Hyperglycemia-induced involuntary movements (a.k.a, C-H-BG syndrome) with bilateral putaminal involvement

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Abstract

Hyperglycemia in uncontrolled diabetes mellitus can present as hemichorea-hemiballismus (C-H-BG syndrome). This condition is mostly reported in older Asian women. Here, we report the case of a 70-year old Caucasian woman with uncontrolled type 2 diabetes mellitus presenting with sudden-onset chorea in all four extremities with greater pronounce on the left side. Laboratory workup showed a serum glucose level of 597 mg/dl and HbA1c of 12.3%. Imaging studies revealed symmetric hyperintense signal abnormalities in the putamina on T1-weighted magnetic resonance (MR) imaging. The patient was treated with insulin for hyperglycemia and achieved reversal of symptoms within one week. To our knowledge, this is the first case report of C-H-BG syndrome in a Caucasian female with bilateral putamina lesions on T1-weighted MR imaging. C-H-BG syndrome should be included in the differential diagnosis in patients with uncontrolled diabetes or hyperglycemia presenting with new-onset chorea, irrespective of race.

Keywords: Hemichorea, Chorea, Putamen, Hyperglycemia, Diabetes, MRI

Introduction

Chorea is an abnormal involuntary movement characterized by brief, asymmetrical, non-stereotypical movements of facial muscles and/or extremities. There are many established causes of chorea, including basal ganglia strokes, Wilson’s disease, thyrotoxicosis, neoplasms and systemic lupus erythematosus [1].

Sudden-onset chorea in patients with uncontrolled diabetes mellitus in a hyperglycemic, hyperosmolar, non-ketotic state is a well-studied, but rare phenomenon [2]. It is most commonly seen in older (>70) Asian women [3]. Magnetic resonance imaging studies in patients with hyperglycemia-induced chorea frequently shows a unilateral hyperintensity in the putamen, notably on T1-weighted images. Bilateral basal ganglia involvement is rare, comprising only 15% of cases [2].

The constellation of findings mentioned above constitutes Chorea-Hyperglycemia-Basal Ganglia Syndrome, or C-H-BG syndrome.

Case Report

Our case describes a 70-year-old Caucasian woman with a significant medical history of uncontrolled type 2 diabetes mellitus, hypothyroidism and depression. The patient presented with abnormal movement of bilateral upper and lower extremities, more prominent on the left side. She also had lip puckering, and oro-mandibular stereotypical chewing movements. These symptoms were associated with imbalance and dysarthria. The patient had an extensive but inconclusive work up at an outside hospital for similar complaints a week prior. During that time, she was started on benztropine and developed an allergic reaction for which prednisone was started. The patient had no prior history of neurological illness, antipsychotic medication use, or any significant cardiac history.

General physical exam was unremarkable. On neurological exam, higher mental functions were normal. The cranial nerve exam was significant for oral buccal dyskinesia. She demonstrated involuntary choreiform movement in upper and lower extremities bilaterally. Significant ataxia was noted on finger-to-nose exam. She was unable to perform heel-to-toe test. She had dysrhythmia on rapid finger tapping. During several repeated examinations, her choreiform movements attenuated and momentarily disappeared with distraction and increased with attention. It had variable magnitude at rest, during posture holding, and during action. These movements included oral-buccal-mandibular-lingual movements. The movements became disrupted with change in frequency during variable rhythms of finger tapping with the unaffected hand at different rates. Patient was tested in both sitting and standing position with similar findings. Motor exam revealed good strength throughout.

Initial laboratory workup showed serum glucose level of 597 mg/dl and HbA1c of 12.3%. She was found to have She was
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found to have weakly positive ANA titer (1:320), and rest of her labs, including ceruloplasmin and ketone bodies were within normal limits. MRI on day 5 showed foci of T2 hyperintense signal abnormality in the central pons, periventricular and deep cerebral white matter suggestive of chronic microangiopathy as well as symmetric T1 hyperintense signal abnormality in the putamina that can be seen with nonketotic hyperglycemia. Our working diagnosis was hyperglycemia-induced involuntary movement supported by imaging abnormalities. The patient was treated with insulin glargine and insulin lispro to correct her elevated serum glucose. On day 6, there was significant improvement in symptoms corresponding to significant reduction in glucose levels (below 250). The symptoms had completely resolved upon discharge. The patient was lost to follow up and a follow-up MRI was never completed (Figures 1 and 2).

Discussion

Involuntary movements, especially chorea, have been associated with nonketotic hyperglycemia in type 2 diabetic patients. Previous case reports have described hyperglycemia-induced choreiform movements in the elderly Asian female population. T1-weighted MR imaging studies typically show a unilateral hyperintensity in the putamen. Our patient has two distinctive characteristics – she is a Caucasian female with bilateral symmetric putaminal lesions.

With the increased prevalence of type 2 diabetes mellitus, physicians must be cognizant of the unusual presentations of hyperglycemia. Early identification and treatment of C-H-BG syndrome can improve outcome.

Author Contributions

Drafting the manuscript: Shitiz Sriwastava, Omar Basha, Meghana Srinivas, Nikita Khetarpal, Aaron Desai.

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References


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