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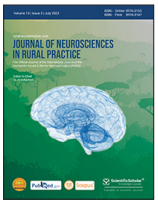

Case Report

Area postrema syndrome: An unusual presentation of neuromyelitis optica spectrum disorder

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ABSTRACT

Isolated area postrema syndrome (APS) is a rare neurological presentation of, neuromyelitis optica spectrums disorder (NMOSD), recognizable by uncontrollable hiccups, nausea, or vomiting. When it occurs as the first presentation of NMOSD, it may present as a diagnostic challenge as the condition may be frequently attributed to gastrointestinal pathology, and the subsequent diagnostic delay may result in debilitating neurological sequelae such as optic neuritis or myelitis. We report such a case of isolated APS in a young woman who presented with a clinical picture of bouts of vomiting and intractable hiccups causing considerable distress and was finally diagnosed to be a case of seronegative NMOSD.

Keywords: Intractable hiccups, Area postrema syndrome, Neuromyelitis optica spectrum disorder

INTRODUCTION

Neuromyelitis optica spectrum disorders (NMOSDs) are a group of immune-mediated inflammatory disorders affecting the central nervous system that commonly targets the optic nerves and spinal cord. It is associated with a pathogenic antibody specific for the astrocytic water channel aquaporin-4 (AQP4). The area postrema (AP), the diencephalon, the brainstem, and the cerebral hemispheres are now included in the diagnostic criteria as additional targets of these autoantibodies.[1] Area postrema syndrome (APS) is a rare neurological condition that manifests as uncontrollable episodes of nausea, vomiting, or hiccups.[2,3] It is easy to mistake APS, which manifests at the outset of NMOSD, for a gastrointestinal (GI) disease. Here, we would like to report a similar presentation in a 25-year-old female, who underwent extensive GI and other evaluation for intractable vomiting and hiccups and was confirmed to have APS on magnetic resonance imaging (MRI).

CASE REPORT

A 25-year-old lady presented with complaints of persistent nausea and vomiting and intractable hiccups causing considerable distress for the past 3 weeks. She also had occasional episodes of headaches. She initially underwent an extensive gastroenterological evaluation including upper GI endoscopy, and contrast-computed tomography scan of the abdomen, all of which were normal. Given the progressive nature of her symptoms along with her headache, she was referred to the neurology department for further evaluation.

On physical examination, vital signs were within normal limits and higher mental functions were intact. Cranial nerves were normal including optic fundi. She had minimal tandem gait ataxia, but no other cerebellar signs. Her motor power, deep tendon reflexes, and sensory system examination were all unremarkable.

Her routine blood investigations were all unremarkable. She underwent a MRI study of the brain with and without contrast to rule out any central causes. It revealed T2-fluid attenuated inversion recovery hyperintense lesion in the dorsal medial medulla at the AP which showed no enhancement with contrast [Figure 1]. There were also mild hyperintensities of the right optic tract and tuber cinereum. Her anti-myelin oligodendrocyte glycoprotein (MOG) antibodies and AQP-4 immunoglobulin G (IgG) were negative, but her classic gastrointestinal symptoms and typical MRI abnormalities were indicative of APS. Her magnetic resonance imaging of the spine and visual evoked potentials were normal. She underwent a lumbar puncture and cerebrospinal fluid analysis was normal.

She received treatment with 1 g/day of intravenous methylprednisolone for 5 days and she reported significant

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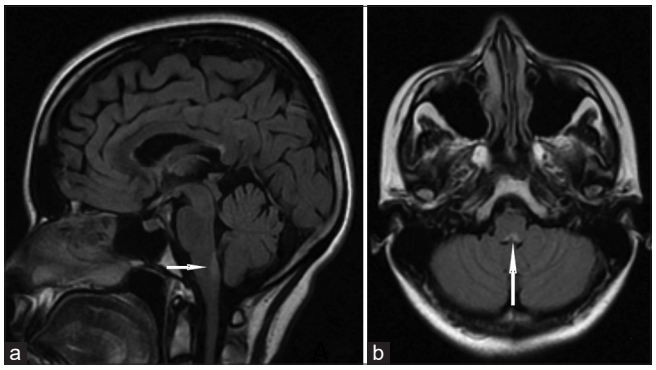

Figure 1: (a) Magnetic resonance imaging (MRI) brain T2 fluid-attenuated inversion recovery (FLAIR) sequence, sagittal view showing (arrow) hyperintensity at area postrema (dorsal medulla) and (b) MRI BRAINT2 FLAIR sequence, axial view showing (arrow) hyperintensity at area postrema (dorsal medulla).

improvement in her symptoms. She was kept on oral steroids with a plan to start on steroid-sparing immunosuppressant therapy.

DISCUSSION

AP is one of the called so-called circumventricular organs located on the dorsal medulla lining the floor of the fourth ventricle. Typically referred to as the brainstem's "vomiting center," it also plays other roles in the control of vasomotor/angiotensin II, somatic development, and hunger.[3]

APS is described as at least 48 h of intractable nausea, vomiting, or hiccups and is included as a core clinical criterion for NMOSD.[1] Unlike spinal and optic lesions seen in classical NMO, lesions in the AP usually demonstrate more inflammation than demyelination and necrosis, thus explaining the potential for complete recovery following treatment. The binding of IgG to AQP4 in the AP lacks the immunoreactivity for complement system activation, instead causing receptor downregulation. This results in alterations to neurotransmitter homeostasis and therefore triggers vomiting.[4]

Only 14% of patients presenting with APS are positive for anti-AQP4 antibodies and MOG positivity is also rare.[5] Our patient was also found to be a case of seronegative APS. Seronegative NMOSD is treated manner similar to seropositive.

As was seen in our case, the digestive symptoms at the forefront of this syndrome often delay the diagnosis of NMOSD as gastrointestinal pathologies are commonly alluded to initially. This can in turn result in delayed diagnosis and treatment. Our patient's case was striking given the considerable distress caused by the nature of her symptoms, rendering her incapable of any food intake without triggering vomiting and therefore causing significant weight loss. This case report emphasizes the importance of raising awareness of APS NMOSD as an early potential diagnosis option, to have a better prognosis.

CONCLUSION

NMO spectrum disorders can present as APS, solely with nausea, vomiting, and hiccups and without other neurological symptoms. Many cases may be seronegative for anti-AQP-4 antibodies. The MRI lesions in the AP are highly diagnostic of NMOSD. Intractable hiccups and vomiting not responding to routine supportive measures must be evaluated for APS.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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