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Radiology

Dysembryoplastic Neuroepithelial Tumor Revealed by Drug-Resistant Focal Epilepsy in an 11-Year-Old Child: A Case Report and Literature Review

Dr M. Mahmoud[1]*, M. Berrada[1], Y. Bouktibe[1], A. Elhajjami[1], B. Boutakioute[1], N. Cherif Idrissi El Ganouni[1]

[1]Département de Radiologie, Hôpital Arrazi , Centre Hospitalier Universitaire Mohammed VI, Marrakech, Maroc

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***Corresponding author:** Dr M. Mahmoud
Département de Radiologie, Hôpital Arrazi , Centre Hospitalier Universitaire Mohammed VI, Marrakech, Maroc

Abstract **Case Report**

Introduction: Dysembryoplastic neuroepithelial tumor (DNET) is a benign glioneuronal tumor of the central nervous system, classified as WHO grade 1. It predominantly occurs in children and young adults and represents a classic cause of chronic drug-resistant focal epilepsy. ***Case Report:*** We report the case of an 11-year-old child with no significant medical history, presenting with focal seizures refractory to medical treatment over the past 2 years. Brain imaging revealed a well-demarcated, lobulated cortico-subcortical lesion of the right posterior temporal lobe, hypointense on T1, markedly hyperintense on T2 with a characteristic microcystic "soap-bubble" appearance, showing heterogeneous FLAIR hyperintensity, central calcifications, and punctate contrast enhancement, without perilesional edema or mass effect. These findings were consistent with a DNET. ***Conclusion:*** MRI is the reference examination for the diagnosis of DNET. Radiological recognition of this benign entity is critical, as it guides surgical resection the reference treatment associated with excellent tumoral and epileptological outcomes, while preventing unjustified adjuvant therapies.

Keywords: DNET; glioneuronal tumor; drug-resistant epilepsy; MRI; child; FGFR1.

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INTRODUCTION

Dysembryoplastic neuroepithelial tumor (DNET), individualized in 1988 by Daumas-Duport *et al.,* from a series of 39 cases, is a benign glioneuronal tumor of the central nervous system. It is classified as grade 1 in the fifth edition of the World Health Organization (WHO) classification of tumors of the central nervous system (WHO CNS5, 2021), within the group of glioneuronal and neuronal tumors.

It is a rare lesion: it accounts for approximately 1% of neuroepithelial tumors in children, but its proportion rises notably up to 5% to 20% depending on the series among tumors resected within the context of epilepsy surgery in young patients. Its clinical presentation is remarkably consistent: focal seizures beginning before the age of 20 in the vast majority of cases, frequently resistant to medical treatments.

The challenge of radiological diagnosis is twofold. On the one hand, DNET requires a specific management strategy: surgical resection, generally curative, without adjuvant treatment. On the other hand, failure to recognize it exposes the patient to an overdiagnosis of higher-grade glial tumors, carrying the risk of aggressive, unjustified therapies. Brain MRI thus plays a central role, with certain signs being highly suggestive.

We report an illustrated case of a temporal DNET diagnosed on imaging in a child with drug-resistant epilepsy, followed by a discussion of current literature data concerning epidemiology, pathophysiology, radiological semiology, differential diagnoses, and management of this entity.

OBSERVATION (CASE PRESENTATION)

The patient is an 11-year-old boy with no notable personal or familial medical history and normal psychomotor development. For approximately 2 years, he has experienced focal seizures with a sensory onset, characterized by an ascending epigastric sensation followed by a 1-to-2-minute loss of contact and brief post-ictal confusion. They occur 2 to 3 times per week, without bilateralization. Neurological examination is normal.

The interictal EEG shows right temporal slow spikes. Despite two successive anti-seizure medications (carbamazepine then valproate) administered at effective doses for more than 6 months each, the seizures persist, thereby defining drug-resistant epilepsy.

A first-line brain computed tomography (CT) scan revealed a well-demarcated, hypodense cortico-subcortical lesion in the right posterior temporal lobe, housing central calcifications, without perilesional edema (Figure 1).

A brain MRI was subsequently performed according to the following protocol: sagittal T1 sequence; axial T2, FLAIR, and T2* sequences; diffusion-weighted sequence; anatomical coronal sequences; and axial and coronal T1 sequences following gadolinium injection.

In the supratentorial compartment, the examination demonstrated a predominantly cortical signal anomaly in the right posterior temporal lobe, extending in places to the subcortical white matter and adjacent periventricular white matter. The lesion, featuring lobulated and well-demarcated contours, appeared hypointense on T1 and markedly hyperintense on T2, with a characteristic microcystic appearance described as "soap-bubble" (Figure 2), alongside a heterogeneous, faint FLAIR hyperintensity. It showed no restricted diffusion (Figure 3). Central signal voids on T2*, corresponding to calcifications, as well as punctate contrast enhancement following contrast administration, were noted. There was neither perilesional edema nor mass effect on adjacent structures; the ventricular system was of normal morphology, and midline structures were in place.

In the infratentorial compartment, no morphological or signal abnormalities of the brainstem, cerebellum, or cerebellopontine angles were identified; the fourth ventricle was of normal morphology.

As a radiological conclusion, the appearance was that of a predominantly cortical cortico-subcortical lesion exhibiting punctate contrast enhancement, primarily suggesting a dysembryoplastic neuroepithelial tumor (DNET).

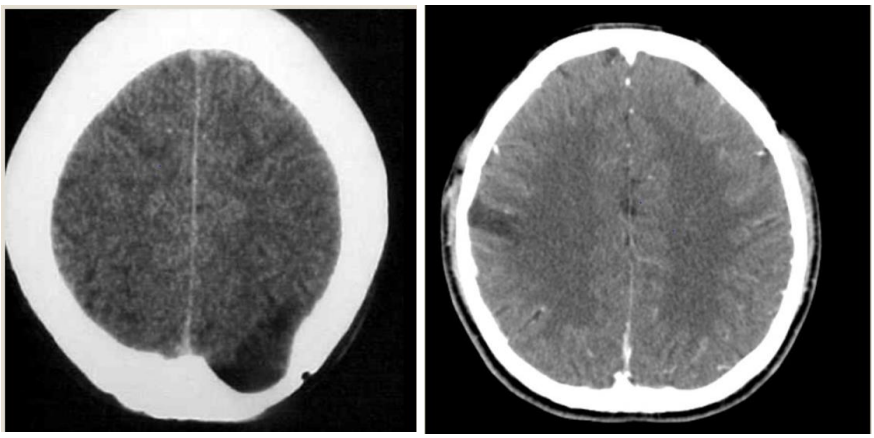


Figure 1: Unenhanced brain CT scan, axial slices: well-demarcated hypodense cortico-subcortical lesion in the right posterior temporal lobe, harboring central calcifications, without perilesional edema.

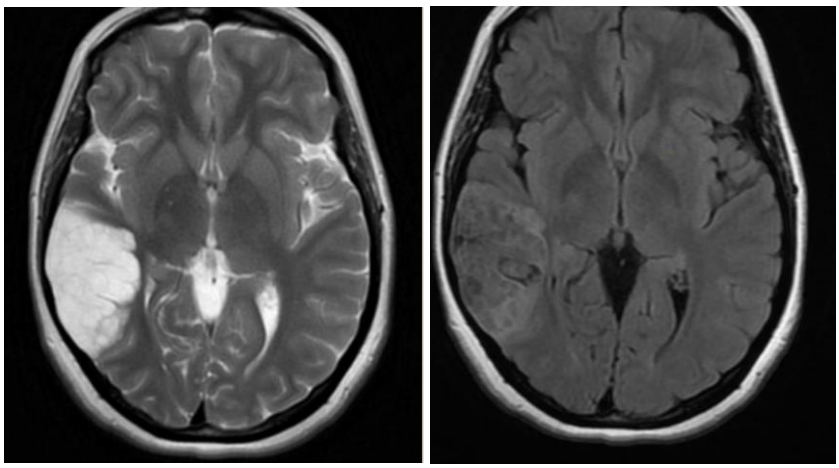


Figure 2: Brain MRI, axial T2 sequences: right posterior temporal lesion exhibiting marked T2 hyperintensity with lobulated contours and a characteristic microcystic "soap-bubble" appearance

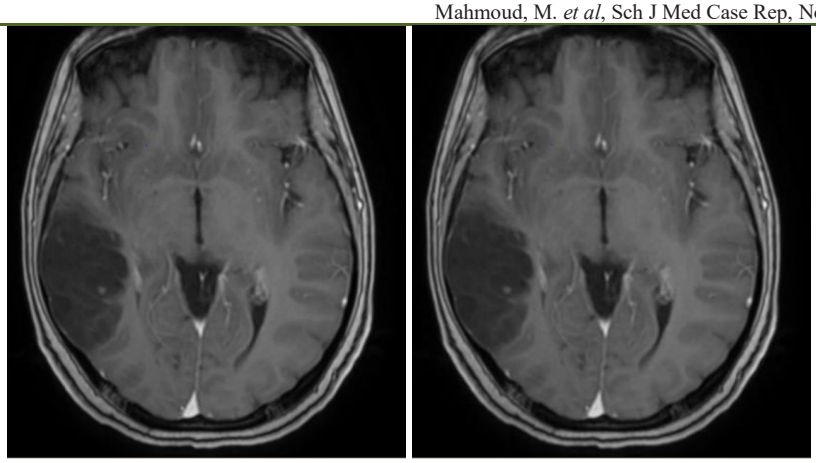


Figure 3: Brain MRI: (a) axial T1 sequence showing the hypointensity of the lesion; (b) diffusion-weighted sequence demonstrating the absence of restricted diffusion

DISCUSSION

Epidemiology

DNET is a rare tumor, accounting for approximately 1% of neuroepithelial tumors in individuals under 20 years of age. However, its relative frequency is significantly higher in selected populations: it ranks among the top three tumor etiologies in operated chronic epilepsy series, alongside gangliogliomas and low-grade astrocytomas. A slight male predominance is reported. The age of seizure onset occurs before 20 years in the vast majority of cases, often in childhood, as observed in our patient.

Definition, Histopathology, and Molecular Data

DNET reflects a developmental cortical anomaly of dysembryoplastic nature, linked to a disruption of neuronal migration and maturation during embryogenesis. Its growth is slow and non-infiltrative; malignant transformation is exceptional and reported only anecdotally.

Histologically, the characteristic element is the "specific glioneuronal element": columns of oligodendrocyte-like cells oriented perpendicular to the cortical surface, associated with astrocytes and mature "floating" neurons within a mucopolysaccharide-rich matrix. Three histological forms are described—simple, complex, and non-specific—with no demonstrated prognostic difference between them. Associated adjacent focal cortical dysplasia is frequent and likely contributes to epileptogenesis.

Molecularly, *FGFR1* alterations constitute the most characteristic signature of DNETs, found in a substantial proportion of cases: internal tandem duplications of the tyrosine kinase domain and point mutations activating the MAPK/ERK pathway. Unlike gangliogliomas, the *BRAF* V600E mutation is rare. These data, integrated into the WHO CNS5 classification, support the individualization of DNETs among low-grade glioneuronal tumors.

Clinical Presentation

The clinical picture is dominated by chronic focal seizures, refractory to medical treatment in more than half of the cases. Three features are suggestive and all present in our patient: early onset in childhood or adolescence; absence of an established neurological deficit or a subtle post-ictal deficit; and a long, stable clinical course without progressive neurological deterioration—reflecting the tumor's very slow growth.

Imaging

General Data

DNET is a predominantly cortical lesion, with potential subcortical extension. It predominantly localizes to the temporal lobe (50% to 70% of cases)—the location in our patient—followed by the frontal lobe (approximately 30%), and more rarely the parietal or occipital lobes or the brainstem. It presents as a lobulated, well-demarcated lesion without significant edema or mass effect; remodeling of the inner table of the cranial vault may reflect its slow growth.

Computed Tomography (CT)

Classically shows a well-circumscribed cortical hypodensity with a pseudocystic appearance, containing calcifications in 20% to 30% of cases, and typically without contrast enhancement. Our case illustrates this appearance: a well-demarcated hypodensity with central calcifications (Figure 1).

Conventional MRI (Reference Examination)

The typical appearance combines:

- **T1-weighted sequence:** Hypointensity relative to the adjacent parenchyma;
- **T2-weighted sequence:** Marked hyperintensity with a multilocular "soap-bubble" organization, a highly suggestive though non-specific sign present in our case (Figure 2);
- **FLAIR sequence:** Heterogeneous signal with partial suppression of the cystic spaces and, characteristically, a hyperintense peripheral rim (*bright rim sign*);

- **T2/GRE/SWI sequences:** Relatively frequent calcifications; hemosiderin deposits are rare, and intratumoral hemorrhages are exceptional;
- **Diffusion:** No restriction, reflecting low cellular density (Figure 3);
- **Post-gadolinium:** Enhancement in approximately 20% to 30% of cases, which may be punctate, nodular, or annular—punctate in our observation.

Advanced Imaging

Complementary techniques reinforce diagnostic confidence when the appearance is atypical: on magnetic resonance spectroscopy (MRS), NAA is normal or moderately decreased without significant elevation of choline (low Cho/NAA ratio), unlike high-grade tumors; on perfusion MRI, relative cerebral blood volume (rCBV) is low; on 18F-FDG PET, the lesion is hypometabolic. In the presurgical evaluation of epilepsy, MRI-EEG correlation (or magnetoencephalography) is essential to delineate the epileptogenic zone, which may extend beyond the boundaries of the visible lesion due to associated cortical dysplasia.

Diagnosis	Features in Favor	Main Distinguishing Feature vs. DNET
Ganglioglioma	Temporal lobe location, child/young adult, epilepsy	Cystic component with an enhancing mural nodule, frequent calcifications, *BRAF* V600E mutation
Low-Grade Astrocytoma	T2 hyperintensity	Infiltrative pattern, ill-defined margins, predominantly subcortical location, absence of multilocular organization
Angiocentric Glioma	Cortical location, pediatric epilepsy	Cortical T2/FLAIR hyperintense rim with a stalk-like extension toward the ventricle, absence of calcifications or contrast enhancement
Oligodendroglioma	Cortico-subcortical location, calcifications	Adult patients, frontal lobe preference, more marked infiltration, 1p/19q codeletion
MVNT (Multivacuolated Neuronal Tumor)	Similar "soap-bubble" appearance	Subcortical / juxtacortical white matter location (rather than purely cortical), classic "leave-me-alone lesion"
Focal Cortical Dysplasia (Type II)	Refractory epilepsy	Transmantle sign (funnel-shaped cortical funneling/thickening), absence of multinodular structure or calcifications

Management and Prognosis

Surgical resection is the gold-standard treatment, prompted most frequently by the pharmacoresistant nature of the seizures. Epileptological outcomes are excellent: 70% to 90% of patients achieve Engel class I (freedom from disabling seizures) following resection, with the completeness of the resection being the primary prognostic factor. There is no indication for radiotherapy or chemotherapy in typical forms. Recurrence after complete resection is exceptional, and spontaneous tumor growth is virtually nil, justifying simple clinical and radiological follow-up. Even in cases of partial resection, satisfactory seizure control is frequently achieved.

CONCLUSION

Dysembryoplastic neuroepithelial tumor is a benign glioneuronal tumor of young individuals, revealed almost exclusively by chronic drug-resistant focal epilepsy. In front of this clinical picture, MRI reveals a well-demarcated cortical lesion with a multilocular "soap-bubble" appearance, lacking significant edema or mass effect, with absent or subtle contrast enhancement. Recognizing this entity is crucial: it guides surgical resection, which is generally curative—ensuring an excellent tumor prognosis and seizure control in the vast majority of cases—while sparing the patient from unjustified adjuvant therapies.

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