DNET AS CAUSE OF SYMPTOMATIC EPILEPSY

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ABSTRACT

Dysembrioplastic Neuroepithelial Tumor (DNET) is a congenital, benign tumor due to a development abnormality of certain embryonic cells of the brain, frequently associated with epilepsy. These lesions typically cause refractory complex partial seizures with onset before age 20 years in neurologically normal individuals who have no evidence of neurocutaneous syndrome. At least 80 % of patients are rendered seizure-free after resection of these lesions. We present the case of a 40 years old woman with complex partial seizures who undervent resective surgery for DNET, and had a good outcome after surgery. We would like to draw attention of the clinicians to this potentially curable type of symptomatic epilepsy.

Key words: Dysembrioplastic Neuroepithelial Tumor (DNET), farmacoresistant epilepsy

INTRODUCTION

Epilepsy is a chronic neurological disorder that affects 0.5 to 1% of the world population. (1)

Although many antiepileptic drugs are available, 30 to 40 % of patients continue to have seizures that are not adequately controlled by pharmacotherapy (2).

Pharmacoresistant epilepsy should be considered if there is persistance of the seizures despite adequate use of antiepileptic drugs, with a minimum of two first-line drugs either as monotherapy or in combinations, as appropiate to the epileptic syndrome.

Surgical treatment for certain types of medically refractory epilepsy is an option of increased interest, especially after the recent advances in neuroimagery. Early surgical intervention, when sucessful, might prevent the disabling psychosocial consequences of uncontrolled seizures, and side effects of the antiepileptic drugs.

Dysembrioplastic Neuroepithelial Tumor (DNET) is a congenital, benign tumor due to a developmental abnormality of certain embryonic cells of the brain, frequently associated with epilepsy.

These lesions typically cause refractory complex partial seizures with onset before 20 years-old age in neurologically normal individuals who have no evidence of neurocutaneous syndrome.

Their intracortical location is best showed by MRI. Histology of the resected cerebral cortex shows the characteristically nodular and intracortical arhitecture of the lesion. The pathology of these tumors consists in glial nodules, foci of cortical dysplasia and a unique glioneural component with a characteristic appearance. Well-differentiated neurons and glial cells are floating in an extracellular matrix. These lesions are biologically indolent and probably hamartomatous. They may resemble both ganglioglioma and true astrocytoma, but distinction from the latter can be made on both clinical and radiological grounds. At least 80 % of patients are rendered seizure-free after resection of these lesions. (3)

CASE PRESENTATION

We present the case of a 40-year-old right handed women admitted to our Department for reccurent

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episodes of "fear and panic attacks". The patient had for 2-3 years ictal episodes started with rising epigastric discomfort, nausea, followed by panic and fear and starring. These had a stereotyped pattern, lasting 2-5 minutes. The frequency of these seizures increased over time to 3-4 attacks per day. There were no tonic-clonic seizures in her history.

There were no febrile seizures in her childhood and no family history of epilepsy.

The physical examinations was unremarkable. The neurological examination was normal.

The electrocardiogram and the biological tests were normal.

The **interictal eclectroencephalogram** (EEG) recorded from scalp electrodes did not show convincing abnormalities.

Cerebral MRI revealed the presence of a left temporal lobe tumor.

Treatment:

As antiepileptic drug we have chosen oxcarbazepine 600 mg/day which decreased seizure frequency.

The patient was reffered to a neurosurgery department and she was sucessfully operated.

The pathological examination of the cerebral tissue disclosed a dysembrioplastic neuroepithelial tumor (DNET). The patient has been seizure free for more than 3 years.

DISCUSSION

Because the attacks were stereotyped we suspected an epileptic disorder, we differentiated from panic attacks and gastrointestinal disorders.

Based on her history a clinical diagnosis of epilepsy with **complex partial seizures** was made. We concluded that it is a **symptomatic temporal lobe epilepsy due to a left temporal lobe tumor.**

Beacause the patient is young, without medical contraindication, with a resectable structural abnormality identified on magnetic resonance imaging she was reffered for resective epilepsy surgery.

We could not perform presurgically long-term video-EEG monitoring, functional MRI or Wadda test to this 40 years old right handed women with a left temporal lobe tumor, by the time this patient was referred to us. However, there are no language or memory difficulties after surgery.

Figure 1 (a-d). *Interictal EEG: University Hospital of Bucharest – Neurology Department – EEG laboratory*

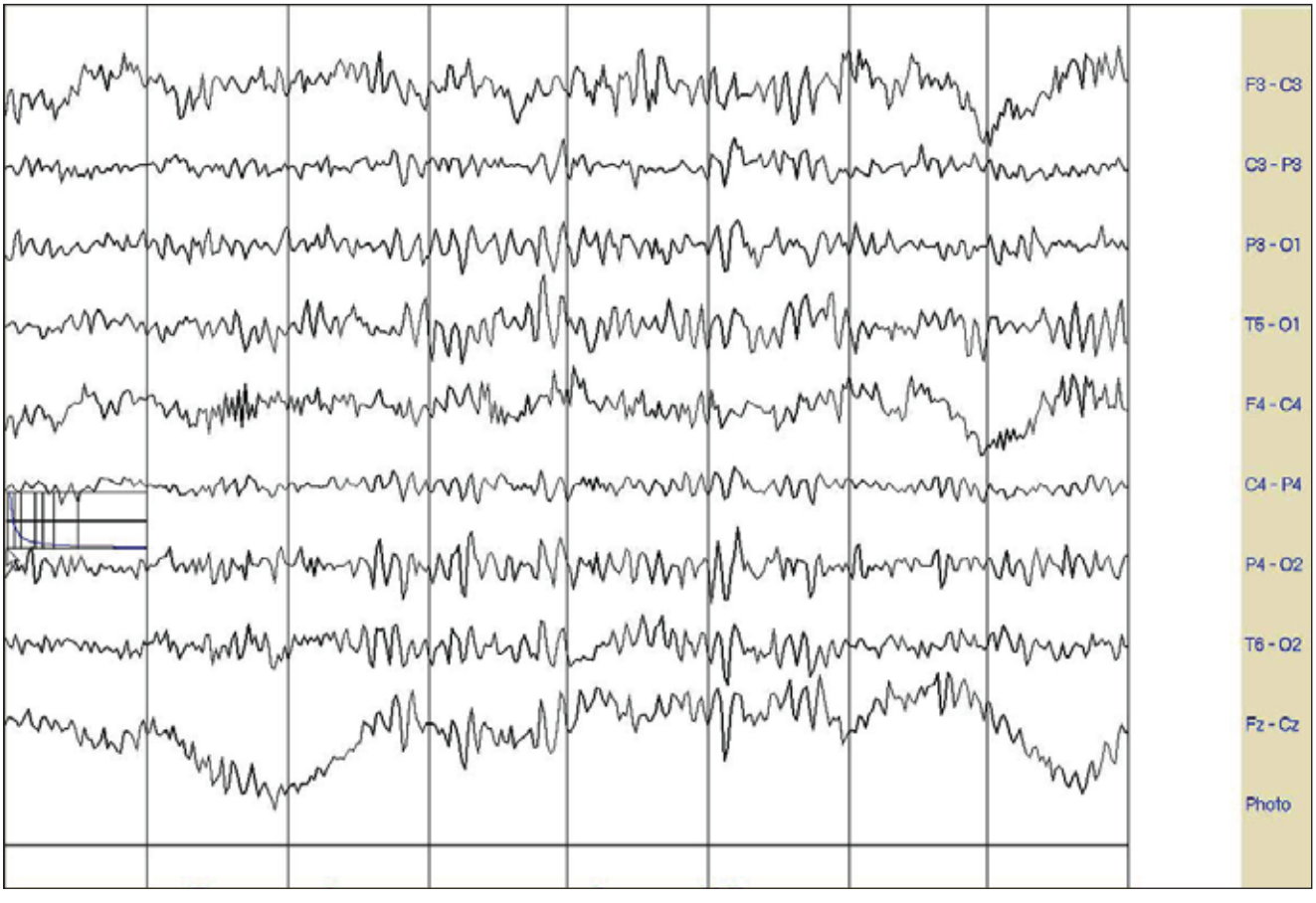
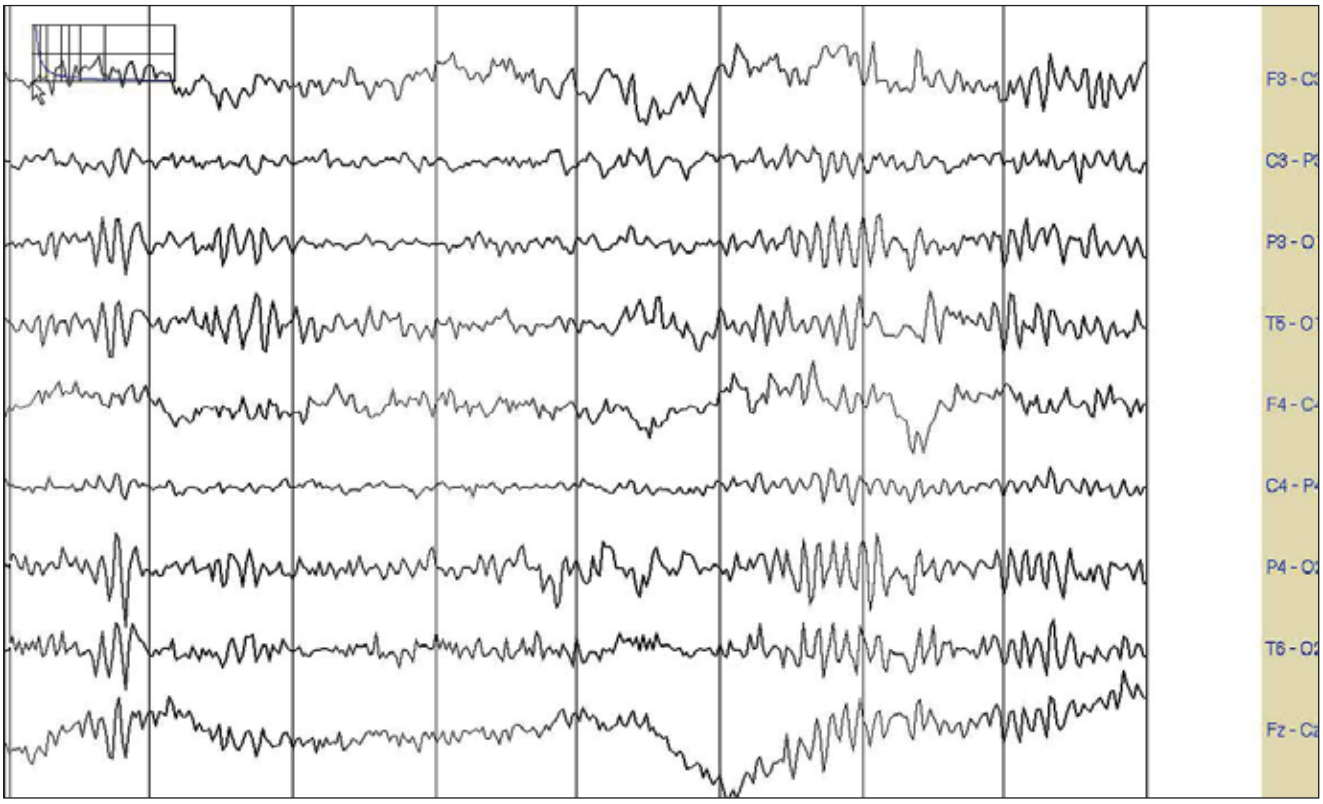


Figure 1 (a-d). *(continuation)*



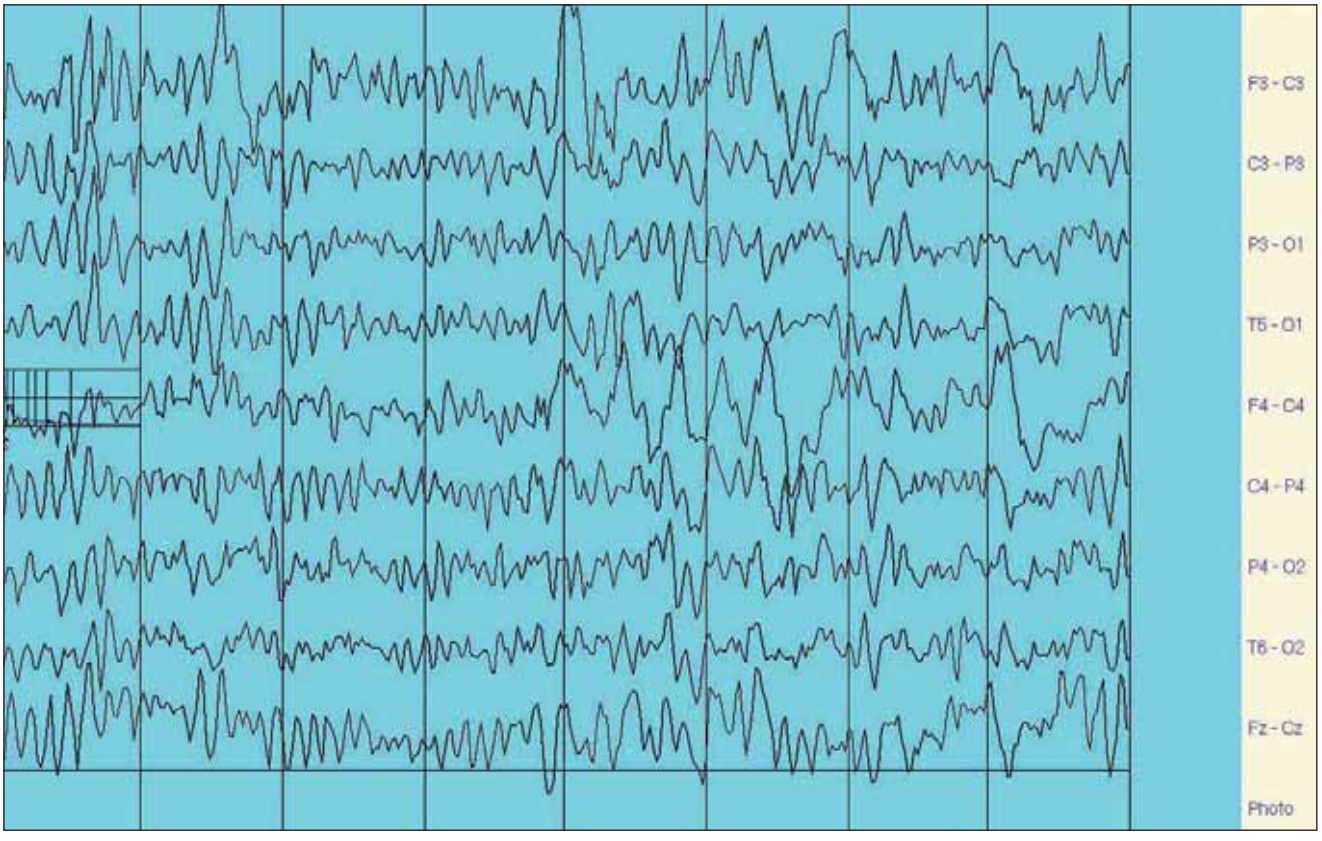
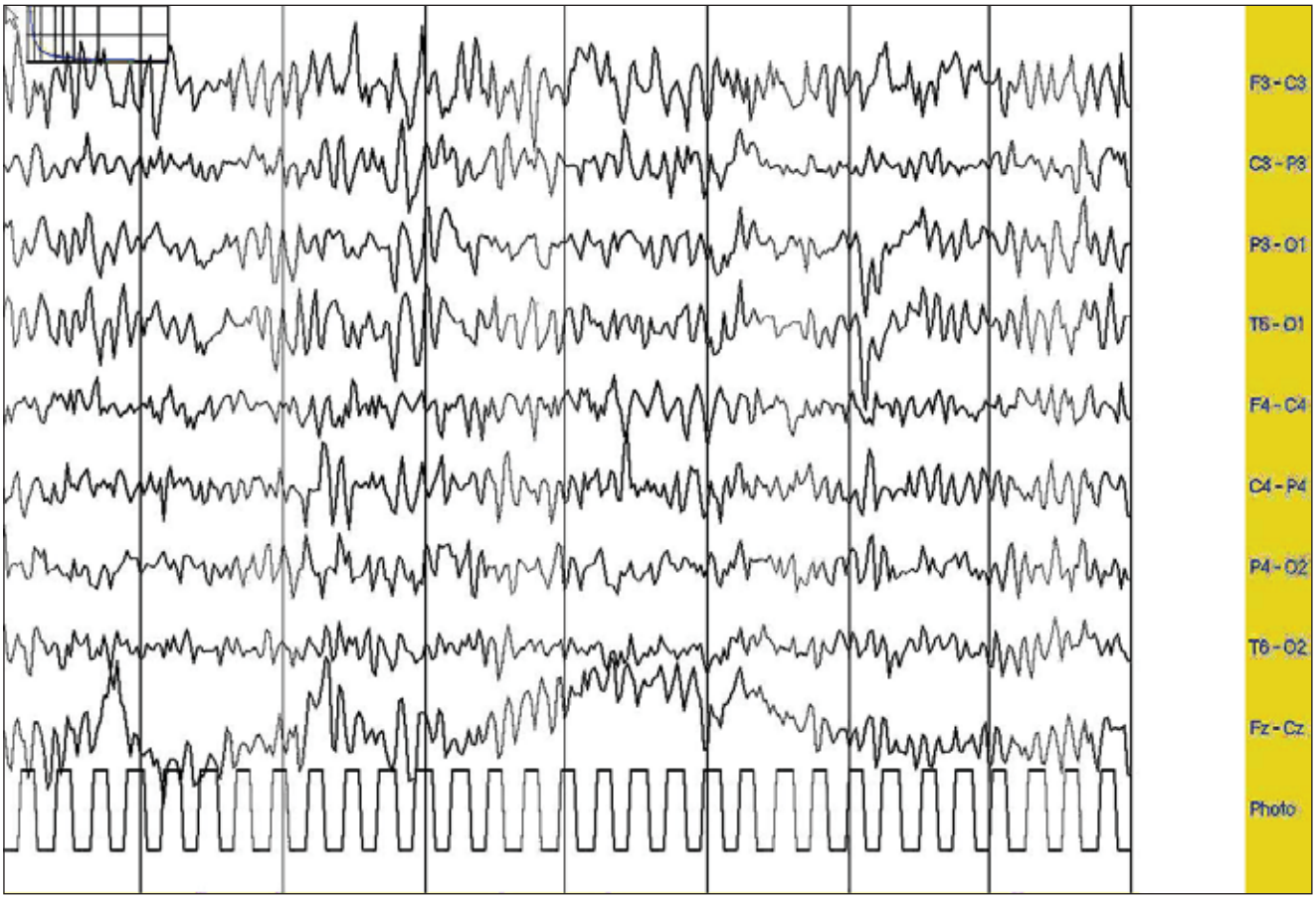


Figure 1 (a-d). *(continuation)*



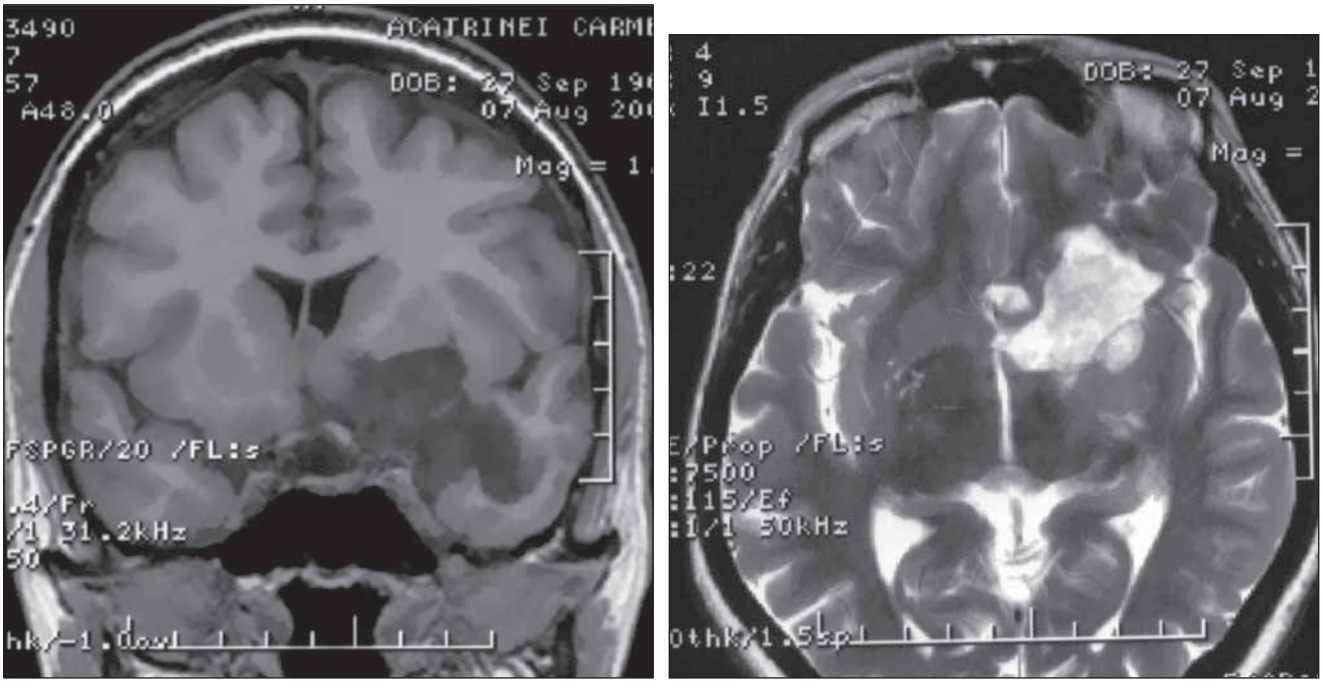


Figure 2. *Brain MRI: University Hospital of Bucharest – Radiology Department: a. T1 – weighted low-intensity b. T2-weighted MRI demonstrates high-intensity signal in the left medial temporal lobe.*

The histological diagnosis of dysembrioplastic neuroepithelial tumor (DNET) was associated with a good outcome: the patient became seizure-free after the surgery.

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