Case Report

MRI findings of autoimmune glial fibrillary acidic protein astrocytopathy involving infratentorial: Case report

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Article history:
Received 28 February 2022
Revised 14 April 2022
Accepted 15 April 2022

Keywords:
Autoimmune glial fibrillary acidic protein astrocytopathy
neuromyelitis optica spectrum disorders
magnetic resonance imaging

Abstract

Autoimmune glial fibrillary acidic protein astrocytopathy (GFAP-A) is a new type of autoimmune astrocytopathy first defined in 2016. Lack of clinical understanding, often misdiagnosed as optic neuromyelitis or multiple sclerosis. We report the clinical and MRI findings of an elderly patient with autoimmune glial fibrillary acidic protein astrocytopathy. With intractable vomiting as the first symptom, the brainstem showed typical vascular enhancement. GFAP-A lacks specificity in clinical and MRI scans. When enhancement reveals paraventricular “vascular-like enhancement” or central spinal cord tubular enhancement, it is important to consider the possibility of this disease.

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Case Report

A 72-year-old male patient had recurrent headache and intractable vomiting for more than half a month and had a history of optic neuritis. MRI showed stripe of long T1 and long T2 signal in the medial ependymal area of the right temporal lobe, brainstem, medulla oblongata and dorsal medulla oblongata, high signal intensity on T2-FLAIR, slightly high signal intensity on DWI, stripe of nodular and linear enhancement, and the length of spinal cord lesions was less than 3 segments (Fig. 1A-D). Laboratory examination showed that the

* Acknowledgments: Written informed consent was obtained from his guardian for publication of this report.

Competing Interests: The authors report no disclosures relevant to the manuscript.

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https://doi.org/10.1016/j.radcr.2022.04.032

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antibody to GFAP was positive (Fig. 2), the level of protein in cerebrospinal fluid was elevated, and the content of sugar and chlorine in cerebrospinal fluid was normal. No specific family history. Combined with imaging and laboratory examination, the clinical diagnosis was GFAP-A.

Discussion

Lennon's team [3] first reported and named autoimmune glial fibrillary acidic protein astrocytomatosi in 2016 and identified the biological marker for this type of disease as specific GFAP antibodies, confirming it as a separate disease from the multiple sclerosis and optic neuritis optica spectrum. The etiology and pathogenesis of GFAP-A is unclear, with some studies suggesting it may be related to tumors and viral infection, about 20% of patients may have concomitant autoimmune diseases such as diabetes mellitus and rheumatoid arthritis [3].

GFAP-A occurs in middle-aged patients over 40 years of age, with slightly more women than men. In foreign studies [3] the clinical presentation was dominated by encephalitis and meningoencephalitis (54.5%), followed by myelitis (10.5%), while in domestic studies [4] most of them were dominated by optic neuritis (63.2%) and myelitis (68.4%), and a few patients could present with motor disorders, autonomic dysfunction, and peripheral neuropathy. In the present case, there was a history of optic neuritis involving the brain parenchyma, brainstem and spinal cord, and the main clinical manifestation was intractable vomiting due to the involvement of the extreme posterior region.

Cerebrospinal fluid examination in GFAP-A shows a marked increase in leukocytes, including lymphocytes, monocytes and polymorphonuclear cells; protein levels in the cere-

Fig. 1 – (A-D) Brain and Neck MRI.
of vascular-like enhancement in the brainstem that traveled horizontally.

The current clinical understanding of GFAP-A is relatively limited and lacks uniform diagnostic criteria, and some scholars have suggested that the possibility of GFAP-A should be considered when patients present with the following conditions that cannot be explained by other diseases [10]: (1) acute or subacute onset with clinical manifestations of brain, meningeal, spinal cord, or optic nerve involvement or a combination of symptoms; (2) MRI findings of intracranial and/or spinal cord multiple lesions with specific vascular-like radial enhancement seen; (3) positive cerebrospinal fluid GFAP antibodies; (4) brain biopsy suggestive of small vessel lesions; (5) effective steroid hormone therapy; and (6) exclusion of other possible diseases.

GFAP-A is mainly distinguished from NMOSD and multiple sclerosis. Intracranial foci of NMOSD are commonly found in the white matter of the cerebral hemispheres and the brainstem, with MR manifestations of fused high signal on T2-FLAIR and T2WI, usually in the lateral ventricles, the ventricular canal layer of the third and fourth ventricles, the corpus callosum, the ventricular canal surface of the mesencephalic region, and the brainstem. Spinal cord lesions are most often located in the cervical and thoracic segments, with lesions ≥3 spinal cord segments in length and more than 50% cross-sectional involvement. In addition, CSF pressure is rarely elevated in NMOSD, leukocyte count may be mildly elevated, and protein levels may be elevated in a few patients. The intracranial involvement of multiple sclerosis is usually manifested by abnormal high signal in the paraventricular white matter perpendicular to the lateral ventricles (Dawson’s sign), and the lesions touch the ventricular canal surface. Spinal cord lesions are usually located in the posterior or lateral part of the spinal cord, with a lesion length of <2 spinal cord segments and <1 of 2 of the spinal cord area in cross-section.

In conclusion, GFAP-A lacks specificity in clinical and MRI scans. When enhancement reveals paraventricular “vascular-like enhancement” or central spinal cord tubular enhancement, it is important to consider the possibility of this disease.

Fig. 1 A and B axial T2-FLAIR and sagittal T2WI showed multiple abnormal signals in the right temporal lobe, medulla oblongata, and medulla oblongata. Fig. 1 C and D coronal and sagittal enhanced T1WI showed strip enhancement of the lesion and horizontal vascular enhancement of the brain stem.

The cytoplasm of Bergmann astrocytes in the cerebral cortex was radial.

**Patient Consent**

Patient agrees to publish.

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