A Case of Absent Brainstem Responses with Electroencephalographic Activity

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In Japan, brain death is diagnosed upon observation of a total absence of all cortical and brainstem responses[1]. An isoelectric electroencephalography (EEG) reading in association with absent brainstem responses is mandatory for the diagnosis of brain death[1]. EEG activity after total cesstion of brainstem function is not a common event[2–5]. This contradictory condition, to our knowledge, has not yet been reported in a patient without distinct infratentorial lesion[2–5]. We described herein a patient with a demyelinating disorder who showed distinct EEG activity in spite of absent brainstem responses during the evolution of his illness.

Case Report

A 7-year-old tracheostomized boy was admitted to our ICU after an episode of respiratory arrest along with consciousness disturbance. He was not deaf. When the attack occurred at the general ward of our hospital, he became cyanotic but cardiac arrest did not occur. He had an approximately 6-year history of congenital deficiency of the E_1 component of pyruvate dehydrogenase (PDH) complex[6]. The first episode of respiratory arrest had occurred when he was 1 year old, when an enzyme study had revealed a severe deficiency of the E_1 component. Since that time, he had suffered many episodes of respiratory arrest and consciousness disturbance.

On arrival, his pupils were reactive to light and were not dilated. However, he was in a deep coma and no spontaneous respiratory movements were observed. Blood pressure was 130/60 mmHg, and pulse rate was 120 beats per minute. The arterial blood gas values were pH of 7.19, Pa_{CO_2} of 42 mmHg, Pa_{O_2} of 401 mmHg, and BE of -12 mEq·l^{-1} at an FI_{O_2} of 1.0 under controlled mechanical ventilation. Blood lactate level was elevated to 8.5 mMol·l^{-1}.

On day 2, diabetes insipidus developed. Neurological examination revealed severe brainstem involvement with absent corneal, oculocephalic, oculovestibular, oropharyngeal, gag, ciliospinal, light, and cough reflexes. No spontaneous respiratory movements were observed. In addition, auditory brainstem responses (ABR) showed no identifiable wave forms.

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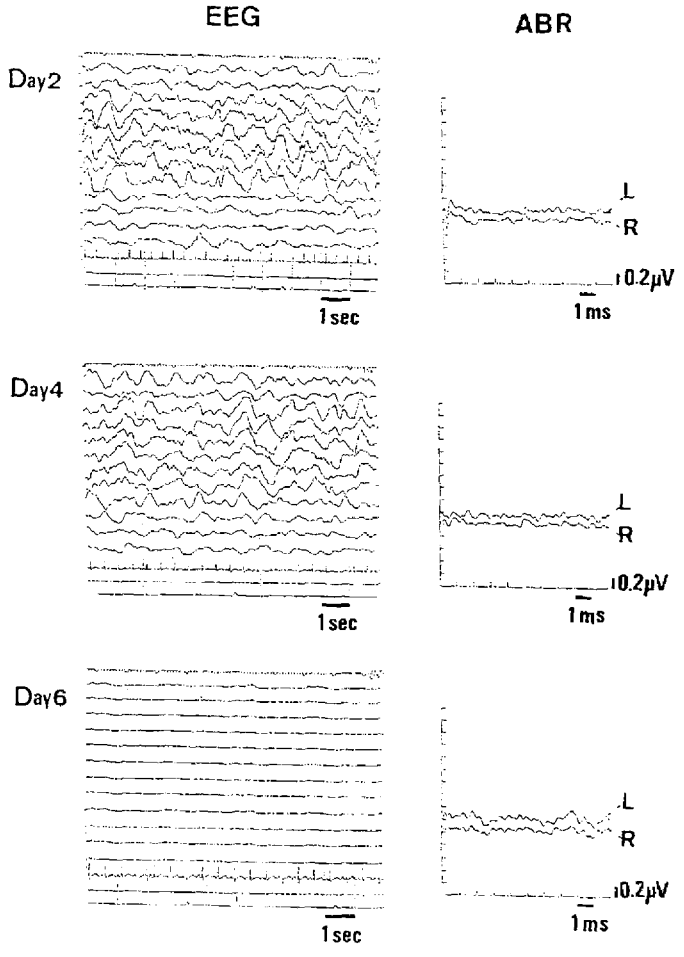

Fig. 1. Representative EEG and ABR recordings on day 2, 4 and 6.

However, EEG revealed θ and δ waves (fig. 1). Cranial computerized tomography revealed a hypodense lesion in the basal ganglia and the deep white matter. However, there was no distinct infratentorial lesion which could be considered to be related to the total absence of brainstem responses. No brain edema ws observed.

On days 3 and 4, the neurological findings did not change; ABR was absent, while EEG still revealed θ and δ waves. On day 5, computerized tomography demonstrated severe brain edema. Although neurological brainstem responses were not observed, spinal cord reflex became obvious. The upper and lower extremities reflectively extended when the body was touched. ABR was absent, but EEG revieled some suggestive biological activities.

On days 6, 9 and 12, ABR was absent and EEG became almost isoelectric. The patient was in a deep coma and no neurological brainstem responses were observed. The apnea test was not performed. Thus we could not diagnose brain death. However, we judged his cerebral and brainstem disturbances irreversible. After obtaining his parents' consent, we transferred him from ICU to the general ward. He survived for about 6 months thereafter in the state of deep coma under mechanical ventilatory support along with

inotropic and vasopressin therapies.

Discussion

Deficiency of the E_1 component of the PDH complex is a congenital demyelinating disease caused by a congenital defect of thiamine pyrophosphate-dependent pyruvate decarboxylase[6]. This disease is characterized by lactic acidemia and neurologic abnormalities consisting of psychomotor retardation, ataxia, lethargy, convulsions, hypotonia and hypertonia, and irritability[6]. The extent and location of nervous system damage may vary from cerebral atrophy to the development of cystic lesions in the cerebal cortex, basal ganglia and/or brainstem[6]. Although most of the patients die in childhood due to overwhelming lactic acidosis, some may survive to adolescence/young adulthood due to recent improvements in therapeutic techniques.

Our patient with PDH complex deficiency manifested distinct EEG activity in spite of the total absence of brainstem function during the evolution of his illness. Brain death is suspected to have occurred when clinical neurological examinations indicated that cerebral and brainstem functions have irreversibly ceased[1]. To confirm the findings of these examinations, the electrophysiologic methods are usually used; isoelectric EEG and the absence of ABR are frequently used criteria.

EEG recordings reflect activity in the postsynaptic dendritic and somatic membranes of neurons in the cerebral cortex[7]. The absence of brain waves indicates the loss of the cortical neuronal function. In deeply comatose patients, the presence or absence of cerebral cortical function cannot be adequately evaluated by the conventional neurologic examinations. In this situation, EEG can be a definite aid[7].

Distinct EEG activity after total cessation of brainstem function is not a common event[2-5]. This contradictory condition has not been reported other than in a few cases of isolated brainstem lesion, such as infratentorial hemorrhage, hematoma, infarction, and encephalitis[2-5]. Thus, in Germany, an EEG is performed in the diagnosis of brain death only in the specific circumstance of a primary infratentorial lesion[8].

In the present case, computerized tomography did not reveal any distinct infratentorial lesion that could be considered to be related to the total absence of brainstem responses. This finding suggests that brain death should be diagnosed cautiously in a patient with demyelinating disease.

According to the definition of "brainstem death" in England[9], a patient with absent brainstem function will die within a few days regardless of whether EEG activity is present[9]. However, this assertion is not consistent with the findings in the present case.

The controversy about when brain death can be diagnosed and which tests are necessary for its diagnosis is on-going. A total absence of all cortical and brainstem responses is mandatory for the diagnosis of brain death in Japan[1]. However, the interpretation of EEG in patients with suspected brain death is extremely complicated due to technical problems. Artifacts produced by both mechanical and electrical interference are often seen and have been well documented[7,10]. Considerable intra-rater variability and inter-rater disagreement in the interpretation of EEG recordings have also been reported[10]. In addition, the absence of ABR has not always confirmed the irreversible cessation of brainstem function[11,12], as there are some case reports of the reappearance of ABR in comatose patients[11,12].

Considering the limitations of both EEG[7,10] and ABR[11,12], and strictly ap-

plying the concept of the death of the entire brain, we think that the total absence of intracerebral blood flow[13,14] should be confirmed before brain death is diagnosed if there is doubt as to whether the damage is reversible or not.

In summary, we have described a patient with a demyelinating disorder who showed distinct EEG activity in spite of absent brainstem function during the evolution of his illness. Computerized tomography did not reveal any infratentorial lesion. These findings presented in this case report suggest that brain death should be diagnosed cautiously in a patient with demyelinating disease.

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