CASO CLÍNICO/CASE REPORT

A Peculiar Diabetic Striatopathy: A Case Report with Normal Basal Ganglia Imaging

Estriatopatia Diabética Peculiar: Um Caso Sem Alterações de Neuroimagem nos Gânglios da Base

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Abstract

Diabetic striatopathy is a rare entity typically associated with signal changes on brain magnetic resonance imaging (MRI) at the level of the basal ganglia. We report a case of a 67-year-old woman with a history of type 2 diabetes mellitus who presented to the emergency department due to the acute onset of hemiballistic movements. The laboratory work-up revealed hyperglycemia without ketoacidosis. A brain MRI (3T) revealed no anomalies in the basal ganglia. Symptomatic treatment and correction of glucose levels led to a favorable outcome. We assumed a presumptive diagnosis of diabetic striatopathy. We hypothesize that the absence of radiological lesions in the basal ganglia could be due to micro-hemorrhages or ischemic lesions below the detection threshold. In an appropriate clinical context, the lack of clinical-radiological concordance should not prevent the diagnosis of diabetic striatopathy.

Resumo

A estriatopatia diabética é uma entidade rara normalmente associada a alterações de sinal na ressonância magnética (RM) cerebral a nível dos gânglios da base. Reportamos o caso de uma mulher de 67 anos com antecedentes de diabetes *mellitus* tipo 2 que consultou o serviço de urgência por movimentos hemibalísticos de instalação súbita. As investigações realizadas revelaram hiperglicemia sem cetoacidose. Uma RM cerebral (3T) não revelou anomalias nos gânglios da base. O tratamento sintomático e a correção dos níveis glicémicos resultaram numa evolução favorável. Assumimos um diagnóstico presumível de estriatopatia diabética. A ausência de lesões radiológicas nos gânglios da base poderá ser devida a micro-hemorragias ou lesões isquémicas abaixo do limiar de detecção. Em contexto clínico apropriado, a falta de concordância clínica-radiológica não deverá impedir o diagnóstico de estriatopatia diabética.

Introduction

Diabetic striatopathy is a rare condition caused by poor glycemic control, typically occurring in cases of acute hyperglycemia ranging from 500 to 1000 mg/dL, without accompanying ketoacidosis. It can present as hyperkinetic movement disorders, including both chorea and ballism. Ballism is characterized by large-amplitude, forceful flinging movements, typically affecting one side of the body. The causal mechanism remains uncertain, although previous reports hypothesize a mechanism related to the presence of reactive astrocytes at the level of the basal ganglia.[1] Chorea refers to random, uncontrollable, involuntary jerking movements, typically more distal and of smaller amplitude than ballism. Brain magnetic resonance imaging (MRI) usually reveals signal changes, particularly in the putamen and/or caudate nucleus, contralateral to the symptomatic side. The most reported imaging finding is T1 hyperintensity, followed by striatal hypointensity on gradient echo-weighted (T2*-GRE) and susceptibility-weighted imaging (SWI).[1,2]

Case Report

We describe the case of a 67-year-old woman with a history of dyslipidemia, hypertension, and poorly controlled type 2 diabetes mellitus with known microvascular complications (diabetic retinopathy and diabetic nephropathy on dialysis), without known macrovascular complications.

She presented to the emergency department (ED) with a sudden onset of hyperkinetic movements affecting the left hemibody, which began approximately 6 hours prior. She denied experiencing headaches. Her medication had not been changed recently, and she denied any drug use. Her usual medications included insulin glargine (28 units in the morning) along with a rapid insulin correction regimen, atorvastatin 40 mg once daily, and perindopril/amlodipine 5 mg/5 mg daily. The most recent dialysis session had occurred the previous day without complications and with no changes to the dialysis prescription or parameters.

According to previous records, the diagnosis of type 2 diabetes mellitus was formally established at the age of 42, when she entered menopause. She continued hormone replacement therapy for 7 years, after which she developed increasing insulin therapy needs, with HbA1c levels consistently above 7.5% since the age of 57. The patient had missed several primary care appointments and had been lost to endocrinology follow-up for the same reason, and she sometimes forgot to administer insulin or did not use the correct dose.

Upon examination, her heart rate was 105 bpm, and her blood pressure was 158/105 mmHg. The patient was sweaty and nauseated but appeared calm and oriented to person, place, time, and situation. She exhibited sudden and irregular involuntary movements, predominantly involving the proximal muscles of the left upper and lower limbs. These movements were characterized by large-amplitude, flinging and throwing-like motions, lacking any purposeful goal, consistent with the classical presentation of hemiballismus. She showed no deficits in muscle strength, and her pain sensitivity was normal. There were no other focal neurological findings, no signs of meningismus, and the general examination was otherwise unremarkable. The arteriovenous fistula in her left arm showed no signs of hemorrhage or infection.

Arterial blood gas analysis upon arrival showed normal pH, pCO2, pO2, bicarbonate, and lactate levels, but glucose was elevated at 475 mg/dL. Cardiac infarction was excluded based on electrocardiogram (ECG) findings and two consecutive sets of troponin measurements within normal limits. The complete blood count revealed a leukocyte count of 11 400 cells/mm³ (normal range: 3600-10 500 cells/mm³), with normal levels of hemoglobin, platelets, international normalized ratio (INR), sodium, potassium, calcium and magnesium. Laboratory tests showed elevated creatinine at 4.67 mg/dL (normal range: 0.55–1.02 mg/dL), urea at 65 mg/dL (normal range: 15–50 mg/dL), and low albumin at 2.9 g/dL (normal range: 3.5–4.8 g/dL), values that are commonly observed in patients undergoing maintenance hemodialysis and not considered unexpected in this setting. Total protein was also below normal at 5.1 g/dL (normal range: 6.6-8.3 g/dL). Liver function tests showed slightly elevated levels of aspartate aminotransferase (35 U/L, normal range: <31 U/L) and alkaline phosphatase (74 U/L, normal range: 30-120 U/L), while alanine aminotransferase, gamma-glutamyl transferase, and ammonia levels were within normal limits. Total bilirubin was elevated at 2.6 mg/dL (normal range: 0.2-1.8 mg/dL), with direct bilirubin also elevated at 1.2 mg/dL (normal range: <0.5 mg/dL). HbA1c was notably high at 9.7%. Thyroid tests (TSH and free T4) were normal. Ceruloplasmin and ferritin values were also within normal range. Antinuclear and antithyroid antibodies were

negative. The antiphospholipid syndrome was excluded. Treponemal testing, human immunodeficiency virus (HIV), quantiferon test, hepatitis B virus, and hepatitis C virus screenings were negative. Blood cultures were negative. Urinary cultures demonstrated the presence of amoxicillin/clavulanic acid-sensitive *Escherichia coli* with counts greater than 100 000 colonies/mL. Chest x-ray was unremarkable.

Brain computed tomography (CT) scan and angio-CT were performed in the emergency department (ED) and revealed mild signs of periventricular ischemic white matter disease and moderate cortical atrophy with enlargement of the cerebrospinal fluid (CSF) spaces, consistent with the degree of atrophy. No ischemic or hemorrhagic lesions in the basal ganglia were visualized. No intra-arterial thrombi or stenoses with hemodynamic significance were visualized in the anterior or posterior circulation. Perfusion maps were not performed at admission.

Although central nervous system infections and autoimmune/paraneoplastic causes were considered unlikely, a lumbar puncture was performed as part of the initial workup to exclude these conditions in a diabetic patient with an acute onset of abnormal movements and no clear explanation for her clinical findings, despite low inflammatory markers. CSF analysis was unremarkable, and bacterial and viral PCR screening for meningitis/encephalitis were negative. A 3T brain MRI was conducted on the third day of hospitalization and included the following sequences: T1, T2, T2 FLAIR, GRE, diffusion-weighted imaging (DWI), and apparent diffusion coefficient (ADC) (**Fig. 1**). No hyperintensities were visualized on T1 sequences. Hyperintensities were seen in the fluid-attenuated inversion recovery (FLAIR) sequence, located in the external capsule and subcortical region bilaterally, corresponding to a moderate degree of periventricular ischemic white matter disease. In the susceptibility-weighted imaging (SWI) and gradient echo sequences (T2*-GRE), hypointensities in the basal ganglia were not visualized. The panel of antibodies for autoimmune and paraneoplastic encephalitis was requested, including anti-cell surface antibodies (anti-NMDAR, anti-AMPA 1/2, anti-DPPX, anti-CASPR2, anti-LGI1, anti-GABABr) and intranuclear antibodies (anti-Hu, anti-Ri, anti-Yo, anti-amphyphisin, anti-Ma2, anti-CMRP-5, anti-recoverin, anti-SOX1, anti-titin, anti-Zic4, anti-GAD65, anti-Tr(DNER)) and was negative. An EEG was performed to exclude epileptic activity as a potential cause of the acute hyperkinetic movements. Although the movements did not appear paroxysmal and the patient exhibited no focal neurological deficits or signs of status epilepticus, differentiating hemiballismus from focal motor seizures or epilepsia *partialis continua* was important to guide appropriate management. Carotid and vertebral artery Doppler and two-dimensional transthoracic echocardiography were also normal.

Insulin was administered in the ED with correction of glycemic values and monitoring of potassium values. Antibiotic therapy with amoxicillin/clavulanic acid was started. Symptomatic treatment of involuntary movements was started with oral haloperidol 0.5 mg three times a day (TID) without response. During hospitalization, a switch from neuroleptics to chlorpromazine 25 mg TID titrated to 50 mg TID was attempted, with an excellent response on the fifth day of hospitalization.

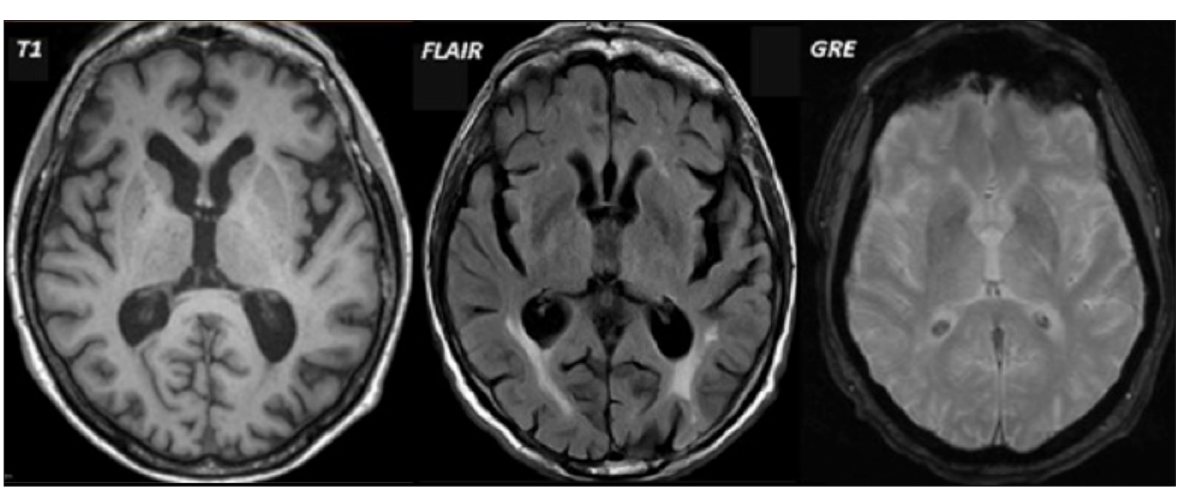

Figure 1. Brain MRI, axial section, from left to right: T1-weighted data, FLAIR and GRE sequences.

Glycemic values were progressively corrected with glycemic adjustment to long-acting insulin 32 U in the morning plus fast-acting human insulin 4U before the meals, accompanied by resolution of the lower urinary tract infection.

The patient was discharged home on the 15th day of hospitalization, presenting with mild hemiballismus with little impact on daily activities. A one-month follow-up consultation confirmed a favorable outcome, with complete resolution of symptoms and successful discontinuation of chlorpromazine.

Discussion

Our patient represents a typical case of type 2 diabetes mellitus with poor metabolic control. As described in the literature, menopause is associated with a decrease in estrogen levels and an increase in insulin resistance, with hormone therapy likely contributing to the stabilization of the glycemic profile during the first years after diagnosis. Additionally, our patient had an acute infection responsible for the non-ketotic hyperglycemia, which is frequently described in diabetic patients alongside diabetic ketoacidosis and hyperosmolar coma. On the other hand, chronic kidney failure under dialysis is known to be associated with extra-renal microvascular disease, also predisposing to cerebral events.[3,5] Additionally, in hemodialysis, there may be other causes of hemichorea not related to diabetes, recently described such as uremic toxin retention, electrolyte disturbances and anemia.[5]

Diabetic striatopathy is a rare entity resulting from acute hyperglycemia without metabolic acidosis, manifested by unilateral choreiform movements. Blood glucose levels and neuroimaging findings are paraclinical tests that support this diagnosis. Hemiballismus is often caused by a lesion in the contralateral subthalamic nucleus. Being a rare entity, before accepting this diagnosis, it is essential to rule out more common causes, including vascular etiologies (stroke, hemorrhage, cerebral venous thrombosis), metabolic causes (hypoglycemia, hyperthyroidism, Wilson's disease), infectious causes (HIV, syphilis, tuberculosis, viral/bacterial encephalitis), autoimmune conditions (systemic lupus erythematosus, antiphospholipid syndrome, multiple sclerosis, paraneoplastic syndromes e.g. anti-Hu/anti-CRMP-5), neoplastic causes (brain tumor), and drug-induced causes (neuroleptic-induced dyskinesia, levodopa, cocaine, amphetamines).[6,7]

The pathophysiological mechanism described in the literature remains speculative and debated. Hyperglycemia induces hyperosmolarity, which reduces cerebral blood flow and directly affects astrocytes, known for their high sensitivity to ischemia (gemistocytes). Gemistocytopathy, accompanied by petechial hemorrhage and methemoglobin deposition, is commonly reported.[2,7]

Previous reports describe radiologically identifiable lesions in the basal ganglia in more than 90% of patients. These lesions are attributed to speculative hypotheses, such as acute infarction, microhemorrhage, hyperviscosity-induced damage, vasogenic edema, or calcium deposits, though definitive proof is lacking. Discrepancies between the expected and actual location of these lesions are rare.[2,7,8] While our patient's glycemic values were below 500 mg/dL, it is important to note that diabetic striatopathy and its associated MRI findings have been reported in cases with borderline values as well. Therefore, despite the relatively lower glucose levels compared to some reported cases, MRI abnormalities

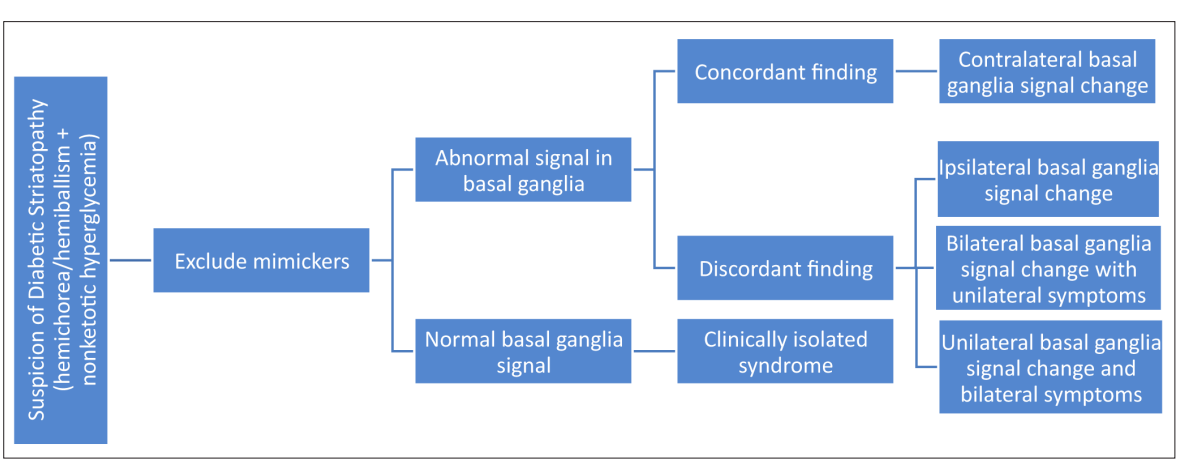

Figure 2. Conceptual descriptive clinical and radiological model in diabetic striatopathy.

would still be expected. Building on the clinical concept of diabetic striatopathy, the lack of clinical-radiological correlation can be categorized into two types: discordant findings (when imaging results are not consistent with the clinical presentation) and clinically isolated diabetic striatopathy (when no imaging abnormalities are present). We present a conceptual model to simplify these discrepancies, shown in **Fig. 2**.

We performed a standard brain CT scan and a 3T brain MRI, which represents the standard approach in most hospital centers. Additional exams, such as 7T brain MRI, MRI spectroscopy, single-photon emission computed tomography (SPECT), and dopamine transporter (DAT) scans, could help align imaging findings with the symptoms. We confidently diagnosed diabetic striatopathy based on the clinical context, the otherwise negative work-up, and the favorable outcome. We hypothesize that the normal findings in this case suggest that microbleeds or ischemia may not be the definitive cause, although they could still potentially exist, albeit below the detection threshold. ■

PATIENT'S PERSPECTIVE

I never thought that these movements would be correlated with diabetes mellitus and I was surprised when I found out that it could be a stroke. This episode in my life drew my attention to the need to better control my disease. I am grateful to the team that took care of me and I am happy to contribute to the advancement of science.

Contributorship Statement / Declaração de Contribuição

EC, MA, MC: Conception and design. Writing of the manuscript and data interpretation.

FM: Conception and design. Critical review of the manuscript. Final approval.

All authors approved the final version to be published.

EC, MA, MC: Conceção e design. Redação do manuscrito e interpretação dos dados.

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Todos os autores aprovaram a versão final a ser publicada.

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