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## Case Report

# Hemichorea-Hemiballism in A 50-Year-Old Man With Newly Diagnosed Diabetes

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### ABSTRACT

**Objective:** Hyperglycemia may cause acute central nervous system dysfunction manifesting as agonizing involuntary movements due to insult to the basal ganglia. We report a case of hemichorea-hemiballism (HCHB) in a patient with diabetes.

**Method:** Clinical assessment of the patient was performed, along with laboratory tests and brain imaging.

**Results:** The patient was a 50-year-old man with newly detected diabetes with persistent involuntary movement of the right upper and lower limbs for few weeks. The involuntary movement was non-rhythmic, nonpatterned, purposeless, and often jerky with variable amplitude and frequency, sometimes wild and flailing in the form of hemichorea with a ballistic component (HCHB). He had a history of poor compliance to prescribed oral antidiabetic drugs. At presentation, although he was hemodynamically stable, random capillary blood glucose level was 18 mmol/L and glycated hemoglobin A1 level was 15.1% (141.5 mmol/mol). Clinical examination did not reveal any focal deficit or positive Babinski sign. There was a hyperintensity in the left basal ganglia region in T1-weighted magnetic resonance imaging (MRI) of the brain, which was iso-to-hyperintense in T2-weighted image and fluid-attenuated inversion recovery sequence. There was no restriction of diffusion on the diffusion-weighted image or blooming on gradient echo sequences, indicating absence of infarction or hemorrhage. Control of hyperglycemia resulted in disappearance of the involuntary movement within 1 month.

**Conclusion:** While there are many differential diagnoses for HCHB, the clinical scenario suggests hyperglycemia as the underlying cause in this patient. This case reiterates that multiple central nervous system manifestations may be attributable to diabetes.

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## Introduction

The central nervous system (CNS) effects of diabetes mellitus (DM) are not uncommon; yet, they are not often highlighted in the literature.<sup>1</sup> In addition to common CNS disorders like stroke and dementia, DM also predisposes patients to seizures and involuntary movements. Hyperglycemia is known to affect the basal ganglia in the setting of the hyperglycemic hyperosmolar state, resulting in hemichorea-hemiballism (HCHB).<sup>2,3</sup> Although stroke is the major

cause of this involuntary movement, hyperglycemia is the second most common cause.<sup>3</sup> A proposed hypothesis of this movement disorder includes hyperglycemia-induced minimal ischemia and hypoperfusion leading to gamma amino butyric acid depletion.<sup>4</sup> The hyperintensity in the basal ganglia region observed on imaging is thought to be the result of protein desiccation in the course of Wallerian degeneration.<sup>5</sup> Correction of hyperglycemia provides dramatic improvement in clinical features. In this article, we describe a case of hyperglycemia-induced HCHB and discuss its clinical implications.

## Case Report

A 50-year-old man presented to the neurology department of National Institute of Neurosciences and Hospital, Dhaka, with persistent involuntary movement in the right upper and lower limbs for the past 20 days. This movement disappeared only

*Abbreviations:* CNS, central nervous system; DM, diabetes mellitus; DWI, diffusion-weighted imaging; HCHB, hemichorea-hemiballism; MRI, magnetic resonance imaging.

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during sleep, and it was so severe that his basic activities of daily living were impaired. He had no family history of such disorders. One month before the onset of this movement disorder, he was experiencing polyuria, polydipsia, and weight loss for several days and was diagnosed with DM at a primary health care facility. His antidiabetic treatment consisted of metformin 1000 mg/day and glimepiride 2 mg/day, apart from lifestyle modification advice; however, his compliance with advice and medications was poor.

Examination revealed an involuntary movement of right upper and lower limbs in the form of hemichorea with a ballistic component—that is, nonrhythmic, nonpatterned, purposeless, and often jerky with variable amplitude and frequencies, sometimes wild and flailing (Fig. 1; scan QR code to see the video; video was recorded and used for scientific purposes only and with informed consent of the patient). He was alert and cooperative but had poor nutritional status. The pulse rate was 90/min with regular rhythm, blood pressure was 120/80 mm Hg, and random capillary blood glucose level was 18 mmol/L. Higher psychic function and cranial nerves were normal. There was no tremor or rigidity. Muscle power and reflexes were normal, and there was no clonus or positive Babinski sign. Cerebellar signs were not present. Stance and gait were unstable. Sensations in all limbs were intact. Other systemic evaluations were unremarkable.

His hemoglobin A1C level was 15.1% (141.5 mmol/mol), and fasting and postprandial blood glucose levels were 14 mmol/L and 19 mmol/L, respectively, while on glimepiride (3 mg/day) and metformin (1 g/day). Complete blood count revealed a hemoglobin level of 12.6 g/dL and erythrocyte sedimentation rate of 21 mm in the first hour. White blood cell and platelet counts were normal. Routine urinalysis revealed presence of reducing substances (++) but absence of albumin. Serum creatinine and electrolytes were within normal limits (Table).

Brain magnetic resonance imaging (MRI) of the patient is shown in Figure 2. Computed tomography scan of the brain was unremarkable, but MRI of the brain revealed a T1 hyperintense area at the left caudate and lentiform nuclei, which was iso- to hyperintense in T2/fluid-attenuated inversion recovery. There was no restriction of diffusion on the diffusion-weighted image or a blooming on gradient echo sequences and no remarkable enhancement after intravenous contrast.

The patient was treated with regular and intermediate-acting human insulin, which were gradually titrated until glycemic targets were achieved. Involuntary movement was treated symptomatically with haloperidol and tetrabenazine. After 1 month of



Fig. 1. Scan the QR code to see the video of involuntary movement of the patient (recorded and used for scientific purposes only and with informed consent of the patient). QR = quick response.

**Table**  
Laboratory Findings of the Patient

Test	Result
HbA1C	15.1% (141.5 mmol/mol)
Fasting plasma glucose	14.2 mmol/L
Plasma glucose 2 h ABF	19.1 mmol/L
Complete blood count	Hemoglobin 12.6 g/dL ESR 21 mm in first h TC $08 \times 10^9/L$ ; DC Neutrophil 57%, Lymphocyte 32% Platelet $245 \times 10^9/L$
Urine examination	Sugar (++) , albumin—nil, ketone—absent
Serum creatinine	0.67 mg/dL
Serum electrolytes	$Na^+$ 140 mmol/L, $K^+$ 3.2 mmol/L, $Cl^-$ 109 mmol/L

Abbreviations: ABF = after breakfast; DC = differential count; ESR = erythrocyte sedimentation rate; TC = total count.

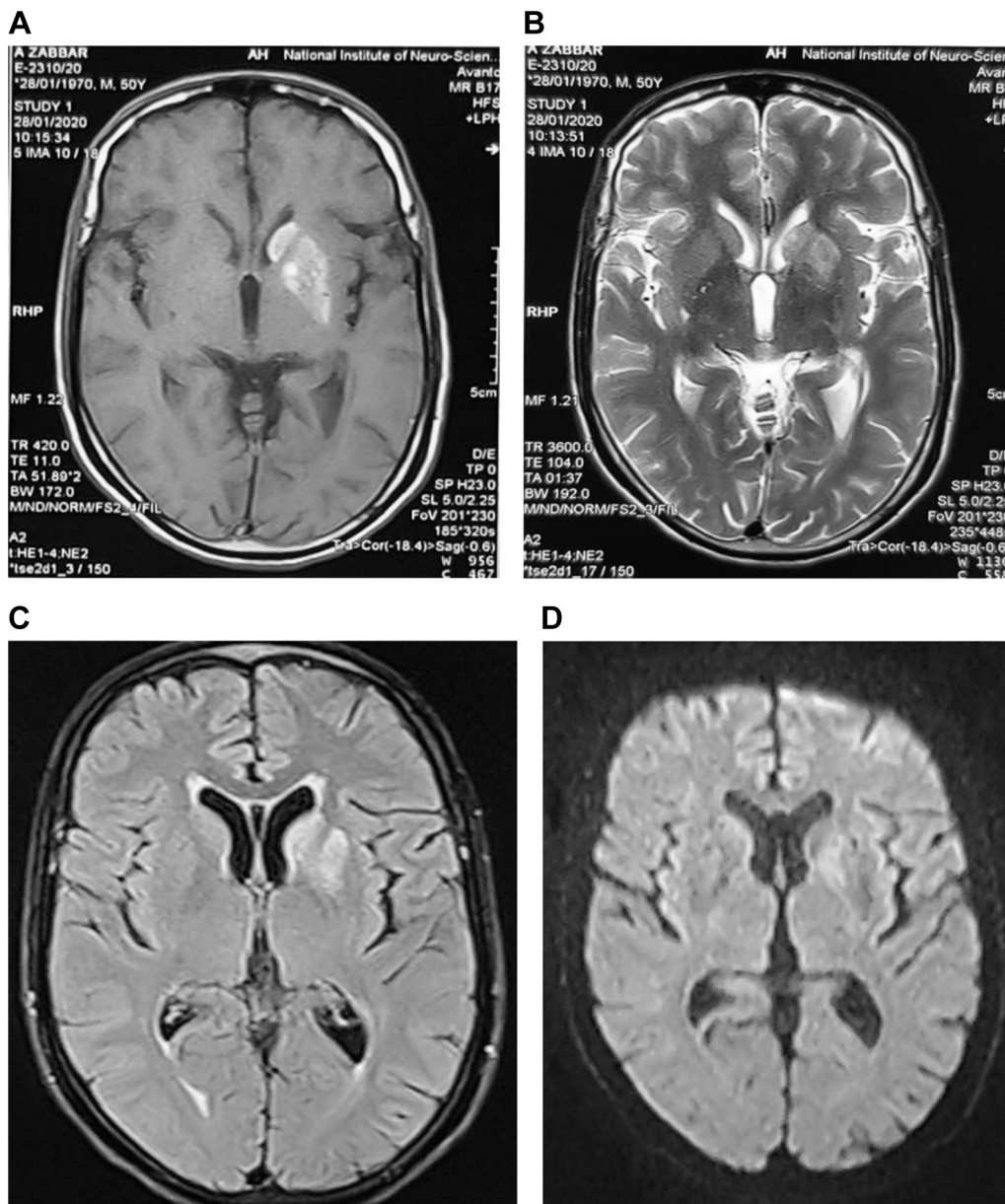
treatment and with proper control of hyperglycemia, the involuntary movements disappeared, and haloperidol-tetrabenazine was gradually withdrawn.

## Discussion

Ballism, chorea, and athetosis represent a spectrum of involuntary, hyperkinetic movement disorders that are often interrelated and may occur in the same patient. It is well known that chorea consists of involuntary, continual, abrupt, rapid, brief, unsustained, irregular movements that flow randomly from one part of the body to another part.<sup>6</sup> When choreic movements are more severe, assuming a flinging, flailing character, it is termed ballism.<sup>7</sup> As sudden onset hemichorea, hemiballism, or hemiathetosis appear to share a common anatomic location for the lesion, it was proposed that these movement disorders represent degrees of severity of the same pathology.<sup>3,8</sup> Therefore, the term HCHB is used to denote a hyperkinetic movement disorder seen in patients with acute or subacute onset of involuntary movements affecting one side of the body.<sup>3</sup>

HCHB is caused by focal lesions in the contralateral basal ganglia. Ischemic or hemorrhagic stroke represents the most common cause, followed by hyperglycemia. Rarer causes include encephalitis, vasculitis, CNS lupus, mass lesions, multiple sclerosis, thyrotoxicosis, and drugs (ie, anticonvulsants, levodopa, oral contraceptives, and neuroleptics).<sup>3</sup> The underlying mechanism is hypoactivity of the subthalamic nucleus and other basal ganglia nuclei, resulting in an abnormal and disorganized firing pattern of globus pallidus pars interna that causes alternative inhibition and disinhibition of motor thalamus and motor cortex, leading to ballism and chorea.<sup>8</sup>

When hyperglycemia is the underlying cause, as in our patient, HCHB usually occurs in the setting of nonketotic hyperglycemia—that is, in the hyperglycemic hyperosmolar state<sup>3,5,9,10</sup>; however, an association with diabetic ketoacidosis has also been reported.<sup>4</sup> HCHB may be the first manifestation of DM or may occur after years of poor glycemic control.<sup>1,10</sup> Hyperglycemia-induced decrease in regional blood flow may be responsible for such manifestations by causing reversible ischemia of the susceptible areas without infarction.<sup>11</sup> Other suggested mechanisms include hyperglycemia- and hyperosmolality-induced hyperviscosity resulting in ischemia and disruption of the blood-brain barrier, which in turn causes intracellular acidosis, regional metabolic failure, and depletion of the neuroinhibitory transmitter gamma aminobutyric acid.<sup>1</sup> Taken together, HCHB seems to be the result of alterations in blood flow and neurotransmitter activity in an area with preexisting structural or vascular abnormality, making it vulnerable to the effects of hyperglycemia. Genetic susceptibility has also been suggested by some authors.<sup>9</sup>



**Fig. 2.** MRI of the brain of the patient revealed (A) T1 hyperintense area at left basal ganglia (caudate and lentiform nuclei). B and C, T2 and FLAIR, respectively, revealing same lesion, which is iso- to hyperintense. D, DWI showing no restriction of diffusion. DWI = diffusion-weighted image; FLAIR = fluid-attenuated inversion recovery; MRI = magnetic resonance imaging.

Such a mechanism is also supported by the resolution of symptoms after correction of hyperglycemia.

Characteristic radiological features of hyperglycemia-induced HCHB include a hyperintense lesion in the contralateral basal ganglia region on T1-weighted MRI.<sup>5</sup> T2-weighted MRI may show variable signal change, ranging from hypo- to hyperintense to gray matter.<sup>12</sup> Brain imaging is necessary to exclude differential diagnoses in such a presentation.

**Conclusion**

Hyperglycemia was the underlying cause of HCHB in our patient, and the presentation makes other diagnoses unlikely. This case serves to reiterate the diversity of CNS manifestations that may result from hyperglycemia. Both neurologists and endocrinologists

should be aware of such presentations to be able to provide appropriate management.

**Disclosure**

The authors have no multiplicity of interest to disclose.

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