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Case Report

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

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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.

Autoimmune glial fibrillary acidic protein astrocytoma (GFAP-A) is a new type of autoimmune astrocytoma first defined in 2016, and the other is neuromyelitis optica spectrum disorders (NMOSD) [1–3]. Both GFAP-A and NMOSD belong to autoimmune astrocytic disease, which can have abnormal vision and myelitis. In addition, there are few reports and lack of clinical understanding, so they are often misdiagnosed NMOSD. Because of its high disability rate, timely and correct diagnosis is particularly important. The clinical and imaging data of a confirmed case of GFAP-A were analyzed retrospectively in order to deepen the understanding and improve the ability of differential diagnosis.

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

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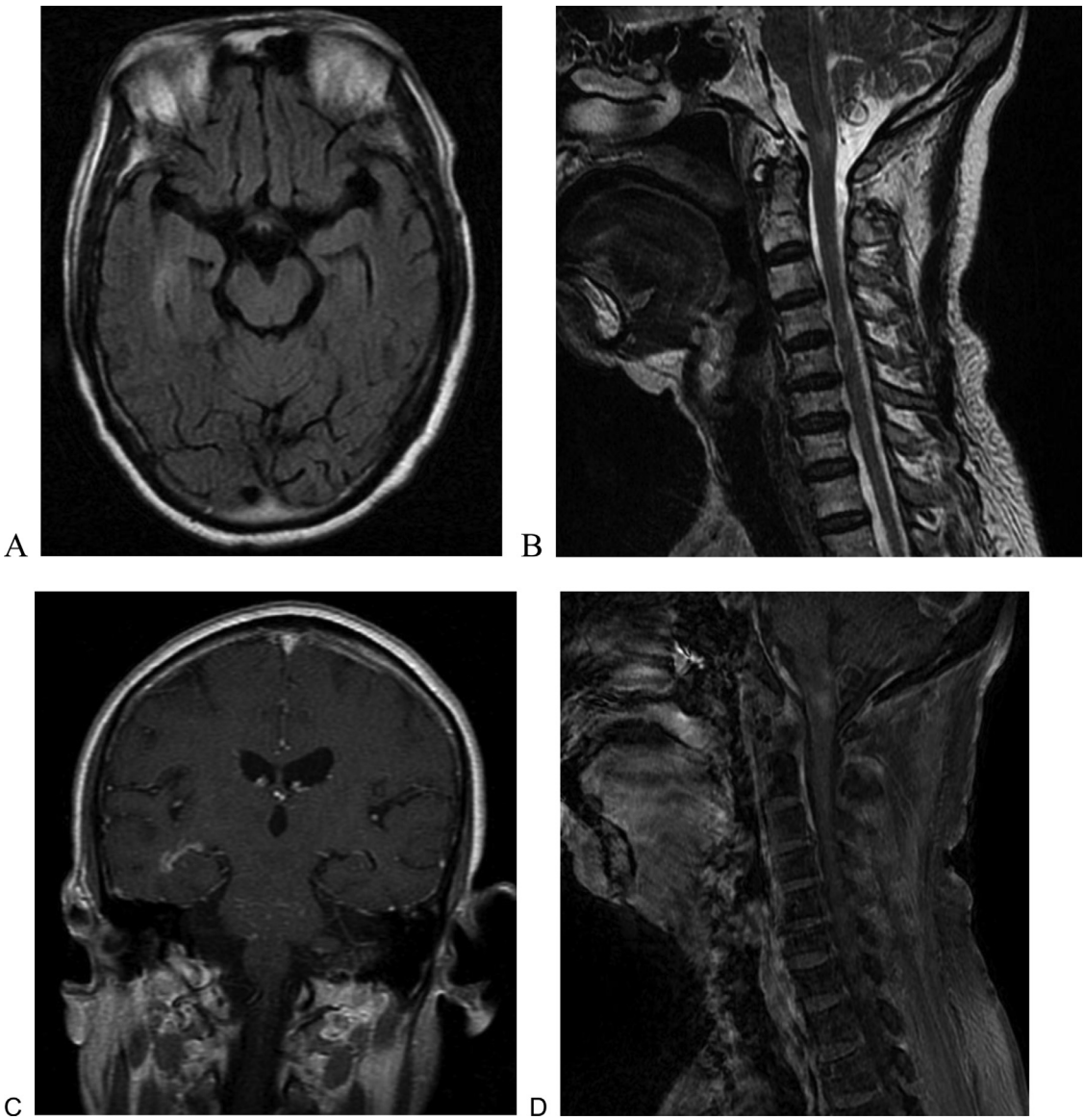

Fig. 1 – (A-D) Brain and Neck MRI.

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 astrocytomatosis 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 neuromyelitis 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-

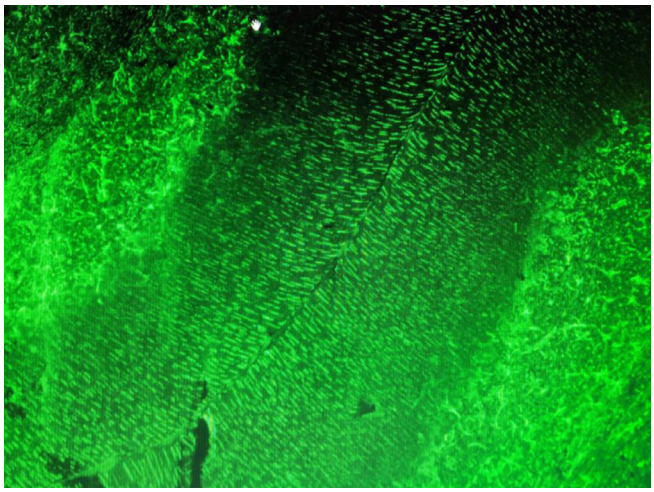

Fig. 2 – Detection of GFAP-IgG in cerebrospinal fluid.

brospinal fluid are elevated and may be >1 g/L in severe cases, cerebrospinal fluid sugar levels may be reduced in a minority of patients, and elevated cerebrospinal fluid pressure may be seen in about 20% of patients [3, 5-7]. Both cerebrospinal fluid GFAP antibodies and serum GFAP antibodies have greater clinical significance for the diagnosis of GFAP-A, but the former has higher specificity and sensitivity than the latter, which often overlaps antibodies associated with other autoimmune diseases [3]. In this case, the cerebrospinal fluid protein levels were all elevated and the cerebrospinal fluid sugar levels were normal.

Most patients with GFAP-A have imaging changes in the brain and spinal cord, and MR is the imaging examination of choice. Combined with recent years of domestic and foreign literature reports [3–10] analysis of GFAP-A patients mainly involved the white matter of the brain and/or spinal cord, MR mainly showed multiple foci, mainly involving the white matter of the brain, basal ganglia area, brainstem and spinal cord, other involved parts also include cerebellum, meninges, ventricles and skull, brain parenchyma and spinal cord mainly showed patchy, striped long T1 long T2 signal shadow. The most specific manifestation is "vascular radiolucency" perpendicular to the lateral ventricles in the white matter of the paraventricular brain, which is seen in about 40%-50% of patients. Intraspinal lesions are usually located in the cervical and thoracic segments, mainly involving the gray matter of the spinal cord, and can be larger than 3 spinal cord segments. The enhanced lesions may disappear after treatment. Analysis may be due to the fact that the main pathology of GFAP-A is perivascular inflammation, and the enhancement is caused by leakage of the enhancing contrast agent from the damaged vascular barrier, and the enhancement disappears after treatment when the blood-brain barrier is repaired. In addition, PET imaging of the brain may show hypermetabolism corresponding to the area of MRI abnormality [6]. In the present case, the deep right temporal lobe and adjacent ventricular canal, brainstem and spinal cord were involved, and abnormal signal in the form of strips and slices was seen, with enhancement showing strips and slices of enhancement, and multiple foci 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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