# Chorea – hyperglycemia- basal ganglia syndrome: A rare entity

Sir,

A 69-year-old male patient presented at the emergency department with a history of abnormal, non-purposeful, involuntary movements involving the right arm from the last 10 hours. It was insidious in onset involving the right shoulder and wrist joint and non-suppressible. The patient had no history of altered sensorium, weakness of any limb or any abnormal movement of any other part of the body.

On examination, his vitals were stable. Neurological examination revealed no abnormality except the hyperkinetic movement which was rapid, semi-purposeful dance-likenon-patterned movement involving the right arm without any sustained abnormal posturing of the involved hand. It was not a stereotyped movement and the patient could not suppress it on its own.

On investigation, his random blood glucose was 894mg/dL, serum sodium was 131 mmol/L and potassium was 5.6mmol/L. Urine examination for ketones was negative. Arterial blood gas (ABG) analysis was within normal limits. Blood urea nitrogen (BUN) was 32mg/dL. The patient was not a diagnosed case of diabetes but his HbA1c was 16.7 and the fundus revealed bilateral non-proliferative diabetic retinopathy (NPDR). The abnormal movement was suggestive of chorea (video 1) and the etiology was kept as hyperosmolar state as his serum osmolality was more than 320 mosm/L.

The patient underwent non-contrast computed tomography head which showed hyperdensity in the left basal ganglia and furthered confirmed on gadolinium-enhanced magnetic resonance imaging (MRI) (Figure1), in which there was T1 hyperdensity in the same region. The patient was managed with IV insulin andhis movement improved after glycemic control. There was no residual disability or recurrence of the movement.

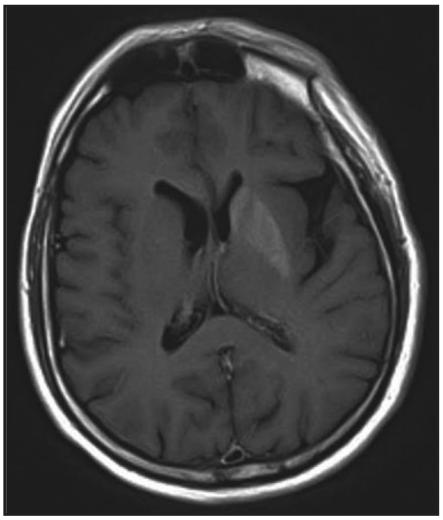

**Figure 1 - MRI showing hyperintensity in basal ganglia.**

Chorea hyperglycemia basal ganglia syndrome (CHBG) is a rare condition which manifests in the presence of non-ketotic hyperglycemia. It is characterized by the manifestation of hemichorea-hemiballism with uncontrolled blood sugar levels. The exaxt etiology is not known but various theories have been postulated. It is proposed that hyperglycemia may impair the cerebral autoregulatory mechanisms of the central nervous system which in turn can lead to hypoperfusion and sequential activation of anaerobic metabolism [1]. This results in the depletion of gamma-aminobutyric acid (GABA) within the basal ganglia neurons. GABA and acetate are depleted rapidly in non-ketotic hyperglycemia which causes a reduction in acetylcholine synthesis [2].

The pathogenesis of this condition is thought to be related to hyperglycemia-induced perfusion changes in the contralateral striatum. It has been reported that hyperglycemia shifts cerebral metabolism to an anaerobic pathway and that striatal GABAergic neurons are particularly vulnerable to ischemia [3]. The selective loss of striatal GABAergic neurons may be related to disinhibition of the thalamocortical pathway, resulting in the motor cortical hyperexcitability [4]. Recently, it was suggested that the presence of acanthocytes in circulating peripheral blood might render people with diabetes prone to develop HCHB [5]. Hyperviscosity induced by hyperglycemia then causes a disruption of the blood-brain barrier and resulting in transientischemia of the vulnerable striatal neurons [6]. Hemichorea is the occurrence of these movements on one side of the body as occurred in our patient [7,8].

The differential diagnosis which can be excluded for this condition and which should be kept in mind are hemorrhagic or ischemic stroke, intracranial neoplasms, systemic lupus erythematosus, Wilson's disease, and thyrotoxicosis. Very few case reports have been published in the literature because of its rarity but it should be kept in mind when dealing with such patients.

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Received - 26 April 2019
Initial Review - 13 May 2019
Accepted - 22 July 2019

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*Funding: None; Conflict of Interest: None Stated.*

**How to cite this article:** Malakar S, Sharma KN, Sharma T, Sharma A, Chorea – Hyperglycemia- Basal Ganglia Syndrome: A Rare Entity. Indian J Case Reports. 2019;5(4):397-398.

Doi: 10.32677/IJCR.2019.v05.i04.035