Neurology Asia 2022; 27(3) : 799 – 802

Hemichorea-hemiballism in a diabetic patient with euglycemia at presentation

Somdattaa Ray *DM*, Vikram V Kamath *DM*, KN Rajesh *DM*

Department of Neurology, Trustwell Hospital, Bangalore, Karnataka, India

Abstract

Hemichorea hemiballismus syndrome (HCHB) is commonly seen in an acute setting of non ketotic hyperglycaemia. Hemichorea hemiballismus has been rarely reported in the setting of normal blood sugars in a diabetic patient. We report an 84-year-old lady who initially presented with uncontrolled diabetes and acute left hemiparesis. Imaging did not reveal any evidence of recent infarction. After sugars were normalised, five weeks later she presented with involuntary movements of left upper and lower limb. On examination, patient had left hemichorea hemiballismus. Repeat MRI brain showed right putamen hyperintensity in T1 weighted images. Venous blood glucose was 123 mg/dl (6.8 mmol/L). Patient improved significantly with haloperidol and clonazepam. In conclusion, hemichorea hemiballismus can be observed in a patient with previously uncontrolled diabetes mellitus after normalization of blood sugars akin to that seen in patients with HCHB associated with hyperglycaemia.

Keywords: Diabetes, normoglycemia, hemichoreaballism, hemiparesis

INTRODUCTION

Hemichorea hemiballismus (HCHB) syndrome is a well-known clinical entity commonly seen in the setting of uncontrolled diabetes. This syndrome has been classically associated with the acute setting of a hyperglycaemic hyperosmolar state and to a lesser extent with diabetic ketoacidosis. The diagnosis is associated with T1 hyperintensity of unilateral putamen and rarely bilateral putamen on magnetic resonance imaging (MRI) and hyperdensity of putamen in CT brain.[1]

HCHB has been rarely reported in the setting of normal blood sugars in a diabetic patient.[2,3] We report a patient with type 2 diabetes mellitus with previously uncontrolled sugars who presented with HCHB and had normal blood glucose at presentation.

CASE REPORT

An 84-year-old hypertensive lady initially presented to another hospital with sudden onset of left upper and lower limb weakness that improved spontaneously after 6 hours. Patient was then diagnosed with diabetes mellitus with a blood glucose of 495 mg/dl (27.5 mmol/L) at admission. Her serum sodium was 118 mEq/L and urine for ketone bodies was negative. MRI brain done then showed bilateral periventricular and fronto-parietal white matter hyperintensities with normal basal ganglia, thalamus and brainstem imaging as well as absence of diffusion restriction in diffusion weighted imaging. There was no acute infarction in the scan. With insulin infusion, her sugars were brought down to 340 mg/dl (18.9 mmol/L) over 2 days that was maintained below 150 mg/dl (8.3 mmol/L) thereafter with oral hypoglycaemic agents. Five weeks later, she presented to our emergency department with involuntary flinging movements of left upper and lower limbs of two day duration. On examination, patient had left hemichorea-ballismus. A possible etiology of hyperglycaemia or basal ganglia stroke was considered.

Further history revealed that on the day of onset of involuntary movements, CBG checked at home was 122 mg/dl (6.8 mmol/L). Blood investigations revealed elevated HbA1c-10.49, while venous blood glucose was 123 mg/dl (6.8 mmol/L). Urine for ketone bodies was negative.

MRI brain without contrast revealed T1 hyperintensity (figure) and T2 iso to hypointensity of the right putamen. Susceptibility weighted images were normal thereby, ruling out calcification or bleed as the cause of T1 hyperintensity. After starting oral haloperidol 0.5 mg and clonazepam 0.25 mg at three times per day each, the patient reported complete resolution of left upper limb symptoms and partial improvement of left lower limb choreo-ballismus.

Address correspondence to: Somdattaa Ray, Department of Neurology, Trustwell hospitals, JC road, Bangalore, Karnataka- 560002, India. Tel: +9180-45666773, Email: somray227@gmail.com

Date of Submission: 27 April 2022; Date of Acceptance: 11 June 2022

https://doi.org/10.54029/2022aus

DISCUSSION

We report a case of severe HCHB who had euglycemia at onset. This is uncommon and has been reported rarely earlier.[2-4] Other unusual presentations of HCHB include ipsilateral basal ganglia abnormalities in imaging[5] and prolonged chorea in patient with hyperglycemia that persisted long after normalization of blood glucose levels.[6] In patients with HCHB with hyperglycaemia, hyperglycemia results in anaerobic metabolism in the brain, thereby leading to depletion of GABA, an inhibitory neurotransmitter. This might lead to disinhibition of subthalamic nucleus, giving rise to the dyskinesia. Hyperviscosity and ischemia of the basal ganglia are other implicated pathogenetic mechanisms.[7,8]

The cause of chorea following attainment of euglycaemia in previously uncontrolled diabetes remains elusive. Most of the patients with HCHB and normal glucose levels at presentation had a history of recent uncontrolled diabetes, as evident by markedly elevated glycated hemoglobin similar to our patient (Table 1). Abe *et al*. postulated that rapid lowering of blood glucose levels might paradoxically produce a detrimental effect on the striatum with pre-existing diabetic striatopathy.[2]

In our patient, acute hemiparesis on the side which further went on to develop HCHB has been described earlier and gives an interesting insight.[2,7] Although, initial basal ganglia imaging done few hours after onset of hemiparesis did not reveal any evidence of basal ganglia ischemia in this patient, it is possible that an ischemic attack was the harbinger of overt basal ganglia dysfunction later as seen with subsequent appearance of HCHB in our patient. Uncontrolled diabetes induces hyperosmotic hyperglycemic state in the striatum[9] that achieves a state of homeostasis when the sugars remain uncontrolled for a long time. This equlibirum gets decompensated following intensive or rapid lowering of blood glucose, resulting in a state of relative glucose deprivation causing sudden energy depletion in the striatum, similar to what is seen in chorea following hypoglycaemia.[9] The paucity of collaterals supplying the external globus pallidus and striatum predispose these nuclei to energy crisis triggered by rapid blood glucose changes leading to worsening of microvascular complications, in patient with microangiopathy in diabetes mellitus.[10,11] Rapid blood glucose control has been reported to trigger diabetic uremic striato pallidal syndrome.[10] The presence of deranged

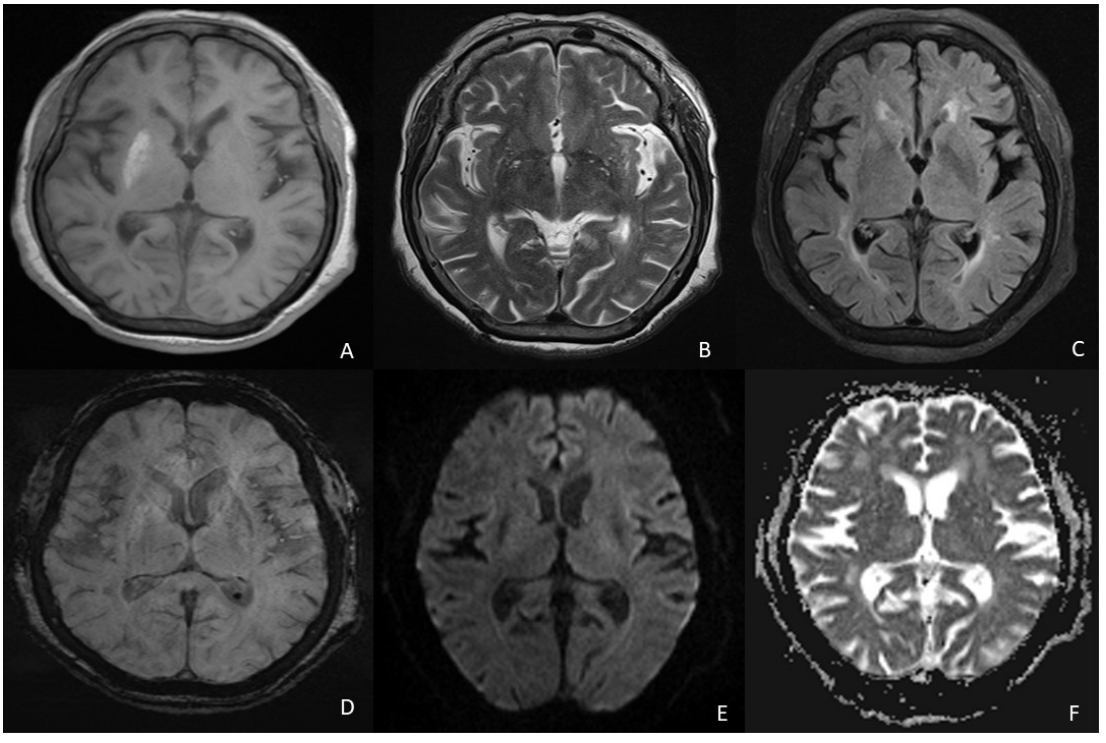

Figure 1. MRI brain of the patient (T1 weighted image-A) shows right putaminal hyperintensity. T2 weighted sequence(B), FLAIR (C) shows periventricular hyperintensity. Susceptibility weighted imaging (D), diffusion weighted imaging (E), and ADC(F) are normal

Table 1: A summary of reports of hemichorea hemiballismus with euglycemia at presentation

Author	Clinical details	Blood glucose correction	HbA1c at presentation for chorea
Abe *et al*., 2009[2]	Patient 1- 73 year old man presented with numbness of right upper and lower limb. Started on oral hypoglycaemic agents in view of HbA1c- 18.9. Two weeks later, developed right hemichorea	At presentation with hemichorea, fasting blood glucose was 151 mg/dl.	17.2%
	Patient 2- 84 year old lady presented with transient weakness of right hand. She was started on OHA since HbA1C – 12.3. Five weeks later patient developed right upper limb ballism.	At presentation, fasting blood glucose was 105 mg/dl and HbA1c dropped from 12.3 to 7.4%.	7.4%
Bizet *et al*., 2014[3]	66 year old lady presented with history of non ketotic hyperglycemia (RBS- 984 mg/dl). Three months later, she developed right sided hemichorea ballismus	984 mg/dl to 84 mg/dl over three months	12.2% (average serum glucose was 294 mg/dl)
Cho *et al*., 2018[4]	A 70 year old gentleman presented with dizziness, RBS was 415 mg/dl. Two days after lowering the blood glucose, patient developed left sided hemichorea	RBS corrected from 415 m/dl to 168 mg/dl	19%

RBS- random blood glucose, HbA1C- glycated hemoglobin

sorbitol pathway as evident by myoinositol peak on MR spectroscopy and decreased regional glucose uptake in the affected striatum in PET studies indicates failure of regional metabolic autoregulatory mechanisms rather than just microcirculatory derangement.[8]

In conclusion, HCHB can be rarely observed in a patient with previously uncontrolled diabetes mellitus after normalization of blood sugars, similar to that seen in patients with HCHB associated with hyperglycaemia. An association could be plausible between recent history of elevated blood sugars and hemiparesis as well as rapid blood glucose control with subsequent development of HCHB. However, this hypothesis requires further investigation.

ACKNOWLEDGEMENT

We would like to thank the patient and her family for their co-operation

DISCLOSURE

Ethics: Informed consent obtained from the patient for the study and publication.

Financial support: None

Conflict of interest: None

REFERENCES

1. Oh SH, Lee KY, Im JH, Lee MS. Chorea associated with non-ketotic hyperglycemia and hyperintensity basal ganglia lesion on T1-weighted brain MRI study: a meta-analysis of 53 cases including four present cases. *J Neurol Sci* 2002;200(1-2):57-62. https://doi.org/10.1016/S0022-510X(02)00133-8
2. Abe Y, Yamamoto T, Soeda T, *et al*. Diabetic striatal disease: clinical presentation, neuroimaging, and pathology. *Intern Med* 2009;48(13):1135-41. https://doi.org/10.2169/internalmedicine.48.1996
3. Bizet J, Cooper CJ, Quansah R, *et al*. Chorea, hyperglycemia, basal ganglia syndrome (CH-BG) in an uncontrolled diabetic patient with normal glucose levels on presentation. *Am J Case Rep* 2014;15:143. doi: 10.12659/AJCR.890179.
4. Cho H-S, Hong C-T, Chan L. Hemichorea after hyperglycemia correction: A case report and a short review of hyperglycemia-related hemichorea at the euglycemic state. *Medicine* 2018;97(10). doi: 10.1097/MD.0000000000010076.
5. Fong SL, Tan AH, Lau KF, Ramli N, Lim SY. Hyperglycemia-associated hemichorea-hemiballismus with predominant ipsilateral putaminal

abnormality on neuroimaging. *J Mov Disord* 2019;12(3):187. doi: 10.14802/jmd.19014.
6. Ahlskog JE, Nishino H, Evidente VG, *et al*. Persistent chorea triggered by hyperglycemic crisis in diabetics. *Mov Disord* 2001;16(5):890-8. https://doi.org/10.1002/mds.1171
7. Taboada GF, Lima GA, Castro JE, Liberato B . Dyskinesia associated with hyperglycemia and basal ganglia hyperintensity: report of a rare diabetic complication. *Metab Brain Dis* 2013;28(1):107-10. https://doi.org/10.1007/s11011-012-9357-z
8. Hsu JL, Wang H-C, Hsu W-C. Hyperglycemia-induced unilateral basal ganglion lesions with and without hemichorea A PET study. *J Neurol* 2004;251(12):1486-90. https://doi.org/10.1007/s00415-004-0571-4
9. Paraskevas GP, Vlachos GS, Vassilopoulou S, *et al*. Hypoglycemia-induced hemichorea in a patient with Fahr's syndrome. *Neurol Sci* 2012;33(6):1397-9. https://doi.org/10.1007/s10072-012-1096-8
10. Feekes JA, Cassell MD. The vascular supply of the functional compartments of the human striatum. *Brain* 2006;129(8):2189-201. https://doi.org/10.1093/brain/awl158
11. Lizarraga KJ, Adams D, Post MJ, Skyler J, Singer C. Neurovascular uncoupling after rapid glycemic control as a trigger of the diabetic-uremic striatopallidal syndrome. *Parkinsonism Related Disord* 2017;39:89-90. https://doi.org/10.1016/j.parkreldis.2017.03.010