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A Rare Case Report: MOGAD with Optic Neuritis-without Spinal İnvolvement

Vaka Takdimi: Optik Nöritin Eşlik Ettiği Spinal Tutulumun Olmadığı Nadir Bir MOGAD Vakası

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Abstract

MOGAD (Myelin Oligodendrocyte Glycoprotein Antibody Disease) is caused by antibodies against the myelin oligodendrocyte glycoprotein (MOG) found on the surfaces of oligodendrocytes and its damage. MOGAD and Neuromyelitis Optica Spectrum Disorders (NMOSD) are very rare autoimmune inflammatory demyelinating diseases of the central nervous system that are often seen with joint involvement of the optic nerves and spinal cord. MOGAD can be confused with Multiple Sclerosis (MS) or NMOSD, which are more common in the community, due to its clinical presentations that may be similar and its characteristic to progress with habitual attacks. Although the clinical course of these three diseases is similar, their diagnosis and treatment are different; It is important to avoid diagnostic confusion, to make differential diagnosis of patients with other diseases and not to waste time for treatment, to prevent possible disability and disability. In addition, Optic Neuritis is an inflammatory disease of the optic nerve and is very common in patients with Multiple Sclerosis, and it can often be the first clinical presentation of the disease. However, being the first and early finding in other very rare demyelinating diseases can cause diagnostic confusion. In this article, we wanted to present the importance of optic neuritis, demyelinating diseases and differential diagnosis in a İnformed consent of the patient was obtained, 51-years-old female patient who had bilateral optic neuritis attacks at different times, in the light of the literature.

Keywords: Optic neuritis; multiple sclerosis; myelin-oligodendrocyte glycoprotein; neuromyelitis optica.

Özet

MOGAD (Myelin Oligodendrocyte Glycoprotein Antibody Disease), oligodendrositlerin yüzeylerinde bulunan miyelin oligodendrosit glikoproteinine (MOG) karşı oluşan antikor ve onun verdiği hasardan kaynaklanır. MOGAD ve NöroMiyelitis Optica Spektrum Disorders (NMOSD), sıklıkla optik sinirler ve omuriliğin birlikte tutulumu ile görülen merkezi sinir sisteminin çok nadir otoimmün inflamatuar demiyelinizan hastalıklarıdır. MOGAD, benzer olabilecek klinik tabloları ve alışılmış ataklarla seyretme özelliğinden dolayı toplumda daha sık görülen Multipl Skleroz (MS) veya NMOSD ile karıştırılabilir. Bu üç hastalığın her ne kadar klinik seyirleri benzer olsa da tanı ve tedavileri farklı olduğundan; tanı karışıklığı oluşmaması, hastaların diğer hastalıklarla ayırıcı tanısının yapılması ve tedavi için zaman kaybetmemesi, oluşabilecek disabilite ve engelliliğin önlenmesi açısından önemlidir. Ayrıca, Optik Nörit, optik sinirin inflamatuar hastalığıdır ve Multiple Skleroz hastalarında çok sık görülmekte, çoğu zaman da hastalığın ilk klinik prezentasyonu olabilmektedir. Ancak çok nadir görülen diğer demiyelinizan hastalıklarda ilk ve erken bulgu olması tanı karışıklığı oluşturabilmektedir. Biz bu yazımızda, aydınlatılmış hasta onamı alınan, farklı zamanlarda bilateral optik nörit atağı geçiren, 51 yaş bayan hasta özelinde optik nörit, demiyelinizan hastalıklar ve ayırıcı tanının önemini literatür eşliğinde sunmak istedik.

Anahtar Kelimeler: Optic neuritis; multiple sclerosis; myelin-oligodendrocyte glycoprotein; neuromyelitis optica.

Introduction

Optic Neuritis is a common symptom in Neurology and Ophthalmology clinics. Sometimes it can be found alone; sometimes it is part of another underlying disease. When evaluated alone and other underlying diseases are neglected, patients cannot be diagnosed and treated; this causes the formation of permanent sequelae. In addition, when very rare diseases are not considered for the diagnosis of another factor, sometimes they may be mistakenly diagnosed with Multiple Sclerosis (MS). This results in patients not being treated. We wanted to draw attention to this issue in this very rare case that we will consider.

Case

Informed written consent from the patient was obtained. 51-years-old female patient came to the neurology clinic with two optic neuritis attacks, which one is in the left eye 1.5 years ago, the other

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one is in the right eye throughtout 2 weeks. She has experienced severe bilateral vision loss. She has had sequelae from vision loss 1.5 years ago. Vision loss has developed in the other eye for 2 weeks. Other neurological examination were found as a normal except for vision loss. The patient was not diagnosed with any kind of diagnosis and she did not receive any treatment for first attack when is 1.5 years ago. Contrast-enhanced and diffusion weighted cranial MRI is observed nodular-patchy, nonspecific T2 hyperintense signal changes in bilateral periventricular deep white matter and centrum semiovale. Long segment diffusion restriction and enhancement is observed in the optic nerve and perineural enhancement left eye and optic atrophy in the right eye. Spinal imaging of the patient is within normal limits. Cerebrospinal fluid tests are found to be normal. Oligoclonal band is not detected. Anti NMO (AQP4-IgG) was negative, while Anti Mog was positive (1/100) in the blood analysis. In the VEP (Visual Evoked Potentials) examination is performed by giving patterns and flash stimulus to both eyes separately; visual evoked potential responses with pattern stimulus with delayed latency on the right, normal amplitude; obtained with normal latency and low amplitude on the left; On the other hand, with flash stimulus, it is obtained with a slightly delayed latency and normal amplitude at the upper limit of bilateral normal. When given Intravenous methylprednisolone pulse therapy (1 g/day for 5 consecutive days; IVMP), the therapy is effected positive way and significantly decreased symptoms. After IVMP therapy is ended, she has had no relapses with subsequent oral prednisolone therapy (20 mg/day) for over 8 weeks. With 100 mg / day azathioprine treatment, she did not have any additional complaints for 2 weeks.

Discussion

Unilateral acute optic neuritis can presented primarly demyelinating central nervous system disease connected to such as MS, NMOSD (NeuroMyelitis Optica Spectrum Disorder) or Myelin Oligodendrocyte Glycoprotein Antibody Disease (MOGAD). It can sometimes be very difficult to distinguish these diseases from each other. However, important point of that the treatments for these diseases are seperated. Consequently, patients who are suffered with optic neuritis, a differential diagnosis should be made quickly as this symptom may result in blindness (1-4). The number of lesions in MRI is more than 3, the size of the lesions is larger than 6 mm, oval shaped lesions with long axes extending perpendicular to the lateral ventricles, and their localization in the periventricular and corpus callosum are typical for MS (2,4,5).

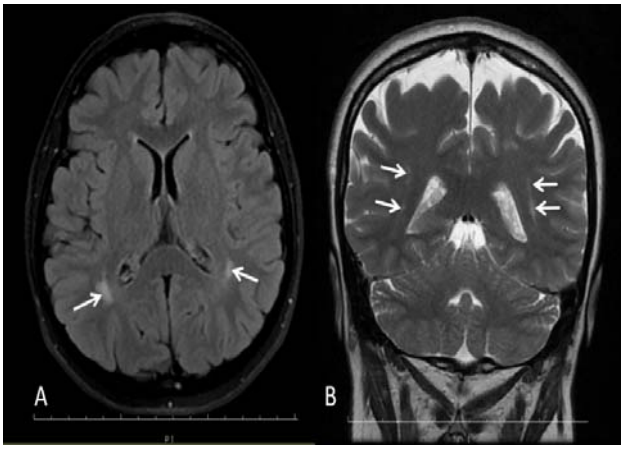

Figure 1. Axial FLAIR (A) and coronal T2 (B) images show areas of T2 signal increase in the periventricular deep white matter with a patchy confluence, marked by thin white arrows. In addition, millimetric nodular T2 signal increase areas were observed in the centrum semiovale, which were not found in the images, and T2 hyperintense signal with callososeptal, juxtacortical or infratentorial location typical for MS was not observed.

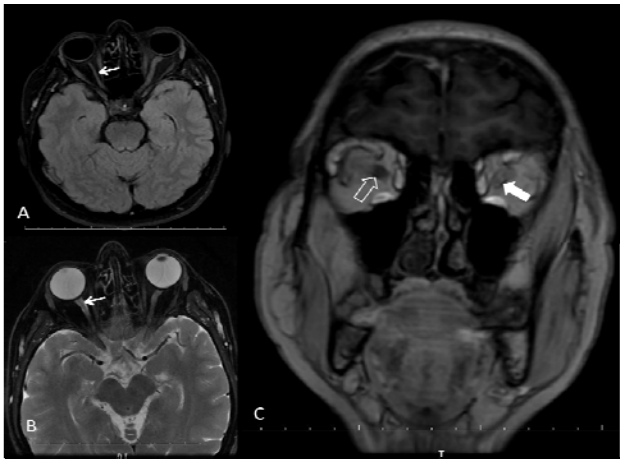

Figure 2. Atrophy of the right optic nerve marked with a thin white arrow in the axial FLAIR (A) and axial T2W (B) images, and enlargement of the peripheral CSF sheath in the T2W image. Left optic nerve volume was normal, and significant T2 signal changes could not be distinguished. In post-contrast coronal T1W images (C), contrast enhancement is not observed in the right optic nerve marked with a light white arrow, while long segment enhancement is observed at the level of the CSF sheath in the left optic nerve marked with a thick white arrow and its periphery, which can be seen in MOGAD.

Notwithstanding, when contrast-enhanced cranial MRI of patient is examined, T2-hyperintense lesion, bilateral, especially on the right, periventricular deep white matter area impairments are found (figüre 1). The volume of the right optic nerve is decreased compared to the

left (figüre 2A), and prominent subarachnoid space around the optic nerves (figüre 2B).

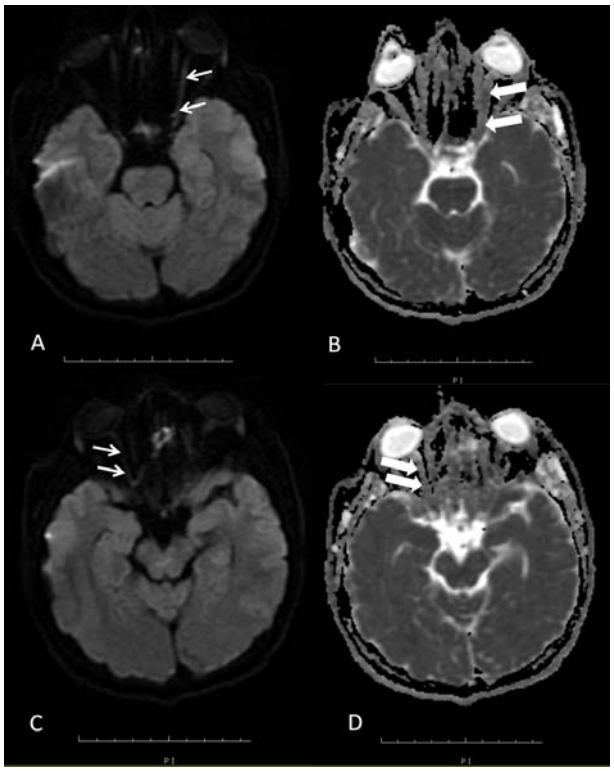

Figure 3. In the diffusion-weighted images taken by giving the axial B1000 value, the left (A) and right (C) optic nerves are marked with white arrows, respectively, and a long-segmented signal increase in diffusion in the left optic nerve is observed. On the ADC map, the optic nerves are marked with thick white arrows, and diffusion restriction is observed in the left optic nerve (B), respectively, while diffusion restriction is not observed in the right optic nerve (D).

Although no significant volume and T2 signal increase were observed in the left optic nerve, long segment diffusion restriction (figüre 3) and contrast enhancement is observed, more prominently in the minimal perineural area after IVCM(IntraVenous Contrast Matter) (figure 2C). Contrast uptake was not detected in other lesions. Distinct imaging patterns may also be seen in optic nerve involvement. Bilateral and preferable anterior optic nerve involvement is commonly described in MOG-IgG–positive patients. Perineural gadolinium enhancement on postcontrast T1-weighted images can be a distinctive imaging feature in these patients(6).AQP4-IgG positive patients with long segment, bilateral and posterior optic nerve involvement with chiasmatic extension. MOG-IgG positive patients usually exhibit long segment, bilateral and anterior optic nerve involvement, with intraorbital optic nerve swelling and usually with perineural gadolinium enhancement. Multiple sclerosis patients classically have unilateral and short segment optic neuritis (Neuromyelitis Optica Spectrum Disorders: Spectrum of MR Imaging Findings and Their Differential Diagnosis). Thus, It is thought that patient may be diagnoses MS variant such as MOGAD or NMOSD rather than MS (2,4,5). Medulla spinalis involvement is seen with a certain frequency in demyelinating diseases of the central nervous system, which are included in MS and its differential diagnosis. The rates of medulla spinalis involvement, especially in MOGAD and NMOSD diseases, have not been determined yet. Interestingly, although medulla spinalis involvement was expected, any involvement of the spinal cord segment was observed considering the age of the patient. At the another point of this, MS is frequently diagnosed in women who are 18-35 years old with only signs of optic neuritis. If optic neuritis is seen in a patient older than 35 years, other demyelinating diseases of the CNS should be considered. Therefore, in this case, as the patient was over 50 years old, it was thought that there might be an MS variant, and diagnostic tests were requested for them. In such cases, the differential diagnosis of NMOSD and MOGAD should be kept in mind (2,4,5). MOG antibody disease, MOGAD or Anti-MOG associated encephalomyelitis is an inflammatory demyelinating disease of the central nervous system. Serum anti-myelin oligodendrocyte glycoprotein antibodies are present in up to half of patients with an acquired demyelinating syndrome (7-9). MOG-IgG is detected by means of so-called cell-based assays. MOG antibodies are the gold standard for anti-MOG antibody testing. Serum is the specimen of choice; cerebrospinal fluid (CSF) analysis is less sensitive compared to serum testing (9-13). İmmunosuppressive therapy (corticosteroids, azathioprine, tacrolimus, mycophenolate mofetil, methotrexate) is the maintaining for treatment to prevent recurrence in MOGAD. In accordance with the literature, immunotherapy was also applied in this case (2,4-13).

Conclusion

We wanted to share this rare case because these patients are often misdiagnosed with MS and mistreated and emphasize the importance of differential diagnosis and early diagnosis of these diseases in the light of the literature.

Conflicts of Interests: The authors declare that they have no conflict of interest.

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References

1. Reindl M, Di Pauli F, Rostásy K, Berger T. The spectrum of MOG autoantibody-associated demyelinating diseases. Nat Rev Neurol 2013; 9(8): 455-461.
2. Eraksoy M, G Akman-Demir. Merkezi Sinir Sisteminin Miyelin Hastalıkları. İçinde: Öge AE, B Baykan, Editörler. İstanbul Tıp Fakültesi Temel ve Klinik Bilimler Ders Kitapları. Nöroloji. 2. Baskı. İstanbul: Nobel Yayınları; 2015. s. 603-648
3. Narayan R, Simpson A, Fritsche K, Salama S, Pardo S, Mealy M, Levy M. MOG antibody disease: a review of MOG antibody seropositive neuromyelitis optica spectrum disorder. Mult scler relat disord 2018; 25: 66-72.
4. Ungureanu A, Seze J, Ahle G. Myelin oligodendrocyte glycoprotein antibodies in neuromyelitis optica spectrum disorder. Rev Neurol 2018; 174(10): 675-679.
5. Louapre C, Papeix C, Lubetzki C. Multiple sclerosis and aging. Geriatr Psychol Neuropsychiatr Vieil 2017; 15(4): 402-408.
6. Dutra BG, da Rocha AJ, Nunes RH, Maia ACM. Neuromyelitis Optica Spectrum Disorders: Spectrum of MR Imaging Findings and Their Differential Diagnosis. RadioGraphics 2018; 38(1): 169-193.
7. Tajfirouz DA, Bhatti MT, Chen JJ. Clinical characteristics and treatment of MOG-IgG–associated optic neuritis. Curr neurol neurosci rep 2019; 19(12): 1-8
8. Ramanathan S, Dale R C, Brilot F. Anti-MOG antibody: the history, clinical phenotype, and pathogenicity of a serum biomarker for demyelination. Autoimmun Rev 2016; 15(4): 307-324.
9. Jarius S, Paul F, Aktas O, Asgari N, Dale R C, De Seze J, et al. MOG encephalomyelitis: international recommendations on diagnosis and antibody testing. J neuroinflammation 2018; 15(1): 1-10.
10. Jarius S, Ruprecht K, Kleiter I, Borisow N, Asgari N, Pitarokoili K, et al. MOG-IgG in NMO and related disorders: a multicenter study of 50 patients. Part 1: frequency, syndrome specificity, influence of disease activity, long-term course, association with AQP4-IgG, and origin. J Neuroinflammation 2016; 13(1): 1-16.
11. Nakashima I. Anti□myelin oligodendrocyte glycoprotein antibody in demyelinating diseases. Clin Exp Neuroimmunol 2015; 6: 59-63.
12. Shahriari M, Sotirchos E S, Newsome S D, Yousem D M. MOGAD: How It Differs From and Resembles Other Neuroinflammatory Disorders. Am J Roentgenol 2021; 216(4): 1031-1039.
13. Takahashi K, Takeuchi H, Fukai R, Nakamura H, Morihara K, Higashiyama Y, et al. Case Report: Anti-MOG Antibody Seroconversion Accompanied by Dimethyl Fumarate Treatment. Front Immunol 2021; 12: 347.