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Case Report

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Diabetic Chorea with Basal Ganglia Lesion on MRI: A Case Series

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Abstract

Non-ketotic hypoglycaemia or hyperglycaemia causing abnormal hyperkinetic movement disorders is a rare condition. This case series illustrates four Asian female patients with poorly controlled type 2 diabetes mellitus who presented with acute onset hemichorea-hemiballismus. Magnetic resonance imaging of the brain was useful in establishing the diagnosis, and all patients recovered following improvement in glycaemic control. Recognising the imaging features of diabetic chorea, as well as the changes in the brain associated with hypo- and hyperglycaemia, is essential for the differential diagnosis of movement disorders and for initiating prompt, appropriate treatment.

Keywords: Hemichorea, Magnetic Resonance Imaging, Basal Ganglia, Non-ketotic hypoglycaemia, Non-ketotic hyperglycaemia

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INTRODUCTION

Hemichorea-hemiballismus is a rare manifestation of nonketotic hyperglycaemic and hypoglycaemic states but is more commonly observed in Asian females with uncontrolled diabetes mellitus.[1-3] The acute onset of hemichorea-hemiballismus is often associated with lesions in the basal ganglia (BG), particularly the subthalamic nucleus (STN). The basal ganglia are a group of subcortical nuclei involved in motor control, and

dysfunction in this region is linked to extrapyramidal syndromes.[4]

Brain magnetic resonance imaging (MRI) is a valuable tool for evaluating the pathogenesis of diabetic chorea. A hyperintense signal lesion in the basal ganglia on T1-weighted MRI is a common finding in both hypoglycaemic and hyperglycaemic states. However, this imaging feature is not specific to diabetic chorea and may be seen in other pathologies. This case series further explores the spectrum of cerebral changes that may occur as a result of glycaemic dysregulation.

CASE SERIES

Four Asian female patients, aged between 49 and 59 years, were admitted to the hospital with complaints of acute-onset, unilateral, involuntary limb movements. All four had a history of long-standing type II diabetes mellitus and hypertension, both of which were being managed with medication. None had a prior history of chorea or kernicterus, and there was no family history of benign hereditary chorea or other neurodegenerative diseases.

Case 1 — A 49-year-old woman presented with sudden onset choreiform movements involving the left upper and lower limbs. On admission, her fasting blood glucose was 15 mmol/L (Normal range 4.5-6.5), HbA1c was 9% (Normal <6.3%), and blood pressure was 140/80mmHg. Urine ketones and arterial blood gases were within normal limits. Neurological examination revealed no other abnormalities, and routine biochemical investigations were unremarkable. Brain MRI showed abnormal hyperintense signals on T1-weighted (T1W), T2-weighted (T2W), and Fluid-Attenuated Inversion Recovery (FLAIR) sequences involving the putamen, globus pallidus, and head of the caudate nucleus in the right internal capsule. These findings were not evident on Gradient Echo (GRE) imaging and showed no restricted diffusion on Diffusion-Weighted Imaging (DWI) or Apparent Diffusion Coefficient (ADC) mapping (**Figure 1**).

The patient was treated with haloperidol 1.25 mg once daily for a few days. The medication was discontinued following the resolution of abnormal movements, which coincided with normalisation of her blood glucose levels.

Case 2 — A 53-year-old woman was admitted with acute-onset choreiform movements affecting the right upper and lower limbs. On admission, her random blood glucose was markedly elevated at 31 mmol/L, with HbA1c of 15.2%. Her blood pressure was 137/82 mmHg. Urinalysis revealed no ketones, and arterial blood gases were within normal limits, ruling out diabetic ketoacidosis.

In addition to limb chorea, she exhibited facial chorea and intermittent ballistic movements on the same (right) side. These involuntary movements subsided during sleep. Neurological examination revealed no additional deficits, and other biochemical parameters

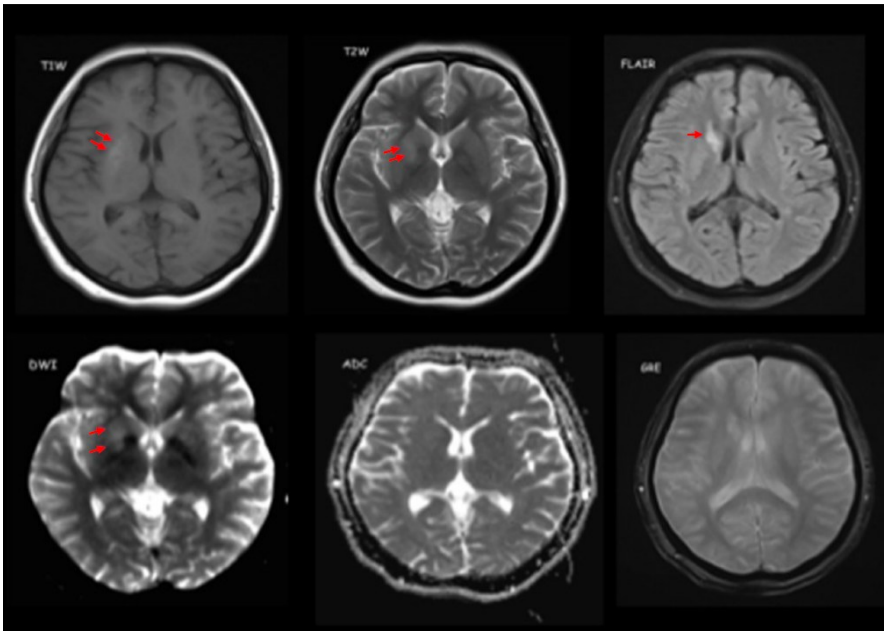

Figure 1: MRI: T1W, T2W and Fluid attenuated inversion recovery (FLAIR) images showed patchy hyperintense signals (red arrows) in the head of caudate nucleus, putamen and globus pallidus of the right internal capsule. Diffusion-weighted Imaging/Apparent Diffusion Coefficient shows no restricted diffusion, thus excludes acute ischaemia. Gradient Resonance Echo reveals no susceptibility artefact which is against acute haemorrhage.

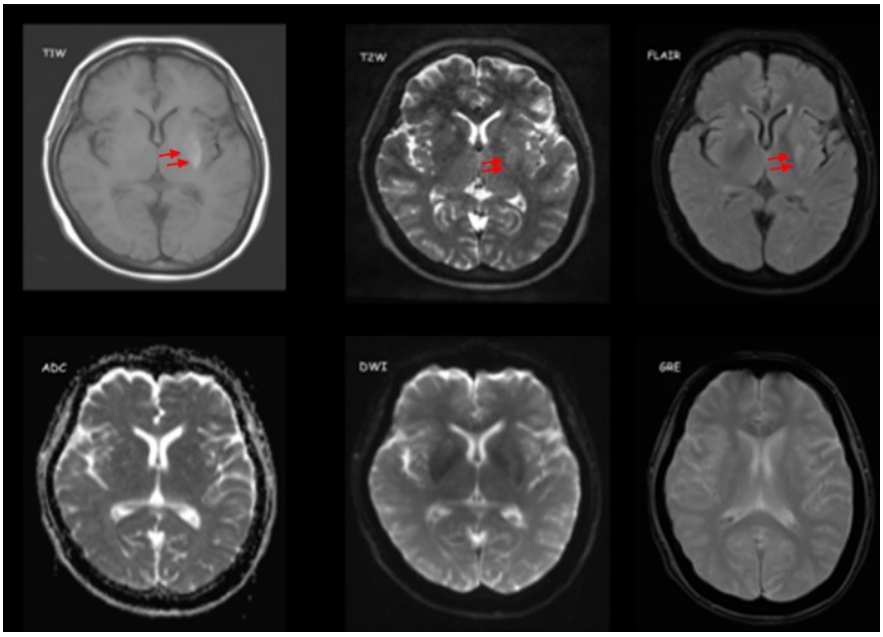

Figure 2: MRI: T1W, T2W and FLAIR showed heterogeneous hyperintense signal (red arrows) in the putamen and globus pallidus of the left lentiform nucleus. No area of restricted diffusion in the diffusion imaging study nor susceptible artefact on GRE were

were within normal ranges.

Brain MRI demonstrated hyperintense signals on T1-weighted (T1W), T2-weighted (T2W), and Fluid-Attenuated Inversion Recovery (FLAIR) sequences involving the putamen and globus pallidus of the left lentiform nucleus. There was no susceptibility artifact on Gradient Echo (GRE) imaging to suggest hemorrhage, and no restricted diffusion was observed on Diffusion-Weighted Imaging (DWI) or Apparent Diffusion Coefficient (ADC) mapping to indicate acute infarction (**Figure 2**).

She was managed with insulin and oral hypoglycaemic agents. No specific medication was administered for the abnormal movements, which gradually resolved with improved glycaemic control.

CASE 3 — An ambulance was dispatched for a 50-year-old woman found in a state of reduced consciousness. On initial assessment, she had a Glasgow Coma Score (GCS) of 12 (E4V4M4), a random blood glucose level of 3.2 mmol/L (hypoglycaemic), and a blood pressure of 140/70 mmHg. She was administered with a stat dose of 20 mL intravenous 50% dextrose, followed by a maintenance infusion of 5% dextrose by paramedics. Upon arrival at the emergency department, she regained full consciousness.

Clinical examination revealed continuous, large amplitude choreiform movements involving the left upper and lower limbs, affecting both proximal and distal muscles. The rest of the neurological examination was unremarkable. Urine analysis and arterial blood gas tests were normal, with no evidence of ketonuria or acidosis.

Brain MRI demonstrated hyperintense signals on T1-weighted (T1W), T2-weighted (T2W), and Fluid-Attenuated Inversion Recovery (FLAIR) sequences in the head of the caudate nucleus, putamen, and globus pallidus of the right internal capsule. No susceptibility artifact was observed on Gradient Echo (GRE) imaging, and there was no restricted diffusion on Diffusion-Weighted Imaging (DWI) or Apparent Diffusion Coefficient (ADC) mapping (**Figure 3**).

Her choreiform movements resolved spontaneously with the normalisation of blood glucose levels prior to discharge.

CASE 4 — A 52-year-old woman presented with a 4-day history of choreiform movements in the right upper limb. She reported no associated weakness or gait disturbances. Notably, she had not taken her usual oral hypoglycaemic medication during that period due to a lapse in prescription refills. Her random blood glucose on admission was 29.0 mmol/L. Urine analysis revealed no ketonuria, and the rest of her neurological examination was unremarkable.

She was treated with insulin in addition to the

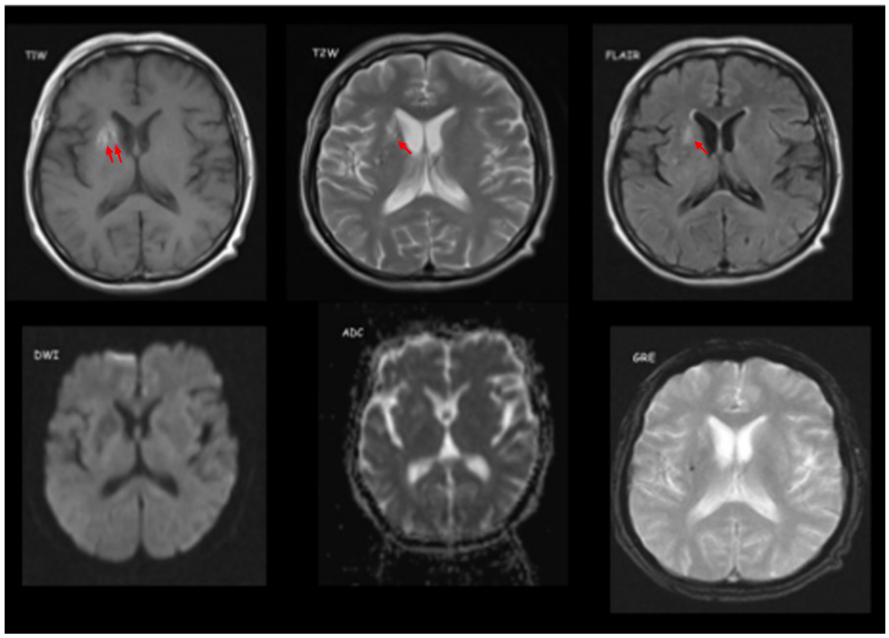

Figure 3: MRI: T1W, T2W/FLAIR images revealed hyperintense signals (red arrows) in the right internal capsule. No susceptibility artefact on GRE or area of restricted diffusion on diffusion imaging.

reinitiation of her regular oral hypoglycaemic agents.

Brain MRI revealed patchy hyperintense signals on T1-weighted (T1W), T2-weighted (T2W), and Fluid-Attenuated Inversion Recovery (FLAIR) sequences in the head of the caudate nucleus, putamen, and globus pallidus of the left internal capsule. There was no susceptibility artifact on Gradient Echo (GRE) imaging, and no restricted diffusion on Diffusion-Weighted Imaging (DWI) or Apparent Diffusion Coefficient (ADC) mapping (Figure 4).

At discharge, her blood glucose had improved from 29.0 mmol/L to 9.8 mmol/L, and her abnormal movements had resolved.

DISCUSSION

Hemichorea-hemiballismus represents a spectrum of continuous, irregular, non-patterned involuntary movements affecting one side of the body. Acute onset hemiballismus is typically associated with lesions in the STN of the basal ganglia and is considered pathogno-

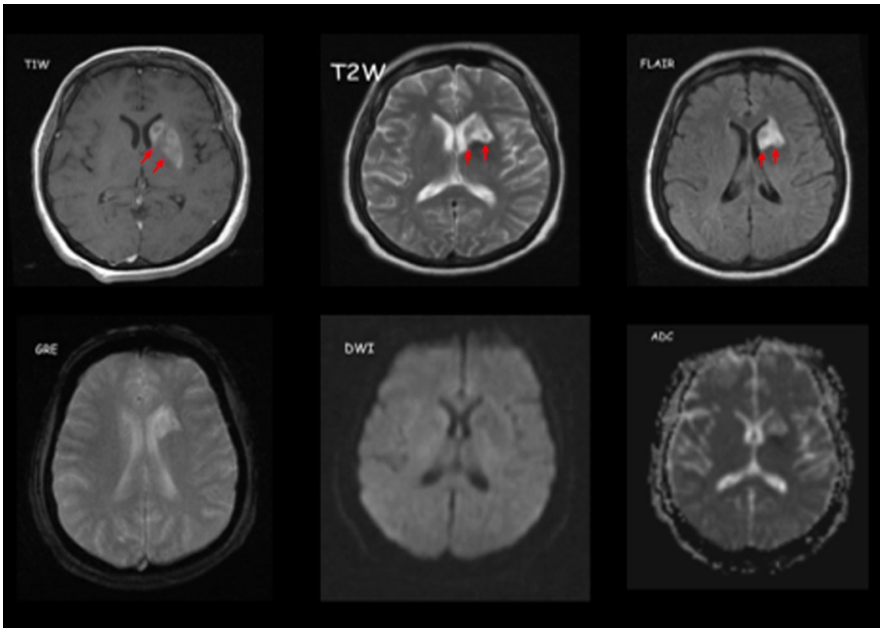

Figure 4: MRI: T1W, T2W and FLAIR images revealed hyperintense signals (red arrows) in the left internal capsule. Diffusion-weighted imaging and ADC imaging showed no evidence of acute ischaemia. Gradient echo imaging revealed no susceptibility artefact.

monic.[5] Established metabolic causes of hemichorea-hemiballismus syndrome include hyperglycaemic hyperosmolar nonketotic states and hypoglycaemia.[6–7] Other etiologies include stroke, neoplasms, vascular malformations, and infections.[7]

Historically, the prognosis for hemichorea-hemiballismus was considered poor, often associated with severe disability or even death. However, more recent studies have shown a more favorable outcome, particularly in cases related to hyperglycaemia or hypoglycaemia, where symptoms often resolve with normalisation of blood glucose levels.[5–8]

Diabetic hemichorea, usually observed in the context of uncontrolled nonketotic diabetes mellitus, most commonly occur during hyperglycaemic episodes. Nonetheless, hypoglycaemia has also been implicated in the development of diabetic chorea.[8] One proposed pathophysiological mechanism involves impaired cerebral autoregulation, leading to hypoperfusion and the activation of anaerobic metabolism due to the inactivation of the tricarboxylic acid (TCA) cycle. As a result, the brain compensates by converting gamma-aminobutyric acid (GABA) into succinic acid as an alternative energy source. Rapid depletion of GABA and acetate subsequently leads to reduced acetylcholine synthesis. This GABA shunting causes metabolic acidosis within basal ganglia neurons, resulting in striatal dysfunction and the emergence of choreiform movements.[9–11]

An alternative hypothesis suggests that hyperglycaemia-induced hyperviscosity disrupts the blood-brain barrier, leading to transient ischemia or hypoxia. This disruption reduces inhibitory input to the thalamus, ultimately contributing to the development of hyperkinetic movement disorders.[11–12]

In hypoglycaemic states, the proposed mechanism involves asymmetrical cerebral perfusion, characterised by decreased blood flow to the basal ganglia on one side and increased perfusion of the contralateral thalamus. The reduced inhibitory signaling to the thalamus on the affected side leads to ipsilateral chorea. This is supported by findings from Single-Photon Emission Computed Tomography (SPECT) studies, which demonstrate decreased perfusion in the basal ganglia of affected patients.[8,13]

All four of our patients developed unilateral hyperkinetic movement disorders, with brain abnormalities located contralaterally to the side of clinical presentation. Three patients presented during hyperglycaemic states, while one developed symptom in the context of hypoglycaemia. In all cases, the abnormal movements resolved completely within one month, coinciding with improved glycaemic control.

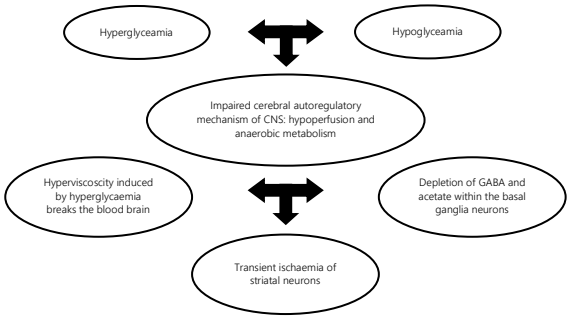

Figure 5. Direct and indirect pathways in cases of hypoglycaemic and hyperglycaemic state induced chorea.

MRI findings were consistent across the cohort. All four patients demonstrated hyperintense signals on T1-weighted (T1W), T2-weighted (T2W), and Fluid-Attenuated Inversion Recovery (FLAIR) sequences involving the putamen and globus pallidus of the lentiform nucleus. Additionally, three of the four patients also showed involvement of the head of the caudate nucleus.

Importantly, none of the lesions demonstrated restricted diffusion on Diffusion-Weighted Imaging (DWI) or Apparent Diffusion Coefficient (ADC) maps, ruling out cytotoxic edema. Similarly, no susceptibility artifacts were observed on Gradient Echo (GRE) sequences, excluding acute haemorrhage (**Figures 1-4**). These imaging findings align with previously reported MRI patterns in patients with uncontrolled diabetes presenting with hemichorea-hemiballismus.[9]

CONCLUSION

This case series highlights hemichorea-hemiballismus as a rare but important complication of poorly controlled diabetes, occurring in both hyperglycaemic and hypoglycaemic states. Recognition of this condition, along with its characteristic radiographic manifestations on MRI, is essential for accurate diagnosis and the selection of appropriate therapy. Prompt identification helps avoid unnecessary investigations and treatments, as well as prevent prolonged morbidity.

Our cases demonstrate that despite the dramatic onset of unilateral hyperkinetic movements, symptoms resolved completely with effective glycaemic control, underscoring the favourable prognosis of this disorder when managed timely. Clinicians should maintain a high index of suspicion for glycaemic-related movement disorders in diabetic patients presenting with acute involuntary movements to ensure prompt and precise intervention.

Take Home Messages

- Hemichorea-hemiballismus is a rare complication of uncontrolled diabetes, seen in both hyper- and hypoglycaemia.
- It presents with sudden unilateral involuntary movements and contralateral basal ganglia MRI abnormalities.
- MRI typically shows no signs of acute stroke or haemorrhage.
- Symptoms resolve completely with proper glycaemic control.
- Early diagnosis prevents unnecessary treatment and guides appropriate management.

Abbreviations

BG	Basal ganglia
STN	Subthalamic nucleus
MRI	Magnetic resonance imaging
GCS	Glascow coma score
TCA	Tricarboxylic acid
GABA	Gamma-aminobutyric acid
SPECT	Single Photon Emission Computed Tomography

Declarations

Conflict of interests

The authors declare no conflict of interests.

Consent

Consents have been obtained from patients for publication.

Acknowledgement

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