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Case Report

Ictal single photon emission computed tomography of myoclonic absence seizures

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Abstract

Background: Epilepsy with myoclonic absences (EMAs) is a rare epileptic disorder characterized by a predominant type of seizures, myoclonic absences (MAs). The pathophysiology of MAs in patients with EMAs remains unknown. Here, we report the first characterization of the ictal phase of MAs by single photon emission computed tomography (SPECT).

Methods: We evaluated 1 male (Patient 1) and 1 female (Patient 2) patient with EMAs, aged 8 and 4 years at first SPECT investigation, respectively. We performed ictal and interictal $^{99\,m}$Tc-ethyl cysteinate dimer (ECD) SPECT. We then generated images of subtraction ictal SPECT co-registered to MRI (SISCOM) from the interictal and ictal data to evaluate topographic changes in cerebral blood flow (CBF) during MAs as compared to the interictal state.

Results: In Patient 1, the CBF increased in the perirolandic areas, thalamus, caudate nucleus, and precuneus, and decreased in the middle frontal gyrus and bilateral orbitofrontal regions. In Patient 2, CBF increased in the thalamus, putamen, and globus pallidus. In contrast to the CBF in Patient 1, CBF was decreased in the precuneus.

Conclusions: Using SPECT, we showed that, in addition to the thalamus and basal ganglia, the perirolandic cortical motor area is involved in MAs. We hypothesize that in MAs the blood perfusion in the perirolandic cortical motor area might have changed under the influence of the cortico-thalamic network oscillation features. The CBF properties observed by means of our SPECT procedure may represent key features of the pathophysiological mechanisms underlying MAs.

Keywords: Basal ganglia; Cortico-thalamic network; Ictal SPECT; Motor cortex; Myoclonic absence; Thalamus

1. Introduction

Epilepsy with myoclonic absences (EMAs) is a rare epileptic disorder characterized by a predominant type of seizure called myoclonic absences (MAs), which were first described by Tassinari et al. [1] and then by Tassinari and Bureau [2,3]. Clinically, the myoclonus in MAs is frequently associated with more or less evident tonic contractions, which differentiates MAs from typical absence seizures (TAs) in EMAs. The pathophysiology of MAs is still unknown [1,3], and functional imaging studies of MAs are limited. Recently, studies using single photon emission computed tomography (SPECT) [4,5] and functional magnetic resonance imaging (fMRI)/electroencephalograms (EEG) [6–8] have revealed various changes in cerebral blood flow (CBF)

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and metabolism in TAs, or spike and wave discharges on EEG.

To identify additional therapeutic interventions for intractable epilepsy and elucidate the pathophysiological mechanisms of EMAs, we here examined the ictal and interictal dynamics of the CBF in 2 such patients. We first obtained ^{99} ^{m}Tc-ethyl cysteinate dimer (ECD) SPECT data and then generated images displaying brain anatomical CBF patterns by subtraction of ictal SPECT co-registered to MRI (SISCOM).

2. Methods

Patients 1 and 2 had MAs with the following clinical and polygraphic criteria: (1) intense rhythmic jerks of the upper limbs, (2) rhythmic and generalized spikes and waves (GSW) at approximately 3 Hz on the EEG, and (3) tonic muscle contraction on surface electromyography (EMG; deltoid, trapezius, or sternocleidomastoideus). Both patients also underwent ictal SPECT imaging. Medical reports and video polygraphic recordings were reviewed retrospectively.

The patients and their parents gave informed consent to undergo all examinations and procedures.

2.1. Imaging protocol

The ECD tracer was injected intravenously during the interictal and ictal phases as confirmed by concurrent polygraphic EEG/EMG recordings.

2.2. Subtraction analysis

The distribution of regions with increased or decreased CBF on SPECT in the ictal state was compared with those in the interictal state by SISCOM.

2.3. Case presentation

2.3.1. Patient 1

Patient 1 was a young boy with intellectual disability noted at the age of 6 years, and his IQ was 79 at 8 years of age. MAs had begun at the age of 5 years. The seizure lasted for 20–40 s. Seizure frequency was 15–20 times a day at the age of 7 years. Subsequently, he developed generalized tonic–clonic seizures (GTC).

Seizures persisted despite treatments with valproic acid (VPA), carbamazepine, or zonisamide, or a combination of these drugs. He was admitted to our epilepsy center at the age of 8 years. Ictal video EEG and polygraphic (EEG and EMG) recordings were examined. Most of the seizures were MAs associated with the predominant elevation of the right arm, a slight version of his head to the right, and orofacial twitching on the right side. The interictal EEG showed GSW at 3–4 Hz. The ictal EEG showed a GSW burst at 3–4 Hz, with a predominance in the right anterior region. MRI revealed no relevant abnormality. SPECT was performed at the age of 8 years because some features were suspected of indicating focal epilepsy. Later the seizures were completely controlled by a combination of ethosuximide (ESM) and VPA.

2.3.2. Patient 2

Patient 2 had a girl with mild intellectual disability and delayed motor development. Her IQ was 64 at the age of 4 years, and she was diagnosed with a pervasive developmental disorder. Her isolated myoclonic seizures started at the age of 2 years and 9 months. One month later, MAs appeared. Each seizure had a duration of 15 s or longer. She had dozens of seizures a day, but no GTC. Treatment with VPA and/or primidone was attempted, but neither drug was effective.

She was admitted to our epilepsy center at the age of 3 years. Ictal video EEG and polygraphic (EEG and EMG) recordings revealed that her seizures were MAs, absence seizures accompanied by intense bilateral rhythmical repetitive jerks; the myoclonus was more intensive on the left side. Some seizures were associated with head version toward the left. The interictal EEG showed bilateral synchronous and almost symmetrical diffuse 3 Hz spikes and waves. The ictal EEG showed generalized spikes and waves of 3–4 Hz, with an anterior predominance. The MRI demonstrated no relevant abnormality. SPECT was performed at the age of 4 years because some features raised the possibility of focal epilepsy. Treatment with ESM, VPA and lamotrigine completely controlled her seizures.

3. Results

3.1. Patient 1

Ictal SPECT was performed during a spontaneous MA that lasted 33 s. Injection of ECD was started at 14.5 s after the onset of the GSW burst and at 10.5 s after the clinical onset (Fig. 1A). The CBF was increased in both the perirolandic areas and the thalamus and in the left caudate nucleus, precuneus, lingual gyrus, and tegmentum, and decreased in the right middle frontal gyrus and bilateral orbitofrontal regions (Fig. 1B).

3.2. Patient 2

Ictal SPECT was performed during a MA that was induced by hyperventilation, which lasted 12 s. The rhythmic myoclonus was subtle. When ECD was administered for the ictal SPECT during seizures, her limbs did not rise as high as before, because of effective medication, although clinically it was the same seizure and the EEG profile was the same as before. The ECD was injected at 3.5 s after GSW burst onset and at 2.5 s after

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