

LETTER TO THE EDITOR

Combined Hemichorea and Seizures in a Patient with Nonketotic Hyperglycemia

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Dear Editor,

Nonketotic hyperglycemia (NKH) is characterized by elevated blood glucose levels associated with hyperosmolarity and dehydration in the absence of ketone formation.¹ NKH may produce various neurological symptoms, including an altered mentality, seizures, and involuntary movements, such as chorea and tremor.¹ Although both chorea and seizures are often observed in patients with NKH, their coexistence in these patients is extremely rare. Herein, we report a patient who concurrently developed hemichorea and seizures secondary to NKH.

A 50-year-old man presented with involuntary abnormal movements of the left upper- and lower-limbs that had developed a few days prior to seeking treatment. He also described three episodes of loss of consciousness with diffuse tonic contractions. The patient had a 6-year history of type 2 diabetes mellitus (DM), which was poorly controlled with oral hypoglycemic agents. He had no history of movement disorders or epilepsy. At the emergency room, he showed left-sided choreiform movements, which were continuous and could not be suppressed voluntarily. Furthermore, he suddenly showed asymmetric tonic limb posturing with extension on the left side and secondary generalization, which lasted for less than 1 minute. Laboratory tests revealed a blood glucose level of 423 mg/dL, with a HbA1c of 14.6%, and the calculated serum osmolality was 290 mosm/kg. Serum ketones were negative. Magnetic resonance imaging of the brain showed an abnormally strong hyperintense signal in the right caudate and lentiform nuclei on T1-weighted images (Figure 1A)

and an isointense signal on T2-weighted images (Figure 1B). Electroencephalography (EEG) revealed periodic rhythmic spike-wave discharges originating from the right temporo-occipital area without clinical signs or symptoms, suggesting subclinical seizures (Supplementary Figure 1 in the online-only Data Supplement). These epileptiform discharges were detected two or three times hourly and lasted for 20–30 seconds. He was diagnosed with NKH-induced hemichorea and seizures. He was started on insulin therapy with fluid replacement. On day 1, his choreiform movements persisted (Supplementary Video 1 in the online-only Data Supplement). On day 4, the patient's serum glucose levels were maintained to below 200 mg/dL and his involuntary movements improved gradually. Follow-up EEG on day 7 did not reveal any epileptiform discharges. The patient was discharged on day 10 after his symptoms had resolved completely.

The clinical, laboratory, electrophysiological, and brain imaging findings of our patient strongly suggested NKH-related chorea and seizures. To the best of our knowledge, this is the second case report of combined chorea and seizures in a patient with NKH. Chung et al.² first reported the co-occurrence of these symptoms due to NKH. Interestingly, the previous case showed hemichorea with focal seizures of temporo-occipital onset, which is consistent with our case.

Chorea related to NKH is mainly observed in elderly Asian women with type 2 DM.^{3,4} In a meta-analysis including 53 NKH-associated chorea patients, more than 80% of the patients showed hemichorea with contralateral putaminal hyperintensity,³ which

Received: August 7, 2019 Revised: September 10, 2019 Accepted: September 23, 2019

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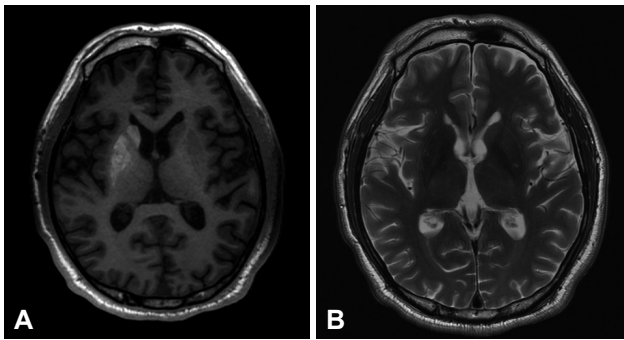


Figure 1. Brain magnetic resonance imaging (MRI) findings. A: MRI revealed increased signal intensities in the right caudate and lentiform nuclei in T1-weighted axial imaging. B: There were no significant signal abnormalities in T2-weighted axial imaging.

is in agreement with our case. Although typical neuroleptic drugs and sometimes benzodiazepines are needed for the management of chorea in many NKH cases,^{3,5} this abnormal movement can be treated only with appropriate control of blood glucose levels, which is supported by our findings. In the abovementioned meta-analysis, 16 patients were also treated only with blood glucose control and showed complete remission within 1 month after symptom onset.³ The favorable prognosis of chorea in our patient may be attributed to the relatively young age at onset. Lee et al.⁵ reported that older age was the sole significant predictor for the persistence of glycemic chorea/ballism one month after onset, and the mean age (standard deviation) of glycemic chorea/ballism patients in their study was 69.8 (13.2) years.

It is known that seizures due to hypoglycemia are mainly generalized, whereas seizures due to hyperglycemia are commonly focal in nature.⁶ The most frequent type of seizures due to hyperglycemia are motor seizures in the form of *epilepsia partialis continua*.⁶ NKH-related seizures are generally resistant to antiepileptic drugs but respond to the correction of glucose levels and appropriate fluid replacement.⁶

The pathophysiology of chorea and seizures related to NKH is not well understood. One possible mechanism is that hyperglycemia-induced autoregulatory dysfunction leads to hypoperfusion and subsequent activation of anaerobic metabolism. This condition results in a decreased level of gamma-aminobutyric acid, which may not only cause basal ganglia dysfunction but also lower the threshold for seizure activity. Another possible mechanism is that hyperviscosity resulting from hyperglycemia causes vascular insufficiency that provokes transient dysfunction of the basal ganglia and some cortical regions. However, these

hypotheses could not explain why, unlike seizures, chorea generally persists after normalization of glucose levels, and this phenomenon needs to be further investigated. A recent study using transcranial magnetic stimulation showed that hyperglycemic hemichorea is related to increased inhibitory activity in the motor cortex.⁷ This finding may reflect a compensatory mechanism to reduce the severity of symptoms.

Supplementary Video Legends

Video 1. The pattern of involuntary movements was continuous, arrhythmic, and dance-like and were confined to left upper and lower limbs.

Supplementary Materials

The online-only Data Supplement is available with this article at <https://doi.org/10.14802/jmd.19058>.

Conflicts of Interest

The authors have no conflicts of interest or financial support to report.

Author Contributions

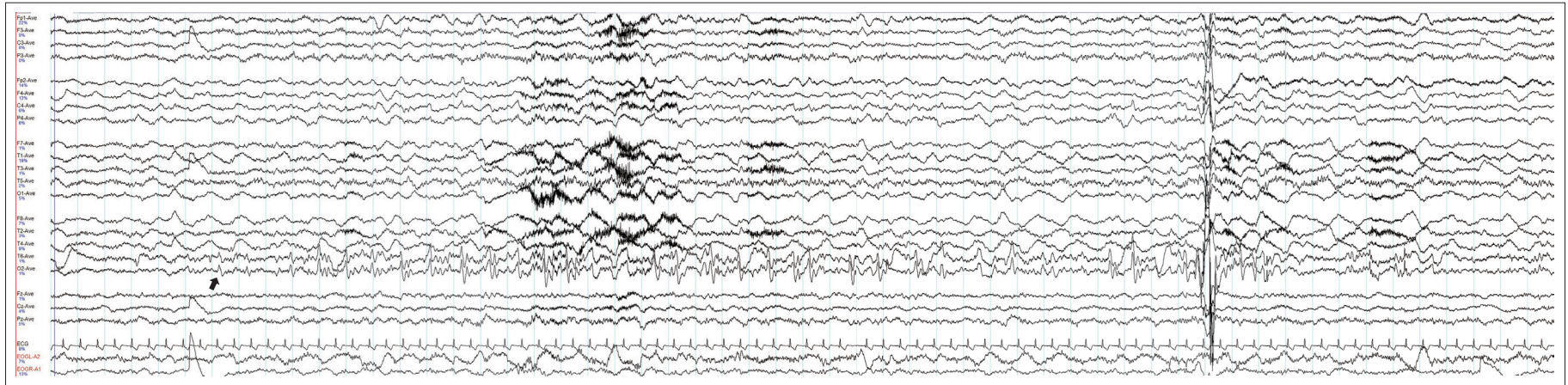
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Supplementary Figure 1. Electroencephalography (EEG) findings. EEG revealed repetitive rhythmic spike-wave discharges originating from the T6 and O2 electrodes (arrow).