the left apex. Chest *x*-ray confirmed left upper zone consolidation with basal and mid-zonal shadowing consistent with aspiration. The patient was then transferred to this institution.

The differential diagnosis at that time included hypoxic cerebral insult and subarachnoid haemorrhage. Computerised axial tomography (CAT) scan was done and showed signs of moderate cerebral oedema. There was no evidence of intracranial bleeding. An urgent neurological consultation was requested. Ventilatory support was continued for 24 hours postintubation and thereafter the patient breathed spontaneously via the endotracheal tube for 48 hours. At this time the patient's clinical condition had improved sufficiently to allow extubation. During this time the patient regained consciousness. She was able to speak clearly and answer questions following extubation.

While in the intensive care unit, the patient was noted to have facial weakness, bilateral ptosis, bilateral sternocleidomastoid weakness and weakness of the hand muscles and extensor muscles of the fingers with a normal hand grip. Frontal balding was also noted. Slit lamp examination of the lens revealed dust opacities in the posterior cortex and a stellate polar opacity in the left lens. A 12-lead electrocardiogram was within normal limits. Electromyographic studies were normal as was a Tensilon (edrophonium) test used to rule out the possibility of myasthenia gravis.

On direct questioning of the patient and her immediate family during this time a history that she had complained of lethargy, malaise and daytime somnolence during the preceding three years was elicited. The patient's presenting complaint of infertility, the hand muscle group weakness, bilateral ptosis and pathognomic ocular lens changes suggested the diagnosis of dystrophica myotonia. The patient was discharged from the intensive care unit on the eighth postoperative day and from the hospital on the twentieth postoperative day.

She returned four months later for pulmonary function testing and full laboratory sleep studies. The spirometry studies showed a mild restrictive and obstructive type breathing pattern with a forced vital capacity (FVC) of 2.2 litres (73 per cent predicted) and a forced expired volume at one second (FEV_1) of 1.55 litres (70 per cent of the FVC). An overnight sleep study was performed using standard polysomnographic techniques.[5] Respiration was measured using respiratory inductance plethyomyography (Respitrace) and arterial oxygen saturation (SaO_2) was also recorded. Awake ventilatory responses to hyperoxic progressive hypercapnia were measured in duplicate by a modified Read rebreathing technique.[6]

The patient maintained long periods of both slow wave and rapid eye movement (REM) sleep. No significant apnoea was recorded during any phase of sleep. During REM sleep SaO_2 fell to less than 70 per cent, compared to greater than 90 per cent during non REM sleep. A reduction in the diaphramatic component of respiration was demonstrated during REM sleep with normal rib cage movement. Awake hypercapnic ventilatory response was within normal range (4.4 L/min/% rise in CO_2) with a correlation coefficient by linear regression analysis of 0.994.

Discussion

Of the three myotonic syndromes, myotonia congenita, paramyotonia and dystrophica myotonia the latter is the most common. It is a familial disease of unknown aetiology inherited as an autosomal domi-

nant trait with a peak incidence in the second to fourth decade. The severity of the disease has been classified into four grades ranging from mild to severe.[8] Had the diagnosis been made preoperatively this patient would have been classified as grade 1 disability. No family history of muscle disease or adverse reactions to general anaesthesia was elicited and a previous uneventful general anaesthetic course was noted. However, if her complaints of lethargy, malaise and drowsiness elicited on direct questioning together with the characteristic muscle group wasting had been noted by the attending physician and anaesthetist preoperatively, the catastrophic complications encountered might have been avoided. The key to the prevention of complications lies in the recognition of the problem preoperatively and in an awareness of all the problems associated with the anaesthetic management of these patients at all stages of the disease.

As the myotonia progresses all muscle types are affected. Skeletal muscle involvement may be widespread throughout the body. When coupled with decreased brainstem neurogenic drive this may lead to terminal respiratory complications.[9,10] Weakness of the pharyngeal and laryngeal muscles and of the oesophageal sphincters can result from smooth muscle involvement.[11] Cardiac involvement is also an integral part of the disease with conduction defects affecting the infranodal system[12] the sinus node and the myocardium.[13,14]

Abnormal neuromuscular responses to depolarizing muscle relaxants are well described in these patients.[15–17] The response to competitive relaxants appears normal.[18] The patient did not receive a depolarizing relaxant during either anaesthetic.

Patients with myotonia dystrophica also have increased sensitivity to the respiratory depressant effects of narcotics, anaesthetic induction agents and volatile agents.[18,19] Dundee[19] suggested that thiopentone may have a peripheral action on the muscle itself. However, it is more probable that the excessive respiratory depression is because of an additive effect to the already diminished respiratory and cardiac reserve in these patients.

Sleep studies performed on these patients have demonstrated decreases in SaO_2 levels, thought to be due to central respiratory dysfunction, with resultant sleep apnoea during REM sleep.[20,21] Our patient had a decrease in SaO_2 to 70 per cent; however, it was not due to sleep apnoea but most probably a fall off in the diaphramatic component of breathing during REM phase of sleep, i.e., a weakness of the actual respiratory musculature. Abnormal ventilatory response to hypercarbia has also been demonstrated;[10] however, our patient showed a normal hypercapnic response on rebreathing. Upper respiratory airway and gastrointestinal involvement must also be considered as possible causative factors of the aspiration in this patient. Incomplete abduction of the vocal cords, incomplete closing of the larynx during deglutition, weakness of the pharynx with complete loss of cricopharyngeal sphincter have all been demonstrated in fully alert patients with severe forms of this disease.[11,22,23]

In this patient we documented mild restrictive and obstructive lung disease and diaphramatic weakness during REM sleep with resultant alveolar hypoventilation. Failure to recognise the diagnosis preoperatively, the above factors, plus the recognised sensitivity of these patients to sedatives and narcotics, probably combined to cause the postoperative respiratory depression, pulmonary aspiration and coma in this case.

In order to avoid anaesthetic management problems in patients with myotonic dystrophy, recognition of all the potential problems is essential. In preoperative assessment particular attention should be paid to the gastrointestinal, respiratory and cardiovascular systems. Premedication with narcotics or sedatives should be carefully titrated. All parenteral solutions should be warmed. Local or regional anaesthesia should be employed where possible. If general anaesthesia is to be used, pre-oxygenation and airway protection on induction is mandatory. Induction agents should be administered in incremental doses and depolarizing relaxants avoided. Volatile agents and narcotics should be used with caution peroperatively.

Postoperatively local or regional anaesthesia should be employed for pain relief if possible and the patient should remain in a high density nursing area for 24 hours after general anaesthesia, where possible.

References

1 *Katz J, Benumof J, Kadis LB.* Anesthesia and uncommon disorders. Philadelphia. WB. Saunders 1981; 530–61.

2 *Hook R, Anderson EF, Noto P.* Anesthetic management of a parturient with myotonia atrophica. Anesthesiology 1975; 43: 689–92.
3 *Mudge BJ, Taylor PB, Banderspeck AFL.* Perioperative hazards in myotonic dystrophy. Anaesthesia 1980; 35: 492–5.
4 *Ravin M, Newmark Z, Saviello G.* Myotonia dystrophica–an anesthetic hazard, two case reports. Anesth Analg 1975; 54: 216–8.
5 *Rechtschaffen A, Kales A.* A manual of standardised terminology techniques and scoring system for sleep states of human subjects. Washington D.C. National Institutes of Health, 1968, Publication No. 204.
6 *Read DJC.* A clinical method for assessing ventilatory response to carbon dioxide. Australas Ann Med 1967; 16: 20–32.
7 *Harper PS.* Myotonic dystrophy. Philadelphia, W.B. Saunders, 1979.
8 *Gillan, PMS, Heaf PJD, Kaufmann L, Lucas BGB.* Respiration in dystrophica myotonica. Thorax 1964; 19: 112–20.
9 *Begin R, Bereau MA, Lupun L, Bernier JP, Lemieux B.* Pathogenesis of respiratory insufficiency in myotonic dystrophy. Am Rev Respir 1982; 125: 312–8.
10 *Carroll JE, Zwillich GW, Weil JV.* Ventilatory response in myotonic dystrophy. Neurology 1977; 27: 1125–8.
11 *Pruzanski W, Profis A.* Pulmonary disease in myotonic dystrophy. Am Rev Respir Dis 1965; 91: 874–9.
12 *Pertoff JK, Stevenson WG, Roberts NK, Cabeen W, Weiss J.* Cardiac involvement in myotonic muscular dystrophy (Steinert's Disease): a prospective study of 25 patients. Am J Cardiol 1984; 54: 1074–81.
13 *Paynes CA, Greenfields JC.* Electrocardiographic abnormalities associated with myotonic dystrophy. Am Heart J 1963; 65: 436–40.
14 *Petkovich NJ, Dunn M, Reed W.* Myotonia dystrophica with A–V dissociation and Stokes-Adams attacks. Am Heart J 1964, 68: 391–6.
15 *Dalel FY, Bennet EJ, Raj PP, Lee DG.* Dystrophica myotonia – a multisystem disease. Can Anaesth Soc J, 1972; 19: 36–44.
16 *Peterson IS.* Generalised myotonia following suxamethonium. Br J Anaesth 1962; 34: 340–3.
17 *Cody JR.* Muscle rigidity following administration of succinylcholine. Anesthesiology 1968, 29: 159–62.
18 *Mitchell MM, Hassen HA, Savarese JJ.* Myotonia and neuromuscular blocking agents. Anesthesiology 1975; 49: 44–8.
19 *Dundee JW.* Thiopentone in dystrophica myotonica. Anesth Analg 1952; 31: 257–60.
20 *Cummiskey J, Lynn Davis P, Guilleminault C.* Sleep study and respiratory function in myotonic dystrophy. Ch. 19: Guilleminault C, Dewent WC. Sleep apnoea syndromes. New York Kock Foundation 1978; 295–308.
21 *Cedieux RJ, Kales A, McGlynn TJ, Jackson D, Manders EK, Simmonds MA.* Sleep apnoea precipitated by pharyngeal surgery in a patient with myotonic dystrophy. Arch Otolaryngol 1984; 110: 611–3.
22 *Pierce JW, Creamer B, MacDermott V.* Pharynx and oesophagus in dystrophia myotonica. GUT 1965; 6: 392–5.
23 *Harvey JC, Sherbourne DH, Siegel CI.* Smooth muscle involvement in myotonic dystrophy. Am J Med 1965; 39: 81–91.

Résumé

Une patiente âgée de 30 ans a présenté un état comateux avec des signes cliniques et radiologiques d'aspiration bronchique seize heures après la chirurgie élective. Une évaluation clinique subséquente ainsi que des investigations ont révélé un faciès caractéristique, une faiblesse des muscles proximaux, une opacification du cristallin, une altération des fonctions pulmonaires, une désaturation artérielle ainsi qu'une respiration anormale durant la période du "rapid eye movement" (REM) souvent associés à la myotonie dystrophique. Même si ces caractéristiques étaient évidentes à l'examen clinique postopératoire elles n'étaient pas notées à l'évaluation préopératoire. L'aspiration bronchique et le coma étaient des signes de présentation peu communs de cette maladie.

La dystrophie myotonique non suspectée doit être considérée dans le diagnostic différentiel d'une dépression respiratoire non expliquée, l'aspiration bronchique, ou un état comateux après la chirurgie. La détection du problème en période préopératoire demeure la clé afin de prévenir les complications durant la période périopératoire.