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Case Report

Hemiconvulsion-hemiplegia-epilepsy evolving to contralateral hemi-Lennox-Gastaut-like phenotype

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Abstract

Background: Hemiconvulsion-hemiplegia-epilepsy (HHE) involves infantile-onset acute hemiconvulsive febrile status epilepticus with subsequent unilateral cerebral atrophy and hemiparesis. Chronic epilepsy later develops, typically involving refractory focal seizures; however, the underlying pathophysiology of this epilepsy is not well understood.

Patient: We present a boy who had a typical acute presentation of HHE at 23 months, but an unusual evolution to chronic epilepsy in which the initially unaffected hemisphere was significantly abnormal. His initial acute presentation was right-sided hemiconvulsive febrile status epilepticus, with subsequent left cerebral hemiatrophy and hemiparesis affecting the right face, arm and leg. Focal seizures began at 5 years and were refractory to medical treatment. At 9 years, video EEG monitoring showed a striking pattern of interictal slow spike-wave and paroxysmal fast activity, maximal over the *right*, initially unaffected, hemisphere. He had primarily focal tonic seizures involving left-sided stiffening, also appearing to originate from the right hemisphere. Following left functional hemispherotomy he became seizure-free and parents reported improved cognitive function, attention and quality of life.

Discussion: This boy had classic features of Lennox-Gastaut syndrome, but expressed almost exclusively over the right hemisphere, which was initially unaffected in his acute presentation of HHE. His evolution to "hemi-Lennox-Gastaut-like phenotype" illustrates the importance of monitoring chronic epilepsy in patients with HHE; early surgical intervention might prevent pathologic recruitment of bilateral secondary networks leading to the refractory seizures and cognitive impairment associated with Lennox-Gastaut syndrome.

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Keywords: Hemiconvulsion-hemiplegia epilepsy; Lennox-Gastaut syndrome; Tonic seizures; Focal epilepsy

1. Introduction

Hemiconvulsion-hemiplegia-epilepsy (HHE) is a dramatic disorder in which infants present acutely with refractory hemiconvulsive febrile status epilepticus, often lasting days, followed by the evolution of a complete or partial hemiparesis [1]. Unilateral cytotoxic cerebral edema occurs acutely, later evolving to hemiatrophy [2]. Months to years later, refractory focal seizures develop [1,3]. Both autoimmune and genetic factors have been hypothesized to contribute but pathogenesis of HHE remains poorly understood. The

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epilepsy syndrome evolution from the initial presentation with HHE has not been well studied.

We present a boy with HHE who developed a contralateral hemi-Lennox-Gastaut-like phenotype. Earlier recognition of an epileptic encephalopathic process affecting the "good" hemisphere is critical as these patients may benefit from early epilepsy surgery to improve long term outcomes.

2. Patient

At age 23 months this previously normal boy presented with fever and seizures involving right eye deviation and predominantly right-sided clonic jerking, involving face, arms and legs, occasionally becoming bilateral. Brain MRI showed left hemispheric cytotoxic edema (Fig. 1A and B). Seizures recurred over four days despite midazolam, phenytoin and phenobarbitone. Four weeks later, he was discharged on carbamazepine, weaned two years later.

At 5 years, he developed focal impaired awareness seizures (FIAS) with 5–120 s of right head and eye deviation. Carbamazepine was re-started but failed to control seizures, nor did the ketogenic diet. Seizure frequency reduced on cannabidiol (Charlotte's Web 50 00 mg/100 ml, 0.65 ml tds).

Early development was normal, but global regression and permanent right hemiparesis, affecting arm, leg and face, occurred following acute status at 23 months. He also had a complete right homonymous hemianopia. He made slow developmental progress but had mild language regression at age 6–7 years with intercurrent illnesses. His neuropsychologic assessments at 7 and 9 years showed a full-scale IQ of 81, with normal language but marked right hemispheric dysfunction (impaired spatial cognition). He had co-morbid tic disorder, anxiety and anger outbursts. There was no family history of seizures, tics or developmental impairment.

Video-EEG monitoring at 9 years captured frequent focal motor seizures involving 1–4 s of right head version and left-sided tonic stiffening, occasionally progressing to bilateral tonic-clonic seizures (video in Supplemental Data). Ictal EEG showed diffuse attenuation with superimposed low amplitude fast activity, of right hemisphere emphasis (Fig. 1G). Interictal EEG showed a striking pattern of right hemispheric 1–3 Hz

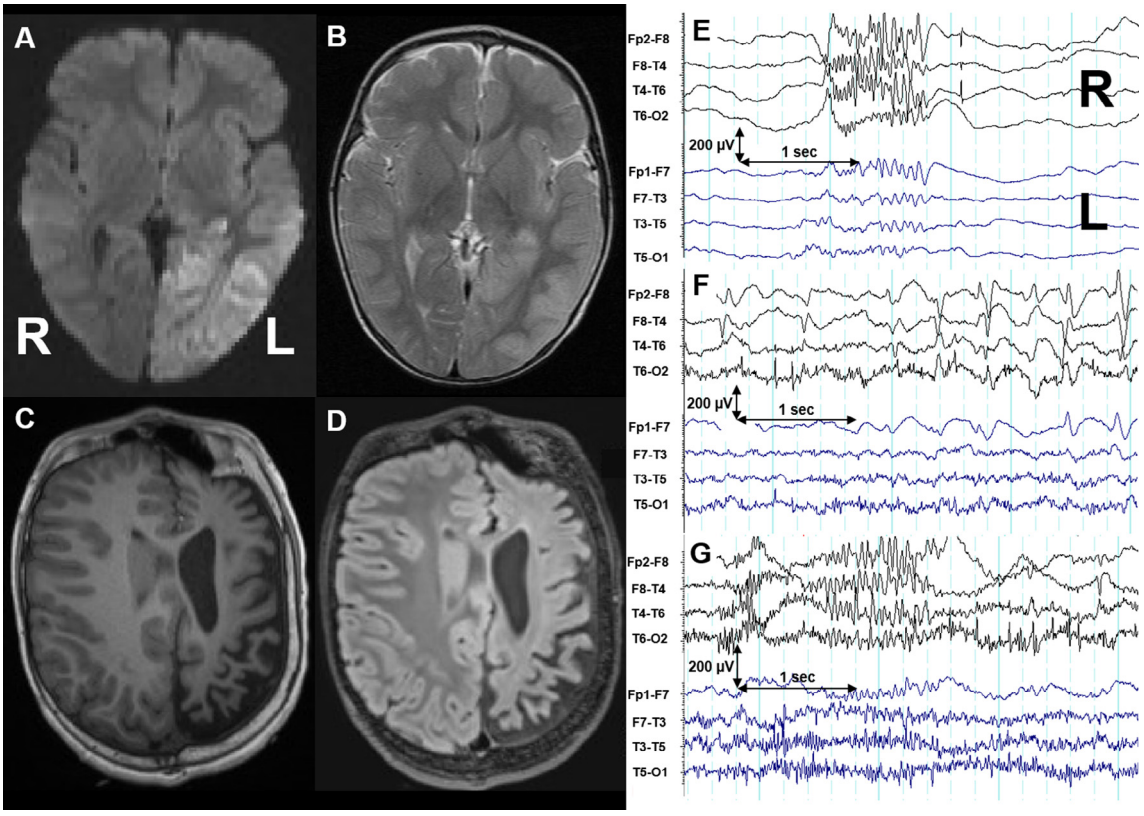

Fig. 1. Magnetic Resonance Imaging and Electrophysiology: Brain MRI three days after initial presentation (23 months of age) shows cytotoxic edema over the posterior left hemisphere demonstrated by restricted diffusion on axial diffusion-weighted imaging (A) and increased signal with swelling on axial T2-weighted sequences (B). At 9 years, diffuse left hemispheric atrophy is seen on axial T1-weighted and fluid attenuation inversion recovery sequences, with normal-appearing right hemisphere (C, D). Interictal EEG shows right hemispheric sharp waves and paroxysmal fast activity (E), and runs of 1–2 Hz frontally-predominant slow spike-wave discharges (F). Ictal EEG during a tonic seizure initially shows diffuse fast activity, maximal over the right hemisphere, evolving to slowing over the right hemisphere (G).

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