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Awake Craniotomy with Neuronavigation Mapping for Parietal Dysembroblastic Neuroepithelial Tumor (Dnet) Causing Intractable Sensory Seizures in An Adolescent - A Case Report

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ABSTRACT

Dysembryoplastic neuroepithelial Tumors (DNET) are a rare, typically benign glial- neuronal cortical tumour that most commonly manifests in children and young adults as intractable seizures. The key benefit of awake craniotomy in multi-drug resistant intractable seizures caused by DNET in eloquent areas especially in and around motor cortex is to do safe maximal resection, preserving motor functions and seizure free outcome. A 16-year-old adolescent presented with refractory sensory seizures for 7 years. MRI Brain was done showing left parietal lesion suggestive of DNET with focal cortical dysplasia (FCD). Awake craniotomy was done with the help of neuro navigation and gross total resection of lesion was achieved. Biopsy reported as DNET with FCD. Post operatively patient was seizure free. DNET associated with FCD in parietal region is very rare. Cortical dysplasia is commonly associated with DNET and appears to contribute to the DNET's epileptogenic activity. For optimal disease treatment, surgical treatment should aim to remove DNET with FCD. We found awake craniotomy with neuro navigation helps to reset the lesion maximally with no or minimal neurological deficits.

Keywords: Awake craniotomy, Sensory seizures, Neuro navigation, Dysembryoblastic neuroepithelial tumor (DNET), Focal cortical dysplasia (FCD)

INTRODUCTION

Dysembryoplastic neuroepithelial tumors (DNET) are a rare, typically benign glial- neural cortical tumor that most commonly manifests in children and young adults as intractable seizures. DNET has a large area of epileptogenic activity that may be related to the presence of cortical dysplasia around it. The type of seizure activity depends on location of the lesion. Most common location of DNET is temporal lobe and least common being parietal lobe. Because of Cortical Dysplasia is frequently associated with DNET, attempting a complete removal of Cortical Dysplasia together with the primary lesion (DNET) may improve seizure free outcome. To achieve effective seizure control in DNET-associated epilepsy, thorough pre-operative studies for the precise identification of epileptogenic activity, meticulous brain mapping, and a very drastic resection of problematic areas may be required.

CASE REPORT

A 16year adolescent male presented with sensory seizures for the last 7 years. Seizure semiology starts with abnormal sensation in his right lower limb lasting for 2 -3 minutes, followed by feeling of numbness, tingling and heaviness over the right lower limb with loss of consciousness for 10 minutes. His first seizure episode started at the age of 9 years for which he was started on single anti-epileptic drug (AED). After 2 years he discontinued AED and was seizure free for 2 years. Again, he developed an episode of sensory seizure, for which he was again started on AED. No imaging was done at that time. Later the frequency of seizure episodes increased 4 to 5 episodes per week and AED was

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increased to four gradually. Then he was referred to our hospital, MRI brain with seizure protocol was done showing a cortical mass with hypointense in T1-weighted images (T1WI) and hyperintense in T2-weighted images (T2WI) and FLAIR sequences without surrounding perilesional edema or signs of mass effect. No contrast enhancement and appear as an enlarged heterogeneous gyrus, with delicate septa-like structures that are visible within the lesion, producing a soap-bubble appearance at the cortical margins reported as DNET with FCD (**Figure 1**). Electroencephalogram (EEG) was done, epileptogenic activity correlating with the lesion. All necessary Preoperative work up was done and planned for awake craniotomy with neuro navigation guidance and safe maximal excision of the lesion.

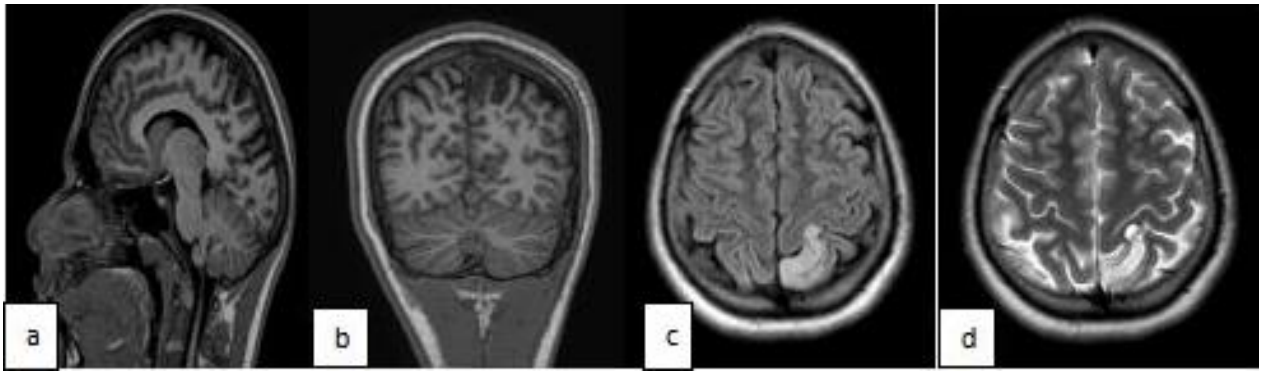

Figure 1. Preoperative images. (a & b) Saggital, Coronal T1-weighted MR image shows a cortical-based superficial lesion in left parietal lobe extending to the cortical margin. Axial FLAIR (c) T2-weighted axial image showing internal focal regions of high signal intensity, compatible with a "soap bubble" appearance (d) FLAIR axial view showing the lesion.

Under scalp block and LA patient in supine position, with the help of neuro navigation, skin incision marked, mini craniotomy done. Pre-operative EEG showed an epileptic focus. Lesion identified with help of neuro navigation. Intra operatively patient was advised to move his right upper and lower limb continuously. At a point where patient developed a mild right lower limb weakness the resection was stopped. Near total excision of lesion was done (**Figure 2**). Postoperative EEG showed no epileptic discharge.

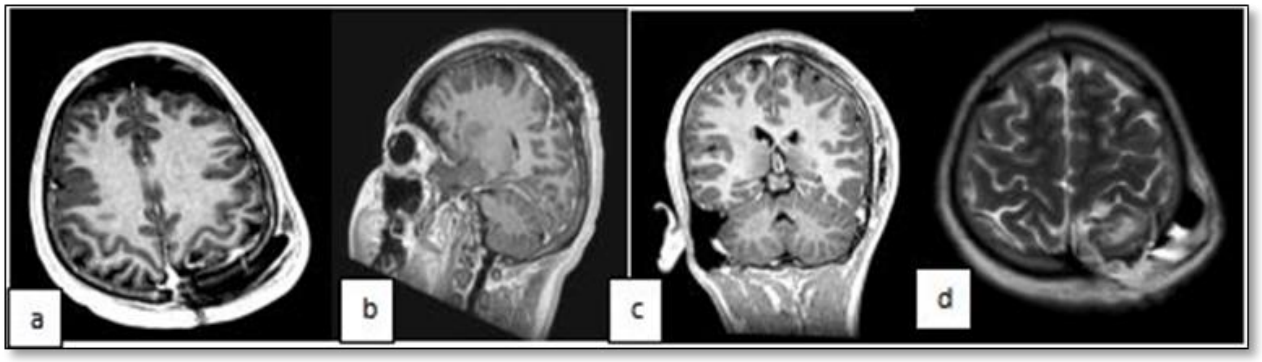

Figure 2. Post-operative images. (a) axial T1 post contrast (b)Sagittal post Contrast (c)Coronal post contrast (d) Axial T2 weighted image showing Gross total excision of lesion.

Histopathology reported as DNET with FCD. IHC markers: S100 - Positive on tumor cells. Synaptophysin - Positive on few floating neurons. GFAP-Negative (**Figure 3**).

DISCUSSION

Dysembryo plastic neuroepithelial tumors (DNETs) are an uncommon, benign type of tumor of neuroepithelial origin that arises from the cortical gray matter. Daumas-Duport coined the phrase "DNET". The temporal lobe (62%) and frontal lobe (31%) are the two most frequent sites [1]. DNETs can develop in the caudate nucleus, cerebellum, or pons, albeit the majority of them are only found in the cortical gray matter [2]. Intractable seizures are the typical presentation in terms of clinical symptoms. Cortical dysplasias are commonly seen with DNETs, occurring in nearly 50 % of cases [3]. Focal cortical dysplasia (FCD) is a diverse group of cortical lesions.

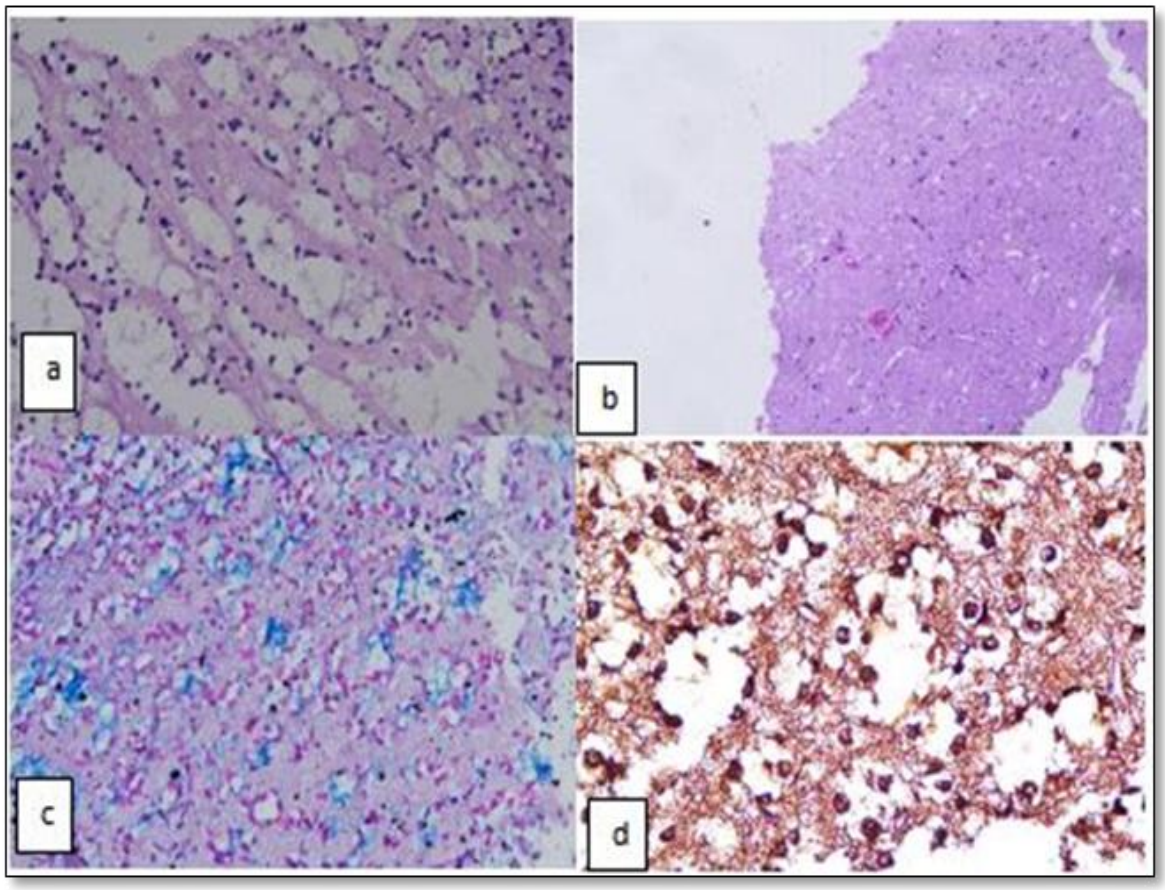

Figure 3. Histopathological findings. (a) Tissue section showing cystic spaces in cortical tissue suggestive of DNET with FCD. (b) Floating ganglion cells in microcystic spaces (c) Alacian blue stain showing mucinous material in cystic spaces. (d) IHC S100 positive.

In the review of literature, Lee [4] in his series of 23 patients only one had lesion in parietal lobe [4]. Takahashi [9] in his series of 24 cases no cases were reported in parietal lobe had sensory seizures. Prayson [5] in the series of 52 patients with cortical dysplasia only 4 had in parietal lobe [5].

The majority of cortical dysplasias are caused by neuronal migration abnormalities that occur before the twenty-fourth week of pregnancy [6].

DNET's are typically cortical lesions that lack significant mass effect or peri-tumoral edema. DNET typically shows up as intracortical masses on MR imaging that are hypointense on T1-weighted images and hyperintense on T2-weighted images without any perilesional edema or mass effect indicators in the surrounding area. Differential diagnosis are oligodendrogliomas, gangliogliomas, and astrocytomas.

DNETs can vary greatly in terms of size and shape, but they typically have a friable consistency and are mucinous, multinodular, or muticystic lesions [7]. Microscopically, DNETs are composed of multiple nodules containing both neuronal and glial components. They are characterized by an admixture of astrocytes and oligodendroglial elements, in association with "floating neurons" and mucinous degeneration [7]. The multinodular pattern and areas of mucinous degeneration observed on histopathology may contribute to a soap- bubble appearance on neuroimaging. Other common histological features include "specific glioneuronal elements" oriented in a columnar pattern, perpendicular to the cortical surface and focal areas of cortical dysplasia [8].

We could see that this epilepsy surgery-oriented approach could cure patients with DNET, which is essentially a benign tumor. Patients can be free of tumor recurrence with complete tumor resection and seizure without antiepileptic drugs with complete resection of epileptogenically active areas in and around the tumor [9].

Complete removal of the lesion (total resection of the tumor microscopically and macroscopically) may be required to

achieve favorable seizure control, but it is not sufficient to achieve cure levels (complete seizure freedom without medication). We believe that DNET, particularly when associated with CD, may have a similar epileptogenic mechanism as intractable epilepsy associated with CD, which we previously reported [10].

Excellent seizure control could be attributed to a combination of thorough preoperative evaluation, radical resection, and meticulous electrophysiological investigation.

CONCLUSION

DNET is commonly associated with Cortical dysplasia and appears to contribute to DNET's epileptogenic activity. For optimal disease treatment, surgical treatment should aimto remove both DNET and FCD. Very few cases of parietal DNET with FCD causing pure intractable sensory seizures were reported in literature so far makes this case rare entity. Awake craniotomy along with neuro navigation helps to achieve safe maximal resection in eloquent areas with minimal or no deficits.

DECLARATION OF PATIENT CONSENT

The authors certify that they obtained all appropriate patient consent forms. In the form the patient has given his images and other clinical information to be reported in journal. The understood that his name and initial will not be published and due to efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

CONFLICT OF INTEREST

The authors declared that there are not conflicts of interests.

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