Clinical Case Report

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Neuromyelitis optica spectrum disorders with vertigo as the initial symptom

A case report

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Abstract

Rationale: Brain abnormalities have frequently been reported in neuromyelitis optica spectrum disorders patients, but vertigo as an initial manifestation has rarely been described.

Patient concerns: A 64-year-old woman who initially presented with vertigo, then accompanied with other brainstem manifestations and spinal cord involvement.

Diagnoses: MRI revealed medulla oblongata, cervical and thoracic spinal cord lesions. NMO-IgG antibody was seropositive. Taken her previous medical history and clinical manifestations into consideration, the patient was eventually diagnosed as neuromyelitis optica spectrum disorders.

Interventions: Before diagnosis, symptomatic treatment and acupuncture were adopted, whereas after diagnosis, steroid, intravenous immunoglobulin, and immunosuppressant were supplemented.

Outcomes: Her dizziness, nausea and vomiting were gradually relieved by symptomatic treatment and acupuncture before the confirmed diagnosis and immunotherapy. After added treatment with steroid, immunosuppressant, especially intravenous immunoglobulin, diplopia and nystagmus disappeared, and superficial sensation was improving. She was fully recovered six months after admission.

Lessons: Vertigo as a rare prodrome of neuromyelitis optica spectrum disorders deserves attention. The symptoms and signs were improved by a combined treatment of steroid, immunosuppressant, acupuncture, and particularly intravenous immunoglobulin.

Abbreviations: IVIG = intravenous immunoglobulin, NMO = neuromyelitis optica, NMOSD = neuromyelitis optica spectrum disorders.

Keywords: acupuncture, intravenous immunoglobulin, neuromyelitis optica spectrum disorders, neuromyelitis optica, vertigo

1. Introduction

Neuromyelitis optica spectrum disorders (NMOSD), which include the neuromyelitis optica (NMO), previously known as Devic's syndrome, are a group of autoimmune conditions characterized by inflammatory involvement of the optic nerve, spinal cord and central nervous system.[1] The first symptoms of NMOSD were reported with nausea and vomiting, diplopia and nystagmus, hearing and balance disorders, but vertigo is rare.[2,3] Here, we report a first-episode NMOSD case with a rare prodrome, highly progressive course and good response to intravenous immunoglobulin.

2. Case report

A 64-year-old Chinese famale initially presented with dizziness, diplopia, and walking instability 2 months before admission. 1 week later she was accompanied with epigastric discomfort, nausea and vomiting, occasional tinnitus, and numbness across the leftcostal arch to the left foot. She went to local hospital. Gastroscopy displayed chronic esophagitis and chronic superficial antral gastritis (moderate). Upper abdominal NMR scan revealed cholecystitis and mild bile duct dilatation. Cranial CT scan showed lacunar cerebral infarction in right basal ganglia and mild myelinopathy considered. Then she took Rabeprazole Sodium Enteric-coated Tablets, Itopride Hydrochloride Dispersion Tablets, Hydrotalcite Tablets, Deanxit and Paroxetine Tablets. But the symptoms persisted, and constipation occurred 1 week before admission.

Then, the woman was admitted to our hospital with above symptoms. Her past medical history was unremarkable. The patient had a history of hypertension and cataract surgery in

Editor: N/A.

We obtained written consent from the patient for publication of this case.

Ethics committee approval is not included, as it is accepted in our hospital that case reports do not require such approval.

This work was supported by the Zhejiang Hospital Health Science and Technology project (No: 2018KY564 to RMA); key Science and Technology Innovation Team of Zhejiang Province (No: 2013TD15 to DH).

The authors have no conflicts of interest to disclose.

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Medicine (2019) 98:34(e16906)

Received: 16 January 2019 / Received in final form: 23 June 2019 / Accepted: 29 July 2019

http://dx.doi.org/10.1097/MD.0000000000016906

the right eye. Her home medications included Irbesartan and Hydrochlorothiazide tablets and Nifedipine Sustained-Release Tablets. No other family member had neurological disease or any autoimmune disease. The patient denied any recent fever, rash, sore throat and diarrhea. She got diplopia. Horizontal nystagmus was positive. The left lower limb was hypalgesic, while bilateral deep sensations were normal. Bilateral pathological signs were negative. Romberg test was positive, while bilateral finger-nose tests were negative. She had staggering gait. Other examinations were normal. The patient was considered as vertigo syndrome and lacunar cerebral infarction. After treatment with Betahistine mesilate Tablets, Alprostadil Injection, Aspirin Enteric-coated Tablets, Atorvastatin Calcium Tablets and acupuncture, her dizziness and left lower limb numbness were gradually relieved.

However, numbness of the right lower limb occurred 1 day after admission and urinary retention appeared the 4th day after admission. On the 4th day after admission, diplopia, and horizontal nystagmus persisted. Bilateral lower extremity strength was 5-/5. The sensation of pain and rough touch were impaired from the right groin to the right foot compared to the contralateral side, while the vibration and motion sense of the right lower limb were dull compared with the contralateral side. The skin temperature of the right lower limb was lower than that of the opposite side. Bilateral Hoffman signs were positive, and right Plantar reflex was absent. Romberg test and bilateral finger-nose tests were positive. Blood tests as well as liver and kidney function were normal. The cervical vascular CTA showed bilateral vertebral arteries were tortuous.

Then, the muscle strength of the lower extremities decreased dramatically. On the 8th day after admission, the muscle strength of lower limbs was graded as 2 out of 5, while the upper limbs were 5-. The sensory disturbance level ascended to the right sixth thoracic level. Bilateral finger-nose tests were strong positive. The Ataxia of upper arms was worse with eyes closed. Thoracic spinal cord MRI showed equal signals on T1-weighted images (T1WI) and long signals on T2-weighted images (T2WI) through the middle and lower segment of thoracic spinal cord, as well as high signals on STIR images. Intense enhancement of that part was showed by MRI enhanced scan (Fig. 1). Cervical spinal cord MRI showed long signals on T1WI and T2WI from medulla oblongata to the 2nd cervical level, where was mild swollen (Fig. 2). Brain NMR revealed multiple lacunar ischemic foci around bilateral semioval center and lateral ventricle. NMO-IgG antibody in serum was positive. The patient was eventually diagnosed as NMOSD based on her previous medical history, clinical manifestations, seropositive NMO-IgG antibody, and exclusion of MS by MRI.

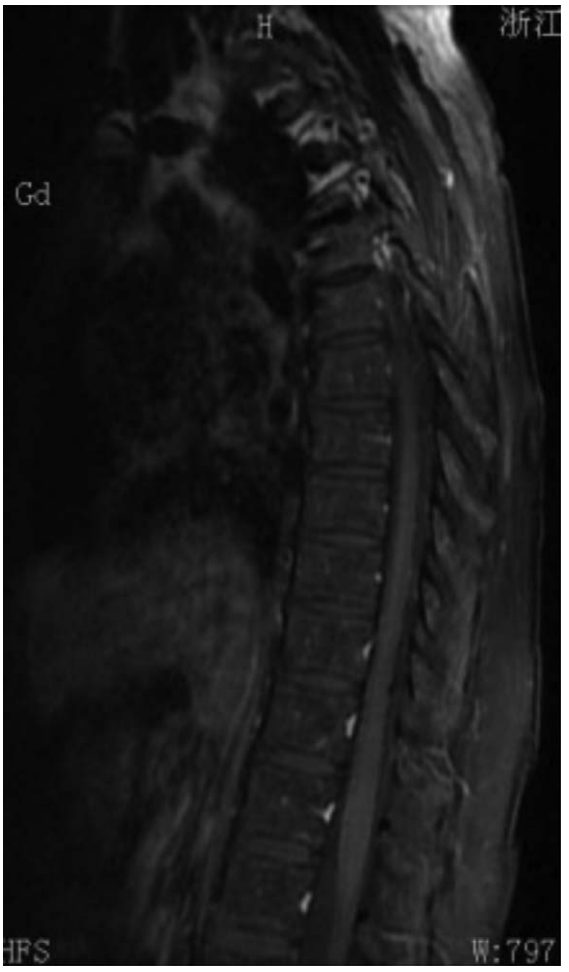

Figure 1. Enhanced scan on Sagittal T2-weighted MRI demonstrates extensive involvement of the thoracic spinal cord.

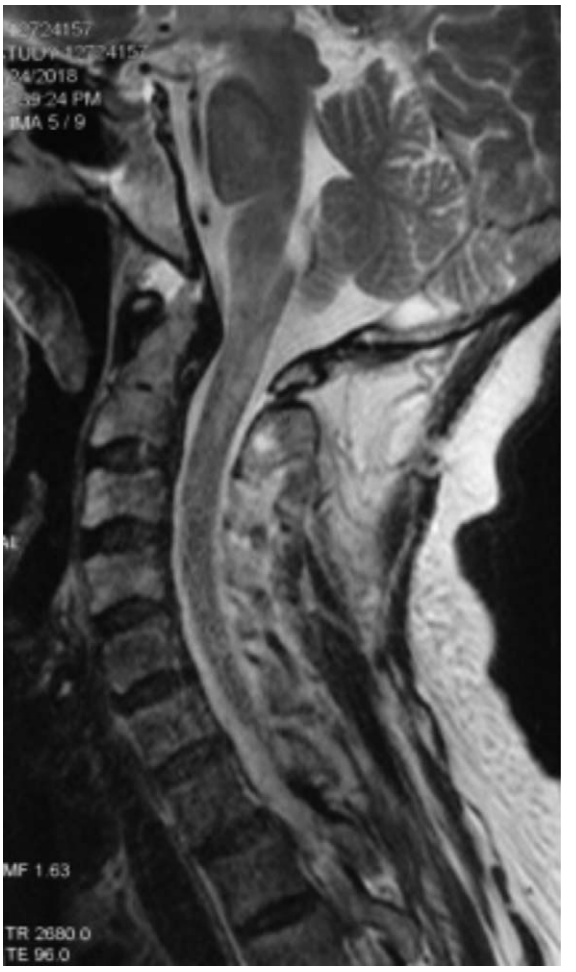

Figure 2. Sagittal T2-weighted MRI shows lesions from medulla oblongata to the 2nd cervical level.

Then, the patient received high-dose intravenous methylprednisolone (0.5 g for 3 days), but did not respond appropriately. Subsequently, intravenous immunoglobulin (0.4 g/kg for 5 days) was adopted, and her symptoms were gradually alleviated. On the 25th day after admission, diplopia and horizontal nystagmus disappeared. The pain and rough touch sensation were less dull with the sensory level descending to the right eighth thoracic level. The Tendon reflexes of bilateral biceps brachii were symmetrically active, whereas the quadriceps femoris Tendon reflexes and ankle reflexes disappeared. Bilateral Hoffman and Babinski signs were positive. The defecation was involuntary, whereas the urine still depended on urethral catheterization. Thereafter, she took Prednisolone Acetate Tablets and Mycophenolate Mofetil Dispersible Tablets as a long-term maintenance. Six months after admission, the patient achieved a complete clinical recovery without any relapses.

3. Discussion

The patient suffered vertigo, diplopia, nausea, vomiting and left lower limb numbness about 2 months before admission. During hospitalization, she developed to have new-onset paraplegia, progressive numbness across right side of the body and bladder dysfunction. MRI revealed medulla oblongata, cervical, and thoracic spinal cord lesions, while NMO-IgG antibody was seropositive. There was no optic nerve involvement in this case. According to the International Consensus Diagnostic Criteria for Neuromyelitis Optica Spectrum Disorders,[4] this patient was diagnosed NMOSD.

The initial presentation of this case was vertigo. NMO frequently follows an infectious prodrome characterized by headache, myalgia, and upper respiratory symptoms. Nausea and vomiting, diplopia and nystagmus, hearing and balance disorders may be seen as the first presentations of NMO, but vertigo is rare.[2,3] A multicenter study showed the incidence of vertigo or vestibular ataxia was 1.7% in NMO patients, while vomiting up to 33.1%.[3] Vertigo and nystagmus could be caused by lesions in the medulla, cerebellum or pons. As for our patient, the involvement of medulla oblongata revealed in MRIs might be the cause. Manifestations including nausea and vomiting as prodromes before the classical optic neuritis or transverse myelitis are thought to be due to vulnerability in the area postrema to serological factors such as NMO-IgG/AQP4-Ab.[5] Owing to the leakage of the blood-brain barrier in this area within the brainstem, it may be a primary target of the process in NMOSD.[5] An awareness of diverse and uncommon primary manifestations is conducive to early and accurate diagnosis of NMOSD.

Her dizziness, nausea, and vomiting were gradually relieved by symptomatic treatment and acupuncture before the confirmed diagnosis and immunotherapy. When NMOSD was diagnosed, we adopted high-dose intravenous methylprednisolone according to the recommendations,[1,6] followed by intravenous immunoglobulin (IVIG) as an alternative to therapeutic plasma exchange (TPE), for lack of response to the steroid approach. We observed significant improvement on superficial sensation and nystagmus after IVIG. To date, IVIG is not covered in standard management. However, several reports including this case and studies of patients with NMOSD on intermittent IVIG all showed potentially beneficial results in the acute attacks or prevention of the relapses with an acceptable side effect.[7,8] Theoretically, IVIG would seem a reasonable therapeutic option for NMOSD, as NMOSD is a humorally mediated neuroimmunological disease and IVIG is known to modulate immune function.[9] IVIG may be a plausible treatment option for relapse treatment and prevention. Further explorations, including prospective randomized controlled trials of larger NMOSD patient cohorts covering longer follow-up periods, are required in order to confirm its efficacy and determine optimal dose and infusion periodicity.

As NMOSD takes a relapsing course in most cases, with incomplete recovery and rapid accumulation of neurological deficits, long-term immunosuppressive treatment is imperative and highly recommended. This patient was taking prednisone and mycophenolate mofetil orally to maintain remission and prevent relapses.

In addition to the standard management, we also adopted acupuncture through the whole course to promote functional recovery. A systematic review and meta-analysis on 12 studies by MA demonstrated that acupuncture could have a beneficial effect on neurological recovery, motor function and functional recovery.[10]

In conclusion, our case, as the first episode of NMOSD, initially presented with dizziness, diplopia and walking instability, then accompanied with other brainstem manifestations and spinal cord involvement. Great improvement was achieved by a combined treatment of steroid, IVIG, immunosuppressant and acupuncture.

Author contributions

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Resources: Jing Liu, Kelin He.
Supervision: Ruijie Ma.
Validation: Dexiong Han.
Visualization: Kelin He.
Writing – original draft: Qin Guo, Ruijie Ma.
Writing – review & editing: Qin Guo, Ruijie Ma.

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