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Original article

Patients with West syndrome whose ictal SPECT showed focal cortical hyperperfusion

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Abstract

To clarify the mechanism of clustered spasms in West syndrome (WS), we examined ictal SPECT and EEG, as well as those during the interictal period, in three patients with symptomatic WS who had apparent focal cerebral lesions. For ictal SPECT and EEG, we monitored the patients with EEG, and as soon as we noticed the occurrence of clustered spasms clinically and electroencephalographically, [^{99m}Tc]ECD was injected intravenously and flushed thoroughly with saline. In these three patients, regional cerebral blood flow (rCBF) increased during ictus and decreased during the interictal period in the area that coincided with the focal cerebral lesion recognized by CT/MRI. The ictal hyperperfusion of bilateral basal ganglia was also detected in two of the three patients. The ictal EEG showed a diffuse slow wave complex corresponding to a clinical spasm. The sharp waves that preceded the diffuse slow wave complex and the spasm appeared in the same area in which rCBF increased during ictus. None of the patients showed partial seizure before or after clustered spasms clinically or electroencephalographically during the ictal SPECT study. Secondary generalization from a cerebral focus may explain the mechanism of spasms in these patients with WS: focal cortical discharge may primarily generate clustered spasms and trigger the brainstem and basal ganglia structures to produce spasms.

Keywords: West syndrome; Spasms; Ictal single photon emission computed tomography; Ictal EEG; Hypsarrhythmia

1. Introduction

West syndrome (WS) is defined as an epileptic syndrome consisting of spasms in clusters and hypsarrhythmia, often associated with the arrest of psychomotor development. Although WS is classified as a generalized epileptic syndrome because of its generalized EEG abnormalities and bilateral clinical manifestations [1], recent neuroimaging studies [2–5] have indicated that pathologic conditions in the cortex play an important role in the pathogenesis of spasms in patients with WS.

We report, here, three patients with symptomatic WS who showed apparent focal cerebral lesions. We used ictal single photon emission computed tomography (SPECT) and EEG, as well as those during the interictal period, and examined the pathophysiology of clustered spasms.

2. Methods

We received informed consent from the families of all of the patients during their first admission. For ictal SPECT and EEG, pediatric neurologists monitored the patients with EEG, and as soon as they noticed the occurrence of clustered spasms clinically and electroen-

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cephalographically, [^{99m}Tc]ethyl cysteinate dimer (ECD) was injected intravenously and flushed thoroughly with saline. The dose of [^{99m}Tc]ECD was 180 MBq. SPECT images were acquired about 30 min after injection using PRISM 3000 (Picker, Ohio, USA). Over a period of 20 min, 120-degree datum points were obtained in a stepwise manner in a 64×64 matrix. Using an Odyssey (Picker) computer system, tomograms two pixels thick (11.67 mm) were reconstructed after a high-frequency cutoff with a Butterworth filter was applied. Axial and coronal slices were constructed parallel and perpendicular to the orbitomeatal plane, while sagittal slices were constructed parallel to the longitudinal fissure. Infants were anesthetized with trichloryl hydrochloride during scanning. Interictal SPECT was performed while the infants were awake, and none showed clinical evidence of seizures during the interictal SPECT study. The mean interval between ictal and interictal SPECT was 10 days.

SPECT images were independently visually analyzed by a pediatric neurologist and a radiologist, and localized cerebral perfusion abnormalities were judged to be present only when both evaluations coincided. Pediatric neurologists monitored the patients with EEG for more than 2 h before the occurrence of clustered spasms and the injection of [^{99m}Tc]ECD, and also monitored the patients with EEG for more than 30 min after the injection. None of the patients showed partial seizure before or after clustered spasms clinically or electroencephalographically during the ictal SPECT study. The ictal EEGs that were recorded at the ictal SPECT study are shown in Figs. 4, 6 and 8. The interictal open-eyed EEGs and natural sleep EEGs that were recorded a few days before the ictal SPECT study are shown in Figs. 3, 5 and 7.

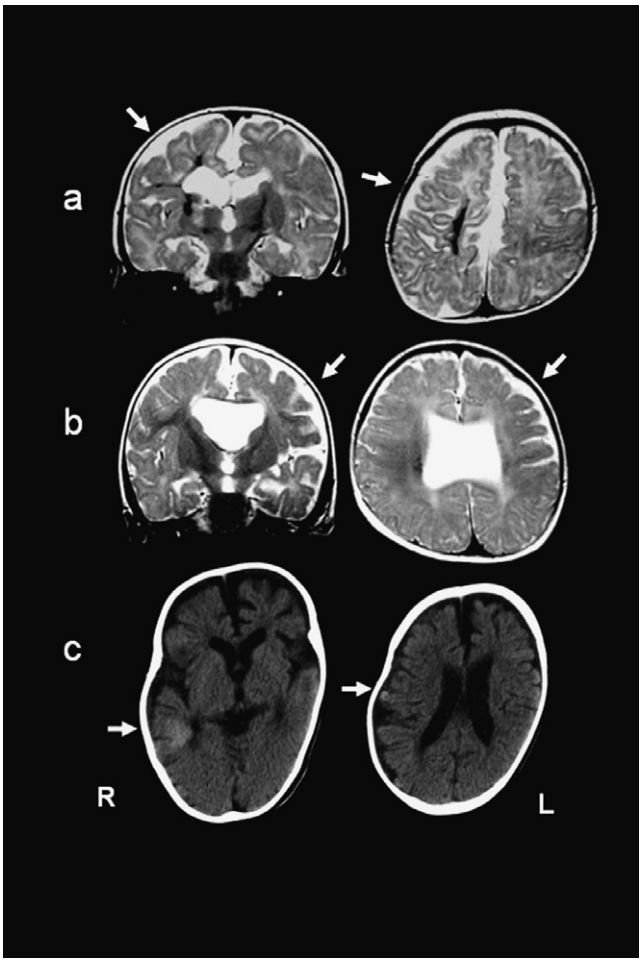

Fig. 1. (a) MRI T_2-weighted images (TR, 2000 ms; TE, 80 ms) in patient 1; (b) MRI T_2-weighted images (TR, 2000 ms; TE, 80 ms) in patient 2; (c) CT in patient 3. The arrows show focal cerebral lesions. R, right; L, left.

3. Case reports

3.1. Patient 1

His mother suffered from idiopathic thrombocytopenic purpura (ITP) during pregnancy. There was no abnormal event at delivery. He was born without asphyxia after 38 weeks of gestation. His birth weight was 2230 g. No abnormal findings were noted during the neonatal period. Head control was seen at age 4 months. Spasms appeared in clusters during periods of wakefulness at age 7 months and he was admitted to Tokushima University Hospital. Mild left hemiparesis was observed and his deep tendon reflexes were exaggerated on the left side, but the spasms showed no asymmetry. Magnetic resonance imaging (MRI) showed atrophic change in the right fronto-parietal region and moderate enlargement of the lateral ventricle on the right side, and T_2-weighted images showed that this was surrounded by a low-intensity area (Fig. 1a). These findings were considered to be due to bleeding during pregnancy and were caused by his mother's ITP. The interictal SPECT showed hypoperfusion in the right fronto-central area and right basal ganglia. On the other hand, ictal SPECT showed hyperperfusion in the right central region. The regional cerebral blood flow (rCBF) of the bilateral basal ganglia was slightly increased on ictal SPECT, but no remarkable change was recognized in the brainstem or cerebellum (Fig. 2a and b). The interictal open-eyed and sleep EEG showed asymmetric hypsarrhythmia in which paroxysmal discharges were prominent on the right side (Fig. 3). The ictal EEG showed a diffuse slow wave complex corresponding to a clinical spasm. Sharp waves of the right temporo-central region preceded the slow wave complex and the spasm (Fig. 4). Administration of clonazepam (CZP) resulted in the complete cessation of spasms. Subsequent EEGs showed diminishing paroxysmal discharges. He is now aged 6 years. Sharp waves now rarely appear in the right temporo-central region. His developmental quotient (DQ) is 55.