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CASE REPORT

Dextromethorphan in the treatment of early myoclonic encephalopathy evolving into migrating partial seizures in infancy

Yin-Hsuan Chien [a], Ming-I Lin [b], Wen-Chin Weng [c], Jung-Chieh Du [d], Wang-Tso Lee [c,*]

[a] Department of Pediatrics, Taipei City Hospital Zhong Xing Branch, Taipei, Taiwan
[b] Department of Pediatrics, Shin Kong Wu Ho-Su Memorial Hospital, Taipei, Taiwan
[c] Department of Pediatrics, National Taiwan University Hospital, Taipei, Taiwan
[d] Department of Pediatrics, Taipei City Hospital Zhongxiao Branch, Taipei, Taiwan

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Epileptic encephalopathy with suppression-burst in electroencephalography (EEG) can evolve into a few types of epileptic syndromes. We present here an unusual case of early myoclonic encephalopathy that evolved into migrating partial seizures in infancy. A female neonate initially had erratic myoclonus movements, hiccups, and a suppression-burst pattern in EEG that was compatible with early myoclonic encephalopathy. The seizures were controlled with dextromethorphan (20 mg/kg), and a suppression-burst pattern in EEG was reverted to relatively normal background activity. However, at 72 days of age, alternating focal tonic seizures, compatible with migrating partial seizures in infancy, were demonstrated by the 24-hour EEG recording. The seizures responded poorly to dextromethorphan. To our knowledge, this is the first reported case of early myoclonic encephalopathy evolving into migrating partial seizure in infancy. Whether it represents another age-dependent epilepsy evolution needs more clinical observation.

Introduction

Early infantile epileptic encephalopathy (EIEE) with suppression-bursts (Ohtahara syndrome) and early myoclonic encephalopathy (EME) are both characterized by

* Corresponding author. Department of Pediatrics, National Taiwan University Hospital, Number 7, Chung-Shan South Road, Taipei, Taiwan.
E-mail address: leeped@hotmail.com (W.-T. Lee).

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suppression-burst pattern in electroencephalography (EEG) and intractable seizures in clinical presentations. EIEE is usually associated with brain structural anomaly, while EME is frequently associated with an inborn error of metabolism.[1] Nonketotic hyperglycinemia is one of the major causes of EME and the response to dextromethorphan (DM) treatment is good.[1,2] DM, a morphine derivative, is a commonly used antitussive drug, and it is also a noncompetitive N-methyl-D-aspartate (NMDA) receptor antagonist and voltage-dependent calcium and sodium channel blocker.[3] Although the use of DM is well established in patients with nonketotic hyperglycinemia, the use of DM in other types of epilepsy is not well known.[4–7]

Epileptic encephalopathy with suppression-burst pattern in EEG frequently evolves into other epileptic syndromes. Of these, EIEE can evolve into West syndrome, severe epilepsy with multiple independent spike foci, Lennox-Gastaut syndrome, and symptomatic partial epilepsy.[1] However, the evolution of EME is less documented. We report one patient with EME that was diagnosed at 40 days of age. The seizures were totally controlled by DM, and the suppression-burst pattern in EEG was changed to a nearly normal background activity. However, she developed alternating asymmetric tonic seizure 34 days later with characteristics of migrating partial seizure in infancy (MPSI) in both the clinical and EEG features. The response to DM was also poor at that time. The seizures were changed to infantile spasms 3 months later with the appearance of hypsarrhythmia in EEG.

Case report

A female baby was born smoothly by cesarean section to a G3P2SA1 mother aged 36 years; the baby had a gestational age of 36 weeks and a birth body weight of 1774 g.The head girth at birth was 30 cm (<third percentile). Myoclonus-like movements with hiccups were noted by the mother while the baby was awake and asleep ever since the patient was 20 days of age. The baby girl was then admitted for evaluation. On physical examination, there was no microcephaly, craniofacial dysmorphism, hepatosplenomegaly, and any other abnormality. The head circumference was 33.8 cm (third percentile). On neurologic examination, she had mild hypotonia, normoreflexia, and normal primitive reflexes, including positive grasping reflex, sucking, rooting reflexes, and Moro reflex. She also had normal cranial nerve examination with bilateral positive light reflex, gag reflex, and symmetric crying face, but she had poor eye contact. There was no optic atrophy or retinitis pigmentosa found during a fundus examination. During the admission period, metabolic evaluations

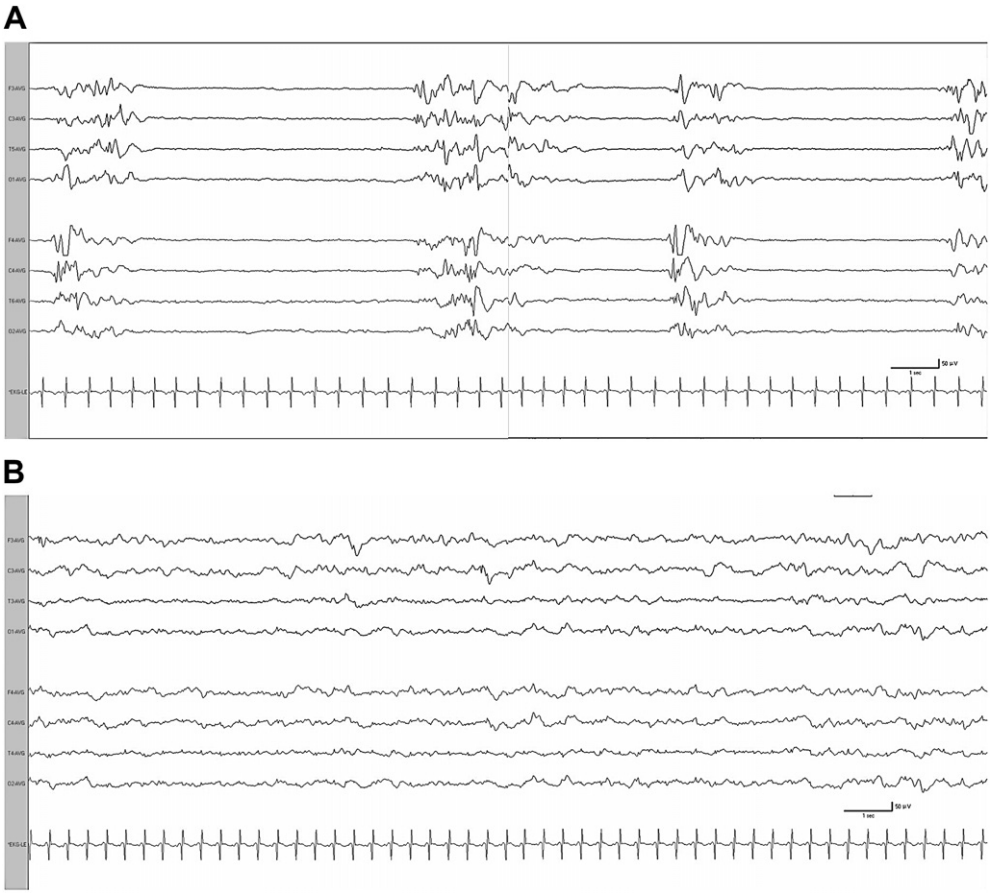

Figure 1 (A) Initial EEG evaluation showing the suppression-burst pattern at 40 days of age; and (B) the disappearance of the suppression-burst pattern with a nearly normalization of the background activity at 52 days of age after treatment with dextromethorphan. EEG = electroencephalography.