DOI: 10.31082/1728-452X-2021-224-2-43-50
УДК 617.731-002+616.832-004.2-071

OPTICONEUROMYELITIS DEVIK: A BRIEF HISTORICAL EXCURSION (REVIEW) AND OWN CLINICAL OBSERVATION

Saule T TURUSPEKOVA[1], https://orcid.org/0000-0002-4593-3053,
Erkin S NURGUZHAEV[1], https://orcid.org/0000-0001-8110-9013,
Bayan K DEMESINOVA[1],https://orcid.org/0000-0002-7302-6178,
Nikolay V STEPUK[2], https://orcid.org/0000-0003-3750-0546,
Togzhan Zh MUKASHEVA[2], https://orcid.org/0000-0002-0471-4137,
Elmira B ATANTAEVA[1], https://orcid.org/0000-0001-8787-1906,
Klara K ALMAKHANOVA[1], https://orcid.org/0000-0002-0498-0566

[1]Asfendiyarov Kazakh National Medical University, Almaty, Republic of Kazakhstan,
[2]City Clinical Hospital No. 1, Almaty, Republic of Kazakhstan

Turuspekova ST

Neuromyelitis optic is an autoimmune, so-called "aquaparinopathy" of the central nervous system (CNS), characterized by the development of inflammatory demyelinating foci primarily located in the spinal cord and optic nerves, clinically manifested by a combination of optic neuritis and longitudinal-transverse myelitis syndromes. In recent years, this condition has generated tremendous interest among scientists and clinical neurologists, aided by the discovery of a highly specific autoantibody of immunoglobulin (Ig)G (NMO-IgG) in serum, targeting the most abundant astrocytic water channel aquaporin-4 (AQP4), which is present almost in 80% of patients with opticoneuromyelitis. This fruitful discovery clearly showed that neuromyelitis optics is not a subform of multiple sclerosis (MS) in most cases, as has been assumed for decades, and emphasizes the need for timely and accurate diagnosis in view of the differences in optimal treatment options between them.

The article highlights the historical data on the study of opticoneuromyelitis, and also considers a clinical case of the disease in a 53-year-old woman of Asian race with a debut in the form of an episode of motor-sensory disorders of the type of right-sided hemsymptomatology with the subsequent development of spinal symptoms in the form of deep lower paraparesis with pelvic disorders. Repeated magnetic resonance imaging of the brain revealed a picture of microangiopathy, discirculatory encephalopathy. MRI examination of the spinal cord revealed a demyelinating lesion of the 3 cervical segments of the type of transverse myelitis. A blood test for antibodies to aquaporin-4 showed a positive result. The study of visual evoked potentials revealed axonal-demyenizing damage. Examination by an ophthalmologist revealed descending atrophy of the optic neurons. Pulse therapy with glucocorticoids with a positive clinical and laboratory response was used as induction therapy. A timely established correct diagnosis, prescribed adequate treatment help to stop exacerbations in a timely manner and improve the quality of life of such patients.

Keywords: opticoneuromyelitis, Devik's disease, autoimmune mechanisms, markers, diagnostics.

For reference: Turuspekova ST, Nurguzhaev ES, Demesinova BK, Stepuk NV, Mukasheva TZh, Atantaeva EB, Almakhanova KK. Opticoneuromyelitis Devik: a brief historical excursion (review) and own clinical observation. *Meditsina (Almaty) = Medicine (Almaty).* 2021;2(224):43-50. (In Russ.). doi: 10.31082/1728-452X-2021-224-2-43-50

Т Ұ Ж Ы Р Ы М
ДЕВИК ОПТИКОНЕЙРОМИЕЛИТІ: ҚЫСҚА ТАРИХИ ЭКСКУРС (ШОЛУ) ЖӘНЕ ЖЕКЕ КЛИНИКАЛЫҚ БАҚЫЛАУ

Сәуле Т. ТҰРҰСПЕКОВА[1], https://orcid.org/0000-0002-4593-3053,
Еркін С. НҰРҒОЖАЕВ[1], https://orcid.org/0000-0001-8110-9013,
Баян К. ДЕМЕСІНОВА[1], https://orcid.org/0000-0002-7302-6178,
Николай В. СТЕПУК[2], https://orcid.org/0000-0003-3750-0546,
Тоғжан Ж. МҰҚАШЕВА[2], https://orcid.org/0000-0002-0471-4137,
Эльмира Б. АТАНТАЕВА[1], https://orcid.org/0000-0001-8787-1906,
Клара К. АЛЬМАХАНОВА[1], https://orcid.org/0000-0002-0498-0566

[1]«С.Ж. Асфендияров атындағы Қазақ Ұлттық медицина университеті» КеАҚ, Алматы қ., Қазақстан Республикасы,
[2]«N1 қалалық клиникалық аурухана» ШЖҚ ҚКЕ, Алматы қ., Қазақстан Республикасы

Оптиконейромиелит – аутоиммунды, атап айтқанда орталық жүйке жүйесінің (ОЖЖ) «аквапаринопатиясы», клиникалық оптикалық неврит және көлденең миелит синдромдарының бірлесуі арқылы көрінеді және жұлын мен көру нервінде біріншілік қабыну-демиелинсіздену ошақтарының орналасуы арқылы дамиды. Соңғы жылдары бұл жағдайлар ғалымдар мен клиникалық невропатолог-тар арасында аса қызығушылық тудырды, оған астроцитарлы су каналы аквапорин-4 (AQP4)-ке бағытталған оптиконейромиелитпен науқастардың 80%-да дерлік қан сарысуында болатын спецификалық аутоантидене иммуноглобулинінің (Ig)G (NMO-IgG) табылуы әсер болды.

Contacts: Turuspekova Saule, Doctor of Medical Sciences, Professor, Head of the Department of Nervous Diseases, Asfendiyarov KazNMU, Almaty, e-mail: turuspekova.s@kaznmu.kz

Контакты: Туруспекова Сауле Тлеубергеновна, доктор медицинских наук, профессор, заведующий кафедрой нервных болезней, НАО «КазНМУ им. С.Д. Асфендиярова», г. Алматы, e-mail: turuspekova.s@kaznmu.kz

Received: 12.08.2021
Accepted in print: 20.09.2021

Бұл жемісті жаңалық соңғы жылдары тек болжам болып келген – оптиконейромиелиттің көп жағдайда шашыранды склероздың (ШС) бір түрі еместігін және уақытылы нақты диагностика мен оңтайлы емдеу варианттарының ерекшелік қажеттігін көрсетті.

Мақалада оптиконейромиетті зерттеу мәселелері бойынша тарихи мәліметтерге шолу жасалған, сонымен қатар азиаттық 53 жастағы науқас әйелдің оңжақтық гемисимптоматика типі бойынша мотосенсорлы бұзылыстары басталған, кейіннен кіші жамбас ағзалары бұзылыстарымен терең төменгі парапарез түрінде спинальды симптоматика дамыған клиникалық жағдайы қарастырылады.

Бірнеше рет өткізілген бас миының магнитно-резонансты зерттеуі микроангиопатия мен дисциркуляторлы энцефалопатияны анықтаған. Жұлынның МРТ-зерттеуі көлденең 3 мойын сегментінің көлденең миелит типі бойынша демиелінсізденіп зақымдануын анықтаған. Аквапорин-4 қарсы антиденелерге қан анализі оң нәтиже берген. Потенциалды шақыру арқылы көру дәрежесін зерттеу аксональды-демиелинсіздену зақымдануын анықтаған. Офтальмолог қарағанда көру нервілерінің төмендеуші атрофиясы анықталған. Индукционды терапия ретінде глюкокортикоидтармен пульстерапия қолданылған және оң клинико-лабораторлы нәтиже көрсеткен. Уақытылы қойылған дұрыс диагноз, адекватты емдеу уақытылы өршуді басып, мұндай науқастардың өмір сүру сапасын жақсартуға септігін тигізеді.

Негізгі сөздер: оптиконейромиелит, Девик ауруы, аутоиммунды механизмдер, маркерлер, диагностикалау.

Р Е З Ю М Е

ОПТИКОНЕЙРОМИЕЛИТ ДЕВИКА: КРАТКИЙ ИСТОРИЧЕСКИЙ ЭКСКУРС (ОБЗОР) И СОБСТВЕННОЕ КЛИНИЧЕСКОЕ НАБЛЮДЕНИЕ

Сауле Т. ТУРУСПЕКОВА[1], https://orcid.org/0000-0002-4593-3053,
Еркин С. НУРГУЖАЕВ[1], https://orcid.org/0000-0001-8110-9013,
Баян К. ДЕМЕСИНОВА[1], https://orcid.org/0000-0002-7302-6178,
Николай В. СТЕПУК[2], https://orcid.org/0000-0003-3750-0546,
Тогжан Ж. МУКАШЕВА[2], https://orcid.org/0000-0002-0471-4137,
Эльмира Б. АТАНТАЕВА[1], https://orcid.org/0000-0001-8787-1906,
Клара К. АЛЬМАХАНОВА[1], https://orcid.org/0000-0002-0498-0566

[1]НАО «Казахский Национальный медицинский университет им. С.Д. Асфендиярова», г. Алматы, Республика Казахстан,
[2]ГКП на ПХВ «Городская клиническая больница №1», г. Алматы, Республика Казахстан

Оптиконейромиелит – аутоиммунная, так называемая «аквапаринопатия» центральной нервной системы (ЦНС), характеризующаяся развитием воспалительно-демиелинизирующих очагов, первично расположенных в спинном мозге и зрительных нервах, клинически проявляющихся сочетанием синдромов оптического неврита и продольно-поперечного миелита. В последние годы это состояние вызвало огромный интерес среди ученых и клинических неврологов, чему способствовало обнаружение высокоспецифичного аутоантитела иммуноглобулина (Ig)G (NMO-IgG) в сыворотке крови, нацеленного на самый распространенный астроцитарный водный канал аквапорина-4 (AQP4), который присутствует почти у 80% пациентов с оптиконейромиелитом. Это плодотворное открытие ясно показало, что в большинстве случаев оптиконейромиелит не является подформой рассеянного склероза (РС), как предполагалось на протяжении десятилетий, и подчеркивает необходимость своевременной и точной диагностики ввиду различий оптимальных вариантов лечения между ними.

В статье освещены исторические данные по вопросу изучения оптиконейромиелита, а также рассматривается клинический случай заболевания у женщины 53-х лет, азиатской расы, с дебютом в виде эпизода мотосенсорных расстройств по типу правосторонней гемисимптоматики с последующим развитием спинальной симптоматики в виде глубокого нижнего парапареза с тазовыми расстройствами. Неоднократно проводимое магнитно-резонансное исследование головного мозга обнаруживало картину микроангиопатии, дисциркуляторной энцефалопатии. МРТ-исследование спинного мозга выявило демиелинизирующее поражение 3-х шейных сегментов по типу поперечного миелита. Анализ крови на антитела к аквапорину-4 показал положительный результат. Проведенное исследование зрительных вызванных потенциалов обнаружило аксонально-демиенизирующее повреждение. Осмотр офтальмолога выявил нисходящую атрофию зрительных нервов. В качестве индукционной терапии применялась пульс-терапия глюкокортикоидами с положительным клинико-лабораторным ответом. Вовремя установленный правильный диагноз, назначенное адекватное лечение помогают своевременно купировать обострения и повысить качество жизни таких пациентов.

Ключевые слова: оптиконейромиелит, болезнь Девика, аутоиммунные механизмы, маркеры, диагностика.

Для цитирования: Туруспекова С.Т., Нургужаев Е.С., Демесинова Б.К., Степук Н.В., Мукашева Т.Ж., Атантаева Э.Б., Альмаханова К.К. Оптиконейромиелит Девика: краткий исторический экскурс (обзор) и собственное клиническое наблюдение // Медицина (Алматы). 2021;2(224):43-50. doi: 10.31082/1728-452X-2021-224-2-43-50

Opticoneuromyelitis (ONM, ICD-10, G36.0) is an idiopathic severe inflammatory demyelinating disease characterized by selective involvement of the optic nerves and spinal cord with relative intactness of the brain structures, leading to profound disability [1]. It is also called Devic's opticomyelitis, Devic's syndrome, or Devic's disease. The disease has significant similarities to multiple sclerosis (MS) in its clinical manifestations and imaging findings and has long been considered a variant of MS [2]. Views on the place of this disease in the classification of neurological diseases, diagnostic criteria, and therapy tactics have long been controversial. Some scientists traditionally considered Devic's disease as one of the malignant variants of MS, some others believed that this pathology is a form of acute disseminated encephalomyelitis (ADEM) [3], and some clinicians considered ONM as a separate nosological entity in the system of nervous diseases [4]. However, the situation changed dramatically since the specific biomarkers, antibodies to aquaporin-4 protein (AQP4-IgG), were identified in an overwhelming number of patients with ONM in 2004, thus producing the first diagnostic criteria [5, 6, 7].

The disease became known as Devic's disease after the seminal 1894 report when E. Devic used the term "Neuromyelitis optica acuta" for the occasion of the French Congress in Lyon [8]. The same year his student F. Gault published a doctoral dissertation entitled *"De la neuro - myélite optique aiguë,"* which included reviewing the existing medical literature and analyzing a clinicopathological case from his practice [7]. However, in their reviews, the authors omitted some early descriptions of probable ONM, probably due to limited access to bibliographic resources at that time.

So, even 26 years before the first case mentioned by Devic E. and Gault F., in 1844, the Genoese physician Giovanni Battista Pescetto reported a 42-year-old man who simultaneously developed acute amaurosis and cervical myelitis, with complete recovery after extensive bloodletting [9]. Attention should also be drawn to a previously forgotten review by the German ophthalmologist Friedrich Albin Schanz (known to ophthalmologists as one of the inventors of the "slit lamp'), where most of the clinical and pathological features currently thought to be characteristic of ONM are indicated [10].

For a long time, the case described in a lecture "On the ophthalmoscopic signs of spinal disease" published in *The Lancet* in 1870 was considered the first description of a patient with possible ONM in the Western literature. The lecture was delivered by Thomas Clifford Allbutt, known to many as the inventor of the clinical thermometer and one of the main initiators of the clinical use of the ophthalmoscope. His idea was the pathogenetic relationship between spinal cord injury and optic nerve damage [11].

However, to be fair in terms of historical chronology, the first description of a patient with vision loss and signs of spinal cord damage was made by King Louis XVIII's first physician, the founder and lifelong president of the National Academy of Medicine, the French pathologist and anatomist Antoine Portal in 1804, when he published an early report on vision loss associated with inflammation of the spinal cord in the Marquis: The case of the Marquis de Causan [12].

Eugène Devis

ONM is a disease of an autoimmune nature that is prevalent in members of the non-European race. Unfortunately, it is currently very difficult to determine the true prevalence and incidence of Devic's disease. This is because, for many years, ONM has been regarded as a variant of MS, and even when there is isolated damage to the optic nerves and/or spinal cord, patients have been treated for MS or other diseases of the nervous system, such as myelitis.

Thomas Clifford Allbutt

Antoine Portal

The incidence of ONM is universal, but there is a higher prevalence in Asian countries [13]. Population-based studies on the prevalence and incidence of ONM are currently very limited and cover only a few countries in Europe, Asia, and South America. For example, according to a study conducted in Denmark, the incidence of ONM was 0.4 per 100,000 population, and the prevalence was 4.4 per 100,000 [14]. Although it is a rare disease in the Caucasian population, a retrospective population-based series of cases has shown the disease to be more common than previously believed [15]. Studies in other countries show the following prevalence figures: 0.5 per 100,000 in the Republic of Cuba, 1.0 per 100,000 in Mexico, and 1.4-2.8 per 100,000 in the United States [13].

ONM is most prevalent among females. The female to male ratio is (2-8):1 (1). According to the Mayo Clinic, the female-to-male ratio is 2.5 to 1 [16]. The age at which the disease begins varies, but it most often begins between 35 and 47 years of age. Thus, the average age of onset is higher in comparison with MS [17]. Single familial cases of Devic's opticomyelitis have been described in the literature. ONM is often associated (50-70%) with other autoimmune diseases – Sjogren's syndrome, autoimmune thyroiditis, rheumatoid arthritis, systemic lupus erythematosus, pernicious anemia, ulcerative colitis, primary sclerosing cholangitis, and thrombocytopenic purpura [18,19].

As mentioned above, ONM is currently the only demyelinating disease where a specific biomarker has been identified. Most early reports described ONM as a monophasic disease, but Beck (1927) and McAlpine (1938) reported patients with a relapsing course (17). In 2004, Lennon and Wingerchuk found neuroopticomyelitis-associated IgG (NMO-IgG) to distinguish ONM from MS [20]. A year later, Lennon and colleagues found that NMO-IgG selectively binds to the aquaporin-4 channel [21]. The NMO-IgG antibodies identified as being specific to ONM confirmed the autoimmune mechanisms of the pathogenesis of Devic's disease. Such antibodies are absent in patients with classical MS and other autoimmune, inflammatory, and non-inflammatory CNS diseases, as well as in healthy individuals [22].

The pathogenesis of the disease is due to the selective association of NMO-IgG with aquaporin-4 (AQP4), one of the major proteins of the CNS water channels localized in the astrocyte stems that form the blood-brain barrier (BBB). The highest concentration of AQP4 in the CNS is found in the grey matter of the spinal cord, hypothalamus, and periventricular areas [22].

A scheme of ONM pathogenesis is being considered, according to which AQP4-IgG circulating in the blood plasma penetrates through the damaged BBB and binds to ion channels located on astrocyte processes such as complement-mediated damage and leukocyte infiltration, leading to astrocyte necrosis [23-27]. In turn, this leads to secondary damage and death of oligodendrocytes and, consequently, secondary demyelination, which eventually leads to neuronal necrosis, i.e., neurodegeneration. Chronic foci of inflammation in the brain, represented by cystic degeneration, gliosis, and nerve tissue atrophy, can lead to the development of secondary syringomyelia [28]. Today, the detection of focal lesions of the nervous system outside the optic nerves and spinal cord is no longer a criterion for excluding the diagnosis of ONM [1].

Devic's disease affects the astrocytes' lipoid coating that protects the optic and cerebrospinal nerves. Therefore, the initial clinical manifestations are visual disturbances, followed by symptoms of severe transverse myelitis after some time, in most cases within three months.

In ONM, optical neuritis that precedes myelitis [29] in 80% of cases is usually bilateral and manifests as visual disturbances in the form of decreased or even complete loss, positive visual phenomena in the form of flickering lights, spots or lines, and pain syndromes in the orbital region. Ophthalmoscopy more often reveals a normal fundus picture, a slight fading of the optic disc borders, mild edema, atrophy, and pallor of the optic discs in chronic cases [30, 31, 32]. However, it is believed that eye damage in Devic's disease is more severe than in MS, with a high incidence of residual effects [32].

However, in 20% of cases, transverse myelitis may precede optical neuritis and/or occur in an isolated form. An acute partial transverse or longitudinal diffuse myelitis with all the symptoms typical of spinal cord damage is a variant of this course. These include, first of all, gross symmetrical motor-sensory disorders, Lhermitte's symptoms, paroxysmal tonic muscle spasms, radicular pain, dyscoordination, ataxia, pelvic organ dysfunction, and others [19, 33].

Both monophasic and recurrent forms of the disease are now accepted (34). However, recurrent attacks are less typical, as well as remissions.

The prognosis for the disease is rather pessimistic, although several possible outcomes are possible. In some

cases, a complete recovery is expected; another variant of the course of ONM is the presence of remissions of the disease. Although the severity of neurological deficits is greater in the relapsing form, the long-term prognosis is more favorable than in the monophasic form, as there is no increase in neurological deficits [35]. The worst outcome is progressive worsening and death [1].

Considering the complexity of diagnosis, the small study of the disease, and the rare occurrence of this pathology***, we describe our clinical observation***, when the debut of ONM was disguised as a vascular pathology and was treated as a stroke.

Patient M-va Z., a Kazakh woman of 53, complained of general weakness, limitation of limb mobility, inability to stand up and walk due to limb weakness, pelvic organ dysfunction such as stool retention, constant feeling of stiffness in her arms and body.

From the anamnesis: the patient considered herself ill since 30 June 2017, when psycho-emotional stress caused weakness, numbness, and tingling in the shoulder and right upper extremity. The above complaints began to escalate, and on 3 July 2017, she was summoned to the emergency hospital and admitted to the CRH. The illness was considered as a stroke (Ischemic stroke). In July 2017, an MRI scan of the brain showed a picture of mixed dyscirculatory encephalopathy. There were manifestations of microangiopathy. In August 2017, her condition worsened, with weakness and numbness in the right lower extremity. She was repeatedly treated in the neurology department. A repeated CT scan of the brain showed a picture of encephalopathy. On 18.09.2017, she noted weakness in both lower extremities, shooting pains in the legs, spasmodic muscle pain, and inability to move independently. She was admitted to the neurology department for a second time and treated for chronic cerebral ischemia and other cerebrovascular syndromes in DEP cerebrovascular disease.

In January 2018, the patient was admitted to the neurology department of Municipal Clinical Hospital No. 1, complaining of weakness in the legs and inability to move independently with episodes of urinary retention.

Her neurological status revealed spinal symptoms in the form of transverse myelitis and pelvic disorder. The doctors and teaching staff of the Department of Nervous Diseases with a course in neurosurgery suspected Devic's disease. A spinal MRI of the spinal cord, consultation with an ophthalmologist, a visual evoked potential study, and a lumbar puncture with further investigation of the spinal fluid for antibodies to AQP4 were prescribed to substantiate the decision made. Differential diagnosis with MS and other autoimmune diseases was also performed.

Neuroimaging changes in the spinal cord revealed a probable demyelinating lesion of the three cervical segments in the form of transverse myelitis, which is consistent with the clinical picture (Fig. 1).

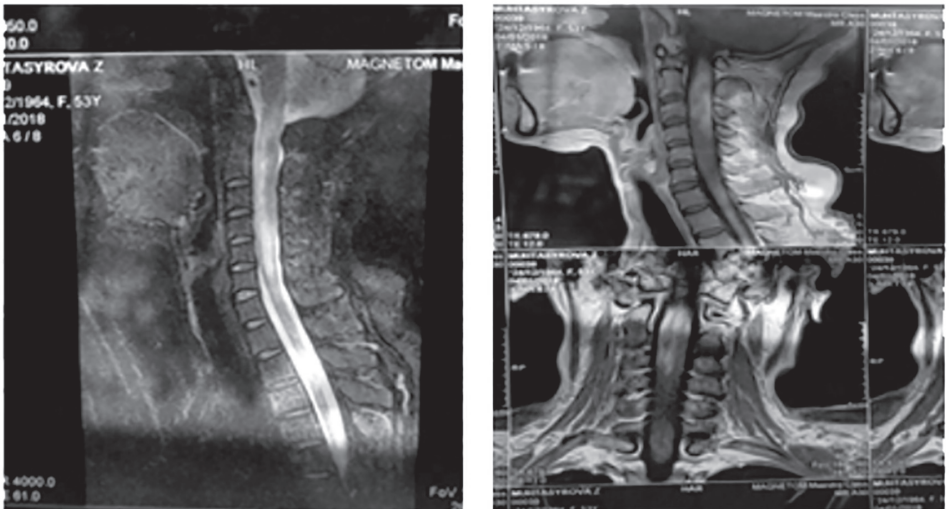

Figure 1 – MR imaging of demyelinating spinal cord disease (debut of MS? spinal form?). An expansive process?

The consultation with an ophthalmologist revealed pale optic discs, clear borders, descending optic atrophy. An examination of the visual evoked potentials revealed that cortical responses are not very stable; amplitudes vary with varying degrees of decrease in several samples, on the left up to unrecorded. The information-conduction to the cortex is impaired on both sides, moderately on the right and markedly on the left, axonal-demyelinating type. Consultation with a rheumatologist ruled out active rheumatologic disease due to the lack of clinical picture and no history of them and laboratory findings of negative tests for systemic and infectious diseases.

Blood serum and cerebrospinal fluid tests for NMO-IgG/antibodies to aquaporin-4 were positive, with minor lymphocytic pleocytosis. In general and biochemical blood tests, there was no significant abnormality.

All the prescribed diagnostic measures made it possible to establish a clinical diagnosis. Pulse therapy and symptomatic supportive treatment were administered, with positive dynamics, complete regression of pain syndrome, pelvic disorders, and a partial improvement of motor and sensory disturbances (Fig. 2).

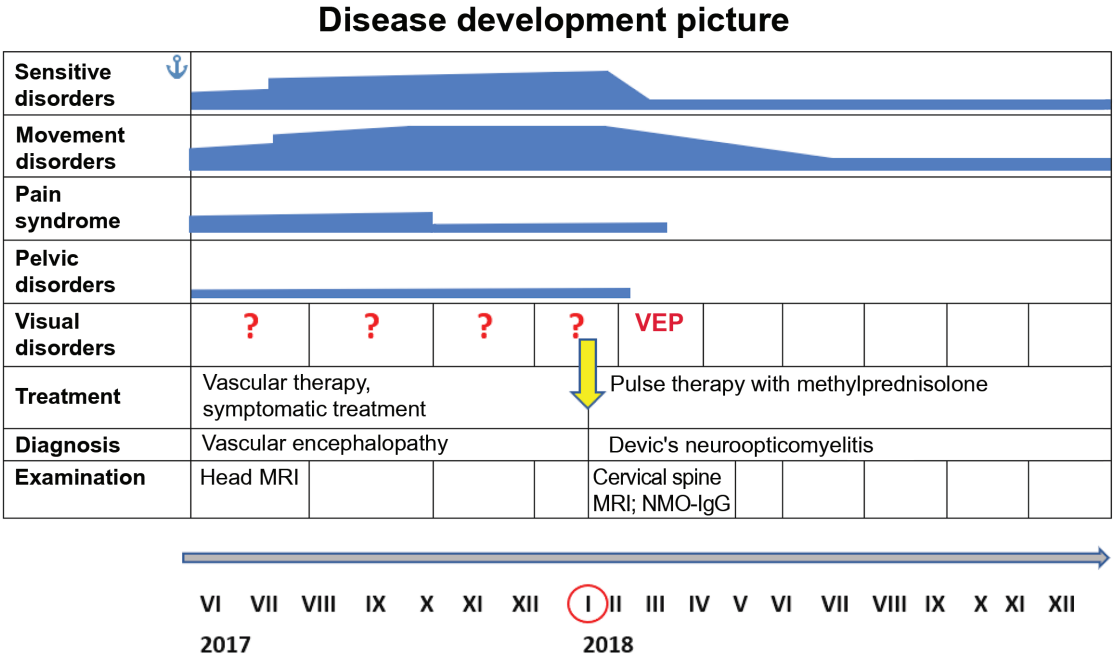

Figure 2 – The clinical case timeline

DISCUSSION

The presented clinical case had transverse, longitudinal myelitis: an extended spinal cord lesion of demyelinating nature at the CI-TVII level, clinically manifesting as partial or complete involvement of the transverse spinal cord. CSF study revealed lymphocytic pleocytosis, and MRI revealed a focus of demyelinating nature accumulating contrast substance, which indicated an inflammatory nature of the disease. After laboratory examination, infectious and systemic diseases were ruled out. MS was also excluded since it is not characterized by the spread of foci in the spinal cord over more than three segments and the absence of focal changes in the brain substance, observed in this clinical case. Devic's optic neuromyelitis was diagnosed based on the clinical picture, contrast-enhanced neuroimaging of the brain and spinal cord, and a positive result of a specific blood test for aquaporin-4 antibodies.

CONCLUSIONS

So, understanding the differences in the clinical course, manifestations, and characteristic changes of the brain and spinal cord according to neuroimaging data in Devic's optic neuromyelitis enables us to choose the right treatment strategy, improve the quality of life, and reduce the degree of disability of patients. We hope that the described clinical case of Devic's disease from our practice will add to our knowledge of this rare and severe disease.

Transparency of the research

The research was not sponsored. The authors are solely responsible for submitting the final manuscript for publication.

Declaration of financial and other relationships

All authors participated in writing the first version of the article and reviewed it critically for important intellectual content. All authors approved the final version of the article before submitting it for publication. The authors did not receive a fee for the research.

Conflict of Interest

The authors declare no conflict of interest.

REFERENCES

1. Mironenko TV, Khubetova IV. Opticomyelitis (Devik's disease). Scientific review and own clinical observation. *Mezhdunarodnyi nevrologicheskii zhurnal = International Neurological Journal.* 2015;1(71):141-147. (In Russ.)

2. Arman Eshaghi, Sadjad Riyahi-Alam, Roghayyeh Saeedi, et al. Classification algorithms with multi-modal data fusion could accurately distinguish neuromyelitis optica from multiple sclerosis. *Neuroimage Clin.* 2015;7:306-314. https://doi.org/10.1016/j.nicl.2015.01.001

3. Modi G, Mochan A, Modi M. Saffer D. Demyelinating disorder of the central nervous system occurring in black South Africans. *J Neurol Neurosurg Psychiatry.* 2001;70(4):500-505. PMID: 11254774. PMCID: PMC1737298. doi: 10.1136/jnnp.70.4.500

4. Wingerchuk DM, Weinshenker BG. Neuromyelitis optica. *In*: McDonald WI, Noseworthy JH, eds. Multiple Sclerosis 2. Woburn, MA, Butterworth-Heinemann, 2003:243–258

СПИСОК ЛИТЕРАТУРЫ

1. Мироненко Т.В., Хубетова И.В. Оптикомиелит (болезнь Девика). Научный обзор и собственное клиническое наблюдение // Международный неврологический журнал. 2015;1(71):141–147

2. Arman Eshaghi, Sadjad Riyahi-Alam, Roghayyeh Saeedi et al. Classification algorithms with multi-modal data fusion could accurately distinguish neuromyelitis optica from multiple sclerosis // Neuroimage Clin. 2015;7:306-314. https://doi.org/10.1016/j.nicl.2015.01.001

3. Modi G., Mochan A., Modi M., Saffer D. Demyelinating disorder of the central nervous system occurring in black South Africans // J Neurol Neurosurg Psychiatry. 2001;70(4):500-505. PMID: 11254774. PMCID: PMC1737298. doi: 10.1136/jnnp.70.4.500

4. Wingerchuk D.M., Weinshenker B.G. Neuromyelitis optica. *In:* McDonald WI, Noseworthy JH, eds. Multiple Sclerosis 2. Woburn, MA, Butterworth-Heinemann, 2003:243–258

5. Barhate KS, Ganeshan M, Singhal BS. A clinical and radiological profile of neuromyelitis optica and spectrum disorders in an Indian cohort. *Ann Indian Acad Neurol.* 2014;17(1):77–81. doi: 10.4103/0972-2327.128559

6. Mehdipour-Dastjerdi R, Ashtari F, Shaygannejad M, et al. The pathologic and diagnostic in magnetic resonance imaging of brain and cervical spine of patients with neuromyelitis optica spectrum disorder. *Iran J Neurol.* 2018 Apr. 4;17(2):58–63

7. Makarov S, Pashkovskyi V, Perveieva K. Neuromyelitis Optica: New Data about Pathogenesis, Methods of Diagnostic and Treatment, Current Views about the Place of this Pathology in Structure of Neurological Diseases and Criteria of Diagnosis. *Nevrologiia i neirokhirurgiia. Vostochnaia Evropa = Neurology and Neurosurgery. Eastern Europe.* 2019;9(1):60-72. (In Russ.)

8. Jarius S, Wildemann B. The history of neuromyelitis optica. *J Neuroinflammation.* 2013 Jan 15;10:8. doi: 10.1186/1742-2094-10-8. PMID: 23320783; PMCID: PMC3599417

9. Jarius S, Wildemann B. 'Noteomielite' accompanied by acute amaurosis (1844). An early case of neuromyelitis optica. *J Neurol Sci.* 2012;313:182–184. doi: 10.1016/j.jns.2011.09.025

10. Jarius S, Wildemann B. Devic's disease before Devic: On the contribution of Friedrich Albin Schanz (1863-1923). *J Neurol Sci.* 2017 Aug 15;379:99-102. doi: 10.1016/j.jns.2017.05.044. Epub 2017 May 23. PMID: 28716289

11. Jarius S, Wildemann B. On the contribution of Thomas Clifford Allbutt, F.R.S., to the early history of neuromyelitis optica. *J Neurol.* 2013 Jan;260(1):100-4. doi: 10.1007/s00415-012-6594-3. Epub 2012 Jul 11. PMID: 22782261

12. Jarius S, Wildemann B. The case of the Marquis de Causan (1804): an early account of visual loss associated with spinal cord inflammation. *J Neurol.* 2012, 259(7):1354–1357. doi: 10.1007/s00415-011-6355-8

13. Drori T, Chapman J. Diagnosis and classification of neuromyelitis optica (Devic's syndrome). *Autoimmun Rev.* 2014 Apr-May;13(4-5):531-3. doi: 10.1016/j.autrev.2014.01.034. Epub 2014 Jan 11. PMID: 24424197

14. Papp V, Illes Z, Magyari M, Koch-Henriksen N, et al. Nationwide prevalence and incidence study of neuromyelitis optica spectrum disorder in Denmark. *Neurology. 2018;* 91(24)e2265-e2275. doi: 10.1212/WNL.0000000000006645

15.Asgari N, Lillevang ST, Skejoe HP, Falah M, Stenager E, Kyvik KO. A pop ulation-based study of neuromyelitis optica in Caucasians. *Neurology.* 2011 May 3;76(18):1589-95. doi: 10.1212/WNL.0b013e3182190f74. PMID: 21536639; PMCID: PMC3269768

16. Wingerchuk DM, Weinshenker BG. Neuromyelitis optica (Devic's syndrome). *Handb Clin Neurol.* 2014;122:581-99. doi: 10.1016/B978-0-444-52001-2.00025-X. PMID: 24507536

17. Wingerchuk DM, Hogancamp WF, O'Brien PC, Weinshenker BG. The clinical course of neuromyelitis optica (Devic's syndrome). *Neurology*. 1999;53(5):1107-1114. doi:10.1212/wnl.53.5.1107

18. *Autoimmunnye zabolevaniia v nevrologii: Klinicheskoe rukovodstvo / pod red. I.A. Zavalishina i dr.* [Autoimmune diseases in neurology: Clinical guidelines / ed. I.A. Zavalishina et al.].Vol. 1. Moscow: Publishing House of ROOI "Human Health", 2014. 392 p.

19. Vinogradova ES, Novikov PI, Moiseev SV. Neuromyelitics optica (Devic's syndrome) is a rare variant of the nervous system impairment in systemic lupus erythematosus. *RMZh = RMJ.* 2018;12(II):103–106. (In Russ.)

20. Lennon VA, Wingerchuk DM, Kryzer TJ, et al. A serum autoantibody marker of neuromyelitis optica: distinction from multiple sclerosis. *Lancet*. 2004;364(9451):2106-2112. doi:10.1016/S0140-6736(04)17551-X

21. Lennon VA, Kryzer TJ, Pittock SJ, Verkman AS, Hinson SR. IgG marker of optic-spinal multiple sclerosis binds to the aquaporin-4 water channel. *J Exp Med*. 2005;202(4):473-477. doi:10.1084/jem.20050304

22. Giovannoni G. Neuromyelitis optics and anti-aquaporin-4 antibodies: widening the clinical phenotype. *J Neurol Neurosurg Psychiatry.* 2006;77(9):1001-2. doi: 10.1136/jnnp.2006.090944

23. Saadoun S, Waters P, Bell BA, Vincent A, Verkman AS, Papadopoulos MC. Intra-cerebral injection of neuromyelitis optica immu-

5. Barhate K.S., Ganeshan M., Singhal B.S. A clinical and radiological profile of neuromyelitis optica and spectrum disorders in an Indian cohort // Ann Indian Acad Neurol. 2014;17(1):77–81. doi: 10.4103/0972-2327.128559

6. Mehdipour-Dastjerdi R., Ashtari F., Shaygannejad M. et al. The pathologic and diagnostic in magnetic resonance imaging of brain and cervical spine of patients with neuromyelitis optica spectrum disorder // Iran J Neurol. 2018 Apr 4;17(2):58–63

7. Макаров С.А., Пашковский В.И., Первеева Е.И. Нейрооптикомиелит: новые данные о патогенезе, методах диагностики и лечения, современные взгляды на место патологии в структуре заболеваний нервной системы и критерии постановки диагноза // Неврология и нейрохирургия. Восточная Европа. 2019;9(1):60-72

8. Jarius S., Wildemann B. The history of neuromyelitis optica // J Neuroinflammation. 2013 Jan 15;10:8. doi: 10.1186/1742-2094-10-8. PMID: 23320783; PMCID: PMC3599417

9. Jarius S., Wildemann B. 'Noteomielite' accompanied by acute amaurosis (1844). An early case of neuromyelitis optica // J Neurol Sci. 2012;313:182–184. doi: 10.1016/j.jns.2011.09.025

10. Jarius S., Wildemann B. Devic's disease before Devic: On the contribution of Friedrich Albin Schanz (1863-1923) // J Neurol Sci. 2017 Aug 15;379:99-102. doi: 10.1016/j.jns.2017.05.044. Epub 2017 May 23. PMID: 28716289

11. Jarius S., Wildemann B. On the contribution of Thomas Clifford Allbutt, F.R.S., to the early history of neuromyelitis optica // J Neurol. 2013 Jan;260(1):100-4. doi: 10.1007/s00415-012-6594-3. Epub 2012 Jul 11. PMID: 22782261

12. Jarius S., Wildemann B. The case of the Marquis de Causan (1804): an early account of visual loss associated with spinal cord inflammation // J Neurol. 2012;259(7):1354–1357. doi: 10.1007/s00415-011-6355-8

13. Drori T., Chapman J. Diagnosis and classification of neuromyelitis optica (Devic's syndrome) // Autoimmun Rev. 2014 Apr-May;13(4-5):531-3. doi: 10.1016/j.autrev.2014.01.034. Epub 2014 Jan 11. PMID: 24424197

14. Papp V., Illes Z., Magyari M., Koch-Henriksen N. et al. Nationwide prevalence and incidence study of neuromyelitis optica spectrum disorder in Denmark" // *Neurology*. 2018;91(24):e2265-e2275. doi: 10.1212/WNL.0000000000006645

15. Asgari N., Lillevang S.T., Skejoe H.P., Falah M., Stenager E., Kyvik K.O. A population-based study of neuromyelitis optica in Caucasians // Neurology. 2011 May 3;76(18):1589-95. doi: 10.1212/WNL.0b013e3182190f74. PMID: 21536639; PMCID: PMC3269768

16. Wingerchuk D.M., Weinshenker B.G. Neuromyelitis optica (Devic's syndrome) // Handb Clin Neurol. 2014;122:581-99. doi: 10.1016/B978-0-444-52001-2.00025-X. PMID: 24507536

17. Wingerchuk D.M., Hogancamp W.F., O'Brien P.C., Weinshenker B.G. The clinical course of neuromyelitis optica (Devic's syndrome) // ***Neurology***. 1999;53(5):1107-1114. doi:10.1212/wnl.53.5.1107

18. Аутоиммунные заболевания в неврологии: Клиническое руководство / под ред. И.А. Завалишина и др. Т.1. Москва: Издательство РООИ «Здоровье человека», 2014. 392 с.

19. Виноградова Е.С., Новиков П.И., Моисеев С.В. Оптиконевромиелит (синдром Девика) - редкий вариант поражения нервной системы при системной красной волчанке // РМЖ. 2018;12(II):103-106

20. Lennon V.A., Wingerchuk D.M., Kryzer T.J. et al. A serum autoantibody marker of neuromyelitis optica: distinction from multiple sclerosis // Lancet. 2004;364(9451):2106-2112. doi:10.1016/S0140-6736(04)17551-X

21. Lennon V.A., Kryzer T.J., Pittock S.J., Verkman A.S., Hinson S.R. IgG marker of optic-spinal multiple sclerosis binds to the aquaporin-4 water channel // J Exp Med. 2005;202(4):473-477. doi:10.1084/jem.20050304

22. Giovannoni G. Neuromyelitis optics and anti-aquaporin-4 antibodies: widening the clinical phenotype // J Neurol Neurosurg Psychiatry. 2006;77(9):1001-2. doi: 10.1136/jnnp.2006.090944

23. Saadoun S., Waters P., Bell B.A., Vincent A., Verkman A.S., Papadopoulos M.C. Intra-cerebral injection of neuromyelitis optica

noglobulin G and human complement produces neuromyelitis optica lesions in mice. *Brain*. 2010;133(Pt 2):349-361. doi:10.1093/brain/awp309

24. Takahashi T, Fujihara K, Nakashima I, et al. Anti-aquaporin-4 antibody is involved in the pathogenesis of NMO: a study on antibody titre. *Brain*. 2007;130(Pt 5):1235-1243. doi:10.1093/brain/

25. Hinson SR, Pittock SJ, Lucchinetti CF, et al. Pathogenic potential of IgG binding to water channel extracellular domain in neuromyelitis optica. *Neurology*. 2007;69(24):2221-2231

26. Papadopoulos MC, Verkman AS. Aquaporin 4 and neuromyelitis optica. *Lancet Neurol*. 2012;11(6):535-544. doi:10.1016/S1474-4422(12)70133-3

27. Ratelade J, Zhang H, Saadoun S, Bennett JL, Papadopoulos MC, Verkman AS. Neuromyelitis optica IgG and natural killer cells produce NMO lesions in mice without myelin loss. *Acta Neuropathol*. 2012;123(6):861-872. doi:10.1007/s00401-012-0986-4

28. De Seze J, Storovic T, Ferriby D. Devic's neuromyelitis optica: clinical, laboratory MRI and outcome protile. *J Neurol Sci.* 2002;197(1-2):57–61. doi: 10.1016/s0022-510x(02)00043-6

29. Blanche P, Diaz E, Gombert B, et al. Devic's neuromyelitis optica and HIV-1 infection. *J Neurol Neurosurg Psychiatry.* 2000;68(6):795-796. doi: 10.1136/jnnp.68.6.795a

30. Weinshenker BG, Wingerchuk DM, Vukusic S. A secondary progressive clinical course is uncommon in neuromyelitisoptica. *Neurology.* 2007;68:603-605

31. Zhuravlev MN, Sivertseva SA, Molokova My, et al. Opticomyelitis or Devik's disease: pathogenesis, clinical picture, diagnosis and treatment experience with the use of betaferon and mitroxan. *Zhurnal nevrologii i psikhiatrii im. S.S. Korsakova = Journal of Neurology and Psychiatry. S.S. Korsakov.* 2007;4:106-112. (In Russ.)

32. Bezbabnaya OA, Golovan VN, Islamova NT, Kilina DA, Feoktistova NO. Damage to the optic nerve in multiple sclerosis and Devik's disease. *Vestnik soveta molodykh uchenykh i spetsialistov Cheliabinskoi oblasti = Vestnik of the Council of Young Scientists and Specialists of the Chelyabinsk Region*. 2016;3(14):7-13. (In Russ.)

33. Shakhov BE, Belova AN, Shalenkov IV, et al. Devik's opticomyelitis: diagnostic issues and case description from practice. *Meditsinskii Almanakh = Medical Almanac.* 2012;1:165-170. (In Russ.)

34. Martynova OA. Opticomyelitis (Devik's disease): Difficulties in differential diagnosis. *Vestnik soveta molodykh uchenykh i spetsialistov Cheliabinskoi oblasti = Vestnik of the Council of Young Scientists and Specialists of the Chelyabinsk Region*. 2016;3(14):49-53. (In Russ.)

35. Matiello M, Weinshenker B. Neuromyelitis optica. In: Lucchinetti CF, Hohlfeld R. Multiple sclerosis 3. *Saunders Elsevier.* 2010;258-275

immunoglobulin G and human complement produces neuromyelitis optica lesions in mice // Brain. 2010;133(Pt 2):349-361. doi:10.1093/brain/awp309

24. Takahashi T., Fujihara K., Nakashima I. et al. Anti-aquaporin-4 antibody is involved in the pathogenesis of NMO: a study on antibody titre // Brain. 2007;130(Pt 5):1235-1243. doi:10.1093/brain/

25. Hinson S.R., Pittock S.J., Lucchinetti C.F. et al. Pathogenic potential of IgG binding to water channel extracellular domain in neuromyelitis optica // Neurology. 2007;69(24):2221-2231

26. Papadopoulos M.C., Verkman A.S. Aquaporin 4 and neuromyelitis optica // Lancet Neurol. 2012;11(6):535-544. doi:10.1016/S1474-4422(12)70133-3

27. Ratelade J., Zhang H., Saadoun S., Bennett J.L., Papadopoulos M.C., Verkman A.. Neuromyelitis optica IgG and natural killer cells produce NMO lesions in mice without myelin loss // Acta Neuropathol. 2012;123(6):861-872. doi:10.1007/s00401-012-0986-4

28. De Seze J., Storovic T., Ferriby D. Devic's neuromyelitis optica: clinical, laboratory MRI and outcome protile // J Neurol Sci. 2002;197(1-2):57–61. doi: 10.1016/s0022-510x(02)00043-6

29. Blanche P., Diaz E., Gombert B. et al. Devic's neuromyelitis optica and HIV-1 infection // J Neurol Neurosurg Psychiatry. 2000;68(6):795-796. doi: 10.1136/jnnp.68.6.795a

30. Weinshenker B.G., Wingerchuk D.M., Vukusic S. A secondary progressive clinical course is uncommon in neuromyelitisoptica // Neurology. 2007;68:603-605

31. Журавлев М.Н., Сиверцева С.А., Молокова М.Ю. и др. Оптикомиелит или болезнь Девика: патогенез, клиника, диагностика и опыт лечения с использованием бетаферона и митроксана // Журнал неврологии и психиатрии им. С.С. Корсакова. 2007;4:106-112

32. Безбабная О.А., Головань В.Н., Исламова Н.Т., Килина Д.А., Феоктистова Н.О. Поражение зрительного нерва при рассеянном склерозе и болезни Девика // Вестник совета молодых учёных и специалистов Челябинской области. 2016;3(14):7-13

33. Шахов Б.Е., Белова А.Н., Шаленков И.В. и др. Оптикомиелит Девика: вопросы диагностики и описание случая из практики // Медицинский Альманах. 2012;1:165-170

34. Мартынова О.А. Оптикомиелит (болезнь Девика): сложности дифференциальной диагностики // Вестник совета молодых учёных и специалистов Челябинской области. 2016;3(14):49-53

35. Matiello M., Weinshenker B. Neuromyelitis optica. In: Lucchinetti C.F., Hohlfeld R. Multiple sclerosis 3. *Saunders Elsevier*. 2010;258-275