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Letter to the Editor

# Convergence spasm due to aquaporin-positive neuromyelitis optica spectrum disorder

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ABSTRACT

A female 27 presented with nausea and diplopia for 1 week. On examination she had normal vertical gaze but would develop convergence with miosis whenever she made horizontal saccades. Pupils were 6 mm and unreactive to light. MRI showed extensive hyperintensity in the dorsal midbrain and thalamus. Spinal MRI and CSF were both normal. Serum aquaporin-4-antibody was positive. She was treated with steroids and plasmapheresis and after 3 months convergence spasm resolved but pupils remained unreactive. Neuromyelitis optica often presents with brainstem signs, rarely a dorsal midbrain syndrome. Convergence spasm is occasionally of organic neurologic origin.



A previously healthy 27 year old female ultrasonographer presented with diplopia and nausea for 1 week. She herself had also noted difficulty moving her own eyes.

On examination she was orthotropic in primary position and had normal vertical eye movements – saccadic, pursuit and vestibular. In contrast whenever she tried to make horizontal saccades her eyes would converge and her pupils would constrict (Video 1). The impression of the first neurologist to see her was that this was convergence-retraction nystagmus but on review it was clear there was in fact no nystagmus in the primary position, no retraction and that the abnormal convergence was triggered only by horizontal and not by vertical saccades. Furthermore in the primary position while fixating in the distance her pupils were 6 mm and unreactive to light (Video 2). Vision was normal and there was no abnormality of limbs, stance or gait.

MRI showed extensive hyperintensity in the dorsal midbrain (Fig. 1) and thalamus that enhanced after one week. MRI whole spine was normal. CSF cell count and total protein was within normal range for a slightly traumatic tap.

Over the next month she developed upward gaze paresis by which time serum aquaporin-4-antibody result returned as positive.

She was now treated with IV methylprednisolone 1 g/day and plasmapheresis for 8 days.

After 3 months convergence spasm and vertical gaze paresis resolved but pupils remained unreactive. She then developed urinary incontinence and 6 months later sudden visual loss in the right eye to <6/60 acuity with only slight improvement on re-treatment with IV methylprednisolone.

Classical presentation of neuromyelitis optica (NMO) is bilateral optic neuritis followed by transverse myelitis, as a monophasic illness [1]. The discovery of aquaporin-4 antibody (AQP4–immunoglobulin G [IgG]) changed our understanding of NMO [2]. NMO, we now know, can have various clinical presentations [3–6].

Concensus criteria for NMO spectrum diseases have been proposed [7]. This patient fulfils the criteria; AQP4 antibody positivity and one of the core symptoms: brainstem syndrome at presentation and 6 months later optic neuritis -dissemination in space.

NMO often presents with brainstem signs, rarely a dorsal midbrain syndrome. A case reported by Lee et al. [8] is similar to ours. Their patient also had a form of convergence spasm ("pseudoabducens palsy") and pupillary light-near dissociation but also had other dorsal midbrain oculomotor abnormalities - impaired vertical saccades, primary position retraction-nystagmus and lid retraction, which our patient did not. Nonetheless the MRI of their patient was similar to the MRI of our patient.

Convergence spasm is the prototypical functional eye movement disorder, especially in previously healthy young females, and so it is, almost always [9]. A few cases of typical convergence spasm have been reported in patients with organic neurological disease [10], particularly focal lesions of the dorsal midbrain [8,11].

In this case pupillary light-near dissociation was the initial clue that the patient's convergence spasm was organic and not functional.

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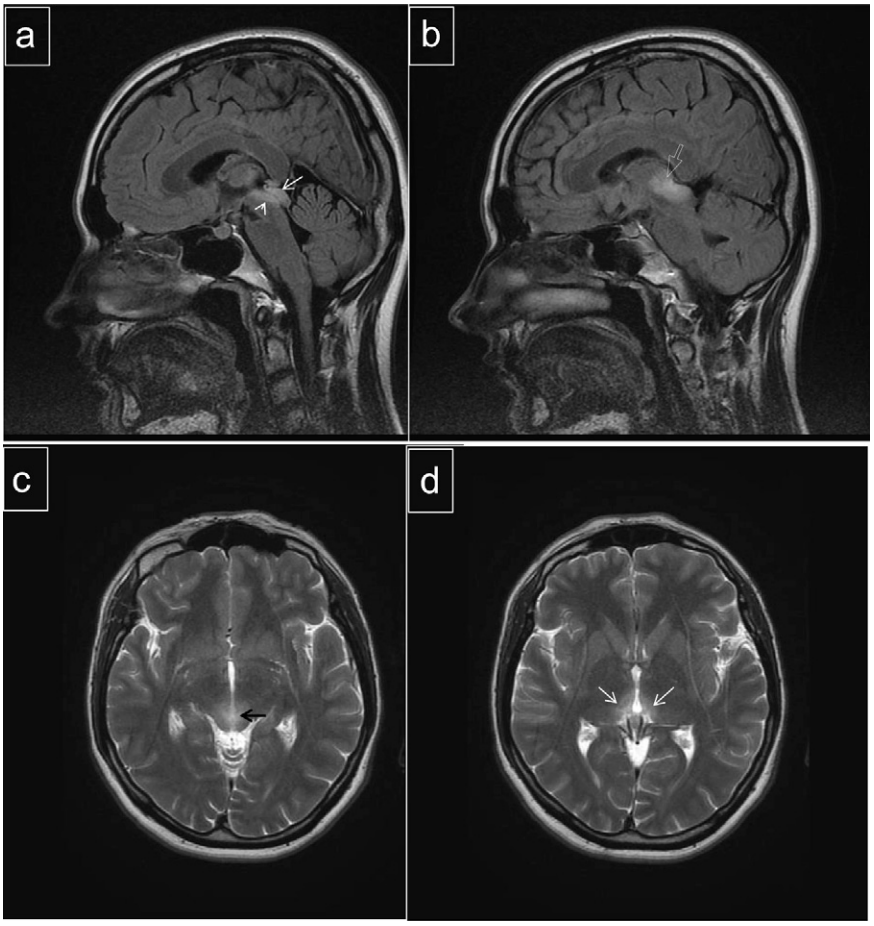


**Fig. 1.** Midsagittal (a) and parasagittal (b) FLAIR images show increased signal in the superior two thirds of the tectum (arrow in panel a), dorsal portion of the mesencephalic tegmentum (arrowhead in panel b). Extension of the abnormal signal from mesencephalon into diencephalon is appreciated on the parasagittal image (hollow arrow in panel b). Panels c and d. Two consecutive axial T2 weighted images delineate involvement of dorsal medial thalamic area (mediodorsal nuclei) (white arrows in panel d), pretectal area and posterior commissure (black arrow in panel c).

Supplementary data to this article can be found online at http://dx.doi.org/10.1016/j.ensci.2017.03.001.

## Conflict of interest

The authors declare that there are no conflicts of interest.

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