ELSEVIER

Brain & Development xxx (2018) xxx–xxx

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

www.elsevier.com/locate/braindev

Case report

Effectiveness of total corpus callosotomy for diffuse bilateral polymicrogyria: Report of three pediatric cases

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Received 25 December 2017; received in revised form 8 March 2018; accepted 21 March 2018

Abstract

Purpose: Polymicrogyria, a malformation of the cerebral cortex, frequently causes epilepsy. Diffuse bilateral polymicrogyria (DBP) is related to poor epilepsy prognosis, but most patients with DBP are not good candidates for resective epilepsy surgery and effectiveness of corpus callosotomy (CC), a palliative surgery, for patients without resective epileptogenic cortices, has not been established in DBP. Because CC might be effective against DBP-related epilepsy, we conducted total CC in three pediatric DBP cases.

Methods: Case 1. A girl developed epilepsy at 3 months of age, with focal versive seizures and epileptic spasms. The electroencephalogram (EEG) showed a suppression-burst pattern. Total CC was performed at 6 months of age.

Case 2. A female infant developed epilepsy on the day of birth, exhibiting epileptic spasms, generalized tonic-clonic seizures, and eye-deviating seizures. She had a history of clusters of tonic seizures. Total CC was performed at 1 year and 2 months of age. After CC, the epileptic focus of the tonic seizures was identified; a secondary resective surgery was conducted.

Case 3. A girl developed multiple types of seizures at 3 years of age. Frequent atypical absence status was refractory to antiepileptic drugs. Total CC was conducted at 8 years of age.

Results: Case 1: Frequencies of both seizure types decreased. The background EEG changed to continuous high-voltage slow waves.

Case 2: Clusters of tonic seizures were well-controlled.

Case 3: Atypical absence seizures completely disappeared.

Conclusion: CC could be effective for patients with DBP, whose habitual seizures include epileptic spasms and absence seizures.

Keywords: Polymicrogyria; Epilepsy; Corpus callosotomy; Drug-resistance; Bilateral cortical malformation; Palliative surgery

1. Introduction

Polymicrogyria is a malformation of the cerebral cortex caused by abnormal postmigrational development

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https://doi.org/10.1016/j.braindev.2018.03.007
0387-7604/

that is associated with a variety of clinical symptoms e.g., epilepsy, intellectual disability, and motor and speech dysfunction [1,2]. Around 60–80% of the patients with polymicrogyria have epilepsy [1]. The appearance of polymicrogyria on magnetic resonance imaging (MRI) is heterogeneous, ranging from focal to generalized distributions [1,2]. Involvement of bilateral hemispheres tends to be a poor prognostic factor for development and motor function [3], as well as poor control of epilepsy [4,5]. Resective surgery for patients with focal or unilateral polymicrogyria was recently reported to be effective [6,7]. Most patients with a bilateral distribution are not candidates for resective procedures, because there are typically multiple and/or extensive epileptic foci bilaterally [2]. The establishment of better seizure management for these patients is an urgent issue.

Corpus callosotomy (CC) is a palliative surgery for intractable epilepsy, especially suitable for patients who cannot be treated with resective surgery [8]. Although CC is reported to be effective against epilepsy in patients with bilateral hemispheric malformations of the developing cortex [9], currently there are no reports on the effectiveness of CC in patients with diffuse bilateral polymicrogyria (DBP). Herein, we report clinical courses of three pediatric patients with DBP and intractable epilepsy who received total CC.

2. Case 1

Case 1 was a 7-year-old girl. The patient showed no developmental progress after birth. She developed epilepsy at 3 months of age and was referred to our hospital. Brain MRI showed DBP that was especially prominent in both frontal lobes (Fig. 1A, B). Her electroencephalogram (EEG) showed a suppression-burst (SB) pattern (Fig. 2A). Seizure semiology indicated focal versive seizures, with frequent evolution to secondary generalized tonic-clonic seizures (GTCS), which occurred in both hemispheres. Epileptic spasms and non-epileptic myoclonus were also confirmed. She was diagnosed with early myoclonic encephalopathy (EME). Her focal seizures occurred over 30 times/day. High-dose phenobarbital reduced her seizure frequency to around 5 times/day, but the EEG abnormalities did not improve at all. Total CC was conducted at the age of 6 months; CC reduced the frequency of focal seizures and epileptic spasms, and ceased secondary GTCS. The SB pattern in EEG changed to continuous slow waves, with multifocal spikes (Fig. 2B). Over 7 years of follow-up, she remained bed-ridden and showed no apparent intellectual development, but came to express comfort/discomfort against environmental stimuli. Seizure semiology grossly remained the same as before surgery, but the frequency decreased to daily or weekly.

3. Case 2

Case 2 was a girl aged 3 years and 6 months. The patient developed hourly seizures from the day of birth. Her EEG showed an SB pattern. Brain MRI revealed DBP with hypertrophic change in the right hemisphere (Fig. 1C, D). Antiepileptic therapies, comprising administration of phenobarbital, clobazam, valproate, zonisamide, levetiracetam, topiramate, clonazepam, phenytoin, and potassium bromide, a ketogenic diet, and intramuscular injection of adrenocorticotropic hormone, were partially effective against her seizures. The patient often developed clusters of tonic seizures that necessitated barbiturate-induced coma.

She was referred to our hospital at the age of 1 year. She was bed-ridden without acquiring control over the head and needed tube feeding. Her habitual seizures comprised epileptic spasms, GTCS, and eye-deviating seizures. EEG revealed hypsarrhythmia (Fig. 2C). With the aim of preventing clusters of tonic seizures, the patient underwent total CC at the age of 1 year and 2 months. Although GTCS disappeared and the frequency of epileptic spasms was ameliorated, right-side dominant tonic seizures with eye blinking appeared every 10 min just after surgery. Ictal scalp/intracranial EEG (Fig. 2D) and ictal single photon emission computed tomography (SPECT) (Fig. 1E) revealed a seizure onset zone in the left temporo-occipital region. Two weeks after CC, left posterior quadrantectomy was conducted. After that, the frequency and severity of tonic seizures were ameliorated. Over the follow up period of 3 years, the patient remained bed-ridden and showed no developmental progress. Even after quadrantectomy, daily tonic seizures with eye-deviation and severe abnormalities on EEG persisted; however, clusters of tonic seizures were well controlled. Vagus nerve stimulation therapy was administered to the patient at the age of 1 year and 9 months. Frequency of residual seizures did not change during the follow-up period.

4. Case 3

Case 3 was an 8-year-old girl. At the age of 10 months, left hemiparesis was noticed. Although she had severe developmental delay, she could walk independently at the age of 3 years and use single words at the age of 4 years. She developed epilepsy at the age of 3 years. She had unresponsive seizures with cyanosis, GTCS, and atonic seizures. Brain MRI revealed DBP and slight asymmetrical brain atrophy of the right hemisphere (Fig. 1F). Most of these seizures were diminished on treatments with antiepileptic drugs. From the age of 6 years, frequent unresponsive seizures emerged followed by falling down on the left side, which were refractory and often needed barbiturate coma therapy.

Please cite this article in press as: Baba S et al. Effectiveness of total corpus callosotomy for diffuse bilateral polymicrogyria: Report of three pediatric cases. Brain Dev (2018), https://doi.org/10.1016/j.braindev.2018.03.007