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Electroencephalography-Guided Resection of Dysembryoplastic Neuroepithelial Tumor

—Case Report—

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Abstract

A 3-year-old girl presented with a dysembryoplastic neuroepithelial tumor in the right cingulate gyrus manifesting as epilepsy refractory to anticonvulsant medication. Computed tomography and magnetic resonance imaging revealed a cystic tumor in the right cingulate gyrus. The tumor was removed under intraoperative electrocorticography guidance. Abnormal spikes recorded adjacent to the tumor disappeared immediately after total removal. Histological examination showed a multinodular, multicystic structure, satisfying the criteria for the diagnosis of dysembryoplastic neuroepithelial tumor. She has remained seizure-free for more than 4 years without complications. In this case, intraoperative electrocorticography was very useful to identify the possible focus and prevent unnecessary resection of the adjacent tissue. Total removal of the tumor resulted in a dramatic reduction of seizure activity.

Key words: cingulate gyrus, complex partial seizure, dysembryoplastic neuroepithelial tumor, surgery

Introduction

Dysembryoplastic neuroepithelial tumor (DNT) is one of the rarest brain tumors and was first described in 1988.[3] DNT is classified as a low-grade tumor, as World Health Organization (WHO) grade I, but causes intractable seizures as the tumor develops in the temporal or frontal lobe in childhood. These seizures are difficult to control with conventional anti-epileptic drugs, but surgical resection is more successful.[1,3,4] We describe a successful surgical treatment of intractable epilepsy caused by DNT with the clinical, neuroradiological, and histological findings.

Case Report

A 3-year-old girl had been born with no complications at the 36th week of pregnancy, weighing 2200 g, as the second child of dizygotic twins. Her development was normal, and her past history was unremarkable. She developed epileptic seizures characterized by violent tossing and turning in her bed, throwing her arms about, and disturbance of consciousness at the age of 2 years 8 months. The seizures continued for several seconds to minutes, initially twice or three times a day, increasing up to 10 times a day 1 month later.

On admission, physical and neurological examinations found no abnormalities. Interictal electroencephalography showed normal background activity, but ictal electroencephalography showed 3 Hz spike-and-wave complexes in the right parietal and central areas. Computed tomography (CT) showed a low density area without contrast enhancement in the right frontal region (Fig. 1A). Magnetic resonance (MR) imaging revealed a cystic tumor (3 cm in diameter) replacing the right cingulate gyrus (Fig. 1B–D). Single photon emission computed tomography (SPECT) with technetium-99m ethyl cysteinate dimer showed attenuated blood flow at the region corresponding to the tumor (Fig. 1E).

Epileptic seizures were not controlled well even after administration of full doses of valproic acid and carbamazepine. Surgical resection was performed through a right frontoparietal craniotomy. The tumor was soft and gelatinous, and was easily distinguished from the surrounding parenchyma. Meticulous intraoperative electrocorticography

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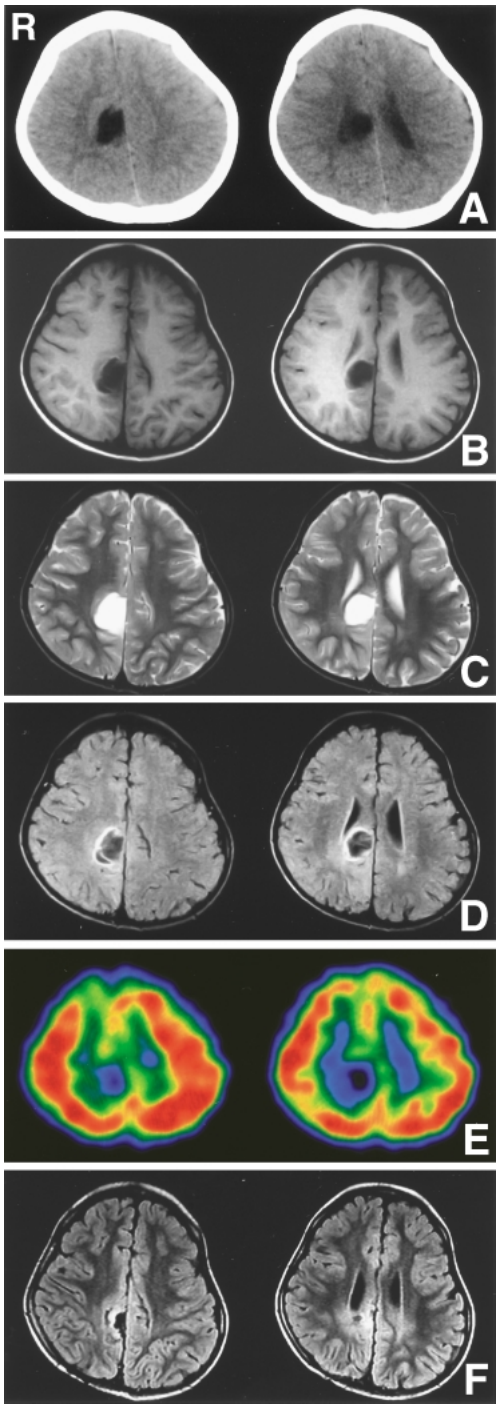

Fig. 1 **A–E: Preoperative computed tomography scans (A), T_1-weighted (B), T_2-weighted (C), and fluid-attenuated inversion recovery (D) magnetic resonance images, and technetium-99m ethyl cysteinate dimer single photon emission computed tomography images (E) showing a tumor mass in the right cingulate gyrus. F: Two years later, postoperative fluid-attenuated inversion recovery magnetic resonance images revealing evidence of resection of the tumor.**

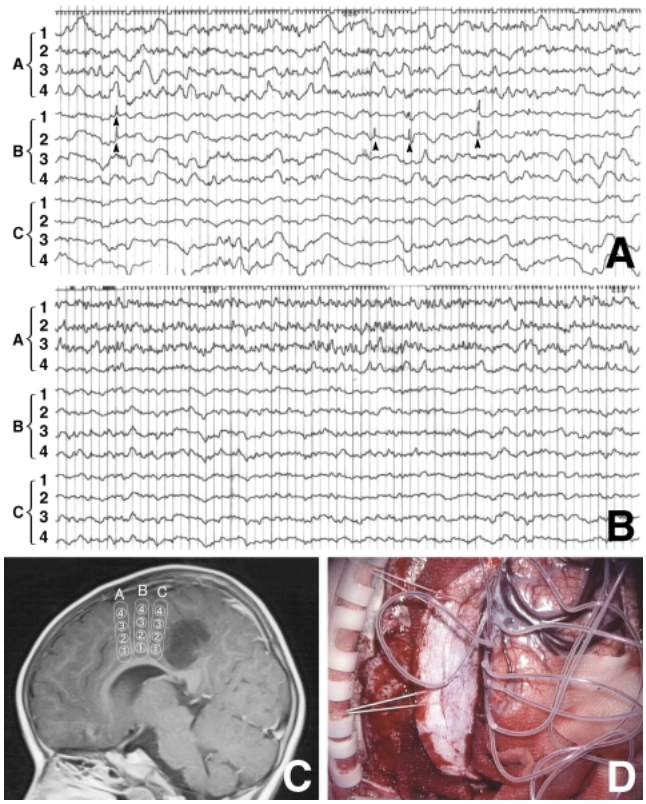

Fig. 2 **A, B: Intraoperative electrocorticograms demonstrating frequent spikes (*arrowheads*) from the right cingulate gyrus subjacent to the anterior side of the tumor (A), which immediately disappeared after resection of the tumor (B). C: T_1-weighted magnetic resonance image showing the spikes originated in regions B-1 and B-2. D: Intraoperative photograph of the electrocorticography procedure during the operation.**

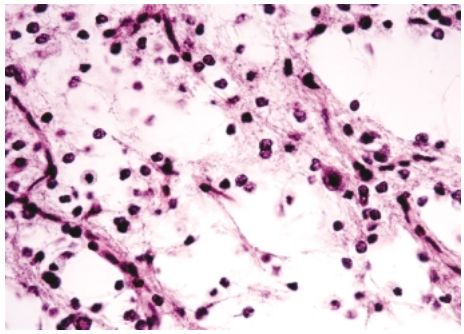

Fig. 3 **Photomicrograph of a characteristic area of the tumor demonstrating abundant mucinous matrix including oligodendrocyte-like cells and prominent neurons. Hematoxylin and eosin stain, original magnification ×400.**

(ECoG) showed frequent spikes only from the right cingulate gyrus adjacent to the anterior side of the tumor (Fig. 2A). These abnormal spikes disappeared immediately after the total removal of the tumor (Fig. 2B), so resection of the adjacent cerebral parenchyma was omitted. The patient's postoperative course was uneventful and she was discharged without complication.

Histological examination of the surgical specimen revealed an alveolar pattern, a mixture of oligodendroglia-like cells as well as neural and glia cells, with abundant mucinous matrix in the tumor (Fig. 3). The histological diagnosis was DNT. Follow-up MR imaging in the outpatient department 2 years after the operation revealed achoresis changes replacing the tumor (Fig. 1F). She has had no seizure for more than 4 years after the surgery.

Discussion

In the present patient, imaging procedures such as CT, MR imaging, and SPECT revealed the brain tumor in the right cingulate gyrus. Fluid-attenuated inversion recovery MR imaging is essential for the detection of DNT, as seen in this case, because of the adequate contrast with the adjacent cerebrospinal fluid.[13] Intraoperative ECoG showed spikes in the right cingulate gyrus adjacent to the tumor. Localization-related epilepsy, with the seizures originating in the cingulate gyrus and surrounding regions, is characterized by complex partial seizures accompanied by specific symptoms of partial dyskinesia such as sudden commencement of walking or running, violent body movements, and throwing about of the arms. These typical seizures in this patient, characterized by violent convulsions in bed accompanied by throwing about of the arms, and with disturbance of consciousness, were almost certainly epileptic seizures originating from the region around the brain tumor in the cingulate gyrus. These seizures disappeared immediately after tumor removal and focal resection was performed at an area adjacent to the tumor. Surgical removal of the lesion is curative in most patients with DNT, and no additional chemotherapy or radiotherapy is needed, so accurate diagnosis and surgical treatment is encouraged.[3,6]

Complete tumor resection is the most important predictor of seizure-free outcome in patients with DNT.[12] However, resection limited to extratemporal lesionectomy, as performed in our case, may spare the residual epileptogenic cortex.[2] Intraoperative ECoG identified the possible focus and prevented unnecessary resection of the adjacent tissue. This sparing of tissue is especially important in or near the eloquent cortex. Intraoperative ECoG can also replace invasive monitoring, which is not indicated for patients who cannot tolerate long-term recording.

The effects of the type and concentration of anesthetics remain controversial for accurate diagnosis of the focus. Sevoflurane with 1.5% mean alveolar concentration (2.5% end-tidal concentration) is now believed to be reliable because the intraoperative spikes are most concordant to the ictal onset zone recorded by invasive monitoring.[5,11]

DNTs are generally considered as benign tumors, but recurrence or rapid increase in tumor size sometimes occur. Therefore, accurate diagnosis and surgical treatment are essential. In our case, the histological findings fulfilled the criteria for the diagnosis of DNT. According to the WHO classification of brain tumors revised in 2000, DNT is included in the neuronal and mixed neuronal-glial tumors of the central nervous system, and is characterized by a multinodular multicystic structure, containing glial and neuronal components with minimal atypia and cortical dysplasia in the adjunct area.[7] Therefore, DNT is considered a hamartoma but is generally classified as a low-grade glioma. The differential diagnosis of DNT includes ganglioma, oligodendroglioma, oligoastrocytoma, and benign cerebral cysts.[3,4,6–10,14,15]

Long-term follow up and observation are essential. Our patient has had an uneventful clinical course for more than 4 years after surgery, with no neurological sequelae and no recurrence of seizures.

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