Dancing with Diabetes: An unusual case of Chorea hyperglycemia basal ganglia syndrome

Anuvanthana S[1], Vigneshvarprashanth Umapathy[2], G. Dominic Rodriguez[3]
[1,2]Resident Internal Medicine, Kauvery Hospital, Tennur, Trichy, India
[3]HOD Internal Medicine, Kauvery Hospital, Tennur, Trichy, India

Correspondence: [1]anuvanthana@gmail.com, [2]worldofuv@gmail.com, [3]domappa1966@gmail.com

Abstract

We report a rare case of chorea hyperglycemia basal ganglia syndrome in a 76-year-old gentleman with uncontrolled ketotic hyperglycemia. His symptoms resolved with strict glycaemic control.

Background

Chorea is characterized by excessive, involuntary, irregular, and non-rhythmic semi-purposeful movements that can be caused by vascular, degenerative, genetic, autoimmune, neoplastic, infectious, or metabolic pathologies [1]. An entity called chorea hyperglycemia basal ganglia (CHBG) syndrome can manifest in patients with uncontrolled non-ketotic hyperglycemia. Here, we report a case of CHBG syndrome in an uncontrolled hyperglycemia patient with positive plasma ketone, which is extremely rare.

Key-words: Chorea, Chorea hyperglycemia basal ganglia syndrome, Ketotic hyperglycemia, insulin, Hemiballismus.

Case Presentation

A 76-year-old gentleman who is known to have systemic hypertension, end-stage renal disease, and uncontrolled diabetes mellitus was brought with complaints of sudden onset of involuntary movements involving the left upper and lower limb with no loss of consciousness or loss of contact with the surroundings. He was initially admitted elsewhere, where he was noted to have a random blood sugar of 460 mg/dL (25.6 mmol/L). He was treated with diazepam, haloperidol, levetiracetam, and split doses of insulin. Although there was some response, he continued to have involuntary movements and was brought to us for further management. There was no history of recent trauma to the head.

On Examination

He was obeying commands, had mild confusion, noted to have left hemichorea, but there was no weakness or sensory loss.

His blood sugar was 301 mg/dL (16.7 mmol/L) and high anion gap metabolic acidosis (HAGMA) due to uremia was noted.

Management

Hyperglycemia-induced seizure was suspected. However, the positive plasma acetone added confusion in making a diagnosis.

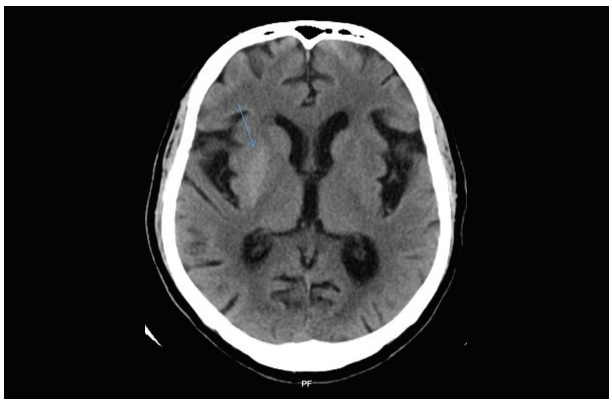
His HbA1c was 13.7. He was started on insulin infusion, midazolam infusion and levetiracetam.

CT brain showed hyperdensity in the right-side basal ganglia region predominantly in the putamen and old infarct of the left external capsule region.

Chorea hyperglycemia basal ganglia (CHBG) syndrome was suspected.

Strict glycaemic control throughout was achieved with intravenous insulin infusion. Gradually involuntary movements subsided. He also had acute on chronic kidney disease which was managed conservatively.

The need for strict glycemic control was explained to the patient and his family.



CT Brain showing (blue arrow) hyperdensity in the right-side basal ganglia region predominantly in the putamen.

Discussion

A diagnosis of Chorea hyperglycemia basal ganglia (CHBG) syndrome was made, as the patient manifested with the triad of hemichorea, hyperglycemia and abnormalities limited to striatum in the CT brain, and the movements subsided with glycaemic control.

CHBG syndrome is a rare condition that usually manifests in the setting of uncontrolled non-ketotic diabetes mellitus, sometimes called non-ketotic hyperglycemia chorea-ballismus. This syndrome manifests with a triad of acute or subacute hemichorea-hemiballismus, hyperglycemia, and unique reversible abnormalities limited to the striatum on neuroimaging. The common differentials for this condition include haemorrhagic or ischemic stroke involving the basal ganglia, neoplasm, Huntington's chorea, Sydenham's chorea, systemic lupus erythematosus causing encephalitis, Wilson's disease, thyrotoxicosis and drug toxicity[1,3]. This patient neither had a family history of chorea, nor a history suggestive of an alternate diagnosis.

This syndrome could easily be misdiagnosed as intracerebral hemorrhage because of its unfamiliarity and radiographic features. The characteristic findings on a T1 weighted MRI are high signal intensity in the contralateral putamen or caudate nucleus. This may be due to the reactive proliferation of glial cells. CT scan also reveals hyper density in the contralateral basal ganglia. However, the involvement of basal ganglia can be bilateral as well. A recent systematic review showed that MRI was more sensitive than CT scan in detecting basal ganglia changes [1, 3].

Although CHBG syndrome occurs mostly in Asian elderly women with non-ketotic hyperglycemia, a few cases of CHBG syndrome in ketotic hyperglycemia patients have also been reported. Yuyan Tan et al from China have described 2 cases with a similar presentation as this patient with positive urine ketones [4].

The exact pathophysiology of CHBG syndrome is still uncertain. However, Carla Battisti et al had put forward that hyperglycemia may directly induce alterations in dopaminergic activity (by upregulation of dopamine receptors and decreased dopamine catabolism) in the striata of predisposed patients and dysregulation of direct and indirect pathways, which ultimately increase the excitatory effect of thalamus on cortex [5].
A chronological relationship between restoration of normal blood glycemic levels and improvement of the chorea has been described [6], which was the same in our patient as well. Although with glycemic control, the involuntary movements subside in a matter of hours to days, the radiological changes have been reported to take about 6 months to resolve after the correction of hyperglycemia.

Conclusion

Sometimes ketotic hyperglycemia also can cause involuntary movements. However, only a meagre of such reports has been described in the literature. Commonly mistaken for seizures, recognizing this syndrome could avoid unnecessary use of antiepileptics and sustained control of hyperglycemia results in remarkable recovery. Unlike, other causes like stroke, neoplasm or encephalitis, chorea due to hyperglycemia can be managed easily with glycaemic control.

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