Epileptic focus in a case of subcortical band heterotopia: SISCOM and ictal EEG findings

Kenjiro Kikuchi, M.D. 1) 2) Shin-ichiro Hamano, M.D. 1), Fumihiro Goto, M.D. 1), Akira Takahashi, R.T. 3), Hiroyuki Ida, M.D. 2)

1Division of Neurology, Saitama Children's Medical Center, 2100 Magome, Iwatsuki-ku, Saitama-city, Saitama 339-8551, Japan
2Department of Pediatrics, Jikei University School of Medicine, 3-25-8 Nishi-Shinbashi, Minato-ku, Tokyo 105-8471, Japan
3Department of Radiology, Saitama Children’s Medical Center, 2100 Magome, Iwatsuki-ku, Saitama-city, Saitama 339-8551, Japan

Key words: antiepileptic drugs, carbamazepine, zonisamide, mechanism of action, calcium-induced calcium releasing systems

Published online November 10, 2010

Abstract

We presented an 11-month-old-girl with subcortical band heterotopia who had focal epilepsy detected by subtraction single photon emission computed tomography (SPECT) co-registered to magnetic resonance (MR) imaging (SISCOM) and ictal electroencephalogram. She manifested cluster of partial seizures composed of asymmetric tonic posturing accompanied by head rotation to the right side. Ictal electroencephalogram showed that paroxysmal discharges were generated from the right parieto-occipital area and spread to sur-
rounding areas. SISCOM revealed hyperperfusion in the overlying cortex of the right superior temporal gyrus, which corresponded to the onset area of ictal epileptiform discharges. These neurofunctional findings corresponded to her clinical seizures. Her seizures were controlled by high-dose phenobarbital therapy. We considered that the patient had focal epilepsy and that the epileptic focus might be in the overlying cortex, but not in the subcortical band.

Introduction

Subcortical band heterotopia (SBH) is a rare neuronal migration disorder, which shows ectopic gray matter separated from the overlying cortex by white matter. Patients with SBH have been reported to have intractable epilepsies and mental retardations [1, 2]. These patients may present with either generalized epilepsies or focal epilepsies [2]. The epilepsies are usually evaluated by ictal and/or interictal electroencephalogram (EEG), and are rarely diagnosed by ictal single photon emission computed tomography (SPECT) or subtraction ictal SPECT coregistered to magnetic resonance (MR) imaging (SISCOM). To our knowledge, there are no reports on SISCOM evaluation of SBH during infancy. Here we report an 11-month-old girl with SBH who was diagnosed with focal epilepsy by SISCOM and ictal EEG.

Case report

An 11-month-old girl was admitted to our hospital because of intractable seizures persisting for 2 weeks. The perinatal and familial histories of the patient were unremarkable. Neurological examinations showed hypotonia in her lower extremities. She was able to sit by herself at 10 months, but could not crawl or stand up by herself at that time. Her seizures were characterized by asymmetric tonic posturing for 30 seconds accompanied by head rotation to the right side, which occurred 4 to 6 times per day.

Interictal EEG demonstrated irregular spike-and-waves and 3-4 Hz high-voltage slow waves (HVS) in the left centro-parieto-occipital area. As background EEG activities, low amplitude (20-30 μV) fast waves (30 Hz) were found in the right hemisphere. Brain MRI revealed a thin overlying cortex and an excessive band of subcortical gray matter, which was split by white matter from the frontal to the occipital region (Fig. 1). Bilateral occipital cortical surfaces were smooth and agyri. These abnormal findings were compatible with diffuse subcortical band heterotopia. Mutation in the DCX gene (178A>G) was found in exon 2.

Her seizure frequencies increased from 4-6 times per day to 4-5 times per hour, despite treatment with valproate. Ictal EEG demonstrated 3-4 Hz high-voltage slow (HVS) waves and sharp waves in the left centro-parieto-temporal area, which subsided and then 14-15 Hz fast activities with 90 μV amplitude appeared in the right mid-temporal area (Fig. 2A). These fast discharges propagated to surrounding areas and their amplitude

Epileptic focus in subcortical band heterotopia

Kenjiro Kikuchi, et al.
increased (Fig. 2B). They were followed by 5-
6 Hz high voltage polymorphic discharge
burst for about 20 seconds (Fig. 2C), and then
1-2 Hz HVS waves in the right hemisphere for
40 seconds (Fig. 2D). Finally these 1-2 Hz
HVS discharges terminated and sharp waves
reappeared in left centro-temporal area, simi-
lar to the interictal EEG findings mentioned
above (Fig. 2E). The total duration of ictal
activities was 75 seconds. Clinical manifesta-
tion of her seizures was recognized 30 seconds
after the 14-15 Hz fast activities appeared.

Technetium-99m-ethyl cysteinate dimmer
(ECD) was used for ictal SPECT. The patient
was injected with 185 MBq 99m-Tc-ECD
immediately after clinical seizure began, and
the seizures lasted for 45 seconds after ECD
injection. After ictal SPECT examination, she
was given high-dose phenobarbital (PB) ther-
apy by rectal suppository at an initial dose of
30 mg/kg/day for the first two days, followed
by a dose of 20 mg/kg/day for 2 days, 10 mg/
kg/day for 2 days, then 10 mg/kg/day by oral
administration for the sequential consecutive
days. Her seizures disappeared after PB ther-
apy.

Interictal ECD-SPECT was performed after
the disappearance of seizures. To elucidate the
ictal hyperperfusion regions, we used the
SISCOM analysis. The SISCOM images
demonstrated hyperperfusion in the overlying
cortex of the right superior temporal gyrus
(Fig. 3), which corresponded to ictal EEG
findings.

The patient had no seizures for more than 3
months after high-dose PB therapy, and was
able to crawl and stand up by herself at the last
follow-up.

Fig. 1. Axial T1-weighted magnetic resonance (MR) imaging
Axial T1-weighted images show diffuse subcortical heterotopic gray matter and mild pachygyria.
A. At the level of cerebral peduncle.
B. At the level of body of the lateral ventricle.
**Discussion**

We performed SISCOM and ictal EEG on an infant with SBH. Ictal EEG indicated that the ictal paroxysmal discharges originated from the right temporal region. SISCOM showed that the hyperperfusion area corresponded to the site of onset of ictal paroxysmal discharges, and that this hyperperfusion area seemed to be corresponded to the overlying cortex of the right superior temporal gyrus. To our knowledge, this is the first report of SISCOM analysis in infant with SBH.

Some experimental studies in SBH rat models reported the correlation between

---

**Fig. 2. Ictal electroencephalogram at the age of 11 months**

(A) 3-4 Hz high-voltage slow (HVS) waves and sharp waves are found in the left centro-parieto-temporal area. The HVS disappeared and burst of 14-15 Hz fast waves is observed in the right mid-temporal area.

(B) At 10 seconds after onset, fast waves have generalized and the amplitude increases.

(C) At 15 seconds after onset, 5-6 Hz high voltage polymorphic discharges burst for about 20 seconds.

(D) From 35 seconds after onset, 1-2 Hz HVS waves in the right hemisphere persist for 40 seconds.

(E) At 75 seconds after onset, HVS waves terminate and sharp waves remain in the left centro-temporal area.
overlying cortex and subcortical band. Chen et al. [3] concluded that normotopic neurons were responsible for initiating seizures in the dysplastic neocortex, because normotopic neurons were more likely to exhibit epileptiform activity than heterotopic neurons, and heterotopic neurons were recruited into spiking by activity initiated in normotopic neurons. These observations are consistent with our findings.

Ictal SPECT has the potential to localize the ictal onset zone accurately in a noninvasive manner, and is particularly useful in MRI-negative localized epilepsy and focal cortical dysplasias [4]. Furthermore, SISCOM is useful to define the onset site of seizure [5, 6]. To our knowledge, this is the first report of SISCOM finding of SBH in infant. SISCOM images showed that the hyperperfusion area corresponded to the onset region of the ictal EEG discharges, and that this region was located in the overlying cortex. These results show that the overlying cortex may be the epileptic focus in our SBH patient.

Fig 3. Subtraction ictal single-photon emission computed tomography (SPECT) coregistered to magnetic resonance (MR) imaging (SISCOM).

(A) In axial view, hyperperfusion is observed in the right frontotemporal area and right medial occipitotemporal area. (B) Coronal view. (C) Sagittal view. (D) Right lateral view. (B)-(D) show that hyperperfusion is located in the right superior temporal gyrus.
Surgical treatment has been performed on focal epilepsy patients with SBH. Bernasconi et al. [7] reported that five of eight patients had no significant improvement by surgical treatment and that temporal resection resulted in especially poor outcome. They suggested that SBH might have a multifocal generator due to widespread structural abnormality, even though clinical and neurofunctional studies proved the existence of focal epileptogenic lesions. Mai et al. [8] reported that one patient with SBH had good outcome by resection of the posterior third of the middle and inferior temporal gyri and part of the fusiform gyrus. However, both groups could not explain the mechanism of seizure generation related to the normal overlying cortex and the subcortical band. Our findings suggest that surgical treatment including corticectomy will be a therapeutic option for patients with SBH.

We demonstrated symptomatic focal epilepsy using SISCOM and ictal EEG in a pediatric patient with SBH, and speculated that the overlying cortex may be the epileptic focus.

Acknowledgement

We would like to thank Dr. Mitsuhiro Kato (Yamagata University School of Medicine) for analyzing the DCX gene, and Emeritus Professor Eric Johnson (Jichi Medical University, Tochigi, Japan) for his assistance with the preparation of the manuscript.

References


