Neuromyelitis Optica

Neuromyelitis Optica or **Morbus Devic** is a rare idiopathic demyelinating disease affecting the CNS. Women suffer from the disease more often. Most cases of this disease can be found in Asia, where it has a worse course and prognosis.

Pathogenesis

In *neuromyelitis optica* (hereinafter referred to as NMO), pathological and highly specific antibodies to aquaporin 4 occur in the patient's body. Aquaporin 4 is a transmembrane protein on CNS support cells. After the antibodies bind to this channel, a complement cascade is likely to be triggered, leading to an inflammatory response and ultimately cell destruction. However, the exact mechanism of this disease is unknown, as it does not explain NMO in patients with negative antibodies to aquaporin 4.

Clinical picture

The course of NMO is characterized by recurrent attacks of unilateral or bilateral inflammation of the n. opticus, which are often combined with a transverse inflammation of the spinal cord (myelitis) exceeding at least three spinal segments. There is a long latent period between attacks. After repeated attacks of the disease, a neurological deficit often remains.

Symptoms

Eye symptomatology

- Swelling of the nerve;
- Pain behind the eye;
- Loss of color vision;
- Visual field failures;
- Complete blindness.

Spinal symptomatology

- Muscle weakness;
- Loss of sensation;
- Dysesthesia;
- Loss of mobility – paraplegia to quadriplegia;
- Impaired sphincter function – urinary incontinence;
- Impaired sexual function;
- Constipation;
- Radicular pain;
- Lhermitt's symptoms;
- Neurogenic respiratory failure due to cervical spinal cord involvement is the most common cause of death in patients with NMO.

Prognosis

The prognosis is not favorable - 50% of patients lose their vision in at least one eye in the first five years of the course of the disease or are unable to walk without support.

Diagnostics

- MRI of the spinal cord to confirm acute myelitis, on the MRI of the brain there are nonspecific changes – most often abnormalities in the hypothalamus and brain stem;
- Examination of antibodies in the liquor (in particular to exclude intrathecal production of IgG, which is typical for multiple sclerosis);
- Serum testing for antibodies to aquaporin 4 – however, the negative finding may not be excretory;

Differentially diagnostically, it is difficult to distinguish NMO from multiple sclerosis. For this reason, NMOs have long been considered a subtype of it.

Therapy

In acute attacks, patients are given high doses of corticosteroids, in a more severe course, plasmapheresis is indicated. In the resting period, immunosuppressants are used. The first choice drugs are azathioprine and rituximab.

Links

Related articles

- Multiple sclerosis
- Multiple sclerosis/PGS
- Causes of sudden blindness
- Lhermitt's symptom

Literature used

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