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Case report

Spontaneous resolution of intractable epileptic seizures following HHV-7 infection

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Abstract

Purpose: We report a three-year-old female with intractable epilepsy post West syndrome whose seizures disappeared following an acute viral infection, without changes in anti-epileptic therapy. *Methods:* The female infant was born at term to a healthy mother after an uneventful pregnancy and delivery. At the age of five months, she developed intractable brief tonic spasms which had a series of infantile spasms, and an electroencephalogram indicated hypsarrhythmia. She was diagnosed with West syndrome. The seizures were uncontrollable with conventional therapy, including ACTH, vigabatrin, sodium valproate, clonazepam, zonisamide, and ketogenic diet. Daily multiple generalized tonic seizures and brief tonic spasms were observed before an episode of viral infection. *Results:* At the age of three years, the intractable seizures disappeared after a febrile rash illness due to human herpesvirus 7 (HHV-7) infection, without changes in anti-epileptic drugs. *Conclusions:* The disappearance of intractable epileptic seizures following acute viral infections might be related to the inflammatory or immunologic processes associated with viral infections. This is the first documented case of spontaneous remission of intractable epileptic seizures following HHV-7 infection.

Keywords: Spontaneous resolution; Intractable epileptic seizures; HHV-7 infection

1. Introduction

Epileptic seizures generally are exacerbated by febrile illness or infection. However, it is well known that in rare instances, epileptic seizures, mostly associated with West syndrome disappear or decrease in severity after an acute viral infection, without changes to anti-epileptic medications. In contrast, patients with intractable epilepsy in infancy, particularly due to West syndrome, rarely show spontaneous remission of seizures. Disappearance of seizures, most often occurring following a viral infection, is an aspect of the natural history of these epilepsies that has not been sufficiently recognized. West first described in his syndrome a patient with such a remission after a brief febrile illness [1]. Certain patients with intractable epilepsy, such as those with West syndrome or Lennox-Gastaut syndrome, respond to therapy with immunomodulatory or anti-inflammatory agents such as high-dose immunoglobulin, ACTH or corticosteroids [2]. We report a three-year-old female with intractable epilepsy post West syndrome whose seizures disappeared following human herpesvirus 7 (HHV-7) infection. This is the first reported case of spontaneous remission of intractable epileptic seizures following HHV-7 infection. The goal of our study was to better characterize this phenomenon through a rare clinical case.

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2. Case report

The patient was born at 40 gestational weeks by normal vaginal delivery. The pregnancy was uneventful. Her birth weight was 3860 g and an initial Apgar score was nine. The family history was negative. Her psychomotor development was slightly delayed, and head control was not acquired at that time. At the age of five months, she had gradually become unwell and intractable brief tonic seizures developed. She did not have any deformity or skin pigmentation including hypopigmented area and her head circumference was within normal limits. Her neck and arms were flexed with legs extended. This condition was repetitive and occurred 10–20 times a day, with each series consisting of 20–30 spasms. Electroencephalogram (EEG) revealed hypsarrhythmia and she was diagnosed with cryptogenic West syndrome. Cranial magnetic resonance imaging (MRI) at the age of five months showed slight bilateral frontal lobe atrophy. However, MRI findings at the age of one year demonstrated severe atrophic changes in the white matter (Fig. 1). The seizures were refractory to conventional anti-epileptic therapies for West syndrome, such as sodium valproate, clonazepam, ACTH, high-dose vitamin B6, vigabatrin, high-dose gammaglobulin, and ketogenic diet. Multiple daily generalized tonic seizures (three to four times a day on awakening) and brief tonic spasms (20–30 times a day) were observed, and the patient was taking daily sodium valproate (400 mg/day) and clobazam (6 mg/day) before an infectious episode. At the age of three years, the intractable seizures disappeared one week after febrile illness with a rash probably due to HHV-7 infection without changes to anti-epileptic drugs. No significant changes were seen in serum concentrations of anti-epileptic drugs before and after this febrile rash illness. After remission, the EEG was also remarkably improved. The EEG before the acute viral infection showed generalized sharp waves and slow components (Fig. 2), whereas the EEG after this infection demonstrated no spikes (Fig. 3). Six months have passed since the remission occurred, the patient is still having the same medication and has no seizures. However, there was no psychomotor developmental catch-up after the seizures disappeared. Levels of HHV-6 IgG and IgM antibodies three weeks after the onset of febrile rash illness were <1:10, respectively. In contrast, the levels of HHV-7 IgG and IgM were >1:160 and >1:20, respectively. The assay was performed on a research based system and not for routine clinical use. The assay system has been established and validated by Focus Technologies for in vitro diagnostic use (Focus Technologies, Cypress, CA, USA). This is the first report in which the spontaneous remission of intractable epileptic seizures following HHV-7 infection was documented.

3. Discussion

Patients with intractable epilepsy in infancy, particularly West syndrome, rarely show spontaneous remission of seizures without changes to anti-epileptic medications [3]. This aspect of the natural history of these epilepsies has been insufficiently recognized. Hrachovy reported that spontaneous remission of West syndrome may occur as early as one month after seizure onset and the remission rate increased to 25% 12 months after onset without therapy, such as ACTH or sodium

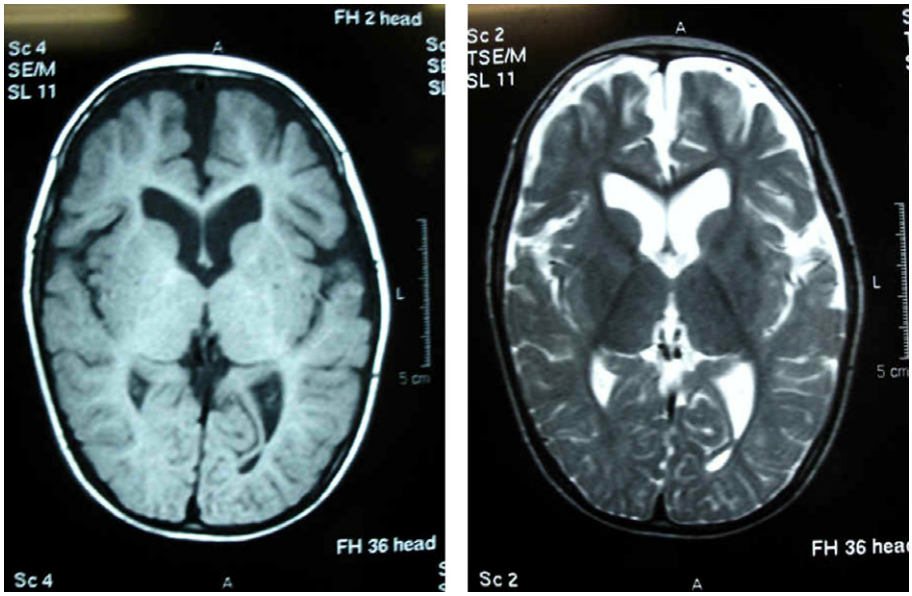

Fig. 1. Cranial MRI at the age of one year showed the severe atrophic changes in white matter and the enlargement of ventricles.