Chronic inflammatory demyelinating polyneuropathy and respiratory failure

Introduction

Neuromuscular respiratory failure is uncommon in chronic neuropathies including inflammatory demyelinating polyradiculoneuropathy (CIDP). In fact, the presence of respiratory failure and mechanical ventilation might question the accuracy of the diagnosis of CIDP. A recent thorough review on several forms of CIDP did not mention respiratory failure in newly proposed criteria nor did a task force of the American Academy of Neurology [6, 7]. We report 4 cases of CIDP and neuromuscular respiratory failure, which required admission to an intensive care unit (ICU).

Case reports

These patients were seen in two medical centers over a 20-year time span. We excluded patients with a CIDP and a concurrent illness and evidence of a monoclonal gammopathy.

Case 1

A 79-year-old woman in previous good health was referred with generalized weakness, prominent in proximal muscles, absent reflexes and mild distal lower limb sensory changes progressing slowly over six weeks. Nerve conduction studies (NCS) showed relatively normal distal amplitudes with marked slowing of distal latencies and conduction velocities. CSF proteins were 55 mg/dl. The patient started prednisone, azathioprine, and monthly intravenous immunoglobulin (IVIG) and improvement occurred. Twelve months later, the patient worsened over 3 months after she had discontinued medication. The muscle strength was markedly reduced in both proximal and distal muscles but no facial or bulbar weakness were noted. Within 48 hours of admission, the patient's level of consciousness declined rapidly. PaCO₂ was 150 mmHg (2 kPa). A chest radiograph was normal. She was quickly intubated and a tracheostomy was placed for one month with subsequent successful weaning. Two courses of intravenous immunoglobulin (IVIG) were given. The patient's strength progressively improved.
Case 2

A woman aged 57 years had presented with a 4-week history of muscle weakness that was initially diagnosed as Guillain-Barré syndrome (GBS) with subsequent improvement. Following numerous relapses, associated with elevated CSF protein and prolonged nerve conduction, the diagnosis of CIDP was made. During the patient’s second admission, the patient developed increasing weakness. Within a week, she was admitted to the ICU because of concern with her breathlessness when supine: a forced expiratory volume in one second (FEV1) was 0.4 and forced vital capacity (FVC) was 1.0 liter. A chest radiograph was normal. No oropharyngeal weakness or facial weakness was noted. The patient was monitored in the ICU for 1 week without requiring intubation or tracheostomy and gradually improved while daily IVIG was given for 5 days.

Case 3

A 71-year-old man presented with paresthesia and upper limb weakness after a respiratory infection. CSF proteins were 94 mg/dl, electrophysiology studies showed slow velocities with preserved amplitudes. He improved following IVIG. Slow return of weakness in the upper limbs led to another treatment with IVIG and corticosteroids were started. The patient’s course fluctuated greatly over 5 months after the initial onset of weakness and he became eventually quadriplegic. Blood pressure fluctuations developed and mechanical ventilation and tracheostomy were needed. During this episode, electrophysiological studies showed severe axonal involvement. The patient received plasmapheresis, IVIG and IV cyclophosphamide. He improved gradually but fully. He was asymptomatic for 8 months until another relapse and required monthly IVIG to maintain mobility and good strength.

Case 4

A 68-year-old white male developed progressive paresthesias and gradual weakness. While receiving prednisone 120 mg daily, an EMG showed a diffuse peripheral neuropathy with absent F-wave latencies, decreased motor conduction velocities, and positive sharp waves and fibrillations on needle EMG. Ten months since his onset, he further deteriorated and was unable to walk. On admission, he had normal cranial nerves; there was diffuse weakness in the proximal and distal musculature. He had a glove and stocking distribution sensory loss, and areflexia. He had difficulty in breathing and marginally abnormal respiratory parameters, FVC of 1.8 liters, PE-max of 110 mmHg, and PI-max of –90 mmHg. The breathing difficulty further declined over the next 3 days, and he eventually was resuscitated for respiratory arrest. His respiratory parameters showed a vital capacity of 750 mL, PE-max of 20 mm Hg, PI-max of –10 mm Hg. He received a tracheostomy and was ventilated for 98 days. A sural nerve biopsy showed decrease in myelinated fibers with perivascular inflammatory cells. Many teased fibers showed segmental demyelination and remyelination consistent with a diagnosis of CIDP.

Discussion

Our cases fulfilled recently proposed criteria for CIDP [6]. While CIDP and GBS are similar in some respects, they are generally regarded as distinguishable by several clinical features. Clinical features that are common in GBS but rare in CIDP include dysautonomia and oropharyngeal weakness and most prominently respiratory failure. Some authors have described overlapping cases; few required ICU support [3, 4]. Rarely phrenic nerve palsy may occur in CIDP requiring mechanical ventilation [8]. Features in the ICU more likely to be found in GBS include a locked-in picture and progression with corticosteroid treatment while multiple relapses might favor CIDP. A recent review of 67 patients with idiopathic and monoclonal protein associated CIDP (including 12 patients with a relapsing GBS pattern) 6 patients had respiratory failure requiring ventilatory support during the course of their illness. Details on respiratory failure were not provided [2]. Occasionally CIDP may present acutely or subacutely, simulating GBS and requiring ventilatory support [3].

Patients with CIDP are at risk of primary pulmonary complications, similar to GBS [5], and other systemic medical conditions that may lead to ICU admission. Treatment may lead to opportunistic infections or plasmapheresis-associated central line complications such as pneumothorax [1]. The use of immunosuppressive agents may predispose these patients to opportunistic infections, some of which may be critical.

Our cases however serve as a reminder that neuromuscular respiratory failure may occasionally occur during the course of CIDP. However, in our practice at the Mayo Clinic, CIDP is commonly diagnosed, and these cases with respiratory failure seem exceptional. It is possible that respiratory failure was not considered clinically in one of our cases and led to hypercarbia. Studies on pulmonary mechanics and electrophysiological studies of the phrenic nerve and diaphragm would be needed in patients with progressive CIDP to determine if neuromuscular respiratory failure is prevalent or truly rare.