

MOVEMENT DISORDER CAUSED BY HYPEROSMOLAR HYPERGLYCEMIC STATE IN AN OLDER ADULT: A CASE REPORT

Distúrbio de movimento por conta de estado hiperosmolar não cetótico em idosa: relato de caso

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ABSTRACT

Diabetes mellitus (DM) and its complications constitute the leading causes of early mortality in most countries. Population aging and the growing prevalence of obesity and sedentary lifestyles, in addition to spreading urbanization, are considered the main drivers of the increasing incidence and prevalence of DM worldwide. This case report describes the acute onset of movement disorder in an older woman secondary to hyperosmolar hyperglycemic state (HHS). The combination of hemichorea–hemiballismus, HHS, and evidence of basal ganglia involvement on neuroimaging is considered a unique syndrome. Movement disorders secondary to HHS respond satisfactorily to administration of neuroleptic agents and proper glycemic control. The lack of published studies on this pathologic entity may lead to clinical and laboratory underdiagnosis, with negative impacts on patient prognosis and follow-up.

KEYWORDS: aged; hyperglycemic hyperosmolar nonketotic coma; dyskinesias; diabetes mellitus.

RESUMO

O diabetes e suas complicações constituem as principais causas de mortalidade precoce na maioria dos países. O envelhecimento da população e a crescente prevalência da obesidade e do sedentarismo, além dos processos de urbanização, são considerados os principais fatores responsáveis pelo aumento da incidência e da prevalência do diabetes *mellitus* (DM) em todo o mundo. Este relato de caso objetiva descrever a presença de distúrbio do movimento em idoso por conta do estado hiperosmolar não cetótico. A combinação de hemicoreia-hemibalismo, hiperglicemia não cetótica e envolvimento dos gânglios da base em exames de imagem é considerada uma síndrome única. Os distúrbios do movimento em estado hiperosmolar não cetótico apresentam resposta terapêutica satisfatória com o uso de neurolépticos e controle glicêmico adequado. A escassez de trabalhos publicados proporciona subdiagnósticos clínico e laboratorial, interferindo no prognóstico e no acompanhamento dos pacientes.

PALAVRAS-CHAVE: idoso; coma hiperglicêmico hiperosmolar não cetótico; discinesias; diabetes *mellitus*.

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Received on: 02/22/2019. Accepted on: 05/13/2019

DOI: 10.5327/Z2447-211520191900015

OBJECTIVE

To describe the natural history of hemichorea–hemiballismus secondary to hyperosmolar hyperglycemic state in an older woman, triggered by chronic decompensation of type 2 DM.

CASE REPORT

A 68-year-old brown woman from a rural area presented to the outpatient geriatrics clinic with sudden onset of involuntary movements in the upper and lower right limbs.

The patient had a history of longstanding type 2 DM and hypertension. Her current medications, to which she was irregularly adherent, included metformin 2550 mg/day, gliclazide MR 30 mg/day, NPH insulin 10 U, losartan 100 mg/day, amlodipine 10 mg/day, hydrochlorothiazide 25 mg/day, and atenolol 100 mg/day. She did not report any history of alcoholism or smoking, and denied any history of movement disorder in her family.

On physical examination, the patient was alert, oriented, cognitively intact, and denied a history of falls. She complained of feeling sad since the onset of illness, especially due to impaired mobility. Neurological examination revealed large-amplitude, non-rhythmic, hyperkinetic involuntary movements of the right limbs, with a proximal predominance.

WORKUP

Blood tests were remarkable for a fasting glucose of 330 mg/dL and glycosylated hemoglobin of 9.9%. No other abnormalities were identified, and the hypothesis of infection was ruled out. Brain magnetic resonance imaging (MRI) showed bilateral hyperintensities in the basal ganglia (Figure 1).

A diagnosis of hemichorea–hemiballismus syndrome secondary to hyperosmolar hyperglycemic state was suspected. The patient's antidiabetic regimen was optimized, and haloperidol and lorazepam were prescribed. After 2 weeks of

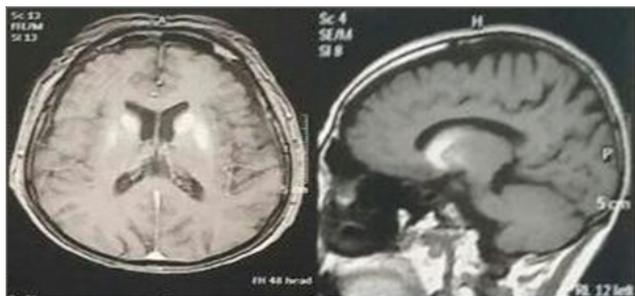


Figure 1 Brain magnetic resonance imaging (MRI) showing bilateral hyperintensities in the basal ganglia.

treatment, a reduction in hyperkinetic movements was evident. At 1-month follow-up, there had been significant further improvement. Glycemic control was also improved. Control brain MRI (Figure 2) performed at 6-month follow-up showed a slight signal abnormality affecting part of the left striatum, consistent with sequelae of a remote metabolic disturbance.

At the time of writing, the patient's medication regimen consisted of alogliptin/metformin 12.5/500 mg twice daily, gliclazide 90 mg/day, haloperidol 1 mg/day and lorazepam 1 mg/day, with a plan for progressive withdrawal and ultimate discontinuation of the neuroleptic agents. Her glycemic control was satisfactory and involuntary movements had been all but abolished, with only slight dyskinesia of the right foot remaining.

The patient provided written informed consent for publication. This report was approved by the Research Ethics Committee of Universidade Estadual de Montes Claros (Unimontes) with opinion number 95299218.9.0000.5146.

DIFFERENTIAL DIAGNOSIS

Ballism (or ballismus) and chorea are hyperkinetic movement disorders distinguished by the amplitude and distribution of affected muscle groups. They may occur at rest or on exertion, and tend to disappear during sleep.¹ These disorders tend to occur as a result of structural injury to the subthalamic nucleus and contralateral striatum. Etiology is variable, but vascular events are especially common.² In the absence of focal vascular injury, metabolic disorders, cerebral neoplasm, and central nervous system (CNS) infections should be considered.³

In the hemichorea–hemiballismus syndrome associated with hyperosmolar hyperglycemic state, neuroimaging often shows striatal hyperdensity (CT) or hyperintensity (T1-weighted MRI).⁴ The main differential diagnoses of these findings are calcifications and hemorrhage.³

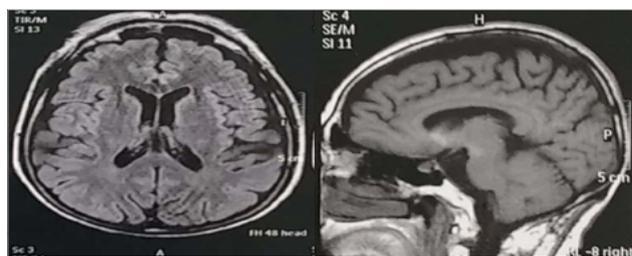


Figure 2 Brain magnetic resonance imaging (MRI) performed at 6-month follow-up showing a slight signal abnormality affecting part of the left striatum.

DISCUSSION

Decompensated diabetes mellitus can trigger a variety of neurological complications. However, movement disorders are rare. In older adults, HHS (formerly known as hyperosmolar hyperglycemic nonketotic coma or hyperosmolar hyperglycemic nonketotic syndrome) may manifest with ballistic-choreic movements. This movement disorder, which has been termed hemichorea–hemiballismus syndrome, constitutes a unique pathologic entity which has been reported only rarely in the scientific literature.⁵ The prevalence of this condition is estimated at less than 1 case per 100,000 population.^{5,6}

Hemichorea–hemiballismus secondary to nonketotic hyperglycemia was first described by Bedwell in 1960.⁷ This syndrome is uniquely associated with HHS, usually occurring at blood glucose levels greater than 300 mg/dL, or glycated hemoglobin (HbA1c) concentrations equal to or greater than 14 mg/dL.^{8,9}

The following criteria must be present to diagnose this syndrome:⁴

- choreiform or ballistic movements of at least two of the following regions: unilateral face, neck, upper limb or lower limb;
- markedly elevated blood glucose level;
- a hyperdense or hyperintense lesion in the contralateral striatum on head CT or brain MRI;
- abrupt cessation of dyskinesia once glycemic control is achieved;
- no evidence of stroke, infection, or inflammatory lesions on CT or MRI; and
- no evidence of other metabolic derangement, drug use, or known history of degenerative disorder.

Women over age 45 are at the highest risk of developing this complication, possibly due to overexpression and hypersensitization of dopaminergic receptors during menopause, as a result of reduced estrogen levels and increased susceptibility to injury during metabolic derangements.⁸ The cerebral changes that occur during HHS remain unclear; however, hypotheses related to energy metabolism and ischemic injury have been proposed.¹⁰

Clinical and imaging findings are potentially reversible and, indeed, usually resolve within 2 to 12 months after treatment of hyperglycemia.^{11,12} In our patient, neuroimaging suggested a metabolic etiology of her movement disorder, with evidence of metabolic disturbance remaining on control imaging even after achievement of blood glucose control and regression of the initial lesions.

The prognosis is good. Glycemic control is the cornerstone of treatment. If dyskinesia persists, medications such as neuroleptics, benzodiazepines, and antiepileptics may be helpful.^{13,14} Recurrence has been reported in up to 17% of cases, particularly as a result of protracted hyperglycemia or irregular adherence to pharmacotherapy. Surgical interventions, such as thalamotomy and deep brain stimulation, are rarely considered.¹

This case report highlights a complication that requires early diagnosis and immediate treatment because of the high associated morbidity. In conclusion, movement disorder secondary to hyperosmolar hyperglycemic state is a unique syndrome with excellent response to glycemic control and, when indicated, appropriate psychotropic agents.

REFERENCES

1. Block H, Scozzafava J, Ahmed SN, Kalra S. Uncontrollable movements in patient with diabetes mellitus. *CMAJ*. 2006;175(8):871. <https://doi.org/10.1503/cmaj.060541>
2. Coral P, Teive HAG, Werneck LC. Hemibalismo: relato de 8 casos. *Arq Neuropsiquiatr*. 2000;58(3A):698-703. <http://dx.doi.org/10.1590/S0004-282X2000000400016>
3. Chang MH, Chiang HT, Lai PH, Sy CG, Lee SS, Lo YY. Putaminal petechial hemorrhage as the cause of chorea: a neuroimaging study. *J Neurol Neurosurg Psychiatry*. 1997;63(3):300-3. <https://doi.org/10.1136/jnnp.63.3.300>
4. Lin JJ, Lin GY, Shih C, Shen WC. Presentation of striatal hyperintensity on T1-weighted MRI in patients with hemiballismus-hemichorea caused by non-ketotic hyperglycemia: report of seven new cases and a review of literature. *J Neurol*. 2001;248(9):750-5.
5. Zetola VF, Verschoor B, Lima FM, Ottmann FE, Doubrawa E, Paiva E, et al. Hemibalismo-hemicoreia em estado hiperglicêmico não cetótico: distúrbio do movimento associado ao diabetes melito. *Arq Bras Endocrinol Metab*. 2010;54(3):335-8. <http://dx.doi.org/10.1590/S0004-27302010000300014>
6. Ondo W. Hyperglycemic nonketotic states and other metabolic imbalances. In: Tolosa WWE, ed. *Hyperkinetic Movement Disorders*. Amsterdam: Elsevier; 2011. p. 768.
7. Bedwell SF. Some observations on hemiballismus. *Neurology*. 1960;10:619-22. <https://doi.org/10.1212/wnl.10.6.619>
8. Lin JJ, Chang MK. Hemiballismus-hemichorea and non-ketotic hyperglycemia. *J Neurol Neurosurg Psychiatry*. 1994;57(6):748-50. <https://doi.org/10.1136/jnnp.57.6.748>
9. Oh SH, Lee KY, Im JH, Lee MS. Chorea associated with non-ketotic hyperglycemia and hyperintensity basal ganglia lesion on T1-weighted brain MRI study: a meta-analysis of 53 cases including 4 present cases. *J Neurol Sci*. 2002;200(1-2):57-62.
10. Guisado R, Arieff AI. Neurological manifestations of diabetic comas: correlation with biochemical alterations in the brain. *Metabolism*. 1975;24(5):665-79.
11. Bekiesinska-Figatowska M, Mierzewska H, Jurkiewicz E. Basal ganglia lesions in children and adults. *Eur J Radiol*. 2013;82(5):837-49. <https://doi.org/10.1016/j.ejrad.2012.12.006>
12. Chokshi FH, Aygun N, Mullins ME. Imaging of acquired metabolic and toxic disorders of the basal ganglia. *Semin Ultrasound CT MR*. 2014;35(2):75-84. <https://doi.org/10.1053/j.sult.2013.09.006>
13. Kranick SM, Price RS, Prasad S, Hurtig HI. Clinical reasoning: a 52-year-old woman with subacute hemichorea. *Neurology*. 2008;71(20):e59-62. <https://doi.org/10.1212/01.wnl.0000334759.72146.39>
14. Sitburana O, Ondo WG. Tetrabenazine for hyperglycemic-induced hemichorea-hemiballismus. *Mov Disord*. 2006;21(11):2023-5. <https://doi.org/10.1002/mds.21100>