Brain & Development 39 (2017) 431–434

BRAIN & DEVELOPMENT
Official Journal of the Japanese Society of Child Neurology

www.elsevier.com/locate/braindev

Case Report

Total corpus callosotomy for epileptic spasms after acute encephalopathy with biphasic seizures and late reduced diffusion (AESD) in a case with tuberous sclerosis complex

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Received 27 October 2016; received in revised form 15 November 2016; accepted 17 November 2016

Abstract

Corpus callosotomy is a palliative therapy for refractory epilepsy, including West syndrome, without a resectable epileptic focus. The surgical outcome of corpus callosotomy is relatively favorable in cryptogenic (non-lesional) West syndrome. Tuberous sclerosis complex (TSC) is a disorder that frequently leads to the development of refractory seizures by multiple cortical tubers. The multiple cortical tubers cause multiple or wide epileptic networks in these cases. Most of West syndrome cases in TSC with multiple tubers need additional resective surgery after corpus callosotomy.

We describe a case of TSC in a boy aged 4 years and 8 months. He had multiple cortical tubers on his brain and developed epileptic spasms. The seizures were controlled with valproate. At the age of 1 year and 4 months, he presented with acute encephalopathy with biphasic seizures and late reduced diffusion (AESD), and had relapsed epileptic spasms one month after the onset of the encephalopathy. The seizures were refractory to multiple antiepileptic drugs. A total corpus callosotomy was performed at the age of 3 years and 8 months. The patient did not show any seizures after the surgery. During 12 months of the follow-up, the patient was free from any seizures. Even in cases of symptomatic WS with multiple lesions, total corpus callosotomy may be a good strategy if the patients have secondary diffuse brain insults.

Keywords: Corpus callosotomy; Epileptic spasms; West syndrome; Acute encephalopathy with biphasic seizures and late reduced diffusion; Tuberous sclerosis complex

1. Introduction

Tuberous sclerosis complex (TSC) is a neurocutaneous disorder caused by mutations in either *TSC1* or *TSC2*, which encode the protein products hamartin and tuberin, respectively. This disorder is characterized by tumors in multiple organs, including the brain [1].

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http://dx.doi.org/10.1016/j.braindev.2016.11.010
0387-7604/

Over 60% of patients develop epilepsy. The multiple cortical tubers cause multiple or wide epileptic networks, which result in medically refractory multifocal seizures or epileptic spasms [2]. These patients sometimes need surgical treatments.

Acute encephalopathy with biphasic seizures and late reduced diffusion (AESD) is a subtype of para-infectious acute encephalopathy frequently seen in Japan [3]. This type of encephalopathy is characterized by a biphasic clinical course. The initial phase consists of febrile convulsions and the second phase is characterized by reduced diffusion of subcortical white matter (bright tree sign) in diffusion weighted imaging (DWI) 2–8 days after the occurrence of the initial convulsion. In the chronic phase, magnetic resonance imaging (MRI) typically shows atrophic changes in the bilateral frontal lobes. The patients develop mild to severe cognitive and motor sequelae. The incidence of epilepsy after AESD is 23% [4].

Here, we describe a case of TSC in a boy, aged 4 years and 8 months, who developed epileptic spasms during infancy. At the age of 1 year and 4 months, the patient presented with AESD and developed epileptic spasms 4 weeks after the onset. He was free from the seizures following a total corpus callosotomy at the age of 3 years and 8 months.

2. Case report

A Japanese boy was delivered at 38 weeks gestation by normal vaginal delivery with a birth weight of 2618 g and a head circumference of 33 cm. Cardiac tumors were detected with ultrasonography. Hypomelanotic macules on the back, hip, and bilateral legs were seen from birth. A head MRI showed subependymal nodules on the walls of the bilateral lateral ventricles and multiple cortical tubers on the bilateral cerebral hemispheres (Fig. 1A). He was diagnosed with TSC based on these findings. He developed daily epileptic spasms starting at the age of 6 months. His electroencephalogram (EEG) showed frequent generalized spikes, polyspikes, and sharp waves, interictally. Valproate was effective for the treatment of the seizures. The patient was able to control his head at the age of 3 months, sit at 8 months, walk at 10 months, and speak some words at 1 year and 3 months.

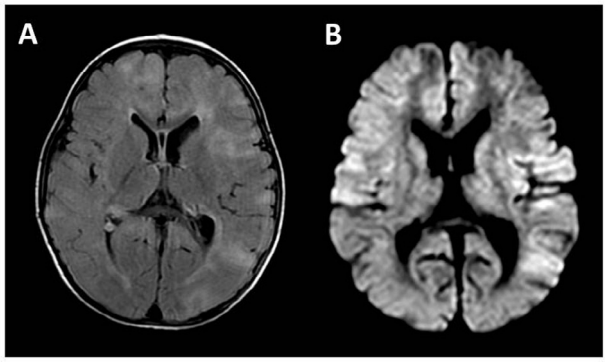

Fig. 1. The fluid attenuation inversion recovery imaging at 12-month old revealed subependymal nodules on the walls of the bilateral lateral ventricles and multiple cortical tubers on the bilateral cerebral cortices (A). The diffusion-weighted imaging on 11 days after the onset showed cortical and subcortical high signals on the bilateral frontal areas (B).

At the age of 1 year and 4 months, the patient presented with a generalized tonic–clonic convulsive status, which lasted for 1 h and 20 min, with a fever and exanthema subitum. Human herpes virus-6 isolation was obtained from his pharyngeal secretion on the day. He did not show attention and response to surrounding stimulations after the convulsive status. Three days after seizure onset, the patient had clustered clonic convulsions in the right upper extremity. He was treated with methylprednisolone pulse therapy (30 mg/kg/day for 3 days) and high-dose immunoglobulin (2 g/kg). A DWI performed 11 days after onset showed high cortical and subcortical signals in the bilateral frontal areas (Fig. 1B). The patient was diagnosed with AESD based on the clinical course and the MRI findings. After the onset of AESD, the patient was unable to sit, walk, or speak any words. He presented with paresis of the right arm. Epileptic spasms recurred 4 weeks after onset. Despite treatments with phenobarbital, zonisamide, and levetiracetam, the seizure frequency increased to 30–40 times per day.

At the age of 3, the patient was referred to Seirei-Hamamatsu General Hospital for the treatment of the refractory seizures. Video-EEG monitoring captured a total of 40 ictal events during 24 h record. The seizure types were epileptic spasms and brief tonic seizures. Ictal EEG showed biphasic or triphasic diffuse slow waves followed by electrodecremental activity. Interictal EEG showed independent or synchronous slow spike–wave bursts in the bilateral temporal or occipital areas, and sporadic focal sharp waves in the right frontal and bilateral temporal areas (Fig. 2A). From the clinical and electrophysiological findings, we diagnosed him as symptomatic West syndrome (WS). Additional treatments with lamotrigine and clonazepam were ineffective. At the age of 3 years and 8 months, we performed a total corpus callosotomy. After the surgery, the patient did not have any seizures. He had 2 weeks of swallowing disturbance as a transient disconnection symptom. EEGs performed 1 and 5 months after the surgery showed sporadic sharp waves in the right frontal to anterior temporal area (Fig. 2B). During 12 months of follow-up, the patient was free from seizures. After the termination of the seizures, his attention to the surroundings and emotional expression had improved.