

Case Report: Frontal Lobectomy with Electrocorticography in a Pediatric Patient with Focal Cortical Dysplasia

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ABSTRACT

A 5 year old male with no past medical history presented with generalized tonic clonic seizures. Brain magnetic resonance imaging (MRI) demonstrated blurring of grey-white matter and lobar hypoplasia of right frontal lobe. Right frontal lobectomy with electrocorticography (ECoG) was performed. The final pathological diagnosis was focal cortical dysplasia (FCD) type IIa. The patient was classified as Engel class I after surgery and postoperative MRI demonstrated preservation of the right lateral ventricle.

Keywords: Cerebral cortical dysplasia, electrocorticography, epilepsy surgery, pediatric.

Abbreviations: FCD: Focal Cortical Dysplasia; MCD: Malformations of Cortical Development; AED: Anti-Epileptic Drug; ECoG: Electrocorticography; GTCS: Generalized Tonic-Clonic Seizures; EEG: Electroencephalography; MRI: Magnetic Resonance Imaging; CSF: Cerebro-Spinal Fluid; ILAE: International League against Epilepsy; FDG-PET: Fluorodeoxyglucose Positron Emission Tomography

INTRODUCTION

Epilepsy is the most common chronic neurological disease, affecting more than 65 million people worldwide [1]. Focal cortical dysplasia (FCD) is a pathological entity that belongs to a group of disorders called malformations of cortical development (MCD). FCD is a common cause of focal epilepsy in children and approximately 75% of the patients suffer from antiepileptic drug (AED) resistant epilepsy [2]. Surgery has been a treatment option for patients with certain types of epilepsy for more than 100 years [3]. FCD often requires surgical intervention to remove the affected area.

We report a patient with lesional epilepsy secondary to FCD type II who had an outstanding improvement following epilepsy surgery with electrocorticography (ECoG).

CASE

This 5 year old male, originally from Brazil, debuted in 2016 with generalized tonic-clonic seizures (GTCS) around the time of awakening. The event lasted for at least 40 s and the patient was admitted for further neurological evaluation. The

patient had no past medical history. During hospital stay, the patient presented 4 more episodes around 5:00 to 6:00 in the morning and myoclonic events were observed at night; levetiracetam was initiated. Electroencephalography (EEG) revealed persistent epileptiform activity of the right frontal lobe (**Figure 1**) and brain magnetic resonance imaging

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(MRI) shows signs suggestive of FCD of the right frontal lobe (**Figure 2**). FCD is a common cause of medically refractory epilepsy in children [4] and surgery is a good treatment option for this pathological entity. Before surgical treatment

the patient presented 6 more crises and clobazam was added to the medications. Neuropsychological testing reported developmental dyspraxia and language delay (dysarthric-dysphasic type), both associated with a developmental disorder secondary to epilepsy.

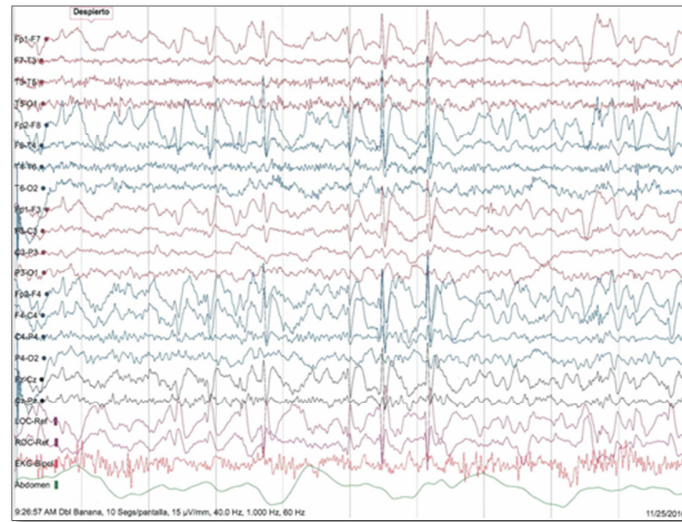


Figure 1. EEG. Shows persistent epileptiform activity in the frontopolar and right frontal lobe.

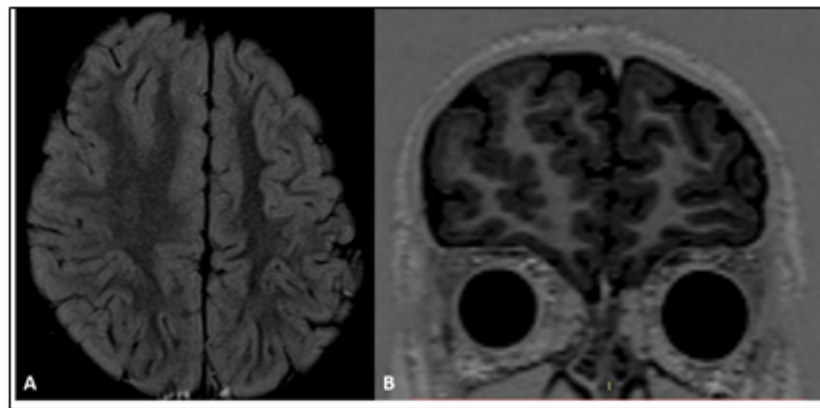


Figure 2. MRI scan of the brain. A) Axial T2 WI showing blurring of grey-white matter in the right frontal lobe. B) Coronal T1 WI revealed medial lobar hypoplasia of the right frontal lobe.

He underwent a frontal lobectomy with intraoperative ECoG and placement of subdural grids (**Figure 3**). During surgery the intracranial monitoring demonstrated the localization of the epileptogenic zone near to the supplementary motor area. When performing the resection of the affected frontal lobe the preservation of the right lateral ventricle was achieved. The postoperative course was uneventful. Cerebral tissue was sent to neuropathology for examination and the final pathological diagnosis of resected right frontal lobe was FCD Type IIa (**Figure 4**) with polymicrogyria and

pachygyria. His follow-up brain MRI, taken 6 months after his surgery, indicated successful resection of the lesion, resect area is filled with cerebro-spinal fluid (CSF) and the right lateral ventricle is preserved (**Figure 5**). The patient remains Engel Class I and showed an improvement in language and cognition. The patient goes to a follow-up clinic, 18 months after surgery, the child understands three languages (Spanish, Portuguese and English), free of seizure, Engel IA.

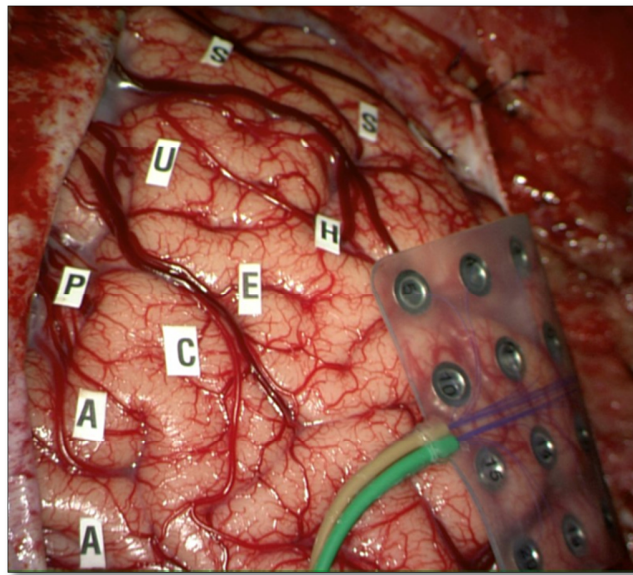


Figure 3. Open right frontal craniotomy window revealing the implantation of invasive monitoring.

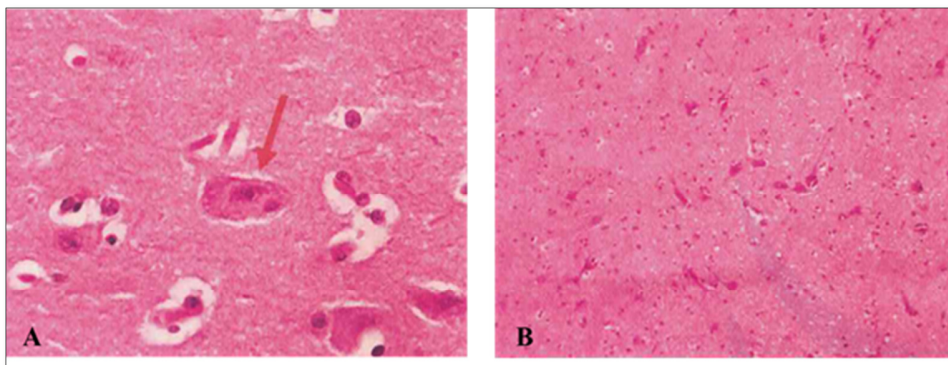


Figure 4. A) Dysmorphic neurons (red arrow) with moderate cytoplasm. B) Ectopic neurons in white matter. The absence of balloon cells and presence of dysmorphic neurons fulfill the criteria for FCD Type IIa.

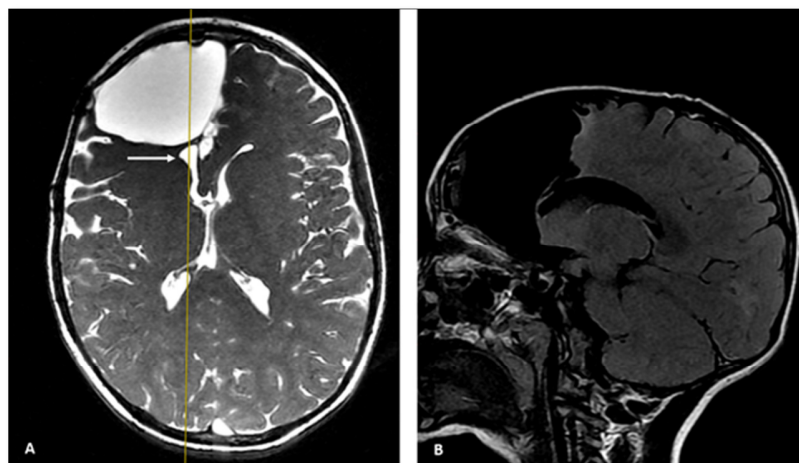


Figure 5. Postoperative MRI scan of the brain 6 months after. A) Axial T2 WI FIESTA sequence showing normal postsurgical changes and resection area is filled with CSF with no communication with the right lateral ventricle (white arrow). B) Sagittal T2 WI FLAIR showing preservation of the right lateral ventricle.

DISCUSSION

Malformation of cortical development (MCD) is a group of lesions in the brain resulting from an abnormal process in cortical development by multiple etiologies [4]. Cortical development of the brain involves a burden of processes that include neuronal stem cell proliferation, migration and differentiation. These pathological entities are a common cause of epilepsy and developmental delay [2]. FCD was first described in 1971 by Taylor et al. as they gave a detailed explanation of a specific malformation that consisted of a disorganized cortex with irregular neurons and balloon cells [5].

In 2011, the International League against Epilepsy (ILAE) Task Force for Neuropathology, proposed the currently used classification for FCD that divides it into 3 groups based on histological findings (**Table 1**). Our young patient presented with a Type IIa FCD, which is characterized by the presence of dysmorphic neurons, an absence of balloon cells, the impossibility to discriminate between cortical layers called cortical dyslamination and the difficulty to delineate the border between gray and white matter; other cortical layer abnormalities might be present [6]. Even though FCD can affect any lobe, Type II seems to have a predilection for the frontal lobe in contrast with Type I predilection for the temporal lobe [7].

Table 1. ILAE classification of focal cortical dysplasias.

FCD type I	Cortical dyslamination mixed with normal neurons and glia.
Ia	Abnormal radial cortical lamination (micro columnar)
Ib	Abnormal tangential cortical lamination
Ic	Abnormal radial and tangential cortical lamination
FCD type II	Presence of dysplastic megalocytic neurons mixed with normal neurons.
IIa	Dysmorphic neurons.
IIb	Dysmorphic neurons and balloon cells
FCD type III	Cortical dyslamination with normal neurons adjacent to another lesion.
IIIa	Associated with HS in the temporal lobe
IIIb	Glial or glioneuronal tumor
IIIc	Vascular malformation
IIId	Any lesion acquired during life (e.g. trauma, ischemic injury)

FCD: Focal Cortical Dysplasia; ILAE: International League against Epilepsy; HS: Hippocampal Sclerosis

Epilepsy is the most common clinical presentation of all types of FCD. Seizures usually begin early in childhood and the anatomical location of FCD will determine the semiology of the crisis, nevertheless, the most common type of seizure at presentation are GTCS followed by tonic seizures, focal onset aware seizures, and focal impaired awareness seizures [8,9]. MRI is an effective tool for identifying FCD, abnormalities were seen in 30% of Type I cases, 55% in Type IIa and 80% in Type IIb in a study by Leach et al. [10]. These abnormalities include cortical thickness changes, signal increase and blurriness of the gray/white matter [5]. The “transmantle sign” in MRI is an important sign for FCD Type II but is almost exclusive for subtype IIb; in fact, Type IIa is harder and not always identified on *in vivo* MRI [6]. Type II FCD frequently shows characteristic EEG changes such as focal rhythmic interictal epileptiform discharges that correlate with the anatomic extent of the lesion [11]. Fluorodeoxyglucose positron emission tomography (FDG-PET) is highly sensitive in

detecting FCD represented by focal regional hypometabolism [8].

FCD II has been recognized as a major cause of AED resistant epilepsy [2]. Patients with drug-resistant epilepsy secondary to FCD are candidates for epilepsy surgery. In patients with FCD a complete resection is necessary, and this can be achieved with the use of intraoperative monitoring [12]. Prior surgical intervention a complete preoperative evaluation should be addressed, and it should include the following: detailed clinical history, video EEG, MR and neuropsychological testing [13]. The use of ECoG is indicated in patients with extra temporal seizures, with discordant results in the preoperative tests, and in bilateral temporal epilepsy [13]. Seizures are not typically captured on the ECoG recording. The “recruiting rhythm” an electrographic seizure on ECoG, has been reported to be a reliable marker of the epileptogenic zone in cortical dysplasia [14]. As mentioned before FCD is a common cause of intractable epilepsy in children, the use of ECoG in pediatric patients is reported in literature and approximately

35% of the patients will benefit from it [12]. Our 5 year old patient had a proper preoperative evaluation and the decision to make use of ECoG with intracranial grids was for establishing the localization of the epileptogenic zone and for determining the extent of the resection.

CONCLUSION

FCD is a pathological entity that may cause disabling seizures especially in children. Intraoperative monitoring allows neurosurgeons to have more precise resection limits with the aim to improve quality of life. We were able to preserve the lateral ventricle despite the size of the resected tissue. Our patient remains Engel class IA and he has shown great improvement in language and cognition.

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