FEATURES OF ELECTROPHYSIOLOGICAL METHODS FOR GUILLAIN–BARRÉ SYNDROME

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Abstract. *Guillain-Barre syndrome recently gained notoriety due to reports of such a diagnosis in former Russian politician Anatoly Chubais, who was admitted to intensive care while on vacation in Italy with "numb legs and arms, a distorted face." Acute autoimmune disease, which is very rare during the pandemic, is widespread, because the main trigger of its development is recent infections (viral and bacterial), including the coronavirus infection caused by SARS-CoV-2.*

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What is Guillain-Barré syndrome?

This is a pathological condition in which a person's immune system begins to attack their own peripheral nerves. Includes several clinical forms. Often this is acute inflammatory demyelinating polyneuropathy - AIDP. This abbreviation reflects the main aspects: the disease begins acutely or subacutely, is inflammatory, but not infectious, affects the myelin sheath of peripheral nerves and causes neurological diseases. The myelin sheath surrounding the peripheral nerves performs many functions, the most important of which is to accelerate the transmission of impulses along the peripheral nerve. Accordingly, clinical signs are associated with the slowing down of nerve impulses. In the typical form of ARDP, primarily the motor nerves are affected, the function of the muscles innervated by them is impaired, motor skills are impaired, and paresis develops. A characteristic feature of Guillain-Barré syndrome is symmetry, that is, both legs are affected, then both arms (paraparesis, tetraparesis). Although there are also casuistic cases with asymmetric opening. In atypical forms, not only the myelin sheath is affected, but also axial cylinders - axons of peripheral nerves, which often leads to a long recovery and recovery of the patient. There are two such forms - acute motor axonal neuropathy (AMAN) and acute motor-sensory axonal neuropathy (ASAN). Another atypical form of Guillain-Barré syndrome is pan dysautonomia, when only autonomic fibers of peripheral nerves innervating internal organs - heart, lungs, intestines, vascular tone and sweating - are affected. Among the atypical forms, the rarest type is Miller-Fisher syndrome, which occurs in 2-3% of cases. This syndrome is characterized by rapid (in a few days) development of oculomotor disorders (ophthalmoplegia), impaired coordination of movements (ataxia) and areflexia.

What is the cause of this acute neurological disease?

The etiology of Guillain-Barré syndrome is unknown. But the triggers and triggers are well-studied. These include various infections - respiratory, gastrointestinal, etc., including the coronavirus infection caused by SARS-CoV-2. The infection disrupts the immune system, which begins to produce antibodies against the myelin of its nerve cells. Triggers for Guillain-Barré syndrome can be other events that activate the immune system, such as trauma, surgery.

International publications describe cases of Guillain-Barré syndrome after vaccination, including some vaccines against coronavirus infection. Therefore, you should be careful, be careful of yourself and your loved ones, and if neurological symptoms appear, you should immediately seek medical help. In most cases, the patient has a history of one or another infectious disease, even very mild (mild cough, diarrhea, short-term fever), 1-2 weeks before the development of neurological symptoms.

How do clinical signs develop?

In the typical form of Guillain-Barré syndrome (acute inflammatory demyelinating polyneuropathy), symptoms begin with discomfort in the lower extremities, then slowly but rapidly spread to the arms over hours or days. For example, the patient complains of numbness and weakness in the legs in the morning, and weakness in the hands in the evening or the next day. If there is no adequate treatment, the cranial nerves that innervate the organs and tissues of the head may be damaged with the development of the disease. The muscles of the skull are involved in the process, the patient's face is distorted, the eyes may not be closed, etc. But the most dangerous option is damage to the 10th pair of cranial nerves with a violation of the respiratory and cardiovascular systems. This condition is called Landry's syndrome and is associated with death mainly in Guillain-Barré syndrome. The danger of Landry's syndrome lies in its rapid development: from involvement of cranial muscles to complete cessation of breathing, less than a day or even hours may pass.

What are the challenges in the process of diagnosing Guillain-Barré syndrome, especially in the outpatient setting, where the patient is usually the first to see?

In fact, in the first stages, when the patient is seen in the clinic by a general practitioner or an emergency paramedic, it is not easy for them to immediately recognize the subtle neurological symptoms of Guillain-Barré syndrome. It is easier to make a diagnosis if there is a typical clinical presentation of the disease - gradual symmetrical damage to the limbs. However, rarely, focal forms appear, when only one arm and / or one leg is weakened. A doctor who sees hemiparesis, including a neurologist, first suspects a stroke. Even here, at the 5th City Clinical Hospital of Minsk, which is the clinical base of the Department of Neurology and Neurosurgery of BelMAPO, such cases occur when patients with hemiparesis and unrecognized Guillain-Barre syndrome are admitted to the vascular department. stroke. But during a stroke, a unilateral lesion becomes unilateral with gradual regression or stabilization (with the exception of a rapidly recurring stroke). And with unrecognized Guillain-Barré syndrome, the asymmetric onset quickly goes to the other side, the clinical picture becomes more detailed, and the diagnosis is no longer in doubt. The patient is transferred to the general neurology department and receives pathogenetic treatment. At the ambulatory stage, diagnostic errors also occur at the beginning of the disease, when emotional disorders, not motor, come to the fore. The patient comes to the polyclinic complaining of back pain in the chest or back area, with radiation to the limbs. That is, it describes radicular pain like radiculitis. As a result, the diagnosis is misdiagnosed based on the pain syndrome: radiculitis, vertebrogenic thoracalgia, myofascial pain syndrome, etc. Addition of motor symptoms - and in Guillain's syndrome - Barre almost always occurs! - will then make you question your initial assumptions and continue your diagnostic search. The most difficult form of recognition is acute pandysautonomia. The patient does not have paresis, pain, emotional disturbances, but he stops keeping pressure, it becomes difficult for him to breathe, bowel movement is disturbed, cardiac arrhythmia appears.

If Guillain-Barre syndrome is suspected, what instrumental and laboratory tests should be performed during the diagnostic search?

Electroneuromyography (ENMG) is the main instrumental method by which we can objectively determine the disturbance of the transmission of impulses along the peripheral nerve, but it shows the slowing down of the transmission of impulses only along the motor and sensory fibers. Unfortunately, sometimes in the first 7 days from the onset of the disease, the device does not recognize disorders and gives the norm. If the autonomic fibers are selectively affected, for example, in the case of pandysautonomia, in ENMG we see a normal picture and this can mislead the clinician in making the correct diagnosis. A characteristic laboratory sign of Guillain-Barré syndrome is an increase in the level of protein in the cerebrospinal fluid with a normal level of leukocytes, which is called protein-cell dissociation. However, this marker appears only at the end of the first week of the disease, sometimes even in 10-14 days. And clinical signs develop within 2-3 days. Therefore, a patient with a typical clinical presentation of Guillain-Barré syndrome is admitted, he undergoes a lumbar puncture, and a normal result, that is, a false negative result, is obtained when analyzing the cerebrospinal fluid. ENMG is also normal. As a result, the doctor, seeing the usual picture of demyelinating polyneuropathy, but not finding instrumental and laboratory confirmation, starts a new diagnostic search. This is a fundamentally wrong approach. My personal opinion: Guillain-Barre syndrome is a clinical diagnosis. If it is confirmed by ENMG - well, if not - Guillain-Barre syndrome cannot be excluded. By repeating the lumbar puncture and repeating the ENMG after a week, you will see the changes characteristic of this disease. How not to make a mistake in making a diagnosis based on clinical neurological symptoms? Especially in the early stages, when is it done by a GP or an emergency paramedic? First of all, it is necessary to determine the nature of paresis: central or peripheral. This is very important for topical diagnosis. Thus, central paresis develops when the brain and spinal cord are damaged, and peripheral paresis develops when the roots, plexuses and peripheral nerves are damaged. In such cases, the symptoms are different. Central paresis is characterized by an increase in muscle tone, the presence of pathological reflexes, and an increase in tendon-periosteal reflexes. In peripheral cases, reflexes are reduced, muscle tone is low, there are no pathological reflexes. Clinically, when we see acute symmetric peripheral paresis, especially of a nature that ascends from the legs to the arms, it is almost always Guillain-Barré syndrome. I repeat, regardless of instrumental and laboratory research. It should also be taken into account that acute pathology of the spinal cord (inflammation of any nature - in the form of stroke, myelitis or injury), the so-called spinal shock is possible in the first few days. And it turns out that the central nervous system is damaged, but when examined clinically, we see peripheral lower paraparesis. At the same time, the spinal symptoms are characterized by a different type of sensory disturbance, dysfunction of the pelvic organs, and the absence of gradual upward paresis. The neurological picture develops immediately from the level of spinal cord injury. Knowledge of these clinical features makes it possible to suspect or at least not rule out Guillain-Barré syndrome with high probability, even with false negative results.

How is Guillain-Barré syndrome treated?

Since the direct cause of the immune disorder is unknown, there is no etiological treatment. But the pathogenesis is very well studied, the autoimmune nature of pathological processes has been proven, so the main methods of treatment are immunosuppression and immunomodulation. The goal is to remove antibodies that destroy myelin. There are two ways. The first is

plasmapheresis, that is, blood purification, washing and removal of pathological antibodies. The second is intravenous administration of immunoglobulins, an expensive but effective method. Intravenous immunoglobulins change the work of the immune system in such a way that it stops the production of pathological antibodies and the disease stops. Myelin gradually begins to recover, and as a result, the symptoms of the disease disappear. It is believed that plasmapheresis should wash 30-50 ml of blood per kg of body weight. The number of sessions is determined by the transfusiologist, but on average it is from 2 to 5 sessions. After each test, tests are carried out in order not to miss complications, because not only pathological autoantibodies, but also proteins and other useful molecules are washed away. If everything is normal, the repeated session is done in 3-4 days. Usually 2 times a week, sometimes 3. Thus, the entire course of plasmapheresis requires 1-2 weeks. When administering immunoglobulins intravenously, we use the standard international regimen: 0.4 g per kg per day for 5 days. The total dose is 2 g per kilogram. The patient's weight is measured, the amount of medicine he needs is calculated, and for 5 days the patient is treated with equal doses in the form of simple drops every day. In addition to pathogenetic treatment, symptomatic therapy is also prescribed. It does not affect the mechanism of the disease, but reduces the symptoms. We prescribe painkillers (if the patient complains of pain), anticholinesterase drugs to improve the conduction of impulses along the nerves, and vascular agents to improve tissue microcirculation and thereby reduce inflammation. B vitamins help regeneration, as well as lipoic acid and others.

What is the prognosis after Guillain-Barre syndrome? How to restore broken functions? Is rehabilitation necessary?

In most cases, the prognosis is favorable; The mortality rate for Guillain-Barré syndrome is approximately 2-3%. The most important thing is the timely appointment of pathogenetic therapy, then even very neurologically severe patients recover well. In addition, clinical recovery was confirmed instrumentally. In follow-up examinations (after six months, one year), we repeat the ENMG and note the improvement of electrophysiological parameters until complete normalization. Recovery speeds vary. Some patients recover completely within a month and do not need rehabilitation. We recommend avoiding respiratory diseases, maintaining a normal work and rest routine, adequate sleep and a balanced diet. But often the recovery of the affected functions is slow, rehabilitation is necessary: physiotherapy, mechanotherapy, acupuncture, repeated physical and symptomatic therapy courses. The patient should always be warned not to worry if, for example, his legs are weak after 2 months. The main thing is that the recovery process has started and it is slowly but developing. In some cases, it can take 2-3 months, six months, a year, and according to some publications, even 2 years. After 2 years, residual impairments are generally irreversible and you have to live with them. Pathogenetic treatment is prescribed once. During the entire period of the pandemic, more than 20 patients with Guillain-Barre syndrome related to COVID-19 were treated in our hospital. In general, in our database there are more than 40 such patients who applied to the department for consultation from Minsk and other institutions of the republic's regions. These are mostly patients over 35-40 years old. According to our observations, the severity of Guillain-Barré syndrome does not correlate with the severity of COVID-19. There are patients with severe coronavirus infection, pneumonia, respiratory support, but in the classic mild version, they suffer from Guillain-Barre syndrome. Conversely, the most severe Guillain-Barré syndrome with a short-term history of discomfort for a week with a slight increase in temperature confirmed by the PCR test of COVID-19, clear recovery and rapid

development of Landry's syndrome after 2-3 days. For resuscitators, I would like to emphasize that if the Guillain-Barré syndrome has a rapidly increasing character, spreads to the cranial nerves, and there is a risk of developing Landry syndrome, it is very important to transfer the patient to the intensive care unit. If the patient has normal blood pressure and breathes well during the examination by the resuscitator, he should be transferred to resuscitation.

In such cases, against the background of apparent well-being, cardiac and respiratory arrest may develop suddenly and immediate connection to mechanical ventilation may be required, and this is problematic when the patient is in a regular ward. And if you do not have time to connect to the ventilator in a few minutes, then a potentially favorable prognosis will be canceled by death. The algorithm is simple: if the cranial nerves are involved and there is a risk of Landry syndrome, the patient should stay in intensive care for 2-3 days. They prescribed pathogenetic treatment, saw stabilization - again transferred him to the somatic department.

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