Case 186: Dysembrioplastic Neuroepithelial Tumor[1]

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History

A 19-year-old man with refractory epilepsy since age 14 years was referred for magnetic resonance (MR) imaging of the brain. During the past 5 years, he had experienced an average of one seizure per month. Each seizure was characterized by a cephalic aura followed by behavioral arrest, tonic left-sided deviation of the head, and bending of the whole trunk to the left side. Current medications included carbamazepine and clobazam. At electroencephalography, epileptiform discharges from both the right centroparietal region and the midline centroparietal region were recorded. The findings of a neurologic examination were unremarkable.

Imaging Findings

Brain MR imaging (Figs 1–5) revealed a cystlike, wedge-shaped well-marginated intraaxial mass located in the right occipital lobe that measured approximately 3.4 × 2.2 × 3.0 cm (anteroposterior, laterolateral, and craniocaudal diameters, respectively).

The lesion had a cortical base and an apex pointing toward the lateral ventricle. With respect to the normal white matter, its signal was homogeneously hyperintense on T2-weighted images and hypointense on T1-weighted images. Some delicate septalike structures were visible within the lesion. Despite its size, neither mass effect nor surrounding parenchymal edema was present. On T2 FLAIR images, the lesion was contoured by a thin markedly hyperintense ring. Susceptibility-weighted images did not depict any hypointense signal in the lesion, which indicated the absence of calcium or blood degradation products. Very high values of ADC were measured inside the mass. Mostly visible in the sagittal sequences was the scalloping of the overlying inner cortex of the parietal bone. No contrast enhancement was noted.

Discussion

The imaging findings are strongly suggestive of dysembryoplastic neuroepithelial tumor (DNET), an entity often discovered in the work-up of focal epileptic seizures in a young patient. The differential diagnosis includes other brain tumors, such as ganglioglioma, angiocentric glioma, low-grade astrocytoma, and pleomorphic xanthoastrocytoma (PXA) (Fig 6).

A diagnosis of ganglioglioma is less likely than a diagnosis of DNET because the most common appearance of gangliogliomas is that of a cyst with a strongly enhancing mural nodule; moreover, gangliogliomas frequently demonstrate some calcification in the lesion, which was absent in this patient (1).

Angiocentric glioma is a recently described pathologic entity (2) that was initially included in the World Health Organization (WHO) classification in 2007. This tumor is radiologically similar to DNET but has some pathognomonic features, including a hyperintense appearance at T1-weighted imaging and a stalk-like extension to the closest section of the ventricular system, both of which were absent in this patient (3).

Part one of this case appeared 4 months previously and may contain larger images.

Published online
10.1148/radiol.12100118 **Content code:** NR

Radiology 2012; 265:317–320

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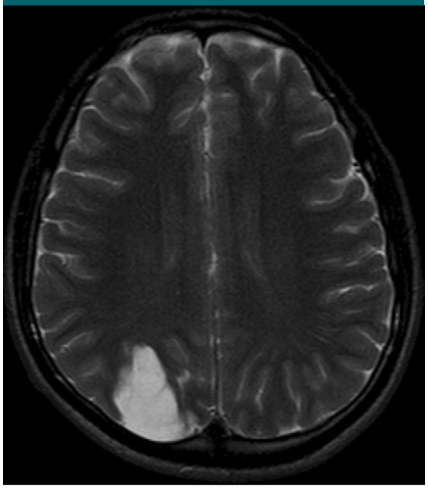
Authors stated no financial relationship to disclose.

Figure 1



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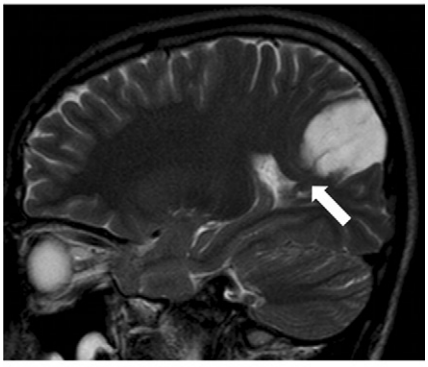


b.

Figure 1: **(a)** Axial and **(b)** sagittal spin-echo T2-weighted MR images (repetition time msec/echo time msec, 5580/110) obtained through the centrum semiovale (**a**) and in the right hemisphere (**b**). There is a wedge-shaped cortical-based lesion in the right occipital lobe, which appears hyperintense with some thin internal septations. The apex of the lesion points toward the trigone of the lateral ventricle, as better seen in the sagittal sequence (arrow). The lesion causes remodeling of the overlying inner cortical parietal bone.

Figure 2

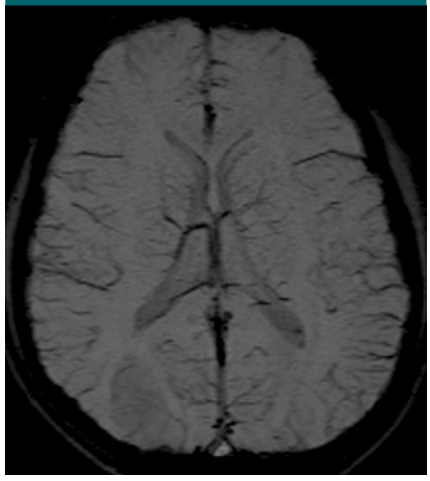
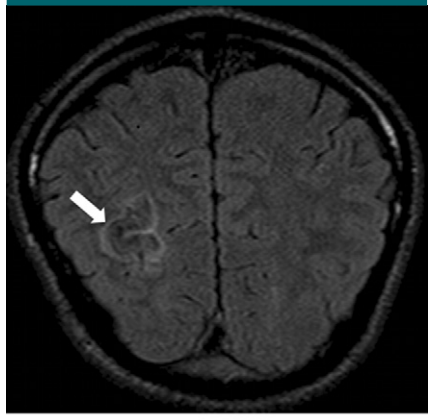


Figure 2: Axial susceptibility-weighted MR image (49/40) obtained through the centrum semiovale. This is a minimum intensity projection image. No abnormal vessels or abnormal signal intensity are seen, suggesting the absence of calcium or blood degradation products.

Figure 3



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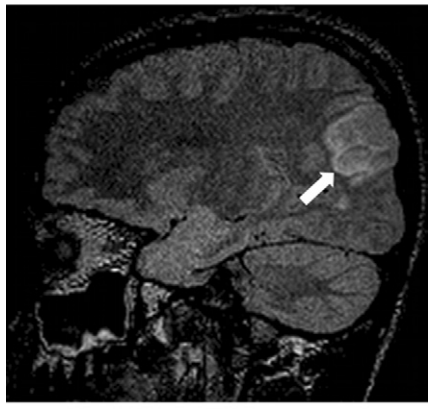

b.

Figure 3: **(a)** Coronal and **(b)** sagittal fluid-attenuated inversion recovery (FLAIR) MR images (repetition time msec/echo time msec/inversion time msec, 8010/109/2500) obtained through the occipital lobes (**a**) and in the right hemisphere (**b**). With respect to normal white matter, the lesion appears isointense with some hyperintense septa inside it; at the periphery, a well-defined hyperintense rim separating the tumor from the surrounding normal brain is visible (arrow). This is referred to as the FLAIR hyperintense ring sign.

Low-grade astrocytoma always should be added in the differential diagnosis of T2 hyperintense intraaxial masses that do not enhance; nevertheless, cortical involvement, a FLAIR hyperintense ring around the lesion, and scalloping of the overlying bone are not features of this entity.

PXA is a WHO grade II astrocytoma variant (4). Similar to DNETs, PXAs usually are superficial tumors. Unlike DNETs, however, PXAs typically manifest as a cystic tumor with a strongly enhancing mural nodule, often with an adjacent dural tail of enhancement (5).

In this patient, the lesion manifested with all of the typical MR characteristics of DNET. The diagnosis was confirmed histologically in the surgical specimen.

DNET is a primary brain neoplasm first described as a distinct entity in 1988 by Daumas-Duport et al (6). In 1993, it was included in the WHO classification in the neuronal and mixed neuronal-glial tumor category, along with ganglioglioma, gangliocytoma, desmoplastic infantile ganglioglioma, central neurocytoma, and ganglioneuroma (7).

DNET is a benign, supratentorial, and predominantly cortical intraaxial lesion, characterized by a multinodular architecture (8). Although a DNET usually is located in the temporal lobe, any lobe within the brain lobes may be involved (9). Histologically, it is composed of heterogeneous cellular components with mature ganglion cells and astrocytes immersed in a myxoid matrix (10).

The differential diagnosis with other brain tumors is important because patients with DNET benefit from complete resection, as the presence of residual tumor is a risk factor for relapse of seizures.

Clinicoradiologic criteria for the diagnosis of DNET are as follows (6,11,12): *(a)* history of partial seizures, with or without generalization, beginning before 20 years of age; *(b)* absence of neurologic deficit; *(c)* cortical location of the lesion and *(d)* absence of mass effect and peritumoral edema.

All these characteristics were pre-

Figure 4

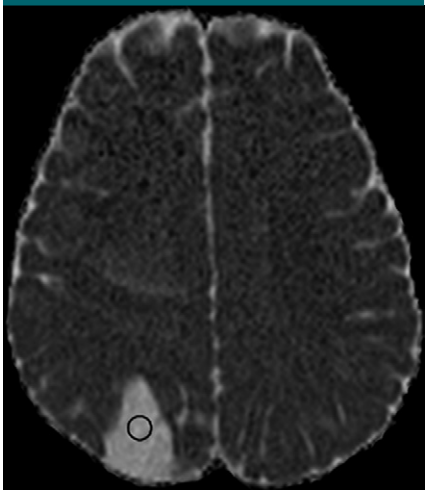

Figure 4: Apparent diffusion coefficient (ADC) map (3500/105) obtained through the centrum semiovale shows homogeneous and abnormally high ADC values inside the lesion, measured by selecting a region of interest (mean ADC, 2.51 × 10^{-3} mm^2/sec ± 0.32).

Figure 5

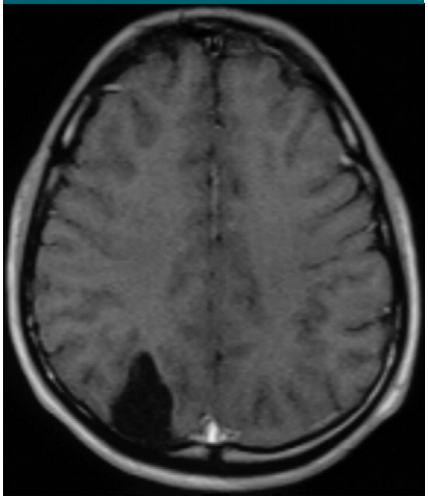

Figure 5: Axial spin-echo T1-weighted MR image (468/11) obtained through the centrum semiovale after intravenous injection of 14 mL of gadodiamide (Omniscan; GE Healthcare, Waukesha, Wis). The occipital lesion appears hypointense at T1-weighted imaging and has some thin septations inside it. No enhancement is noted.

sent in this patient, along with additional typical radiologic features of DNET, namely the wedgelike shape (Fig 1), presence of internal septations (Figs 1, 3, 5), absence of calcification (Fig 2), absence of contrast enhancement (Fig 5), and scalloping of the overlying cortical bone (Fig 1b) (8,9,13).

Two further features of DNET recently have been deemed useful for distinguishing DNET from low-grade glioma and ganglioglioma, namely the FLAIR hyperintense ring sign and the high ADC values within the tumor.

The FLAIR hyperintense ring sign was described by Parmar et al (14) as a thin rim of well-defined hyperintensity at the borders of the DNET, separating it from the surrounding normal brain. This feature is apparent in this patient (Fig 3). The pathologic correlate of this imaging sign was loosely packed glioneuronal elements at the margin of the lesion (14); at follow-up MR imaging, three of 10 patients had a partial residual ring, and two patients had continued symptoms of seizures (14), leading to speculation that removal of the hyperintense ring around the lesion can be relevant to the avoidance of recurrence.

Figure 6

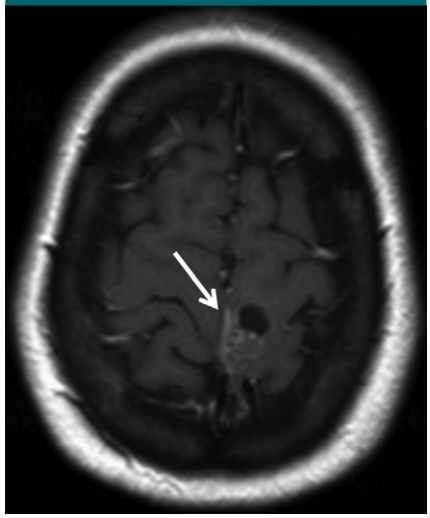

Figure 6: Axial spin-echo T1-weighted magnetic resonance (MR) image (repetition time msec/echo time msec, 11/5) obtained through the upper frontal lobes after intravenous injection of 12 mL of gadodiamide shows PXA. There is a left-sided cortical-based lesion in the frontal lobe, localized in the medial part of the precentral gyrus. The lesion appears as a cyst with an enhancing mural nodule and is accompanied by an adjacent dural tail (arrow). The lesion was removed surgically, and pathologic analysis indicated it was a PXA.

The ADC values of DNET have previously been detailed by Yamasaki et al (15). These authors observed that, among patients with WHO grade 1 or 2 tumors, those with a DNET had the highest ADC value (mean, 2.54 ×10^{-3} mm^2/sec ± 0.13 [standard deviation]; mean ADC of the region of interest in Figure 4, 2.51 ×10^{-3} mm^2/sec ± 0.32), possibly due to both a larger extracellular space and a lower cellularity compared with that of other brain tumors (Fig 4).

In summary, we made the diagnosis of DNET in this patient on the basis of a combination of findings: A cortical-based mass was seen in the brain on MR images in a young patient with refractory epilepsy. It had a cystlike appearance, without perilesional edema or contrast enhancement. Furthermore, the FLAIR sequence revealed a hyperintense ring sign, and the ADC value inside it was remarkably high. MR features of DNET can be highly specific, as in this patient, and use of MR imaging is important for preoperative planning and to determine the extent of resection.

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Congratulations to the 111 individuals and four resident groups that submitted the most likely diagnosis (dysembrioplastic neuroepithelial tumor) for Diagnosis Please, Case 186. The names and locations of the individuals and resident groups, as submitted, are as follows:

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