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Clinical features of different forms of myasthenia depending on structural changes in thymus

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Annotation: This article presents the features of the clinical picture of myasthenia gravis on the background of the presence or absence of structural changes in the thymus, based on the use of classical neurological examination, pharmacological tests. Assessment of the severity of the patient's condition was performed on the MGFA scale (2001) using neurological data as well as the quantitative QMGS test [5].

Keywords: myasthenia gravis, thymus, muscle weakness, MGFA scale, QMGS test.

Myasthenia remains an urgent medical and social problem due to the predominant lesion of the motor system and rapid disability of the patient. Despite current achievements in practical medicine and science in the study of etiopathogenesis, clinical features and approaches to myasthenia treatment, many issues remain unresolved [1, 2, 6].

According to various authors, the number of myasthenia cases increases annually and reaches at present 14.2 - 20.3 cases per 100,000 population. The disease is observed predominantly in women (3:1), and the start of it is at a young age (90% - up to 40 years) [3, 5]. In women, the debut of the disease occurs earlier and its course is more severe than that of men [1, 4, 6].

Development of myasthenia is traditionally associated with thymus pathology. According to the American National Institute of Neurological Disorders and Stroke, 10% of patients with myasthenia are thymoma and 70% have thymic hyperplasia. In this case, thymectomy, as one of the most common methods of treating myasthenia, does not always lead to sustained remission [2, 3, 5].

Materials and methods. 102 people aged 18 to 69 with myasthenia have been comprehensively surveyed and their data have been analyzed and studied. Diagnosis of myasthenia was established according to the tenth edition of the International Classification of Diseases ICD-10 (WHO, 1992) - G 70.2 (Congenital and acquired myasthenia). All patients independently signed an informed consent to participate in a scientific clinical research.

The exclusion criteria were: the age of patients younger than 18; refusal of the patient to participate in the scientific clinical research at any stage and reluctance to sign an informed consent to participate in it; available decompensated cardiovascular pathology and respiratory diseases that were not a result of muscle weakness in myasthenia; Lambert-Eaton syndrome and other myasthenic syndromes against the background of somatic and endocrine pathology.

General examination complex was conducted according to a single scheme. Clinical peculiarities of disorders in patients with myasthenia were determined by generalizing the data of the disease anamnesis and their life, patient complaints (according to the card of the inpatient patient); data of somatic and thorough neurological examination. Assessment of the severity of the patient's condition was performed on the MGFA scale (2001) using neurological data as well as the quantitative QMGS test [4]. For diagnostic purposes, a classical pharmacological test with subcutaneous administration of 2 ml 0,05% solution of neostigmine was conducted. 30 minutes later muscular strength of the patient was evaluated, limited to the gradual assessment of the test. Structural condition of thymus in patients with myasthenia was evaluated using the apparatus of the spiral computer tomography (SCT) SeleCT SP (Marconi).

Taking into account the purpose of the study and the data of SCT of chest organs, patients with myasthenia were divided into three groups. The first group consisted of 35 patients with myasthenia without structural thymus disorders (group M), the second group consisted of 37 patients with myasthenia against the background of thymic

hyperplasia (group MH), the third group included 30 patients with myasthenia against the background of thymomas (MT group).

Analysis of clinical and anamnestic data has shown that in patients with myasthenia without structural thymus disorders the average age of the debut disease was in women - 31.7 ± 8.3 years, in men - 32.1 ± 9.4 and manifested by ocular disorders in 20% patients, in those with bulbar disorders - 11.4%, generalized muscle weakness of 68.6%.

In patients with myasthenia against the background of thymic hyperplasia the disease manifested earlier in women than in men (24.4 ± 7.2 and 33.4 ± 12.1 years, respectively) and was characterized by the appearance of local forms: ocular - 37.8%, pharyngeal -facial - 46%; and only in 16.2% of patients the disease debut manifested as a generalized muscle weakness. Also, patients in this group were characterized by rapid progression of muscular symptoms (increased symptoms and the transition of local forms to generalized in 3-6 months).

In patients with myasthenia against the background of thymomas, there was a late disease debut regardless of sex (men - 48,7 ± 8,3, women 52,9 ± 9,7). In 70% of patients in this group myasthenia manifested by generalized muscle weakness. The bulbar form was diagnosed at the beginning of the disease in 20% of the patients, the ocular form - in 10%. Subsequently, the disease was slightly unstable and difficult. This was confirmed by the fact that only in this group 17% of patients had urgent states in the form of myasthenic crisis according to anamnesis of the disease given in the medical documentation provided by the patients under study.

Generalized neurological status data showed that oculomotor disorders were found in M group most often in the form of nystagmus - 40.0% of patients, ptosis was observed in 11.4% of cases, and diplopia and obliquity - in 14.3% of patients. In MH group, oculomotor disorder manifested with the following frequency: ptosis in 18 (9.0%) patients, diplopia, nystagmus, and strabismus in 45 (9.0%). In MT group, only 10% of patients had ptosis, but the highest frequency of nystagmus (83.3%), diplopia and strabismus (50.0%) was noted.

Dysarthria and dysphagia, as a result of the oropharyngeal muscle weakness, in MT group were found in 60% of patients, in MH group - 54%, and in M group- in 40% of patients.

In MH and MT groups, respiratory failure was recorded in (5.6% and 36.6% of patients respectively) but was not found in M group. Unlike patients in MH group, 13.3% of patients in group MT prior to being included into the study, needed artificial ventilation to compensate for respiratory distress (after that they were included in the study). This phenomenon was not observed in other groups.

In all patients we observed a reduced muscle tone and decreased muscle strength in the limbs. Thus, in patients from M and MT groups upper limb muscle lesions (54.3% and 53.4% of patients, respectively) predominated, unlike in MH group patients, where lower extremities were more often affected (54.0% of cases). However, maximum reduction in muscle strength in general was observed in MT group patients. Thus, in 16.6% of patients in this group muscle strength was 1.0-1.5 points, 50.0% - 2.0-2.5 points and 33.4% - it corresponded to 3.0-3.5 points. In MH group, 16.2% of the patients experienced a decrease in muscle strength to 2.0-2.5 points, in 70.2% - 3.0-3.5 points and in 13.6% of patients to- 4.0 - 4.5 points. The slightest changes in muscle strength were observed in M patients group, whose indicators were not less than 3.0 points (3.0-3.5 points - 71.4% of the cases, 4.0-4.5 points - 28.6 % of cases).

The fact that 87.0% of patients in these three groups had hyperreflexia maybe explained by theories of autoimmune myasthenia gravis and similar pathophysiological mechanisms of other diseases of the nervous system with the autoimmune mechanism’s development. According to the data obtained by MGFA classification, the severity of the patients with myasthenia status without structural thymus disorders was in line with grade IIA in 21 patients (60.0%), IIB grade -9 patients (25.7%), grade IIIB - in 5 patients (14.3 %)

In patients with myasthenia against the background of thymic hyperplasia, the severity of the patients' status corresponded to grade IIA in 5 patients (13.5%), grade IIB - 12 patients (32.4%), grade IIIA - 13 patients (35.2%) IIIB grade - in 5 patients

(13.5%), in IVA grade - in 2 patients (5.4%). The most severe course of the disease according to MGFA classification was observed in patients with myasthenia against the background of thymomas.

The severity of the patients' condition corresponded to grade IIIA in 5 patients (16.7%), IIIB grade - 14 patients (46.7%), IVA grade - 7 patients (23.3%), IVB grade - in 4 patients (13.3%)

A quantitative scale of myasthenia clinical manifestations (QMGS) was performed to objectivize the severity of the patients' condition with different forms of myasthenia. The average severity of the disease in patients with myasthenia without structural thymus disorders was 15.4 ± 4.5 points, in patients with myasthenia against the background of thymic hyperplasia 19.4 ± 4.5 points, and in patients with myasthenia against the background of thymomas, maximum index was 29.8 ± 4.36 points, which is 1.9 times higher than that of M group and 1.5 times higher than that of MH group. The data obtained quantitatively confirm the most severe course of myasthenic symptoms in patients with myasthenia against the background of thymomas, and the lighest - in patients with myasthenia without structural thymus disorders.

When conducting a pharmacological test with neostegmine in patients with myasthenia without structural thymus disorders, a sharp positive reaction was obtained in the form of complete subjective and objective disappearance of muscle weakness in 60% of patients, a positive reaction in the form of complete subjective and objective disappearance of muscle weakness symptoms in a state of rest and its reduction to 4 points with standard physical activity in 40% of patients. In patients with myasthenia against the background of thymic hyperplasia, a sharp positive reaction was observed in 64.9% of patients, positive in 29.7%, and slightly positive in the form of incomplete elimination of muscle weakness in the state of rest and reduction of strength to 3 points in the middle of the standard load in 5.4% of patients.

In patients with myasthenia against the background of thymomas a sharp positive reaction was noted in 50%, positive - in 20%, and in 30% there was a slightly positive reaction.

7. References

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