Anti-NMDAR encephalitis combined with a subependymoma

Sir,

Anti-N-methyl D-aspartate receptor (NMDAR) encephalitis is a novel NMDAR-mediated form of autoimmune encephalitis.[1] The main clinical manifestations include psychiatric symptoms, epilepsy, movement disorders, disturbance of consciousness, autonomic nervous system disorders, and central hypoventilation.[2] It is necessary to detect anti-NMDAR antibodies as early as possible to confirm the diagnosis of anti-NMDAR encephalitis.[2] Anti-NMDAR encephalitis is usually associated with a teratoma;[1] however,

anti-NMDAR encephalitis combined with a subependymoma has still not been reported.

A 34-year-old male patient presented with neurological symptoms of blurred and magnified vision, paroxysmal amaurosis, and malaise. A heterogeneous space-occupying lesion was detected in the patient's right lateral ventricle, with slight hypointensity on T1 weighted imaging (WI), heterogeneous hyperintensity on T2-WI and T2-FLAIR imaging, without gadolinium contrast enhancement [Figure 1]. Anti-NMDAR antibodies were present in the cerebrospinal fluid (CSF), but not in the serum.

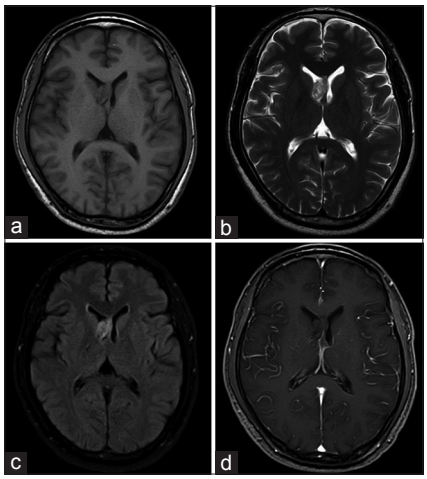

Figure 1: A heterogeneous space-occupying lesion was detected in the patient's right lateral ventricle on MRI. (a) Slight hypointensity on T1-WI. (b) Hyperintensity on T2-WI. (c) Slight hyperintensity on T2-flare. (d) No enhancement with gadolinium contrast on T1-WI

Six months after conservative treatment, the patient's symptoms had almost resolved. The mass was removed by surgical resection, and a series of histopathological examinations were performed. Hematoxylin and eosin staining showed that there were clusters of cellular neoplastic proliferation with islands of high nuclear density and abundant fibrillary matrices [Figure 2a-d]. Immunofluorescence staining revealed that there were sporadic NMDAR-positive cells distributed in the glial fibrillary acidic protein (GFAP) positive cells and neuropil [Figure 2e-h]. The tumor was histopathologically confirmed to be a subependymoma (WHO grade 1).

The patient was, therefore, diagnosed as the first case of anti-NMDAR encephalitis associated subependymoma. Anti-NMDAR antibodies were detected in the CSF, but not in the serum, which strongly indicated that the anti-NMDAR antibodies were synthesized intrathecally. As the only intracranial neoplasm expressing immunogenicity to NMDAR in the central nervous system, the presence of a subependymoma may be closely related to the intrathecal composition of anti-NMDAR antibodies. Since the resection of subependymomas, the patient has not had a relapse at 1 year of follow-up observation.

A subependymoma is considered to be a variant or subtype of an ependymoma and may be originate from subependymal cells.[3] The cell of origin of a subependymoma is controversial and its ultrastructural features exhibit both astrocytic and ependymal differentiation.[3] The activated tumor cells of the subependymoma express functional NMDAR.[4] As the patient's immune allergens stimulate the body to produce anti-NMDAR antibodies, anti-NMDAR encephalitis may be induced.

This is the first reported case of anti-NMDAR encephalitis associated with a subependymoma, with NMDAR-positive

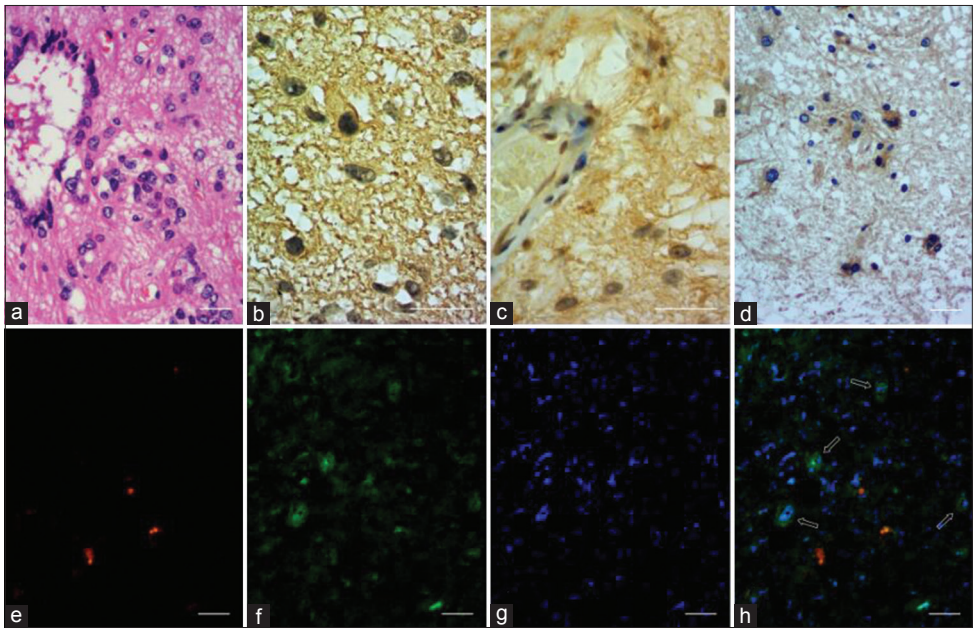

Figure 2: Histopathological observations. (a) Hematoxylin and eosin staining; (b) GFAP-positive cells; (c) S100-positive cells; (d) Immunopositive reaction; (e) NMDAR-positive cells; (f) GFAP-positive cells; (g) Nuclear staining with DAPI; (h) The merged image (e-g). (Bars = 50 μm)

immunoreactive cells and possible intrathecal anti-NMDAR antibody synthesis. The observations of this case would contribute towards ascertaining the mechanisms of anti-NMDAR encephalitis.

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Conflicts of interest

There are no conflicts of interest.

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