



Hyperglycemia-induced Hemichorea and Simultaneous Acute Ischemic Stroke: A Case Report

Hiperglisemi ile İlişkili Hemikore ve Eş Zamanlı Akut İskemik İnme: Bir Olgu Sunumu

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Dear Editor,

Hemichorea associated with hyperglycemia (HCAH) is a rare, specific syndrome with non-ketotic hyperglycemia, hemichorea and hyperintense appearance in the basal ganglia in T1-weighted brain magnetic resonance imaging (MRI) (1). It is thought that hyperglycemia-induced decrease in gamma-aminobutyric acid (GABA) levels (2), hyperviscosity, and vascular insufficiency lead to incomplete, transient dysfunction in the striatum and results in chorea (3). The most common cause of hemichorea is ischemic lesions affecting the basal ganglia (4). Striatal ischemic lesions are frequently seen in patients with HCAH. However, very few patients with HCAH have concomitant acute striatal infarctions (3).

In this article, our aim was to present a patient who was admitted with hyperglycemia and hemichorea and who had hyperdense appearance on cranial computed tomography (CT) scan and ipsilateral acute infarction of the corona radiata. We discuss the relationship between hyperglycemia, ischemic stroke, and hemichorea in light of the literature.

A 52-year-old male patient presented with right-sided numbness and involuntary movements that started acutely 1 week ago. His blood glucose level was 556 mg/dL at the time of admission. It was learned that the patient did not receive treatment for diabetes. The serum osmolarity was 285 mmol/kg and the pH in the blood gas test was 7.32. Ketone was negative in complete urinalysis. The glycated hemoglobin (HbA1c) value was 14. On neurologic examination, hypoesthesia and choreiform movements were observed in the right upper extremity. Cranial

CT revealed a hyperdense appearance in the left striatum. Cranial MRI revealed restricted diffusion in the left corona radiata compatible with acute infarction (Figure 1). The hyperdense lesion in cranial CT was hyperintense on T2-weighted, fluid-attenuated inversion recovery (FLAIR), diffusion-weighted and apparent diffusion coefficient (ADC) sequences, but not visible on T1-weighted sequences (Figure 2). Acetylsalicylic acid treatment was initiated. The blood glucose level was reduced by insulin treatment. Carotid MR angiography revealed 40% stenosis in the right internal carotid artery (ICA); no stenosis was seen in the left ICA. Normal sinus rhythm was seen on electrocardiography (ECG). Echocardiogram (ECHO) and 24-hour rhythm Holter were evaluated as normal. The patient was discharged with antiplatelet treatment and insulin therapy, following diminished choreiform movements with normalized blood glucose levels. At the follow-up after ten days, involuntary movements were considerably reduced, but continued.

The high blood glucose level and presence of the hyperdense lesion in the left putamen on cranial CT gave rise to the diagnosis of HCAH in the patient who presented with acute onset right-sided choreiform movements in the upper extremity.

In a review of patients with HCHA, it was reported that T1-weighted hyperintense lesions were observed in the basal ganglia contralateral to the chorea in the majority of patients and putamen was involved in all cases. MRI lesions and involuntary movements were ameliorated completely or improved within 6 months. Blood glucose control was sufficient in the treatment of movement disorder in about one third of patients. Haloperidol monotherapy

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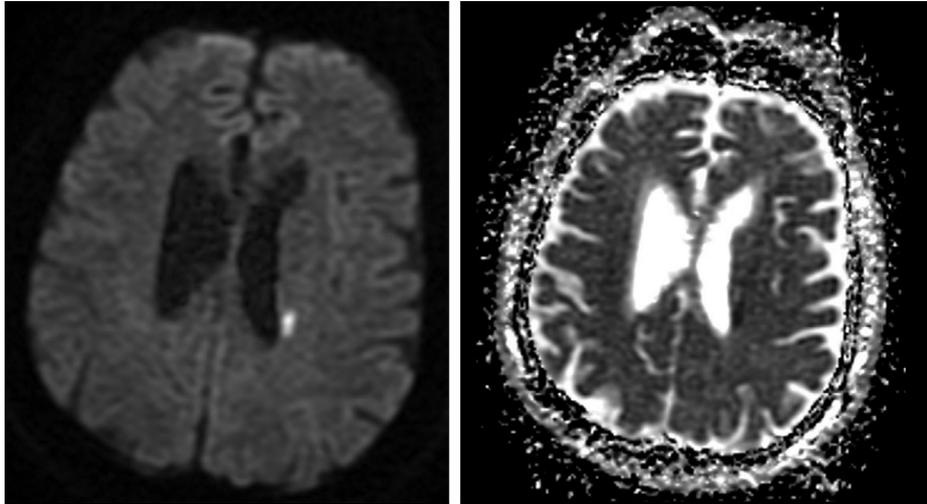


Figure 1. Restricted diffusion in the left corona radiata compatible with acute ischemia on diffusion- apparent diffusion coefficient sequences.

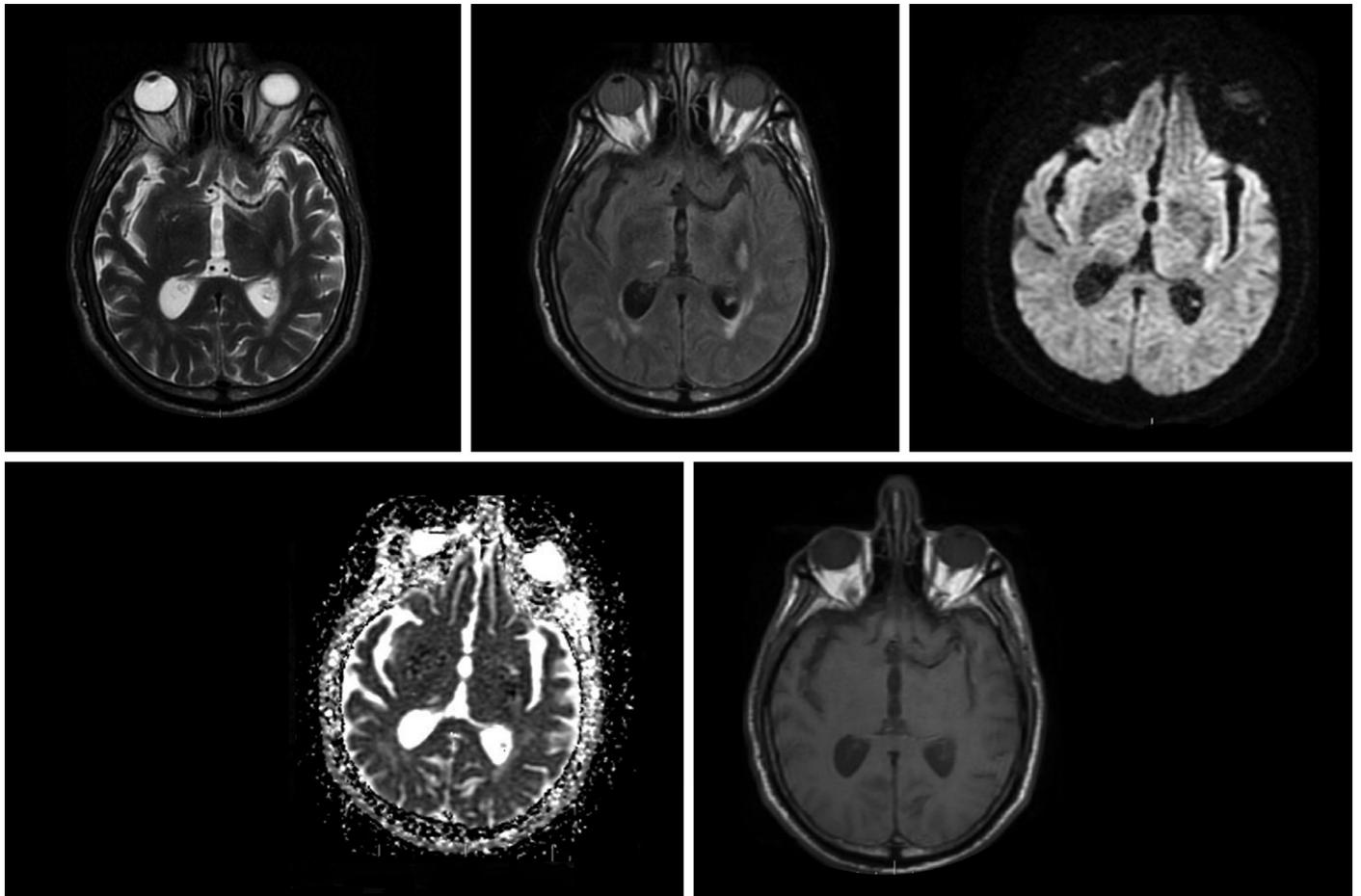


Figure 2. The lesion in the left putamen is hyperintense T2-weighted, fluid-attenuated inversion recovery, diffusion-weighted and apparent diffusion coefficient sequences (from left to right, respectively), but it is not seen on T1-weighted sequence (rightmost).

was given in one third and was given in combination with other drugs in one third of patients. Involuntary movements can recur after complete amelioration (1).

For the patients the mean blood glucose level, HbA1c value, and serum osmolarity were reported as 481.5 mg/dL, 14.4%, and 305.9 mmol/kg, respectively. The characteristics of our

patient were consistent with the literature, except the absence of hyperintense lesion on T1-weighted sequences.

Based on signal changes on diffusion-weighted, ADC, and T2-weighted sequences seen in patients, the underlying mechanism of HCAH has been suggested to be hyperviscosity and vasogenic edema (3). Although the serum osmolarity of our patient was normal, the presence of a hyperintense lesion in the left striatum on T2-weighted, FLAIR, diffusion-weighted and ADC sequences suggested vasogenic edema.

Other mechanisms underlying HCHA are vascular insufficiency with decreased GABA and acetylcholine. Both acetates and GABA are rapidly consumed during non-ketotic hyperglycemia. Depletion of acetate reduces acetylcholine synthesis. Decreases in GABA and acetylcholine, metabolic acidosis, and energy deficiency lead to dysfunction in the basal ganglia, and consequently to chorea (2).

The most common cause of hemichorea is ischemic lesions affecting the basal ganglia. Although hemichorea is most commonly caused by subthalamic nuclear lesions, the involvement of the striatopallidal pathway due to lesions affecting other basal ganglia regions results in the disinhibition of motor thalamic structures by reducing inhibitor signals to pallidal neurons, which leads to hemichorea (4).

Acute striatal infarction rarely accompanies HCHA (3). As in our case, HCHA with hyperdense striatal lesion contralateral to hemichorea in cranial CT and concomitant acute ischemic infarction is very rare and there is only one example in the literature. In this case report, a patient with HCHA with T1 hyperintensity in the right basal ganglion and acute infarction in the right corona radiata was presented (5). Two hypotheses have been proposed; first, stroke leads to decompensation of diabetes and increased cerebral permeability, and hyperglycemia lead to hemichorea. Second, hyperglycemia triggers stroke through ischemic damage due to hyperviscosity. The close proximity of the ischemic lesion and T1 hyperintensity reinforces the second hypothesis, but it was emphasized that the first hypothesis would be true if the T1 hyperintensity was included in the penumbra of the ischemic lesion. In addition, it was also emphasized that the probability of ischemic lesions of the basal ganglion and surrounding white matter causing vascular hemichorea-hemiballismus could not be excluded (5). Carotid MR angiography

of the patient showed no stenosis of the carotid artery on the symptomatic side. Cardioembolism was also excluded with ECG, ECHO, and 24-hour rhythm Holter. As the MR angiography was normal, we think that a striatal lesion cannot be included in the penumbra of the acute infarction. As in the first hypothesis in the aforementioned article (5), we believe that acute ischemic stroke decompensates diabetes and also increases cerebral permeability, thus hyperglycemia leads to hemichorea on this basis.

In conclusion, acute ischemic stroke may occur concomitantly in patients with HCHA; therefore, diffusion-weighted and ADC sequences should be included in imaging modalities in the examination of these patients.

Ethics

Informed Consent: Not taken (case reports are exempt research).

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Authorship Contributions

Surgical and Medical Practices: M.Ç.A., M.M.A., Concept: M.Ç.A., Design: M.M.A., Data Collection or Processing: M.Ç.A., Analysis or Interpretation: M.Ç.A., M.M.A., Literature Search: M.Ç.A., Writing: MMA

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