HHS Public Access

Author manuscript

Am J Med Case Rep. Author manuscript; available in PMC 2020 May 19.

Published in final edited form as:

Am J Med Case Rep. 2020 ; 8(6): 159–161.

Hemichorea-Hemiballismus as an Unusual Presentation of Hyperosmolar Hyperglycemic Syndrome

Javier Ticona,

Victoria Zaccone,

Unaiza Zaman,

Daniel Kashani,

Zachary Chung,

Isabel M. McFarlane*

Department of Internal Medicine, State University of New York, Downstate Medical Center, Brooklyn, NY 11203 USA

Abstract

Diabetes mellitus can lead to a diverse array of systemic complications. Poorly managed hyperglycemia can result in serious neurological consequences ranging from peripheral neuropathy to seizures and coma. A rare neurologic disorder seen in acute decompensated type 2 diabetes mellitus (T2DM) is hemichorea-hemiballismus (HCHB). HCHB is a movement disorder primarily associated with cerebrovascular accidents of infarct or hemorrhagic origin. It is a condition that can occur in a diabetic patient, especially when no other signs or symptoms of hyperglycemia are present. It is urgent to recognize HCHB movement disorder quickly as it may be the only presenting sign of hyperglycemia and can alert medical personnel to a possible hyperosmolar hyperglycemic state (HHS). We report an unusual case of HCHB in a patient with HHS, whose only presenting sign was unilateral hyperkinesis, which completely resolved after adequate blood glucose control. Prompt treatment and management of hyperglycemia yields an excellent prognosis in HCHB.

Keywords

type 2 diabetes mellitus; movement disorders; chorea; hemiballismus; hyperosmolar hyperglycemic syndrome; basal ganglia

1. Introduction

Chorea is a well-known movement disorder in which the neural connections between the basal ganglia and frontal motor areas are dysfunctional, leading to an uninhibited flow of involuntary, spontaneous muscle contractions. [1] Presence of hemi-ballistic movements may indicate pathology associated with underlying structural defects, metabolic/endocrine

*Corresponding author: Isabel.McFarlane@downstate.edu.

derangements, autoimmune/inflammatory conditions, infections or drug/toxin mediated damage [2]. Commonly, hemiballismus is associated with lacunar strokes. Rarely, HCHB presents as a sign of hyperglycemia, especially in elderly females with T2DM and a long-term, poor glycemic control [3,4]. HHS is a serious complication of poorly managed T2DM and typically presents as polyuria, polydipsia and glycosuria followed by a progressive decline in mentation and eventual coma. The diagnostic criteria for HHS includes glucose over 600 mg/dL, arterial pH over 7.30, low serum ketones and an elevated effective serum osmolarity over 320 mOsm/kg. Stupor and coma may equally be seen at even greater blood glucose levels over 1,000 mg/dL [5]. HHS is the second most common cause of acute hemiballismus and although described in the literature, hemiballismus is unusual to be the first and only presentation of HHS. We present an interesting case of HHS, where HCHB was the initial and only sign of uncontrolled hyperglycemia.

2. Case Report

A 71-year-old Black male with a past medical history of hypertension, chronic kidney disease and type II diabetes mellitus (T2DM) presented to our hospital after experiencing involuntary, continuous, sporadic movements of his right upper and right lower extremities for the previous four days. Vitals on admission were: BP 166/69, T: 98.5, HR: 64, RR: 18, and SaO2: 99 on RA. He was alert and oriented x 3. Cardiovascular and lung exams were unremarkable. Abdomen exam was significant for distention and suprapubic tenderness. Cranial nerves were intact, motor strength in the upper and lower extremities was 5/5 bilaterally, and reflexes were 2+ in the upper and lower extremities. Sensation was also intact bilaterally. The right upper and lower hyperkinetic episodes were spontaneous and lasted for 2-3 minutes. The patient's review of systems revealed a dry cough of few days duration, however there were no fevers, chills, dizziness, headaches, shortness of breath, chest pain, facial droop, limb weakness or numbness. Of note, patient had been voiding more frequently than usual. He stated that he awakes multiple times at night to void, but he feels "satisfied" once he finishes voiding. The patient had T2DM for many years, managed with metformin. However, he had recently ran out of metformin and had not taken it for a "few days". He has never experienced any movement disorders in the past. His home regimen consisted of chlorothiazide, atorvastatin and metformin. Family history was negative for movement disorders.

Laboratory data was significant for leukocytosis, chemistries were consistent with HHS and acute kidney injury on chronic kidney disease; hemoglobin A1C (HbA1C) was 8% (Table 1). EKG showed normal sinus rate & rhythm. A CXR demonstrated a 1.1 cm left upper pulmonary nodule with a differential diagnosis of malignancy and benign etiology. Computer tomography (CT) of the head showed chronic microvascular ischemia with normal gray-white matter differentiation with evidence of chronic microvascular ischemia; no evidence of hemorrhage, and no acute/abnormal intracranial findings (Figure 1A, B, C). The patient also had an elevated WBC on admission which was attributed to a possible URI due to a recent cough that he had experienced a few days earlier.

The patient received aggressive resuscitation with normal saline, regular insulin and calcium channel blockers for blood pressure control. The patient was also found to have urinary

obstruction, which was relieved with an indwelling urinary catheter. Normoglycemia and complete resolution of the hyperkinetic movements were achieved 24 hours after admission. The patient was monitored to ensure resolution of the AKI; the hyperkinetic movements did not reappear during the hospitalization.

3. Discussion

HCHB is a hyperkinetic disorder that results from damage to the basal ganglia regulatory pathways which leads to excessive dopamine activity. Chorea is a hyperkinetic movement disorder which is characterized by brief, random and irregular contractions. The movements can be involuntary and vary in speed and direction. Chorea has low-amplitude contractions, that usually affect the distal limbs. Less commonly, it can also affect the face and trunk. Hemiballism refers to ballism that is unilateral in nature. It is involuntary movements that more commonly affect proximal areas and have large-amplitude contractions (in contrast to chorea). HCHB typically manifests with a high signal intensity in the basal ganglia on a computed tomography (CT) or T1 weighted magnetic resonance imaging (MRI). HCHB can be seen in strokes affecting the circulation of the basal ganglia. [6] More specifically, the structures that can be involved in HCHB include the: putamen, caudate nucleus, thalamus, subthalamic nucleus, and their interconnecting pathways. A second common cause of HCHB has been associated with metabolic imbalances such as hyperglycemia in acute decompensated T2DM. It is postulated that the hyperosmotic state seen in HHS promotes breakdown of the blood-brain barrier, resulting in increased passage of white blood cells through the capillaries and formation of the subsequent lesion [7]. Other hypotheses suggest derangements in perfusion to the basal ganglia secondary to hyperglycemia which leads to ischemia of gamma-aminobutyric acid (GABA), the chief inhibitory compound in the central nervous system neurons (GABAergic) and uninhibited excitatory stimulation, resulting in consequent hyperkinesis. Furthermore, hyperglycemia has been shown to alter the cerebral metabolism and promote an anaerobic state, resulting in additional ischemia to GABAergic neurons [8]. It is possible that the patient’s inadequate blood glucose regimen, in addition to the use of thiazides resulting in volume contraction, could have precipitated HHS which in turn led to the development of hyperkinetic movements.

Our patient presented with acute hemiballismus, which completely resolved only after adequate blood glucose was achieved. Current literature suggest resolution of HCHB after several weeks, and sometimes even months after achieving blood glucose control [3]. Of note, the rapid resolution of HCHB can be attributed to the correction of an endocrine derangement, as opposed to a structural brain dysfunction, however delays in treatment can potentially lead to permanent brain damage [9,10]. In the elderly with T2DM, HCHB has been described as the presenting symptom of HHS and diabetes [8–12]. Furthermore, the classic basal ganglia hyperintensities were not appreciated on CT scan [11]. This may help to support the heterogeneous nature of this condition.

Awareness of HCHB is of paramount importance in the management of HHS. Prompt recognition allows a practitioner to diagnose HHS and manage quickly, preventing further decompensation and possible death. Recognition of this movement disorder also helps in ruling out intracranial pathology, ordering of unnecessary testing and preventing the use of

inefficient therapeutic regimens. Finally, presentation of HCHB without a history of diabetes warrant screening for T2DM, in order to prevent additional long-term complications.

Acknowledgements

This work is supported, in part, by the efforts of Dr. Moro O. Salifu M.D., M.P.H., M.B.A., M.A.C.P., Professor and Chairman of Medicine through NIH Grant number S21MD012474.

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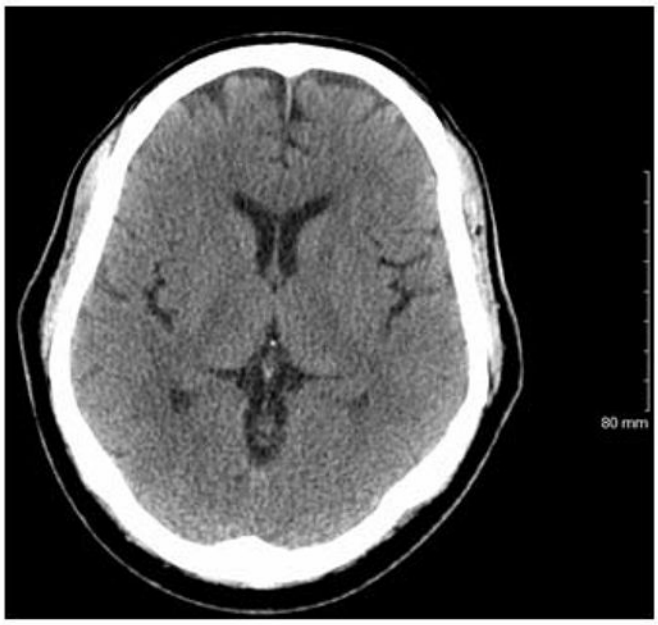

Figure 1A: Axial Basal Ganglia

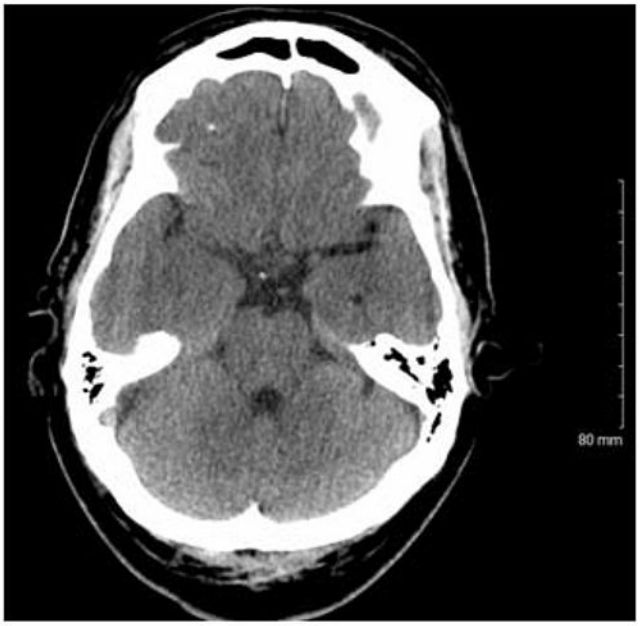

Figure 1B: Axial Substantia Nigra

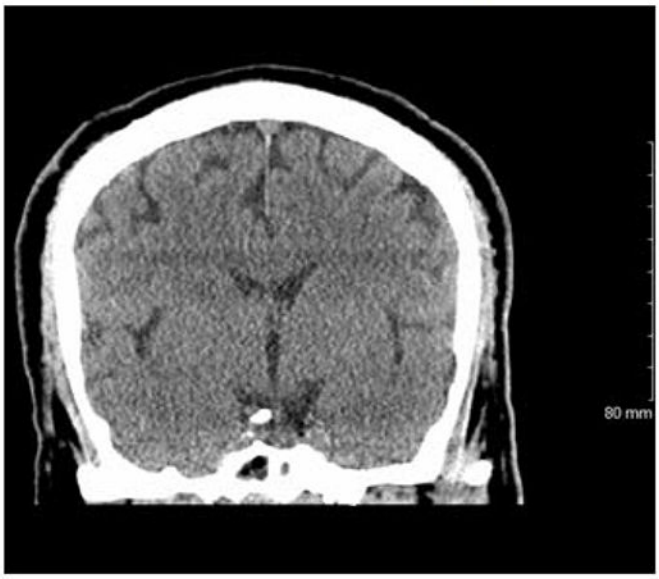

Figure 1C: Coronal Basal Ganglia

Figures 1.

Minimal atrophic changes consistent with age. Normal gray-white matter differentiation with some evidence of chronic microvascular ischemia. Normal lateral ventricles. Midline third and fourth ventricles. Basal cisterns patent. Normal basal ganglia and posterior fossa structures, no evidence of focal lesions in brainstem or cerebellum. No evidence of mass, hemorrhage or fracture

Table 1.

Laboratory Data

Serum	On admission	HD # 2	Reference Range
WBC (K/uL)	14.68	11.95	4.5-10.9
RBC (M/uL)	3.12	3.39	4.2-5.4
Hemoglobin (g/dL)	9.6	10.7	12.0-16.0
Hematocrit (%)	26.9	30.5	37.0-47.0
Platelets (K/uL)	260	317	130-400
Sodium (mmol/L)	141	142	136-146
Potassium (mmol/L)	5.2	4.8	3.5-5.0
Chloride (mmol/L)	103	103	98-106
CO2 (mmol/L)	21	26	24-30
BUN (mg/dL)	53	28	6-20
Creatinine (mg/dL)	4.4	2.5	0.4-1.2
Calcium (mg/dL)	9.3	10.6	8.4-10.3
Total Protein (g/dL)	7.0	7.7	6.0-8.5
Albumin (g/dL)	4.0	4.0	2.8-5.7
AST (U/L)	34	29	10-35
ALT (U/L)	46	38	0-31
Alk. Phos (U/L)	117	115	25-125
Total Bilirubin	0.4	0.93	0.0-1.2
Glucose (mg/dL)	545	146	70-99
Serum Osmolality (mOsm/kg)	331.2	302.1	285-295