

Case Report

Neuromyelitis Optica with a Good Response After Low Doses of Corticosteroid Therapy: A Case Report

Zylfije Hundozi¹, Granit Xhiha^{1*}, Jehona Rrustemi¹, Bujar Gjilkolli² and Fisnik Jashari²

¹Department of Neurology, University Clinical Center of Kosovo, Prishtina, Kosovo, 10000, Albania

²Department of Radiology, University Clinical Center of Kosovo, Prishtina, Kosovo, 10000, Albania

*Corresponding author: Granit Xhiha, Department of Neurology, University Clinical Center of Kosovo, Prishtina, Kosovo, 10000, Albania, Tel: +37744385682; E-mail: xgranit@hotmail.com

Received April 13, 2017; Accepted May 19, 2017; Published May 22, 2017

Abstract

Neuromyelitis optica (NMO), also known as Devic disease is heterogeneous condition consisting of simultaneous inflammation and demyelination distinguished by the combination of Optic Neuritis and myelitis.

We report a case of a 54-year-old women, with vision loss and pain on the right eye worsened with eye movement, headache, vertigo, mono paresis of right hand, sphincter dysfunction. Patient met the criteria for diagnosis of NMO by having transverse myelitis, optic neuritis, confirmed by MRI imaging and high level of aquaporin-4-antibodies (AQP4-Ab). Symptoms improved after treatment with low doses of corticosteroids.

Keywords: Neuro myelitis optica; Transverse myelitis; Aquaporin-4 antibodies; Corticosteroid therapy

Introduction

Neuromyelitis optica, also known as Devic disease or Devic's syndrome is an immune mediated chronic inflammatory disease of central nervous system. Clinically, it presents with optic neuritis (ON) and myelitis, often characterized by poor or no recovery [1]. It can be monophasic or recurrent. The prevalence and incidence of Devic's disease has not been established, because it is underdiagnosed and also confused with sclerosis multiplex. Its prevalence is estimated to range from less than 1 to 4.4/100.000 in western World [2]. More women than men have NMO (ratio 9:1, compared to Sclerosis multiplex which is 2:1) [3]. Specific etiological causes of NO have not been identified, although connective tissue disorders, tuberculosis, and acute disseminated encephalomyelitis have been associated with it. Although originally considered as variation of multiple sclerosis, clinical radiological pathological and especially immunological data have led to a novel definition of this clinical entity [4]. It is currently considered autoimmune disease (autoimmune astrocytopathy or autoimmune astrocytic channelopathy) based on presence of autoantibodies against AQP4 [5]. NMO-IgG seropositive patients with a history of optic neuritis or transverse myelitis who do not fulfill all of the clinical criteria are classified as having NMO spectrum disorder but are clinically treated identically as definite NMO. The clinical hallmarks of NMO are acute neuritis that is often bilateral and transverse myelitis that is often longitudinally extensive. Commonly reported symptoms include unilateral and bilateral loss of visual acuity, ocular pain, and severe paraplegia, a symmetric sensory level, bladder dysfunction, and paroxysmal tonic spasm of the trunk and limbs, and Lhermitte's phenomenon [6].

Magnetic resonance imaging (MRI) reveals involvement of more than three vertebral segments, which is synonymous with longitudinally extensive transverse myelitis (LETM) [7-9].

Case Presentation

A 54 years old women was hospitalized in the department of Neurology with loss of vision and pain on the right eye, progressive weakness and numbness on her right hand, headache, vertigo, vomiting, and bladder dysfunction. She had anxiety accompanied by

poor endurance. Complaints started two days before admission in Neurologic department. This was second hospitalization in department of Neurology. First time she had left side hemiparesis. Family history was unremarkable. During Neurological assessment muscle power grade 3/4 on the Medical Research Council (MRC) scale on right hand with painful spasticity and brisk deep tendon reflexes. Sensory deficit to fine touch and pressure on the right hand. An afferent pupillary defect on right eye, and eye pain worsened with eye movement. She had urinary incontinence and urinary catheter was placed *in situ*.

The full blood count showed the following: WBC: $10.2 \cdot 10^3 / \text{mm}^3$, RBC: $4.92 \cdot 10^3 / \text{mm}^3$, lymphocytes: 24, 1%, Monocytes: 5, 9%, Granulocytes: 70%, HGB: 13.1 g/dl, HTC: 41.0%, PLT: $356 \cdot 10^3 / \text{mm}^3$, PCT: .230%. It was detected a high level of autoantibodies directed to AQP4 qualitative ++ and quantitative (RIA): 2, 4 nM (normal values ranged 0.0 to 0.4 nM). A lumbar puncture was performed and showing normal glucose and protein level lactate level, with WBC count of 5 cells/uL, IgG, IgA and IgM CSF/serum ratios were normal. The presence of oligoclonal bands in liquor was negative. Brain MRI revealed hypertrophy of right optic nerve (Figure 1) and cervical spine MRI revealed multiple hyper intense signals involving cervical cord levels C1, C2, C3, C4 and C6 on T2-weighted images (Figure 2).

Since in the first few days after admission the diagnosis of NMO was not yet confirmed, the treatment was started with low-dose intravenous methylprednisolone of only 120 mg daily for 5 days



Figure 1: Axial T2-weighted MRI showing a mild hypertrophy of right optic nerve. Asymmetry in size of optic nerves (left optic nerve 3-4 mm and right optic nerve 5-6 mm).

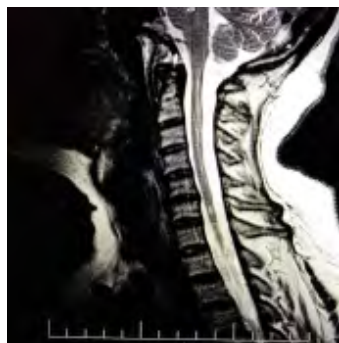


Figure 2: Sagittal T2-weighted MRI showing hyper intense lesions involving the medulla oblongata and involvement of cervical spinal cord (C1-C5).

and was followed by an oral prednisone taper 60 mg every other day together with azathioprine 2.5 mg/kg/day. She had a good response with her vision improved after second day of therapy administration.

Discussion

Neuromyelitis optica (NMO) is severe idiopathic autoimmune disorder of the central nervous system. For reasons not completely understood, NMO preferentially affects the optic nerves and spinal cord causing relapses of optic neuritis and transverse myelitis associated with a long lesion in the spinal cord, typically extending three or more spinal segments (longitudinally extensive transverse myelitis). NMO was considered as variant of multiple sclerosis, but now is regarded as a distinct disease entity since Aquaporin-4 antibodies were first described in 2004.

Our case presentation shows a detailed medical history, physical examination, laboratory investigations; brain MRI, cervical and thoracic spine MRI together with serologic evaluation. All findings fulfill criteria for diagnosis of NMO: optic neuritis, acute myelitis, NMO-IgG seropositive status, contiguous spinal cord MRI lesions extending over ≥ 3 segments in the context of an acute myelitis attack. Our focus was on not only on primary symptoms but also on symptoms that could suggest alternative diagnosis or concomitant autoimmune disorders which are frequently present in patients with AQP-4-Ab positive NMO but finally they were excluded. There are many case reports in literature of NMO associated with another autoimmune condition such as autoimmune thyroiditis, myasthenia gravis and Sjogren syndromes [10-12].

Following an attack of NMO, high-dose IV Methylprednisolone (1 g for 5 days) is considered standard treatment and should be initiated as soon as it is clear that a patient is having an attack [8,13]. Long-term immunosuppressive treatment should be initiated once the diagnosis of NMO has been confirmed to prevent relapsing course in most cases [11]. Azathioprine and Rituximab are currently the most widely used first line therapies in NMO [1].

In our case reported here, low-dose of corticosteroids was started on the second day after admission. She was treated with Methylprednisolone 120 mg IV for 5 days and was followed by an oral prednisone taper every other day. For long term relapse prevention, we have started Azathioprine 2.5 mg/kg/day. She had a good response after second day of therapy administration, with her vision improved and also pain in the eye diminished. To our knowledge, it is only one

case of NMO reported in the literature, which showed improvement after low corticosteroid doses [14-16].

Conclusion

NMO is diagnostic challenge in every-day clinical neurologic practice because of similarity with multiple sclerosis. Early differentiation between NMO and MS is particularly important because of course of NMO is more severe and treatment strategies for attack preventions are different. Disease modifying therapies for NMO have not been rigorously studied; we have reported a case with good response after treatment with low doses of corticosteroid therapy.

Conflict of interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

References

1. Trebst C, Jarius S, Berthele A, Paul F, Schippling S, et al. (2014) Update on the diagnosis and treatment of neuromyelitis optica: recommendations of the Neuromyelitis Optica Study Group (NEMOS). *J Neurol* 261: 1-16.
2. Jacob A, Panicker J, Lythgoe D, Elson L, Mutch K, et al. (2013). The epidemiology of neuromyelitisoptica amongst adults in the Merseyside county of United Kingdom. *J Neurol* 260: 2134-2137.
3. Wingerchuk DM (2009) Neuromyelitisoptica: effect of gender. *J Neurol Sci* 286: 18-23.
4. Beiruti K, Saleh AS, Daitzman M, Shahien R (2016) Neuromyelitis Optica Spectrum disorders: A Case report. *J Mult Scler* 3: 192.
5. Lucchinetti CF, Guo Y, Popescu BFG, Fujihara K, Itoyama Y, et al. (2014) The pathology of an autoimmune astrocytopathy: lessons learned from neuromyelitis optica. *Brain Pathol* 24: 83-97.
6. Kimbrough DJ, Fujihara K, Jacob A, Lana-Peixoto MA, Leite MI, et al. (2012) Treatment of neuromyelitis optica: review and recommendations. *Mult Scler Relat Disord* 1: 180-187.
7. Roy U, Saini DS, Pan K, Pandit A, Ganguly G, et al. (2016) Neuromyelitis optica spectrum disorder with tumefactive demyelination mimicking multiple sclerosis: a rare case. *Front Neurol* 7.
8. Matiello M, Weinschenker BG (2014) Multiple Sclerosis and CNS inflammatory Disorders. John Wiley & sons.
9. Jarius S, Paul F, Franciotta D, de Seze J Munch C, Salvetti M, et al. (2012) Neuromyelitis spectrum disorders in patients with myasthenia gravis: ten new aquaporin -4 antibody positive cases and review of the literature. *Mult Scler* 18: 1135-43.
10. Sudalangunta Rao S, Sodalanguta Babu M, Khorram H, Gonivada J, Noroozpour Z, et al. (2015) Autoimmune thyroiditis associated with neuromyelitisoptica (NMO). *Ger Med Sci* 13: 22.
11. Carvalho DC, Tironi TS, Freitas DS, Kleinpaul R, Talim NC, et al. (2014) Sjogren syndrome and neuromyelitisoptica spectrum disorders coexist in a common autoimmune milieu. *Arq Neuropsiquatr* 72: 619-24.
12. Wingerchuk DM, Weinschenker BG (2008) Neuromyelitis optica. *Cur Treat Options Neurol* 10: 55-66.
13. Jarius S, Aboul-Enein F, Waters P, Hauser A, Berger T, et al. (2008) Antibody to aquaporin- 4 in the long-term course of neuromyelitisoptica. *Brain* 131: 3072-3080.
14. Costanzi C, Matiello M, Lucchinetti CF, Weinschenker BG, Pittock SJ, et al. (2011) Azathioprine tolerability, efficacy, and predictors of benefit in neuromyelitis optica. *Neurology* 77: 659-666.
15. Bichuetti DB, de Oliveira EML, Oliveira DM, de Souza NA, Gabbai AA (2010) Neuromyelitis optica treatment: analysis of 36 patients. *Arch Neurol* 67: 1131-1136.
16. Mitsuyama T, Yoneyama T, Suzuki S, Kawamata T (2012) Neuromyelitisoptica spectrum disorder: a case report. *No Shinkei Geka* 40: 337-42.