Intractable Hiccups in a Middle-Aged Female

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Figure 1. Axial T2 (A), FLAIR (B), and coronal T2-weighted (C) MRI sequences showing hyperintense signal in the region of dorsal medulla. FLAIR indicates fluid-attenuated inversion recovery; MRI, magnetic resonance imaging.

A previously healthy 50-year-old female was admitted with recurrent hiccups and nausea for 2 weeks. She had previously taken several consultations from physicians and gastroenterologist without getting any relief. She denied any abdominal symptoms, visual impairment, motor weakness, incoordination, gait imbalance, or abnormal sensation. General physical and neurological examinations were normal.

A magnetic resonance imaging (MRI) scan of the brain showed a hyperintense signal in the region of dorsal medulla on T2-weighted and fluid-attenuated inversion recovery sequences (Figure 1A-C) without diffusion restriction or post-gadolinium enhancement. Screening of the spine MRI and visual evoked potentials were normal. Serum aquaporin-4 (AQP4)/neuromyelitis optica (NMO)-IgG antibody test (by indirect immunofluorescence assay) was positive. A diagnosis of neuromyelitis optica spectrum disorder (NMOSD) with isolated medullary involvement and without optic neuritis and transverse myelitis was made. The patient was given intravenous methylprednisolone 1 g daily for 5 days and showed clinical improvement. Subsequently, azathioprine was initiated to prevent further attacks.

Neuromyelitis optica or Devic disease is an immune-mediated demyelinating disorder of the central nervous system

that predominantly affects the optic nerves and spinal cord. The term NMOSD is used to include AQP4-IgG-seropositive patients with limited or atypical forms of NMO (eg, isolated longitudinally extensive transverse myelitis, isolated recurrent optic neuritis, or area postrema syndrome).¹ Intractable hiccups, nausea, and vomiting (area postrema syndrome) could be the initial or isolated presence of NMOSD.² Brain lesions in NMOSD are present in more than half of the patients at onset and usually seen at sites of high AQP4 expression, such as the diencephalon, the hypothalamus, and the periaqueductal region.² The NMOSD, sarcoidosis, Sjogren syndrome, systemic lupus erythematosus, tumor, and Behcet disease are the main differential diagnosis for solitary brain stem white matter lesion on brain MRI.

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Declaration of Conflicting Interests

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