Neuromyelitis optica (NMO, Devic’s disease) is a rare inflammatory, demyelinating disease of the central nervous system that predominantly affects the spinal cord and optic nerves [1]. The discovery of a specific NMO immunoglobulin started a new era in the classification and understanding of the pathogenesis of NMO [2]. NMO - IgG (AQP4 - Ab) binds to aquaporin 4 that is the main channel that regulates water homeostasis in the CNS. Diagnostic criteria for NMO with AQP4 - Ab are: at least one core clinical characteristic (optic neuritis, acute myelitis, acute brainstem syndrome, symptomatic narcolepsy or symptomatic cerebellar syndrome with typical NMO brain lesions), a positive AQP4 - Ab and exclusion of alternative diagnoses [3].

The incidence and prevalence of NMO are not well known. Studies from Europe, South East and Southern Asia, Caribbean and Cuba suggesting that the incidence and prevalence are from 0.05-0.4 to 0.5-4.4 per 100 000 respectively [4]. The proportion of NMO patients among non-Caucasians is much higher. NMO predominantly affect females with female to male ratio of 9:1 [5].

We describe a case of NMO presented with an acute onset of paraparesis.

Clinical Details. A 65 years old woman was admitted to our hospital due to the sudden onset of acute pain in thoracic part of the spine that was accompanied with leg weakness. Symptoms started three days before during her physical treatment for mild rheumatoid arthritis (RA). The weakness in legs constantly progressed during those three days so that at the admission she was unable to walk and she became incontinent. Besides mild RA she was previously treated for hypertension and hypothyroidism, so she was taking small doses of methylprednisolone, pantoprazole, levothyroxine, ramipril and bisoprolol. Examination at the day of admission revealed normal somatic status (no fever, regular heart beats and normal blood pressure), normal neurological function of all cranial nerves and upper extremities but her right leg was plegic, left was severely paretic, she had hypoesthesia below Th10, diminished deep tendon reflexes and bilateral Babinski sign. Laboratory evaluation showed elevated leukocytes (WBC; 21.4), neutrophils (86%) and C reactive protein (CRP; 262.2). In urine: RBC/Hgb 3+, nitrites +, leukocyte esterase 3+, in sediment: WBC 15-20, many RBC, many bacteria. Analysis of spinal tap showed 9/3 WBC, 6/3 RBC, proteins 0.59 g/l with normal glucose and lactate. Brain CT scan revealed few lacunar vascular lesions, MRI of her cervical spine was normal. The MRI (1.5T) of the thoracic spine reviled the hyperintense T2 lesion with edema from Th1-9, and post contrast ring enhancement at the level Th5-7. There was also the syrinx at the level of Th4-7. The radiologist concluded that those changes were due to the inflammatory process. The specialist for infective diseases was consulted; he prescribed antibiotics (gentamycin and clindamycin) and recommended hemocultures that come back positive for Staphylococcus aureus and her antibiotics were changed according to antibiogram to rimactane.

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and vancomycin. During her hospital stay she was constantly afebrile, her neurological condition was unchanged and it was decided that she should have a 3T MRI of her thoracic spine. The 3T MRI (Fig 1) reviled hyperintensive lesions at level Th1-5 and according to the radiologist those changes were in accordance with changes caused by spinal cord infarction in subacute fase. After 35 days of her hospital stay our patient had severe flaccid paraparesis - she was able to perform only minimal leg movements on the bed surface. Her deep tendon reflexes were still diminished, she had no Babinski sign, had hypoesthesia below Th6 and incontinency. The acetylsalicylic acid was added to her therapy and she was transferred to special hospital for medical rehabilitation. Three months later she was readmitted because of the strong sensation of the compression around her thorax, she felt dizzy, she was vomiting and her legs were again plegic. According to the laboratory results she had a urinary tract infection caused by E. coli and she was treated with antibiotics (nitrofurantoin). On the brain MRI there were multiple, small, non-specific demyelinating lesions (Fig.2). The new MRI of her

Figure 1a) Sagital MRI thoracic spinal cord showing swolen thoracic medula with syrinx and hypertensive signal from Th1-Th5. 1b) Axial T2 MRI thoracic spinal cord showing hypertensive lesion of more than a half of spinal cord transection.

Figure 2. Coronal T2 brain MRI showing multiple nonspecific hyperintensive white matter lesions
thoracic spine reviled regression of earlier lesion (Th1-7) but a new lesion from Th7-L1 has appeared. This time radiologist suggested the diagnosis of transverse myelitis. We tested her for the AQP4 - Ab and they were positive (1:320). She was treated with high doses of methylprednisolone, plasmapheresis and immunoglobulines but her condition stayed unchanged. At the discharge she was paraplegic and incontinent; azathioprine was added in her treatment.

Discussion. We presented this case because we wanted to point out the importance of thinking on NMO as differential diagnosis in every patient with longitudinally extensive myelitis. Especially these days when we can easily perform the confirmation test with AQP4 - Ab. We also wanted to point out the fact that though the neuromyelitis optica spectrum disorders and neuroimaging is of exquisite importance for managing neurological patients we must not forget that our diagnosis should always be an integration of history, clinical and neurological findings and neuroimaging. In this particular case we should have doubt the diagnosis of spinal cord infarction at the beginning. The presentation of spinal cord infarction is usually apoplectic - evolving over minutes. And if we look again at the MRI of her spinal cord (axial T2, Fig. 1b) we can see that the changes are not in concordance with any vascular supply territory - neither of anterior nor posterior spinal arteries.

It is known that the course of NMO is quite severe; common relapses and poor outcome are its characteristics [5,6]. Therapy of NMO should be initiated early. Azathioprine and rituximab are now suggested as first-line treatments and results are promising [7,8].

REFERENCES


XÜLASƏ

65 YAŞLI XORVİ QADINDA KƏSKİN YARANAN PARAPAREZ - KLİNİK HADİSƏ

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Optikomieliit (NMO, Devik xastalıyi) - mərkəzi sinir sisteminin nadir iltilahi demielinizasiyaedici xəstalıyi olub, așanson onurğa beynini və görmə sinirlərini zadalaşdır. NMO-IgG seropositivlik (akvaporin 4-ə qarşı antitelmələr) və onurğa beyninin boylama geniş zadağalərini (3 və daha artıq seqmentlər) NMO üçün xarakterdir. Taqdim edilmiş maqlədən koskin başlanğıç paraparezə 65 yaşlı qadın haqqda məlumat verilib, xastalıyan başlanğıçında NMO diaqnozu qoyulmuşdur. Diaqnoz iç açı ayrı, xəstə xəstəsədən gərək təkrar qəbul edildikdə qoyulmuşdur. Metilprednisolonun yüksak dozaları ilmüssiləyi, plazmaferes və immunoqlobulinlərlə müalicə baxmayaq, xastanın vaziyəti dayışmamışdır - paraplegiya və sidik saxlamazlığı qalmışdır. Açar sözlər: optikomieliit (NMO), Devik xastalıyi, klinik hadisə, müalicə
ОПТИКОМИЕЛИТ (NMO), БОЛЕЗНЬ ДЕВИКА - РЕДКОЕ ВОСПАЛИТЕЛЬНОЕ ДЕМИЕЛИНИЗИРУЮЩЕЕ ЗАБОЛЕВАНИЕ ЦЕНТРАЛЬНОЙ НЕРВНОЙ СИСТЕМЫ, КОТОРОЕ ПРЕИМУЩЕСТВЕННО ПОРАЖАЕТ СПИННОЙ МОЗГ И ЗРИТЕЛЬНЫЕ НЕРВЫ. СЕРОПОЗИТИВНОСТЬ ДЛЯ NMO-IgG (АНТИТЕЛА К АКВАПОРИНУ 4) И ПРОДОЛЬНЫЕ ОБШИРИНЫЕ ПОРАЖЕНИЯ СПИННОГО МОЗГА (3 ИЛИ БОЛЕЕ СЕГМЕНТОВ) ЯВЛЯЮТСЯ ХАРАКТЕРНЫМИ ДЛЯ NMO. МЫ ОПИСАЛИ 65-ЛЕТНЮЮ ЖЕНЩИНУ С ОСТРЫМ НАЧАЛОМ ПАРАПАРЕЗА, КОТОРАЯ ВНАЧАЛЕ НЕ БЫЛА ПРИЗНАНА КАК NMO. ДИАГНОЗ БЫЛ ПОСТАВЛЕН ТРИ МЕСЯЦА СПУСТЬ, КОГДА ОНА БЫЛА ПОВТОРНО ГОСПИТАЛИЗИРОВАНА ИЗ-ЗА РЕЦИДИВА. НЕСМОТРЯ НА ЛЕЧЕНИЕ ВЫСОКИМИ ДОЗАМИ МЕТИЛПРЕДНИЗОЛONA, ПЛАЗМАФЕРЕЗА И ИММУНОГЛОБУЛИНОВ, ЕЕ СОСТОЯНИЕ ОСТАВАЛОСЬ НЕИЗМЕННЫМ - У НЕЕ ОТМЕЧАЛАСЬ ПАРАПЛЕГИЯ И НЕДЕРЖАНИЕ МОЧИ.

**Ключевые слова:** оптикомиелит (NMO), болезнь Девика, клинический случай, лечение.