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Case Report Open Access

Dysembryoplastic Neuroepithelial Tumor (DNET) in Parietal Lobe: Is It Different?

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Abstract

Dysembryoplastic neuroepithelial tumor (DNET), a benign, mixed neuroglial tumor that most commonly occupies the temporal lobe of the cortex. The spectrum of clinical presentation varies from simple partial seizure to generalized seizure in some cases. This is a case of a 20-year-old man who presented with a drug resistant epilepsy and was found to have a DNET located in the partial lobe. Therefore, based on radiological finding the diagnosis of DNET was made. Surgical resection of the tumour was done, and the patient recovered uneventfully. The patient was discharged with antiepileptic medications, and showed a complete seizure-free for one year follow up. To the best of our knowledge, this case report brings attention to a rare location of DNET in parietal lobe, also, discusses the clinical presentation, pathological findings, different diagnostic modalities, and surgical approach to such tumour.

Keywords: Dysembryoplastic Neuroepithelial Tumor (DNET); Seizure; Pharmacoresistant epilepsy; Parietal lobe

Introduction

Dysembryoplastic neuroepithelial tumor (DNET) is a benign uncommon mixed glial–neuronal tumour. Initially described by Daumas-Duport et al. in 1988 [1]. Commonly appears in the temporal lobe of the cortex and associated with medically intractable epilepsy [2]. Several diagnostic modalities have been used to diagnose DNET, such as electroencephalogram (EEG), brain Magnetic Resonance Imaging (MRI), brain Computed Tomography (CT) and histopathological findings. The management, of DNET is mainly surgical that reveals good outcomes and low rates of recurrence.

Case Report

A 20-year-old man known to have a drug resistant epilepsy for the last 3 years, which started with dizziness, behavioral arrest and tonic of the upper extremities mainly the right followed generalized tonic-clonic seizure. He was on Levetiracetam (1500 mg) twice daily and Topiramate (100 mg) twice daily, he was previously on Valproate and Carbamazepine. His seizure is intractable not controlled by medications. The seizure frequency is around once per week mostly nocturnal and no proceeded aura. Neurological examination was unremarkable. Visual field examination showed loss of visual field over the nasal and anterior part of the right and it was normal visual field over the left side.

An EEG 24- hour study recording showed two seizure semiology in the form of complex partial seizures and atonic seizure accordingly without proceeded aura. On brain MRI flair axial showed cortically based well-defined left parasagittal frontal lobe lesion with no restricted diffusion or enhancement (Figure 1). A positron emission tomography-Computed Tomography (PET-CT) revealed symmetrical global physiological metabolic activity noted throughout the study. Radiological diagnosis of DNET was made. Extensive evaluations in Epilepsy Monitoring Unit with scalp recording and subdural invasive recording of bilateral strips covering mesial frontal and left parasagittal regions, that showed lesion in the left parietal lobe drug-resistant epilepsy which is corresponding and congruent to brain MRI findings with congruent data that warrants epilepsy surgery intervention.

The tumour was attached to the cortex mesially invaginating the white matter deep to the ventricle. Gross total resection of the tumor along with the thickness of the cortex mesially were taken. The tumor was grayish and soft in the center of and light grayish at the peripheral with thickness. There was transient seizure activity during intraoperative stimulation stimulation. The hand area was localized. Cavitron Ultrasonic Surgical Aspirator (CUSA) was performed from the left mesial frontal tumour. It consists of multiple fragments of soft tan tissue measuring in aggregate 2.5 × 2 × 0.3 cm (Figure 2).

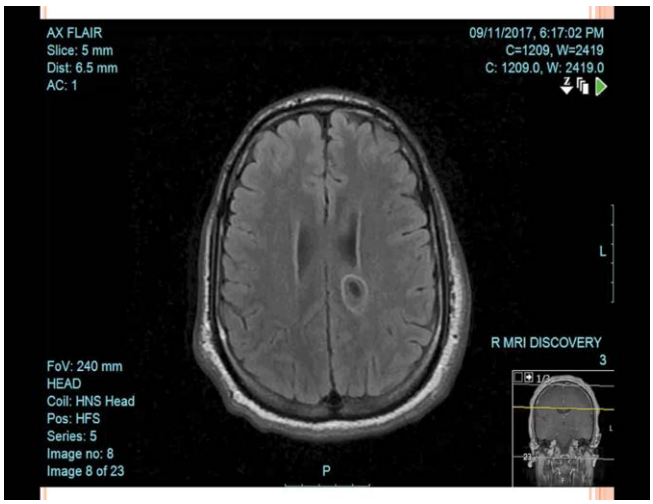

Figure 1: Brain MRI showed left parasagittal isointense lesion surrounded by hyperintense rim with a hypointense center.

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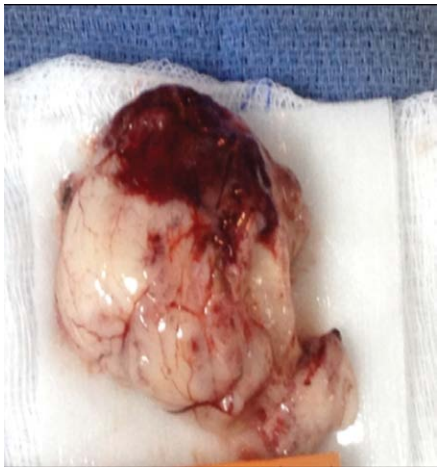

Figure 2: Multiple fragments of soft tan tissue measuring in aggregate 2.5 × 2 × 0.3 cm.

Therefore, diagnosis of dysembryoplastic neuroepithelial tumour (DNET) GRADE I based on World Health Organization (WHO). Post-operative brain Computed Tomography (CT) without contrast revealed no evidence of residual tumor.

Postoperatively, the patient recovered uneventfully. The patient was discharged with antiepileptic medications. No seizures, weakness or tumour recurrence, and no visual field defect confirmed by confrontation test. And the patient has been observed in one-year follow-up.

Discussion

DNET is a benign intracortical supratentorial neoplasm tumor. A classical characteristic of DNET is intracortical location, multinodular architecture, and heterogeneous cellular composition [3]. Based on World Health Organization (WHO) Classification of the Brain Tumors 2007, DNET is classified as WHO grade 1 in the group of neuronal and mixed neuronal–glial tumors [4]. Moreover, temporal

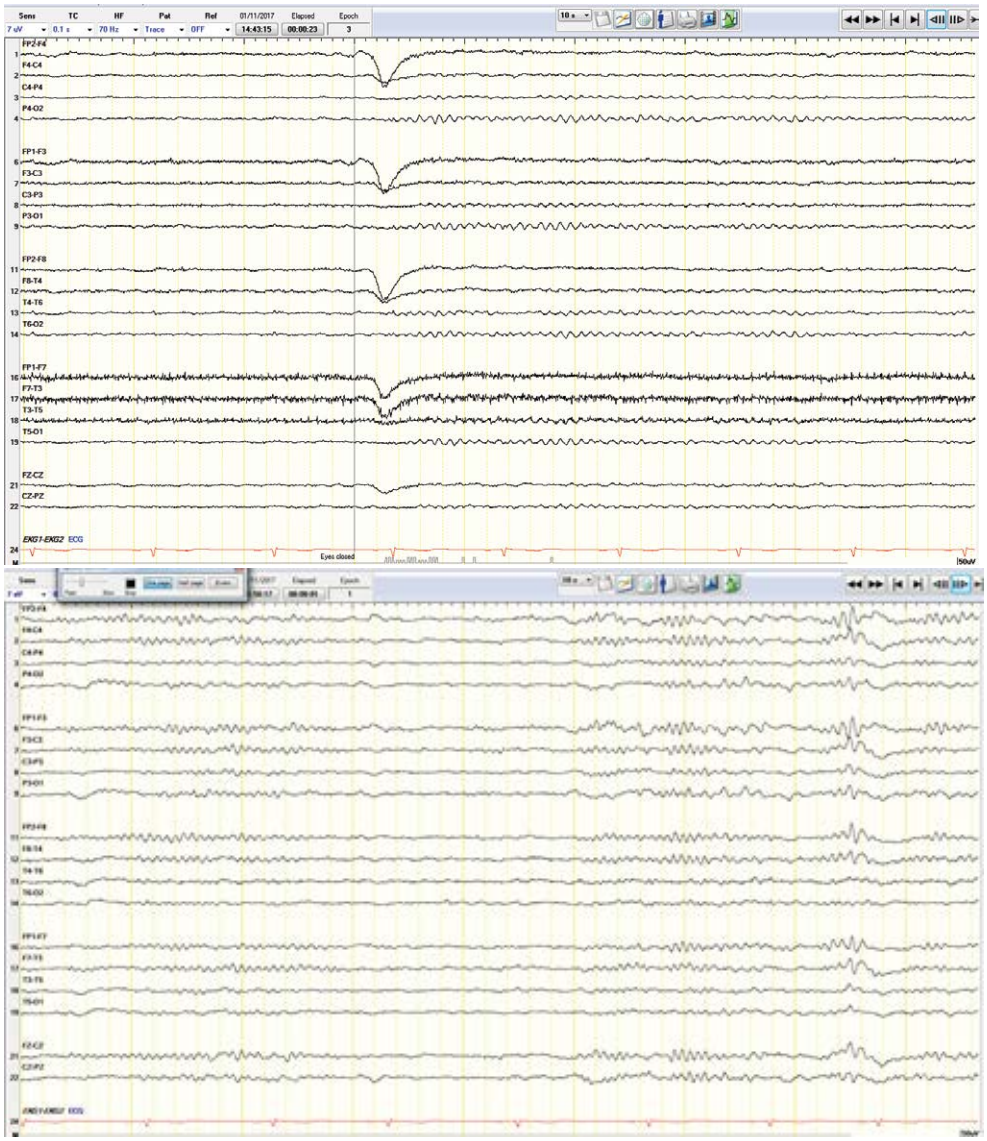

Figure 3: (A and B) No interictal epileptiform discharges were seen during the recording.

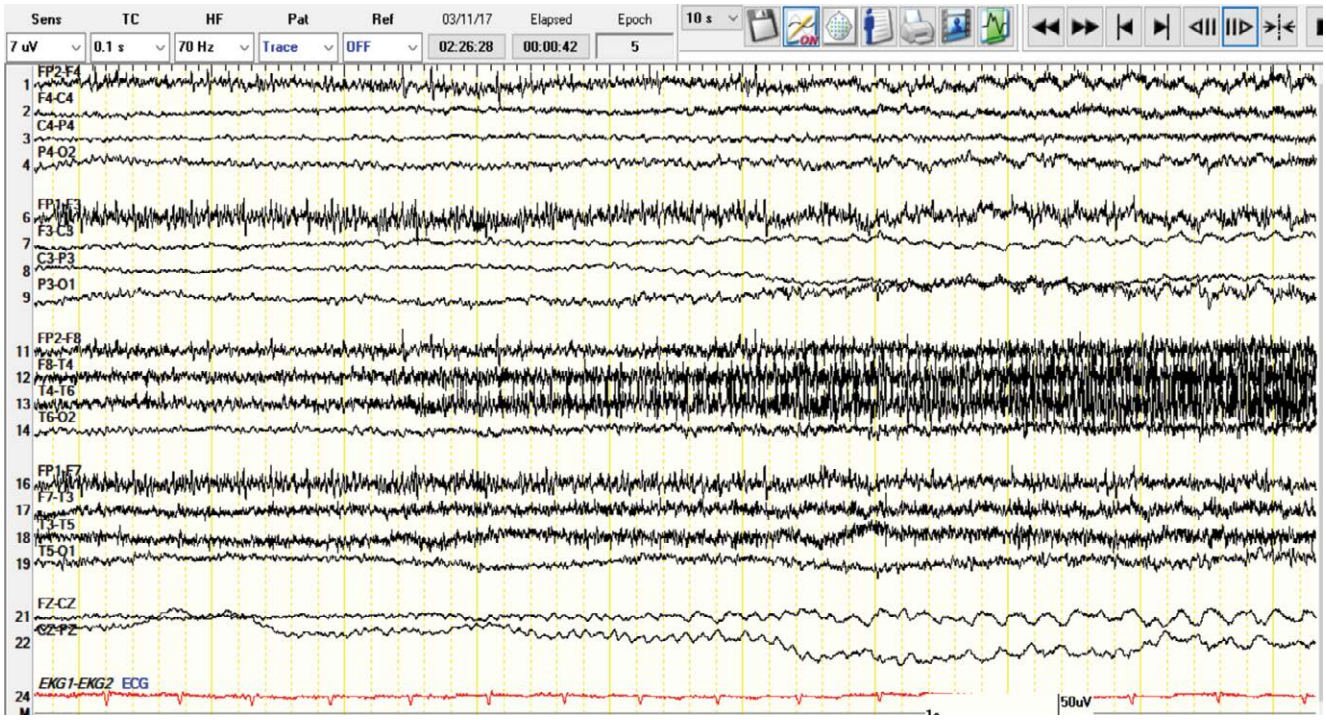

Figure 4: The ictal recording showed a parasagittal ictal pattern more toward the left side (Cz and C3) which evolved later after the onset by around 5-10 seconds.

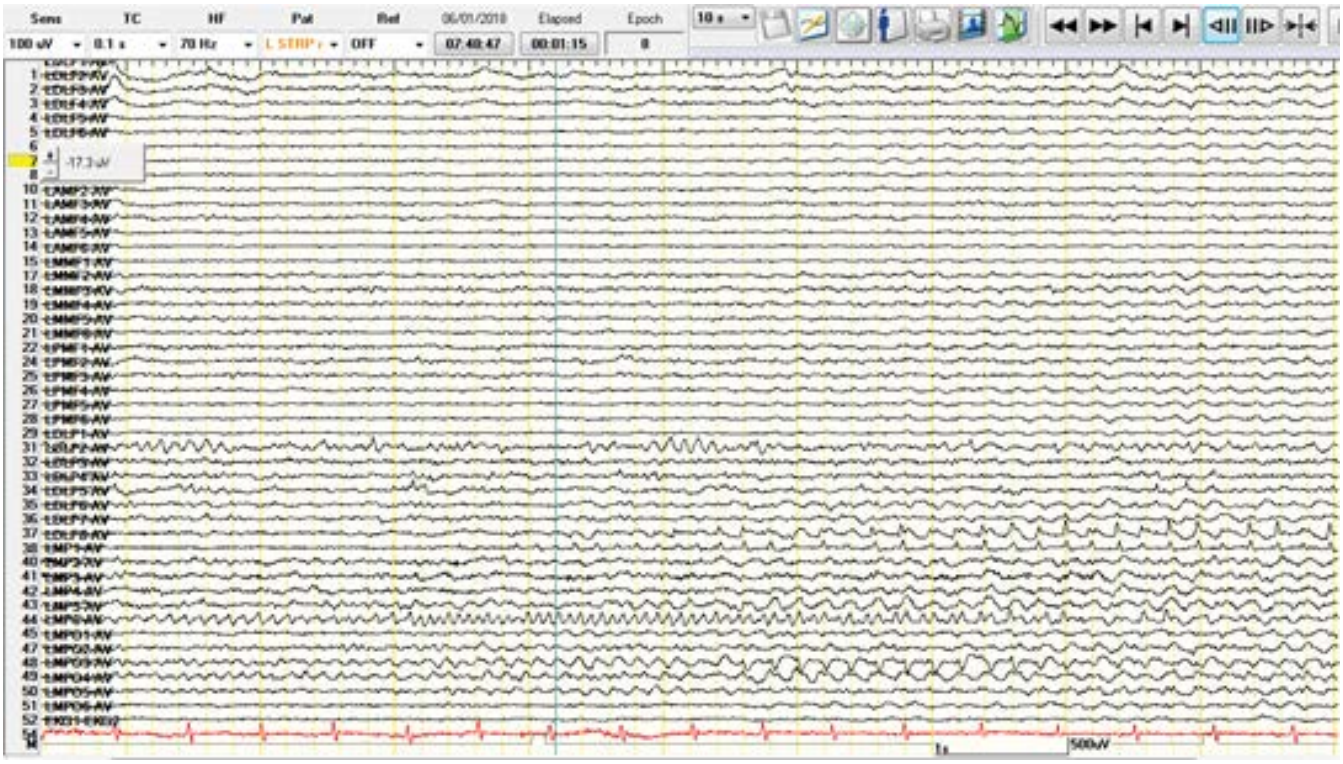

Figure 5: Seizures arising from the left mesial parietal head region. (44 LMP6 AV)

lobe is most commonly invaded by DNET in about 94% of cases, however, it rarely occurred in the parietal lobe with only few cases been previously reported in the literature [5]. Other rarer locations of DNET are Infratentorial such as in the tectum, cerebellum, pons and brainstem have been reported [6]. The clinical features of DNET vary from simple seizure to generalized seizure. Burneo et al., reported a retrospective cohort study of 23 patients seen at two major epilepsy centers which concluded that out of 23 patients, 13 patients (57%) had

simple partial, 21 patients (91%) had complex partial, 16 patients (70%) had secondarily generalized seizures, and 5 patients had only simple partial seizures [7]. Chan et al., reported that all the patients in the study presented with pharmacoresistant seizures as the predominant feature and none had an abnormal neurological examination. Figures 3A and 3B.

DNET is diagnosed mainly by EEG, imaging and pathological findings. EEG features elaborates the understanding of the epileptogenesis associated with DNET tumors, however, specific features of DNET are not well described, for instance, temporal lobe DNET is commonly associated with interictal EEG abnormalities, interestingly, 44% of the patients found to have EEG abnormalities in a location discordant to the tumour [8]. In addition, brain MRI typically demonstrated a cortical mass of hypointensity on T1 and of marked hyperintensity on T2 weighted images with occasional gadolinium enhancement [2]. Correspondingly, brain MRI of the patient showed left parasagittal isointense lesion surrounded by hyperintense rim with a hypointense center. Furthermore, computed tomography (CT) of DNETs characterized by well-demarcated, hypodense cortical lesions [9]. Additionally, histopathological features of DNET are classically described as bundles of axons lined by oligodendroglia-like cells, forming columns in a pale mucoid matrix in which isolated neurons float [6]. Comparatively in this case, histopathological findings showed that the tumor consists of multiple fragments of soft tan tissue with grayish soft center and light grayish at the peripheral with thickness (Figure 4).

The mainstay of treatment is surgical resection of the tumor. In the case, the patient underwent Cavitron Ultrasonic Surgical Aspirator (CUSA) of the tumor located in the left mesial frontal lobe. Favorable seizure outcome immediately following surgical resection in the form of complete recovery to baseline. Complete surgical resection without adjuvant radiotherapy or chemotherapy is the recommended treatment in DNET with specifically pharmacoresistant complex partial seizures for both children and young adults as it results in an excellent control of seizures [10]. Bruneo et al, demonstrated that the surgical treatment yield an excellent reselision of seizure with no significant associations with the age at seizure onset, duration of epilepsy and age at surgery [7] (Figure 5).

Conclusion

In conclusion, DNET is a benign tumor, composed of neuroglial cell, most probably confined to the temporal lobe. It affects children and adults, and it results in seizure varying in severity from simple partial to generalized seizures. Only a few number of patients were found to have partial lobe DNET, which can be demonstrated by the EEG. Treatment is mainly by surgery; however, each case is different and treatment is best individualized depending in the presentation and the patient case.

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