Vanishing Hemichorea-Hemiballismus Due to Non-Ketotic Hyperglycemia: An Unusual Presentation



Background

Hemichorea-hemiballismus is a rare presentation of severe hyperglycemia without ketosis. We present a case of hemichorea in a patient with uncontrolled, newly-diagnosed diabetes mellitus.

Case Presentation

A 66 year-old Bengali female presented to the ED department of Jamaica Hospital Medical Center with steadily worsening purposeless involuntary movements of the right upper and lower extremities over the last two weeks. Movements were present throughout the day and not diminished with sleep, although they did not prevent the patient from walking.

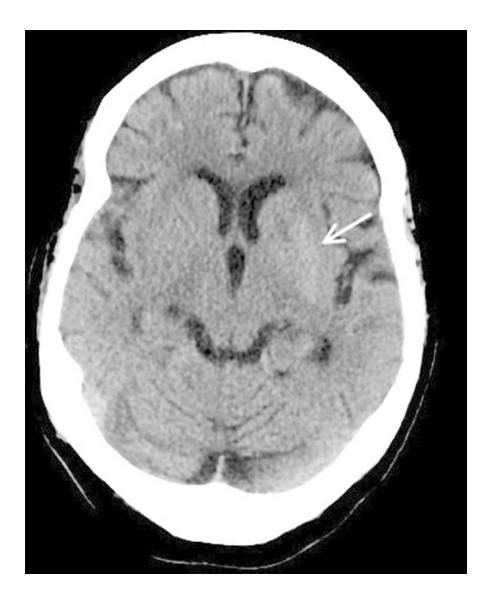
Two days prior to the onset of symptoms, the patient had been referred to the ED from office due to finger-stick glucose of > 600 mg/dL. The patient had no ketonemia at that time. She briefly received insulin and fluids but was sent home on metformin and sitagliptin since she declined insulin. This was the first time the patient was diagnosed with diabetes mellitus.

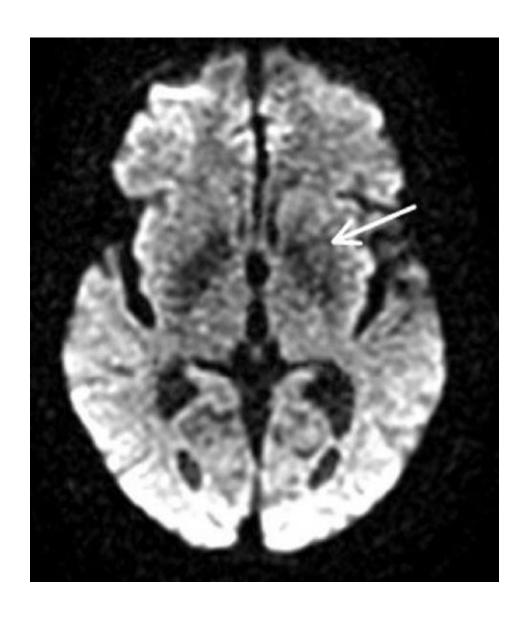
On exam, the patient was awake, alert, and well oriented. Choreiform movements of the right upper and lower extremities were noted. Power was 4/5 on the right side and 5/5 on the left. Laboratory work-up again showed elevated glucose levels in the range of 400 – 500 mg/dL without ketonemia.

The patient was initially admitted with impression of basal ganglia stroke, but CT of head showed high intensity in the area of left lentiform nucleus. MRI of brain with diffusion-weighted imaging and MR angiography did not confirm the presence of acute ischemia, but high intensity in the same area of left basal ganglia on T1-weighted view, suggesting hemichorea secondary to non-ketotic hyperglycemia.

Insulin was used aggressively to control hyperglycemia. Over the next few days, the purposeless movements started to improve. However, patient required almost a month before all abnormal movements disappeared.

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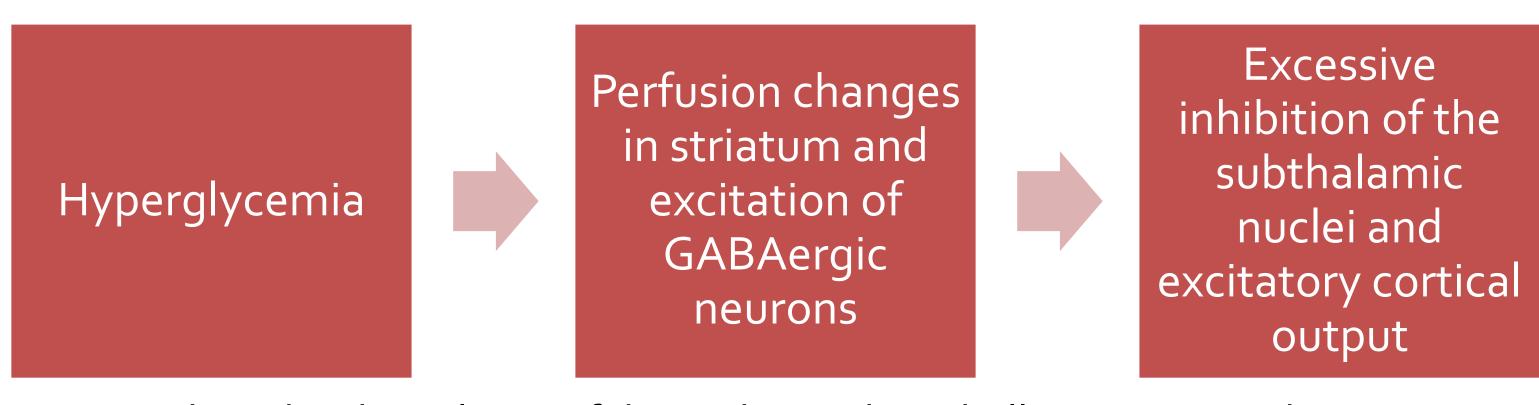


Left

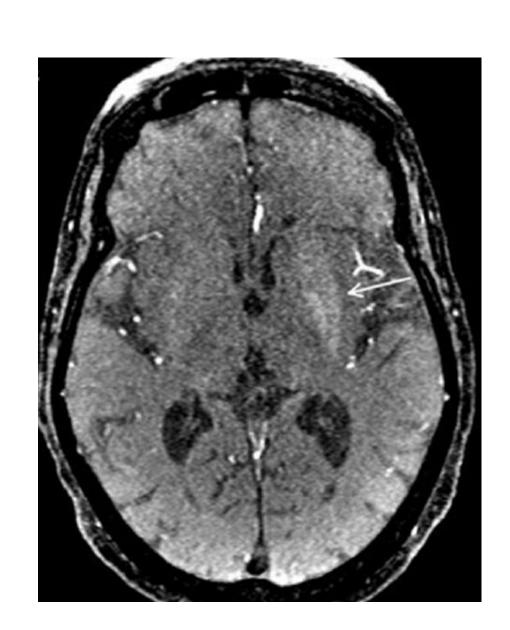
CT of head without contrast showing high intensity in the left lentiform nucleus Middle

Diffusion-weighted MRI of brain showing only minimal increase in signal intensity in the left lentiform nucleus, not typical of acute ischemic stroke Right

T1-weighted MRI of brain showing high-intensity signal in the left lentifom nucleus, suggesting hemichorea secondary to non-ketotic hyperglycemia



Proposed pathophysiology of hemichorea-hemiballismus secondary to nonketotic hyperglycemia.



Discussion

Hemichorea-hemiballismus secondary to non-ketotic hyperglycemia is a very rare complication of uncontrolled diabetes mellitus, characterized by unilateral, involuntary, poorly patterned movements developing over a period of hours. Many patients are initially admitted with the impression of acute stroke. The mean age of onset is 72 years, with women affected more commonly than men. This may be the first manifestation of decompensated diabetes mellitus, although it can also occur after years of poor glycemic control.¹

The condition should be suspected in the context of choreiform or ballistic movements in a patient with severe hyperglycemia but no ketosis.¹

Diagnosis is supported and differentiated from acute stroke with observation of an area of hyperdensity in basal ganglia on CT, which can be differentiated from hemorrhage by absence of edema or mass effect and sparing of internal capsule. The lesion is best seen on T1-weighted MRI of brain, which shows high signal intensity in the basal ganglia contralateral to the symptomatic side, especially the putamen.^{1,2}

Recognition of this condition is particularly important since the symptoms usually resolve over days or weeks if timely glycemic control is achieved. The MRI findings may persist for months in spite of complete resolution of symptoms.^{1,2}

References



1. Padmanambhan S, et al. A case of hemichorea-hemiballismus due to nonketotic hyperglycemia. *Diab Care* 2013; 36 (4): e55–6.

2. Nagai J, et al. Cranial magnetic resonance imaging and angiography findings in a patient with hyperglycemic hemichorea-hemiballism. J *Clin Endocrinol Metab* 2015; 100 (1): 11–2.