Unusual Magnetic Resonance Imaging Abnormality in Nonketotic Hyperglycemia – related Epilepsia Partialis Continua

Jui-Ping Tsai[1], Jau-Jiuan Sheu[1,2], Kevin Li-Chun Hsieh[3,4]

Departments of [1]Neurology and [3]Medical Imaging, Taipei Medical University Hospital, [2]Department of Neurology, School of Medicine, College of Medicine, Taipei Medical University, [4]Research Center of Translational Imaging, College of Medicine, Taipei Medical University, Taipei, Taiwan

Abstract

Epilepsia partialis continua (EPC) is a rare epileptic syndrome, presenting as continuous focal motor seizures for a period of minutes, hours, or days. EPC may develop in patients with cerebral cortical lesions and occasionally may develop in patients with metabolic disorders, such as nonketotic hyperglycemia (NKH). Here, we report a case of EPC following NKH, showing an unusual magnetic resonance imaging (MRI) finding of concurrent hypointensity on susceptibility-weighted image (SWI) and T2-weighted image (T2WI) with leptomeningeal and cortical enhancement, which have never been reported. A 68-year-old woman presented to our emergency department with a 3-day history of involuntary repeated contraction of the right side of the face and upper limb. Laboratory data revealed NKH of diabetes mellitus. Electroencephalography (EEG) was unremarkable. Brain MRI revealed focal cortical and leptomeningeal enhancement together with subcortical T2 shortening and SWI hypointensity of the left frontal operculum. She responded well for hyperglycemia and antiepileptic drug therapy. Follow-up brain MRI performed 1 week later showed complete resolution of the abnormal signal and enhancement in the same region. Although EPC caused by NKH occurs rarely, it may result in an MRI abnormality of subcortical hypointensity on SWI and T2WI with leptomeningeal and cortical enhancement, which may be misinterpreted as other brain pathologies. Rapidly recognition is important because timely treatment with hydration and correction of hyperglycemia can lead to better outcome. We recommend such cases of metabolic disorder (such as hyperglycemia) for early consideration, particularly in the elderly.

Keywords: Epilepsia partialis continua, magnetic resonance imaging, nonketotic hyperglycemia, susceptibility-weighted image

Introduction

Epilepsia partialis continua (EPC) is defined as a form of focal status epilepticus, presenting as continuous focal motor seizures confined to one part of the body for a period of minutes, hours, or days.[1] EPC may develop in patients with cerebral cortical lesions and occasionally may develop in patients with metabolic disorders, such as nonketotic hyperglycemia (NKH). Radiological abnormalities in NKH related to EPC are characterized by asymmetry or unilateral hyperintensity on T2-weighted image (T2WI) and fluid attenuation inversion recovery (FLAIR) images.[2] Transient T2 shortening in the subcortical white matter on T2WI has been reported in previous cases.[2-4] However, concurrent hypointensity on susceptibility-weighted image (SWI) and T2WI with leptomeningeal and cortical enhancement have never been presented simultaneously. Here, we report a case of EPC following NKH, showing an unusual magnetic resonance imaging (MRI) finding of leptomeningeal and cortical enhancement together with a subcortical hypointense signal change.

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Website: www.annalsofian.org

DOI: 10.4103/aian.AIAN_386_17

Case Report

A 68-year-old woman with diabetes mellitus and hypertension treated at the clinic for more than 10 years presented to our emergency department with a 3-day history of sudden and repetitive right-side partial motor seizures affecting her face (mouth angle and eyelid twitching) and the right upper limb. The ictal phase occurred every 1–2 min and increased in frequency. She was fully conscious during ictal and interictal periods. She did not report a history of fever, head trauma,

Address for correspondence: Assistant Prof. Kevin Li-Chun Hsieh, Department of Radiology, School of Medicine, College of Medicine, Taipei Medical University, Taipei, Taiwan.
E-mail: kevinh9396@gmail.com

How to cite this article: Tsai JP, Sheu JJ, Hsieh KL. Unusual magnetic resonance imaging abnormality in nonketotic hyperglycemia – related epilepsia partialis continua. Ann Indian Acad Neurol 2018;21:225-7.

Todd's paralysis, or a similar episode. Her neurological examination was grossly intact. There was no sign of any infection identified.

Laboratory blood tests revealed hyperglycemia (371 mg/dL), hyperosmolality (299 mOsm/L), a high level of hemoglobin A1c (11.4%), and hyponatremia (133 mOsm/L). Ketone bodies were not present in serum and urine. EEG did not show epileptiform discharge. Lumbar puncture was recommended but refused by the patient. Brain MRI was performed on the 5th day after first seizure attack and revealed focal cortical ribbon enhancement together with T2 hypointensity change at the subcortical white matter of the left frontal operculum. The involved regions also showed subcortical hypointensity on SWI [Figure 1]. This finding is consistent with that of epilepticus partialis continua with transient disruption of the brain–blood barrier. Together with the clinical presentation and laboratory findings, a diagnosis of hyperglycemia-induced epilepticus partialis continua was established. After normalizing the blood glucose level with insulin therapy, along with hydration and administration of valproic acid (1200 mg intravenous daily) and oxcarbazepine (600 mg oral daily), no seizure episode was detected 5 days after the treatment.

After 1 week, follow-up MRI study revealed nearly complete resolution of the aforementioned signal abnormality [Figure 2]. The patient remained seizure free, and we discontinued antiepileptic agents before discharge.

Discussion

Many conditions have been implicated in the pathogenesis of EPC, including various cortical lesions and metabolic disorders, such as hyperglycemia and hyponatremia. NKH may be the first sign of diabetes mellitus, which subsequently results in EPC, particularly in elderly patients with diabetes.[5] However, the pathophysiology of EPC secondary to NKH is unclear. It was hypothesized that hyperglycemia, a proconvulsant, may precipitate EPC by inhibiting the Kreb cycle and reducing the gamma-aminobutyric acid (GABA) level. Hyperglycemia also induces intracellular hyperosmolality and dehydration, contributes to neuronal hyperexcitability, lowers the seizure threshold, and then triggers seizures.[5,6]

Transient subcortical hypointensity changes in the subcortical white matter on T2WI and FLAIR images have been documented in ischemia, multiple sclerosis, leptomeningeal metastasis, and meningitis, rarely reported in those with prolonged seizures following NKH.[6,7] The leptomeningeal enhancement is an unusual manifestation in hyperglycemic state but has been reported in cases with hyperglycemia-related seizures.[4,8] The associated differential diagnosis of leptomeningeal enhancement with subcortical T2 hypointensity includes focal patchy meningitis, encephalitis, hemorrhagic infarcts, malignancy, and paraneoplastic process.[6] Almost these etiologies can be excluded in our case on the basis of patient's history and clinical manifestations with a shortly reversible clinical course and MRI abnormalities.

To the best of our knowledge, SWI and T2 hypointensities in addition to leptomeningeal and cortical enhancement, like our case, have never been reported in patients with EPC secondary to NKH. We surmise that the enhancement of leptomeninges may be due to vasodilatation caused by seizure-induced hypermetabolism and an increase in brain perfusion. Prolonged ictal activity may result in hypoxia and acidosis, which lead to disruption of the blood–brain barrier, permitting leakage of contrast medium and subsequent cortical enhancement.[1-3] In addition, the reason for subcortical T2 shortening on imaging is not fully understood. Postulated mechanisms may include ischemia, mineral deposition, free radical, or iron accumulation due to excitotoxicity of axonal damage during seizures.[2] The hypointense signals at the matching areas on SWI in our case also support this hypothesis since low signal in SWI may represent hemorrhage, mineral accumulation, increased vascularity, or increased oxygen extraction related to ischemia.[9] Striatal hypointensities in SWI have also been reported in hyperglycemia-induced hemichorea–hemiballism,[10] which was proposed to be related to manganese and zinc deposition within the gemistocytes (reactive astrocytes response to acute injuries). Therefore, it is possible that the two disease entities may share a similar pathway in their pathogenesis.

Because the abnormal signals regress after normalization of hyperglycemia, it is more likely to be related to the underlying hyperglycemic state.[11] According to Hung's report, the abnormal signal in brain images is more related to prolonged moderate

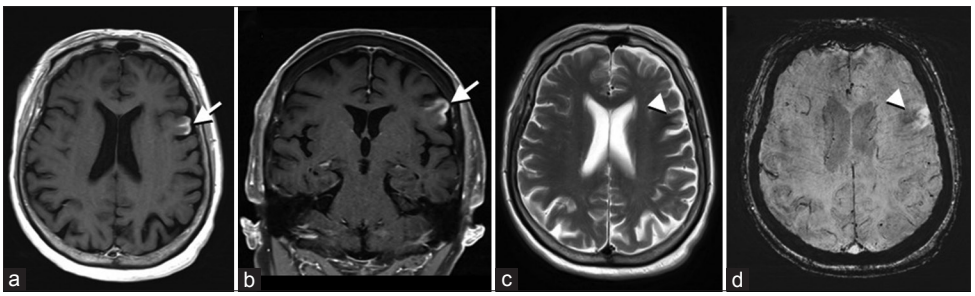

Figure 1: Brain magnetic resonance imaging revealed focal cortical and leptomeningeal enhancement over the left inferior and middle frontal gyri (Brodmann area 44, arrows) on contrast-enhanced T1-weighted image (a: axial, b: coronal) along with abnormal hypointensity at the subcortical white matter (arrowheads) on T2-weighted image (c) and susceptibility-weighted image (d). No abnormal signal was noted at the affected cortices on T2-weighted image

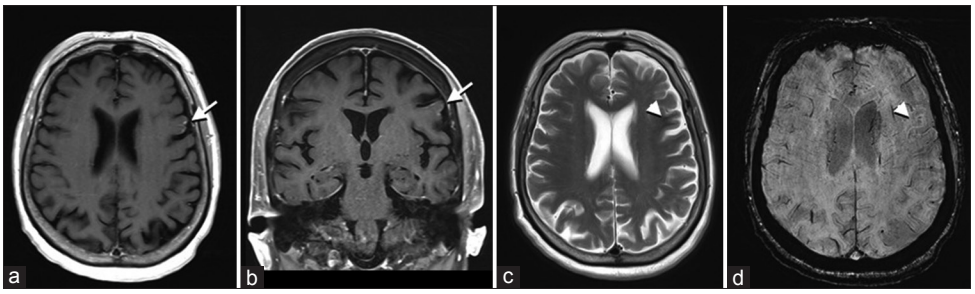

Figure 2: After 1 week, follow-up brain magnetic resonance imaging showed nearly complete resolution of the abnormal enhancement (arrows) in contrast-enhanced T1-weighted image (a: axial, b: coronal) with some residual subcortical T2 shortening (arrowhead in c). Complete regression of subcortical hypointensity on susceptibility-weighted image is depicted (arrowhead in d)

hyperglycemia (presented by markedly elevated glycated hemoglobin) than the hyperglycemia level in the acute stage.[12]

The time to lesion disappearance in MRI is quite variable, depending on the types of epilepsy, different locations, extent of signal, and morphological characteristics. The sequelae of these insults may range from focal atrophy to complete normalization.[13] The lesion of our patient involved Brodmann area 44 [Figure 1], which is a part of Broca's region and is considered a part of specialized parieto-premotor network devoted to motor control of oropharyngeal movements and speech production.[14]

Individuals with Broca's aphasia typically present with a nonfluent aphasia. However, our patient was able to understand the language. Her lingual expression was also correct and meaningful at initial interview. More aphasia tests may be warranted in the future to further clarify this issue.

Regarding treatment, hyperglycemia-related seizures resolve after the normalization of the blood sugar level and adequate hydration. Although anticonvulsants are not necessary in most cases with EPC following hyperglycemia, a short-term use of valproic acid, a GABAergic drug, showed benefit and improved seizure control in our patient.

Conclusions

Although NKH-related EPC is uncommon, it may result in an MRI abnormality of subcortical hypointensity on SWI and T2WI with leptomeningeal and cortical enhancement, which may be misinterpreted as other brain pathologies. Being aware of this etiology of EPC is crucial for the timely diagnosis and appropriate management, especially in elderly patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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