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Case Report

Effective Epilepsy Surgery for Post-Traumatic West Syndrome Following Abusive Head Trauma

Hiroki Tsuchiya[a]*, Takashi Shibata[a], Tatsuya Sasaki[b], Takushi Inoue[c], Isao Date[b], Tomoyuki Akiyama[a], and Katsuhiro Kobayashi[a]

Department of [a]Pediatrics (Child Neurology), [b]Neurological Surgery, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences and Okayama University Hospital, Okayama 700-8558, Japan, [c]Department of Pediatrics, National Hospital Organization Okayama Medical Center, Okayama 701-1192, Japan

West syndrome, an infantile developmental and epileptic encephalopathy with a deleterious impact on long-term development, requires early treatment to minimize developmental abnormality; in such cases, epilepsy surgery should be considered a powerful therapeutic option. We describe a 10-month-old female admitted with West syndrome associated with a hemispheric lesion following abusive head trauma. Her seizures were suppressed by hemispherotomy at 12 months of age, leading to developmental improvement. Surgical treatment of West syndrome following traumatic brain injury has not been reported previously but is worth considering as a treatment option, depending on patient age and brain plasticity.

Key words: abusive head trauma, developmental and epileptic encephalopathy, epilepsy surgery, epileptic spasms, hemispherotomy

West syndrome is characterized by epileptic spasms (ESs), a serious type of pediatric seizure associated with very abnormal scalp electroencephalography (EEG) patterns, or hypsarrhythmia. West syndrome is a developmental encephalopathy (DEE) that is known to affect subsequent long-term development and requires early treatment with a short lag time to minimize developmental abnormalities including progression to Lennox-Gastaut syndrome [1]. Post-traumatic epilepsy (PTE) is an important chronic complication following traumatic brain injury (TBI), in which the development of West syndrome has also been previously reported. The specific risk for West syndrome is high when a severe TBI occurs during infancy [2]. Although first-line treatment of West syndrome is medical, accumulating reports have shown the effectiveness of epilepsy surgeries, such as hemispherectomy or lobectomy, for West syndrome caused by a resectable brain lesion [3] and corpus callosotomy for West syndrome without resectable lesions [4]. Successful surgical treatment can dramatically improve developmental outcomes if the non-resected areas of the brain are functioning normally [5].

Here, we report the case of an infant with PTE in the form of West syndrome attributable to abusive head trauma (AHT) who underwent hemispherotomy [6-8], a surgical technique that functionally realizes the effects of hemispherectomy. This technique disconnects an entire unilateral hemisphere from other parts of the brain by resecting the commissural and unilateral projection fibers and results in immediate seizure control and improvement in long-term development, albeit sacrificing all remaining hemispheric function. To the

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*Corresponding author. Phone: +81-86-235-7372; Fax: +81-86-235-7377
E-mail: pxf46gle@s.okayama-u.ac.jp (H. Tsuchiya)

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best of our knowledge, there have been no previous reports of epilepsy surgery, including hemispherotomy, for West syndrome caused by TBI. There have been a limited number of reports of West syndrome or ESs associated with AHT [2, 9, 10]. In previous reports, all patients with West syndrome caused by AHT have been treated medically; the present report is the first case of epilepsy surgery for West syndrome resulting from AHT. Sharing our experience may provide a new, powerful option for treating West syndrome in the context of TBI, including AHT.

Case Report

The patient was a girl who at the time of writing was 3 years and 4 months of age, and had been healthy during early infancy after an uneventful 38-week-long pregnancy and a normal vaginal delivery with a birth weight of 2,558 g. At 5 months, however, she presented with right hemi-convulsive status epilepticus after a brief period when her father cared for her alone. Cranial computed tomography (CT) revealed a left-sided acute subdural hematoma (Fig. 1A). Emergency surgery was performed at the Okayama Medical Center, Japan to remove the hematoma and reduce intracranial pressure by decompression craniotomy. Retinal hemorrhage was also noted. The case was therefore highly suggestive of an AHT, based on the judgment of the hospital's Abuse Prevention Committee and the police. Right hemiparesis and right visual field defect or hemispatial neglect, suggested by a lack of attention to objects on the right side, remained after the operation. At 9 months of age, prophylactic medical treatment with sodium valproate was started because of the emergence of epileptic discharges on electroencephalography (EEG).

ESs in clusters began to occur in association with a change in the EEG pattern into near-hypsarrhythmia despite prophylactic treatment. The patient was admitted to Okayama University Hospital at 10 months of age (5 months after the trauma) with a diagnosis of West syndrome. The clinical manifestations of her ESs were slight twitching movements of the extremities and minimal nodding when held in an upright posture. Her ictal clinical symptoms might have been more evident had the patient been able to sit still at that time. Cranial magnetic resonance imaging (MRI) revealed marked left hemisphere-dominant cerebral atrophy (Fig. 1B). Interictal EEG showed near-hypsarrhythmia with a relative paucity of activity in the left hemisphere, suggestive of hypofunction in this part of the brain (Fig. 2A, B). Ictal EEG of the ESs episodes showed preceding 15-25 Hz low-amplitude fast activities in the left occipital region and subsequent slow waves with superimposed fast activities predominant over the right hemisphere (Fig. 3). Associated electromyography (EMG) activity that was dominant in the right deltoid muscle was suggestive of right-side dominance of the muscle contraction of seizures. Iomazenil single photon emission computed tomography (SPECT) revealed reduced benzodiazepine receptor-binding potential in the left parietal and occipital lobes. Studies of flash-visual evoked potentials and upper limb short-latency somatosensory evoked potentials indicated hypo-responsiveness and hypofunction of the left hemisphere. The preoperative developmental quotient (DQ) was assessed as 62 using

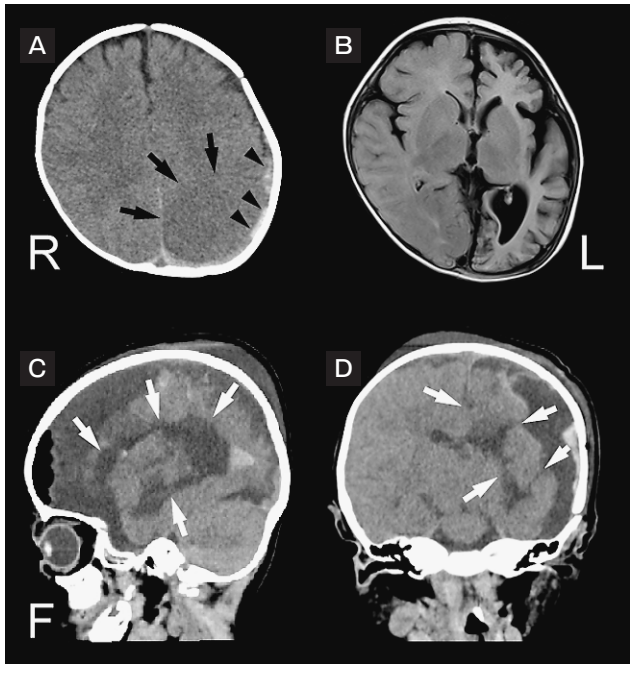

Fig. 1 Neuroimaging. (**A**) A computed tomography (CT) scan taken immediately after the status epilepticus shows an acute subdural hematoma in the left parietal region (black arrowheads), and a broad low-density area with blurred corticomedullary borders in the left parietal and occipital lobes (black arrows). (**B**) A magnetic resonance imaging (MRI) fluid-attenuated inversion recovery (FLAIR) image taken at 10 months of age shows marked cerebral atrophy of the left hemisphere and enlargement of the left lateral ventricle. (**C**, **D**) A CT scan taken after the hemispherotomy shows the fiber disconnection (white arrows) (**C**, sagittal view; **D**, coronal view) and atrophy of the left hemisphere.

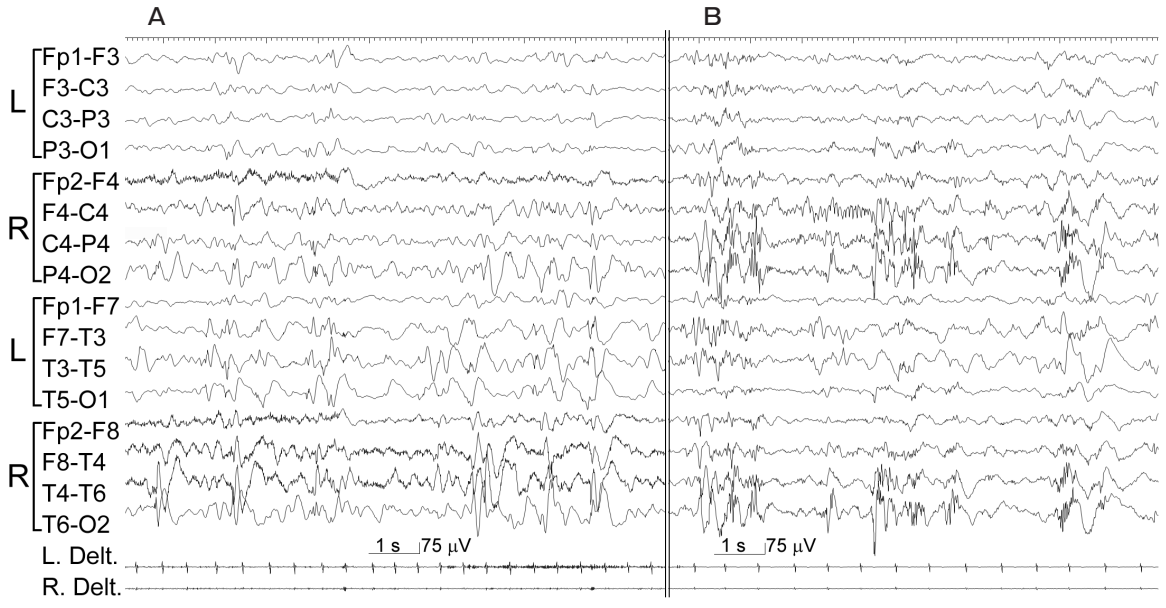

Fig. 2 Interictal scalp electroencephalography (EEG) at 10 months of age. (**A**) Waking record showing near-hypsarrhythmia with multifocal spike discharges mixed with high-amplitude irregular slow waves dominating the bilateral temporal-parietal-occipital regions. Activities in the left central region were relatively poor compared with those in the contralateral counterpart region. (**B**) Natural sleep record showed fragmentation abnormalities with polyspike waves in the right hemisphere. Sleep spindles were observed only in the right hemisphere.

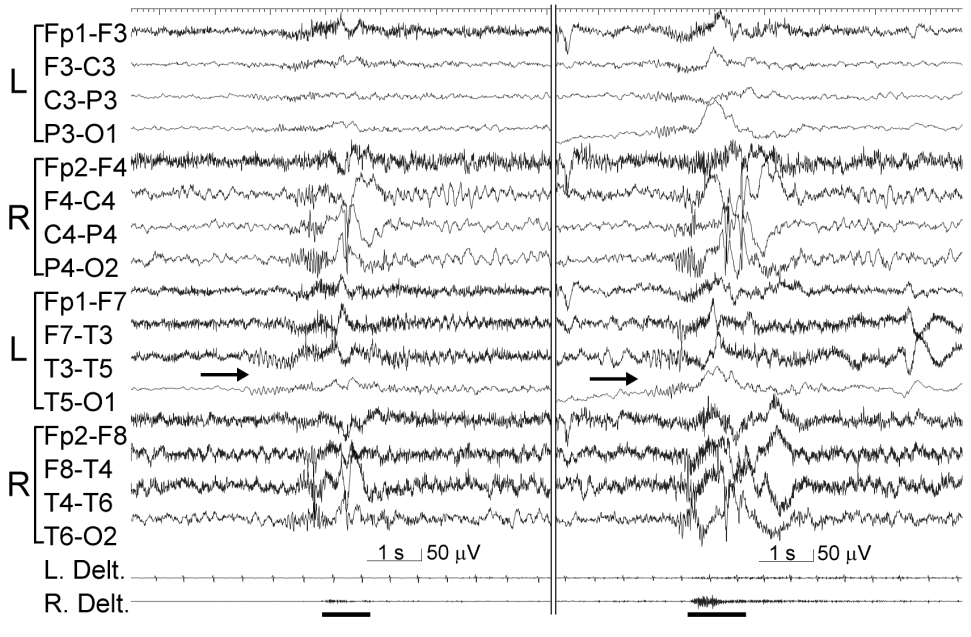

Fig. 3 Ictal scalp electroencephalography of epileptic spasms. Ictal EEG revealed the preceding 15–25 Hz low-amplitude fast activities in the left occipital region (arrows) and subsequent high-amplitude slow waves with superimposed fast activities that were dominant in the right hemisphere and associated with the clinical spasms indicated by right-side-dominant muscle activities in the deltoid muscles (two spasms indicated by underlines).

the Enjoji Scale. Adrenocorticotropic hormone (ACTH) therapy was avoided due to the risk of induction of further hemorrhage, and vigabatrin (VGB) was administered with no effect. Other antiepileptic drugs were considered to be even less effective.

These findings led us to consider the therapeutic option of epilepsy surgery. The seizure focus was considered to involve the major part of the left hemisphere. We aimed to stop the progression of developmental deterioration induced by the epileptic encephalopathy of West syndrome and to prevent the development of epileptic abnormalities in the normal right hemisphere. In infants and toddlers, plasticity in the right upper and lower extremity functions and language function can be expected; hence, the benefit of the suppression of West syndrome was considered far greater than the disadvantages of post-surgical right hemiplegia. A left hemispherotomy including corpus callosotomy (horizontal approach) was performed at 12 months of age (2 months after the ES onset) to completely suppress seizures, following consent from the patient's mother, who was not involved in the original abuse.

There has been no seizure recurrence for more than 2 years after the surgery. Immediately after surgery, the patient displayed complete paralysis of the right upper and lower extremities. However, she gradually regained the ability to run, speak brief sentences, and use her right hand to some extent, and her DQ improved to 70. CT scan showed dissection of nerve fibers due to hemispherotomy and associated atrophy of the left hemisphere (Fig. 1C, D). EEG showed epileptic discharges from the dissected left frontal region and some right frontopolar discharges that were probably volume-conducted potentials from the left hemisphere (Fig. 4).

Ethics statement. Written informed consent was obtained from the patient's mother for the publication of this case report and the accompanying images.

Discussion

Post-trauma epilepsy (PTE), and post-traumatic West syndrome in particular, is a serious consequence of childhood TBI. The overall incidence of PTE following pediatric TBI is reported to be 10%, and occurrences of early seizures, severe TBI, and intracranial hemorrhage increase the risk of PTE [11]. Six out of 47

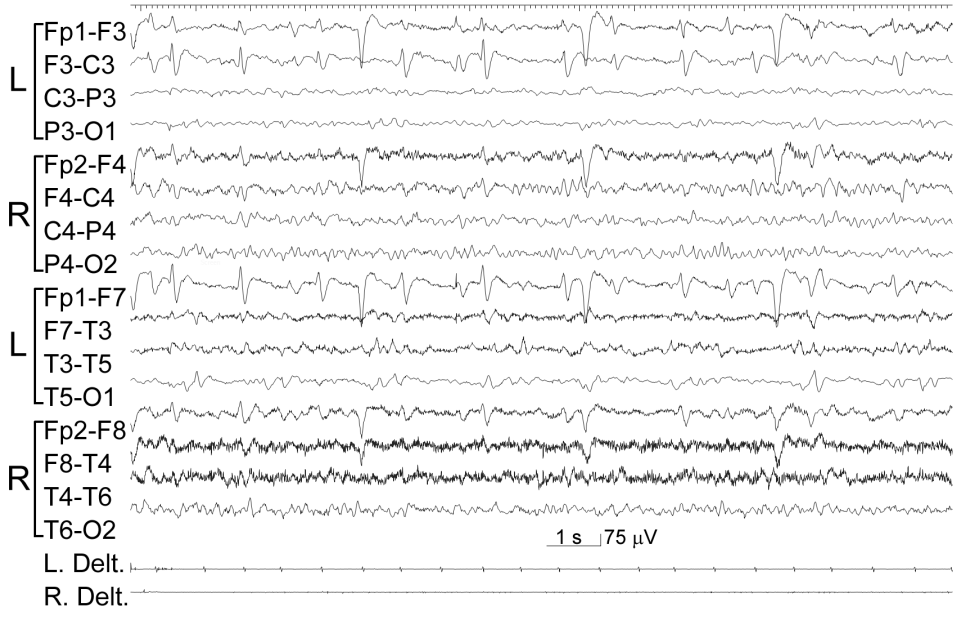

Fig. 4 Interictal scalp electroencephalography recorded at exactly 3 years of age. Waking record showing alpha rhythm dominance in the right hemisphere with asymmetry in the background activity. Epileptic discharges (spikes and sharp waves) are frequent and dominate over the left frontal and frontopolar regions. Discharges over the right frontopolar region are probably due to volume conduction from the left hemisphere.

(12.8%) pediatric patients diagnosed with PTE had ESs that occurred between 2 months and 2 years after trauma; the risk of ESs is likely to be much higher if a severe TBI occurs during infancy [2]. The present case had these predictive features of ES occurrence, and close EEG follow-up enabled the detection of the emerging West syndrome, which allowed early treatment.

It is known that a shorter lag time to treatment commencement and early ES suppression leads to better subsequent developmental outcomes [1]. Although medical treatment, including ACTH and VGB, is the first-line therapy for post-traumatic West syndrome and other types of West syndrome [2], the responses in some patients are insufficient [10], as in the present report. When medical treatment options are ineffective for ESs, other treatment modalities, including epilepsy surgery, should be sought. Success in resective epilepsy surgery is expected when epileptogenic lesions are present in MRI [3]. Suppression of ESs by surgical resection of the epileptogenic focus is known to contribute to recovery from developmental delays [12], and younger infants, particularly patients with ES, are known to have a higher increase or recovery of DQ after surgery [13]. Even in reports limited to patients undergoing hemispherotomy, early surgery for infants with epileptic encephalopathy has shown the potential for better developmental prognoses [5].

Conversely, poor functional outcome has been associated with a longer duration of epilepsy and older age at the time of surgery [14]. In the present case, after medical treatment had failed, hemispherotomy to disconnect the pathologic left hemisphere and minimize developmental delay was undertaken immediately, since prolongation of ineffective medical treatment would have resulted in further developmental deterioration. In addition, the entire left hemisphere was severely damaged and epileptogenic with impaired function, and at the patient's young age, postoperative functional recovery could be expected. All these factors were considered carefully when deciding on the surgical technique. This successful case suggests that epilepsy surgery may be performed early after the onset of ESs following an AHT, as for ESs with other etiologies such as cerebral dysplasia, and that such surgeries are meaningful for recovery from developmental delays.

EEG findings are essential to determine the seizure focus, and curative surgery is best performed in patients with lesional abnormalities on MRI with EEG concordance [3]. In the present case, ictal EEG showed spasm-associated slow waves with superimposed fast activities dominant over the relatively healthy right hemisphere. Despite this EEG finding, the injured left hemisphere was considered to be the epileptogenic hemisphere because leading low-amplitude fast activities were seen there first. Clinical movements of spasms were right-side dominant; the left hemispherotomy disclosed the background EEG activity in the right hemisphere. This paradoxical false lateralization of EEG findings has been observed when the pathological cortex is atrophic and does not generate much EEG activity [15], as in the present case. It teaches us that the correct epileptogenic cortical regions should be deduced from EEG findings and consideration of the condition of the brain structure.

To the best of our knowledge, this is the first report of a case of hemispherotomy for West syndrome or ESs developing after AHT. Because epilepsy surgery can significantly impact subsequent developmental prognosis, when DEE is resistant to medical treatment and the epileptogenic focus is confined within a unilateral hemisphere, epilepsy surgery, including hemispherotomy, should be considered without hesitation to achieve seizure control and improve the chances of healthy development.

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