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Case Report

Internal Medicine Section

Hemichorea Hyperglycaemia Basal Ganglia Syndrome in a Patient with Type 1 Diabetes Mellitus: A Rare Case Report

SWATHI GURURSAMY[1], KALAIMANI SIVAMANI[2], SELVARAJ PITCHAI[3], AMEYA S CHOGLE[4]

ABSTRACT

Hyperglycaemia induced involuntary movements are uncommon in clinical practice though Chorea Hyperglycaemia Basal Ganglia syndrome (CHBG) or Non-ketotic Hyperglycaemic Hemichorea (NHH) is being reported with increasing frequency due to the clinical awareness and widespread availability of neuroimaging. Prompt recognition of CHBG is essential, since correction of hyperglycaemia usually leads to early resolution of the involuntary movements. It is usually seen in elderly patients with uncontrolled Type 2 diabetes mellitus who present acutely with hemichorea or hemiballismus. It is rarely reported in Type 1 diabetes mellitus. Here, the author presents a 25-year-old male patient diagnosed with Type 1 diabetes mellitus with persistent chorea involving the left upper and lower extremity in whom the movement disorder persisted despite correction of hyperglycaemia.

Keywords: Blood glucose, Dyskinesia, Movement disorders

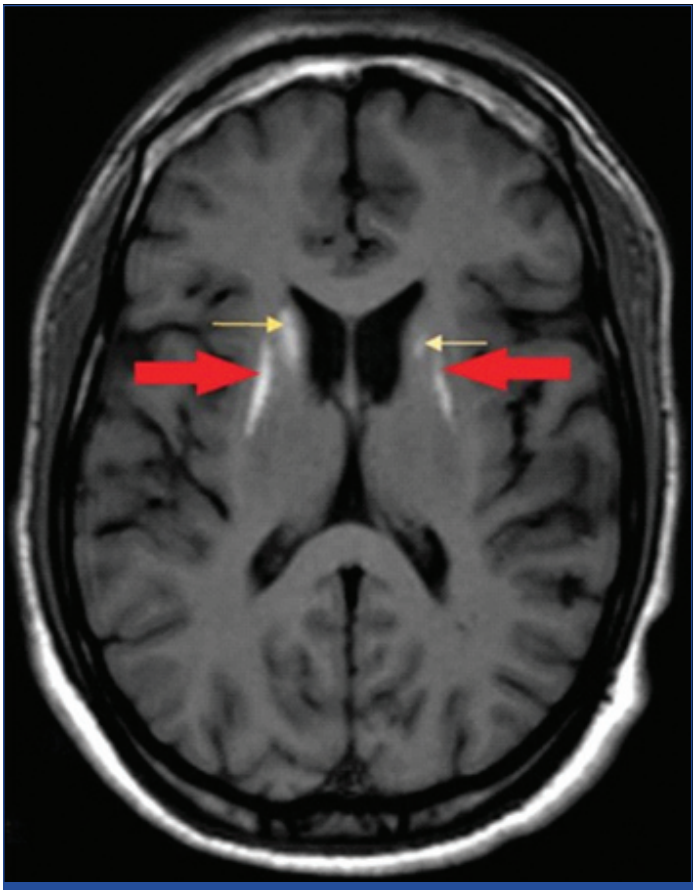
CASE REPORT

A 25-year-old male, diagnosed with Type 1 diabetes mellitus since 5 years of age, on irregular insulin therapy, presented with involuntary movements of left upper limb and lower limb on and off for the past 4 years. The diagnosis of Type 1 diabetes was based on his age of onset, though serological tests for anti-Glutamic Acid Decarboxylase (GAD) and other autoantibodies were not done.

The involuntary movements started in the left upper limb and left lower limb which were brief and jerky. It was not present during sleep. There was no history of fever, chest pain and joint pain. There was no history of cardiac illness. There was no family history of chorea. Patient was found to have blood sugar levels above 500 mg/dL when the symptoms first started 4 years back though he did not adhere to the prescribed dose of insulin and the involuntary movements continued for the subsequent years before his current presentation.

General physical examination revealed diffuse lipomatosis. His vitals were stable. Neurological examination revealed normal higher mental functions, normal tone, power of 5/5 in all 4 limbs and normal deep tendon and superficial reflexes. There were brief involuntary jerky movements involving left upper limb and occasionally in left lower limb. The investigations revealed fasting blood sugar of 600 mg/dL, post-prandial blood sugar of 718 mg/dL, HbA1c of 14.7%, urine ketones were negative. His cardiac, renal and liver parameters were within normal limits. Fundus examination revealed mild nonproliferative diabetic retinopathy in the right eye and slit lamp examination did not reveal any Kayser-Fleischer (KF) ring. A 2D echocardiogram was found to be normal. Antinuclear antibody and Human Immunodeficiency Virus (HIV) tests were negative. His thyroid profile was normal. MRI brain was done which showed bilateral symmetrical hyperintense signals at both globus pallidi and caudate nuclei on T1 weighted imaging [Table/Fig-1] and no restriction on Diffusion Weighted Imaging (DWI). He was admitted for glycaemic control with a diagnosis of hemichorea hyperglycaemia basal ganglia syndrome.

Blood sugar levels were controlled with biphasic isophane insulin injection (30% regular insulin and 70% isophane), the dose of which was titrated according to blood sugar levels. He was given 25 units of pre-mixed insulin in the morning and 15 units at night and regular monitoring of Capillary Blood Glucose (CBG) pre- and post-meals was done.



[Table/Fig-1]: T1 weighed MRI of Brain showing bilateral symmetrical hyperintensities at both globus pallidi (red arrows) and caudate nuclei (yellow arrows).

After 3 days of admission all his random blood sugar values were below 200 mg/dL. There was a decrease in the frequency of the involuntary movements, but it persisted. Hence, he was started on T. Quetiapine 25 mg HS as advised by neurologist. The patient was followed-up after one month and there was reduction in choreic movements but he reported that it increased whenever he skipped his insulin doses. A repeat MRI was not taken during follow-up as there was no complete resolution of the symptoms.

DISCUSSION

Chorea Hyperglycaemia Basal Ganglia syndrome (CHBG) is a rare condition that occurs in uncontrolled Type 2 diabetes mellitus and characterised by hemichorea/hemiballismus with non-ketotic hyperglycaemia. CHBG is pre-dominantly seen in women especially in Asian women indicating genetic predisposition [1-3]. Only few cases of hyperglycaemia induced chorea have been reported in Type 1 diabetes mellitus with one of the patients having ketotic hyperglycaemia and the remaining having non-ketotic hyperglycaemic chorea [4-9]. The index case had non-ketotic hyperglycaemia.

The exact aetiopathogenesis of CHBG syndrome is incompletely understood, with multiple theories suggesting impaired cerebral autoregulatory mechanism, activation of anerobic metabolism and depletion in the GABAergic neurons and reduction in acetyl choline synthesis leading to dysfunction of the striatum producing the clinical manifestations of this syndrome. Histological findings may suggest neuronal loss/atrophy within corpus striatum [10,11].

Hemichorea is a hyperkinetic movement disorder due to lesions of contralateral striatum [12]. There are various causes like haemorrhagic or ischemic stroke, neoplasm, systemic lupus erythematosus, hyperosmolar hyperglycaemic non-ketotic syndrome, Huntington's disease, Sydenham's chorea, HIV, hyperthyroidism, trauma and drug toxicity. CHBG can be seen both in patients with long standing uncontrolled diabetes mellitus as well as the initial manifestation of hyperglycaemia leading to the diagnosis of diabetes mellitus [13-16]. This patient was considered to have CHBG syndrome after excluding other secondary causes of chorea like Wilson's disease; (Huntington's disease usually start in the fourth decade), negative family history, there was no recent head injury and patient was not on any antipsychotics or antiepileptics drug. A 2D echocardiography was normal and there was no evidence of rheumatic heart disease. HIV causes chorea by direct effects as well as opportunistic infections like toxoplasmosis involving the basal ganglia [17]. Hyperthyroidism has also been reported to cause chorea probably due to functional alteration in the dopamine turnover in the corpus striatum [18]. HIV serology was negative and thyroid profile was normal in this patient.

The radiological findings in CHBG syndrome include hyperintensities in striatum and globus pallidus in T1 weighed imaging without contrast enhancement. In a report of 20 cases of chorea associated with hyperglycaemia, almost all the patients had striatal abnormalities on neuroimaging which was either contralateral to the hemichorea or bilateral in some cases [16]. The index case had bilateral striatal abnormalities though he had unilateral symptoms. The typical triad of chorea with striatal abnormalities on neuroimaging and hyperglycaemia in a patient with diabetes mellitus clinches the diagnosis of CHBG syndrome, as in this patient.

The treatment of CHBG syndrome or NHH is to achieve optimal glycaemic control with insulin and oral hypoglycaemic drugs. Complete resolution of the movement disorder can occur within 48 hours of glycaemic control, though it may continue for few weeks in many patients [16]. In a case series of 5 adult patients with non-ketotic hyperglycaemia, chorea persisted despite optimal control of blood sugars for more than 6 months during follow-up [19]. In the reported cases of Type 1 diabetes mellitus with chorea, there was complete resolution of the involuntary movements in all cases except one [4-9]. In the index patient the movement disorder persisted for more than 4 years probably due to noncompliance to insulin therapy. In those patients with persistent chorea despite blood sugar control, neuroleptics like haloperidol, benzodiazepines like clonazepam and antiepileptics like sodium valproate have been tried with variable response [9,16,19].

CONCLUSION(S)

CHBG is a rare complication of non-ketotic hyperglycaemia which has a good prognosis if promptly recognised and optimal glycaemic control is achieved. Clinicians should be aware that this entity can occur in both Type1 and Type 2 diabetes mellitus.

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PARTICULARS OF CONTRIBUTORS:
1. Postgraduate, Department of Internal Medicine, Mahatma Gandhi Medical College and Research Institute, Pillaiyarkuppam, Puducherry, India.
2. Ex-Assistant Professor, Department of Internal Medicine, Mahatma Gandhi Medical College and Research Institute, Pillaiyarkuppam, Pondicherry, India.
3. Professor, Department of Internal Medicine, Mahatma Gandhi Medical College and Research Institute, Pillaiyarkuppam, Pondicherry, India.
4. Postgraduate, Department of Internal Medicine, Mahatma Gandhi Medical College and Research Institute, Pillaiyarkuppam, Puducherry, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Kalaimani Sivamani,
No. 26, Bheeshmar Street, Kennedy Garden, Lawspet, Puducherry-605008, India.
E-mail: kalaimani.sivamani@gmail.com

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