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CASE REPORT

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Intraventricular dysembryoplastic neuroepithelial tumor in the temporal horn with Broad involvement of the ependyma

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Key Clinical Message

Though typical dysembryoplastic neuroepithelial tumors (DNETs) are located in the cerebral cortex, an atypical DNET could occur in the temporal horn of the lateral ventricle and broadly involve the ependyma. Awareness of this atypical form of DNET is of value for the wright diagnosis and management of atypical DNETs.

KEYWORDS

DNET, dysembryoplastic neuroepithelial tumor, epilepsy, lateral ventricle, seizure, surgery

1 | INTRODUCTION

The intracortical location of dysembryoplastic neuroepithelial tumors (DNETs) has been a critical characteristic for diagnosis. We report a case of DNET in the temporal horn of the lateral ventricle. Awareness of this atypical form of DNET is of value for the wright diagnosis and management of atypical DNETs.

Dysembryoplastic neuroepithelial tumor (DNT, DNET) is a relatively newly identified and rare type of brain tumo,r which was first described around 3 decades ago.[1] DNET contains both neuro components and glial components histologically and is classified as a Grade 1 tumor by the 2016 World Health Organization Classification of tumors of the Central Nervous System. It has been related to abnormal embryological cortical development of the brain, and its occurrence is often linked to focal cortical dysplasia (FCD). This type of tumor resides typically in the brain cortex and often involves only one gyrus, with neuronal elements that form columnar structures perpendicular to the cortical surface.[2]

DNET is primarily found in patients with epilepsy. Confident diagnosis based on magnetic resonance imaging (MRI) can often be made due to its signal profile and its relationship with the cortex. With wright diagnosis, surgical treatment of typical DNETs could often achieve satisfactory seizure control.[3]

While the contemporary management strategy of typical DNETs is mainly straightforward, atypical ones could still pose diagnosis, and management challenges.[4] Intraventricular DNETs are very rare. Reported intraventricular DNETs are often located at the vicinity of the third ventricle, sometimes associated with hydrocephalus.[5] Given the established correlation of DNETs with epilepsy, wright noninvasive diagnosis of DNETs would facilitate the clinical decision for surgical treatment of the associated epilepsy.[6] In this respect, knowing that atypical forms of DNETs could occur inside the ventricular

system is of great value. This could be especially true for DNETs in the temporal horn, as lesions in the temporal lobe in general are more likely to be associated with epilepsy.

However, intraventricular DNETs located in the temporal horn of the lateral ventricle have not been reported in the literature. Therefore, in order to demonstrate that epileptogenic DNETs suitable for surgical treatment could also occur in the temporal horn, we report this surgical case of an intraventricular DNET in the temporal horn of the lateral ventricle.

2 | CASE

A 25-year-old man presented with two seizures within 2 months before admission. Both seizures occurred while the patient was awake and performing routine daily activities in a sitting position. The patient did not recall any aura. Seizure started with a sudden halt of ongoing activity, blank staring, and unresponsiveness to the witness who tried to communicate. The seizure then quickly evolved to right-hand automatism and left arm twisted posturing, which lasted for around 2 minutes. The patient then fell suddenly. Secondary generalized tonic-clonic attack occurred and lasted for approximately another 2 minutes. The interval between the two seizures was about 3 weeks. The two seizures were stereotypical in semiology and featured a right temporal lobe epilepsy. MRI with T1 (with and without gadolinium contrast) and T2, T2 fluid-attenuated inversion recovery (FLAIR) sequences were scanned. The image showed a polycystic-like lesion primarily located in the right temporal horn of the lateral ventricle, with broad involvement of the ependyma. The tumor demonstrated hypointensity signal on T1, without gadolinium enhancement. On T2, it showed mild hyperintensive signal. T2 FLAIR demonstrated mild hypointensive signal. The signal profile supported a diagnosis of dysembryoplastic neuroepithelial tumor, in spite of that the lesion was located in the temporal horn of the lateral ventricle, and primarily involved the ependyma rather than the cortex (Figure 1). Temporal lobe epilepsy secondary to an intraventricular DNET was therefore diagnosed. We performed a tumor resection with anterior temporal lobectomy as a treatment for the patient's epilepsy. A postoperative pathology study confirmed the diagnosis of a DNET (Figure 2). The operation went well with no postoperative deficit of language, memory, or other neurological functions. The patient was followed up and has been seizure free for 1 year since the operation.

3 | DISCUSSION

Intraventricular DNET is rare. Though large DNETs originated from the hippocampus may extend to the temporal horn of the lateral ventricle, they are free from the ependyma of its lateral wall and roof. MRI of the tumor is usually conspicuous enough to make a diagnosis.[3] Our case shows that DNETs also occur in the temporal horn and broadly involve the ependyma.

Around thirty intraventricular DNET cases have been reported so far. They were generally linked to the supratentorial middle line structures. Mostly reported types were described as tumors arising from septum pellucidum, caudate nucleus, and the third ventricle.[7,8] Anecdotal reports of DNET found

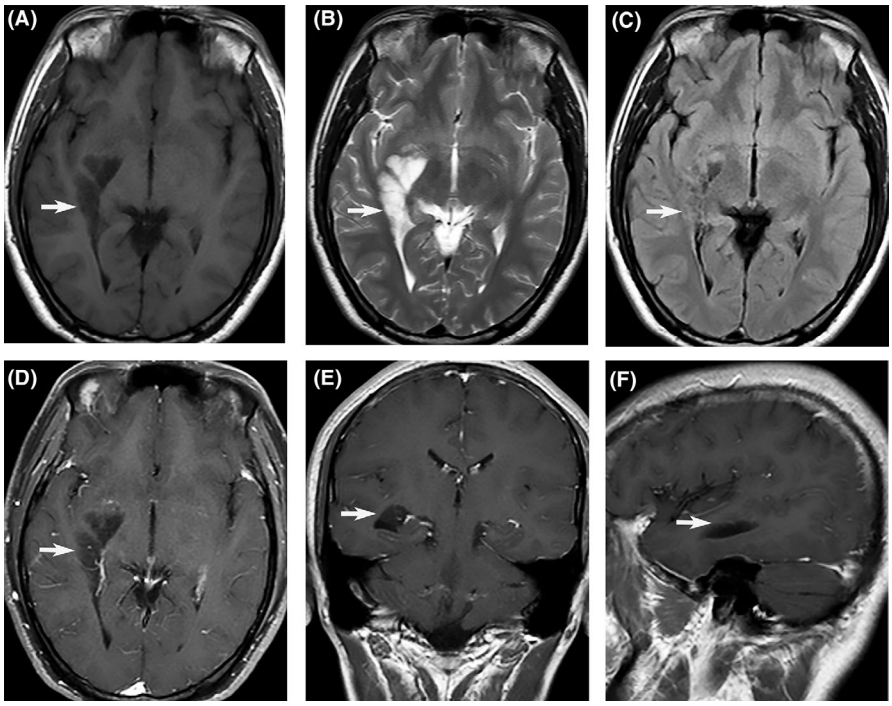

FIGURE 1 MRI showing an intraventricular dysembryoplastic neuroepithelial tumor in the temporal horn. MRI with T1 (A), T2 (B), T2 fluid-attenuated inversion recovery (FLAIR) (C) sequences, and T1 sequence with Gadolinium injection (D, E, F) showing signal features of an intraventricular polycystic-like dysembryoplastic neuroepithelial tumor (arrow), which extends to the trigone of the lateral ventricle along the roof and lateral wall of the temporal horn

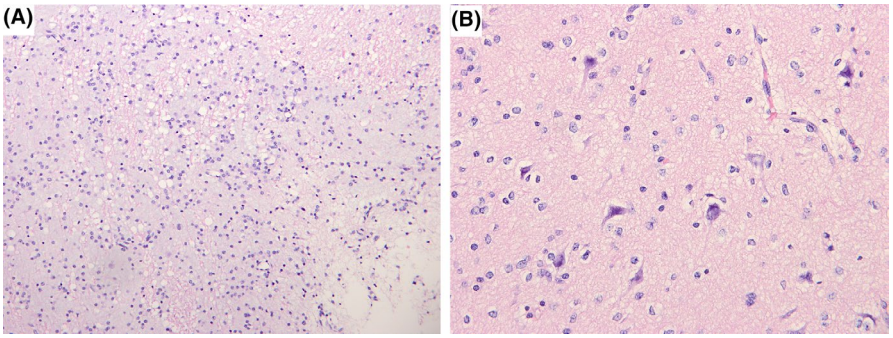

FIGURE 2 Histology of an intraventricular dysembryoplastic neuroepithelial tumor in the temporal horn. The small oligodendrocyte-like cells are present, but the typical columns as seen in cortical DNETs are not prominent (A, ×200); Mucin-filled microcysts and floating neurons in the mucin-filled tumor are evident, which support the diagnosis of a DNET (B, ×400)

in the anterior[9] and occipital horn[10] of the lateral ventricle also exist in literature. Even DNET with whole ventricle and spinal dissemination have also been reported.[11,12] However, we are not sure whether any case has been reported regarding an intraventricular DNET broadly involving ventricular wall and roof of the temporal horn. Building on the reports of intraventricular DNETs mentioned above, our case may thereby support the notion that intraventricular DNETs occurs in all parts of the supratentorial ventricular system.

The Relationship of DNET with the cortex is an important characteristic not only for imaging diagnosis but also for histological diagnosis. Intraventricular types of DNET lack this feature. However, the typical multicystic appearance is another feature of DNETs, and this feature pointed to the right diagnosis and management of our case.

Though evidence from high-quality RCTs specifically on surgical treatment of DNET-related epilepsy is not available, a Cochrane review has comprehensively reviewed available evidence on surgical treatments of epilepsy. This review concluded that abnormal preoperative MRI and the presence of tumor are prognostic factors associated with better postsurgical seizure outcomes.[13] This evidence supports surgical treatment of DNET-related epilepsy. The key lies in wright preoperative diagnosis of the tumor. Our case shows that an epileptogenic DNET could occurs in the temporal horn of lateral ventricle. Meanwhile, surgical treatment of this atypical DNET is safe and effective for seizure control.

4 | CONCLUSION

Epileptogenic DNET could occur in the temporal horn of the lateral ventricle. Awareness of this atypical form of DNET is of value for the wright diagnosis and management of atypical DNETs.

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CONFLICT OF INTEREST

None declared.

AUTHOR CONTRIBUTION

LL: wrote the manuscript. XH: collected the MRI images. JL: collected histological images. HZ: supervised and revised the report.

ETHICAL APPROVAL

This study is approved by our Institutional Review Board.

DATA AVAILABILITY STATEMENT

Data are available on request due to privacy/ethical restrictions.

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