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Analysis of two cases of myasthenic crisis

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ABSTRACT

Introduction: Myasthenia gravis is an autoimmune disease of the postsynaptic part of the neuromuscular junction. The disease is chronic, manifesting itself in the fatigue of various muscle groups, and the first manifestation of the disease often affects the muscles that move the eyeball and eyelid, although MG can lead to weakness of any muscle group. In turn, myasthenic crisis is a severe worsening of muscle weakness that often results in the urgent need of mechanical ventilation of the patient, due to the respiratory failure.

Case reports: The first patient, 64 years old, female was admitted to the Neurology Department due to breathing difficulties and weakness of the limbs, which suddenly worsened within a dozen of hours before admission. During the stay, steroid therapy was used, as well as six times plasma exchange took place at the Nephrology Department with good treatment tolerance. The dosage of pyridostigmine was also modified, with no adverse effects. During the stay, the patient's condition improved - respiratory disorders resolved, ptosis was significantly withdrawn, and limb muscle tension increased globally. The second patient, aged 67, also female, with a history of myasthenia gravis as well, was admitted to the Neurology Department due to increased fatigue, ptosis, dysphagia, and difficulties in speaking and breathing. Steroid therapy was used, but due to high blood glucose, the steroid dose was reduced and a 2-day cladribine treatment was initiated, with good tolerance.

Discussion: Myasthenic crisis is often a life-threatening condition that requires immediate treatment. Steroid therapy, pyridostigmine, IVIG and plasma exchange are standard approaches in myasthenic crisis. Cladribine is not an ordinary approach, but can be beneficial in some patients.

Keywords: myasthenia gravis; myasthenic crisis; plasma exchange; cladribine.

INTRODUCTION

Myasthenia gravis is an autoimmune disease of the postsynaptic part of the neuromuscular junction, usually characterized by the presence of anti-acetylcholine receptor antibodies as well as some other antibodies e.g. against muscle specific tyrosinase kinase. The disease is chronic, manifesting itself in the fatigue of various muscle groups, and the first manifestation of the disease often affects the muscles that move the eyeball and eyelid, although myasthenia gravis can lead to weakness of any muscle group, and in the most dangerous form respiratory muscle involvement occurs.

In turn, myasthenic crisis is a severe worsening of muscle weakness that often results in the urgent need of mechanical ventilation of the patient, due to the respiratory failure. [1-4]

CASE REPORTS

The first patient, 64 years old, female was admitted to the Neurology Department due to breathing difficulties and weakness of the limbs, which suddenly worsened within a dozen of hours before admission. A patient was diagnosed with myasthenia gravis many years ago. The patient was admitted to the neurological intensive care

subunit. In the neurological examination on the day of admission, the patient was conscious, auto- and allopsychic-oriented, with dysartic speech, but logical contact was maintained, bilateral ptosis, other cranial nerves were normal, global weakness in strength and tension of upper and lower limb muscles was found, Babinski's symptom was negative. During the stay, steroid therapy was used, as well as six times plasma exchange took place at the Nephrology Department with good treatment tolerance. The dosage of pyridostigmine was also modified, with no adverse effects. During the stay, the patient's condition improved - respiratory disorders resolved, ptosis was significantly withdrawn, and limb muscle tension increased globally. The patient was discharged home in a good general condition, without respiratory difficulties, and in a state of significant neurological improvement.

The second patient, aged 67, also female, with a history of myasthenia gravis as well, was admitted to the Neurology Department due to increased fatigue, ptosis, dysphagia, and difficulties in speaking and breathing. In the neurological examination on the day of admission, the patient was conscious, auto- and allopsychically oriented, in logical verbal contact, bilateral ptosis was found, dysartic speech was present, without evident paresis and ataxia in the limbs, but with global muscle fatigue and a decrease in muscular strength after a small effort. During hospitalization, steroid therapy was used, but due to very high blood glucose values (above 300 mg/dl [the patient admits that she is being treated for type 2 diabetes, but she is

not taking insulin regularly]), the dose of the steroid had to be reduced and a two-day cladribine treatment was initiated, with good treatment tolerance. During the stay, an attempt was also made to perform EMG, nerve conduction study and myasthenic test, but during the examination the patient refused to continue it, and therefore the study was discontinued. The patient was discharged home in good general state and improved neurological condition.

DISCUSSION

Myasthenic crisis is often a life-threatening condition that requires immediate treatment. In the case of respiratory disorders, which are often a manifestation of a developing crisis, proper management and supportive treatment are important: ensuring proper respiratory function (if necessary, intubation of the patient and mechanical ventilation), controlling acid-base balance parameters and starting glucocorticoid therapy as soon as possible. Pyridostigmine application should be also considered. In severe cases, IVIG administration or plasma exchange can be also performer. Cladribine is not a standard way of dealing with myasthenic crisis. Some of the papers suggests a highly cautious attitude when introducing cladribine in patients suffering from myasthenia gravis. However, as this case shows, in some patients this kind of approach can be beneficial. [5-8]

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