Finding the missing link in neuromyelitis optica presenting with recurrent transverse myelitis flares

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ABSTRACT

Neuromyelitis optica (NMO) is a distinct clinical entity from multiple sclerosis with its own clinical, laboratory and pathological characteristics. Definitive diagnosis of NMO is challenging at times as there can be a long interval between the occurrence of the index event and other neurological deficits which would fulfill the diagnostic criteria. Detection of NMO antibody could serve as an early marker in the disease progression. We present a young woman previously identified to have NMO antibody with recurring episodes of transverse myelitis for 3 years before manifesting with optic neuritis.

1. Introduction

Neuromyelitis optica (NMO) or Devic's disease is a severe idiopathic inflammatory demyelinating disease that selectively targets optic nerves and the spinal cord. Previously thought to be monophasic in presentation consisting of bilateral simultaneous optic neuritis (ON) and acute myelitis, the NMO spectrum now includes relapsing disorder of ON and myelitis, both of which could be separated months or even years apart, thus potentially delaying an accurate diagnosis. The presence of NMO antibodies can distinguish NMO as a distinct entity. We report a young woman who was previously identified as having NMO antibody positivity and had recurring episodes of transient transverse myelitis for a year before manifesting with unilateral ON and new brainstem lesions.

2. Case presentation

A 26-year-old woman presented with blurring of vision in her left eye. She reported a 5 day history of pressure-like pain on the left eye during lateral gaze associated with headache, characterized as pulsating pain on the temporal area. Two days prior to admission, she started to have blurring of vision of the left eye which was gradually worsening with lateral visual cuts on the left eye and decreased color perception. Within the past year, she had three previous admissions for symptoms attributed to transverse myelitis flares, manifesting as numbness or weakness on the lower extremities and left upper extremity with improvement after treatment with intravenous (IV) steroids. A positive NMO antibody was detected a year before her current presentaiton.

Ophthalmologic examination revealed decreased visual acuity in the left eye (oculus sinister, faint hand movement and finger counting) while the right eye was normal (oculus dexter, 20/20). There was positive afferent pupillary defect of the left eye. Extraocular muscle movement was full in all directions of gaze on both eyes. Visual confrontational testing revealed lateral visual field cut on the left eye. Slit lamp and dilated fundoscopic exams were unremarkable. MRI of the brain revealed increased T2 signal in the right cerebral peduncle (Fig. 1) extending to the right thalamocapsular region (Fig. 2) without surrounding vasogenic edema or mass effect, and mild enhancement of left optic nerve on post-contrast T1-weighted imaging (Fig. 3). Previous brain MRI performed a year ago was unremarkable. The NMO antibody returned positive (>160 U/mL). During the hospital stay the patient's vision on the left eye had no improvement after 5 days of IV steroids. She did not develop new neurological deficits. She was discharged with follow-up at a neurology clinic and was given prednisone *per os* [PO] to be taken every day in a gradually tapering dose and azathioprine 50 mg PO once a day with complete blood count monitoring weekly.

3. Discussion

Our patient exemplifies the importance of considering NMO as a differential in patients with transverse myelitis flares even in the absence of ON which can manifest later, as observed in this patient. Obtaining NMO antibody has both diagnostic and prognostic value [1]. This young woman fulfilled only one major criterion in presenting 1 year prior to developing ON with three episodes of numbness and weakness of lower extremities and two minor criteria, namely extensive spinal cord lesions and positivity for NMO antibody. The major criteria were not fulfilled until a year later with the appearance of unilateral retrobulbar ON. In a retrospective study in 1999 by Wingerchuk et al., it was reported that NMO could either be monophasic or relapsing. Most relapsing patients manifested with isolated ON or myelitis separated from other index events over a period of more than 3 months [2].

In 2006, the revised NMO diagnostic criteria emphasized the specificity of longitudinally extensive spinal cord lesions and NMO immunoglobulin (Ig) G seropositivity in diagnosing NMO while the absolute restriction on optic nerves and spinal cord has been removed. According to the latest diagnostic criteria, the two absolute requirements for diagnosis are ON and acute myelitis and at least two of the three minor criteria, which are (1) contiguous spinal cord MRI of lesion extending over $\geqslant 3$ vertebral segments, (2) brain MRI not meeting diagnostic criteria for multiple sclerosis, and (3) NMO-IgG seropositive status [3].

Lennon and colleagues reported that the presence of NMO IgG could distinguish NMO from multiple sclerosis with a sensitivity

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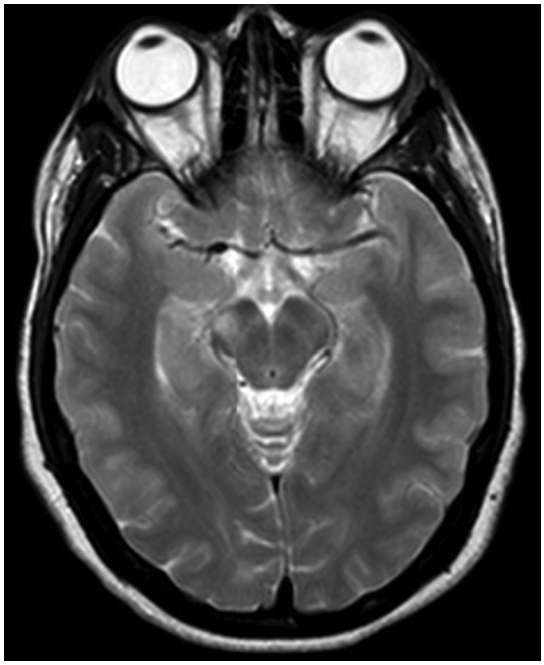

Fig. 1. Axial T2-weighted MRI of the brain showing increased T2 signal in the right cerebral peduncle.

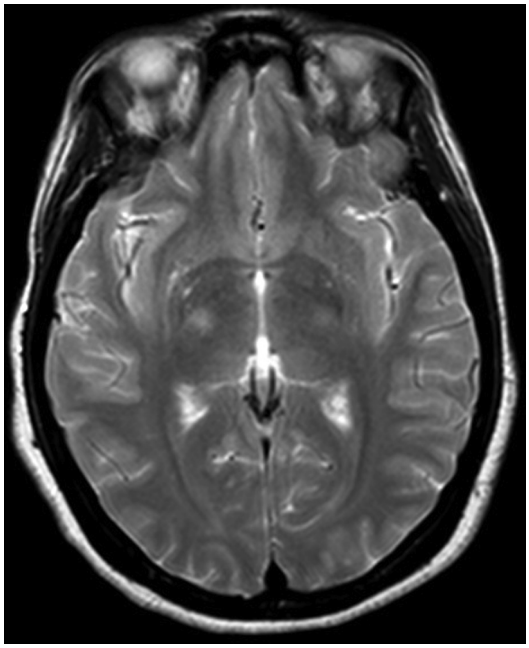

Fig. 2. Axial T2-weighted MRI of the brain showing involvement of the right thalamocapsular region.

and specificity of 73% and 91% for NMO, respectively [4]. It has been suggested that autoantibodies to aquaporin 4 are derived from peripheral B cells causing the activation of complement, inflammatory demyelination and necrosis [5]. NMO IgG antibody was detected in 52% of recurrent transverse myelitis patients in one retrospective study [4].

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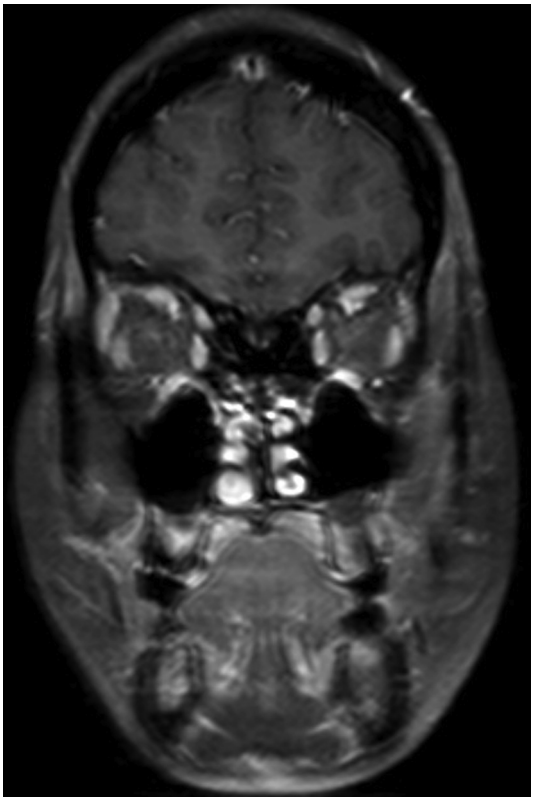

Fig. 3. Coronal T1-weighted post-contrast MRI of the brain showing mild enhancement of the left optic nerve.

4. Conclusion

Our patient highlights that the two major criteria (ON and acute myelitis) can occur separately as distinct clinically isolated syndrome. A high index of suspicion for NMO should be entertained in patients with isolated transverse myelitis even in the absence of ON on initial clinical presentation. Obtaining diagnostic biomarkers for NMO will facilitate earlier diagnosis and initiation of appropriate treatment to prevent complications. Early and accurate diagnosis is important because NMO carries a poorer prognosis than multiple sclerosis [3].

Conflicts of interest/disclosure

The authors declare that they have no financial or other conflicts of interest in relation to this research and its publication.

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