FEATURES OF THE COURSE OF MYASTHENIC CRISIS IN LATE DEBUT DISEASE

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Introduction. Myasthenia gravis (MG) is autoimmune neuromuscular disorder, clinically characterized by pathological weakness and skeletal muscle fatigue, associated with damage to acetylcholine receptors of the postsynaptic membrane of the striated muscles by specific complement-fixing antibodies. If earlier myasthenia gravis was considered a rare disease with a prevalence of 0.5-5 cases per 100,000 people in the 60s of the twentieth century, now the disease is considered a fairly frequent phenomenon with a prevalence of 4.8-5.0 to 17.5-20 , 3 per 100,000 people with an annual increase in the number of patients by 5-10% in all age groups. [1,2]

Purpose: to analyze the course of myasthenic crisis with late onset of myasthenia gravis.

Materials and methods. During the observation, 13 cases of myasthenic crisis were investigated. All patients with crisis were divided into 2 groups. The first group included patients at a young age of up to 35 years - 4 people. The second group included elderly patients (60 years and older) - 9 people.

All patients underwent a neurological examination, multispiral computed tomography of the mediastinal organs

Results. In the period from 2013 to 2015, 13 patients with a crisis were registered. All patients had a generalized form of myasthenia gravis with severe respiratory and bulbar disorders.

Among the observed patients, persons aged 60–80 years prevailed - 9 people and only 4 people, in 1/3 of cases - myasthenic crisis developed at the age of 19–35

years. There were no patients with myasthenic crisis aged 35 to 60 years. Among the patients, women predominated - 11 people.

The duration of the disease in all patients was 5.77 ± 1.84 years, among young patients it was 9 ± 5.1 years; in patients aged 60 years and older - 4.33 ± 1.46 years.

In 2 people, a myasthenic crisis was observed, which was manifested by pronounced bulbar disorders, increasing respiratory failure, rapidly increasing weakness of skeletal muscles, which entailed hospitalization of patients in the intensive care unit and their transfer to mechanical ventilation.

Cholinergic crisis was observed in 2 people, which was associated with an overdose of anticholinesterase drugs. Outwardly, the cholinergic crisis resembles myasthenic crisis and is characterized by a deterioration in the condition, generalization of muscle weakness, the appearance or aggravation of bulbar disorders, and respiratory disorders. The muscles of the face, the chewing muscles, and the muscles of the shoulder girdle are always severely affected. Unlike myasthenic crisis, in this case, there are distinct autonomic disorders characteristic of muscarinic and nicotine effects: increased salivation and separation of bronchial mucus, sweating, violent intestinal peristalsis, diffuse abdominal pain, often profuse loose stools, frequent urination, repeated vomiting may occur ... In addition, constriction of the pupils, bradycardia, hypotension, widespread fasciculations in the muscles, sometimes cramps, and a feeling of spasm in the throat are characteristic. Loss of consciousness, involuntary urination may develop.

The remaining 9 people experienced a mixed crisis, which is most common in clinical practice. It combines all the clinical features of myasthenic and cholinergic crises described above. This is the most severe variant of vital disorders in patients with myasthenia gravis. The peculiarity of the distribution of movement disorders during the mixed crisis is that with complete functional incompetence of the cranio-bulbar and respiratory muscles, the strength of the muscles of the arms and legs can be reduced slightly. In addition, attention is drawn to the unequal reversibility of movement disorders in various muscle groups while taking anticholinesterase drugs. It is this type of crisis that most often leads to death in patients with myasthenia

gravis.

According to the results of the study, the average age of onset of myasthenia gravis in the age group 60–80 years was 64.5 ± 7.7 years, the average duration of the disease was 3.7 ± 0.8 years, in one patient the disease debuted with myasthenic crisis at the age of 60 years.

A provoking factor in the development of a crisis in six out of thirteen people was an acute respiratory viral disease, five out of six people took anticholinesterase drugs uncontrollably; in three, exacerbation was provoked by surgical treatment (caesarean section, hip arthroplasty), in three people - psycho-emotional stressful situations, while two independently increased the dose of anticholinesterase drugs, in one patient, physical activity caused decompensation.

Acute respiratory viral diseases and stress conditions were common for both age groups. However, in the group of elderly patients, the predominant factor was an uncontrolled independent increase in the frequency of administration and dosage of anticholinesterase drugs. Thus, seven out of nine elderly patients independently increased the dose of anticholinesterase drugs (pyridostigmine) to 6–8 tablets per day.

In patients with late onset of myasthenia gravis, there was a mixed crisis with a predominance of the cholinergic component, which manifested itself in the form of increased salivation, diarrhea, abdominal pain, sweating, convulsions, fascicular twitching. The cholinergic nature of the crisis was a natural consequence of a previous overdose of anticholinergic drugs. Young patients developed a myasthenic crisis with a characteristic clinical picture.

All patients with a developed crisis were admitted to the intensive care unit, where they underwent the following set of therapeutic measures: breathing with mechanical ventilation; placement of a nasogastric tube to ensure swallowing; withdrawal of anticholinesterase drugs; pulse therapy with metipred at a dose of 1000 mg intravenous drip once a day for 5 days, followed by a switch to oral glucocorticosteroid therapy with the calculation of an individual dose of the drug; plasmapheresis 3-5 procedures.

The period of withdrawal of anticholiesterase drugs in elderly patients was

longer (5 days) than in young patients (3 days), which was associated with the nature of the crisis. It is known that cholinergic and mixed crises, which in our observational study were more common in elderly patients, have a more severe and protracted course compared to myasthenic crisis. Accordingly, the period of respiratory support was also longer in patients of the older age group.

Conclusion. Thus, crisis is a fairly common complication of myasthenia gravis in patients with late onset of the disease. The crisis course of myasthenia gravis was more common in elderly patients than in young patients. At the same time, a feature of the course of myasthenic crisis in the elderly is the predominance of the cholinergic component, which requires a longer withdrawal of anticholinesterase drugs. Independent uncontrolled increase in daily dosages of anticholinesterase drugs by patients is an important provoking factor in the development of crises in patients with late onset of the disease, which requires more careful dynamic monitoring of the patient's condition, additional monitoring of the clinical manifestations of myasthenia gravis and extreme caution when increasing the dose of anticholinesterase drugs.[3]

LIST OF LITERATURE:

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