Anorexia Heralding the Onset of Neuromyelitis Optica

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Abstract

Neuromyelitis optica (NMO) is an inflammatory disease of the central nervous system (CNS) that preferentially affects the optic nerves and spinal cord. An autoantibody (NMO-IgG) targeting the aquaporin-4 water channel distinguishes NMO from other inflammatory disorders of the CNS. Recent studies have demonstrated that the area postrema and other circumventricular organs (CVOs) can be targeted in NMO. We herein report the case of a 12-year-old girl who experienced anorexia six months before the onset of NMO. Anorexia caused by hypothalamic or CVO dysfunction may herald the onset of NMO.

Key words: aquaporin-4, circumventricular organs, anorexia

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Introduction

Neuromyelitis optica (NMO) is an idiopathic inflammatory disease of the central nervous system (CNS) that preferentially affects the optic nerve and spinal cord. An immunoglobulin G (IgG) autoantibody (NMO-IgG) targeting the water channel protein aquaporin-4 (AQP4) is the biomarker that distinguishes the NMO spectrum disorders (NMOSD) from multiple sclerosis (MS) and other CNS demyelinating disorders (1, 2). Compelling evidence suggests that this IgG plays a pathogenic role in NMOSD. AQP4 is concentrated in astrocytic foot processes at interfaces between the CNS parenchyma and fluid compartments, including both cerebrospinal fluid (CSF) and blood (1, 2), and in areas involved in the regulation of fluid balance and energy homeostasis, including the hypothalamus and sensory circumventricular organs (the subfornical organ, the organum vasculosum of the lamina terminalis and the area postrema). Neuroimaging has revealed NMO-typical lesions in these AQP4-rich areas (3). To date, two cases of young female patients with NMO who developed anorexia have been reported. In one case, the anorexia was associated with intractable nausea and vomiting and an inflammatory lesion involving the area postrema (4). In the other patient, the anorexia was accompanied by psychotic symptoms and was associated with an inflammatory lesion of the hypothalamus (5).

We herein describe the case of a young girl who experienced isolated anorexia six months before the onset of NMO and discuss the involvement of the circumventricular organs (CVOs) in AQP4 autoimmunity.

Case Report

A 12-year-old girl developed a fever that lasted for three days. When the fever subsided, the patient began to lose her appetite. Her baseline height was 161 cm and her baseline weight was 56 kg. During psychological counseling, she denied dieting or inducing vomiting. She also experienced irregular menstrual periods. She never developed nausea, vomiting, hiccups or symptoms of autonomic dysfunction. She lost 12 kg in six months. Thereafter, she experienced acute bilateral visual loss in the setting of a demyelinating disease diagnosed as acute disseminated encephalomyelitis at a local hospital. After six months, she developed dizziness and upper limb paresthesias and was referred to our clinic. Brain MRI revealed multiple hyperintense lesions on T2-weighted images, all showing ring-enhancement after gadolinium administration (Figure A-C). Spinal cord MRI showed a longitudinally extensive lesion involving C1-C2-C3-C4 (Figure D). A cerebrospinal fluid analysis revealed no pleocytosis along with normal glucose and protein levels and the presence of oligoclonal bands. The visual evoked potentials showed a bilateral increase in P100 latency (P100 right eye: 133 msec; P100 left eye: 166 msec). The somatosensory evoked potentials were altered in the upper limbs.
The authors state that they have no Conflict of Interest (COI).

References

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