Chorea Hyperglycemia Basal Ganglia Syndrome: A Case Report of a Rare Diabetes Complication

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Abstract

Introduction. We present a patient with long-standing uncontrolled type 2 diabetes mellitus (T2 DM) who developed sudden onset of choreiform movement, which rapidly resolved after insulin therapy and haloperidol.

Case Description. A 53-year-old Filipino male, with T2DM and hypertension for more than 10 years, presented with sudden onset of hyperkinetic, involuntary, non-patterned, continuous movements of the left upper and lower extremities. Investigations revealed severe hyperglycemia without acidemia and ketonuria. Cranial computed tomography scan showed hyperdensity on the right caudate and lentiform nuclei. On cranial magnetic resonance imaging, there was T1-weighted hyperintense and T2 - weighted hypointense signal involving the right putamen, globus pallidus and caudate. Cranial magnetic resonance angiography showed stenosis on the cavernous segment of the right internal carotid artery (ICA), left ICA and middle cerebral artery (MCA) junction, the A1 segment of the left anterior communicating artery and proximal P2 segments of the bilateral posterior cerebral arteries. The patient was managed with a basal-bolus insulin regimen to control the blood glucose and haloperidol to manage the extrapyramidal symptoms. Consequently, there was complete resolution of the involuntary movements.

Conclusion. This case illustrates the importance of a systematic approach to movement disorders and early recognition of this rare diabetes complication known as chorea hyperglycemia basal ganglia syndrome or diabetic striatopathy.

Keywords. Diabetic striatopathy, movement disorders, diabetes complication, chorea hyperglycemia basal ganglia syndrome

Introduction

Chorea hyperglycemia basal ganglia syndrome (CHBG) otherwise known as diabetic striatopathy (DS) is characterized by unilateral involuntary choreiform movements, non-ketotic hyperglycemia, and contralateral basal ganglia hyperintensity on T1-weighted magnetic resonance imaging (MRI) or high density on computed tomography (CT) scans. CHBG is more frequently seen among Asians and elderly women with poor blood glucose control.[1] The actual prevalence is thought to be underestimated because it is frequently misdiagnosed as the more common intracerebral hemorrhage. Compared with other diabetes complications, an excellent prognosis can be achieved through prompt recognition of this movement disorder and correction of hyperglycemia and anti-chorea drugs.[2] Hence, the purpose of this report is to present a rare sequalae of acute hyperglycemia, discuss the approach to a patient presenting with dyskinesia, and its pathophysiology secondary to hyperglycemia.

Case Presentation

A 53-year-old-male, Filipino, right-handed, presented at the emergency room with a chief complaint of involuntary movement of left upper and lower extremities. His symptoms had been present for six days, which started as twitching of his fingers on his left hand. This gradually progressed to involve the left upper and lower extremities, described as involuntary, irregular, purposeless, and non-rhythmic movements. It was noted that the said movements disappear while patient was asleep. The condition did not affect his vision, face, trunk, motor strength nor sensation of his limbs. Review of systems revealed no loss of consciousness, dizziness, chest pain, frothy urine, weakness nor numbness.

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Table I. Laboratory Results of the Patient on Admission

Blood Chemistry			Arterial Blood Gas			Urinalysis		
Parameter	Patient's Result	Reference Range	Parameter	Patient's Result	Reference Range	Parameter	Patient's Result	Reference Range
CBG	328		pH	7.54	7.35 – 7.45	Glucose	2+	(-)
HbA1c	15.4%	4.5 – 6.5	pCO_2	26	35-45	Ketones	Negative	(-)
BUN	6.8	3.2 – 7.4 mmol/L	pO_2	187	80-100	Albumin	3+	(-)
Crea	148	64 -110 umol/L	HCO_3	22.2	22-26	WBC	3.8	0-2
Na	137	136 -145 mmol/L	Oxygen Saturation	100%		RBC	0.5	0-2
K	3.7	3.5 – 5.1 mmol/L	FiO_2	Room Air		Bacteria	21.4	0-2
iCa	1.11	1.05 – 1.3 mmol/L	Interpretation: Uncompensated respiratory alkalosis with more than adequate oxygenation					
Mg	0.81	0.85 – 1.10 mmol/L						

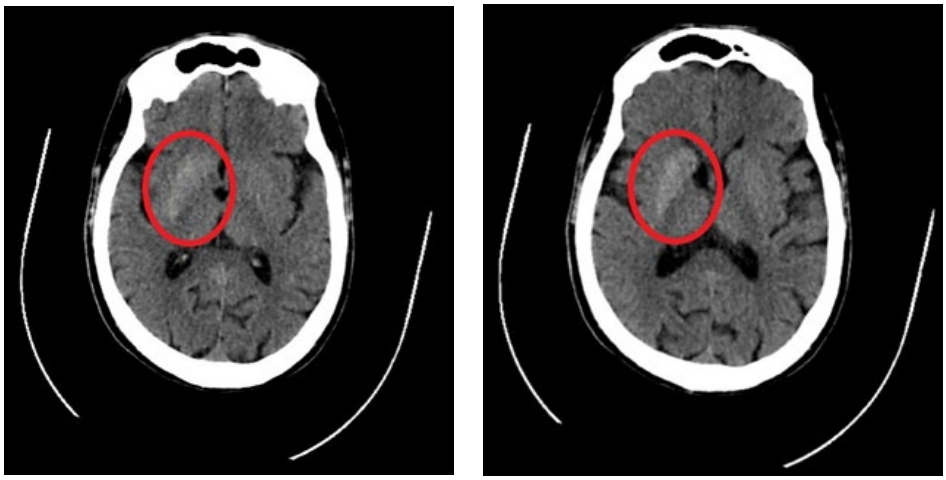

Figure 1. Plain Cranial CT scan of the patient showing asymmetrically hyperdense caudate nuclei and lentiform nuclei, appearing more hyperdense on the right, suggestive of systemic metabolic abnormalities such as non-ketotic hyperglycemia.

In 2002, he suffered from cerebral infarction, Modified Rankin Scale (mRS) = 0, and was diagnosed with T2DM and hypertension. In 2011, he underwent below-knee amputation (BKA) of the right leg and was likewise diagnosed to have chronic kidney disease (CKD) secondary to diabetic kidney disease (DKD). In 2020, because of the pandemic, he was not able to have regular consultations and was not compliant with his maintenance medications, which included metformin 500 mg tablet once a day and amlodipine 5mg tablet once a day. He does not drink alcoholic beverages, nor does he smoke.

The systemic physical examination was unremarkable except for BKA, with right leg with prosthesis. Vital signs were stable. On neurological examination, the patient had Glasgow Coma Scale of 15, with intact cerebral functions and no cranial nerve deficits. His motor

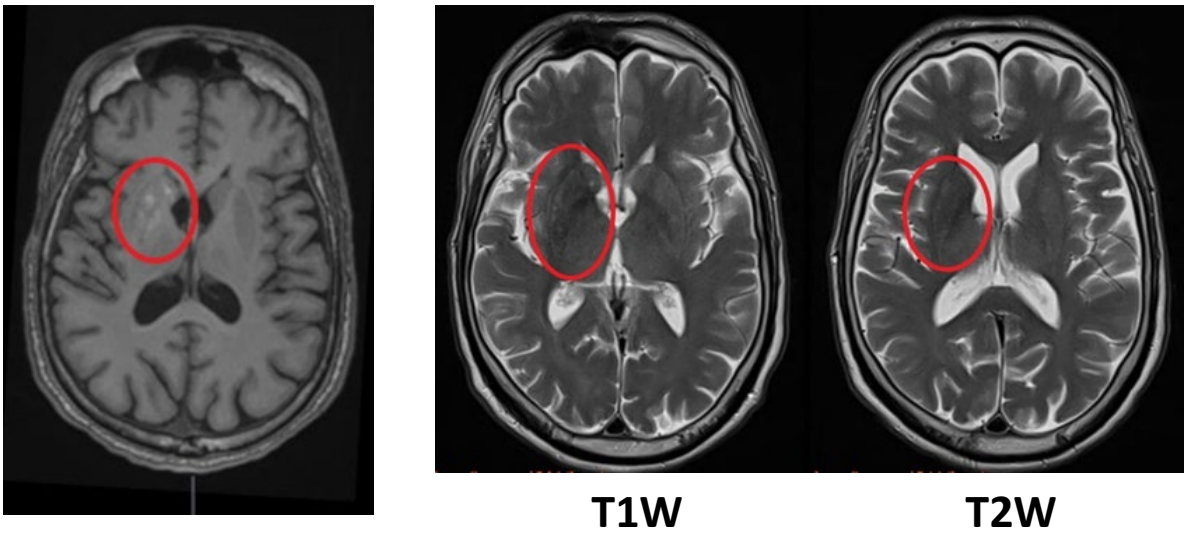

Figure 2. Plain Cranial MRI of the patient showing T1W hyperintense, T2W hypointense involving the right putamen, globus pallidus, and caudate nucleus, seen in cases of diabetic hyperglycemia

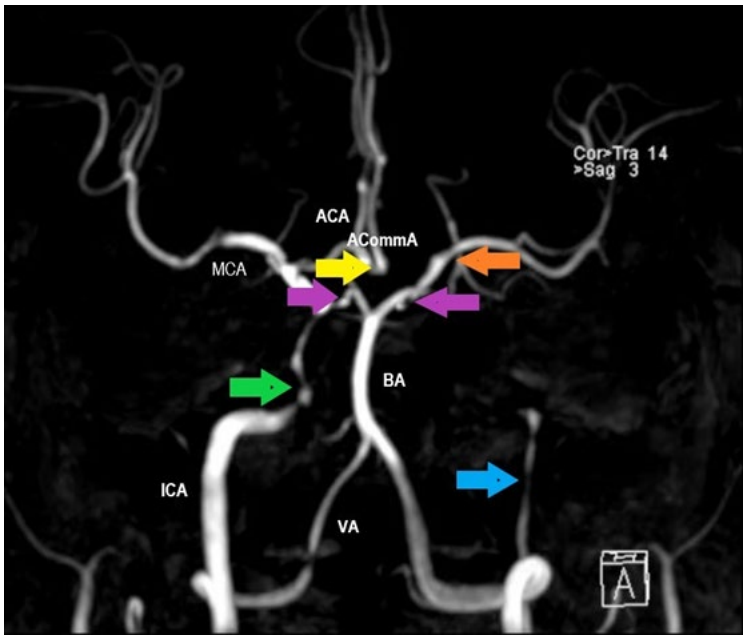

Figure 3. Cranial Magnetic Resonance Angiography showing stenosis on the cavernous segment of the right internal carotid artery (ICA) (green arrow). There was also noted stenosis involving the left ICA (blue arrow) and MCA junction (orange arrow), the A1 segment of the left anterior communicating artery (ACA) (yellow arrow) and proximal P2 segments of the bilateral posterior cerebral arteries (PCA) (purple arrow).

strength was 5/5 on all extremities with full and intact sensation. Reflexes were normal, no long tract sign, nor meningeal signs. However, there were involuntary, irregular, purposeless, non-rhythmic and non-suppressible movements of the left upper and lower extremities.

Laboratory examinations, as shown in *Table I*, revealed elevated capillary blood glucose at 328 mg/dL and glycosylated hemoglobin (HbA1c) of 15.4%. Arterial blood gas showed respiratory alkalosis. Urine ketone was negative. Serum creatinine was elevated at 148 umol/L with blood urea nitrogen level at 6.80 mmol/L. The computed eGFR was 45.9 mL/min/1.73 m^2.

A cranial CT scan was requested as shown in *Figure 1*. It revealed a hyperdensity on the right caudate and lentiform nuclei. A follow up cranial MRI is presented in *Figure 2* which showed T1-weighted hyperintense signal, and corresponding T2-weighted hypointense signal

involving the right putamen, globus pallidus, and caudate nucleus. Cranial magnetic resonance angiogram (MRA) was also done as shown in *Figure 3*. There was stenosis on the cavernous segment of the right internal carotid artery (ICA), left ICA, and middle MCA junction, the A1 segment of the left ACA and proximal P2 segments of the bilateral PCA.

The patient was then admitted as a case of CHBG, hyperglycemic crisis secondary to T2DM, uncontrolled, hypertensive atherosclerotic cardiovascular disease, CKD stage IIIB secondary to DKD, status post cerebrovascular disease mRS 0, status-post BKA, right leg. He was started on basal-bolus insulin regimen to control the hyperglycemia and haloperidol for the dyskinesia. There was total resolution of his involuntary movements on the fourth day of hospitalization.

Discussion

We report a patient with long standing uncontrolled T2DM who presented with a classic clinical presentation of CHBG which includes the triad of acute hemichorea, severe hyperglycemia in the absence of keto-acidosis, and a T1 hyperintensity signal in the contralateral basal ganglia on neuroimaging. The prevalence of this condition has been reported to be 1 in 100,000 which is believed to be underestimated because it could be misdiagnosed as the more common intracerebral hemorrhage.[2] The condition is more prevalent among Asians, particularly elderly women.[9] Our patient, however, was male and was relatively young.

In patients with movement disorders, a systematic approach is recommended to establish the correct diagnosis. Four key questions need to be addressed: (1) Which type of movement disorder is present, (2) What the dominant movement disorder is, (3) What the associated features are, including both neurologic and non-neurologic features, and lastly, (4) What the differential diagnosis is.[5]

Our patient manifested with involuntary choreic movement of the left limbs, which was a manifesting symptom of a disorder in the extrapyramidal system located in the right basal ganglia.[4] CHBG is classified as a movement disorder emergency defined as any movement disorder which evolves over hour to days and failure to appropriately diagnose and manage can result in mortality or morbidity.[3]

After clinical localization, cranial CT scan and MRI were done showing hyperdensities and T1 weighted hyperintense signals in the right basal ganglia, respectively. These imaging findings are often mistaken with hemorrhages. However, the absence of mass effect and the sparing of the internal capsule, as observed in our patient, is consistent with CHBG.[2] These neuroimaging findings are reversible upon correction of the underlying cause. Unfortunately, a repeat MRI was not performed in our patient.

The MRA findings seen in our patient was also observed by Nagai et al. where perivascular signal intensities were found along with stenosis of the MCA in a patient with CHBG.[6] These lesions have vascular fragility and are vulnerable to ischemia and metabolic disorders associated with hyperglycemia. The lesion can be secondary to a variety of diseases including genetic, neurodegenerative, metabolic, infectious, autoimmune, drugs, and vascular disorders among others.[4] These disorders were carefully ruled out in our patient due to the absence of family history of Huntington's disease and no intake of drugs. With the history and initial work-up available, hyperglycemia was thought to be the most likely cause.

The basal ganglia is vulnerable to various metabolic abnormalities. Physiological high activity with high glucose and oxygen uptake, rich vascular supply, large number of mitochondria and chemical substances make the basal ganglia very sensitive to metabolic, chemical, or ischemic trauma.[7] The exact pathophysiology of CHBG is still unclear but there are several theories.[1,6,9,10] According to the ischemic injury/ion deposition theory, hyperglycemia leads to hyperviscoscity, diabetic vasculopathy, and cytotoxic edema. The ischemic injury leads to the proliferation of astrocytes and the expression of zinc-friendly metalloproteins which would explain the high signal in T1-weighted MR images.[1,9]

By achieving control of blood glucose levels and using dopamine receptor antagonists such as haloperidol, most patients with DS have a good prognosis. In a review by Chua et al., successful treatment rates of chorea with glucose-control only was 25.7% while glucose control and additional dopamine receptor antagonists was 76.2%. Haloperidol is the most used dopamine receptor antagonist.[2]

The clinical symptoms of our patient were significantly improved with the control of blood glucose and the use of haloperidol. Aside from CHBG being an acute complication of non-ketotic hyperglycemia, our patient also manifested with chronic complications of diabetes. He had macrovascular complications which resulted to stroke and to amputation, and developed microvascular complication leading to diabetic kidney disease. Hence, diabetes education and compliance to medications are also very important to prevent further complications.

Conclusion

CHBG is a rare complication of T2DM. A patient with hemichorea may have a lesion in the contralateral basal ganglia but this may be due to numerous etiologies. The presence of non-ketotic hyperglycemia and T1-weighted hyperintense signal on cranial MRI makes the diagnosis of CHBG likely. Aggressive control of blood sugar and anti-chorea drug such as haloperidol are the mainstays of treatment. Early recognition and management of this movement disorder emergency is essential to avoid morbidity and mortality.

Conflict of Interest. The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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