Two Cases of Conus Medullaris Lesions with Neuromyelitis Optica Spectrum Disorders

Yanqiang Wang, Yuge Zhang, Yilong Shan and Zhengqi Lu

Department of Neurology, Hospital of Sun Yat-sen University, Guangzhou, China

Abstract

Neuromyelitis optica spectrum disorders (NMOSD), an autoimmune astrocytopathic disease associated with anti-aquaporin-4 (AQP4) antibody, is characterized by inflammatory lesions in the optic nerves, spinal cord, and central parts of the brain. These symptoms, urinary and fecal incontinence about conus medullaris involvement in NMOSD, are usually not appreciated as the possible manifestation of NMOSD. Missing diagnosis at this early stage will lead to a delay in the treatment, and hence, irreversible complications. We present two cases of conus medullaris involvements with neuromyelitis optica spectrum disorders.

Keywords: Neuromyelitis optica spectrum disorders; Conus medullaris; Anti-aquaporin-4 antibody (AQP4-Ab); Multiple sclerosis (MS)

Introduction

Neuromyelitis optica spectrum disorders (NMOSD) is a group of inflammatory and demyelinating syndrome of the central nervous system. With the growing recognition of the pathophysiological mechanisms of NMO-IgG/Antibodies to aquaporin-4 (AQP4-Ab), numerous studies have consistently shown that NMOSD lesions tend to be localized to the site of AQP4 expression, such as spinal cord, white matter, hypothalamus, thalamus, third or fourth periventricular, midbrain, medulla oblongata etc. Among the spinal cord involvement, the most common locations were the cervical or thoracic spinal cord. However, there are few reports on the conus medullaris lesions in NMOSD. Here we report two anti-AQP-4 anti-body-positive NMOSD patients with conus abnormalities on Magnetic Resonance Imaging (MRI).

Cases

Case 1

A 25-year-old Chinese woman, with no previous history of optic neuritis, myelitis or comorbidities, complained of six months of repeated nausea, vomiting, and progressive numbness in the lower extremity. Meanwhile progressive visual loss initially only affecting the right eye but involving the left before four months. During the disease onset, developed loss of consciousness, dysuria, and incontinence. On admission, general physical examination results were unremarkable. Neurologically, Ophthalmoscopy showed bilateral optic atrophy, her best-corrected visual acuity (BCVA) was 0.3 in her right eye and 0.2 in her left eye. She present with marked hypotonia, generalized muscle weakness (grade 4/5 proximally of the proximal and distal in the upper and lower extremity limbs). And had hypoesthesia below the C3 vertebra, absent joint position and vibration sensations. The EDSS score was 4.0 (out of 10). An abdominal X-ray was normal. Cerebrospinal fluid (CSF) examination showed 5*10^6 white cells, glucose level of 2.81 mmol/L (with a corresponding serum glucose level of 5.2 mmol/L) and protein level of 0.32 g/L (normal <0.4 g/L), no oligoclonal bands. Brain MRI showed bilateral frontal lesions, without gadolium enhancement; spinal magnetic resonance imaging (MRI) showed highsignal lesions over C2-3, C6-8 and L1-3 on T2-weighted images. Serological investigations revealed NMO-IgG/AQP-4 antibody. The patient presented seropositivity for anti-SSA and anti-SSB. Antibodies to other immunological antibodies were undetectable.

Discussion

Conus medullaris is the cone-shaped terminal end of the spinal cord that is usually located between the 12th thoracic (T12) vertebra and the 3rd lumbar (L3) vertebra. Although, Spinal cord lesions is one of the predominant characteristics in NMOSD. Few reports describe the conus medullaris lesions in neuromyelitis optica spectrum disorders (NMOSD). Takai Y et al reported the lesions in the conus medullaris were 1.58% in NMOSD, and 9.6% in anti-AQP4 antibody positive cases [6-8], Kitley J et al reported the myelin oligodendrocyte glycoprotein antibody-positive (MOG-Abs) and AQP4-Ab-positive NMO/NMOSD have conus involvement on spinal magnetic resonance imaging 75% or 17%, respectively, Joanna Kitley et al reported the

Case 2

A 47-year-old woman presented with unrelenting abdominal cramps and lower extremity numbness. Since 3 months before admission, she had experienced abdominal pain with belt-shaped numbness, recurrently. And suffered from severe left-sided to right-sided. There was no family history of autoimmune disease. 2 months later, developed sensory impairment followed by weakness of bilateral lower limb leading to difficulty of her gait. Accompanied with urinary and fecal incontinence. The major abnormalities of a physical examination were a distended, diffusely tender and painful abdomen and periumbilical tenderness. And had hypoesthesia below the C6 vertebra, absent joint position and vibration sensations. The EDSS score was 4.0 (out of 10). An abdominal X-ray was normal. Cerebrospinal fluid (CSF) examination showed 5*10^6 white cells, glucose level of 2.81 mmol/L (with a corresponding serum glucose level of 5.2 mmol/L) and protein level of 0.32 g/L (normal <0.4 g/L), no oligoclonal bands. Brain MRI showed bilateral frontal lesions, without gadolium enhancement; spinal magnetic resonance imaging (MRI) showed highsignal lesions over C2-3, C6-8 and L1-3 on T2-weighted images. Serological investigations revealed NMO-IgG/AQP-4 antibody. The patient presented seropositivity for anti-SSA and anti-SSB. Antibodies to other immunological antibodies were undetectable.

*Corresponding author: Yanqiang Wang, Department of Neurology The Affiliated Hospital of Wei fang Medical University, No 2428 uhe Road, Weifang, Shandong 261031, China, Tel: 8657188165697, E-mail: wffwywy2006@126.com

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lesions in the conus medullaris were 19.4% in NMO with longitudinally extensive transverse myelitis (LETM), Aquaporin 4 antibody (AQP4-Ab) negative and AQP4-Ab positive with longitudinally extensive transverse myelitis (LETM) have conus involvement, respectively, 34% or 6% [3,4]. Factors influencing variation may due to the differences in ethnic, disease susceptibility, diagnostic criteria, sample size. Secondly, magnetic resonance imaging of the cervical, thoracic is the common regions, the lesions in the conus medullaris are rare in NMOSD, this has not attracted attention. Although bladder and bowel dysfunction is the most common symptom of cone involvement, Because of the pathway involved in sacral cord, pontine reticular formation, Para central lobule neuron pathways, It is easy to neglect the lower motor neuron pathways involvement [2-4].

NMOSD pathogenesis involves the autoantibodies (NMO-IgG) against astrocyte water channel aquaporin-4 (AQP4), which initiates complement dependent cytotoxicity (CDC) and antibody-dependent cell-mediated cytotoxicity (ADCC). The consequences of astrocyte damage and downstream inflammation including the cytokine release and infiltration of granulocytes and macrophages, disruption of the blood-brain barrier, and injury to oligodendrocytes and neurons [1,6,9]. Therefore, these suggest that the conus involvement have been associated with the titer of NMO-IgG/AQP4, the integrity of blood spinal cord barrier and blood brain barrier. An early diagnosis is essential to start treatment at an early stage and prevent relapses and disability progression in NMOSD. In acute attacks, high-dose intravenous corticosteroids and plasmapheresis, are of prime importance for the prevention of the inflammatory damage, next one is a maintenance treatment to avoid relapses including low-dose corticosteroids and non-specific immunosuppressants (azathioprine, tacrolimus, mycophenolate mofetil). The new biological agents such as rituximab: an anti-CD20 monoclonal antibody, and eculizumab: an anti-C5 monoclonal antibody [2,7].

To our knowledge, to avoid any diagnostic dilemma, it is significant to understand the rare presentation with incontinence and sexual dysfunction in NMOSD with conus involvement, therefore, timely lumbosacral MRI scan, and conus medullaris in NMOSD may be significantly valuable for for early diagnosis and prognostic evaluation.

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Competing Interests

All of the authors declare that there is no conflict of interest. All of the authors have not any sources of support or conflicts of interests in regards to this article

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