

Dysembryoplastic Neuroepithelial Tumors: MR Appearance

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PURPOSE: Dysembryoplastic neuroepithelial tumor (DNT) is a newly described, pathologically benign tumor arising within the supratentorial cortex and having a 100% association with partial complex seizures. We reviewed the MR appearance of the brains of six patients with DNT, without and with administration of gadolinium-DTPA, emphasizing the clinical and radiologic features. **METHODS:** The MR images in six patients (five male, one female) with pathologically proved DNT were reviewed. Five had partial complex seizures and one had partial simple seizures. Age at onset of partial seizures ranged from 2 to 19 years. Scans were obtained with conventional T1 and T2 weighting without and with the administration of gadolinium-DTPA. All patients had craniotomies for medically refractory seizures and pathologic examination of all specimens was available for review. **RESULTS:** MR demonstrated a focal cortical mass in all patients. Five were located in the temporal lobe and one was located within the occipital lobe. Two showed enhancement with gadolinium-DTPA. Calcification occurred in one lesion. Common features included very low signal intensity on T1-weighted images and high signal on T2-weighted images, similar to cerebrospinal fluid. Proton density images demonstrated slightly higher signal intensity in the lesion than cerebrospinal fluid. The margin of the tumor is well-circumscribed and may remodel the adjacent calvarium. **CONCLUSIONS:** DNT, a newly described pathologic entity resulting in chronic, often medically, intractable seizures, has characteristic features on MR that allow it to be suggested in the differential diagnosis. These lesions may simulate benign cysts; however, increased signal intensity on proton density images should prompt further investigation in the proper clinical setting. Differentiation from low-grade astrocytomas and ganglioglioma is not possible by MR.

Index terms: Brain neoplasms, magnetic resonance; Brain neoplasms, computed tomography; Seizures

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Neoplasms account for approximately 15% of all temporal lobe epileptogenic foci (1). Dysembryoplastic neuroepithelial tumor (DNT) has been described as a neoplasm associated with partial complex seizures (2). These tumors are patholog-

ically benign and occur within the cortical regions of the brain, typically in the temporal lobes (2). The computed tomography (CT) appearance of these lesions has been previously reported (2). The magnetic resonance (MR) experience with these lesions is limited (2). We reviewed retrospectively the noncontrast and postcontrast MR examinations of six patients who had pathologically proved DNTs.

Materials and Methods

The MR studies of seven patients and the CT studies of three patients who had pathologically proved DNT were reviewed retrospectively by the authors. Five of the patients were male and two were female. The age at onset of the partial seizures ranged from 2 years to 19 years. The age at the time of imaging ranged from 7 years to 28 years. One patient, a 12-year-old boy with an 8-year history of seizures and pathologically proved DNT, was excluded from the study because no preoperative post-gadolinium images

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TABLE 1: Patient data in six cases of dysembryoplastic neuroepithelial tumors

Patient No.	Sex	Age	Age of Onset	Location	Signal Intensity			Enhancement
					T1W	PDW	T2W	
1	F	18 yr	15 yr	Temporal	hypo	hyper	hyper	None
2	M	26 yr	19 yr	Temporal	hypo	hyper	hyper	Mild (rim)
3	F	22 yr	7 yr	Occipital	hypo	hyper	hyper	Minimal?
4	M	28 yr	3 yr	Temporal	iso	hyper	hyper	None
5	M	7 yr	2 yr	Temporal	hypo	hypo	hyper	None
6	M	10 yr	2 yr	Temporal	hypo	hypo	hyper	None

Note.—T1W = T1-weighted; T2W = T2-weighted; PDW = proton density-weighted; hyper = hyperintense to gray matter; hypo = hypointense to gray matter; iso = isointense to gray matter.

were obtained. The patients' sex, age of onset of symptoms, and imaging characteristics are summarized in Table 1. Five patients presented with partial complex seizures and one patient presented with partial simple seizures. All were refractory to medical therapy and had craniotomies for surgical resection of epileptogenic foci. Since DNT is a new classification, and since there may be considerable difficulty in distinguishing this neoplasm from other neoplasms (especially low-grade astrocytomas and ganglioglioma) the surgical specimens were "double-reviewed," once by our institution's neuropathologists and secondly by a separate institution's neuropathologists.

All patients had MR scans obtained at 1.5 T (GE Signa, Milwaukee, WI) using 256 × 192 or 256 matrix. Sequences included sagittal 4- or 5-mm (1-mm gap) spin-echo (SE) 600/20/1 (TR/TE/excitations) images followed by axial 5-mm (1- or 2.5-mm gap) SE 600-800/11-20/2 images. Axial SE 2000-3000/30-35,70-90/1-2 images with 5-mm section thickness (2.5-mm gap) were obtained. Coronal SE 400-800/20/1-2 images with 4- or 5-mm section thickness (1-mm gap) images precontrast were obtained. Following contrast (Magnevist, Berlex, Cedar Knolls, NJ) injection, axial 5-mm (1- or 2.5-mm gap) SE 500-800/20-30/2 and coronal SE 400-800/20/1-2 images with 4- or 5-mm thickness (1-mm gap) were obtained. Coronal SE 2069-2353/30,80/1 gated images with 5- or 6-mm thickness (2.5-mm gap) were obtained in two patients. Coronal SE 2000-2069/30-35,70-80/1-2 nongated images with 5- or 6-mm thickness (2.5 mm gap) were obtained in two patients.

Representative Cases

Case 1

An 18-year-old girl had partial complex seizures for 3 years. The episodes were characterized by garbled speech and left-hand numbness, lasting 30 to 45 seconds. A CT scan demonstrated a nonenhancing hypodense mass of the right temporal lobe cortex (Figs. 1A and 1B). An MR scan showed a well-defined cortical right temporal lobe mass with increased signal intensity compared to gray matter on proton-density (PDW) and T2-weighted (T2W) images (Fig. 1C) and decreased signal intensity on (T1-weighted) T1W images. No enhancement after administration of gadolinium-DTPA was noted (Fig. 1D). A cerebral angiogram with bilateral selective internal carotid injections of amobarbital

(Wada test) demonstrated normal vascularity without mass effect. Her neurologic examination was normal. A right frontotemporal craniotomy was performed. While cortical electroencephalogram (EEG) electrodes demonstrated normal activity, intraoperative depth electrodes confirmed the site of the lesion. Grossly, a single area of the superior temporal gyrus was softer and had a salmon-colored appearance compared to the remainder of the gyrus. A near total resection of the mass was achieved. Frozen section of the resected specimen was consistent with protoplasmic astrocytoma.

Postoperatively, the patient did well without any seizure activity. Following review of the resected specimen, the final pathologic diagnosis was DNT based on the presence of a unique specific glioneuronal element and the history of partial complex seizures.

Case 2

A 26-year-old right-handed man presented with a history of complex partial seizures since 19 years of age. They were associated with stress and sleep deprivation. There were no associated headaches. The seizures began with an aura of *deja vu*, epigastric fluttering, lightheadedness, followed by clenched teeth, biting of the right cheek, staring, and drooling. Later in his course, he began to take progressively longer to clear postictally. He denied any history of head trauma or drug use. Family history was unremarkable. The initial work-up included a CT scan that showed a right hippocampal mass. Initial therapy included dilantin that poorly controlled his seizures, which averaged approximately two episodes per month, often within 24 hours of each other. Carbamazepine (Tegretol, Ciba-Geigy, Basel, Switzerland) and valproic acid (Depakene, Abbott, North Chicago, IL) were added to the patient's regimen without improvement and his job performance was suffering. An MR scan showed a right hippocampal mass that was hyperintense to gray matter on PDW and T2W images and hypointense to gray matter on T1W images with a 1.2 × 1.2 × 0.75 cm peripheral focus of extremely low signal intensity consistent with either hemorrhage or calcification (Figs. 2A and 2B). Mild rim enhancement and enhancement of the adjacent tentorium with gadolinium-DTPA was noted (Fig. 2C). Upon referral to our institution, he underwent video telemetry with tapering of anticonvulsant therapy

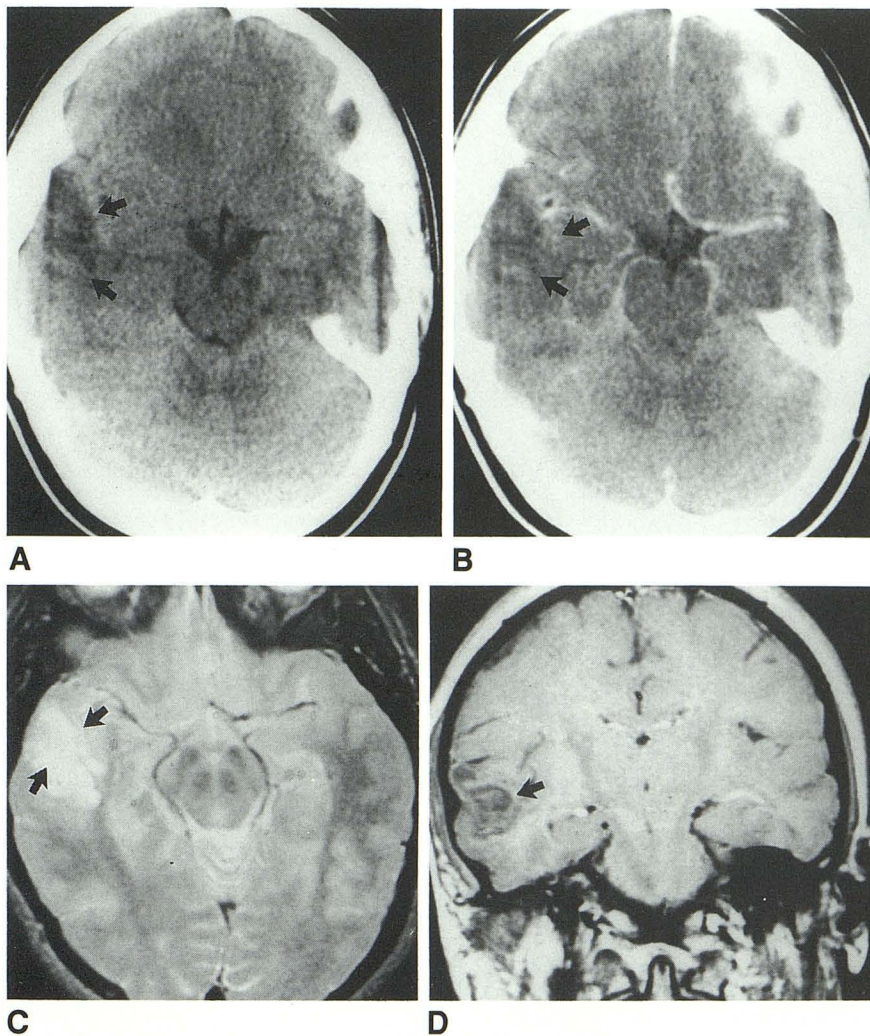


Fig. 1. Case 1; 18-year-old girl with 3-year history of partial complex seizures. (Courtesy of Santa Rosa Memorial Hospital, Santa Rosa, CA.)

A and B, Nonenhanced (A) and enhanced (B) CT scan sections show hypodense nonenhancing mass of right temporal lobe cortex (arrows).

C, Axial SE 3000/80 section through right temporal lobe shows well-defined cortical mass with signal intensity higher than that of gray matter (arrows).

D, Coronal SE 800/20 postcontrast MR shows a hypointense mass (arrow) without evidence of enhancement.

and had a single seizure with abnormal EEG activity from the right mesotemporal area. After having a cerebral angiogram with Wada test, which showed normal vascularity, he underwent a craniotomy and nearly total resection of the right temporal lobe mass. A rock-hard nodule measuring approximately 1 to 1.5 cm \times 0.75 cm was noted along the periphery of the mass at the time of surgery. The mass had firm lateral margins and a friable vascular medial margin. Intraoperative EEG recording demonstrated frequent epileptogenic spikes from the lateral and inferior cortex of the right temporal lobe, from the depth electrodes before resection, and showed enhancement following injection of methohexital for cortical mapping. Following partial resection, no epileptogenic activity was evident. With the exception of mild clinical depression that resolved prior to discharge, the patient did well postoperatively without any further seizure activity. He has been followed for 11 months since surgery and has been seizure-free.

Pathologic examination of the surgical specimens revealed cortical nodules with a columnar arrangement of oligodendrocytes and neurons and surrounding cortical dysplasia. The slides were independently reviewed by a

separate institution's neuropathologists who confirmed the diagnosis of DNT.

Case 3

A right-handed 22-year-old woman had frequent intermittent partial simple seizures marked by blurred vision, dizziness, eye deviation, and occasional clenching of teeth. Each episode lasted approximately 30 seconds and the patient had significant fatigue postictally. Vision was otherwise normal interictally. The episodes were associated with exposure to bright lights. Therapeutic levels of Tegretol failed to control the seizures. An EEG showed mild diffuse slowing and occasional sharp waves bitemporally. An initial MR scan was reported as normal, but a follow-up MR 2 years later demonstrated a well-margined cortical mass in the left occipital lobe that was hypointense to gray matter on the T1W images and hyperintense on the T2W images (Fig. 3A). Minimal postcontrast enhancement along the medial margin of the mass was noted (Fig. 3B). A cerebral angiogram with Wada test showed normal vascularity without mass effect or abnormal blush.

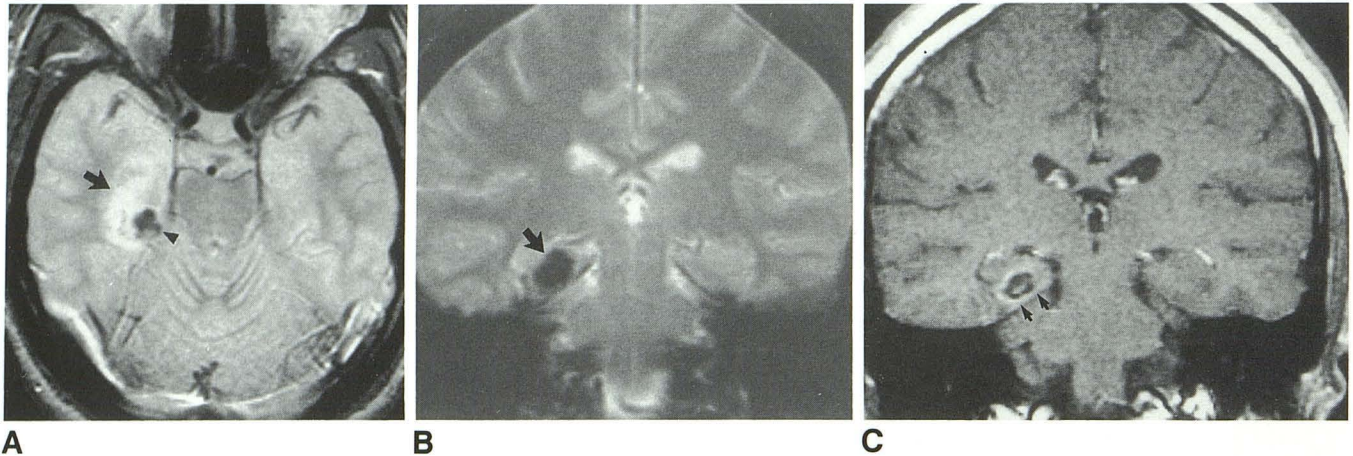


Fig. 2. Case 2; 26-year-old man with partial complex seizures for 8 years.

A, Axial SE 2800/30 section shows region of hyperintensity (arrow) in the medial right temporal lobe (arrow) with a peripheral area of marked decreased signal intensity (arrowhead).

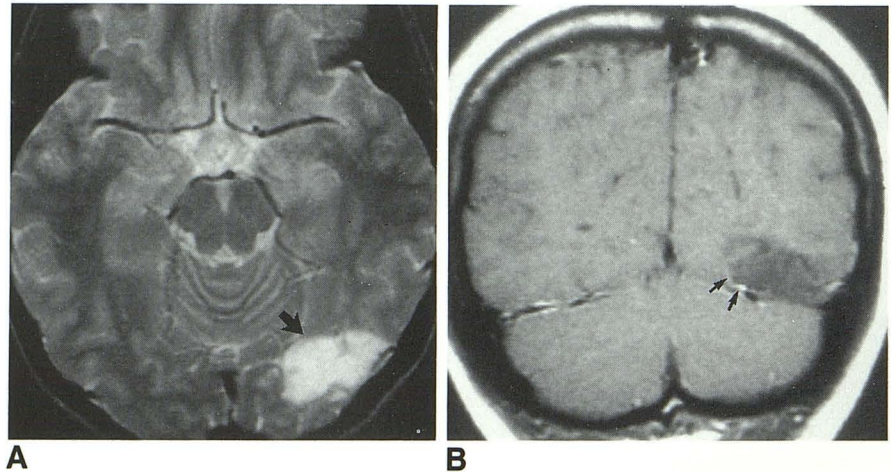
B, Coronal multiple planar gradient recalled 500/35 section through temporal lobes shows a $1.2 \times 1.2 \times 0.75$ cm well-defined area of marked hypointensity of the periphery of the mass (arrow). This reflects magnetic susceptibility artifact and the location and dimensions correlated with calcification noted within the pathologic specimen.

C, Coronal SE 400/20 section (postcontrast) shows a focal area of marked hypointensity with a surrounding rim of enhancement and enhancement of adjacent tentorium (arrows).

Fig. 3. Case 3; 22-year-old woman with a 15-year history of partial simple seizures.

A, Axial SE 2800/80 section through occipital lobes shows a well-margined hyperintense mass in the left occipital cortex (arrow).

B, Coronal SE 600/20 postcontrast section posterior to the ventricular atria shows minimal enhancement along medial margin (arrows).



A left temporoparietal occipital craniotomy revealed a mass which was paler than adjacent brain at the base of the temporo-occipital region near the tentorium. Intraoperative EEG evaluation revealed epileptogenic activity immediately superior to the lesion with little or no activity at any distance from the mass. Adjacent large veins made total resection of the mass impossible. The resected specimen had two components, one very soft and friable and the other somewhat firmer. The superior and medial margins of the mass blended imperceptibly into the surrounding white matter in the region of the optic radiations. Following resection of the mass, EEG revealed no residual epileptic activity.

Technical difficulty prevented an en bloc resection. This, in addition to the friable nature of part of the mass, caused fragmentation of the specimen. Therefore, pathologic examination of the resected specimen showed only a glioneu-

ronal component, one of the characteristics of DNT, in one of the fragments. Given the long history of seizures and the pathologic appearance, the diagnosis of "probable DNT" was made.

Postoperatively, the patient has been seizure-free.

Results

All six patients demonstrated a single focal cortical abnormality on MR scan. Five lesions were located in the temporal lobe and one was located in the occipital lobe. Each lesion was well-margined. Five of the six were hypointense to gray matter on T1W images. A single lesion (case 4, Fig. 4A) was isointense to gray matter on T1W images. Four lesions were hyperintense to gray

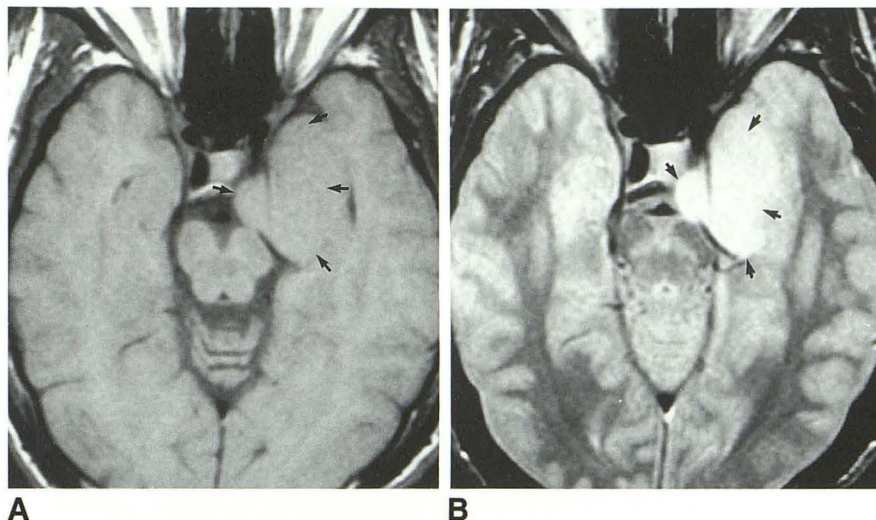


Fig. 4. Case 4; 28-year-old man with partial complex seizures since 3 years of age.

A, Axial SE 600/30 noncontrast section shows a left temporal isointense and slightly exophytic mass (arrows). This was the only case of DNT that demonstrated isointensity on T1W imaging.

B, Axial SE 2800/30 section at comparable level shows hyperintensity (arrows) of a well-defined cortical left temporal lobe mass, consistent with DNT.

matter and two lesions were hypointense to gray matter on PDW images. All lesions were hyperintense to gray matter on T2W images. One lesion (case 2, Fig. 2C) mildly enhanced in a rim-like fashion following the administration of gadolinium-DTPA. This lesion also demonstrated magnetic susceptibility phenomenon that was confirmed on pathologic examination to be calcification. Another lesion (case 3, Fig. 3B) showed minimal enhancement. Four patients had cerebral angiograms and Wada tests. Normal vascularity without mass effect or abnormal blush was noted in all four.

Discussion

Epilepsy occurs in approximately 1% of the general population (3). One third of these patients have epileptogenic foci within the temporal lobes and of these, about half are medically refractory (3-5). Since the 1930s, surgical resection of the temporal lobe in patients with medically refractory epilepsy has evolved from subpial aspiration to en bloc resection of the temporal lobe, which affords pathologists adequate tissue for more refined histologic diagnosis (3, 4). This procedure is a proven therapeutic measure in patients with medically refractory temporal lobe epilepsy, with approximately 50% of operative patients being seizure-free and another 25% having at least better seizure control (5-7). In general, patients are operative candidates when several criteria are met. These include a clinical diagnosis of temporal lobe seizures (often with video telemetry), seizure activity despite documented therapeutic anticonvulsant serum levels, decreasing cognitive-intellectual performance, and documented

epileptogenic focus within the temporal lobes on EEG. Prior to surgery, identification of the dominant hemisphere is often confirmed by means of a selective internal carotid amobarbital test (Wada test). The advent of CT and MR imaging has allowed the radiologist to identify mass lesions within the temporal lobe. MR has been shown to be superior to CT in the evaluation of temporal lobe seizures (8-11).

Pathologic study has revealed that mesial temporal sclerosis is the most common abnormality in patients with medically refractory temporal lobe epilepsy (1, 12, 13). Focal mass lesions of the temporal lobe occurs in about 24% of cases. These include gliomas (52% of mass lesions), nonspecific cerebral injury secondary to infection or trauma (24%), vascular malformations (9%), hamartomas (7%), nonglial tumors (5%), and tuberous sclerosis and formes frustes (2%) (1).

DNT is a benign tumor frequently associated with medically refractory partial complex seizures (2). Our study evaluated six patients, five with partial complex seizures and one with partial simple seizures who had pathologically proved DNT. None of our patients had headaches, history of head trauma, central nervous system infection, or neonatal anoxia. However, one patient (case 3) had a history of febrile seizures and rubella as a child. The age at time of onset of symptoms ranged from 2 to 19 years with a mean age of 7.8 years. Males were more commonly affected than females. All of the lesions were located in the supratentorial cortex with the temporal lobe (80% in our study) most commonly affected. One lesion was located within the occipital lobe.

A previous study reviewed retrospectively the pathologic diagnoses of over 265 patients, all of

whom had surgery for medically refractory partial complex seizures; because of certain common pathologic features (see below), 39 cases were reclassified as DNT (2). While partial complex seizures were present in all patients, other symptoms, including headache and papilledema, and associations with head trauma, neonatal anoxia, and measles encephalitis were noted. The age at time of onset of seizure activity ranged from 1 to 19 years of age, with the mean being 9 years. Males were more commonly affected. The temporal lobe was the most common location (62% of all cases). Other locations included the frontal lobe (31%) and, rarely, the parietal and occipital lobes.

Pathologically, DNTs exhibit a high degree of cellular polymorphism, are occasionally cystic, and show at least one of three characteristics: a specific glioneuronal element, a nodular component, and an association with cortical dysplasia (2). These features help distinguish DNTs from oligoastrocytomas, which superficially resemble DNTs and are not to be confused with embryonal tumors which, by convention, are currently considered to be malignant with immature components (2, 14). However, because of the location of many of these lesions to adjacent critical structure (ie, cranial nerves or vessels), frequently only partial resection is possible. The use of subpial aspiration is often required to preserve adjacent anatomic structures. As a consequence, the possibility arises that all three specific characteristics of DNT may not be present within the surgical specimens obtained. While the etiology is unknown at present, it may originate from the secondary germinal layers, most likely the subpial granular layer (2). They may be related to microcystic cerebellar astrocytomas and, if DNTs do originate from secondary germinal layers (as is postulated), it is reasonable to expect that they may occur in the cerebellum.

Patients with DNTs have an excellent prognosis with surgical resection. In the study of Daumas-Duport (2), no recurrences were identified in 37 patients (two patients died of other causes not related to the tumor) in follow-up ranging from 1 to 18 years (mean, 9 years). Even if only partial resection was obtained, all 39 patients in the previous study experienced at least a significant reduction in seizure frequency and usually were seizure-free. In our study, all six patients had subtotal resections. With follow-up ranging from 6 to 40 months, five patients are seizure-free and one is considerably improved with better control

of his seizures since surgery. None has received chemotherapy or radiation therapy, based on the experience of the previous study.

The CT appearance of DNT, as reported by Daumas-Duport, usually consists of hypodense masses before and following administration of iodinated contrast (2). Calcification, enhancement, and calvarial erosion may be present. Surrounding white matter edema has not been reported. CT scans may in fact be normal in 10% of cases (2). In our study, four patients had CT scans and we had a similar experience with one patient (case 5) having multiple scans (which were not available for review) reported as normal by another institution and three patients having contrast CT scans which showed hypodense nonenhancing masses. With conventional angiography, the majority of DNTs will be undetectable without evidence of neovascularity, mass effect, or hypovascularity, a finding corroborated by our study.

Daumas-Duport reported three cases of DNT imaged with noncontrast MR scans (2). Each lesion had prolonged T1 and T2 relaxation times on MR. While most of our cases demonstrated well-marginated low signal intensity on T1W images, one case demonstrated an isointense mass. All six cases showed lesions with increased signal intensity on T2W images. The characteristics on PDW images were more variable, as one case was hypointense, one case was hypointense with a rim of increased signal intensity, and four cases were hyperintense to gray matter. In general, and in agreement with previously published CT data, the majority of these lesions show minimal or no enhancement with gadolinium-DTPA. Calcification was rare in our study and in the previous study by Daumas-Duport.

While DNT may resemble a benign cyst, the increased signal intensity on proton density images should suggest the presence of a more complicated lesion. The MR appearance of ganglioglioma or low-grade astrocytoma may demonstrate a similar appearance to DNT with a focal mass that is almost always iso- to hypointense on T1W images and hyperintense on T2W images (15, 16). Gangliogliomas may calcify and may enhance on CT. While enhancement with gadolinium-DTPA on MR has not been documented in the literature (16), we have seen a case at our institution of an enhancing ganglioglioma. Calvarial erosion has not been observed with gangliogliomas. In contrast to the experience with DNT, gangliogliomas and low-grade astrocyto-

mas may recur and this possibility may necessitate the use of chemotherapy or even radiation therapy (17). Of note, prior to the report of DNT by Daumas-Duport, our neuropathologists would have classified the tumors herein described as low-grade astrocytomas or gangliogliomas. This underscores the importance of including DNT in the radiologic differential diagnosis of a well-circumscribed, low intensity (T1W) cortical mass found in the patient with the proper clinical history.

In summary, DNT is a pathologically benign neoplasm which is usually supratentorial in location, or usually in the temporal lobe. While the MR appearance may mimic that of ganglioglioma or low-grade astrocytoma, the presence of a focal cortical lesion with hypointensity on T1W images and hyperintensity on T2W images in patients who have a history of partial seizures (usually complex) with onset prior to young adulthood should suggest the diagnosis of DNT. It may superficially resemble a benign cyst or have increased signal intensity on PDW images. Post-contrast enhancement and calcification occur occasionally.

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