

CHOREA IN NON-KETOTIC HYPERGLYCEMIA

Authors: Michael Modica, Kevin Nesh, Najy Masri Mail

Section 1: Introduction

Chorea is an involuntary, irregular, poorly patterned movement that often times can take on a worm-like appearance termed athetosis. This movement disorder is associated with a wide variety of illnesses ranging from primary hereditary disease, such as Huntington's, to secondary causes including cerebrovascular, para-neoplastic, metabolic, inflammatory and immunologic diseases. Of the metabolic causes, hyperglycemia is the most common [2].

Section 2: Case Presentation

A 71-year-old African American female with past history of diabetes mellitus type II presented to the emergency room with a chief complaint of uncontrollable spasm and odd movements in her left wrist for the past five days. She also noted that her blood sugar had been elevated over the last several days with her glucometer reading "Hi" at home after a recent change in medication from Metformin to Victoza and Glipizide. Physical exam showed

spontaneous choreiform movement of the left wrist and left arm with no other significant findings on the rest of her neurologic examination. Labs at the time of admit were significant for a blood glucose of 1013 mg/dL with a normal anion gap and without ketosis in the blood or urine.

The patient was started on a basal-bolus insulin regimen and neurology was consulted. At the time of neurology's exam, the patient's blood glucose had been reduced to 563 with insulin and hydration. MRI/MRA of the brain were performed which showed no acute findings and were only significant for chronic microvascular changes (Figure 1). Hemoglobin A1c returned the next day at a value of 16.3%. The choreiform movements initially seen were resolved upon improvement in the patient's hyperglycemia. The patient's blood glucose continued to be managed with subcutaneous insulin and the patient was discharged the next day with no noted choreiform movements and with an insulin regimen to be optimized by her primary care physician as an outpatient.

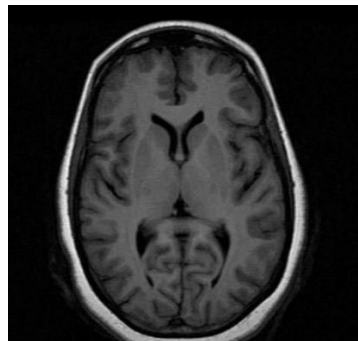


Figure 1. MRI brain of the patient showing chronic microvascular changes in an otherwise normal study.

Section 3: Discussion

Choreiform movements can be a rare presenting symptom of non-ketotic

hyperglycemia, perhaps more commonly in elderly women. In some cases, hyperintense lesions can be seen in

the putamen on T1 sequences of brain MRI (Figure 2), although this finding is less sensitive [4]. The pathophysiology of this finding is unclear, although several suggestions have been made, including postmenopausal dopamine hypersensitivity or depletion of GABA in the brain [3,5]. This finding may also be related to reports of focal seizures in cases of non-ketotic hyperglycemia, which are hypothesized to have similar mechanisms [1]. Other diagnoses that should be considered with this symptom are Huntington's Disease,

Wilson's Disease, and Sydenham chorea. Monitoring for these diseases can be done via pedigree and DNA testing in the case of Huntington's Disease and serum copper, urine copper, and serum ceruloplasmin in Wilson's Disease. Sydenham chorea, which can be a manifestation of rheumatic fever, can be suggested by an elevated antistreptococcal antibody titer. Treatment consists of blood glucose management with hydration and insulin therapy while in the hospital and then optimization of diabetes management as an outpatient.

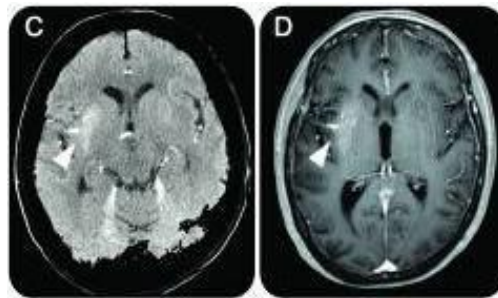


Figure 2. Examples of MRI enhancement of the putamen in other cases of hyperglycemia-induced chorea.

This case was reported to illustrate the fact that although MRI can sometimes help with the diagnosis of hyperglycemia-induced chorea, it is not consistently sensitive. In this instance, the patient presented with choreiform movements directly related to hyperglycemia without classic, previously described MRI findings which subsequently resolved immediately upon improvement in glycemic control. While MRI of the brain is warranted in suspected cases of hyperglycemia-induced chorea, it is not always diagnostic. Consideration should therefore be given to the diagnosis of non-ketotic hyperglycemia chorea in patients with new onset choreiform movements in the setting of significant hyperglycemia without classically described MRI findings.