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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

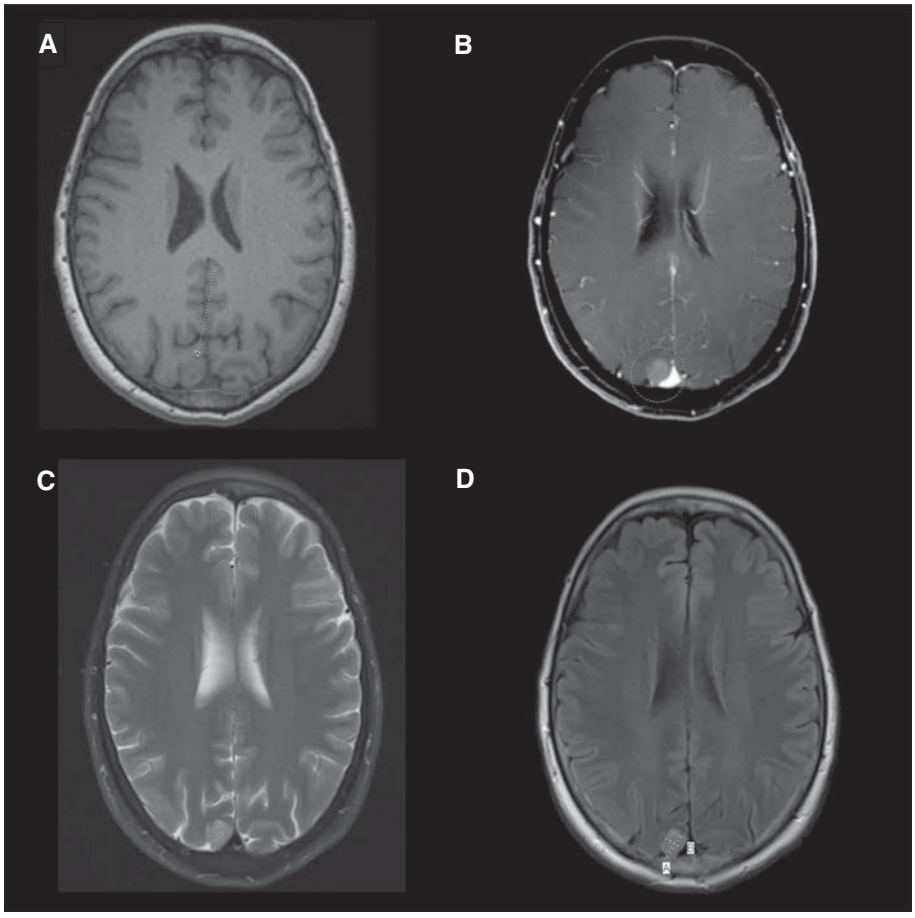

Fig. 1. MRI T1 axial view (A) showing rounded hypointense lesion. MRI T1-post (B) showing lesion with minimal abnormal homogenous enhancement. MRI T2 axial view (C) and FLAIR axial view (D) showing rounded hyperintense signal abnormality in the parasagittal right occipital lobe measuring 1.3 cm × 1.0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