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Case Report

Permanent Chorea In A 76-Year-Old Female With Poorly Controlled Diabetes Mellitus

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Abstract

Background: Chorea hyperglycemia basal ganglia syndrome (CHBS) is a rare disorder accounting for only 1% of the total chorea-ballism cases. It is characterized by an acute or subacute, most often unilateral evolvement of chorea/ballism after nonketotic hyperglycemic episode, MRI characteristic features mainly of T1-hyperintense signal in the basal ganglia, and alleviation of symptoms as glycemic control is achieved. **Case presentation:** A 76-year-old poorly controlled diabetic female, presented with acute gait disorder. During her admission, she had a nonketotic hyperglycemia episode with blood glucose levels up to 700 mg/dl. A few days later she developed bilateral choreiform limbs' movements more pronounced in her lower limbs. A magnetic resonance imagining (MRI) scan of the brain revealed T1 hyperintense and T2 hypointense signals in the basal ganglia bilaterally suggesting CHBS. A gradual control of her blood sugar was successful, achieving a HbA1C of 7%. During an 18-month follow-up, there was no improvement in her chorea despite strict glycemic control. **Conclusions:** The emergence of chorea in uncontrolled diabetic patients should raise the suspicion of CHBS in the setting of characteristic T1 MRI hyperintensity in the basal ganglia. Our report adds to the database a case of CHBS with an unfavorable outcome, emphasizing the importance of awareness of this diagnosis and glycemic control.

Keywords: Chorea; CHBS; Hyperglycemia

Introduction

Chorea hyperglycemia basal ganglia syndrome (CHBS) is a rare disorder that manifests with new-onset chorea in patients with uncontrolled type 2 diabetes mellitus [1], and less frequently in type 1 diabetes mellitus. Here, we report a case of a poorly controlled diabetic patient who developed permanent CHBS showing no improvement during a long-term follow-up, despite a strict glycemic control.

Case Presentation

A 76-year-old female with a history of essential hypertension, dyslipidemia, heart failure, and poorly controlled type 2 diabetes mellitus presented to the emergency department with an acute gait disorder and a fall causing a traumatic injury to her left upper limb. Her initial neurological examination revealed dysarthria and unsteady gait. A CT scan of the head was unremarkable except

for chronic ischemic changes in the left parietal lobe that were considered unrelated to her acute symptoms. A stroke workup was negative. Her HbA1C on admission to the ER was 12.5%. During her admission, she had a nonketotic hyperglycemia episode with blood glucose levels up to 700 mg/dl. A few days later she developed bilateral choreiform limbs' movements more pronounced in her lower limbs. A magnetic resonance imagining (MRI) scan of the brain revealed T2 widespread hyperintense lesions in the white matter consistent with chronic ischemic changes, T1 hyperintense and T2 hypointense signals in the basal ganglia bilaterally suggesting a metabolic etiology (Figure 1). An extensive workup including routine biochemical blood tests and lumbar puncture were all unremarkable as well as neoplastic and paraneoplastic workups. There was no history of exposure to neuroleptic medications. CHBS was diagnosed. A gradual control of her blood sugar was successful, achieving a HbA1C of 7% three months after admission. During an 18-months of follow-up, there was no improvement in her chorea despite strict glycemic control. She was treated symptomatically with Tetrabenazine resulting in a partial response.

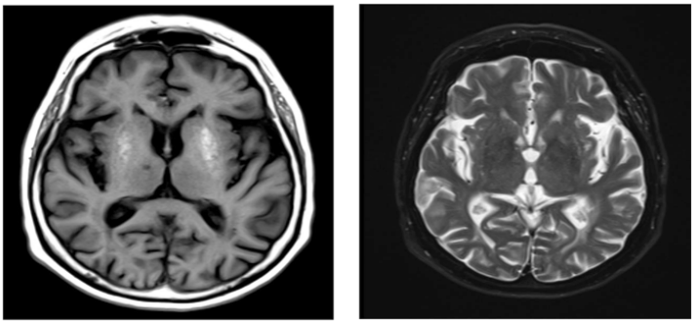

Figure 1: Brain MRI showing bilateral abnormal signal in the basal ganglia with high T1W signal (left image) and low T2W signal (right image).

Discussion

CHBS is a rare movement disorder associated with uncontrolled diabetes mellitus. The classical clinical presentation of CHBS is characterized by acute or subacute unilateral chorea or ballism that most often affects the upper more than the lower limbs and develops following a nonketotic hyperglycemic episode [2]. Brain MRI usually shows characteristic features of a T1-weighted hyperintense signal and restricted diffusion in the basal ganglia [3]. The pathophysiology of CHBS is poorly understood, some hypothesized mechanisms include hyperviscosity secondary to hyperglycemia or decreased gamma-aminobutyric acid availability in the striatum [4].

The clinical presentation of our patient has several unique features: she presented with bilateral choreatic movements that were more pronounced in her lower limbs. Based on previous reports, this clinical distribution is less common; however, involvement of the lower limbs has been reported as well [5].

The chorea in our patient did not resolve during 18 months of follow-up in spite of good glycemic control. Although most CHBS cases show complete, or at least partial recovery, as the glycemic control is achieved, persistent or recurring symptoms were described [6,7] indicating possible irreversible damage to the basal ganglia. This raises the question whether early treatment of hyperglycemia improves the outcome of CHBS. However, given the paucity of cases, little is known about risk factors for persistent cases. A recent meta-analysis of CHBS cases reported complete resolution of chorea in 76.2% of the patients who received glucose-control and anti-chorea treatment and an 18.2% recurrence rate [8]. The time interval between symptoms onset and treatment was similar between patients who had a complete response and those who had partial or no response. However, the time interval between the failure of glycemic control to relieve chorea and the administration of anti-chorea treatment was not mentioned in most of the studies that were included in this meta-analysis. Animal models might help identify the long-term effect of prolonged hyperglycemia on the basal ganglia.

Undoubtedly, hyperglycemia should be managed tenaciously to prevent systemic complications. It is unclear whether the type of diabetic treatment (e.g. oral vs. subcutaneous or intravenous) affects CHBS outcome. Nevertheless, Insulin should be considered the treatment of choice in all patients with severe hyperglycemia. In terms of chorea-control, most of the published cases were treated with haloperidol [8].

CHBS is a rare disorder but probably underdiagnosed in the western population [9], accounting for only 1% of the total chorea-ballism cases [10]. The knowledge of this disorder is based on case reports and case series sharing distinctive clinical and radiological features. The emergence of chorea in an uncontrolled diabetic patient should raise the suspicion of CHBS in the setting of characteristic T1 MRI hyperintensity in the basal ganglia. Our report adds to the database a case of CHBS with an unfavorable outcome, emphasizing the importance of awareness of this diagnosis and glycemic control. Whether early treatment influences prognosis remains unsettled. However, given the many systemic complications of hyperglycemia, it seems essential to treat these patients as soon as possible.

Availability of data and materials: Authors declare that all data are available.

Conflict of Interest: No specific funding was received for this work, and the authors declare that there are no conflicts of interest relevant to this work.

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Ethical Compliance Statement: The authors confirm that the approval of an institutional review board and patient consent was not required for this work.

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