Intractable Vomiting and Hiccups as a Presenting Symptom of Neuromyelitis Optica Spectrum Disorder

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Sir,

Intractable vomiting and hiccups are symptoms common to a wide variety of diseases lasting for more than 48 hours. Patients with this complaint first seek consultation with internists and gastroenterologists who do the necessary workup to exclude systemic and gastroenterological causes. That this may be the first manifestation of a neurological illness is not common knowledge amongst the physicians. It is only when other symptoms suggestive of a neurological illness appear, they are directed to a neurologist.

A 39 year old man presented with persistent vomiting and hiccups for 2 months with no other symptoms. He was initially seen and treated symptomatically by a general practitioner and gastroenterologist with no relief. His routine blood tests, USG abdomen and Endoscopy were normal. It was only when he developed diplopia followed by gait imbalance and hesitancy of micturition that he was referred to a neurologist. MRI brain was normal. MRI of the spinal cord showed long segment lesion extending from cervico-medullary junction up to C6 with cord swelling suggestive of demyelination. CSF showed proteins 57 mg% with 40 lymphocyte/mm3. His NMO IgG test was positive which suggested the diagnosis of Neuromyelitis Optica Spectrum Disorder (NMOSD). He received IV methylprednisolone for 5 days. He had marked relief from persistent vomiting and hiccups along with improvement in other neurological symptoms. He was started on Azathioprine and oral steroids. During his follow-up visits over next 18 months, he had no clinical or radiological recurrence.

Several neurological conditions may result in intractable vomiting and hiccups especially those involving the medullary region. Neuromyelitis Optica (NMO) was considered to be a monophasic illness affecting optic nerves and spinal cord before the landmark discovery of Aquaporin-4 IgG antibody by the Mayo hospital group. NMO antibody targets the regions in the central nervous system rich in astrocytic water channel aquaporin-4.

The area postrema located on the medullary floor of the fourth ventricle expresses AQP-4 abundantly and is a preferential target for NMO lesions. It is the chemosensitive receptor zone of the emetic reflex pathway. This explains the occurrence of intractable vomiting and hiccups in NMO. Such specific symptoms and MRI findings have resulted in the grouping of all these conditions with positive NMO antibody under the term NMOSD1 even if these occur in isolation like optic neuropathy, myelitis, intractable hiccups and vomiting.

Intractable vomiting and hiccups occur frequently both during the course of the disease and as an initial presenting feature. Researchers at Mayo Clinic found intractable vomiting as a presenting symptom in 12% of their seropositive AQP-4 patients.2 Only a few isolated case reports of such symptoms have appeared recently from India3,4 and that too only in the neurological literature. However, that NMOSD is an important cause of intractable vomiting and hiccups needs special emphasis. It is desirable to include MR imaging and testing for Anti-Aquaporin 4 (NMO antibody) when investigating a case of persistent vomiting and hiccups.

References