

## CASE STUDY

**DEVIC'S OPTICOMYELITIS: A CASE REPORT FROM THE AUTHORS' CLINICAL PRACTICE**

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The aim was to analyze the contemporary scientific literature on Devic's opticomyelitis and to present a case report from our clinical practice.

Based on the patient's complaints, case history and features of clinical course, objective neurological status, clinical laboratory and additional examination methods, characteristic MR-patterns, consultations of related specialists and differential diagnostics, we made the clinical diagnosis according to ICD-10: G36.0 Devic's opticomyelitis, exacerbation, with a sustained bilateral lesion of the optic nerves in the form of retrobulbar neuritis with the development of partial atrophy of the optic nerves in both eyes, spinal cord lesions with common cystic, cicatricial and atrophic alterations at C1-Th8 level with moderate lower paraparesis, expressed by sensory ataxia, sensory disturbances by the descending conductive type from Th10, impaired function of pelvic organs by the type of acute urinary retention, asthenic and neurotic syndrome.

Widespread cases of demyelinating pathology in medical practice and complexity of differential diagnostics determine the need for a specific diagnostic algorithm. This algorithm should consider anamnestic data along with the course of the disease, clinical, laboratory and instrumental examination, including neuroimaging, analysis of CSF for oligoclonal bands, analysis for IgG antibodies to AQP4, which will allow to carry out diagnostics and to decide on tactics for further management of patients of this cohort. Further research is needed to conduct additional studies for optimization of tactics for dynamics monitoring and improvement of diagnostic, treatment and rehabilitation measures in patients with Devic's opticomyelitis, including appropriate immunological control, given the complexity of differential diagnostics and the affinity of this pathology to multiple sclerosis.

**KEY WORDS:** Devic's opticomyelitis, diagnostic algorithm, differential diagnostics, treatment, aquaporin-4, IgG antibodies

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**INTRODUCTION**

Opticomyelitis (neuromyelitis optica, Devic's syndrome / disease) is a severe idiopathic demyelinating disease of the central nervous system, characterized by the prevailing lesion of the optic nerves and spinal cord with relative intactness of brain structures [1]. For a long time, this disease was considered within the framework of malignant variants of multiple sclerosis. However, the progress in the pathogenesis study on demyelinating diseases of the central nervous system (CNS) at the end of the 20th century allowed the researchers to distinguish opticomyelitis as a separate nosological form [2].

**CASE REPORT**

The aim of the research was to analyze the contemporary scientific literature on Devic's opticomyelitis and to present a case report from our clinical practice.

Opticomyelitis is a disease, which is autoimmune by nature. The ratio of affected women to men is (2-8):1. The age of the disease onset varies from 1 to 77 years, most often starting at the age of 35-47 years, more commonly in the non-Caucasian representatives [3; 4]. The

pathogenesis of the disease is based on the formation of NMO-IgG autoantibodies to aquaporin-4 (AQP4) (the protein of water-conveying canals of cell membranes), which localizes at the peduncles of astrocytes forming the blood-brain barrier. The highest concentration of AQP4 in the CNS is observed in the gray matter of the spinal cord, hypothalamus, periventricular areas [5].

Therefore, the foci in the brainstem and hypothalamus may be considered relatively characteristic and specific; the cerebral foci by their localization tend to those areas of the brain that display a high level of immunoreactivity to AQP4 [6]. Pathophysiologically, demyelination and necrosis of the white and gray matters occur in opticomyelitis. Chronic foci of inflammation in the brain are represented by cystic degeneration, gliosis, and nerve atrophy, which can lead to the development of secondary syringomyelia [7]. Initially, in the clinical presentation of the disease, there is visual impairment in the form of reduction, until its complete loss, and after a while the symptoms of severe transverse myelitis develop – para- and tetraparesis, impaired function of the pelvic organs. It is now assumed that opticomyelitis can have both a single-phase and a remittent type of the course; however, repeated attacks are less typical than remissions [8].

Optic neuritis is one of the main symptoms of opticomyelitis. During ophthalmoscopy, a normal pattern of the fundus is more often observed, a slightly blurred optic discs, slight edema, atrophy and pallor of the optic nerves in chronic cases. In opticomyelitis, optic neuritis is usually bilateral, commonly preceding myelitis (in 80% of cases). In a few weeks, less frequently in a few months, severe transverse myelitis develops, whose typical symptoms are muscle weakness, spasticity, discoordination, ataxia, Lhermitte's sign, urinary retention, autonomic dysfunction, possible sensory disorders below the level of the lesion of the spinal cord. In most cases, myelitis occurs less than 3 months later. However, in 20% of cases, transverse myelitis may precede optic neuritis [9].

Changes in the optic nerves can be detected by neuroimaging, since they involve the optic nerves over a greater length, unlike changes in multiple sclerosis. In almost all cases, opticomyelitis has to be differentiated from multiple sclerosis. In opticomyelitis, either brain MRI does not reveal any pathological changes, or in almost half of cases there are non-specific, often asymptomatic, foci of demyelination. On MRI in multiple sclerosis, foci in the spinal cord usually do not exceed one segment in length, whereas in opticomyelitis, foci exceeding three or more segments are visualized.

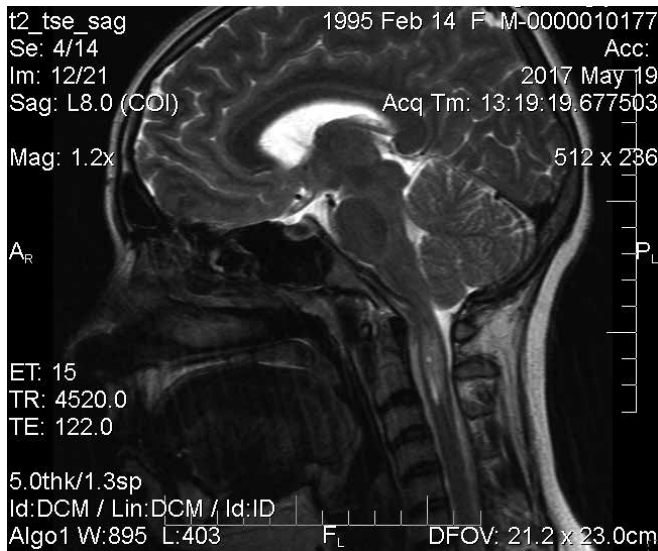
The analysis to determine IgG antibodies to AQP4 is diagnostically important. Additional features are the results of CSF analysis, its study for the presence of oligoclonal bands. In 2008, the international work group reviewed and formulated the diagnostic criteria for optic neuromyelitis (by D.H. Miller et al., 2008): 1) the "major" criteria (the presence of all essential criteria is required, with an indefinite time interval between them): optic neuritis with the lesion of one or both eyes; transverse myelitis, clinically complete or incomplete, but associated in the acute period with the presence of radiologically confirmed lesion of the spinal cord that extends longer than 3 segments on T2-weighted MRI images and is hypo-intensive on T1-weighted images; lack of data on sarcoidosis, vasculitis, systemic lupus erythematosus, Sjogren's syndrome, or other explanation for the disorder; 2) the "minor" criteria (at least one must be relevant): the latest brain MRI should display no pathology or detect only pathological changes that do not meet Barkoff's criteria as reflected in McDonald's criteria (2005); positive serum or cerebrospinal fluid test for NMO-IgG / antibodies to AQP4.

In terms of therapeutic tactics, there is currently no common standard in the treatment of this pathology. Symptomatic and restorative therapies are used to support the existing neurological functions. For the treatment of myelitis and optic neuritis attack, high doses of corticosteroids are used (methylprednisolone 1000 mg daily intravenously No.5 consecutively), then it is recommended to administer prednisolone maintenance therapy at a dose of 1 mg / kg / day as part of initial immunosuppressive therapy to prevent recurrent attacks [10]. Unfortunately, myelitis often poorly yields to such therapy, and sometimes it is even aggravated. In these cases, plasmapheresis is recommended (seven

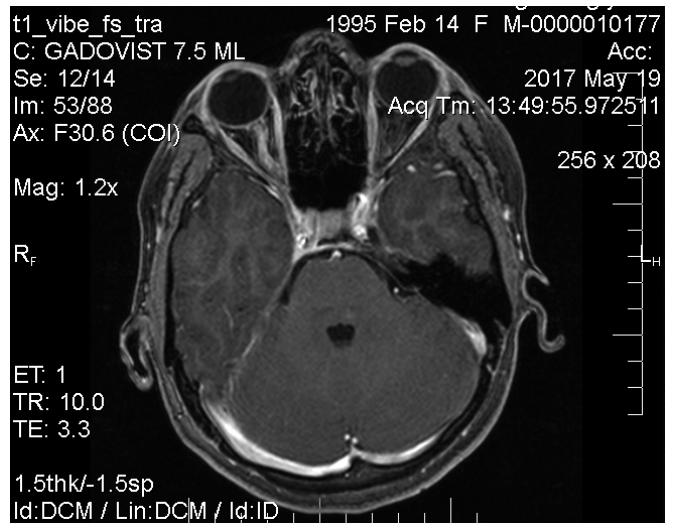
sessions a day, in 55 ml / kg per metabolic transfusion) [11]. For long-term treatment of Devic's opticomyelitis, it is recommended to apply immunomodulatory therapy, rather than the immunosuppressive one. Most practitioners consider the combination of oral prednisolone and azathioprine to be a therapy of choice, followed by gradual reduction of corticosteroids to the lowest effective dose or their complete withdrawal and azathioprine monotherapy [12]. Despite therapy, Devic's opticomyelitis in some cases leads to lethal outcome, often as a result of severe attack of myelitis with involvement of the cervical spinal cord and development of respiratory disorders [13].

Case presentation: Patient O., born in 1995, in March 2018 presented with urinary disorders (urinary retention), weakness in the legs, numbness and impaired sensitivity in the legs and the lower body, grogginess when walking, decreased vision, pain in the thoracic spine, excessive fatigue and severe weakness. The patient considered herself ill since December 2015, when she had noticed a sharp deterioration of vision in the right eye. She had been treated at the ophthalmology department of the regional hospital (retrobulbar neuritis on the right) with a positive dynamics. In May 2017, she again had suffered from visual impairment in the right eye. The patient had undergone MRI of the brain with intravenous contrast (MR signs of the focal lesions of the spinal cord at the level of C2-C3, the right optic nerve, probably of demyelinating character (Fig. 1, 2)), pulse therapy with corticosteroids (methylprednisolone 1000 mg intravenously, by drop infusion No.5) with a tendency to positive dynamics, but in 2 weeks there had been a decrease in vision in the left eye as well. Plasmapheresis course had been conducted at the end of May 2017, and there had been a slight positive dynamics. In August 2017, the condition had aggravated again: visual impairment and sensitivity in the legs and lower left trunk with weakness in the left leg had developed. MRI of the brain, cervical and thoracic spine had been performed (no data on the volumetric, focal processes of the brain had been detected; MR signs of demyelinating changes in the spinal cord at the levels of C1-C3, C7-Th4, Th7-Th8 (Fig. 3)), the course of pulse therapy with corticosteroids (methylprednisolone 1000 mg intravenously by drop infusion No.5) and neurometabolic therapy had been conducted with a positive dynamics. It was also known that the patient had been periodically self-treated with non-steroidal anti-inflammatory drugs for pain in the spine, mostly in the cervical and thoracic regions. Marked deterioration of the condition had been observed about 3 days before addressing a doctor after contracting acute respiratory viral infection with hyperthermia, when there had been a significant increase in leg numbness, weakness, and urinary retention had joined the abovementioned complaints.

At the time of admission: skin and visible mucous membranes were pale pink; blood pressure was 115/70 mm Hg, heart rate was 74 beats per minute; heart tones were rhythmic, sound; abdomen was soft on palpation, sensitive in the lower parts, the bottom of the bladder was determined by palpation and percussion – urinary retention. Neurological



**Fig. 1.** MRI of the brain of patient O. (May 19, 2017)



**Fig. 2.** MRI of the brain of patient O. (May 19, 2017)



**Fig. 3.** MRI of the cervical and thoracic spine of patient O. (August 07, 2017)



**Fig. 4.** MRI of the cervical and thoracic spine of patient O. (March 23, 2018)

status: palpebral fissures D=S, pupils D=S. Photoreactions were preserved. There is no nystagmus. The exit points of the V pair were painless. The face was symmetrical, the tongue was along the middle line. Reflexes from the back of the pharynx and soft palate were preserved. Swallowing was not impaired, the voice was loud. The speech was preserved. The muscular tone was dystonic in hands, and increased by spastic type in feet. Barré test was “+” in the legs. The strength in hands was retained, and reduced in the legs to 3.0 points. Babinski's symptom was (+) on two sides. Hand reflexes D=S were high, with extended reflex zones, abdominal abs reflexes, knee reflexes D=S, high, Achilles reflexes D=S, polykinetic. There is a pronounced descending hypesthesia by the conductor type from Th10 with gross disturbance of vibration sensitivity in the legs. There were no meningeal signs. Coordination tests were performed with ataxia in the lower extremities. The patient was anxious, with distal hyperhidrosis, hypothermia; dysfunction of the pelvic organs by central type (urinary retention).

The comprehensive clinical laboratory and instrumental examination was conducted along with consultations from related specialists (urologist – acute urinary retention; ophthalmologist – partial atrophy of the optic nerves in

both eyes), which allowed us to confirm the absence of systemic vasculitis, other rheumatological and infectious pathologies. During the examination, the following results attracted special attention and served as a confirmation of the correctness of our diagnostic search: analysis for IgG antibodies to AQP4 (21.03.18): 1:320 (positive result); MRI of the brain, cervical and thoracic spine with intravenous contrast (23.03.18): no data on volumetric, demyelinating processes of the brain were found, the effects of previously sustained myelitis with extensive cystic, cicatricial and atrophic changes at the level of C1-Th8. There has been a negative dynamics as compared to MRI as of August 2017.

Based on the patient's complaints, case history and features of clinical course, objective neurological status, clinical laboratory and additional examination methods, characteristic MR-patterns, consultations of related specialists and differential diagnostics, we made the clinical diagnosis according to ICD-10: G36.0 Devic's opticomyelitis, exacerbation, with the sustained bilateral lesion of the optic nerves in the form of retrobulbar neuritis (May 2017) with the development of partial atrophy of the optic nerves in both eyes, spinal cord lesions with common cystic, cicatricial and atrophic alterations at C1-Th8 level

(according to MRI data as of 23.03.2018 (Fig.4)) with moderate lower paraparesis, expressed by sensory ataxia, sensory disturbances by the descending conductive type from Th10, impaired function of pelvic organs by the type of acute urinary retention, asthenic and neurotic syndrome.

The patient underwent a course of pulse therapy with corticosteroids (methylprednisolone 1000 mg intravenously by drop infusion No.5) with subsequent transition to oral methylprednisolone according to the scheme of gradual dose de-escalation, angio-, neuroprotective, antioxidant and physiotherapy. The patient refused from therapy in combination with azathioprine. During inpatient stay, no intolerance to medicines was observed. As a result of the conducted treatment, a positive dynamics was observed: urination resumed (acute urinary retention regressed to the neurogenic bladder), strength in the legs increased (moderate lower paraparesis regressed to the mild one), ataxia and general weakness decreased, sensitivity resumed in the lower part of the trunk and improved up to the level of the upper third of the thighs.

## CONCLUSIONS

Thus, taking into account the widespread cases of demyelinating pathology in medical practice, and keeping in mind the cases of Devic's opticomyelitis and complexity of their differential diagnostics, requiring clear clinical thinking, the necessity to follow a specific diagnostic algorithm becomes obvious. This algorithm should consider anamnestic data along with the course of the disease, clinical, laboratory and instrumental examination, including neuroimaging, analysis of CSF for oligoclonal bands, analysis for IgG antibodies to AQP4, which will allow to carry out diagnostics and to decide on tactics for further management of patients of this cohort. Further research is needed to conduct additional studies for optimization of tactics in dynamics monitoring and improvement of diagnostic, treatment and rehabilitation measures in patients with Devic's opticomyelitis, including appropriate immunological control, given the complexity of differential diagnostics and the affinity of this pathology to multiple sclerosis.

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## Conflicts of interest:

*Authors declare no conflict of interest.*

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