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Case Report

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Associated with hyperglycemia: a rare cause of hyperkinetic movement disorders

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ABSTRACT

Diabetic striopathy, a neurological disorder associated with diabetes mellitus, is characterized by specific changes in basal ganglia structures, particularly the striatum. Although its precise etiology remains elusive, chronic hyperglycemia, microvascular dysfunction, oxidative stress, and inflammation are implicated. Diagnosis relies on clinical assessment and neuroimaging, revealing characteristic basal ganglia abnormalities. Management focuses on optimizing glycemic control and alleviating symptoms. We present an 83-year-old woman with abrupt-onset unilateral chorea-ballismus as a case illustration, showcasing diagnostic and therapeutic approaches. Understanding the underlying mechanisms and refining management strategies are crucial for effectively addressing this complex neurological complication of diabetes mellitus.

Keywords: Hyperglycemia, striatopathy, diabetic ketoacidosis

INTRODUCTION

Diabetic striopathy, also known as diabetic striatal syndrome or diabetic striatal degeneration, is a neurological disorder characterised by specific changes in the basal ganglia structures, particularly the striatum, in individuals with diabetes mellitus.[1,2] It is considered a relatively rare complication of diabetes, predominantly affecting individuals with long-standing and poorly controlled diabetes.[3]

The pathogenesis of diabetic striopathy is not yet fully understood, but it is believed to be multifactorial. Chronic hyperglycemia, microvascular dysfunction, impaired blood-brain barrier integrity, oxidative stress, and inflammation have been implicated as potential contributors to the development of this condition.[4,5]

It has usually been described in elderly females with hyperglycemic hyperosmolar states, but instances in patients with diabetic ketoacidosis are few. Clinically, diabetic striopathy presents with various neurological symptoms, including movement disorders such as chorea, dystonia, or parkinsonism. These movement abnormalities may be accompanied by cognitive impairment, psychiatric symptoms, and autonomic dysfunction. The severity and progression of symptoms can vary among individuals.[6]

The diagnosis of diabetic striopathy is typically based on clinical evaluation, neuroimaging studies. It can be shown to have reversible hipodensity in the corpus striatum on computer tomography (CT) and hyperintensity on brain magnetic resonance imaging (MRI), with the exclusion of other possible causes of basal ganglia dysfunction.[7,8] Neuropathological studies have revealed characteristic findings in the striatum, such as gliosis, neuronal loss, and vascular changes.[9]

Management of diabetic striopathy primarily focuses on optimising glycemic control and addressing associated comorbidities. Symptomatic treatment may include the use of medications to manage movement disorders or psychiatric symptoms. However, the effectiveness of specific therapies remains limited, and further research is needed to establish optimal treatment strategies.[6]

CASE

A 83-year-old woman with a history of type 2 DM presented with abrupt-onset unilateral chorea-ballismus. Symptoms started with abrupt-onset choreiform movements on the left-sided extremities lasting three weeks.

Her medical history included hypertension, atrial fibrillation, and a previous stroke with no residual disability. She doesn't have a history of movement disorders or epilepsy. Records revealed capillary blood glucose (CBG) of 565 mg/dl, urine glucose +++, urine ketone was negative, and a venous blood pH value of 7.4 with a high serum osmolarity of 303 mOsm/kg and normal renal and liver function tests. The patient was diagnosed with non-ketotic hyperglycemia. A brain CT showed slightly high attenuation of the right putamen and globus pallidus Figure 1.A. Brain MRI revealed an area of high signal intensity in T1-weighted images Figure 1.C and low signal intensity in FLAIR images Figure 1.B, involving the right putamen and globus pallidus. Diffusion-weighted MRI brain images did not reveal any abnormal restricted diffusion in the right basal ganglia Figure 1.D.

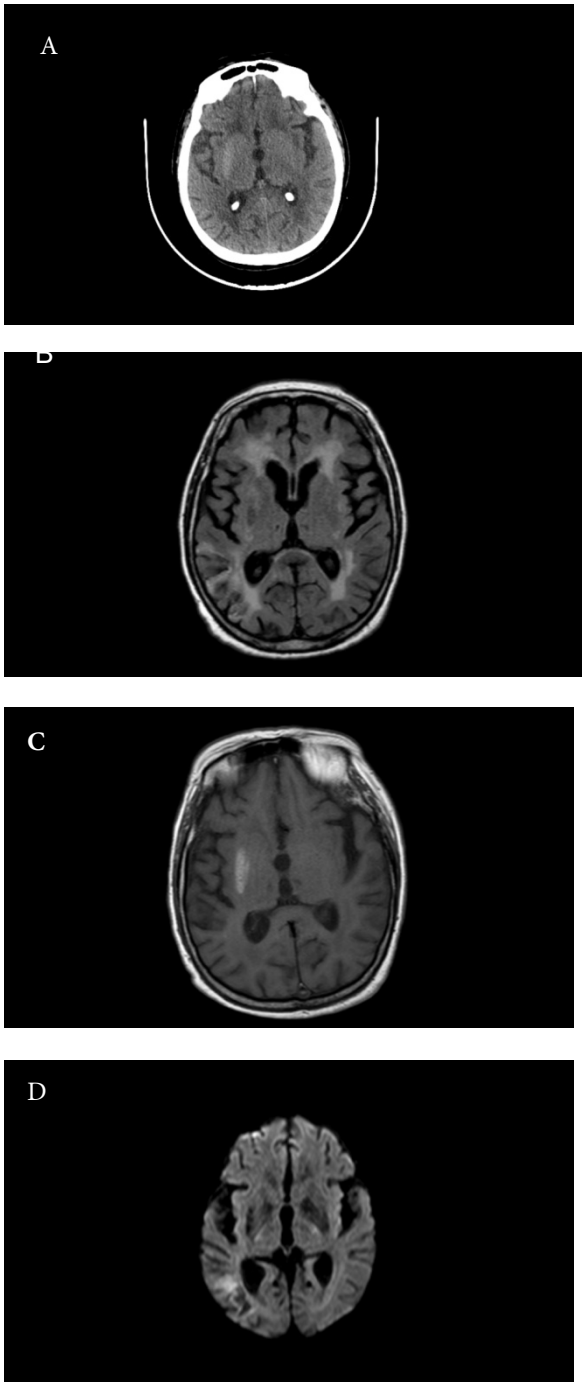

Figure 1. (A-D) (A) A brain CT scan revealed slight attenuation of the right putamen and globus pallidus shows with white arrow (B) FLAIR images showing hypointensities and (C) T1 weighted images with white arrow shows hyperintensities in the right lenticular nucleus (D) Diffusion Weighted MR image demonstrating no diffusion restriction.

According to the blood sugar measurements, insulin treatment has been planned for the patient. Moreover, the initial treatment for the involuntary movements is 5 mg of haloperidol twice a day. But it wasn't efficient, so we added ketiapin 25 mg twice a day. After this change, involuntary movements decreased, but they were still continuing. Two weeks after the admission, involuntary movements totally disappeared, and we stopped haloperidol and planned to reduce the ketiapin dosage in a 1-3 month follow-up. We are scheduled for a revaluation and repeat imaging after 3 months. A follow-up brain MRI after 3 months of treatment showed improved signal abnormalities at the right globus pallidus and putamen Figure 2, B-C. Also, a control brain CT showed a decrease in the size of the hyperdensity Figure 2.A.

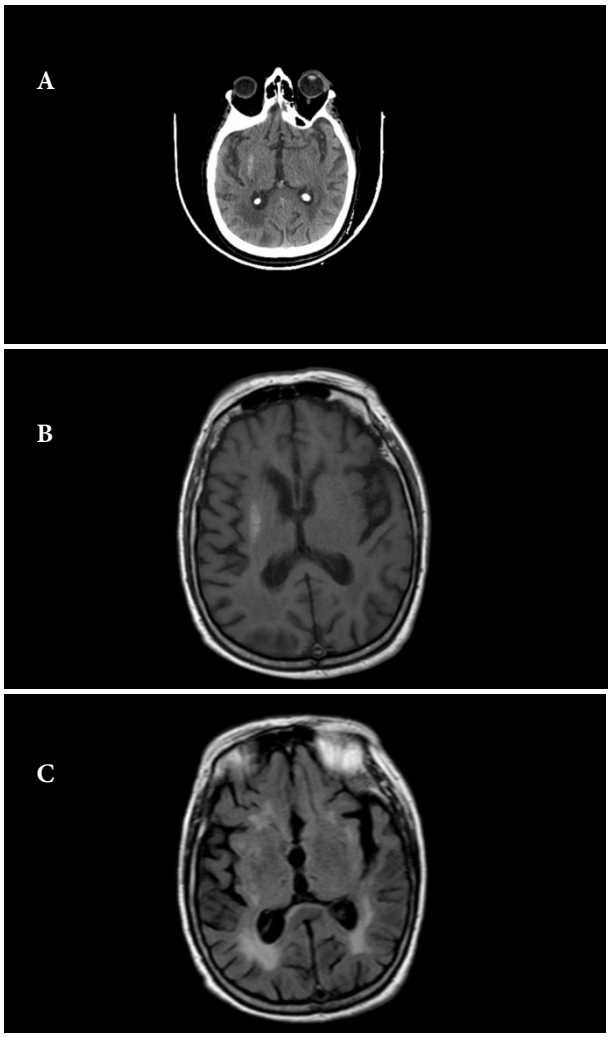

Figure 2. (A-C) (A) Control CT image showing the decrease in the size of hyperdense lesion in the right basal ganlia. (B-C) Control brain MRI (T1 wegihted image: B, FLAIR image: C) showing slight decrease in abnormal signal intensities.

DISCUSSION

Diabetic striopathy is a complex neurological disorder associated with diabetes mellitus, characterised by specific changes in the basal ganglia structures, particularly the striatum. Although the exact pathogenesis of diabetic striopathy is not fully understood, several mechanisms have been proposed based on existing literature.

Chronic hyperglycemia has been identified as a key factor in the development of diabetic striopathy. Prolonged exposure to high glucose levels can lead to oxidative stress and the

formation of advanced glycation end products (AGEs), which contribute to neurovascular dysfunction and neuronal damage within the basal ganglia structures.[10] Microvascular dysfunction, including endothelial dysfunction and blood-brain barrier impairment, has also been implicated in the pathogenesis of diabetic striopathy.[11] These vascular abnormalities can lead to compromised perfusion and nutrient supply to the basal ganglia, resulting in neuronal loss and gliosis.

Inflammation is another potential contributor to the development and progression of diabetic striopathy. Chronic low-grade inflammation is commonly observed in diabetes mellitus and has been linked to neuroinflammation and neurodegeneration.[12] Inflammatory mediators, such as cytokines and chemokines, may play a role in the disruption of the basal ganglia structures and the development of movement disorders seen in diabetic striopathy.

Genetic factors may also influence the susceptibility to diabetic striopathy. Studies have identified specific gene variants associated with an increased risk of developing movement disorders in individuals with diabetes.[13] However, further research is needed to elucidate the genetic underpinnings of this condition and their interaction with environmental factors.

The diagnosis of diabetic striopathy relies on clinical evaluation, neuroimaging findings, and the exclusion of other potential causes of basal ganglia dysfunction. Neuroimaging techniques, such as MRI or CT, can reveal characteristic changes in the striatum, including atrophy, signal abnormalities, and vascular lesions.[14]

Management of diabetic striopathy focuses on optimising glycemic control and addressing associated comorbidities. Intensive glucose management and control of other cardiovascular risk factors may help slow the progression of neurological symptoms.[15] Symptomatic treatment with medications targeting movement disorders, such as dopaminergic agents or antipsychotics, may be considered on a case-by-case basis, taking into account the individual patient's needs and risks.

CONCLUSION

Diabetic striopathy is an intriguing neurological complication of diabetes mellitus, characterised by distinct changes in the basal ganglia structures. Management involves optimising glycemic control and addressing associated comorbidities. Understanding the underlying mechanisms and improving diagnostic and therapeutic approaches are crucial for better management of this condition.

ETHICAL DECLARATIONS

Informed Consent:

The patient signed and free and informed consent form.

Referee Evaluation Process:

Externally peer-reviewed.

Conflict of Interest Statement:

The authors have no conflicts of interest to declare.

Financial Disclosure:

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Author Contributions:

All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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