Case report

Successful treatment of dermatomyositis complicated by ventilatory failure

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SUMMARY A case is reported in which ventilatory failure complicating dermatomyositis was successfully managed with artificial ventilation for 18 days while remission was achieved with methotrexate and prednisolone. Such ventilatory failure is reversible and requires an active treatment.

The development of respiratory failure during the course of dermatomyositis is often regarded as a sinister event with a very poor prognosis. Barwick and Walton¹ described a patient who was artificially ventilated on 2 separate occasions but who eventually died of the disease. Rose and Walton² reported the case a woman who had rapidly progressive weakness and recovered after artificial ventilation for several days. This is a report of a case of dermatomyositis in which ventilatory failure was successfully treated with intermittent positive pressure ventilation (IPPV) while remission was achieved with prednisolone and methotrexate.

Case report

A 45-year-old man was admitted with a history of weakness and shortness of breath of a month's duration. For 6 months he had had a rash on the arms and arthralgias. On examination there was a heliotrope discolouration of the eyelids and a scaly rash on the extensor aspect of the elbows, wrists, and hands. There was marked proximal muscle weakness.

A diagnosis of dermatomyositis was made on the basis of a raised creatine kinase (5767 IU/l: normal <100); an abnormal electromyographic pattern consisting of fibrillation at rest, with a polyphasic pattern of small units; and a muscle biopsy which showed a widespread inflammatory exudate.

Treatment was started with prednisolone 80 mg daily. His condition continued to deteriorate and intravenous methotrexate was added to the regimen.

Soon afterwards he developed increasing dyspnoea and dysphagia. Apart from a poor chest expansion there was no abnormality on examination of the chest, and chest x-ray was normal. The peak expiratory flow rate (PEFR) was 200 l/min. Arterial blood gases on 60% oxygen were: P_{O_2} 23.12 kPa (175 mmHg), P_{CO_2} 9.67 kPa (73.7 mmHg), pH 7.30.

IPPV and fine-bore nasogastric feeding were begun. He developed a pneumococcal chest infection, which was successfully treated with erythromycin. His condition then gradually improved and it was possible to discontinue IPPV after 18 days and nasogastric feeding after a further 3 weeks. On discharge from hospital 10 weeks later he could climb a flight of stairs unaided.

Pulmonary function tests on recovery showed no evidence of intrinsic pulmonary disease.

Discussion

The respiratory complications of dermatomyositis include ventilatory failure due to muscle weakness; pulmonary fibrosis; and pneumonia, both due to hypostasis and aspiration. During the acute illness the patient was too unwell to undergo pulmonary function testing. Clinically there was no evidence of intrinsic pulmonary disease at the time of his developing respiratory failure, and this was confirmed by pulmonary function tests on recovery. The respiratory failure was therefore considered to be due to muscle weakness.

It is of interest that at the time of his requiring artificial ventilation the PEFR was 200 l/min and therefore failed to indicate the severity of the

respiratory failure as shown by the arterial blood gases. There is no substitute for blood gas estimation in this situation.

This case illustrates the reversibility of the muscle weakness in severe dermatomyositis, and shows that active management, including assisted ventilation and treatment of chest infections, is worthwhile regardless of the severity of the muscle weakness.

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References

- 1 Barwick D D, Walton J N. Polymyositis. *Am J Med* 1963; **35**: 646–60.
- 2 Rose A L, Walton J N. Polymyositis: a survey of 89 cases with particular reference to treatment and prognosis. *Brain* 1966; **89**: 747–68.