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.
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 tetraparesis, paroxysmal muscle spasms, radicular pain, and Lermite'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 optocmielitis 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 aquaporin-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 drugs of choice, but there are cases of positive dynamics in the treatment with methotrexate.

References / Список литературы


