

Hemichorea - A Rare Complication of Hyperglycemia

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All authors approved the submitted manuscript and are responsible for reported research. There are no conflicts of interest associated with the publication of the article. The corresponding author is João Pinto Machado, whose contacts follow:

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Abstract: *Hemichorea is characterized by involuntary non-rhythmic movements on one side of the body and can be due to brain vascular or traumatic insults and neoplasm. Hyperglycemic state is a rare, reversible cause of this condition. We present an 87-years-old-woman with a poorly controlled Type 2 Diabetes Mellitus, that complained of erratic, uncontrollable limb that lasted for 6 days. She had high glycemc values, despite her medication. The patient presented with pronounced choreiform movements of both arms and legs, more marked on the left hemibody. Her blood glucose level was 257mg/dL and had A1c hemoglobin test of 13.9%. A brain computerized tomography showed spontaneous hyperdensity at the basal nuclei, more evident at the right, suggestive of hyperglycemic hyperosmolar sequel. She was admitted with suspect of hyperglycemic hemichorea. Brain Magnetic Resonance that showed no evidence of acute ischemic lesions and showed loss of signal in T2- weighed acquisitions in areas in the internal capsulae of the striae region, compatible with paramagnetic substances deposition. She started an intensive insulin therapy with symptom improvement. Hyperglycemia results in the progression of cerebral ischemia and consequent injury to central nervous system and can justify the most common magnetic resonance image changes: hyperintense signals in the contralateral putamen on T1weighted images. Despite of its unclear mechanism, the role of glycemc control to ameliorate or treat the symptoms is proved and part of the diagnosis criteria. Hyperglycemic hemichorea is mostly a clinical diagnosis that is of crucial important as it's a reversible state and shouldn't be overlooked.*

Keywords: Chorea; Hemichorea; Hemyballismus; Hyperglycemia; Diabetes Mellitus

1. Introduction

The major causes of unilateral hemichorea–hemiballismus include brain vascular or traumatic insults and neoplasms. However, metabolic disturbances can be a reversible cause of this symptoms^{1,2}. Hyperglycemic state is an uncommon etiology of hemiballismus, but one to be aware of, as it's frequently misdiagnosed.

Hyperglycemic hemichorea is characterized by clinical, imaging and treatment criteria. Patients may present with involuntary random-appearing, irregular, movements on one side of the body and contralateral basal ganglia lesions on imaging study. These movements improve with glycemc control^{3,4}.

2. Case Report

An 87-years-old-woman with personal history of hypertension and Type 2 Diabetes Mellitus, presented in the emergency room complaining of erratic, uncontrollable limbs movements for 6 days.

She reported that both her arms and legs made involuntary movements, more pronounced on the left limbs, which she futilely tried to control.

She had an history of uncontrolled hyperglycemia, and although she was previously treated with oral antidiabetics, she had been put on insulin therapy 4 days prior to admission.

Her glycaemia values were improving, according to the patient, but self-monitoring identified several results above 300mg/dl.

She denied fever, respiratory, urinary or gastrointestinal complains.

On examination, the patient presented with pronounced choreiform movements of both arms and legs, more marked on the left hemibody. The movements were involuntary and non-rhythmic. It only affected the limbs, with no extension to facial mimic. Although it was difficult to evaluate, muscle strength seemed to be preserved bilaterally and no other focal neurological deficit was identified.

Her blood glucose level at time of admission was 257mg/dL. We could not find a recent value for A1c hemoglobin.

ER analytic study had no evidence of infection or hydroelectrolytic disorders.

A brain Computerized Tomography (CT) scan showed spontaneous hyperdensity at the basal nuclei, more evident at the right, suggestive of hyperglycemic hyperosmolar sequel (figure 1).

The patient was admitted with suspect of hyperglycemic hemichorea.

A more detailed study showed an A1c hemoglobin test of 13.9%.

She did a Cranial Magnetic Resonance that showed no evidence of acute ischemic lesions and showed loss of signal in T2- weighed acquisitions in areas in the internal capsulae of the striae region, compatible with paramagnetic substances deposition (figure 2 and 3).

She started an intensive insulin therapy scheme (20 units of isophane insulin twice daily, and 20 units of neutral insulin divided among 4 meals) and risperidone 0.5mg twice daily. Her symptoms improved over 6 days.

The patient was discharged 12 days later with no symptoms and prescribed NPH insulin 18 units in the morning and 16 units at night and risperidone 0,5mg twice daily.

3. Discussion

Although hyperglycemia is the most common metabolic cause of hemichorea, the clinical characteristics and mechanisms are still unclear⁵.

The main hypothesis suggests that depletion of gamma-aminobutyric acid (GABA) and acetylcholine, both used as an alternative energy source during nonketotic hyperglycemia, leads to a decreased inhibitory signal to the thalamus resulting in hyperactive movements. Though, there are some cases of hemichorea described in ketotic patient, which makes us think that this may not be the sole mechanism^{3,5}. Recently, the role of microvascular ischemia has been discussed. This theory proposes that hyperglycemia leads to an alteration of blood-brain barrier with altered transport of glucose, insulin, choline, amino acids and oxidative stress in central nervous system (CNS)

micro-capillaries. Hyperglycemia also results in the progression of cerebral ischemia and consequent injury to CNS⁶. Both conditions can justify the most common MRI changes: hyperintense signals in the contralateral putamen on T1 weighted images.

Despite of its unclear mechanism, the role of glycemic control to ameliorate or treat the symptoms is proved, and is part of the diagnosis criteria. Early recognition of this condition is also extremely important to improve prognosis. We present a case of a hyperglycemia chorea, with movements on both sides of the body, though more pronounced on the left body, compatible with the more intense hypersignal found on CT scan. The MRI scan excluded other causes of chorea. As expected, the patient improved with glycemic control.

4. Learning Points

Hyperglycemic hemichorea is a rare presentation of hyperglycemic state. It is mostly a clinical diagnosis that is of crucial importance as it's reversible. Physicians should think of this entity when presented with unilateral hemichorea-hemiballismus on a patient with uncontrolled hyperglycemia.

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Figure 1: Brain CT Scan

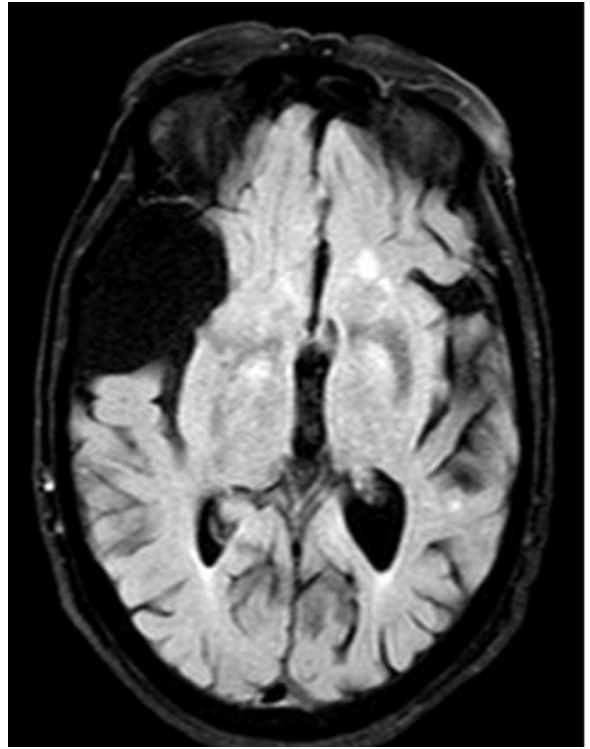


Figure 3: Brain MRI Scan

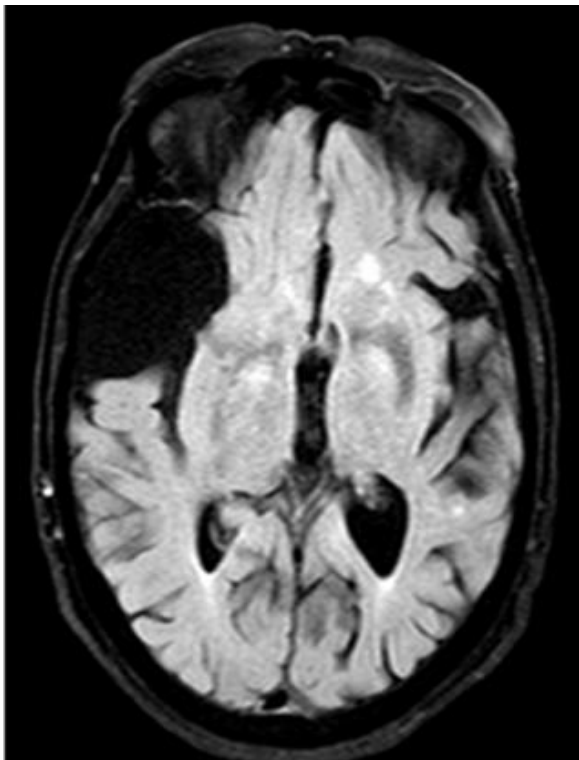


Figure 2: Brain MRI Scan