결신발작 이후 확인된 일시적인 자기공명영상 이상 1예

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Transient Abnormalities on Magnetic Resonance Imaging after Absence Seizures

Magnetic resonance imaging (MRI) is recommended for patients with epileptic seizures to rule out an underlying focal lesion. However, abnormalities in idiopathic generalized epilepsy, including childhood absence epilepsy, cannot usually be identified using brain imaging modalities such as MRI. Peri-ictal MRI abnormalities have been most commonly reported secondary to status epilepticus and are rarely observed in patients with focal seizures and generalized tonic-clonic seizures. Transient peri-ictal MRI abnormalities in absence epilepsy are extremely rare. A five-year-old girl presented with a three-day history of absence seizures that persisted despite continued treatment with sodium valproate. Electroencephalography showed bursts of generalized 3-Hz spike-and-wave discharges, during and after hyperventilation. Abnormal cortex thickening in the left perisylvian region was detected on T2-weighted brain MRI, and cortical dysplasia or a tumor was suspected. The patient started treatment with lamotrigine and was seizure-free after one month. The abnormal MRI lesion was completely resolved at the two-month follow-up. We report on a patient with childhood absence epilepsy and reversible brain MRI abnormalities in the perisylvian region. To our knowledge, this is the first report of transient MRI abnormalities after absence seizures. Transient peri-ictal MRI abnormalities should be considered for differential diagnosis in patients with absence seizures and a focal abnormality on brain MRI.

Key Words: Epilepsy, Absence epilepsy, Peri-ictal MRI abnormalities, Differential diagnosis

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Introduction

The underlying cause of idiopathic generalized epilepsy (IGE) is unknown. By definition, IGE refers to cases without structural abnormalities on conventional magnetic resonance imaging (MRI) but with bilateral synchronous epileptic discharges on ictal or interictal electroencephalography (EEG). IGE in childhood includes several conditions such as childhood absence epilepsy (CAE) and epilepsy with generalized tonic-clonic seizures.

MRI is recommended for the detection of structural lesions and affected brain regions in patients with epileptic seizures. Transient peri-ictal MRI abnormalities (TPMA) due to seizures were first reported in 1987¹⁾ and several subsequent

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studies have reported MRI signal changes following focal or generalized status epilepticus. The signal changes on brain MRI during status epilepticus and seizures are caused by physiological changes occurring during brain seizures. These changes ultimately cause cerebral edema, rupture of the blood-brain barrier, and increased blood flow to the epileptogenic focus²⁾ TPMA show complete or partial regression within a few days. The differential diagnosis of seizure-induced TPMA should include acute ischemic stroke, encephalitis and venous infarction, posterior reversible encephalopathy syndrome, and brain tumors.

TPMA is most commonly reported secondary to status epilepticus and is rarely observed in patients with focal seizures, secondary to generalized tonic-clonic seizures, or absence seizures³⁾. We describe a patient with CAE with TPMA in the perisylvian region.

Case Report

A girl aged five years and three months, with allergic rhinitis, had a seizure where she suddenly stopped moving and speaking for a few seconds, and returned to normal when the seizure ended. The patient visited another hospital because of these symptoms and started treatment with sodium valproate (150 mg/day) after being diagnosed with CAE. After three days of medication, the patient visited our outpatient clinic owing to persistent symptoms lasting for a month.

The neurological examination revealed no abnormalities, and her development had been normal. There was no family history of seizures or delayed development. Laboratory examinations,

including blood and urine tests, and thyroid function tests revealed normal results. EEG revealed three episodes of electroclinical and electrographic seizures accompanied by bursts of generalized 3-Hz spike-and-wave discharges, during and after hyperventilation (Fig. 1). She underwent MRI to confirm the structural abnormality of persistent seizures. T2-weighted images (T2WI) showed abnormal cortical thickening in the left perisylvian region (Fig. 2), and a cortical dysplasia or a tumor was suspected. The patient started treatment with lamotrigine (1 mg/kg/day). The number of seizures decreased after 20 days, and seizures were completely absent after one month of treatment with lamotrigine (4 mg/kg/day) monotherapy. Seventy-three days after treatment, brain MRI revealed the complete resolution of the abnormal signals in the left perisylvian area; no abnormalities were observed (Fig. 3). And 9 months later, EEG showed no interictal epileptiform discharges

Fig. 2. T2-weighted brain magnetic resonance images obtained three days after the onset of absence seizures show abnormal cortical thickening in the left perisylvian region; (A) axial and (B) coronal view.

Fig. 1. Electroencephalography at three days after the onset of absence seizures reveals generalized 3-Hz spike-and-wave discharges during hyperventilation.

Fig. 3. Brain magnetic resonance imaging (MRI) after 73 days of treatment. (A) Axial T2-weighted and (B) coronal T1-weighted MRI revealed no abnormalities.

and normal records during awake and sleep. The patient continued receiving lamotrigine. Two years later, she was still seizure-free. We decided to withdraw the antiepileptic medication. She remained seizure-free for over four years.

Discussion

Brain MRI is the preferred imaging tool for patients with generalized or focal seizures and is widely used to rule out any structural cause of epileptic seizures. However, radiological changes during the peri-ictal period have rarely been reported. There have been several reports of partial status epilepticus where patients developed peri-ictal focal cerebral edemas that showed spontaneous regression on serial computerized tomography scans^{4,5}. These ictal-related MRI changes were identified after status epilepticus, cluster seizures, and single seizures⁶. However, these conditions are uncommon in clinical settings and are sometimes misdiagnosed as cortical dysplasia, tumors, and other structural abnormalities. Some studies reported an incidence as low as 0.007%⁷, and others report an incidence as high as 20.6%⁸. Studies on TPMA describe the variations in location, seizure type, and mechanisms, and there is no consensus on why only some patients develop TPMA.

The pathophysiological mechanism of TPMA is still unclear. TPMA lesions usually appear hypointense on T1WI and hyperintense on T2WI⁹. According to diffusion-weighted imaging (DWI)-MRI studies, increased cellular metabolism leading to increased regional blood flow is a possible etiology for post-ictal partially or totally reversible DWI impairments. These events lead to cytotoxic edema of neurons, which correlates to hyper-intensity on DWI and T2 and T2-fluid

attenuation inversion recovery imaging¹⁰. The report on TPMA in absence seizures is extremely rare. In a study of 21 IGE patients with absence seizures with perfusion MRI study, the mean cerebral blood flow of IGE patients was significantly increased in the left middle temporal gyrus, left parahippocampal gyrus and left fusiform gyrus and cortical dysfunction in the temporal lobe and fusiform gyrus has been reported to be related to epileptic discharges¹¹. Using SPECT, the cerebral blood flow in patients with myoclonic absence increased in thalamus, basal ganglia and the perirolandic cortical motor area¹².

In our patient, abnormal cortical thickening was observed in the left perisylvian region. The differential diagnosis included cortical dysplasia or a tumor. All laboratory examinations and the patient's development were normal. Accurate verification of TPMA is important because this condition is easily misdiagnosed as the primary epileptogenic lesion. Follow-up MRI can provide an accurate diagnosis of the lesions. The reason of TPMA in the perisylvian region is not clear. We hypothesized that the cerebral blood flow in perirolandic area might have been changed by the cortico-thalamic network oscillations in this patient.

To our knowledge, this is the first report of TPMA associated with CAE. It is important to recognize the potential presence of TPMA in patients with epileptic disorders and to differentiate between TPMA and potential epileptogenic lesion via radiological examination. It is important for clinicians to be aware of these reversible seizure-related changes on MRI. Of particular clinical significance is the need for close imaging follow-up to document the regression of acute changes and to monitor long-term sequelae. Therefore, understanding the characteristics of this phenomenon is essential to avoid the use of other unnecessary and more invasive diagnostic techniques.

요약

뇌전증 환자에서 자기공명영상은 기질적인 병변을 확인하기 위해 권장된다. 그러나 청소년기 결신 발작을 포함한 특발성 전신 발작에서 이상소견은 대개 자기공명영상과 같은 뇌 영상 기법을 사용하여 확인할 수 없다. 발작기 동안 자기공명영상 이상은 뇌전증 지속증에 의한 이차성으로 가장 흔히 보고되었으며, 부분발작과 전신 강직간대 발작 환자에서 거의 나타나지 않는다. 결신 발작 동안 자기공명영상의 이상은 매우 드물다. 5세 소녀가 sodium valproate로 계속 치료를 했음에도 불구하고 3일간 발생한 결신 발작으로 내원하였다. 뇌파 검사에서 과다호흡 이후 3 Hz 극서파를 보였다. T2 강조 뇌 자기공명영상에서 좌측 외측구 주변의 비정상 피질 비후가 확인되었고, 피질 형성 이상이나 종양이 의심되었다. 환자는 lamotrigine으로 치료를 시작하

었고 이후 발작은 없었다. 2개월 후 추적관찰로 뇌 자기공명영상을 시행하였고 이전의 비정상 부분은 확인되지 않았다. 저자들은 유년기 결신 발작 시 확인된 외측구 주변의 가역적인 뇌 자기공명영상 이상에 대해 보고하는 바이며 이것은 결신 발작 후 일과성 자기공명영상 이상 소견의 첫 번째 보고로 의미가 있다. 결신 발작과 부분 발작에서 자기공명영상 이상은 일시적인 이상 일 수 있으며 다른 질환과 감별 시 고려해야 할 것이다.

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