



Diabetic Striatopathy with Ketoacidosis: A Case Report

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Abstract

Diabetic Striatopathy (DS) is an uncommon complication of diabetes mellitus (DM), characterized by the manifestation of dyskinesias and associated with imaging abnormalities in the basal ganglia. In this case study, we present a 39-year-old female patient who exhibited chorea due to severe hyperglycemia and diabetic ketoacidosis (DKA). Neuroimaging assessments, including computed tomography (CT) and magnetic resonance imaging (MRI) of the brain, confirmed the diagnosis of DS. Notably, the choreic symptoms resolved following the restoration of glycemic control and the administration of benzodiazepines (BZDs). This case report highlights the diagnostic approach and clinical presentation in a patient with concurrent DS and DKA, and it emphasizes current therapeutic protocols.

Keywords

Diabetic Striatopathy, Diabetes Mellitus, Benzodiazepines, Chorea

Abbreviations

DS: Diabetic Striatopathy; DM: Diabetes Mellitus; DKA: Diabetic Ketoacidosis; CT: Computed Tomography; MRI: Magnetic Resonance Imaging; BZDs: Benzodiazepines; HCHB: Hemichorea-hemiballismus; RR: Reference Range; CSF: Cerebrospinal Fluid; GABA: Gamma-aminobutyric Acid; DAT-SPECT: Dopamine Transporter Single Photon Emission Computed Tomography; VMAT2: Vesicular Monoamine Transporter Type 2

Introduction

Diabetic striatopathy, alternatively recognized as hemichorea-hemiballismus (HCHB) syndrome and non-ketotic hyperglycemic hemichorea, is distinguished by the coexistence of hyperglycemia with choreiform or ballistic movements, accompanied by reversible abnormalities within the basal ganglia detectable via neuroimaging assessments [1]. Typically, DS manifests

in patients with acute or subacute onset, presenting as the emergence of rapid, irregular, non-purposeful, and involuntary motor activities involving one or both extremities, along with facial dystonias such as ocular protrusion, pursing of the lips, and tongue protrusion [2]. In 2011, the prevalence of DS was approximated at 1 case per 100,000 individuals [3], with a predilection for elderly Asian females, exhibiting a male-to-female

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ratio of 1:1.76 [4]. Notably, Asian individuals comprised up to 91% of cases in 2002 and 71.6% in 2020, although the proportion of other racial groups has been rising in recent years [4,5]. The majority of DS patients exhibit a hyperglycemic hyperosmolar state, with diabetic ketoacidosis being a less frequent occurrence [6].

Case Presentation

A previously healthy 39-year-old female was admitted to the emergency department with a one-week history of polyuria, polydipsia, and abdominal pain, which deteriorated with dyspnea, vomiting, and unconsciousness subsequent to ingesting a significant quantity of fruit for a day. Further history revealed she had had an unproductive cough before the onset of the illness. During this period, she exhibited choreiform movements lasting 20 seconds, which responded to a 5 mg dose of Midazolam, and subsequently lost consciousness.

On physical examination, tachypnea, tachycardia, rhonchi, and unlocalized crackles were noted. There was no marked positive sign that could be found. GCS score was E3V5M6/15. Laboratory workup revealed hyperglycemia with a blood glucose level of 17.8 mmol/L (reference range, 3.9–6.1 mmol/L), hypokalemia with a potassium level of 3.0 mmol/L (RR 3.5–5.5 mmol/L), and severe metabolic

acidosis in routine blood tests. Arterial blood gas analysis demonstrated a pH of 7.0 (reference range, 7.35–7.45), a bicarbonate level of 2.3 mmol/L (RR 22.0–27.0 mmol/L), a base excess of -29.9 (RR -3.0 to +3.0 mmol/L), an anion gap of 15.6 mmol/L (RR 8–16 mmol/L), and a normal lactate level. Urinalysis indicated extremely high levels of glucose and ketone bodies, exceeding the test limits at 55 mmol/L and 7.8 mmol/L, respectively (**Table-1**).

Due to unconsciousness, a lumbar puncture was performed on the first day of admission. The intracranial pressure was recorded as 120 cmH₂O. Analysis of the cerebrospinal fluid (CSF) showed normal results in routine tests, but revealed elevated levels of protein and glucose, with values of 11.0 mmol/L (RR 2.5–4.5 mmol/L) and 0.48 g/L (RR 0.15–0.45 g/L), respectively. To date, no clinically significant pathogenic findings have been identified. Subsequently, a CT scan of the brain was conducted. The CT scan (**Fig-1**) demonstrated a high-density lesion in the right basal ganglia, which exhibited a clear boundary and was devoid of any edema.

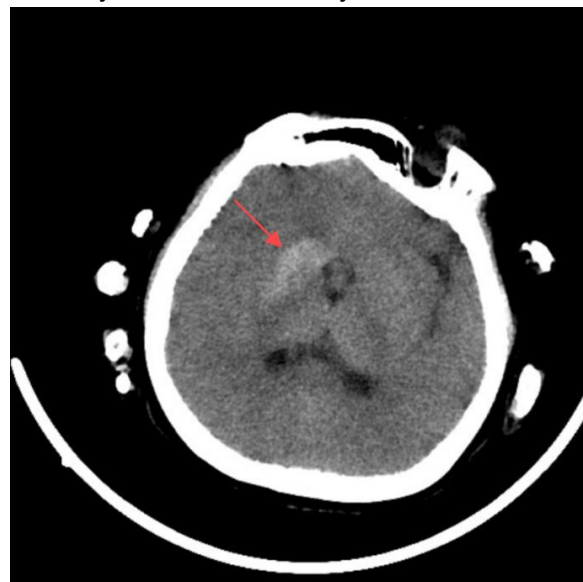


Fig-1: A High-Density Lesion in the Right Basal Ganglia in CT Scan

The presence of high intensity in the basal ganglia on the brain CT scan suggested the possibility of cerebral hemorrhage, which could not be definitively excluded in this case. Consequently, mannitol was prescribed to reduce intracranial pressure. Additionally, insulin therapy was initiated to lower blood glucose levels, and antibiotics were administered to treat

Table-1:

Parameter	Value	Normal Range
Hemoglobin	149g/L	110-150 g/L
Platelet count	187×10 ⁹ /L	100-350×10 ⁹ /L
Leukocyte count	14.32×10 ⁹ /L	3.50-9.50×10 ⁹ /L
BUN	4.03mmol/L	2.78-7.14 mmol/L
Serum creatinine	49μmol/L	45-84μmol/L
ALT	13U/L	7-40 U/L
Sodium	137mmol/L	135-145 mmol/L
Potassium	3.0mmol/L	3.5-5.5 mmol/L
Chloride	106mmol/L	96-111 mmol/L
Bicarbonate	2.3mmol/L	22.0-27.0 mmol/L
CRP	56.0mg/L	0-8 mg/L
ESR	9mm/h	0-20 mm/h
BS	17.8	3.9-6.1mmol/L
GA%	46.20%	10.8-17.1%

BUN: Blood Urea Nitrogen, ALT: Alanine Aminotransferase, CRP: C-Reactive Protein, ESR: Erythrocyte Sedimentation Rate, BS: Blood Sugar

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pneumonia. However, despite these interventions, the patient experienced recurrent continuous, regular, and involuntary chorea/ballismus after two days of treatment.

A review of the cranial MRI (**Fig-2**) showed abnormal signal in the right basal ganglia and the head of the caudate nucleus on T1-weighted images, while no significant changes were observed on T2 images (**Fig-3**). These imaging characteristics were indicative

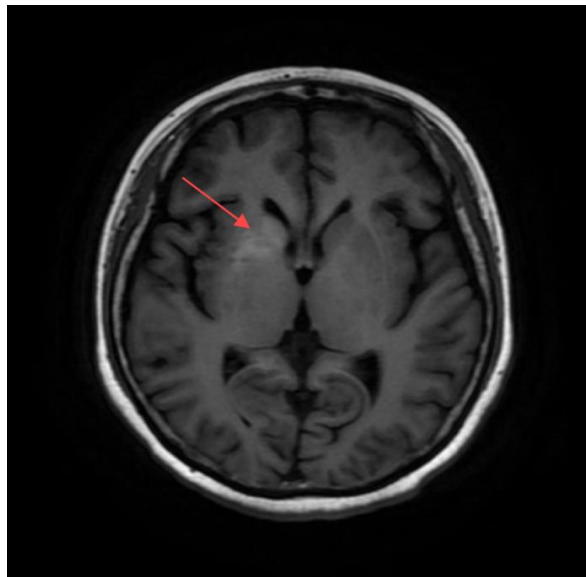


Fig-2: High Signal in the Right Basal Ganglia on T1-Weighted MRI

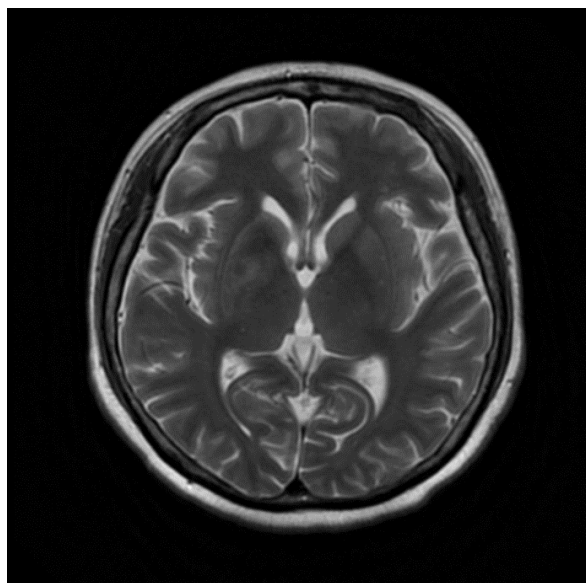


Fig-3: Normal Signal on T2-Weighted MRI

of or compatible with DS. Based on these findings, a diagnosis of DKA and DS was made. The chorea was attributed to hyperglycemia-induced impairment of the basal ganglia, manifesting as rapid, involuntary, and

irregular movements in the contra-lateral limb. Tiapride 150 mg three times daily and clonazepam 0.5 mg per night were administered, resulting in the alleviation of neurological symptoms.

Discussion

DS is defined as a hyperglycemic condition that is associated with at least one of the following manifestations: (1) the presence of chorea or ballismus; (2) striatal hyperdensity on CT or hyperintensity on T1-weighted magnetic resonance imaging [1].

The precise pathogenesis of DS remains incompletely understood. At the microscopic level, the depletion of gamma-aminobutyric acid (GABA) is implicated in the disease process. R. Guisado and A.I. Arieff [7] postulated in 1975 that hyperglycemia might shift brain metabolism towards anaerobic pathways, leading to the inactivation of the Krebs cycle. This, in turn, results in the shunting of GABA into succinic acid to generate additional energy. The rapid depletion of GABA and acetate leads to an imbalance in neurotransmitter distribution within the central nervous system, causing dysfunction in the subthalamus and basal ganglia, and subsequently presenting as chorea. They also revealed that GABA can be resynthesized in ketotic hyperglycemia [7], which explains why a large number of DS cases occur with non-ketotic hyperglycemia, whereas few cases are associated with DKA [6]. However, the aforementioned theory does not fully elucidate our case, and the involvement of other mechanisms needs to be considered.

Apart from GABA, an uneven distribution of dopamine may also contribute to DS. A case report discusses that choreiform movements in postmenopausal women may be attributed to hormonal alterations that affect dopamine functionality [8]. A recently published case in Japan provides further insight [9]. The patient with DS developed symptoms eight days after discharge, accompanied by exacerbated striatal hyperintensity on T1-weighted MRI. Haloperidol was restarted, and the dose was gradually increased, but it failed to improve the ballismus. Dopamine transporter single photon emission computed tomography (DAT-SPECT) revealed diminished bilateral striatal uptake and a positive response to a

L-dopa challenge test. With incremental increases in L-dopa, the ballismus significantly improved. This suggests that presynaptic dopaminergic dysfunction may be one of the causes of DS.

In macroscopic terms, structural alterations may also result in lesions. The basal ganglia, also known as the basal nuclei, exhibit a close association with DS. These subcortical structures are located deep within the white matter of the brain and consist of five pairs of nuclei: the caudate nucleus, putamen, globus pallidus, subthalamic nucleus, and substantia nigra, collectively forming part of the extrapyramidal motor system. Damage to the basal ganglia may manifest as behavioral changes and movement disorders, presenting as hypokinesia, hyperkinesia, or a combination of both, contingent upon the location and extent of the affected structure. Seung-Hun Oh et al. [4] examined a total of 53 cases of DS, all of which demonstrated high signal intensity lesions in the basal ganglia, particularly involving the putamen, on T1-weighted brain MRI. Among these patients, 24 cases exhibited low signal intensity, and 17 cases showed iso-signal intensity lesions in the basal ganglia on T2-weighted imaging, respectively [4]. These findings suggest that damage to or imaging changes within the basal ganglia may contribute to the development of DS.

In the clinical setting, a variety of medications are utilized for the treatment of DS. To address the underlying issue, management of hyperglycemia is essential. For the alleviation of choreic movements, dopamine receptor blockers are generally regarded as the most efficacious agents. Second-generation antipsychotic medications, such as risperidone and olanzapine, are commonly prescribed to assist in reducing chorea. Additionally, dopamine-depleting agents, particularly inhibitors of the presynaptic vesicular monoamine transporter type 2 (VMAT2), including valbenazine and deutetrabenazine, represent viable therapeutic options for the treatment of chorea [10]. In this particular case, beyond the aforementioned therapies, tiapride, clonazepam, and midazolam were prescribed, which were also utilized in this case, with a resultant alleviation of involuntary movements. Benzodiazepines (BZDs) are ubiquitously employed for their anxiolytic, hypnotic, anticonvulsant, and even

antiseizure properties through their binding to GABA(A) receptors, which are ligand-gated chloride ion channels. When GABA binds to these receptors, it enhances the chloride current generated by the receptor. Theoretically, BZDs potentiate GABA's inhibitory effect by increasing the frequency of chloride channel openings, thereby exerting a mild antichorea effect. However, the documentation concerning the use of such agents is limited [11].

Conclusion

In the presence of an elevated blood glucose level, choreiform movements, and T1-weighted brain MRI demonstrating high signal changes in the basal ganglia, with or without evidence of ketoacidosis, diabetic striatopathy should be considered as a highly probable diagnosis. Despite the current limitation of supportive evidence, BZDs may offer therapeutic benefits in alleviating chorea and potentially treating DS, particularly when used in conjunction with glycemic control measures.

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Authors' Contributions

Y. N. drafted the manuscript, collected the patient's information, researched literature and conceived the study. Y.L. involved in protocol development, gaining ethical approval, patient recruitment and data analysis. Y.N. wrote the first draft of the manuscript. All authors reviewed and edited the manuscript and approved the final version of the manuscript.

Data Availability

All data generated or analyzed during this study are included in this published article.

Ethical Approval and Consent to Participate

Not applicable.

Consent for Publication

Written Informed consent was obtained from the patient for publication.

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Potential Conflicts of Interest

The authors declare that they have no competing interests.

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