ANAESTHESIA AND ACUTE DERMATOMYOSITIS/ POLYMYOSITIS

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Dermatomyositis, a condition of unknown aetiology characterized by dermatitis, oedema and inflammation of muscle, is deemed to be a clinical variant of the same pathological process as that producing polymyositis [1,2]. Many patients date the onset of their condition from an infection, and in 20% of cases there is an associated neoplasm [3,4]. Dermatomyositis/polymyositis associated with neoplasia is normally acute in onset and is said to be reversible with successful treatment of the neoplasm. Clinically, the condition has the characteristics of a connective tissue disorder and may have features in common with other diseases of this group such as involvement of the joints. In dermatomyositis there may be facial rashes and erythematous cutaneous changes similar to those seen in scleroderma.

The onset of the condition may be acute or insidious. The principal feature is muscle weakness, which may be generalized, but it most commonly presents with bilateral involvement of the pelvic, shoulder and neck muscles. The onset of weakness may be preceded by prodromal symptoms of muscle aches, fever, joint pains and, in dermatomyositis, facial oedema. Involvement of other muscle groups can produce diplopia, dysphagia, ventilatory impairment and facial weakness. Heart failure may occur as a result of myocardial involvement. Death is usually caused by ventilatory failure, heart failure or a complicating infection. Treatment is with corticosteroids.

The problems that dermatomyositis/polymyositis are said to present for the anaesthetist are outlined in the standard textbooks as muscle weakness resulting in ventilatory insufficiency, myocardial involvement and dysphagia with the potential for soiling of the ventilatory tract [5,6]. However, the basis for these statements appears to be largely surmise, since there are only three published case reports of anaesthesia in acute polymyositis. These were all in the Japanese literature and two were published after the above statements were made [7-9].

The anaesthetic management is reported of two patients, one with acute dermatomyositis associated with a neoplasm of his gastrointestinal tract, the other with acute polymyositis and a neoplasm of the epiglottis. Both had severe muscle weakness and presented for resection of their malignancies.

CASE REPORTS

Patient No. 1

A 70-yr-old male (weight 80 kg) gave a 3-month history of weakness of the shoulder and thigh muscles. The onset was apparently sudden, but had been preceded for several days by aching of both muscle groups, and during the ensuing 3...
months the weakness had become worse. By the time he presented for surgery he was unable to rise from the sitting position or to elevate his arms above shoulder level.

He was a type II diabetic controlled on diet and for the previous 6 months had been treated for hypertension with nifedipine and frusemide. On admission his arterial pressure was 160/90 mm Hg. There was nothing else of note in his medical history.

Investigations

Serum concentrations of hydroxybutyrate dehydrogenase, aldolase, alkaline phosphatase, alanine amino transferase, and glutamyl transferase were increased (table I). Measurement of quadriceps strength [10] showed his maximum voluntary contraction strength to be 35.3% of that expected for his body weight. Vital capacity (VC) was 3.2 litre (predicted 3.95 litre) and the ratio of forced expiratory volume in 1 s (FEV₁) to VC was 82%.

Chest x-ray was normal, although on CT scan there was a suggestion of an opacity, “possibly a metastasis” in the posterior basal segment of the right lung. ECG showed some evidence of an old posterior myocardial infarction. Full blood count, glucose tolerance test and blood-gas tensions were normal. Barium enema and colonoscopy revealed a stricture at the junction of the sigmoid and descending colon. He had had no symptoms from this and was scheduled to undergo sigmoid colectomy.

Anaesthesia

Diazepam 10 mg was given by mouth 2 h before anaesthesia was induced with alfentanil 0.5 mg and etomidate 16 mg. Atracurium 20 mg was given initially, then a further 10 mg because the 20 mg proved to be clinically inadequate. Ninety seconds after the second dose the trachea was intubated.

The lungs were ventilated with nitrous oxide and oxygen in a ratio of 2:1, and 0.6% enfurane. The operation lasted 2 h 15 min. Increments of alfentanil 0.5 mg were given every 30 min (total dose 2.5 mg). Neuromuscular transmission was monitored using a peripheral nerve stimulator (750 Digital-Bard Biomedical). Three incremental doses of atracurium 10 mg were given during the course of the operation when neuromuscular function started to recover. Systolic arterial pressure had been 200 mm Hg on arrival in theatre and heart rate 100 beat min⁻¹. The arterial pressure settled to between 140 and 160 mm Hg systolic for the duration of the operation and heart rate to between 85 and 100 beat min⁻¹. No irregularities were noted on the ECG. Enflurane was discontinued 10 min before the end of the operation, spontaneous ventilation was established and residual neuromuscular blockade antagonized with neostigmine 2.5 mg (and glycopyrrolate 0.6 mg). There was a rapid return of full neuromuscular function as denoted by a vigorous sustained contraction in response to a supramaximal stimulus of 100 Hz and no evidence of “fade” in response to sequential stimuli at 2 Hz. Lung volumes were measured before extubation using a Wright respirometer. Tidal volume was 600–800 ml and vital capacity 1200 ml. Negative inspiratory pressure was 20 cm H₂O. The trachea was extubated and the patient transferred to the recovery ward where his cardiovascular and ventilatory state remained stable.

He developed a chest infection in the immediate postoperative period, but this responded to physiotherapy and ampicillin. By the 6th day after operation his muscle weakness had improved dramatically; he was able to stand from the sitting position and raise his arms above his head. He was discharged home 13 days after the operation. He died at home 5 months later from bronchopneumonia. A post-mortem was not carried out.

Patient No. 2

The patient was a 47-yr-old man weighing 85 kg who was admitted for the treatment of bronchopneumonia. Two weeks previously he had had an influenza-like illness, with aching muscles and a headache. This was followed by the progressive onset of muscle weakness and ventilatory symptoms over the week preceding admission. His
pneumonia responded to antibiotics, but a clinical and radiological diagnosis of fibrosing alveolitis was made. The muscle weakness persisted and affected both arms and legs, but was worse in the upper limbs. Although he was able to lift his arms above his head he was unable to hold them there and was able to rise from a squatting position with extreme difficulty. He complained also of hoarseness, and on examination was found to have a carcinoma of the epiglottis.

**Investigations**

Chest x-ray showed evidence of fibrosing alveolitis; ECG was normal. Arterial Po$_2$ was 10.1 kPa, but blood-gas tensions were otherwise normal. Vital capacity was 3.25 (predicted 4.75) litre and FEV$_1$/FVC 72%. Serum concentrations of creatine kinase, hydroxybutyrate dehydrogenase and alanine amino transferase were increased (table I). Muscle biopsy was performed and a diagnosis of acute polymyositis was made.

Prednisolone 80 mg on alternate days was started, with some improvement in his muscle weakness. On examination before surgery his arterial pressure was 140/90 and heart rate 80 beat min$^{-1}$. Ventilatory rate was 20 b.p.m. and he was slightly short of breath at rest. He was scheduled to undergo partial laryngectomy.

**Anaesthesia**

Lorazepam 2 mg by mouth and glycopyrrolate 0.4 mg i.m. were given 1 h before operation. Anaesthesia was induced with thiopentone 350 mg and the patient breathed nitrous oxide, oxygen and isoflurane spontaneously until anaesthesia was deep enough to allow intubation of the trachea using a fibreoptic laryngoscope. A test dose of atracurium 12 mg was given, with insignificant clinical effect. A further 25 mg of atracurium was given, followed by an infusion at a rate of 0.36 mg kg$^{-1}$ h$^{-1}$. Neuromuscular block was monitored as in the first patient. Isoflurane was discontinued and alfentanil 4 mg given over 10 min, followed by an infusion of 0.5 µg kg$^{-1}$ h$^{-1}$. Artificial ventilation with nitrous oxide and oxygen in a 2:1 ratio and 0.6% enfurane was maintained throughout the procedure. During the course of the operation attempts were made to reduce the rate of the infusion of atracurium, but these were unsuccessful and two further doses of atracurium (10 mg and 15 mg) had to be given at different times to maintain satisfactory neuromuscular blockade. Arterial pressure was monitored using an indwelling cannula, and central venous pressure through a catheter inserted to the basilic vein. The cardiovascular system was stable over the course of the operation and no ECG abnormalities were observed.

Surgery lasted 4 h and both infusions were stopped 30 min before the end. Neuromuscular blockade was antagonized with neostigmine 2.5 mg and glycopyrrolate 0.6 mg, and naloxone 0.2 mg was given to counteract mild ventilatory depression. Tidal volume on recovery was 800 ml and VC 2.5 litre. The trachea was extubated and the patient transferred to the intensive care unit at 16.00 h, breathing 25% oxygen. His Pa$_2$ was 12.0 kPa and Pa$_{CO_2}$ 5.1 kPa. At 23.00 h that evening he was still breathing 25% oxygen; Pa$_2$ was 15.7 kPa and Pa$_{CO_2}$ 4.9 kPa. Tidal volume at this stage was 800 ml and VC 3 litre. He remained stable and was discharged to the ward the following day, where his recovery continued uneventfully. He was discharged from hospital 12 days later, receiving prednisolone 70 mg daily. His muscle weakness had improved and on discharge he was able to rise from the squatting position and to hold his arm above his head, although subjectively he felt tired and weak.

**DISCUSSION**

In common with three previous case reports [7–9], none of the problems outlined in the standard textbooks [5, 6] was experienced in the management of either of these two patients. In neither instance was there any clinical evidence of undue sensitivity to neuromuscular blocking drugs, ventilatory function appeared to recover in a normal fashion after surgery, and there was no evidence of myocardial dysfunction.

Feldman [11] stated that some improvement in the muscle weakness seen in polymyositis occurs with the administration of an anticholinesterase drug, but gave no supportive evidence. He also reported that two of a series of 10 patients with the condition showed evidence of a myasthenic response to decamethonium, but with no reference to published reports. The fact that the condition is sometimes associated with a malignancy, and that malignancy itself can be associated with defects in neuromuscular transmission, does mean that these patients could be particularly sensitive to neuromuscular blocking agents [5]. In both instances, a relatively small dose of atrac-
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Curium was given initially and a further dose only when the original proved to have little effect. In view of the nature of the disease—an acute inflammation of muscle and the possibility of hyperkalaemia—it seemed wise not to give suxamethonium.

The disease in both patients was of recent onset. In the first it did not involve the pharyngeal muscles, so the question of pulmonary aspiration did not arise. The second patient presented with pneumonia in addition to his muscle weakness, but also proved to have carcinoma of his larynx. Polymyositis involving the pharyngeal muscles or local pathology of the laryngopharnyx could both potentially cause aspiration. This patient also had chronic chest disease, so any of these three conditions could have been the cause of his pneumonia.

One difficulty in these patients is to determine if ventilatory function is impaired by the muscle weakness. Although the first patient was not breathless at rest and had normal blood-gas tensions, an individual who is so weak that he cannot stand is incapable of stressing his respiratory system, so any assessment of, for example, exercise tolerance is impossible. The second patient was breathless at rest, but had chronic chest disease. The FEV₁/FVC ratio does give some indication of ventilatory power, and in both patients the ratio was normal. Tests such as maximum breathing capacity and peak expiratory flow rate may have provided more sensitive indices of ventilatory reserve, but these were not performed. After operation, ventilatory function in both cases appears to have recovered normally. No formal measurements were made in the first patient beyond the immediate postoperative period as there were no indications to do so, but lung volumes and inspiratory force at that stage were satisfactory and progress thereafter appeared to be normal. Lung volumes and blood-gas tensions in the second patient were measured later and these had improved.

The newer short acting neuromuscular blockers and analgesic agents have obvious advantages, with a wider margin for error over the traditional agents such as morphine or tubocurarine when anaesthetizing patients whose cardiorespiratory reserve is impaired or patients who may respond unpredictably to anaesthetic or analgesic drugs. They can also be given by continuous infusion, allowing control of analgesia and neuromuscular blockade at a lower dose than with repeat bolus administration [12]. No objective measurement was made, in either patient, of neuromuscular transmission, but it was monitored in the conventional fashion, observing the power of the contraction elicited in response to tetanic stimuli and the presence of fade in response to repeated stimuli. Neither patient appeared to be unduly sensitive to neuromuscular blocking drugs, and the doses of these drugs in the three subjects reported previously [7–9] were all within the normal range, so again there was no evidence of undue sensitivity.

In the three previous reports of anaesthesia for patients with dermatomyositis [7–9] extradural analgesia was used during operation and for postoperative pain relief. The patient reported by Kato, Aruga and Wakasugi [7] had severe muscle weakness and was unable to stand, but ventilatory function was normal. Anaesthesia was induced with a mixture of fentanyl and droperidol, suxamethonium was given to facilitate intubation of the trachea and pancuronium 6 mg was given in 1–2 mg increments for muscle paralysis. They recommended avoiding suxamethonium in the acute form of the disease, stated that long-acting myoneural blockers should be given in “small divided doses” and that it should be assumed that all patients have myocarditis. Their patient had no ventilatory problems in the postoperative period. The patient described by Kashimoto and colleagues [9] had pre-existing chronic chest disease with FEV₁/FVC = 44%. They used a similar technique and, despite the pre-existing ventilatory impairment, no problems were experienced. Ogino’s patient [8] was given thiopentone, pancuronium and extradural analgesia and, again, no immediate problems occurred. The patient died 3 months later from bronchopneumonia precipitated by the immunosuppression produced by chemotherapy. One of the subjects of the present report also died several months after the operation. In all likelihood this was coincidental, although it is not possible to rule out entirely any connection between his death and his dermatomyositis.

From the report of these two patients and the three previous ones it appears that, provided elementary routine precautions are taken, the severe muscle weakness seen in acute dermatomyositis/polymyositis in itself need present few problems for the anaesthetist. More reports would have to be published, however, before the many other potential problems associated with
this condition that are outlined in the standard textbooks could be discounted.

REFERENCES