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Case Report

Occipital dysembryoplastic neuroepithelial tumor presenting as adult-onset temporal epilepsy☆

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ABSTRACT

Dysembryoplastic neuroepithelial tumor (DNET) is a benign brain tumor which commonly presents as childhood-onset temporal lobe epilepsy (TLE). We present a case of histologically proven DNET with a clinical presentation and scalp EEG suggestive of adult-onset TLE. MRI showed an occipital lesion. PET showed abnormal metabolism of the occipital lesion and the ipsilateral temporal lobe; raising concern for an abnormal functional network reorganization. Intracranial EEG showed interictal spikes and seizures originating from the occipital lesion with no seizures emanating from the temporal lobe. Occipital DNET due to their chronic nature can reorganize the network and mimic TLE.

1. Introduction

Dysembryoplastic neuroepithelial tumor (DNET) was first described in 1988 as a rare benign tumor which typically presents as childhood-onset, medically drug-resistant complex partial epilepsy [1]. We present a case of adult-onset epilepsy due to an occipital DNET, rare in this age group and location. Because of incongruence in the clinical presentation (temporal lobe semiology) and imaging, we discuss the importance of intracranial EEG in localization and surgical planning for the best possible outcome in extratemporal DNETs.

2. Case

A 55-year-old woman presented with drug-resistant epilepsy since the age of 30. Her most common seizure semiology was characterized by episodes of blinking and staring followed by several minutes of confusion, sometimes evolving to a bilateral convulsion, with variable weekly frequency. She had a past medical history of orthostatic hypotension, premature menopause, and thrombocytopenia. She had tried multiple antiseizure drugs including topiramate, levetiracetam, lamotrigine, zonisamide, lacosamide, and valproic acid and had failed due to persistent seizures or intolerable side effects. Neurological exam was normal. MRI demonstrated an abnormal rounded focus of T2 and FLAIR hyperintensity with minimal enhancement in the right superior parasagittal occipital lobe (Fig. 1). PET imaging demonstrated a concurrent hypermetabolic focus in the occipital lobe as well as a hypometabolic region within the right temporal lobe (Fig. 2). During a 6-day continuous scalp EEG, she had three clinical episodes of blinking and staring followed by several minutes of confusion, during which EEG showed bitemporal slowing (maximal over the right hemisphere) and no clear ictal or interictal epileptiform discharges. Based on the discordant clinical, neurophysiological and imaging data, implantation with intracranial depth electrodes for stereoelectroencephalography (sEEG) was performed, with seven 12-contact depth electrodes "caging" the occipital lesion to best define the boundaries of the seizure-onset zone, and one 12-contact depth electrode per hippocampus/mesial temporal region to assess the extent of the hypothesized epileptic network. sEEG demonstrated innumerable subclinical seizures originating from electrodes within the occipital lesion, and immediately inferior medial, superior medial and inferior lateral to it (Fig. 3). None of the seizures originated from or spread to the

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hippocampal electrodes. Mapping was done to delineate areas of eloquent cortex. She underwent a right occipital craniotomy with resection of the occipital lesion guided by intraoperative electrocorticography. Gross pathology showed a firm, rubbery lesion surrounded by grossly normal brain. Histology showed proliferation of oligodendrocyte-like cells (OLCs) with scattered neurons without dysplastic features. No specific glioneuronal element, adjacent cortical dysplasia or dystrophic calcifications were noted (Fig. 4). The immuno-morphologic features and molecular information was consistent with nonspecific form of DNET (WHO grade I). One year after the resection, the patient remained seizure free on zonisamide and lacosamide with her only deficit being a partial left homonymous hemianopia.

3. Results

3.1. Imaging

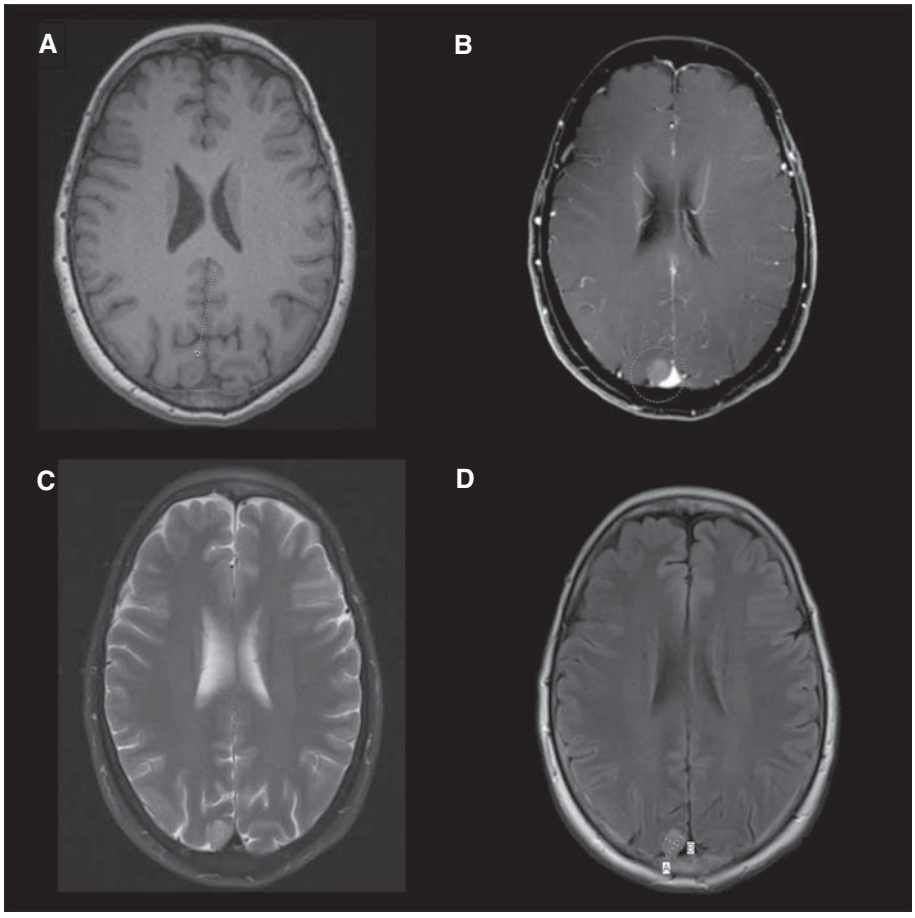

Fig. 1. MRI T1 axial view (A) showing rounded hypointense lesion. MRI T1-post (B) showing lesion with minimal abnormal homogenous enhancement. MRI T2 axial view (C) and FLAIR axial view (D) showing rounded hyperintense signal abnormality in the parasagittal right occipital lobe measuring 1.3 cm × 1.0 cm × 1.3 cm.

3.2. sEEG

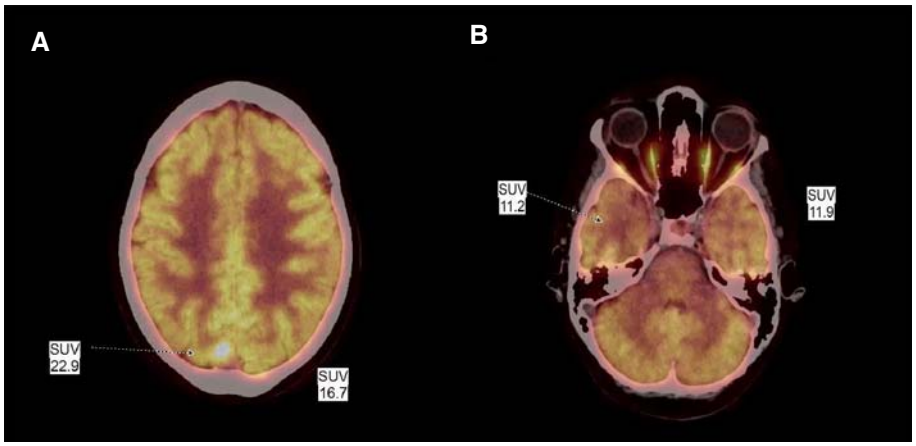

Fig. 2. (A) Axial PET scan showing focal increased activity in the right parasagittal occipital lobe with maximum standardized uptake values (SUV) of 22.9 in the occipital lesion, compared to the contralateral side with SUV of 16.7. (B) Axial PET scan of bilateral temporal lobes showing slightly hypo-metabolic region in the right temporal lobe medially with SUV of 11.2, compared for the left side with SUV of 11.9.

3.3. Histopathology

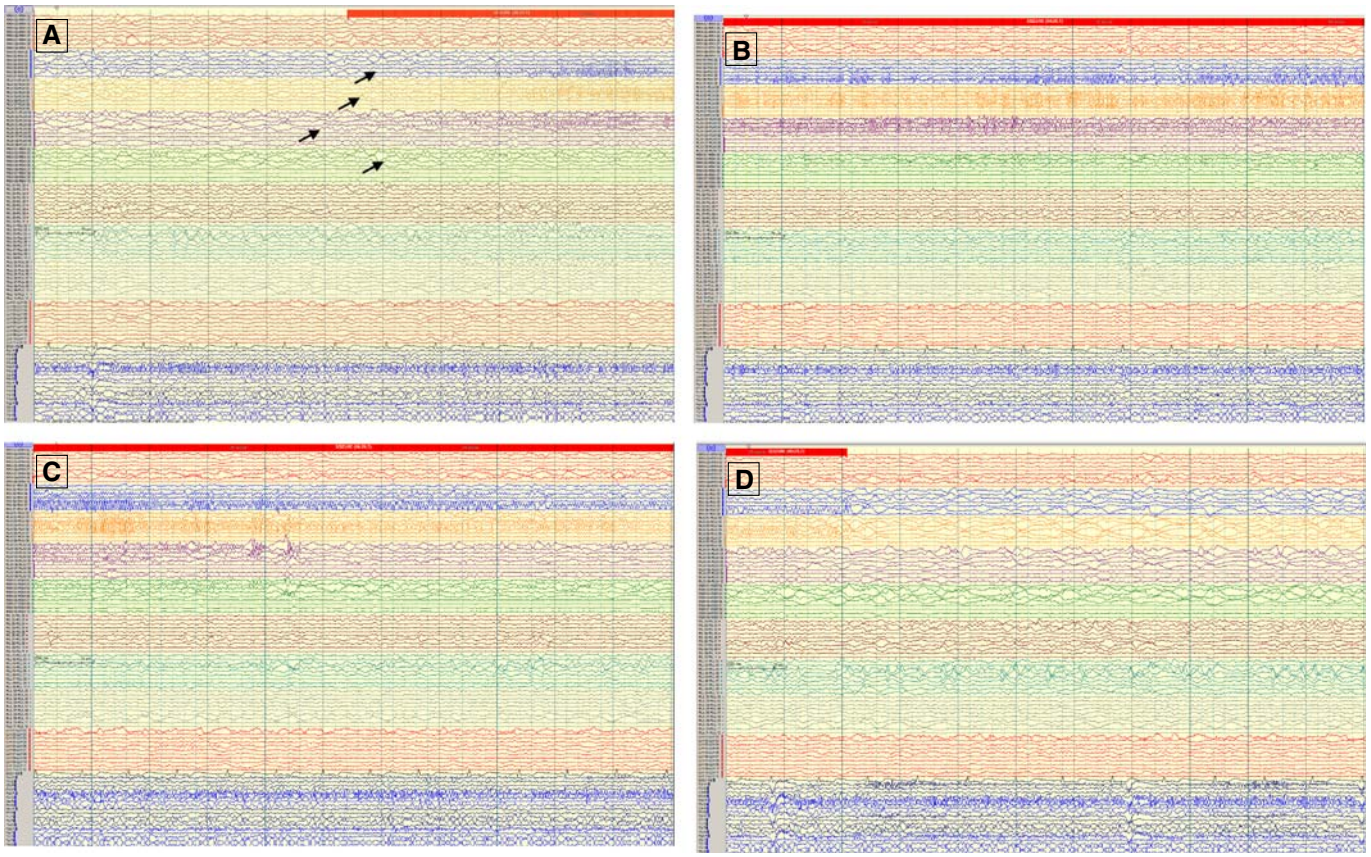

Fig. 3. (A–D) sEEG recording showing electrographic seizure (red bar) originating (onset marked with black arrows in A) from electrodes inferomedial to (RIM), within (RLM, RUM), and superomedial to (RSM) the occipital lesion.

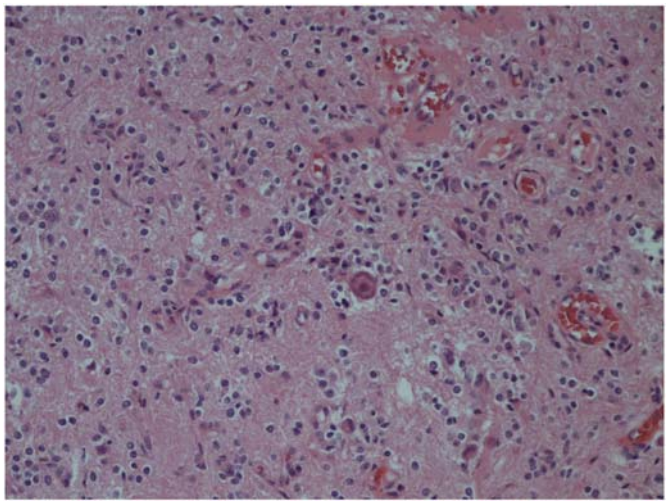

Fig. 4. Hematoxylin and Eosin X40 showing proliferation of Oligodendroglia like cells and neuron without dysplasia. The neurons were negative for CD34 (usually present in DNET in 61% cases). The oligodendroglia like cells had wild type IDH1 and IDH2 genes (by sequencing) and did not show 1p/19q codeletion (by FISH). The Ki-67 proliferation rate was low (<1%).

4. Discussion

Dysembryoplastic neuroepithelial tumors present as epilepsy with a mean age of onset of 18 years as found in review of multiple studies better representative of the patient population [2]. Studies suggesting an older mean age of onset were limited by sample size and higher number of adult non-DNET cases in their cohort [3]. Adult-onset epilepsy associated with DNET is not much different from its childhood-onset counterpart except for the low prevalence of cortical dysplasia in adult cases [3]. The temporal lobe is the most common location, followed by the frontal lobe, with the parieto-occipital region being a relatively rare location [1,3]. In our case, onset of epilepsy was age 30, and the lesion was entirely occipital. On the basis of MRI, DNETs have been classified into 3 types: type 1 (well-delineated cystic-like), type 2 (nodular-like), and type 3 (dysplastic with poor delineation) with higher association with cortical dysplasia [4]. The presence of an adjacent dysplastic cortex in DNET has been shown to require extensive resection [5] due to wider area of epileptogenicity demonstrated in sEEG [6]. The epileptogenic zone (EZ) on sEEG has been shown to correlate with MRI types: colocalized in type 1, perilesional in type 2, and extensive in type 3; thus MRI-based appearance of tumor can be used in guiding resection ranging from lesionectomy in type 1 to corticectomies in type 3 [4]. Our patient had features consistent with a type 2 DNET based on MRI, which corresponds to the nonspecific histological type with a perilesional EZ. sEEG confirmed a perilesional EZ and absence of any adjacent epileptogenic dysplastic cortex. However, interictal PET showed hypometabolism in the ipsilateral temporal lobe in addition to an active occipital lesion, raising the suspicion of right temporal network involvement the EZ of this patient given the semiology of TLE. sEEG in temporal DNETs has been shown to detect additional epileptogenic zone in the ipsilateral hippocampal area [4] and rarely in the contralateral hippocampal area [3]. In our case, sEEG showed seizures of exclusively right occipital origin with no spread to right hippocampus justifying a perilesional cortex removal. DNETs usually have good surgical outcome with mean seizure freedom rate of 86% on a median follow-up of 4 years [2] irrespective of age of onset, duration of epilepsy and age at surgery [3] and no need of postoperative radiation therapy [1]. In extratemporal DNET resections, the overall surgical outcome has been reported poor as compared to temporal resections [5]; however larger systematic reviews found no significant difference between temporal versus extratemporal resections [7,8]. However, in extratemporal resections, the seizure-freedom rate was superior in occipital DNETs as compared with frontal or parietal DNETs if incompletely resected when in the frontal and parietal lobes [9]. It is unclear whether these occipital DNETs presented with TLE semiology as in our case. Based on a resection tailored to the MRI and the EZ, as well as pathology confirming a nonspecific DNET without dysplastic cortex in occipital region, we expect our patient to maintain a good outcome.

We have described an unusual case of occipital DNET presenting with adult onset seizures; the clinical manifestations were characteristic of temporal rather than of occipital lobe seizures.

Disclosure

The authors declare no conflict of interest.

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