NEUROMYELITIS SPECTRUM DISORDERS

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Abstract: *neuromyelitis spectrum disorders are actual medical and social problem, the nosologies of this group are the second most common cause of disability in young age. This article discusses Neuromyelitis Optica. It has a progressive course, can quickly end in death. For this reason an extremely important question is the early diagnosis establishment and the administration of treatment. To confirm the diagnosis, it is necessary to use a whole range of laboratory and instrumental methods of research. The article presents modern diagnostic criteria and options for adequate therapy.*
Keywords: *neuromyelitis spectrum disorders, neuromyelitis optica, multiple sclerosis, differential diagnosis.*

ЗАБОЛЕВАНИЯ ГРУППЫ ОПТИКОМИЕЛИТА

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Аннотация: *заболевания группы оптикомиелита являются актуальной медицинской и социальной проблемой, нозологии этой группы являются второй по частоте причиной инвалидности в молодом возрасте. В этой статье обсуждается оптический нейромиелит. Он имеет прогрессирующее течение, может быстро приводить к летальному исходу. По этой причине чрезвычайно важным вопросом является разработка способов ранней диагностики и лечения. Чтобы подтвердить диагноз, необходимо использовать целый ряд лабораторных и инструментальных методов исследования. В статье представлены современные диагностические критерии и варианты адекватной терапии.*
Ключевые слова: *заболевания группы оптикомиелита, оптический нейромиелит, рассеянный склероз, дифференциальная диагностика.*

Neuromyelitis optica is an inflammatory idiopathic disease of the central nervous system, the leading manifestations of which are damage to the optic nerves, the spinal cord and extensive transverse myelitis at the level of the thoracic or cervical spinal cord, with minimal brain damage. [1]

Epidemiology. Neuromyelitis Optica is a rare pathology, the incidence and prevalence of which has not yet been studied. According to studies in European countries, the incidence of Neuromyelitis Optica is about 2% of all demyelinating diseases. The Mayo Clinic (USA) studied materials from 71 patients. It turned out that with the remitting form the ratio of men and women was 1: 4, and with monophasic 1: 1. The age of debut of the disease was in the range of 40-50 years, which is 10 years later than the classic debut of multiple sclerosis. [2]

Etiology. Previously Neuromyelitis Optica was classified as a malignant variant of multiple sclerosis. But recently scientists have found that the pathogenesis of multiple sclerosis and opticomielitis has significant differences. In Neuromyelitis Optica there is a synthesis of the antibody NMO-IgG to the membrane protein aquaporin-4, which acts as a water channel. In large quantities it contains the membranes of astrocytes, the gray matter of the spinal cord and paraventricular areas. These antibodies can only be formed at the periphery. But, nevertheless, their concentration in the cerebrospinal fluid is 500 times greater than the concentration in plasma, which indicates the ability of NMO-IgG to penetrate the BBB. In addition, these antibodies were not detected in the blood of patients with MS and other autoimmune diseases. This allows to allocate Neuromyelitis Optica as an independent nosological form. In addition to NMO-IgG, this disease also produces antibodies that damage myelin proteins, oligodendrocyte glycoproteins (anti-MOG antibodies). This causes oligodendrocyte damage and axonal demyelination. This autoimmune process leads to degenerative disorders in one or two optic nerves and in at least three adjacent segments of the spinal cord. [3]

Clinical picture. The clinical picture of Neuromyelitis Optica is characterized by optic neuritis and myelitis. In 80% of cases symptoms of optic neuritis are the first to occur. Spinal cord lesions usually occur after several months or years.

Damage to the optic nerve is severe. There are both single and bilateral lesions. Optical neuritis is characterized by a sharp decrease in visual acuity, which may be preceded by an attack of misting for several hours. The cause of visual impairment is most often the central (in 90% of cases) and paracentral slopes. They are accompanied by changes in color vision, manifested by loss of tone and color, the predominance of gray color and the narrowing of the visual fields to red and green. On the ophthalmoscopic picture, the optic nerve discs are not changed or their slight blurring and puffiness are visible. With repeated exacerbations there is a blanching of the discs and their atrophy. [4]

Monophasic and recurrent forms of this disease are distinguished. Demyelination of the optic nerve causes blindness: as a rule, in one eye in 50% of patients with recurrent and in 25% of patients with monophasic form of the disease. The monophasic form is represented by myelitis and optic neuritis, after which the attacks do not recur (symptoms may appear at the same time, but with a difference of no more than 30 days). In the recurrent form of OM between the first attacks, there may be a large gap of several months or not. But later the course of the disease becomes constantly recurrent in nature.

Myelitis often occurs in severe form, develops acutely (within a few hours or days). It is characterized by symmetrical gross violations of the motor sphere, sensory abnormalities and dysfunction of sphincters. For the majority of patients (about 80%), incomplete recovery of function after recurrence of the disease is typical. In case of recurrent myelitis, para-or tetrapraresis, paroxysmal muscle spasms, radicular pain, and Lermitte's symptom are considered classic symptoms. Due to focal myelitis, motor patients are observed (flaccid and spastic paresis, discoordination, ataxia), sensory impairments below the level of the lesion, impaired defecation and urination, and vegetative disturbances. Permanent monoplegia or paraplegia is observed in 50% of patients with recurrent and in 30% of patients with monophasic disease. The most severe manifestation is respiratory failure, occurring in a third of patients and leading to a fatal outcome in 93% of patients suffering from this form of the disease. [2]

Diagnostics. Neuromyelitis Optica is characterized by a clinical combination of optic neuritis and myelitis, but it is impossible to judge the presence or absence of opticomielitis based on clinical data.

The most accurate diagnostic method is MRI of the spinal cord. The majority of patients on an MRI performed during the period of exacerbation of myeloma, revealed a large focus of lesion of the spinal cord, spreading over more than 3 segments of the spinal cord. In the period of exacerbation, the substance of the spinal cord is swollen, the lesion can over-absorb the contrast for a long time. [4]

It is highly likely that foci of the brain stem and hypothalamus may be considered as diagnostic signs; cerebral foci are most often found in areas of increased immunological sensitivity to aquaporin-4.

Diagnostic criteria are used to confirm the diagnosis of Neuromyelitis Optica (by: D.H. Miller et al., 2008)

Big criteria (presence of all criteria is necessary, they can be revealed at different times): • optic neuritis with lesion of one or two eyes;• transverse myelitis, clinically complete or incomplete, but associated during an exacerbation with a radiologically confirmed lesion of the spinal cord that extends into three spinal segments on T2-weighted MRI images and is hypo-intensive on T1-weighted images; • lack of evidence for SLE, sarcoidosis, vasculitis, Sjogren syndrome or other diseases.

Small criteria (requires at least one criterion): • non-specific changes in T2 mode that do not meet the Barcoff criteria given in the McDonald criteria; • foci in the dorsal regions of the medulla oblongata, combined with foci in the spinal cord or isolated; • foci in the hypothalamus and / or brainstem; • "linear" foci located periventricular or in the corpus callosum, but not ovoid and not spreading into the parenchyma of the cerebral hemispheres in the shape of Dawson's fingers; • positive serum or cerebrospinal fluid test for MNO-IgG / antibodies to aquoporin-4. [5]

Treatment. For the treatment of attacks of myelitis and optic neuritis, large doses of glucocorticosteroids are administered (1000 mg per day, intravenous drip, for 5 days); Supplemental therapy with prednisone at a dose of 1 mg / kg per day is additionally recommended as an initial immunosuppressive therapy aimed at preventing the recurrence of the disease. However, treatment of myelitis with glucocorticosteroids often does not give the desired result, and in some cases it can provoke worsening of the condition. [3]

The effectiveness of preventive immunomodulatory therapy based on MS (interferons beta, Glatira mera acetate) has not been formally studied in patients with OM. There is conflicting information about the effectiveness of interferon beta-1b, and about its possible negative effects in relation to the increase. Therefore, for long-term treatment of OM, it is recommended not immunomodulating, but immunosuppressive therapy. Most specialists of choice therapy consider a combination of orally administered prednisone and azathioprine, tested in patients with OM in the late 90s of the last century. Azathioprine is prescribed in a daily dose of 2.5–3 mg / kg, prednisone - in a dose of 1 mg / kg per day. Over time, the dose of prednisolone is gradually reduced to the minimum supporting dose, or even canceled altogether, leaving only mono-therapy with azathioprine. [1]

In 2014, a clinical case of effective use of methotrexate at a dose of 20 mg IV drip together with 20 mg of prednisolone was described. Therapy was carried out for a year with a three-month break. A year later, the EDDS score dropped from 9.0 to 2.5. [3]

Conclusions:

1) as a result of the analysis of scientific literature, it was revealed that the main difficulty in the diagnosis of Neuromyelitis Optica is related to the similarity of its clinical manifestations with such neurological pathologies as multiple sclerosis, recurrent transverse myelitis, recurrent retrobulbar neuritis, systemic lupus erythematosus, sarcoidosis

2) in connection with this diagnostics of Neuromyelitis Optica should be comprehensive and based on the clinical picture, laboratory and instrumental data of the study

3) currently diagnostic criteria - D.H. Miller and co-authors, 2008.

4) the majority of specialists recognize the combination of prednisolone and azathioprine as the grugs of choice, but there are cases of positive dynamics in the treatment with methotrexate.

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