



Case Report

Neurosurgical treatment of nonconvulsive status epilepticus due to focal cortical dysplasia



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ABSTRACT

We present a rare case of focal cortical dysplasia (FCD) and nonconvulsive status epilepticus (NCSE) treated successfully with early surgical intervention. Our case is a 9-year-old boy whose seizures, characterized by short episodes of loss of consciousness, appeared at the age of 7, and he showed progressive cognitive decline in the following years. NCSE was diagnosed, and his MRI revealed FCD in the left frontal region which was the same side as his EEG abnormality. Following lesionectomy, his NCSE disappeared and cognitive functions improved. Histopathologic analysis of the resected tissue revealed type-II B FCD. This case illustrates the importance of early surgery to help restore cognitive functions by eliminating the clinical and electrophysiological features of NCSE.

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1. Introduction

Nonconvulsive status epilepticus (NCSE) presents with clinical signs such as unexplained changes in behavior and mental status, confusion, or even a severe tendency to sleep, accompanied by almost continuous epileptiform activity in electroencephalography (EEG) [1]. NCSE is observed mainly during the clinical course of patients with drug-resistant epilepsy. Recognition of NCSE especially in pediatric patients with epilepsy is very important for optimal mental development. We present a case of NCSE, which developed in a 9-year-old male with focal cortical dysplasia (FCD) and was successfully treated with neurosurgery.

2. Case report

A 9-year-old boy was born at term without any remarkable perinatal problems. There was no history of epilepsy in his family. His motor development was also normal. He began to have episodes of unconsciousness lasting a few minutes when he was 7 years old. Despite the administration of valproate, carbamazepine, phenytoin, levetiracetam,

clobazam alone or in combination, seizures recurred 2 or 3 times a month. At 8 years of age, sharp wave discharges were seen over the frontal regions predominantly over the left side in his EEG but the report for his magnetic resonance imaging (MRI) was normal. On The Wechsler Intelligence Scale for Children-Revised (WISC-R), he scored 81 on his total intelligence quotient (TIQ) testing which is in normal range of intellectual functioning at that time. Since he was 8 years old, he had been taking levetiracetam, clobazam and phenytoin. His family indicated that he had episodes of loss of consciousness lasting for 20 min, 5 to 6 times daily and he began to have a marked decline in school achievement at the age of 9. On the WISC-R, given at the time, he achieved a TIQ of 68, which is indicative of mild mental retardation. This finding suggested that the episodes of NCSE had a negative impact on mental activity even over a short time. Originally a patient at the pediatrics department, he was coincidentally referred to our laboratory for EEG. His EEG was recorded during wakefulness and showed almost continuous generalized spike-wave discharges compatible with left frontal secondary bilateral synchrony, consistent with NCSE (Fig. 1A-B). During this EEG recording, his communication was limited and he was slow in responding to verbal commands. His spontaneous speech was not fluent and he could not even do simple mathematical calculations. He was able to perform only some basic tasks. After intravenous diazepam administration abnormal electrophysiological activity was suppressed, although there was no prominent clinical change. Although

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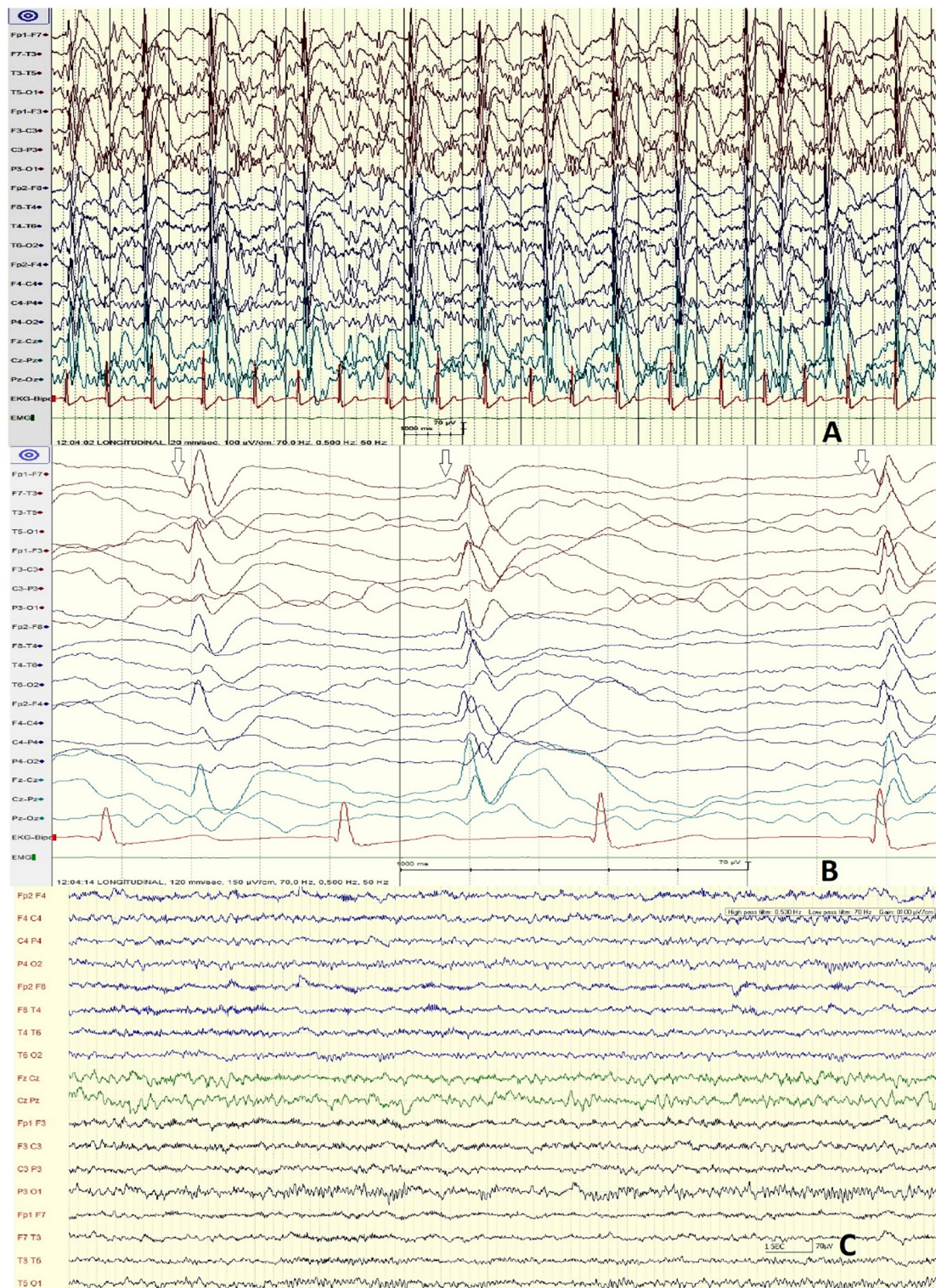


Fig. 1. Preoperative EEG (A–B). A: Continuous generalized spike–wave discharges with left frontal secondary bilateral synchrony and high amplitude. B: Left frontal discharges antecedent and extending to the right hemisphere when amplitude is decreased. C: Postoperative EEG showed no epileptiform discharges.

there was no evidence of electrical status epilepticus during sleep, spikes and/or sharp waves were observed in the frontal regions predominantly on the left side in NREM sleep.

His MRI demonstrated a lesion suggestive of FCD in the anterior cingulate gyrus of the left frontal lobe (Fig. 2A). There were no abnormalities in his blood tests. After preoperative evaluations, he underwent complete lesionectomy (Fig. 2B). Neuropathology of the resected brain tissue revealed FCD type IIB. The specimen was cut into 2–3 mm slices,

perpendicular to the cortical surface. Histopathological examination showed a distorted cortical lamination and absence of important amount of the neurons; besides, there were large, bizarre shaped dysplastic neurons and balloon cells with eosinophilic glassy cytoplasm at unusual locations. Immunohistochemically, using the neurofilament (NF) antibody, dense accumulation of NF was observed in these large neurons (Fig. 2C–D). During the postoperative EEG recording, his communication was better, his speech was more fluent and he was giving

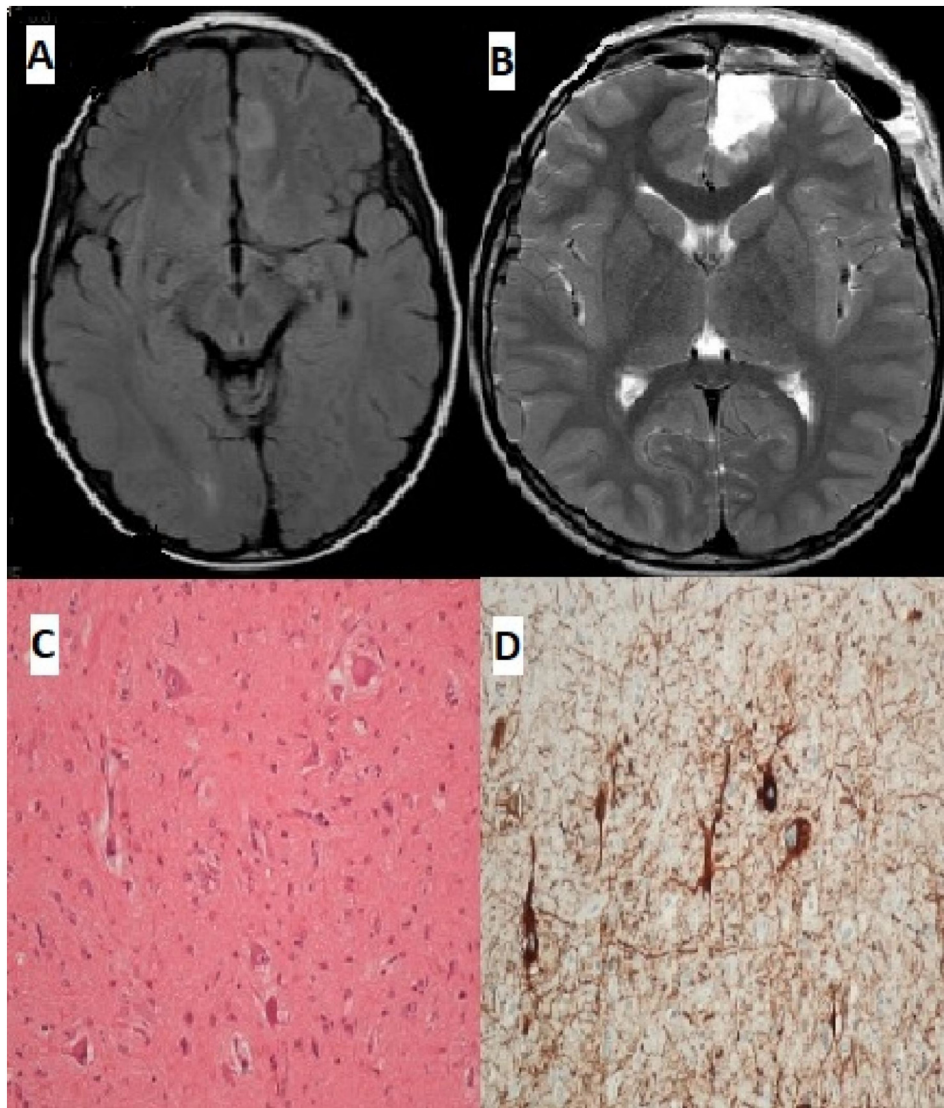


Fig. 2. Preoperative fluid attenuated inversion recovery (FLAIR) axial (A) MRI image showing increased signal, cortical focal thickening, blurring of the gray–white matter junction, an abnormal sulcal and gyral pattern in the region of anterior cingulate gyrus of the left frontal lobe compatible with focal cortical dysplasia. Postoperative T2 weighted axial (B) MRI image showing that the lesion has been totally removed. (C) Cortical dysplastic neurons and balloon cells at abnormal locations. HE X200. (D) Neurofilament accumulation in cytoplasm of the abnormal neuronal cells. Neurofilament, immunoperoxidase X200.

correct answers to questions. His awake and sleep EEG showed no spike discharges interictally (Fig. 1C). NCSE was not seen after surgery and he was seizure-free at 5 months follow-up examination.

3. Discussion

Our case demonstrates that NCSE may rarely develop and be successfully treated with surgery in children with epilepsy due to FCD. Most cognitive deficits resulted from FCD located in the frontal lobe, NCSE may exacerbate cognitive and behavioral regression if not prevented.

In our case, the exact time of the clinical onset of status epilepticus is not known. However, there was probably a long delay between the clinical onset of NCSE and EEG confirmation. Similarly, Miyama reported a 6-year-old patient, whose EEG documented continuous paroxysmal discharges in the left frontocentral/anterior temporal areas for almost 3 years before the development of NCSE was discovered due to focal cortical malformation in the left frontal lobe [2]. We considered that the rapid cognitive decline of the patient in the last one year may be related to frequent seizures and NCSE.

In the previous EEG report of our case recorded elsewhere, sharp wave discharges were detected in the anterior regions of bilateral hemispheres predominantly over the left side, suggesting that NCSE is associated with the left frontal lobe. The epileptiform discharges were seen to be generalized predominantly over the left side in our recording as well. When the amplitude of the discharges was reduced and (Fig. 1B), the discharges originated from the left frontal region, which is known as secondary bilateral synchrony [3]. Even when the MRI is normal, similar severe EEG findings, should suggest a hypothetical epileptogenic region for surgery. Accurate diagnosis of epilepsy, appropriate medical treatment, and potential surgical candidacy rely on distinguishing secondary bilateral synchrony from primary bilateral synchronous discharges. Various noninvasive methods such as independent component analysis of EEG and magnetic source imaging can be used to identify and analyze the focus of secondary bilateral synchrony [4,5]. In our case, it was easy to decide on the treatment approach due to his FCD lesion.

In addition, in children, there may be variants of electrical status epilepticus during slow wave sleep (ESES). Epilepsy syndromes such as CSWS, and LKS should be considered in this age group of epileptic children who have continuous cognitive decline. LKS is a syndrome

characterized by acquired receptive aphasia and epilepsy. They are both age-related epileptic encephalopathies that represent the most severe childhood seizure susceptibility syndrome. Their characterizing features are seizures, interictal epileptiform activity that becomes prominent during sleep leading to the EEG pattern of ESES and neurocognitive regression. Our case had none of these clinical or electrophysiological findings that align with either ESES or LKS.

FCD is a well-recognized etiology of drug-resistant epilepsy in the pediatric population but successful surgery for NCSE has rarely been described in FCD. NCSE was documented in one of the 120 patients with FCD reported in a study [6]. Similar to our case, Yoshimura reported a 6-year-old patient with FCD in the left frontal region who developed “generalized” NCSE [7]. Unlike our case, their patient was treated medically. Ng et al. reported on a pediatric patient with complex partial status treated with resection of epileptogenic area associated with FCD [8]. However, they also performed multiple subpial transection (MST) with focal resection. Therefore, in their case MST may have contributed to treatment of status epilepticus in addition to resection. Duane et al. reported a 7-year-old boy with NCSE treated with hemispherectomy [9]. In these case reports, more complicated operations such as MST, focal resection, hemispherectomy were applied as treatment for status epilepticus. However, we performed only lesionectomy following which NCSE was terminated and there was no seizure recurrence as well as cognitive improvement.

4. Conclusion

While NCSE is difficult to treat, early Intervention is crucial for the prevention of cognitive decline. Our case highlights a less recognized presentation of FCD which was treated successfully with surgical intervention as well as outlining the importance of early surgery in children

with NCSE if an epileptogenic focus can be found. In such cases, early recognition and control of NCSE contribute to cognitive recovery.

Ethical statement

Informed consent was obtained from the patient.

Conflict of interest

The authors have no conflicts of interest to report.

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