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Case report

Early total corpus callosotomy in a patient with cryptogenic West syndrome

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1. Introduction

West syndrome is a generalized epileptic syndrome of infancy, that is characterized by clinical spasms and hypsarrhythmia and usually has an onset age before two years old. Children affected by West syndrome have a high risk of severe psychomotor developmental delay. Infants with West syndrome without identification of underlying neurological disorder after adequate investigations are classified as "cryptogenic" and usually have better prognoses than children with "symptomatic" West syndrome.[1] Early seizure cessation, particularly among cryptogenic cases, may cure the disease and allow a good neurodevelopmental outcome.[2]

Corpus callosotomy was first introduced as a palliative treatment for epilepsy in 1940 and is performed in patients whose seizures are not amenable to surgical focal resection. West syndrome is often medically intractable and has features of "generalized" epilepsy, but corpus callosotomy has not been discussed widely as a treatment strategy. Given the catastrophic nature of West syndrome, an early indication for total corpus callosotomy may be appropriate. Here, we describe a case that was diagnosed as cryptogenic West syndrome, in which the epileptogenic focus was defined after total corpus callosotomy.

2. Case

An 8 month-old boy was referred to our hospital following the appearance of repetitive spastic movements. He was born naturally at 38 weeks of gestation with no complications and reached all developmental milestones until 7 months-old. About one week before referral he started having clusters of 5–7 sudden, brief spastic movements, during which he dropped his head and raised both arms. He became unable to sit and lost his smile. His interictal waking and sleep EEG showed typical hypsarrhythmia consisting of random high-voltage slow waves and spikes. His ictal EEG showed voltage attenuation superimposed with bursts of low voltage, fast activity that was often contaminated with movements artifacts (Fig. 1A). MRI showed no abnormalities and other tests were also within normal limits.

The patient was diagnosed with cryptogenic West syndrome. After VitB6 administration (40 mg/kg/day), adrenocorticotrophic hormone (ACTH) (0.0125 mg/kg/day = 0.5 IU/kg/day) was started. Because of several series of epileptic spasms, ACTH dosage was increased to 0.025 mg/kg/day and valproic acid (VPA) (40 mg/kg/day; serum concentration 80 μg/dl) was added. Seizures ceased completely after five days and an EEG showed age-appropriate background activity without any epileptiform discharges. At one year old, the patient started having a series of similar epileptic spasms and he again showed developmental regression. His EEG showed typical hypsarrhythmia, but MRI and interictal and ictal ECD-single photon emission computed tomography (SPECT)findings were unremarkable (Fig. 2A-1). Increasing the VPA dose (serum concentration 120 μg/dl), nitrazepam, zonisamide, and lamotrigine were all tried with no response.

At the age of 14 months, total callosotomy was performed at the National Hospital Organization Nagasaki Medical Center. After corpus callosotomy all spasms lateralized to the left side of the body. During seizures, he suddenly looked up and raised his left arm. Ictal EEG showed polyspikes and waves followed by brief voltage attenuation predominantly over the right hemisphere (Fig. 1B). Interictally, polyspike-wave discharges were expressed in the right fronto-central region. An ictal ECD-SPECT study demonstrated hyperperfusion in the right frontal lobe (Fig. 2A-2). Interictal FDG-positron emission tomography (PET) showed hypometabolism in the right hemisphere (Fig. 2B). Based on these localized findings, right frontal resection was performed two months after corpus callosotomy. The resected lesion was

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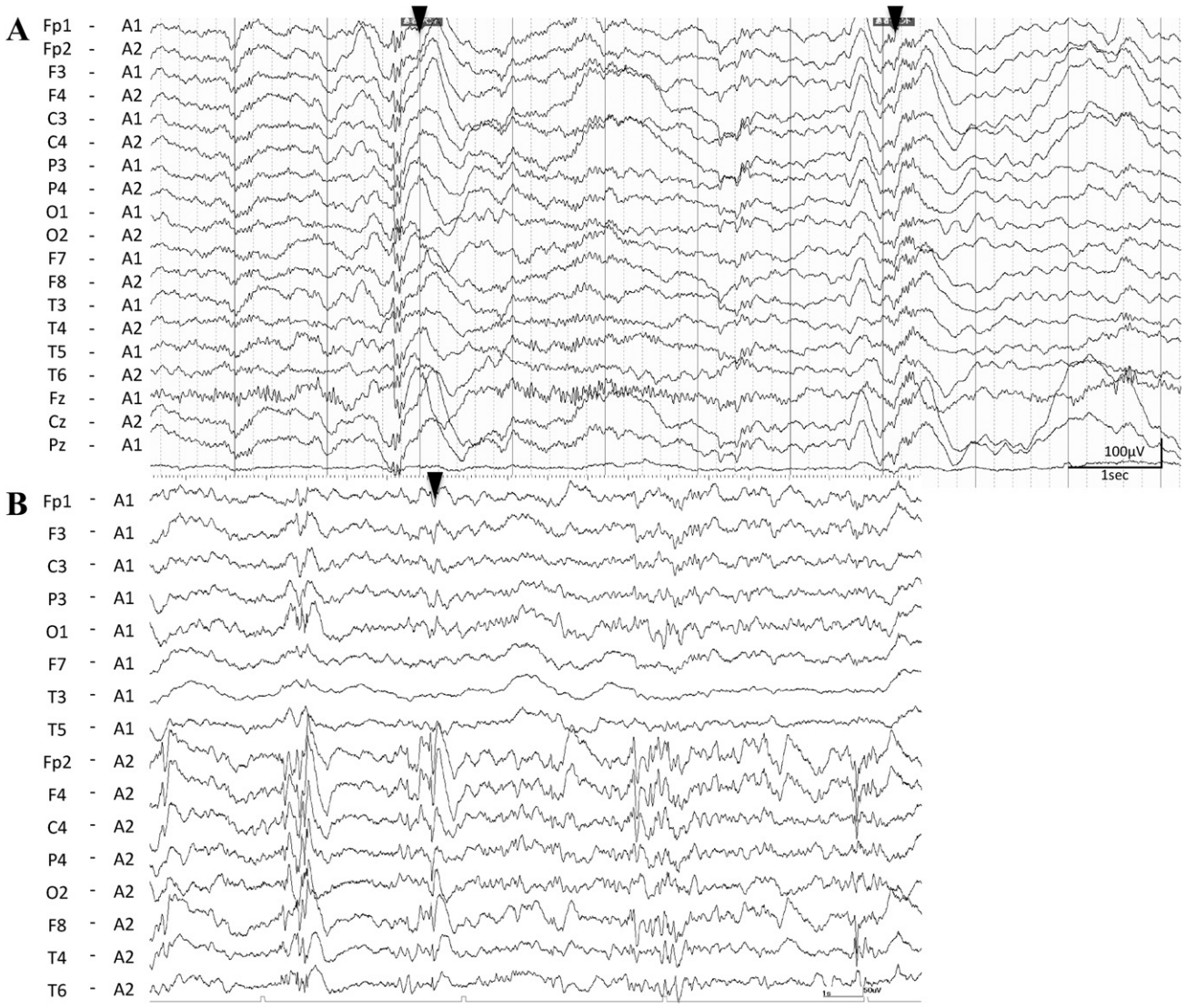

Fig. 1. (A) Example of an ictal EEG recorded during preoperative EEG-video monitoring. The ictal pattern is characterized by diffuse voltage suppression superimposed with bursts of low voltage beta range activities. The EEG was recorded by 21 electrodes using the 10–20 system and is represented in monopolar reference. (B) Example of an ictal EEG recorded during routine EEG-video monitoring after corpus callosotomy. The ictal pattern is characterized by high voltage polyspike wave discharges followed by brief voltage attenuation over the right hemisphere. The EEG was recorded by 19 electrodes using the 10–20 system and is represented in monopolar reference. Arrowheads indicate times at which an ictal event was noted by observers.

determined by intraoperative electrocorticography. Surgical specimens showed preserved cortical lamination and prominent gliosis in the subcortical areas. Subsequently, he has been seizure-free for three years.

Before corpus callosotomy at 16 months old his developmental age was 10 months, he had no language, showed autistic behavior, and his developmental quotient (DQ) was 62.5 on the Kinder Infant Development Scale. Six months after surgery, his behavior clearly changed and he was able to sit still for 30 min watching TV, speak several words, and follow simple verbal commands, although the DQ was only 66.7. Post surgically, he showed "catch-up" developmental progress, and his DQ improved to 72.7, 80.4, and 81.8 yearly over three years. At his current age of 4 years 7 month old, 3 years after surgery, his language is beyond the telegraphic stage with some Japanese grammatical errors.

3. Discussion

Total corpus callosotomy revealed previously unperceivable seizure foci in our patient diagnosed with cryptogenic West syndrome, and led to successful surgical lesional resection. Surgical treatment for symptomatic West syndrome is effective for controlling seizures and cessation of spasms, and early surgery for catastrophic epilepsy such as West syndrome has been shown to have positive effects on developmental outcomes.[2] Structural abnormalities also strongly influence the prognosis in patients with West syndrome.[1,3] Therefore, the importance of evaluation to determine resectable epileptogenic lesions in medically intractable West syndrome cannot be stressed enough.

Corpus callosotomy is most effective in patients with atonic seizures with drop attacks, and has not been established as a standard surgical strategy for cryptogenic West syndrome. Clear improvements with corpus callosotomy also occur for other types of seizures, such as tonic seizures and secondarily generalized seizures.[4] Pinard et al.[5] first reported seizure reductions and developmental improvements in patients with post-West syndrome after corpus callosotomy. Baba and colleagues performed early total corpus callosotomy in patients with West syndrome without a clear resectable region, of whom 75% showed significant developmental improvement during three-year follow-up.[6]