Resection of a dysembryoplastic neuroepithelial tumor in the precentral gyrus

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Background: **Dysembryoplastic neuroepithelial tumors (DNTs) are common causes of intractable epilepsy in pediatric epilepsy patients. The effect of surgical intervention is often limited when the tumor is located in the precentral gyrus. Furthermore, complete surgical resection is often not performed in order to avoid permanent neurological deficits.**

Methods: **Here, we present a pediatric patient with intractable epilepsy caused by a simple DNT located in the precentral gyrus. Intracranial electrodes were implanted and used in combination with magnetic resonance imaging, video-electroencephalography and electrical cortical stimulation to assess neurological function, and where the epileptogenic zone was located.**

Results: **The results of intracranial electrode monitoring suggested that the epileptogenic zone was located in the tumor area and that cortical function had been reorganized. We completely resected the tumor based on these findings. The patient has been seizure free after the surgery and has not had any neurological deficits.**

Conclusions: **Simple form DNTs in the precentral gyrus can be completely resected with careful preoperative assessment of cortical function. Cortical reorganization could partly explain the functional preservation after surgery.**

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Key words: **cortical reorganization;**
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Introduction

Dysembryoplastic neuroepithelial tumors (DNTs) were first described by Daumas-Duport et al[1] in 1988 and are benign glioneuronal tumors. These tumors are usually located in the supratentorial cortex. There is a three-tier classification of DNTs based on pathological findings. The simple form consists of a specific glioneuronal element (SGNE). The complex form consists of an SGNE with associated glial nodules and focal cortical dysplasia (FCD).[2] Nonspecific form can be identified as a glial tumor with FCD characteristics but no SGNE.[3]

We report a pediatric patient with intractable epilepsy and a DNT in the precentral gyrus. This tumor was identified by neuroimaging and pathological studies. We discuss the epileptogenicity of DNTs and functional organization in the precentral gyrus based on the results of electrical cortical stimulation (ECS).

Case report

History and examination

The patient was a 6-year-old boy with intractable epilepsy since 6 months after birth. After entering the ward, we observed over 15 simple seizures each day by video-electroencephalography (EEG) monitoring. The seizures always occurred in the morning and presented as paroxysmal clonus and spasms of the patient's left face. The spasms extended to the upper arm. The patient was treated with 150 mg of oxcarbazepine (every 12 hours), 25 mg of lamotrigine (every 12 hours), and 1 mg of clonazepam each night. Neuropsychological examination showed borderline intelligence quotient and retardation of language development. The patient could only speak simple words such as "papa" and "mama". Physical examination was normal, and no abnormal facial or limb movements were noted. Magnetic resonance imaging (MRI) results suggested an abnormal signal in the right premotor area or precentral gyrus (Fig. 1). Thus, the preliminary diagnosis was DNTs. The video-EEG demonstrated slow waves in the right C4, P4 and midline electrodes.

Operation

The patient was subjected to craniotomy. At the first

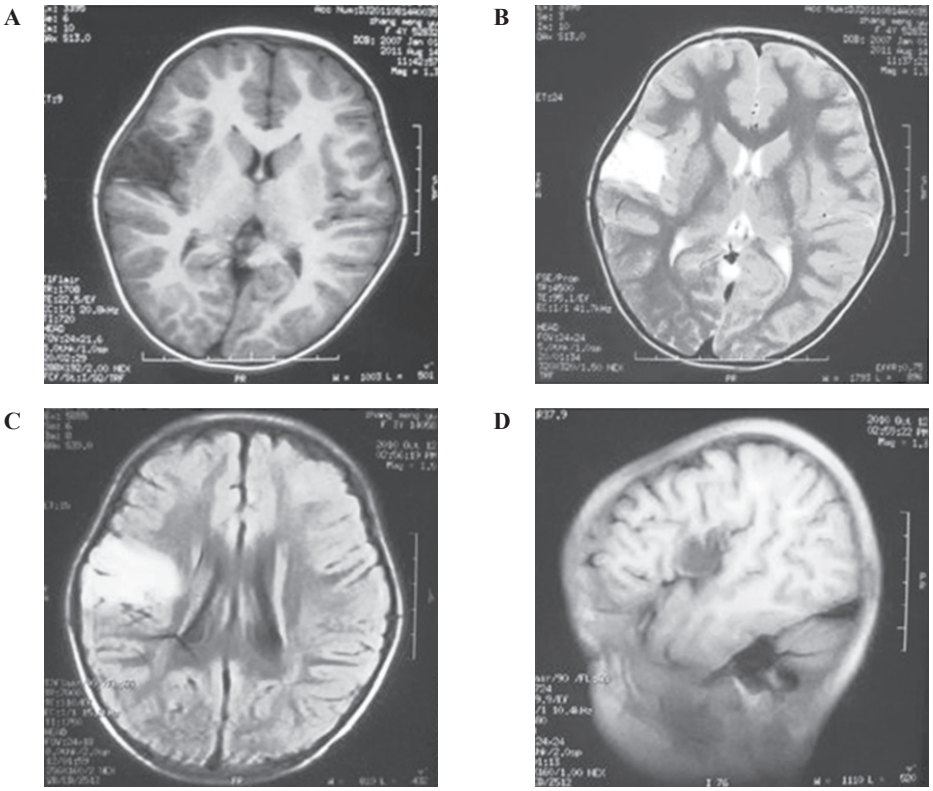

Fig. 1. MRI T1 (**A**), T2 (**B**), FLAIR (**C**), and axial (**D**) images of the patient. MRI changes in the precentral or central area corresponding to a type 1 (cystic or polycystic like) DNT. The lesion consisted of a well-delineated, strongly hypointense signal on T1 and a hyperintense signal on T2 and FLAIR. There was a well-defined gray-white matter demarcation. There was no occupying effect or peri-tumoral edema. These MRI changes usually indicate a simple form DNT.[5] MRI: magnetic resonance imaging; FLAIR: fluid attenuated inversion recovery; DNT: dysembryoplastic neuroepithelial tumor.

surgery, we confirmed that the suspicious tumor was located in the inferior precentral gyrus and had destroyed the normal structure and distorted the vessel distribution (Fig. 2A). We implanted intracranial electrodes to further localize the epileptic zone (EZ) and to define the function of the lesion and the surrounding cortex. We then conducted invasive EEG monitoring (IEM) and functional mapping with ECS. Low-amplitude fast activities were observed inside the suspicious tumor during IEM at the initiation of seizures (Fig. 2B). ECS was performed on the 7th day after the first surgery. The ECS parameters included a pulse width of 0.2 ms, a frequency of 50 Hz and an intensity of 1 mA-15 mA. This test demonstrated the lack of function in the lesion area (Fig. 2A). Based on these results, we performed a complete lesionectomy and carefully preserved the cortical movement function during the resection.

Postoperative course

The patient presented with left facial paralysis 2-3 days after the second surgery. There was also a slight involvement of the left arm. His arm and hand movements recovered within 10 days. The facial movements became normal after 3 weeks of follow-up in an outpatient clinic. The pathological diagnosis

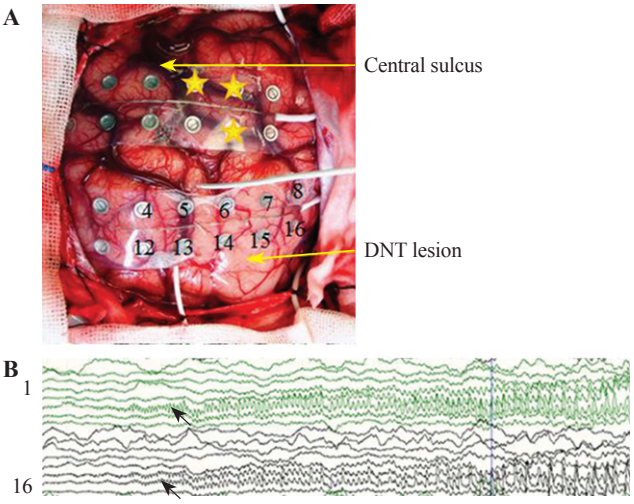

Fig. 2. A: The suspicious DNT lesion was located in the inferior central area and had a pale color. The tumor had destroyed the structure of the normal central gyrus and had clear margins with the normal gyrus. The vein system was distorted backward. We detected 10 seizure attacks after implanting the electrodes. Low-amplitude fast activities were detected in electrodes 6, 7, 14, 15. ECS detected left hand and forearm movements in the electrode with a yellow star and left facial movements in electrodes 4 and 12. The facial movement shifted backward from the central area; **B**: Low-amplitude fast activities can be seen within the DNT lesion (black arrow, corresponding to electrodes 6, 7, 14, 15 in Fig. 2A). DNT: dysembryoplastic neuroepithelial tumor; ECS: electrical cortical stimulation.

was a simple form DNT. A typical columnar architecture was identified in the cortex, and it was lined with oligodendroglial-like cells that were glial fibrillary acidic protein negative. Normal hexaribonucleotide binding protein-3-positive clusters of differentiation 34-negative neurons were scattered throughout the tumor. No changes were indicated for FCD. The patient has been seizure free for 2 years. He has discontinued all antiepileptic drug treatment and is performing well at school.

Discussion

DNTs defined as World Health Organization grade I[4] are neuronal-glial neoplasms affecting children and patients in early adulthood with drug-resistant epilepsy. Surgical series report favorable outcomes in 70% to 90% of cases.[5,6]

Before the surgery, we discussed the MRI characteristics of the lesion and concluded that it was a simple form DNT.[5] This diagnosis was confirmed pathologically after operation. The IEM results suggested that the epileptogenic activity originated from the tumor tissue in the form of low-amplitude fast activity, which was indicative of EZ localization.[7] The functional roles of DNTs in the primary motor cortex have been rarely reported. The ECS results of our patient suggested the absence of functional tissue inside the tumor. The facial movement cortex seemed to be reorganized from the precentral gyrus. No movement deficit was found during the initial physical examination, which indicated the possibility of a "functional reorganization" or "functional shift" of the motor cortex. Devaux et al[8] reported 7 patients with DNTs and one patient with ganglioglioma in the precentral area. They evaluated the cortical function by stereoelectroencephalographic exploration. In the seven patients, stimulation of electrodes within the tumor tissue did not elicit any motor response. After surgery, six (75%) patients became seizure free and no permanent deficits were observed. This result indicated a possible functional reorganization. We speculated that the lesion developed during an early embryonic stage. Thus, there is a possibility to reorganize the cortex structure and preserve function.

In our patient, preoperative MRI aided in locating the tumor and indicated the DNTs subtype, and IEM and ECS estimated epileptogenicity and functional reorganization. Hence even if the simple form DNTs are located in the primary motor area, there is a greater possibility of favorable seizure control after surgery. These tumors show a clear tumor borderline that permits complete tumor removal and preserves neurological function. In complex form and nonspecific form DNTs (with surrounding FCD), the epileptogenic area may involve the peri-lesional area or a remote area.[5,7] Besides, a functional MRI or magnetoencephalography may help demonstrate preserved function in lesions located in an eloquent area.[9]

The optimal time for surgical intervention in children with glioneuronal tumors remains unclear. Ramantani et al[10] studied 29 patients including 13 DNTs patients. They found that shorter seizure duration may lead to a better cognitive outcome. Pediatric patients can benefit from surgery because of a favorable seizure outcome and can discontinue the use of antiepileptic drugs. Furthermore, it is critical to perform surgery as soon as possible to avoid deterioration of cognitive function.

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Contributors: Li YJ contributed to the study design. Xue H undertook information collection, data analysis and literature writing. Sveinssen O was responsible for literature structure and language revision.

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