RAPIDLY GROWING DYSEMBRYOPLASTIC NEUROEPITHELIAL TUMOR: CASE REPORT

**OBJECTIVE:** During the past 15 years, the concept of dysembryoplastic neuroepithelial tumors has continued to evolve. We present an interesting case of dysembryoplastic neuroepithelial tumor that showed rapid growth during a short period of time.

**CLINICAL PRESENTATION:** A 9-year-old boy had been experiencing intractable complex partial seizures since the age of 7 years. Magnetic resonance imaging scans demonstrated a well-demarcated 3.5-cm lesion with a 1.5-cm ring-enhanced core in the left temporal lobe. One month later, the lesion had rapidly grown to occupy three times more space than on the first evaluation, with the ring-enhanced core reaching approximately five times its initial volume.

**INTERVENTION:** A combined tumor removal and epileptogenic focus resection surgery was performed immediately. In the pathological examination, the presence of the specific glioneuronal element with a Ki-67 labeling index of lower than 1%, as well as the glial component with a Ki-67 labeling index of 8%, led to a postoperative diagnosis of dysembryoplastic neuroepithelial tumor, complex form. No adjuvant therapy was performed. Five years after surgery, there is no evidence of any recurrence and the boy continues to be seizure free without antiepileptic drugs.

**CONCLUSION:** The lesion did not behave as a stable benign entity as it is generally accepted, and is, therefore, presented as an argument in favor of an early and complete resection.

**KEY WORDS:** Benign tumor, Dysembryoplastic neuroepithelial tumor, Glial component, Temporal lobe epilepsy

**CASE REPORT**

A 9-year-old, right-handed boy had been experiencing intractable complex partial seizures since the age of 7. His seizures were characterized by apnea during sleep, with no response to wakening, sometimes followed by stiffness of all extremities.

Despite more than 2 years of treatment with various anticonvulsants, he continued to experience seizures several times a week. The comprehensive physical examination was negative for peripheral stigmata of phakomatosis, congenital malformations, and focal neurological signs.

Magnetic resonance imaging (MRI) scans demonstrated a well-demarcated 3.5-cm lesion in the left temporal lobe. The lesion had no surrounding edema and appeared hyperintense on T2-weighted scanning (Fig. 1B). On
T1-weighted scanning, the lesion was hypointense and had a 1.5-cm ring-enhanced core (Fig. 1, A and C). The left hippocampus showed no signs of atrophy and no abnormal signal.

Repeat electroencephalograms were positive for interictal epileptiform discharges in the left temporal lobe. Ictal discharges started from the mid- and post-temporal lobe. Interictal single-photon emission computed tomography showed hypoperfusion of the tumor.

During 1 month of preoperative investigations, the lesion rapidly grew to occupy three times more space than on the first evaluation (Fig. 1, D and E). The ring-enhanced section, although continuing to be contained within the hypointense part, reached approximately five times its initial volume (Fig. 1F).

A combined tumor removal and epileptogenic focus resection surgery was immediately performed. The tumor was entirely located within the left temporal lobe and did not involve the middle cerebral artery. We found two different components in the tumor. One was soft, red tumor containing yellowish fluid, but containing no hemorrhage, which corresponds to the ring-enhanced core and intratumoral cyst on MRI scans. The other part was slightly hard tumor that existed surrounding the soft, red tumor, which corresponds to well-demarcated and non-enhanced mass lesion. After tumor removal, we removed the surrounding epileptogenic cortex, which showed spikes on intraoperative electrocorticography. The hippocampus, which showed no spikes on intraoperative electrocorticography, was spared.

Five years after surgery, there is no evidence of any recurrence and the boy continues to be seizure free without anticonvulsants. He has no neurological deficits, his intelligence has not been affected, and he is developing normally and according to his age group.

**Pathological Findings**

On pathological examinations, the specimen from the section that appeared hypointense on the T1-weighted image proved to be abundant in microcystic nodules containing a mucinous matrix (Fig. 2A). The microcysts exhibited alveolar appearances with bands of oligodendrocyte-like cells (OLCs) (Fig. 2B). So-called floating neurons, which showed synaptophysin immunoreactivity (Fig. 2C), were occasionally observed in the microcysts. The Ki-67 labeling index was lower than 1%.

**FIGURE 1.** MRI scans at admission showing that the lesion is a hypointense mass on the T1-weighted image (A) and a hyperintense mass on the T2-weighted image (B). The T1-weighted image with gadolinium reveals a ring-enhanced core (C). MRI scans taken 1 month later show a threefold volume increase on both T1-weighted (D) and T2-weighted (E) scans. The ring-enhanced core increased fivefold (F). MRI scans taken 5 years later show a dead space in the left temporal tip (G, H) and that tumor recurrence was not observed (I).
The tumor consisted of many mucinous microcystic nodules showing alveolar appearances (A, hematoxylin and eosin; original magnification, ×33). B, so-called floating neurons in the cystic nodules were outlined by OLCs (hematoxylin and eosin; original magnification, ×66). C, floating neurons were immunoreactive for synaptophysin (original magnification, ×66). D, septum-like vascular proliferation in the ring-enhanced lesions (hematoxylin and eosin; original magnification, ×132). E, vascular proliferation with endothelial proliferation was noted (hematoxylin and eosin; original magnification ×66).

In contrast, the microscopical examination of the ring-enhanced section revealed a high density of glial cells and pronounced vascular (Fig. 2D) and endothelial proliferation (Fig. 2E), with a Ki-67 labeling index of 8%. The presence of the specific glioneuronal element led to a postoperative diagnosis of DNT, complex form.

**DISCUSSION**

The pathological findings in our patient were similar to DNTs of previous reports (1, 2). A DNT, regardless of its subtype, has a myxoid matrix and multinodular architecture. The simple form of a DNT consists mainly of the “specific glioneuronal element,” bundles of axons oriented perpendicular to the cortical surface and lined by a population of OLCs.
The majority of OLCs, along with the normal neurons floating in the pale, eosinophilic matrix, distinguish DNTs from gangliogliomas and glioneuronal hamartomas, which include highly differentiated ganglion cells and bizarre neurons. Even the complex form, with additional glial nodules and foci of cortical dysplasia, can be differentiated from gangliogliomas by the lack of abundant connective tissue stroma and perivascular lymphocytic infiltration.

**Pathological Characteristics of the Present Case**

The surgical specimen in this case exhibited the “specific neuronal element,” as well as regions of glial proliferation. Both within and between microcysts, there were OLCs with small, uniform, hyperchromatic nuclei and scant cytoplasm. They failed to express glial fibrillary acidic protein, and the Ki-67 index was less than 1%. The ring-enhanced section, with glial fibrillary acidic protein-positive areas and a Ki-67 index of 8%, had the features of a Grade III astrocytoma. Previous reports have classified DNTs with polymorphic parts as “nonspecific forms of DNT,” or DNTs “resembling astrocytomas” (2).

However, in our case, the “nonspecific” section was completely contained within the DNT and had a clinical growth rate that indicated a high mitotic activity. We thought that the rapid growth of tumor was mainly caused by cyst enlargement within the glial component. We, therefore, considered it as a distinct malignant entity.

**Clinical Importance of the Present Case**

As an epilepsy-associated tumor, DNT has a double therapeutic goal: the removal of the tumor and the control of the epilepsy (8, 9). It is generally accepted that even subtotal tumor removal is effective for seizure control and that radical total removal or postoperative adjuvant therapy are usually not indicated in patients with a DNT. In our case, we discussed the clinical treatment of the glial section that resembled a Grade III astrocytoma. Because our patient was a young boy who is susceptible to side effects of radiation therapy and chemotherapy, we decided to follow the patient carefully without adjuvant therapy. After a 5-year follow-up period, we are now convinced that total removal of the lesion along with the surrounding epileptogenic cortex was a necessary treatment.

Considering that, for DNTs, the patient population consists mainly of adolescents and young adults, avoiding neoadjuvant therapy with its sometimes deleterious side-effects is just as important as avoiding recurrence. With only a few known cases of composite tumors (4, 10) or malignant transformations (3, 11), the obvious choice would be the most minimally invasive treatment. However, if malignancy is a possibility, complete excision is desirable. Studying the clinical behavior of DNTs may, as in this case, refine the criteria for surgical treatment.

**CONCLUSION**

This case is an example of a rapidly growing DNT in its complex form. The lesion did not behave as a stable benign entity, as is generally accepted, and is, therefore, presented as an argument in favor of an early and complete resection.

**REFERENCES**


**Acknowledgment**

We thank Dr. Takashi Komori (a tumor neuropathologist at Department of Clinical Neuropathology, Tokyo Metropolitan Institute for Neuroscience and a consultant neuropathologist in the Japanese Society of Neuropathology) for his support of pathological diagnosis in this case.

**COMMENTS**

W e continue to learn more about dysembryoplastic neuroepithelial tumors (DNTs). In essence, DNTs are a newly recognized brain tumor that is frequently associated with epilepsy. The removal of a DNT has frequently, but not always, been associated with a good outcome. There are a few reports in the literature of DNTs that behave in a more aggressive fashion, and the present case fits into this category. The authors have convincingly shown a DNT that increased in size dramatically over a short period of time, and the removal of this lesion in toto, however, was associated with a good outcome.

It may not be surprising that DNTs behave differently in some situations. Exactly what predisposes a DNT to behave in an aggressive manner is still not known. The authors’ report provides an impetus for treating neurosurgeons to re-image patients at intervals that would be compatible with detecting growth changes that may occur in such cases.

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The authors present a case of a young boy with a rapidly growing tumor that, at the time of resection, was reported by the pathologist as being a DNT. This is a most unusual behavior for a DNT, and the authors explain the rapid growth of the tumor due to cyst enlargement within the glial component. Clearly, our understanding of DNTs continues to evolve, and this particular case adds to our understanding of this entity.

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The authors report an interesting case of a dysembryoplastic neuroepithelial tumor (DNT) with anaplastic features, which progressed significantly over a short time interval. They argue, appropriately, that this case is important to highlight because it demonstrates an unusual biological behavior for DNTs. These tumors are generally considered stable, low-grade lesions. The 9-year-old boy’s tumor, as presented here, clearly did not behave this way, and these authors from Tokyo present an argument in favor of an aggressive surgical approach. This case report adds to the growing literature on DNTs, which now also includes other recent reports, also cited by the authors, demonstrating possible malignant transformation of these generally indolent neoplasms. The pathology of the tumor here clearly revealed two distinct components: one with the characteristic features of DNT and a second resembling anaplastic astrocytoma contained within the first. The authors should be congratulated for adding to our understanding of the range of potential biologic behaviors of these increasingly recognized neoplasms.

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