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Case Report

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Hemichorea as The First Manifestation of Type 2 Diabetes Mellitus: A Case Report

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ABSTRACT

Introduction: The development of dyskinetic movements in hyperglycemic states is a rare condition, and nonketotic hyperglycemia is an uncommon cause of hemichorea. The presence of these involuntary and abnormal movements as the first presentation of diabetes mellitus (DM) has been poorly described in the literature. Case report: A 53-year-old female patient arrives at the emergency department (ED) presenting an involuntary movement in the left upper limb that had been going on for three days. Conclusion: This case report alerts the medical community to pay close attention to this disorder and addresses the need for further studies to guide its treatment accordingly.

Keywords: Hyperglycemia; Dyskinesia; Case Report; Emergency Care

Introduction

Chorea is characterized as an involuntary, unilateral, or bilateral, irregular, and brief movement of the limbs. In some cases, ballism may also occur in muscles of the face, jaw, and tongue. Many pathologies can trigger this condition, such as neurodegenerative, infectious, and metabolic diseases.[1,2] The development of dyskinetic movements in hyperglycemic states is a rare condition, occurring in 1 over 100,000 patients. Nonketotic hyperglycemia is an uncommon cause of hemichorea/hemiballismus; the presence of these involuntary and abnormal movements as the first presentation of diabetes mellitus (DM) has been poorly described in the literature. Acknowledgment of this disease is important, considering that it is a treatable disorder with a good prognosis when recognized in time.

Due to the rarity of the event, the objective of this case report is to present the clinical history of a 53-year-old female patient without a previous diagnosis of type 2 DM, who was admitted to the Emergency Department of a hospital in the city of São Paulo with hemichorea in the left upper limb that had been going on for three days associated with non-ketone hyperglycemia,

Case Report

A 53-year-old female patient arrived at the emergency department (ED) presenting involuntary movements in the left upper limb that had been going on for three days, with insidious onset one day after using meloxicam 15 milligrams to treat lumbar pain. The movements gradually worsened in frequency and pain. However, it ceased when she held the limb, and during the night. Simultaneously, she reported a pulsatile frontal headache and some episodes of vomiting.

Previous medical history included hypertension, gout, insulin resistance, astigmatism, and a biological aortic valve replacement fourteen years before. Regular medications include diltiazem, olmesartan, chlorthalidone, and acetylsalicylic acid. In addition, she had a positive

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first-degree family history for type 2 DM.

When arriving at the physical examination, she was in good general condition, her blood pressure was 109 over 69 millimeters of mercury (mmHg), her heart rate was 100 beats per minute (bpm), and her oxygen saturation was 95%. Cardiovascular, pulmonary, and abdominal apparatus did not present relevant changes to the case. Capillary glucose demonstrates high (HI) error.

The neurology team reported that the only relevant sign on the neurological exam was arrhythmic chorea on the left hand and left forearm happening every 10 seconds. Her mental state, campimetry, strength, tonus, sensitivity, reflexes, coordination, equilibrium, and march were reported as normal. There were no meningeal or radicular signs. The Fundoscopy exam was not considered possible because of her degree of astigmatism.

The significant laboratory data were glucose 904 milligrams/dL (mg/dL) (reference range: 70-99mg/dL); glycated hemoglobin (HbA1C) 16.6 % (reference range 4,5-5,6%); type 1 urine with glycosuria 10.0; ketonuria 2 crosses (reference range: absent) and proteinuria 0.25 grams per liter (g/L) (reference range: inferior than 0,15 g/L) and total leukocytes of 11520 per milliliter (/mL) (reference range: less than 10.000/mL). Other altered laboratory findings included: total calcium 10.6 mg/dL (reference range: 8,6-10,2 mg/dL); creatinine 2.47 mg/dL (reference range: 0,5-0,9 mg/dL); alkaline phosphatase 125 units per liter (U/L) (reference range: 35-105 U/L); phosphorus 4.5; gamma glutamyl transferase 82 U/L (reference range: until 40U/L); aspartate aminotransferase 35 U/L (reference range: until 32 U/L); alanine aminotransferase 42 U/L (reference range: until 33 U/L); urea 104 mg/dL (reference range: 10-50); sodium 123 mmol/L (reference range: 137-148 mmol/L); Troponin T 40 (reference range: less than 50).

The patient was then submitted to cranial computerized tomography, which revealed that she had a calcified subcentimeter image close to the inner face of the skullcap in the frontal region, which could correspond to intracranial osteoma or calcified meningioma. In addition, the patient presented thickening of the left maxillary sinus (Image 1)

However, none of these findings matched her clinical condition. As no central neurological cause was found, the neurology team left the follow-up only with the emergency medicine team for the investigation and treatment of metabolic causes of chorea.

The initial approach was to start the hyperglycemia protocol, with 0.9% saline hydration and insulin in a continuous infusion pump. After partial improvement of the condition, a regular and Neutral Protamine Hagedorn (NPH) insulin regimen was started.

The patient had an improvement in chorea after hyperglycemia was resolved. Due to the first decompensation of DM, she was admitted to the Intensive Care Unit (ICU) for medical monitoring, glycemic control, and to receive instructions of the disease by the Endocrinology team.

The dyskinetic movement disorder was controlled with DM treatment, so after eleven days in the hospital, the patient was then discharged. After leaving the hospital, the patient was admitted to a neurology clinic, where an MRI was done on neurology follow-up, with no changes regarding the symptoms of chorea. She was then discharged from the neurology clinic a month after being hospitalized, with no need for neuroleptic medications.

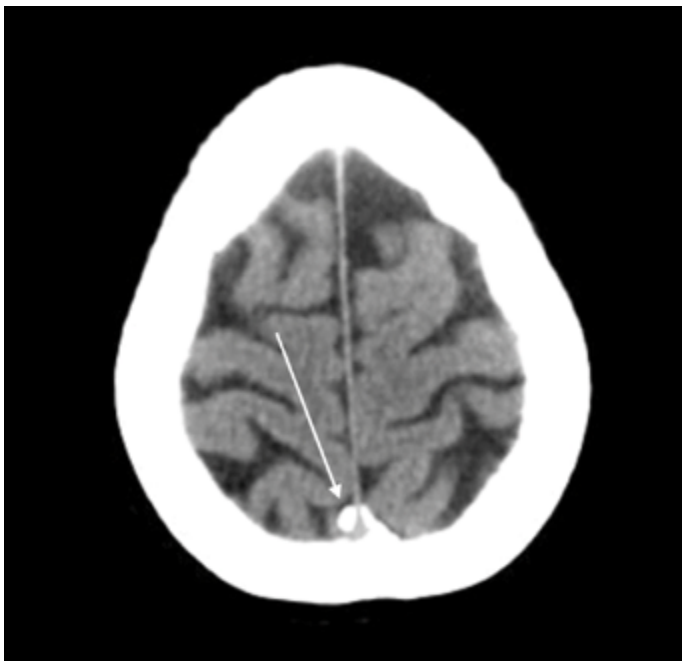

Image 1. Cranial computerized tomography

Discussion

The combination of hyperglycemic nonketotic hemichorea or hemiballism, with or without brain involvement in imaging studies, has been considered a unique syndrome and generally occurs as a complication of long-standing type 2 DM, but has also been described as the first manifestation of DM.[2] Other terms such as diabetic striatopathy (DS), diabetic hemiballism/hemichorea, and basal ganglia syndrome are used to describe the unusual presentation of hyperkinetic movements associated with hyperglycemic states.

According to the literature, the prevalence of this syndrome is reported as 1 in 100.000, which may be overestimated, since most doctors are unaware of this

condition.[4] There is a predisposition in the elderly (mean age 71.1), but with an age range between 15.9 to 67.6 years. The female sex is the most affected (56.6%) and about 96.6% of the patients had a previous diagnosis of type 2 DM. All reported cases had high levels of serum glucose (about 481 mg/dL) and HbA1c (about 14.4%). Most patients (88.1%) had hemichorea/hemiballism on the arrival and about 73% had a complete recovery.[5]

Characteristic radiological findings are hyperdensity on CT and hyperintensity in T1-weighted magnetic resonance imaging (MRI) in the contralateral striatum of the presentation.[2] Chorea and image changes did not appear concurrently in about one-tenth of the patients. The origin of ballistic movements in chorea may be related to a reduction in pallidal activity due to lesions in the subthalamic, caudate, and putamen nuclei, with consequent loss of thalamic inhibition and over-activation of the direct path.[6] Some systemic changes, such as systemic lupus erythematosus, Wilson's disease, and non-ketotic hyperglycemia can also cause the movement disorder.

The patient in question had non-ketotic hyperglycemia as the main cause for the hyperkinetic disorder. It is believed that in this condition, the decrease in brain levels of gamma-aminobutyric acid (GABA) has an important role in the pathogenesis of hemiballism. The hyperglycemic state causes changes in anaerobic metabolism, which then leads to a secondary decrease in GABA, which acts as an alternative energy substrate during anaerobiosis, contributing to the dysfunction of the basal ganglia. In ketotic cases, GABA can be resynthesized, which explains the lack of association between ballism/chorea and diabetic ketoacidosis. In addition, hyperkinetic disorder can also be attributed to reduced blood flow in the brain ganglia, petechial hemorrhage, acetylcholine depletion, or metabolic acidosis.[5]

The patient's clinical improvement was obtained after controlling blood glucose levels, which corroborates that the neurological changes were secondary to the metabolic disorder. Despite previous studies indicating that correction of hyperglycemia generally resulted in complete or partial resolution of the condition, specific anti-chorea drugs may be needed as an adjuvant in non-respondents' patients.

Conclusion

Nonketotic hyperglycemic state is a systemic and unusual condition that can mitigate neurological conditions, such as hemichorea/hemiballism, as reported in this case.

Hemichorea is a condition that can lead to a variety of treatment options, therefore, it's important to recognize the differential diagnoses to provide the best medical care and correct treatment, assuring patient safety.

Because of its rarity, there is a lack of well-established guidelines to treat the condition, also, there are few studies about this matter in the literature. This case report alerts the medical community to pay close attention to this disorder and addresses the need for further studies to guide its treatment accordingly.

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