

Hemichorea associated with non-ketotic hyperglycemia: report of two cases and brief review of the literature

Hemicoreia associada à hipoglicemia não cetótica: Relato de dois casos e breve revisão da literatura

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Abstract

Introduction: Chorea is characterized as a hyperkinetic movement disorder that is caused by lesions on the basal ganglia. Many etiological factors have been described, including ischemia, infection, neoplasm and neurodegenerative disorders, such as Huntington's disease. Non-ketotic hyperglycemia induced chorea is a rare, but reversible, condition usually associated with contralateral striatal radiological abnormalities. **Case Report:** Case 1: A 64 year-old Brazilian woman was admitted concerning about a 4-day history of involuntary, erratic and continuous movements with sudden onset affecting the left arm, leg and, partially, the left hemiface. Her past medical history was remarkable for arterial hypertension and type 2 diabetes mellitus. Neurologic examination revealed the presence of choreic movements of the left hemiface, arm and leg. Case 2: A 63 year-old Brazilian woman was admitted concerning about an 1-day history of involuntary, without purpose, assymmetric, irregular and abrupt movements of the upper limbs and face associated with difficulty of speaking and swallowing. Her past medical history was remarkable for arterial hypertension and coronariopathy with two episodes of acute myocardial infarction four months before. There was no history of diabetes mellitus. On neurological examination, she presented severe dysarthry and difficulty of swallowing associated with choreic movements of the upper limbs and face. **Conclusion:** In the present study, we describe two cases of chorea as a complication of non-ketotic hyperglycemia. A high index of clinical awareness and early suspicion of this possible association are extremely important, once the correct management and normalization of glycemia may result in complete recovery of neurologic symptoms.

Descriptors: Chorea; Hyperglycinemia Nonketotic.

Resumo

Introdução: Coréia é caracterizada como um distúrbio de movimento hiperkinético causado por lesão dos gânglios da base. Muitos fatores etiológicos tem sido descritos, incluindo isquemia, infecção, neoplasia e desordens neurodegenerativas, como a Doença de Huntington. Hiperglicemia não cetótica induzindo coréia é uma condição rara, embora reversível, usualmente associada a anormalidades radiológicas estriatais contralateral. **Relato de Caso:** Caso 1: Mulher brasileira de 64 anos foi admitida com queixa de ha 4 dias apresentar movimentos involuntários, aleatórios e contínuos de início súbito afetando braço, perna e parcialmente a face, a esquerda. Portadora de HAS e DM tipo 2. No exame neurológico apresentava movimentos coreicos em braço, perna e face a esquerda. Case 2: Mulher brasileira de 63 anos com queixa ha 1 dia de movimentos involuntários, assimétricos, sem propósitos, irregular e abruptos em membros superiores e face associado a disartria e disfagia. Portadora de HAS e coronariopatia com IAM ha 4 meses atrás, sem historia de DM. Ao exame neurológico, apresentava severa disartriam, disfagia, associado a movimentos coreicos em membros superiores e face. **Conclusão:** No presente estudo, nós descrevemos dois casos de coréia como complicação de hiperglicemia não cetótica. A percepção clinica e a suspeita precoce da possibilidade da associação são extremamentes importantes, uma vez que o correto manejo e a normalização da glicemia podem resultar na complete resolução dos sintomas neurologicos.

Descritores: Coréia; Hiperglicinemia não Cetótica.

Introduction

Hemichorea is defined as an involuntary, erratic, abrupt and non-stereotyped hyperkinetic movement disorder in one side of the body varying on intensity and topography⁽¹⁻⁴⁾. It can be caused by a wide variety of etiologies, including ischemia,

infection, neoplasm and Huntington's disease. Non-ketotic hyperglycemia induced chorea is a rare, but reversible, condition associated with contralateral striatal radiological abnormalities usually resolving within 6 months following correction of hyperglycemia⁽⁵⁻⁶⁾.

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Conflito de interesses: Não

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In the present report, we describe two cases of type 2 diabetic patients manifesting chorea as a complication of non-ketotic hyperglycemia and discuss the current knowledge about possible pathophysiological mechanisms and treatment.

Case Report

Case 1

A 64 year-old Brazilian woman was admitted concerning about a 4-day history of involuntary, erratic and continuous movements with sudden onset affecting the left arm, leg and, partially, the left hemiface. She reported that the movements ceased during sleep and progressively worsened until hospitalization. Her past medical history was remarkable for arterial hypertension and type 2 diabetes mellitus. On clinical assessment, blood pressure was 180x110mmHg, no cardiac murmur sounds and lungs clear. Neurologic examination revealed the presence of choreic movements of the left hemiface, arm and leg. The remaining of neurologic exam was essentially normal. Laboratorial blood exams revealed: Hemoglobin 13; Hematocrit 42%, leucocytes 11800, reactive protein-C 0,6, K:3,8, Na:132, creatinine 1,2, glucose 485mg/dl. Brain computed tomography revealed hyperdense signal on the right putamen and caudate. Brain magnetic resonance showed hyperintense signal on T1-weighted image on the right putamen and caudate. She had the blood-glucose level controlled with insulinotherapy and presented a remarkable improvement on involuntary movements. After one week the patient was completely symptom free.

Case 2

A 63 year-old Brazilian woman was admitted concerning of a 1-day history of involuntary, without purpose, asymmetric, irregular and abrupt movements of the upper limbs and face associated with difficulty of speaking and swallowing. She reported that the erratic movements disappeared during sleep and worsened during emotional stress with greater intensity on the left side. She also referred to had presented similar symptoms seventeen days before. Her past medical history was remarkable for arterial hypertension and coronariopathy with two episodes of acute myocardial infarction four months before. There was no history of diabetes mellitus. On clinical assessment, blood pressure was 140x90 and cardiac systolic murmur sounds were present. On neurological examination, she presented severe dysarthry and difficulty of swallowing associated with choreic movements of the upper limbs and face. The remaining of neurologic exam was essentially normal. Laboratorial blood exams revealed: Hemoglobin 12,5; Hematocrit 38%, leucocytes 6400, reactive protein-C 0,4, K:3,9, Na:136, creatinine 1,0, glucose 327mg/dl. Brain computed tomography revealed hyperdense signal on the putamen and caudate bilaterally. Brain computed tomography showed hyperdense signal on the putamen and globus pallidus bilaterally (Figure 1). She had the blood-glucose level controlled with insulinotherapy and presented a remarkable improvement on involuntary movements. After four weeks the patient was completely symptom free.

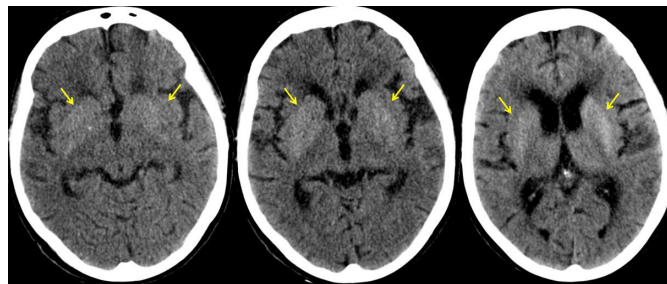


Figure 1. Brain computed tomography image showing hyperdense signal on the putamen and globus pallidus bilaterally. São José do Rio Preto/SP, 2015

Discussion

The hemichorea is a hyperkinetic movement disorder and has been described as one of the semiologic signs that result from lesions in the subthalamic nucleus, corpus striatum, cerebral cortex, thalamus, and brainstem⁽⁷⁻⁹⁾. The nature of the lesion varies considerably and can be a consequence of vascular disease, infections, drugs, metabolic abnormalities, neurodegenerative diseases, immunologic disorders and tumors⁽¹⁰⁾. Therefore, the focus of neurologic diagnostic relies on the investigation of clinical features from the patient, its epidemiological nuances and previous risk factors.

Generally, chorea is classified as primary, when a responsible gene is identified or no other etiological cause is defined; and secondary, when it results from a preliminary condition. Mendes and colleagues, in 1996, reported that, in Brazil, the most frequent cause of chorea is Sydenhan's chorea. Patients over the age of 50 years represent around 15% of all patients presenting chorea and metabolic abnormalities is a rare related cause in Brazil⁽¹¹⁾. Non-ketotic hyperglycemia is a metabolic disturbance on the glucose balance and is usually associated with neurological manifestations, such as changes on mental status, seizures and motor deficits; however it is an uncommon, but reversible, cause of chorea in type 2 diabetes mellitus patients^(5-6, 10, 12-15).

Many hypotheses have been raised to explain the pathological mechanisms involving chorea and hyperglycemia, including vascular insufficiency, dopaminergic hyperactivity, depletion of gamma-aminobutyric acid (GABA) and acetylcholine, acute dysfunction secondary to hyperglycemic or hyperosmolar insult, hyperviscosity, petechial hemorrhage and genetic predisposition^(6, 16-20). Elder female patients with type 2 diabetes mellitus from East Asian are more prone to develop chorea, suggesting a possible genetic predisposition⁽¹⁶⁾. Acanthocytes in circulating peripheral blood have been described as a predisposing factor to chorea in patients with non-ketotic hyperglycemia⁽²¹⁾. In the present reports, we found no acanthocytes in circulating peripheral blood.

Neuroimaging findings also contribute to the debate about the pathophysiology of chorea in patients with non-ketotic hyperglycemia. The majority of reported cases show a high signal on T1-weighted magnetic resonance imaging in the putamen, globus pallidus and caudate contralateral to the involuntary movement⁽²²⁾. Occlusive vasculopathy of the arterioles with patchy necrosis, neovascularization and perivascular inflammation on

the striatum and lentiforme nucleus may be responsible for the radiological abnormalities observed in patients with non-ketotic hyperglycemia and chorea⁽²³⁾. In our reported cases, we noticed a precise correlation of the site of radiological change to the contralateral clinical manifestation of involuntary movements.

Conclusion

The present study highlights that hemichorea is a rare, but potentially reversible cause of non-ketotic hyperglycemia. A high clinical awareness and early suspicion contribute to a precocious diagnosis and treatment.

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