folia medica

Folia Medica 61(4): 639-42
DOI: 10.3897/folmed.61.e47947

Case Report

Neuromyelitis Optica and Systemic Lupus Erythematosus Association – the Chicken or the Egg Dilemma: a Case Report

Orlina Chaneva, Ekaterina Viteva, Anastasia Trenova, Georgi Slavov, Krasimir Shukerski, Maria Manova, Zahari Zahariev

Department of Neurology, Medical University of Plovdiv, Plovdiv Bulgaria

Corresponding author: Ekaterina Viteva, Department of Neurology, Medical University of Plovdiv, 66 Peshtersko shosse Blvd., 4000 Plovdiv, Bulgaria; E-mail: eiviteva@abv.bg; Tel.: 00359887752235

Received: 14 Feb 2019 ♦ **Accepted:** 24 Apr 2019 ♦ **Published:** 31 Dec 2019

Citation: Chaneva O, Viteva E, Trenova A, Slavov G, Shukerski K, Manova M, Zahariev Z. Neuromyelitis optica and systemic lupus erythematosus association – chicken or the egg dilemma: Case report. Folia Med (Plovdiv) 2019;61(4):639-42. doi: 10.3897/folmed.61.e47947.

Abstract

We present a case report of a 32-year-old woman diagnosed with opticomyelitis of Devic (OMD) and systemic lupus erythematosus (SLE). The onset of neurological symptoms was with optic neuritis. Five months later the neurological deficit progressed within a few days to lower paraplegia and upper paraparesis, retention of urine and faeces, impaired somatic and deep sensation below the level of Th_1 dermatome. The results from laboratory investigations confirmed anaemic syndrome, increased urea and creatinine, hypoproteinemia and severe proteinuria. The results from CSF investigations demonstrated hyperproteinorachia with extremely high Ig fractions. Serum and CSF oligoclonal bands and positive serum Aquaporin IgG 32 times higher than the upper referent limit were found. The association with SLE was confirmed by the increased levels of total ANA and anti-ds-DNA ANA. MRT visualized the spinal cord as non-homogenously hypointense on T_1 and extremely hyperintense on FLAIR sequences through its whole length up to the bulbar-pontine region. The MRT findings and the serum Aquaporin IgG confirmed the diagnosis OMD. The patient was treated with intravenous immunomodulating agents.

We consider the presented case of special interest because of the comorbidity of an aggressive autoimmune systemic and an organ-specific disease of the central nervous system.

Keywords

aquaporin IgG, opticomyelitis of Devic, optic neuritis, systemic lupus erythematosus, transverse myelitis

INTRODUCTION

Opticomyelitis of Devic/Neuromyelitis optica (NMO) is an inflammatory demyelinating disease of the central nervous system (CNS), which is associated with optic neuritis, transverse myelitis, involving at least three or more, continuous spinal cord segments, seropositivity for antibodies against aquaporin 4 of astrocytes. Systemic lupus erythematosus (SLE) is a multiorgan autoimmune disorder and its pathophysiology may affect all CNS components. The association of both diseases is rare, but not unusual.

CASE REPORT

We present a case of a 32-year-old woman without preceding diseases. The disease onset was in March 2016 with blurred vision. The patient was diagnosed with bilateral optic neuritis. Pyramidal signs occurred in August 2016: pathological hyperreflexia, clonus of both feet, abdominal areflexia. The Brain MRI with contrast matter showed no pathological findings. In November 2016 the patient was hospitalized in the Clinic of Neurology at St George University Hospital in Plovdiv, Bulgaria. Over the next several days the neurologi-

cal deficit progressed to quadriparesis with lower paraplegia, left severe and right latent upper paraparesis, distal hypesthesia below Th_1 dermatome and urine retention. The laboratory investigation showed the following pathological results: a moderate anemic syndrome; severe inflammatory activity, hypoproteinemia with proteinuria; total ANA diffusely /+/ in titer 1:160 and anti-DNA positive, rheumatoid factor in titer increased 10 times than the upper referent limit, positive anti-SS-A and anti-SS-B. ***Table 1*** *presents the results from the CSF investigation and serum anti-aquaporin 4 antibodies.*

The contrast MRI investigation visualized the spinal cord as a non-homogenously and diffusely hypointense on T_1, relatively hyperintense on T_2 and hyperintense on FLAIR sequences, up to the level of the bulbar-pontine transition ***(Fig. 1)****.*

In December 2016, after being diagnosed with SLE (based on the availability of five of the Systemic Lupus International Collaborating Clinics criteria – presence of proteinuria, neurological symptoms, hemolytic anemia, positive anti-DNA and positive ANA), the patient was treated with monthly pulse corticosteroid therapy.[1] In May 2017 the neurological examination showed: increased deep tendon reflexes; dystonic positions of the left hand fingers, lower severe paraparesis; preserved somatic sensation with impaired proprioreception;

Table 1. Results from CSF investigation and serum anti-aquaporin 4 antibodies

Indicator	Result	Deviation - times higher than the upper reference limit
Total protein	3.96 g/l	6.5
IgG	974.21 mg/l	32.5
IgM	17.2 mg/l	57
IgA	78.16 mg/l	26
Leucocytes	24 x 10^6/l	4
Neutrophils	32%	32
Serum oligoclonal bands	(+) positive	
CSF oligoclonal bands	(+) positive	
IgG (L)	4140 mg/dl	2.5
IgG (CSF)	142 mg/dl	35.5
IgG CSF/S ratio	34.30	7
Neuromyelitis optica IgG/ Aquapurin IgG ratio	1:320	32

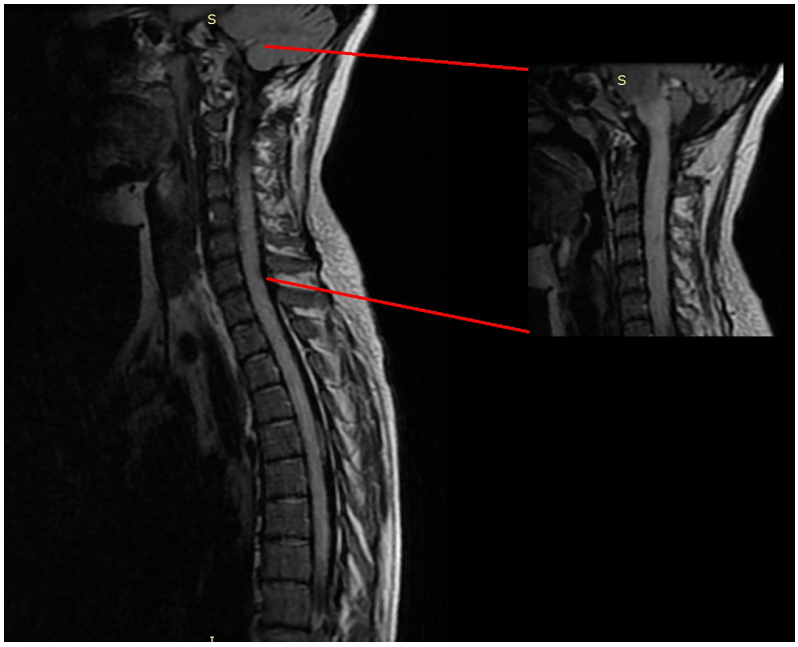

Figure 1. Sagittal plane MRT FLAIR image: diffuse hyperintensity of the spinal cord up to the level of bulbo-pontine transition (magnified image included).

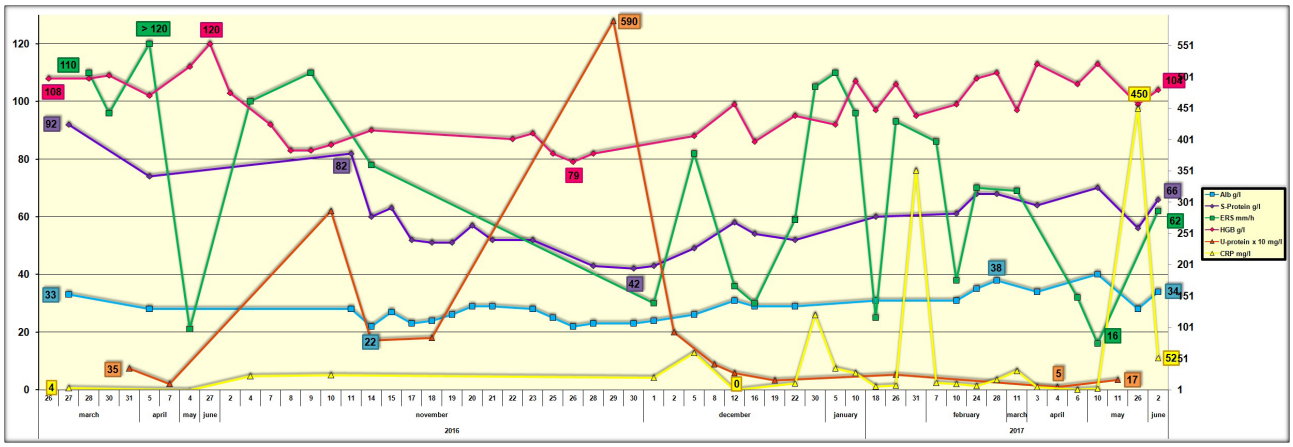

Figure 2. Dynamic assessment of serum albumin and total protein (S-Protein), urine protein (U-Protein), hemoglobin, ESR and CRP.

bilateral optic nerve atrophy; urine retention persistence. ***Fig. 2*** *presents the dynamic assessment of serum albumin and total protein (S-Protein), urine protein (U-Protein), hemoglobin, ESR and CRP.*

In the disease course, as a result of the treatment and neurological complications, various complications have been reported: chroniosepsis (Staphyl. coagulase (-) and E. cloacae); pyelonephritis/ urosepsis (C. glabrata, P. aeruginosa, E. cloacae, E. coli); infected decubitus ulcer (E. coli, A. baumannii); acute psychosis and drug-induced Cushing syndrome.

Specific medications were applied: pulse methylprednisolone 2500 mg i.v. for 5 days and prednisolone 120 mg p.o for 1.5 moth in March-April 2016; pulse therapy with methylprednisolone 8000 mg i.v. for 15 days, cyclophospfamide 400 mg i.v. for 5 days and pulse therapy with immunovenin 15 g i.v. for 3 days in November 2016; monthly pulse therapy with methylprednisolone 1500 mg i.v. for 3 days and cyclophosphamide 200 mg i.v. for 1 day, medrol a total monthly dose of 350 mg in December 2016 - May 2017, a monthly dose of azathioprine 1500 mg p.o. since April 2017.

DISCUSSION

On the one hand, in up to 30% of the cases, NMO is associated with another autoimmune disease, on the other hand, in 40% of patients with confirmed diagnosis of NMO, various antibodies may be positive, without the presence of another autoimmune disease.[2] The presence of antibodies typical for primary Sjogren syndrome – anti-SS-A and anti-SS-B in combination with negative anti-ds-ANA, typical for SLE, sets the question about the necessity of a differential diagnosis among the group of systemic diseases. A significant association of anti-SS-A and AQP4 in patients with NMO has been proven in the literature[3] and it is also present in the reported case. CSF oligoclonal bands have been confirmed in our patient – a typical finding for another organ specific autoimmune disorder – multiple sclerosis, therefore a differential diagnosis with autoimmune CNS diseases, is also required.

Only 2 cases with NMO preceding SLE have been described in literature by now.[4] Their clinical course, the therapeutic approach and prognosis are similar to those of our case, the latter however, is unique with the largeness of spinal cord impairment and the onset with optic neuritis.

CONCLUSION

We consider the presented clinical case of special interest because of the comorbidity of an aggressive autoimmune systemic and an organ-specific disease of the central nervous system. Based on the clinical and laboratory findings, as well as the disease course, this case puts a blurred line instead of a distinct border between autoimmune disorders, no matter what their systemic or specific characteristics are. The exact cause and effect relation between SLE and NNO remains unclear whether NMO is just a piece of a giant puzzle in SLE symptoms, or SLE is a result of an invasion of a CNS autoimmune disease to a far broader spectrum of manifestations. The common feature in both is autoimmunity, a disorder even far less understood.

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Связь между зрительным нейромиелитом и системной красной волчанкой - дилемма „яйца или курицы“: клинический случай

Орлина Чанева, Екатерина Витева, Анастасия Тренова, Георги Славов, Красимир Шукерски, Мария Манова, Захари Захариев

Кафедра неврологии, Медицинский университет - Пловдив, Пловдив, Болгария

Адрес для корреспонденции: Екатерина Витева, Кафедра неврологии, Медицинский университет - Пловдив, бул. „Пещерско шоссе” № 66, 4000 Пловдив, Болгария E-mail: eiviteva@abv.bg; Тел: 00359887752235

Дата получения: 14 февраля 2019 ♦ **Дата приемки:** 24 апреля 2019 ♦ **Дата публикации:** 31 декабря 2019

Образец цитирования: Chaneva O, Viteva E, Trenova A, Slavov G, Shukerski K, Manova M, Zahariev Z. Neuromyelitis optica and systemic lupus erythematosus association – the chicken or the egg dilemma: a case report. Folia Med (Plovdiv) 2019;61(4):639-42. doi: 10.3897/folmed.61.e47947.

Абстракт

Мы представляем случай 32-летней женщины, которой поставили диагноз: болезнь Девика - оптикомиелит (ОМД) и системная красная волчанка (СКВ). Начало неврологических симптомов было в форме неврита зрительного нерва. Спустя пять месяцев неврологический дефицит усиливается в течение нескольких дней с более низкой параплегией и верхним парапарезом, ретенцией мочи и кала, нарушенной соматической и глубокой чувствительностью ниже уровня дерматома Th1. Лабораторные данные подтвердили анемический синдром, увеличение мочевины и креатинина, гипопротеинемию и выраженную протеинурию. Результаты исследований CSF выявили гиперпротеинорragию с чрезвычайно высокими фракциями Ig. Сывороточные и олигоклональные полосы CSF и положительный уровень аквапоринового IgG в сыворотке были обнаружены в 32 раза выше верхнего контрольного предела. Ассоциация с СКВ была подтверждена повышенными уровнями общего ANA и анти-ds-ДНК ANA. При МРТ-исследовании спинной мозг был неоднородно гипоинтенсивным в T1 и чрезвычайно гиперинтенсивным на изображениях FLAIR на всём протяжении его расширения до бульбарно-понтинной области. Результаты МРТ и сывороточного аквапоринового IgG подтвердили диагноз ОМД. Пациентке вводили внутривенные иммуноглобулиновые препараты.

Мы считаем, что данный случай представляет особый интерес из-за сопутствующей патологии агрессивных аутоиммунных заболеваний, специфичных для органов центральной нервной системы.

Ключевые слова

оптикомиелит Девика, системная красная волчанка, аквапорин IgG, неврит зрительного нерва, поперечный миелит