# Case report

## Focal cortical dysplasia with underlying dysembryoplastic neuroepithelial tumor: A Case Report

Ashwini Nagda[1], Poorvi Agrawal[1], Poonam Wade[1]

**Author's Affiliation:**
1- Topiwala National Medical College and BYL Nair Ch hospital, Mumbai, Maharashtra, India.

**Correspondence:**
Ashwini Nagda, Email: nagdaashwini@gmail.com

Received on: 29-Sep-2021 Accepted for Publication: 10-Sep-2025

**Abstract**

Epilepsy associated with slow growing brain tumors may frequently be the cause for pharmaco-resistant focal seizures in young children. Dysembryoplastic neuroepithelial tumor is one such entity which may cause epilepsy resistant to multiple antiepileptic agents. Focal cortical dysplasia, a congenital cortical developmental anomaly which may be seen associated with DNET, independently acts as an epileptogenic focus. Early diagnosis of such lesions is necessary. Long term use of antiepileptic drugs (AEDs), needed for control of tumor related epilepsy, might affect the cognitive development of the child. Complete surgical removal of the tumor along with overlying dysplasia is one of the modalities for making the patient seizure free. However, the risks of surgery and neurological development post-surgery should also be considered while deciding on the management of LEATs in pediatric patients.

### Introduction

The occurrence of DNETs is 0.03 person-year per 100,000 with highest incidence between 10 and 14 years.[1] It is a mixed neural glial WHO grade 1 neoplasm causing early onset, drug resistant, partial epilepsy in children. Early diagnosis using imaging provides the patient with an additional option of surgical treatment.

**Keywords**: Dysembryoplastic neuroepithelial tumor (DNET), Focal cortical dysplasia (FCD), long term epilepsy associated brain tumors (LEAT), pharmaco-resistant epilepsy, epileptogenic tumor.

### Case report

8-year-old developmentally normal female child, first by birth order, born of non-consanguineous marriage, presented with complaints of 3 episodes of tonic clonic movements of left upper and lower limbs with twitching of eyelids, with preserved consciousness over a span of 6 hours. Patient did not lose consciousness in between these episodes. MRI brain was suggestive of focal cortical dysplasia. Electroencephalogram (EEG) was normal. Patient was started on Levetiracetam 20mg/kg/day. Patient was compliant to medications. Therapeutic drug monitoring (TDM) levels sent for Levetiracetam were 24mg/L which was within the therapeutic range.

Patient again had convulsions in the form of colonic movements of both upper limbs with preserved awareness after an asymptomatic period of 2 months. Dose of Levetiracetam was increased to maximum

therapeutic dose and Oxcarbazepine was added at 6mg/kg/day. In spite of dual antiepileptics, convulsions persisted. Patient was loaded with a third AED, Valproate, at 20mg/kg and dose of Oxcarbazepine was increased to maximum therapeutic dose. A fourth AED, Clobazam was also added as seizures were persistent. Electrolyte and sugar disturbances were ruled out. EEG was repeated which was normal. Repeat MRI brain showed features suggestive of dysembryoplastic neuroepithelial tumor (DNET) with overlying focal cortical dysplasia in inferior frontal gyrus of left side. Seizures were controlled on polytherapy, hence no active neurosurgical intervention taken. Patient was discharged successfully on 4 AEDs.

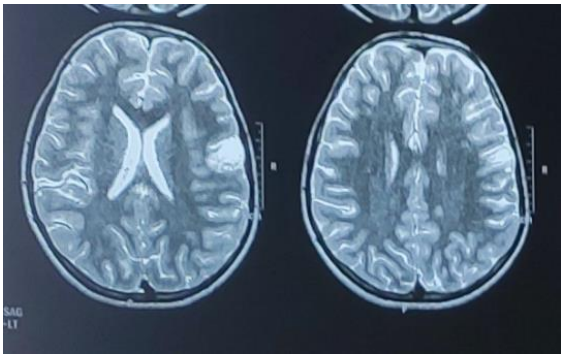

Figure 1. T2 weighted MRI showing hyperintense lesion

## Discussion

Focal cortical dysplasia (FCD) is a congenital abnormality of cortical development is one of the most common causes drug resistant focal epilepsy in children.[2, 3, 4] It is classified into 3 main types and 9 subtypes according to Blumcke et al, where lesions associated with developmental tumors, such as DNET areclassified as IIIb. These lesions cause a positive mass effect, have a cystic component, various calcifications and show contrast enhancement.[5]

A variety of brain tumors have the potential to cause early epilepsy in children and young adults collectively called 'long term epilepsy associated brain tumors (LEAT). LEAT encompasses astrocytoma, ganglioma, glioma and DNET. DNETs are benign, supratentorial, cortical low-grade tumors (WHO grade I or WHO grade II).[6] FCDs are usually peripheral to the tumor but may also be present within the tumor.

A study conducted by Chan et al. evaluated outcomes of epilepsy in children undergoing surgical resection of temporal DNETs. Complete tumor resection showed favorable outcomes and recurrence was not seen. Persistent seizure activity was frequently seen in cases where the tumor was incompletely resected. Repeat surgery in such children showed improved control of epilepsy.[7] Another study has also shown that children respond better to surgery than adults. Earlier the surgery performed, the better the seizure control.[8]

Brain tumors disrupt interneuronal networks locally as well as in distal areas of cerebral hemisphere by the phenomenon of neuroplasticity. Low-grade, slow growing tumors, hence, show greater disruption of cortical networks as they have more time for neuroplasticity to take place. This explains the existence of

epileptogenic foci distant to the tumor location.[9] This poses a challenge to surgical treatment as the surgical excision would have to include other epileptogenic foci. This can be achieved by intraoperative EEG recording.

It is necessary to bear in mind the risks of surgery in young children as well as risk of perioperative mortality. Neurosurgical procedures can have adverse effects on the neurological development and function of the young child.[10]

In the last decade or so, many new AEDs have been introduced which show promise, both, in terms of efficacy and tolerability. Large scale studies further evaluating these AEDs are needed for better control of tumor-related epilepsy.[11]

**Conclusion**

Convulsions caused by DNETs and FCDs are difficult to treat and may require polytherapy with multiple AEDs which may affect the cognition and learning of the child. Complete surgical excision of the tumor and all epileptogenic foci may be considered in pharmaco-resistant, difficult to treat epilepsies while keeping in mind the risks associated with surgery. Newer AEDs have proven relatively more tolerable and may be used to overcome these challenges.

**References**:


1. Luzzi S, Elia A, Del Maestro M, Elbabaa SK, Carnevale S, Guerrini F, Caulo M, Morbini P, Galzio R. Dysembryoplastic neuroepithelial tumors: what you need to know. World neurosurgery. 2019 Jul 1;127:255-65.
2. Piao Y-S, Lu D-H, Chen L, et al. Neuropathological findings in intractable epilepsy: 435 Chinese cases. Brain Pathol. 2010;20:902-908.
3. Daumas-Duport C, Scheithauer BW, Chodkiewicz JP, Laws ER, Vedrenne C. Dysembryoplastic neuroepithelial tumor: a surgically curable tumor of young patients with intractable partial seizures. Report of thirty-nine cases. Neurosurgery. 1988;23:545-556.
4. Aronica E, Becker AJ, Spreafico R. Malformations of cortical development. Brain Pathology. 2012 May;22(3):380-401.
5. Blümcke I, Thom M, Aronica E, Armstrong DD, Vinters HV, Palmini A, Jacques TS, Avanzini G, Barkovich AJ, Battaglia G, Becker A. The clinicopathologic spectrum of focal cortical dysplasias: a consensus classification proposed by an ad hoc task force of the ILAE Diagnostic Methods Commission 1.
6. Louis DN, Perry A, Reifenberger G, et al. The 2016 World Health Organization classification of tumors of the central nervous system: a summary. Acta Neuropathol. 2016;131:803-820.
7. Chan CH, Bittar RG, Davis GA, Kalnins RM, Fabinyi GC. Long-term seizure outcome following surgery for dysembryoplastic neuroepithelial tumor. Journal of neurosurgery. 2006 Jan 1;104(1):62-9.
8. Giulioni M, Marucci G, Pelliccia V, Gozzo F, Barba C, Didato G, Villani F, Di Gennaro G, Quarato PP, Esposito V, Consales A. Epilepsy surgery of "low grade epilepsy associated neuroepithelial tumors": a retrospective nationwide Italian study. Epilepsia. 2017 Nov;58(11):1832-41.
9. Esposito R, Mattei PA, Briganti C, Romani GL, Tartaro A, Caulo M. Modifications of default-mode network connectivity in patients with cerebral glioma. PLoS One. 2012 Jul 9;7(7):e40231.

10. Ranger, Adrianna, and David Diosy. "Seizures in children with dysembryoplastic neuroepithelial tumors of the brain--A review of surgical outcomes across several studies." Child's nervous system : ChNS : official journal of the International Society for Pediatric Neurosurgery vol. 31,6 (2015): 847-55. doi:10.1007/s00381-015-2675-9
11. Rudà R, Trevisan E, Soffietti R. Epilepsy and brain tumors. Current opinion in oncology. 2010 Nov 1;22(6):611-20.