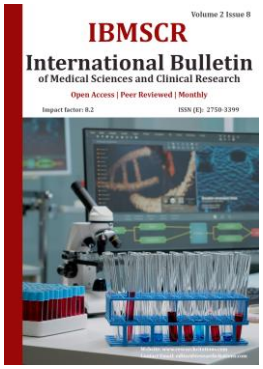

GUILLAIN-BARRE SYNDROME WITH BULBAR DISORDERS

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Summary: Bulbar disorders in the onset of Guillain-Barre syndrome (GBS) are rare and create diagnostic difficulties. A 50-55-year-old patient was hospitalized due to the development of dysarthria, ataxia, nasolalia, paresthesia and increased drowsiness, which reached the greatest severity after a day. An atypical variant of GBS was diagnosed. According to its manifestations, it corresponded to "acute bulbar paralysis plus". The peculiarity was the absence of an increase in protein in CSF by the end of the 2nd week of the disease and the absence of antibodies to gangliosides in the blood. Treatment included plasmapheresis. After a month, all clinical symptoms regressed. GBS in atypical cases can manifest with bulbar disorders and ataxia, which must be taken into account in the differential diagnosis with a stem stroke.

Key words: Guillain-Barre syndrome, acute bulbar palsy–plus; Bickerstaff stem encephalitis.

Guillain-Barre syndrome (GBS) is traditionally attributed to self-limiting diseases characterized, in most cases, by spontaneous recovery. The prevalence is 1-2 cases per 100,000 population per year. Despite a good prognosis for recovery in general, some patients have some residual phenomena that prevent them from returning to normal professional activity. It is known that the more severe the disease is in the phase of increasing neurological symptoms, the longer the recovery period lasts. It is generally believed that demyelinating forms of GBS have the best prognosis, whereas axonal forms, especially acute motor-sensory axonal neuropathy, are characterized by a longer and sometimes incomplete recovery.

Bulbar disorders in Guillain Barre syndrome (GBS) are noted in about a third of patients [1], however, at the very beginning of this disease they occur rarely, mainly with such atypical variants as pharyngocervico-brachial form, M. Fischer syndrome and Bickerstaff stem encephalitis [2-5]. In 2016, J.K. Kim et al. proposed to single out an independent atypical variant of GBS, designated by them as "acute bulbar paralysis plus" (OBPP) [6]. Its distinctive feature is the acute development in the onset of the disease of pronounced bulbar syndrome in the absence of motor deficiency in the extremities. This form of GBS always creates diagnostic difficulties, since it requires differentiation with another pathology, the early manifestations of which may be acute/subacute bulbar disorders (stem stroke, encephalitis, myasthenia gravis, botulism, etc.). Under our supervision there was a patient who was hospitalized in the Russian Academy of Medical Sciences with a suspected stroke. 2 weeks before hospitalization, the patient suffered an acute respiratory infection complicated by bronchitis. She was treated on an outpatient basis at the place of residence (took antibiotics). On the eve of hospitalization, she felt that she was staggering when walking. I went to rest at home, and when I woke up after a long day's sleep, I noticed a slight nasal voice. The next morning, the nasal twang increased, the speech became slurred, and when drinking water

began to pour out through the nose. There was general weakness and increased drowsiness, coordination of movements in the left hand worsened. The patient was taken to the hospital with a suspected stroke. There were no foci of acute ischemia in the brain according to multispiral microcomputer tomography (MSCT) and magnetic resonance imaging (MRI), however, based on clinical data, the patient was diagnosed with lacunar stem ischemic stroke. In substantiating the diagnosis, the sufficiently acute development of bulbar symptoms and ataxia were taken into account, as well as the fact that the sensitivity of MRI in the diagnosis of stem lacunar strokes is not one hundred percent. The patient was hospitalized, and a day later transferred to the department of emergency neurology. During the examination, the patient complained of slurred speech, nasal voice, difficulty drinking ("water is poured through the nose"), instability when standing, general weakness, increased drowsiness. Her condition was regarded as severe due to the presence of bulbar disorders associated with a potential threat to life. Objectively: body temperature, heart rate and respiration, blood pressure — within normal values. The patient is oriented in place, time and her own personality, adequately answers questions, although she concentrates her attention with difficulty, quickly depletes, periodically falls asleep. There are no meningeal signs. The movement of the eyeballs in full, nystagmus is not detected. Medium-sized pupils, their reactions to light and convergence are preserved. There are no changes from the V–VIII pairs of cranial nerves. There are nasolalia, decreased mobility of the soft palate with phonation, more pronounced on the left, bilateral decrease in pharyngeal and palatal reflexes, dysarthria (speech is slurred, non-flowing, with pauses, with distortion of articulation of both consonants and vowels, approaching "chanted" speech). Swallowing solid food is not disturbed, tongue movements are preserved, the tongue is located along the middle line when sticking out. The finger test on the left is performed with a miss and intention, the finger test on the right and the heel-knee test on both sides are performed correctly. The volume of active movements in all joints is complete. Resistance in the study of all major muscle groups of the arms and legs is sufficient, there are no paresis in the extremities. Tendon reflexes from the hands of the usual amplitude, symmetrical; knee and Achilles reflexes are low. There are no pathological hand and foot signs. On the same day in the evening, the patient felt discomfort in the neck – according to the patient, "the neck seemed to swell", and tingling in the left, and a few hours later — in the right hand ("as if the hands were lying down"), although no decrease in superficial and deep sensitivity was detected during examination. Paresthesia persisted for the next three days. Laboratory and instrumental examination was carried out in accordance with the regulations for the provision of medical care to patients with acute disorders of cerebral circulation. Along with this, other possible causes of bulbar syndrome were excluded, since the diagnosis of ischemic stroke was doubtful due to the increase in nasal voice and dysarthria noted throughout the day; the absence of dysphagia and paresis of the muscles of the tongue in the presence of unilateral paresis of the soft palate; the absence of stroke risk factors. The results of general clinical and biochemical blood tests (cellular composition, ESR, hemoglobin, glucose, total protein and protein fractions, C-reactive protein, transaminases, creatinine, lipoprotein fractions, electrolytes, thyroid hormones), indicators of the blood coagulation system, blood gas composition and blood electrolytes were within normal limits. Monitoring of blood pressure and ECG revealed no pathology. There were no changes in the lungs according to radiography. According to the results of duplex scanning, there were no hemodynamically significant violations of blood flow through the extracranial arteries of the brachiocephalic

system. CT angiography revealed no pathology of cerebral vessels. On the 2nd day after the onset of the first symptoms of the disease, a lumbar puncture was performed. The cellular composition and the results of biochemical examination of cerebrospinal fluid (CSF) were not changed. During the first three days, the patient retained pronounced nasal voice, dysarthria, ataxia, paresthesia, general weakness and pronounced drowsiness – not only at night, but also most of the day, she slept, and when she got up and tried to walk, she held on to an assistant to keep her balance. Heart rate and respiration, swallowing, muscle strength in the extremities remained intact. From the end of the third day of the disease, rapid improvement began in the form of a decrease in the severity of ataxia, nasal voice and general weakness / drowsiness. After 5 days, the mobility of the soft palate was restored with phonation, instability in the Romberg test and mimopadence disappeared when performing the finger-nasal test on the left. However, on the 6th day of the disease, during a neurological examination, the disappearance of knee and achilles reflexes was noted. During this period, the examination of the patient continued to identify/exclude antiphospholipid syndrome and vasculitis as possible causes of "young stroke", as well as myasthenia gravis, neuroinfections, porphyria as alternative diagnoses to stroke. The results of the electroneuromyographic (ENMG) examination of the nerves of the upper extremities, performed on the 7th day of the disease, were within normal values. The standard decrement test with tetanization, conducted on the same day, did not reveal a decrement in the amplitude of the M-response of the circular muscle of the right eye, the right deltoid muscle and the muscles of the right hypotenar during electrical stimulation of the facial, axillary and ulnar nerves, respectively, with a frequency of 3 pulses per second, as well as significant changes in the amplitude of M-responses during periods of postactivation relief and postactivation exhaustion, which indicated the absence of neuromuscular transmission disorders. On the 9th day of the disease, against the background of complete restoration of balance and coordination of movements and almost complete regression of the paresis of the soft palate, the patient experienced severe pain in the temporal regions and the lower half of the face. Following this, a few hours later, weakness of the facial muscles of the left half of the face developed, and on the next (10th) day - and its right half. As the patient recalled later, her "face turned to stone, her lips hung, her eyelids did not close, and tears flowed from her eyes." A neurological examination revealed bilateral paralysis of facial muscles. The strength in the extremities, including in the fibular muscles and the muscles extending the foot and toes, remained intact. Tendon reflexes were triggered from the hands, although their amplitude was reduced, and knee and Achilles reflexes were absent. There were no pathological signs. There were no violations of coordination and sensitivity. Lasega's symptom was negative. On this day, the diagnosis of stroke was removed and an atypical variant of GBS syndrome was diagnosed. Its clinical characteristics corresponded to the signs of BPP described in 2016 by J.K. Kim and co-authors: bulbar disorders, ataxia and paresthesia in the hands at the onset of the disease; suppression of tendon reflexes in the absence of paresis of the muscles of the extremities; delayed onset of facial diplegia; an increase in symptoms for less than four weeks, followed by its regression [6]. The results of ENMG on the 13th day of the disease were as follows: median (motor and sensory portions), tibial, calf nerves are normal, M-responses from the facial and ulnar nerves are slightly reduced, significantly from the peroneal nerves, which may be with axonal neuropathy of these nerves. Results of repeated lumbar puncture on the 13th day of the disease: cytosis of 19 cells (18 neutrophils, 1 lymphocyte) in 1 ml; protein 0, 26 g/l; glucose

3.60 g/l; chlorides 118.0 mmol/l. The detection of mild pleocytosis and the absence of protein-cell dissociation in CSF were not a reason to revise the diagnosis of GBS. The final diagnosis was formulated as follows: Guillain-Barre syndrome: acute motor-sensory axonal neuropathy with bulbar disorders, ataxia and hypersomnia at the onset of the disease, facial diplegia, mild sensory disorders. Immunological analysis (line-blot), carried out on the 17th day of the disease, did not reveal antibodies of the JgG/M classes to the gangliosides GM1, GM2, GM3, GM4, GD1a, GD1b, GD2, GD3, GT1a, GT1b, GQ1b in the blood serum. After the diagnosis of GBS, the patient was treated with plasmapheresis; a total of 3 sessions were conducted every other day. Facial diplegia regressed 3 weeks after the onset of the disease. Upon repeated examination of the patient after 7 months, only mild hypomimia, a slight nasal tinge of the voice and the absence of tendon reflexes from both hands and feet were detected in the neurological status. The results of repeated ENMG 7 months after the onset of the disease did not differ from the indicators obtained on the 13th day of the disease. Discussion Guillain-Barre syndrome, along with the classical form, includes rare clinical variants that can be clearly delineated, reduced, atypical or cross [7, 8]. It is the atypical variants of GBS that present the greatest difficulties for diagnosis, predisposing to errors in the recognition of this pathology and the delayed start of specific therapy [9]. This situation also developed in the described case, when the atypical for GBS onset of the disease with bulbar disorders initially led to an incorrect diagnosis of stroke and delayed initiation of adequate treatment. Atypical variants of GBS are Miller Fisher syndrome, pharyngocervico-brachial form, paraparetic motor form with selective involvement of the legs; facial diplegia or paresis of the diverting nerves in combination with paresthesia; sensory atactic variant; variant with pandisautonomy [1, 4, 5, 7]. To this list in 2016, J.K. Kim and co. proposed to add also the OBPP [6]. The basis for the recognition of OBP as a variant of GBS can be their pathophysiological similarity and the presence of such common clinical signs as inhibition of tendon reflexes, damage along with the bulbar group and other cranial nerves, ataxia, decreased nerve conduction rate according to ENMG, albuminocellular dissociation in CSF, the presence of antibodies to gangliosides in blood serum, monophasic the nature of the course of the disease with the progression of symptoms for no more than 4 weeks [6]. At the same time, according to J.K. Kim et al., the manifestations of OBPP do not meet the diagnostic criteria of other atypical forms of GBS [10], which served as the basis for describing it as an independent rare variant of GBS. According to J.K. Kim et al., the absence of oculomotor disorders distinguishes OCD from M. Fischer syndrome, characterized by weakness of extraocular muscles, ataxia and areflexia [11]. As for the acute development of bulbar disorders, they cannot serve as a differential diagnostic sign, since they can be noted (although rarely) in M syndrome. Fischer [5, 11]. The difference between OBPP and the pharyngocervico-brachial variant of GBS is the absence of paresis of the muscles of the neck flexors and the muscles of the proximal parts of the arms [12]. According to J.K. Kim et al., OBPP differs from the mild manifestations of the classical form of GBS, as well as from its "intersection" with M. Fischer syndrome, in the absence of weakness of the limb muscles [6]. After analyzing the medical histories of 184 patients treated in Korea in 2012-2013 for Guillain-Barre syndrome, J.K. Kim et al. regarded 11 cases as OBPP. The first symptoms in these patients most often (in 6 out of 11 or 55% of cases) were dysarthria and/or nasolalia, as well as instability when walking (in 2 out of 11 or 18% of cases); dizziness, tingling sensation in the extremities, diplopia were less common.

Thus, GBS can manifest in rare atypical cases with bulbar disorders, ataxia and hypersomnia, which must be taken into account in the differential diagnosis of stem stroke in the practice of vascular centers. Knowledge of the clinical features of atypical variants of GBS is determined by the importance of early initiation of its specific therapy necessary to stop the autoimmune process

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