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Research Article

Hemichorea as an Atypical Manifestation of Diabetes Mellitus

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Abstract

Non-ketotic hyperglycemic hemichorea (NHH), also known as chorea, hyperglycemia, basal ganglia syndrome or diabetic striatopathy, is a rare neurological complication of non-ketotic hyperglycemia. We describe a case of 65 year old woman with previously undiagnosed diabetes who presented to the emergency room with continuous, involuntary, writhing movements on the left side of her body for one weeks duration. Exam revealed choreiform movements involving the left arm and leg. Laboratory testing revealed elevated blood glucose at 250 mg/dl and elevated HbA1C at 6.5%. MRI brain showed T1 hyper intensity in right basal ganglia without GRE correlate. Patient was diagnosed with non-ketotic hyperglycemic hemichorea and started on metformin. Follow-up MRI brain showed resolution of T1 hyper intensity with normalization of blood glucose. NHH is rare syndrome characterized by sudden onset of hemichorea in poorly controlled diabetics. Early detection and intervention is critical in management of disease morbidity and mortality.

Keywords: Hemichorea, hemiballismus, non ketotic hyperglycemia, diabetic striatopathy, basal ganglia

Abbreviations

T2DM: Type 2 Diabetes Mellitus; HCHB: Hemichorea Hemiballism; NHH: Non-ketotic Hyperglycemic Hemichorea; C-H-BG: Chorea, Hyperglycemia, Basal Ganglia Syndrome; CT: Computerized Tomography; MRI: Magnetic Resonance Imaging; FLAIR: Fluid-attenuated Inversion Recovery; DWI: Diffusion Weighted Imaging; HbA1C: Glycosylated Hemoglobin; GRE: Gradient Echo.

Introduction

Diabetic striatopathy or non-ketotic hyperglycemic hemichorea (NHH) is a rare and debilitating manifestation of diabetes mellitus type 2 (T2DM). It has a tendency to occur in the elderly population, is slightly female predominant and frequently occurs in patients of Asian descent [1]. It is characterized by unilateral involuntary, continuous, non-patterned movements, associated with contralateral basal ganglia lesions on CT or MRI, and clinical recovery with normalization of blood glucose levels. The involuntary movements typically occur at the onset of hyperglycemia and subside with euglycemia. However, occasionally patients develop NHH several weeks after the hyperglycemic event, even when blood sugar is normal [1]. We report a case of hyperglycemic hemichorea as an initial manifestation of T2DM.

Materials and Methods

A 65 year old African American woman with no significant past medical history presented to the emergency room with worsening continuous, involuntary, writhing movements on the left side of her body for one weeks duration. The symptoms started in distal lower extremity and progressed to involve left upper and lower extremity. There was no history of associated weakness, sensory changes, alteration of consciousness, trauma, recent illness, alcohol or increased caffeine intake. She denied family history of abnormal movements. Exam revealed choreiform movements involving the left proximal and distal upper and lower extremities. There was

no evidence of associated weakness, numbness, increased tone or hyper-reflexia. Laboratory testing revealed elevated blood glucose at 250 mg/dl and elevated HbA1C at 6.5%. B12 and thyroid levels were unremarkable. MRI of the brain revealed T1 hyper intensity in right basal ganglia involving putamen without GRE correlate (Figure A). Patient was diagnosed with non-ketotic hyperglycemic hemichorea and started on oral hypoglycemic agent, metformin. The involuntary movements resolved within one week and follow-up MRI of the brain one month later showed resolution of T1 hyper intensity (Figure B) with normalization of blood glucose.

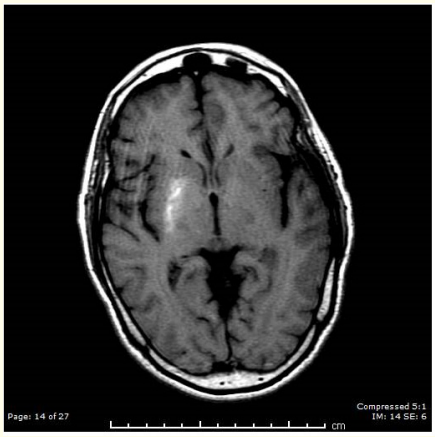

Figure A: Axial T1 weighted MRI of the brain with right basal ganglia hyperintensity.

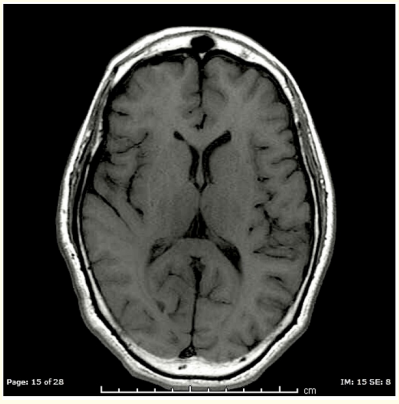

Figure B: Axial T1 weighted MRI of the brain with resolution of right basal ganglia hyperintensity.

Results and Discussion

T2DM can lead to a wide range of systemic complications. Neurological complications of poorly managed T2DM can range from peripheral neuropathy to seizures and life threatening coma. HCHB is a rare neurologic disorder that can be seen in acute decompensated T2DM. NHH, also known as chorea, hyperglycemia, basal ganglia (C-H-BG) syndrome or diabetic striatopathy is a rare neurological complication of T2DM [2]. We report an unusual case of NHH, who's only presenting sign was unilateral hyperkinesis, which completely resolved after correction of blood glucose levels.

NHH is a spectrum of involuntary, continuous non-patterned movement involving one side of the body. Possible etiologies of NHH include ischemic or hemorrhagic stroke, malignancy, thyrotoxicosis, systemic lupus erythematosus, and Wilson's disease [1,2]. Our case elucidates the need to be cognizant of hyperglycemia as a cause of NHH. This condition typically occurs in elderly females of Asian descent and often heralds a new diagnosis of diabetes [1]. The underlying pathophysiology remains unclear. It is hypothesized that failure of cerebral blood flow autoregulation in combination with hyerglycemia induced basal ganglia metabolic derangement can contribute to this syndrome. Other postulated mechanisms include [3]:

- Decreased availability of gamma-aminobutyric acid in the striatum secondary to non-ketotic state
- hyperglycemia leading to hyperviscosity with subsequent regional blood-brain barrier disruption and metabolic damage
- The increased sensitivity of dopaminergic receptors in postmenopausal state.

Imaging of the brain is very helpful in the acute phase and tracking the prognosis. CT head is initially normal, but subsequently demonstrates subtle hyperdensities in the striatum, primarily involving the caudate and putamen. These observations tend to occur contralateral to the side affected by hemiballistic, hemichoreic movements. MRI of the brain is the primary modality of choice for diagnosing NHH and classically demonstrates signal abnormalities localized to putamen and/or caudate contralateral to the symptomatic side. It is associated with contralateral striatal T1 hyperintensity, T2/FLAIR hypointensity and diffusion restriction on DWI sequences [4]. Overall, the T1 hyperintensity is the most consistent

finding of the disease. Other associated findings do not present to the same frequency and tend to vary [5]. Imaging findings gradually resolve after hyperglycemia correction. However, they return to baseline much more slowly when compared to clinical features [6]. Although movements typically resolve within one week of hyperglycemia correction, additional therapies including haloperidol, risperidone, and tetrabenazine may be required in certain cases [7].

Conclusion

We report an unusual case of hemichorea as an initial manifestation of T2DM. Larger case series are quintessential to gain a better understanding of the pathophysiological mechanisms. It is critical to establish the diagnostic criteria of NHH. The most important diagnostic feature is the correction of the underlying hyperglycemia leading to a swift improvement of the involuntary movements.

In a patient with unilateral involuntary movements in the setting of uncontrolled diabetes, NHH syndrome should be on the top of the differential especially when the MRI has characteristic features of hyperintensities in the contralateral basal ganglia. This is a rare disease that warrants consideration because it is easily treatable with timely correction of hyperglycemia. Thus, prompt recognition and early management is critical in avoiding adverse outcomes.

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Conflict of Interest

Author reports no financial interest or any conflict of interest exists.

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