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Diabetic Striatopathy (Hyperglycemic Hemichorea-Hemiballismus Syndrome) in a Young Patient with Type 1 Diabetes Mellitus in Dar es Salaam, Tanzania: A Case Report

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Keywords
Diabetic striatopathy · Hyperglycemic hemichorea-hemiballismus · Nonketotic hyperglycemia · Type 1 diabetes mellitus · Caudate and putamen hyperdensity

Abstract
Introduction: Diabetic striatopathy, or nonketotic hyperglycemic hemichorea-hemiballismus syndrome, is a rare movement disorder linked to poorly controlled diabetes mellitus. It predominantly affects older women with type 2 diabetes mellitus and presents with characteristic basal ganglia abnormalities on computed tomography (CT) and magnetic resonance imaging (MRI). Even rarer is the presentation in a young patient, which may pose diagnostic and management challenges. ***Case Presentation:*** We report a 17-year-old male with poorly controlled type 1 diabetes mellitus presenting with left-sided hemichorea-hemiballismus of acute onset associated with hyperglycemia without ketoacidosis. Brain imaging revealed increased attenuation in the right caudate and putamen on CT and hyperintensity on T1-weighted MRI, consistent with diabetic striatopathy. The abnormal movements abated after 1 month through dietary counseling, increased insulin dosage, and anti-chorea therapy. ***Conclusion:*** Diabetic striatopathy may occur in young patients with type 1 diabetes mellitus. In resource-limited settings, its management can be challenging. There is a need for increased awareness among physicians of this potentially reversible condition, especially when seeing atypical patient populations. Strict glycemic control is an essential part of treatment.

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Introduction

Diabetic striatopathy, also referred to as a nonketotic hyperglycemic hemichorea-hemiballismus syndrome, is a rare neurological movement disorder associated with poorly controlled diabetes mellitus (DM) [1]. It manifests with hyperglycemia and the acute onset of chorea and ballism [1]. These movements are continuous, rapid, non-rhythmic, and irregular, resembling flinging or throwing actions, typically involve the proximal and distal muscles of the upper and lower extremities, most frequently unilateral, and cease during sleep [1, 2].

This condition has distinctive, reversible basal ganglia abnormalities visible on computed tomography (CT) and magnetic resonance imaging (MRI) scans [1]. Due to similar radiological presentations, diabetic striatopathy is often mistaken for hemorrhagic stroke as both can show hyperdense and hyperintense lesions on brain CT and T1-weighted MRI, respectively [1].

Although striatopathy is predominantly documented in individuals with type 2 DM, its manifestation in type 1 DM, as observed in this instance, is exceptionally infrequent due to the advantageous metabolic impact of ketosis. We present a case of diabetic striatopathy in a young patient with type 1 DM who presented with acute onset of left-sided hemichorea-hemiballismus and nonketotic hyperglycemia. This report reviews the characteristic features of this uncommon movement disorder and its neuroimaging findings.

Case Report

A 17-year-old male with a history of poorly controlled type 1 DM since 11 years of age attended Muhimbili National Hospital in Dar es Salaam, Tanzania. The hospital is the largest referral facility in the country.

His diabetes was characterized by inconsistent medical follow-up. He had been on a regimen of short-acting insulin in the morning and before lunch, in conjunction with intermediate-acting insulin in the morning and evening. He presented with a 1-month history of acute-onset, involuntary, dance-like movements affecting the left upper and lower limbs. Initially, the movements involved the left upper limb and progressively involved the left lower limb after 3 days, sparing the face. The patient experienced increased intensity of symptoms, which hindered his capacity to engage in routine activities, sit, stand, or walk without assistance. These movements persisted during wakefulness, intensified with voluntary actions, and ceased during sleep. He had no other neurologic symptoms, nor did he report chest pain, heat or cold intolerance, skin rashes, joint pain, previous history of pharyngitis, or renal or hepatic insufficiency. There were no features of rheumatic fever. Additionally, he had no history of using medications that could trigger abnormal movements and no personal or family history of seizures, movement disorders, or mental illness.

Upon physical examination, all vital signs were within normal limits. The patient's weight was 49 kg and height was 156 cm, resulting in a body mass index (BMI) of 20.1 kg/m^2. Cranial nerve functions were unremarkable. The patient could not walk, stand, sit, or maintain an upright posture due to continuous, rapid, non-rhythmic, irregular flinging movements on the left side involving both the proximal and distal muscles of the upper and lower limbs, sparing the face. He displayed the milkmaid's sign. The power of the left and right upper and lower limb muscle groups was normal, as were tendon reflexes and coordination. Sensory examination and other systemic examinations were unremarkable.

Laboratory tests showed a random blood glucose level of 29.2 mmol/L, glycosuria without ketonuria, and a normal blood gas analysis. His glycated hemoglobin (HbA1c) was 14.1%. Thyroid, liver, and renal function tests were normal. A CT scan of the brain showed

increased attenuation in the right basal ganglia area involving the caudate and putamen with areas of calcification, sparing the internal capsule (shown in Fig. 1a). T1-weighted MRI revealed hyperintensity in the same region, with no mass effect or surrounding edema (shown in Fig. 1b) and without diffusion restriction. The electroencephalogram indicated the absence of epileptiform discharges. Furthermore, parathyroid hormone levels, serum electrolytes, and erythrocyte sedimentation rate were all within normal parameters. Antinuclear antibody and antistreptolysin O serology were also negative.

A diagnosis of nonketotic hyperglycemic hemichorea-hemiballismus syndrome was made. During the first week of hospitalization, clonazepam was administered orally at 2 mg twice daily and added to the subcutaneous insulin regimen (details of which are given in Table 1), but there was no relief. In the second week, baclofen was substituted for clonazepam, starting at a dose of 5 mg administered orally 3 times a day and increasing to 10 mg twice daily from day 4, but again without relief. In the third week, the patient was switched to amantadine 50 mg administered orally, which resulted in a significant improvement in symptoms. By the fourth week, the movements had become less frequent and less forceful, thus allowing him to regain the ability to stand and walk. However, his blood glucose levels remained relatively high (Table 1), exacerbated by poor dietary control. Dietary counseling was employed, and the total daily insulin dose was increased to 58 units, resulting in better glycemic control. The patient was discharged home with a follow-up planned through the neurology clinic. The CARE Checklist has been completed by the authors for this case report, attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000542452).

Discussion

This report describes a rare case of hemichorea-hemiballismus in a young patient with type 1 DM. Chorea and ballism are hyperkinetic movement disorders typically associated with lesions in the basal ganglia, which can be either bilateral or unilateral; the latter is referred to as hemichorea-hemiballismus [3, 4]. These disorders are most commonly caused by vascular events, with ischemic or hemorrhagic strokes in the contralateral basal ganglia being the primary culprits [3, 4]. Metabolic disorders also play a significant role, with diabetic striatopathy being the leading metabolic cause. Other metabolic conditions that can contribute to these movement disorders include thyrotoxicosis, hypoparathyroidism, uremia, and hepatic encephalopathy [1, 3–5]. Infections such as neurocysticercosis, tuberculosis, and toxoplasmosis, as well as immune-mediated conditions like systemic lupus erythematosus and acute rheumatic fever, can also present with chorea and ballism [1, 3–5]. Additionally, drug-induced causes, deposition disorders like Wilson's disease and Fahr's syndrome, and degenerative diseases such as Huntington's disease are known to contribute to these symptoms [3, 4].

Diabetic striatopathy, a neurological complication predominantly observed in patients with poorly controlled type 2 DM, accounts for more than 90% of cases in the context of nonketotic hyperglycemia [1, 5]. It occurs in approximately 1 in 100,000 diabetic patients, with a higher prevalence in older women of Asian descent [1, 5].

The precise pathophysiology of diabetic striatopathy is still not fully understood. However, it is believed that hyperosmolarity and hyperviscosity, resulting from hyperglycemia, reduce cerebral blood flow and cause damage to the striatal astrocytes, which are particularly vulnerable to ischemia, disrupting basal ganglia circuitry involved in regulating movements, resulting in a hyperkinetic movement disorder. This can manifest as microinfarcts, petechial hemorrhages, post-ischemic calcifications, myelinolysis, gliosis, and atrophy in the basal ganglia [1, 5]. Furthermore, heightened sensitivity of dopaminergic receptors and diminished availability of gamma-aminobutyric acid in the striatum due to the nonketotic state have been proposed as

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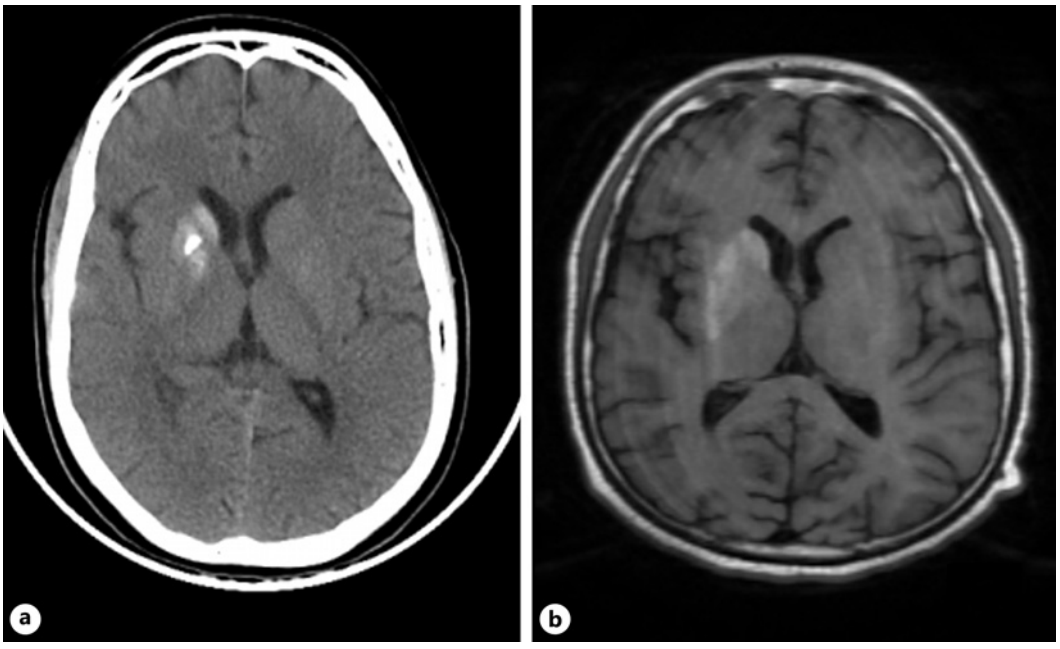

Fig. 1. a Axial CT scan of the brain showing hyperdense lesions in the right basal ganglia involving the caudate and lentiform nuclei with calcifications. **b** Axial T1-weighted MRI of the brain showing hyperintense lesions on the corresponding basal ganglia area.

Table 1. Interventions, capillary fasting blood glucose, and clinical response during admission

Intervention	Week 1	Week 2	Week 3
Insulin SC	Short-acting	Short-acting	Short-acting
	AM: 8 units	AM: 10 units	AM: 12 units
	Pre-lunch: 4 units	Pre-lunch: 6 units	Pre-lunch: 8 units
	PM: 6 units	PM: 8 units	PM: 10 units
	Intermediate-acting	Intermediate-acting	Intermediate-acting
	AM: 10 units	AM: 12 units	AM: 16 units
	PM: 8 units	PM: 10 units	PM: 12 units
Clonazepam PO	2 mg BD	Stopped	–
Baclofen PO	–	10 mg BD	Stopped
Amantadine PO	–	–	50 mg OD
Diet	Non-adhering	Non-adhering	Non-adhering
FBG, mmol/L	15–20	12–18	8.1–11
Movements	Present	Present	Subsiding

BD, twice a day; FBG, fasting blood glucose; OD, once a day; PO, orally; SC, subcutaneously.

additional contributing factors [1, 5]. The precise pathophysiology underlying the unilateral presentation of basal ganglia lesions or unilateral chorea and ballism within the context of a systemic metabolic condition remains poorly understood.

Other metabolic disorders, including thyrotoxicosis, hypoparathyroidism, uremia, and hepatic encephalopathy, may also result in similar movement disorders and basal ganglia alterations. Infections such as neurocysticercosis, tuberculosis, and toxoplasmosis, as well as immune-mediated conditions like systemic lupus erythematosus and acute rheumatic fever, have been associated with these movement disorders.

Diabetic striatopathy is even rarer in type 1 DM patients, likely because of the formation of ketone bodies, which can serve as an alternative energy source for the brain in a hyperglycemic state [1, 5–7]. In contrast, nonketotic hyperglycemia leads to gamma-aminobutyric acid utilization and eventual depletion, contributing to chorea and ballism [1, 5]. One review of 176 patients with diabetic striatopathy reported an average blood glucose level of 414 mg/dL (23 mmol/L) and an HbA1c level of 13.1% [5].

Neuroimaging findings in diabetic striatopathy are variable, ranging from the absence of radiological abnormalities to distinct, reversible basal ganglia alterations discernible on CT and MRI scans, with hyperdensity and hyperintensity on CT scan and T1-weighted MRI, respectively [1, 5, 8]. Because of similar radiological presentations, diabetic striatopathy may therefore be mistaken for a hemorrhagic stroke.

Our patient is a 17-year-old male with poorly controlled type 1 DM who presented with sudden onset of hemichorea and hemiballism, nonketotic hyperglycemia, and an HbA1c level of 14.1%. Neuroimaging revealed characteristic right-sided basal ganglia lesions on CT and MRI, consistent with previously reported cases [1, 2, 5–7]. The patient was admitted for 1 month and treated with insulin therapy, intravenous hydration, and a sequence of medications: clonazepam, baclofen, and eventually amantadine. Despite counseling, achieving euglycemia was challenging because of poor dietary adherence. The patient showed significant but partial recovery by the third week and was discharged in the fourth week.

Achieving euglycemia with insulin and hydration is considered the mainstay of treatment for diabetic striatopathy, with anti-chorea medications as adjuncts [1, 5]. In some refractory cases, invasive interventions such as pallidotomy and deep brain stimulation have been reported [1]. The prognosis is generally favorable, although the resolution of involuntary movements can vary widely, from a few days to several months, with some patients experiencing only partial improvement over months to 5 years [1, 5]. Close follow-up is crucial due to a recurrence rate of 17%–20% [1, 5].

Conclusion

This case report showcases a rare neurologic complication of poorly controlled diabetes in a young patient with type 1 DM, which we believe is the first reported case in Tanzania. It underscores the importance of a high index of suspicion for diabetic striatopathy, a potentially reversible condition, in patients presenting with chorea and ballismus. It is important to consider other metabolic conditions, stroke, or infections as part of the differential diagnosis, particularly when neuroimaging results indicate basal ganglia involvement. Furthermore, the case highlights the importance of achieving euglycemia during management, which is the mainstay of treatment.

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Statement of Ethics

This case report was reviewed and approved by the Clinical Research, Training and Consultancy Unit of the Muhimbili National Hospital, approval reference no. [MNH/HTRCU/2024/67]. Written informed consent was obtained from the patient's mother for the publication of this case report. A copy of the consent form is available for review by the Editor upon request.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

B.T., T.P., M.A., L.L., K.O., E.K., and M.B. wrote the main manuscript. B.T. formatted the table, figure, and main text. All authors have reviewed the manuscript and agreed to its publication.

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

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