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Early Myoclonic Epileptic Encephalopathy in a Newborn: Successfully Control Seizures and Suppression-Burst EEG with Levetiracetam

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1. Abstract

Early epileptic encephalopathy with Suppression-Burst (SB) in Electroencephalography (EEG) comprises two epileptic syndromes, Early Infantile Epileptic Encephalopathy (EIEE) and Early Myoclonic Encephalopathy (EME). The SB EEG abnormalities are believed to contribute to the progressive disturbance in cerebral function. The patients with the epileptic syndromes with onset either immediately after birth or in the first months of life and characterized by erratic, fragmentary and massive myoclonus, partial seizures, and late tonic spasms. Those newborns with SB patterns in EEG, the outcomes usually are grave and seizures are difficult to be treated. We report a neonatal EME, in whom, seizures and SB activities were successfully controlled by intravenous levetiracetam.

2. Introduction

Early epileptic encephalopathy with Suppression Burst (SB) comprises two distinct epileptic syndromes, Early Infantile Epileptic Encephalopathy (EIEE) and Early Myoclonic Encephalopathy (EME). Suppression-Burst (SB) activity of Electroencephalography (EEG) was defined the EEG as a pattern of high-amplitude and slow-waves discharges with or without spikes, being alternating with periods of minimal activity of low amplitude (less than 10 uV) [1-3]. The epileptiform EEG abnormalities themselves are believed to contribute to the progressive disturbance in cerebral function and dysfunction of cognition. Literatures have been published that describe an epileptic syndrome with onset either immediately after birth or in the first months of life and characterized by erratic, fragmentary and massive myoclonus, partial seizures, and late tonic spasms [2-4]. Those newborns with SB patterns in EEG, the outcomes usually are grave and seizures are difficult to be treated [5-6]. We report a female neonatal EME. Her seizures and EEG SB activities were successfully controlled by intravenous levetiracetam.

3. Case Report

A female patient was born from a normal mother with 39 weeks gestational age after an uneventful pregnancy, with birth weight of 2800 gm. Her family history was unremarkable. She was referred from local clinics due to poor feedings and seizure-like behaviors on day 3 after birth. The first day after admission, she exhibited many times of tremble-like movements of the lower limbs and trunk. Her activity was depressed. In addition, she had poor sucking power, and was difficult to be orally fed more than 10 ml of breast milk. On second day of admission, she started frequent apneas and myoclonic seizures, particularly during sleep. A blood count showed her hemoglobin at 14.3 g/dL (reference range, 13.88 ± 1.34 g/dL), white blood cell count at 13000 mm^3 μL (reference range, 9100–34000 mm3 μL), and platelet count at 259 mm3 μL(reference range, 84–478 mm3 μL). A c-reactive protein test was at 0.15 mg/dL (reference range, 0.09-1.58 mg/dL). The Cerebral Spinal Fluid (CSF) examination demonstrated normal finding. A blood gas analysis exhibited no metabolic acidosis. Her serial blood lactates varied from 4 to 16 mg/dL (reference range, 4.4 to

14.4 mg/dL). The amino acid and organic acid study exhibited normal glycine level in blood and CSF, and the glycine ratio of CSF to blood was 0.04 (reference rang<0.08). The lactic acid in blood and CSF were unremarkable. Her brain Magnetic Resonance Imaging (MRI) exhibited mild corpus callosum hypoplasia (Figure 1). Because of suspection of mitochondrial disorder, we performed muscle biopsy, in which, her bicep muscle biopsy exhibited both significant variation in the muscle sizes, and with fibrosis (Figure 2). A spectrophotometric analysis of the patient's respiratory chain complexes was performed and showed normal result. EEG manifested a marked suppression-burst pattern during sleep (Figure 3) at day 21. We used phenobarbital intravenously (6mg/kg/day) for her massive seizures, but it was fail to inhibit seizures. Her apnea with desaturation (oxygen saturation downed to 60 % -70% at the apnea associated with bradycardia) continued to occur frequently and with poor sucking ability. Intravenous levetiracetam was added and titrated from 30 mg/kg/day to 60 mg/kg/day, myoclonic seizure was significantly reduced, however, the apnea was still. SB EEG pattern was obviously reduced on the day 30 after 9 days of intravenous levetiracetam. Because her clinical manifestations improved significantly, we added oxycarbamazepine (30mg/day). On her day 35 of age, her apnea was improved after adding on bi-level positive airway pressure for her breathing. On 45 days of age, we replaced intravenous form levetiracetam to oral form (60 mg/kg/day), her sucking ability achieve a partial improvement. She was discharge depending on BiPAP. Her EEG changed to multiple focal spikes in both hemispheres with continuous background at 3 months old. At 4 months old, she could be smiling to parents but with BiPAP and tube-feeding. At the age of 1 year old, she had no seizure, and he was without apnea even without use BiPAP. At 4 years old, she could not walk, and he had severe cognitive disability without development of any language. Her chromosome analysis was normal and whole-exome sequencing for genetic study was unremarkable.

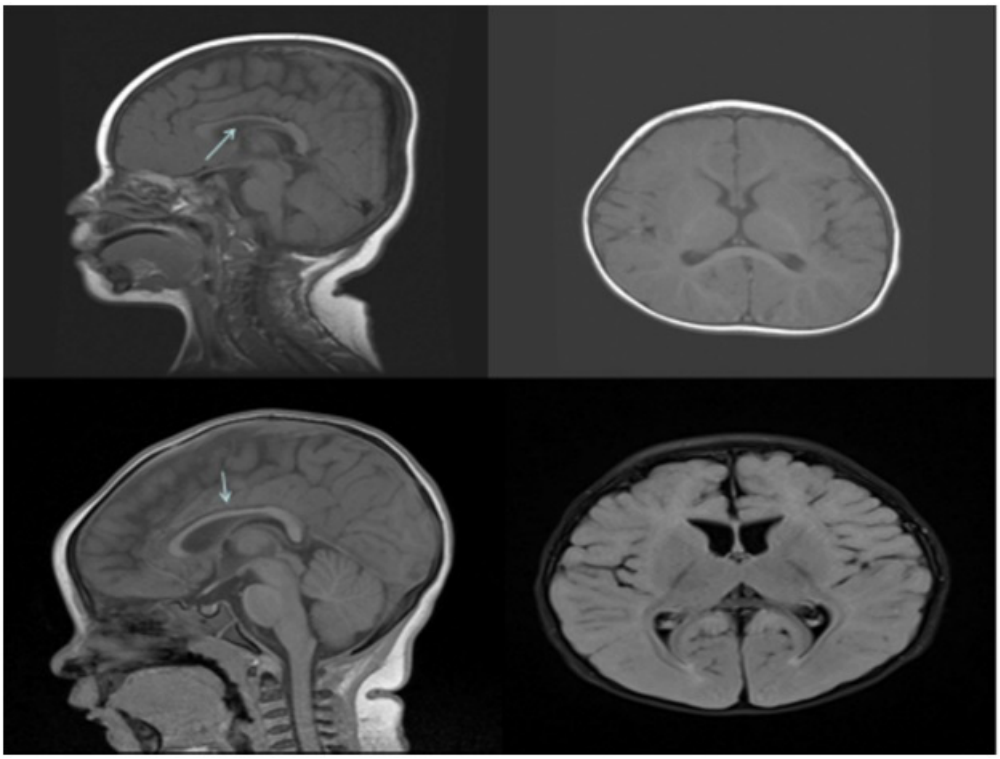

Figure 1: Her MRI show a thin corpus callosum (arrows).

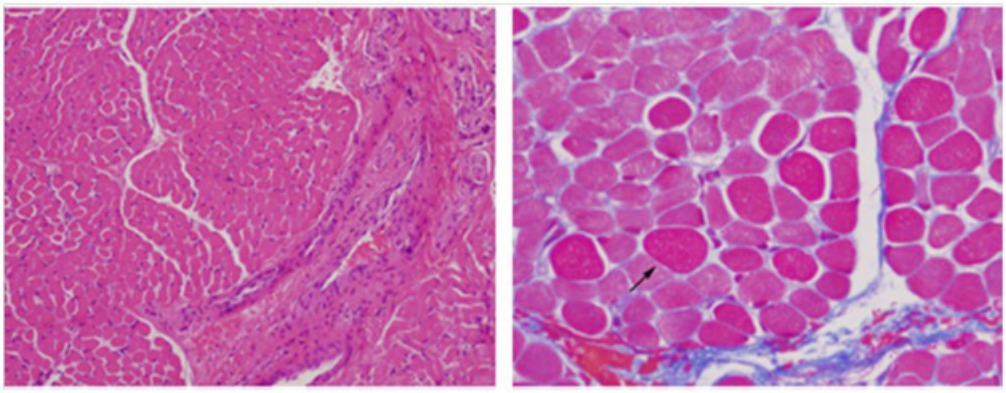

Figure 2: Her bicep muscle biopsy exhibited variation in muscle sizes (right, arrow) and mild fibrotic change (left).

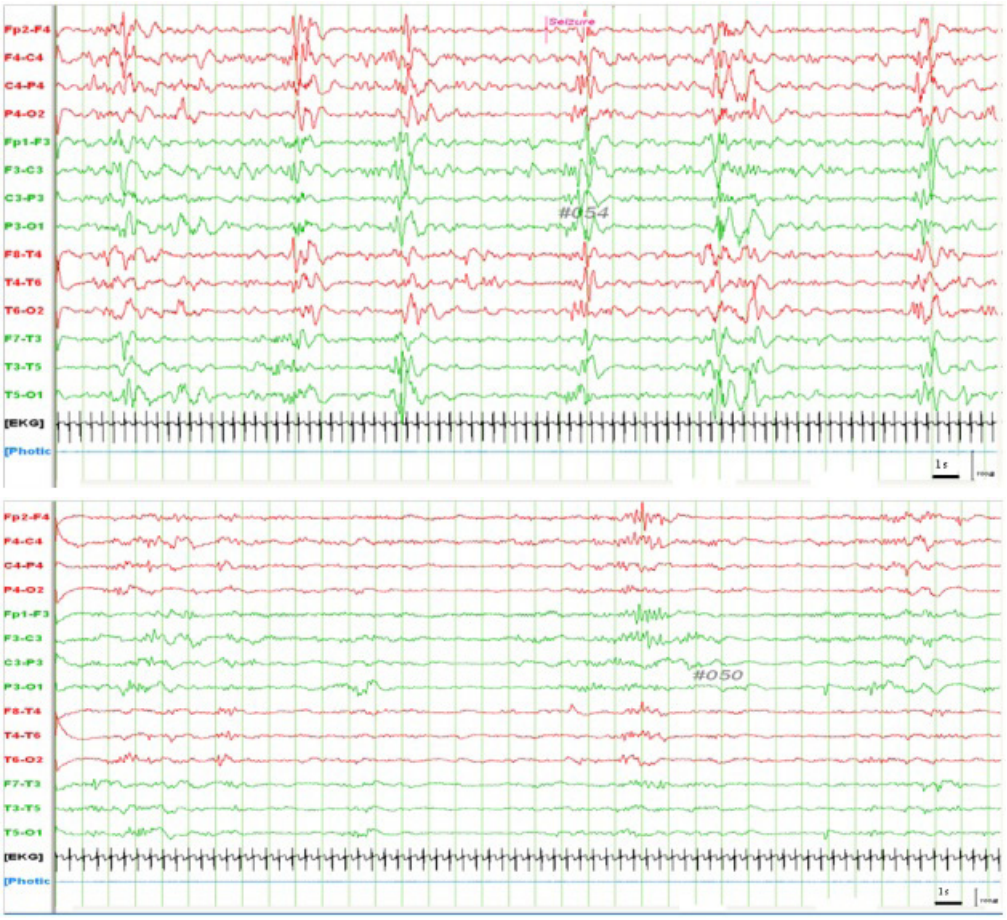

Figure 3: The EEG exhibited marked suppresion-burst activities (upper, arrows) at day 21, which achieved a marked response (lower) after intravenous levetiracetam up to 60mg/kg/day at day 30.

4. Discussion

Most EME cases had poor response to antiepileptic drugs. The patient received phenobarbital as first-line drug, however, failed to have response. We added intravenous levetiracetam and got improvement in her EEG and clinical seizures despite her apneas were still. Her apneas were probable not epileptic origin. The patient should be, to our best

knowledge, rare EME cases being treated with levetiracetam and achieved a favorable improvement in EEG and clinical seizures. We suggest levetiracetam could be a choice for treating the neonatal seizures with EEG SB activities.

5. Acknowledgments

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