CASE REPORT

Non-ketotic Hyperglycemia Presenting with Acute Hemichorea and Ballism
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Non-ketotic hyperglycemia is a complication of poorly controlled diabetes mellitus. Rarely, it can present like an acute neurological syndrome with unilateral choreiform and ballistic movements. Such a presentation usually raises the suspicion of a cerebrovascular event and prompts more workup. Moreover, the neuroimaging in this condition also suggests a variety of potential possibilities. Identification of this rare presentation of non-ketotic hyperglycemia helps with the appropriate management and avoid unnecessary investigations. In this case report, we report the case of an elderly woman who presented with hemichorea-ballism due to non-ketotic hyperglycemia and discuss the literature on this presentation. We also highlighted the differential diagnosis based on neuroimaging.

INTRODUCTION

Hemichorea/hemiballism is a neurological syndrome characterized by violent involuntary movements on one side of the body. These movements mainly involve the upper extremity. This syndrome is usually associated with acute vascular insults in the vicinity of the subthalamic nucleus. Lesions in different sites of the striatum were also reported to result in a similar syndrome1. In the last three decades, several patients with this syndrome associated with elevated blood sugars were reported. Most of these patients have a fairly quick recovery from their symptoms with the correction of hyperglycemia. However, we report a case of prolonged hemichorea/hemiballism that started in the setting of poorly controlled diabetes mellitus and associated with T1-weighted putaminal hyperintensity on magnetic resonance imaging (MRI) scan of her brain.

CASE PRESENTATION

A 64-year-old Caucasian female presented to the outpatient neurology clinic with an almost eight-month history of abnormal movements of the left upper and lower extremities that started suddenly while she was at church. They were present constantly during the daytime and would subside during sleep. These movements were choreiform in nature most of the time and occasionally would become ballistic with reports of self-injury during the flailing. Patient reported that her diabetes was under poor control before and around the time her clinical symptoms started. Her other significant medical problems included chronic kidney disease and peripheral neuropathy.

Her examination did not show rigidity or bradykinesia in the limbs. She also did not have any focal weakness. The deep tendon reflexes were slightly
diminished in the lower extremities symmetrically which may have been due to her peripheral neuropathy. The MRI of the brain showed a T1 hyperintensity in the right putamen without any evidence of acute infarction or hemorrhage (figure 1). Magnetic resonance angiography did not show any significant stenosis of the brain or neck vasculature. A trial of haloperidol resulted in significant improvement of her abnormal movements but caused severe sleepiness and the medication was stopped. She was started on aripiprazole which also helped significantly with the abnormal movements. During this time, the patient started to get her blood sugars under control. Repeat MRI of the brain in three months showed reduction in the T1 hyperintense signal in the right putamen (figure 2). Her neuroleptic treatment was complicated by

![Initial MRI scan at the time of presentation showing T1 weighted hyperintensity in the right putamen.](image-url)
the appearance of buccolingual and copulatory dyskinesias that required gradual discontinuation of the neuroleptic therapy. Her symptoms gradually resolved in the next few months only to become manifest during times of emotional stress as the patient went off the medication completely.

**DISCUSSION**

Although rare, nonketotic hyperglycemic hemichorea-ballism is a potential complication of poorly controlled blood sugars and usually presents in patients with no previous history of diabetes or hyperglycemia. Sometimes, the same presentation can be seen in the setting of...
diabetic ketoacidosis. Most of the affected patients are in the 50-80 age range with usually no history of diabetes mellitus\textsuperscript{2}. The usual onset of the symptoms is acute to subacute and typically over a period of hours. Hemichorea and ballism associated with hyperglycemia was first described in 1960 by Bedwell\textsuperscript{3}. Interestingly, patients affected by this disease entity are mostly females and abnormal movements often get significantly better with the correction of hyperglycemia. A case of non-ketotic hyperglycemia with vague non-localizing symptoms without an associated movement disorder has also been described in the past\textsuperscript{4}. Most of the cases reported in the past had significant resolution of symptoms in 1-2 days. Our patient had her symptoms for almost 8 months which is unusual.

The pathophysiology of this acute syndrome is unclear. It is thought that hyperglycemia might result in a greater utilization of gamma-aminobutyric acid (GABA) as an alternative energy source resulting in its depletion. GABA is a major inhibitory neurotransmitter in the brain and the basal ganglia circuitry. This reduction in GABA might disinhibit the thalamus which can result in abnormal movements\textsuperscript{2}. Various other mechanisms were suggested that include acute basal ganglia dysfunction in the setting of hyperosmolar state\textsuperscript{5}, hyperviscosity\textsuperscript{6}, petechial hemorrhage\textsuperscript{7}, inflammation\textsuperscript{8}, and cerebrovascular insufficiency\textsuperscript{9}. Post-menopausal alterations of the GABA and/or dopamine receptors in elderly females may be the reason for higher incidence in this group\textsuperscript{2}.

While the characteristic findings on neuroimaging may vary to some degree, the pattern of basal ganglia involvement sparing the internal capsule along with the sudden onset of abnormal movements in the setting of markedly elevated blood sugars should prompt the consideration of this disease entity. The blood sugar level above which this condition gets triggered is not clear. The condition is typically easily treatable by correcting blood sugar with rapid resolution of abnormal movements. Computed tomography (CT) scan of the brain commonly shows hyperdensity of the putamen and caudate nucleus sparing the internal capsule\textsuperscript{10} which sometimes can be mistaken for basal ganglia hemorrhage\textsuperscript{11}. The MRI of the brain typically shows T1-weighted hyperintensity in the lentiform nucleus sparing the internal capsule with some cases reported to be showing restricted diffusion of striatum\textsuperscript{12}, high T2-weighted signal in globus pallidus, low T2-weighted signal in the striatum and contrast enhancement\textsuperscript{8}.

Most patients with this condition recover with optimization of hyperglycemia and the recovery ranges anywhere from one day to few months. A small minority of the patients can have persistent chorea\textsuperscript{13}. Radiological improvement is seen in almost all the cases including those with persistent chorea. Along with management of blood sugar, treatment options include D2 receptor blockers like haloperidol though their prolonged use is associated with increased risk of tardive dyskinesias as seen in our patient. Tetrabenazine has been successfully used in some patients\textsuperscript{14} and is not associated with long term risk of tardive dyskinesias. Successful long-term treatment with thalamic deep brain stimulation (DBS) has also been reported in a case of hemichorea-hemiballism in the setting of diabetes mellitus\textsuperscript{15}.

**CONCLUSION**

Hemichorea/ballism in the setting of non-ketotic hyperglycemia is a rare but treatable neurological condition. The sudden appearance of unilateral symptoms usually raises the suspicion of an acute vascular event which might result in unnecessary
diagnostic studies. Identification of the characteristic MRI findings in the appropriate clinical setting will help in early recognition of this rare syndrome and prompt the initiation of treatment. Compared to a cerebrovascular event causing a movement disorder, this syndrome usually has a good prognosis with resolution of symptoms after correction of elevated blood sugar levels.

**Notes**

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**References**