Commentary

Myasthenia gravis is the prototype autoimmune disease of the nervous system, predominantly affecting young women and older men. A small proportion of patients with generalized autoimmune myasthenia gravis may develop acute neuromuscular respiratory failure due to myasthenic crisis. Very rarely, myasthenic crisis may be the clinical presentation of a patient with myasthenia gravis.

In this issue of the journal, Sharma and colleagues[1] report a case of acute respiratory failure in a 22-year-old young woman with autoimmune myasthenia gravis. This patient had bulbar symptoms of unrecognized myasthenia gravis for 3 months before presenting with respiratory failure. The usual cause of myasthenic crisis is neuromuscular weakness affecting respiratory muscles and upper airway muscles because of bulbar myasthenia or a combination of weakness affecting both groups of muscles.[2] The case illustrates an important learning point that failure to recognize and treat bulbar myasthenia could lead to a potentially life-threatening condition requiring respiratory assistance. The overall in-hospital mortality rate of myasthenia crisis even in the best possible setting of intensive care units in the West is about 4.5%,[3] similar to the mortality of acute respiratory failure associated with Guillain Barre syndrome.

In a recent publication from the Mayo Clinic, it was observed that among patients with neuromuscular failure without known diagnosis before admission have poorer outcomes.[4] In this study, nearly a third of all patients admitted to the intensive care unit with acute neuromuscular respiratory failure were finally diagnosed with myasthenia gravis; this number was over twice higher than the number of patients who had a final diagnosis of Guillain Barre syndrome. Clearly, the diagnosis of myasthenia gravis should be on the top of the list in a young woman presenting with acute neuromuscular respiratory failure and well-preserved tendon reflexes without any previous history of muscle disease or toxic exposure (e.g. organophosphate poisoning, snake envenomation) and normal serum level of creatine kinase (CK).

Early diagnosis of myasthenic crisis is always important and high-dose corticosteroids in combination with plasma exchange or human intravenous immunoglobulin (IVIg) remain the cornerstone of treatment.[2] The trend of using IVIg for myasthenia crisis has significantly increased as compared to plasma exchange in recent years,[3] partly due to the ease and convenience of administration and timeliness of availability of IVIg in most medical centers. In the reported case,[1] the therapeutic response to steroids and IVIg was highly satisfactory, leading to a happy outcome.

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References

1. Sharma SR, Sharma N, Yeolekar ME. An undiagnosed myasthenia gravis presenting as isolated recurrent acute respiratory failure. J Neurosci Rural Pract 2012;3:80-2.
2. Chaudhuri A, Behan PO. Myasthenic crisis. QJM 2009;102:97-107.
3. Alshekhlee A, Miles JD, Katriji B, Preston DC, Kaminiski HJ. Incidence and mortality rates of myasthenia gravis and myasthenic crisis in US hospitals. Neurology 2009;72:1548-54.
4. Serrano MC, Rabinstein AA. Causes and outcomes of acute neuromuscular respiratory failure. Arch Neurol 2010;67:1089-94.

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