Case Report

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A Case Report of Diabetic Striatopathy: Rare and Unusual Complication of Hyperglycaemia

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ABSTRACT

Non-Ketotic Hyperglycaemic (NKH) Hemichorea-Hemiballismus Syndrome/Diabetic Striatopathy (DS) is a rare neurological complication of diabetes mellitus. Hemichorea can present as a manifestation of diabetes mellitus or occur in patients with poorly controlled diabetes. This case report highlights the importance of differentiating this syndrome from other intracranial pathologies, as adequate glycaemic control can lead to complete resolution of the symptoms. The authors present a case of a 65-year-old female who presented to the Emergency Department (ED) with a sudden onset of involuntary movements in the left upper and lower limbs for 30 minutes. The patient was conscious and oriented, with a General Random Blood Sugar (GRBS) of 502 mg/dL, and ketone levels of 0.1 mmol/L (normal value: less than 0.6 mmol/L). The Computed Tomography (CT) scan revealed high attenuation in the right basal ganglia, and the Magnetic Resonance Imaging (MRI) of the brain indicated T1 hyperintensity in the right basal ganglia. The patient was managed with insulin infusion and hydration. Involuntary movements completely resolved after blood sugar levels were controlled with insulin. Hyperglycaemia should be considered as a differential diagnosis in patients presenting to the ED with hemichorea.

Keywords: Hemichorea, Hyperintense basal ganglia, Non ketotic hyperglycaemia

CASE REPORT

A 65-year-old female presented to the ED with a sudden onset of involuntary movements in her left upper limb and left lower limb for 30 minutes. She also experienced increased frequency of urination for one day. The involuntary movements were continuous and non-patterned. There was no history of fever, drug intake, head injury, frothing from the mouth, visual disturbances, headache, giddiness, limb weakness, altered sensorium, increased appetite, weight loss, fatigue, or increased thirst. The patient did not have a known case of diabetes mellitus or hypertension, and there was no history of similar illness in the past. Upon arrival at the ED, the patient was conscious and oriented. Her vital signs were as follows: Pulse Rate (PR)- 80 beats per minute, Respiratory Rate (RR)- 20 breaths per minute, Blood Pressure (BP)- 130/90 mmHg. The Glasgow Coma Scale (GCS) score was 15/15. The General Random Blood Sugar (GRBS) level was 502 mg/dL, and Serum Ketones were 0.1 mmol/L (normal <0.6 mmol/L). Arterial blood gas analysis showed normal results (pH=7.34, Partial Pressure of Carbon Dioxide (PCO_2)- 38.4 mmHg, Bicarbonate (HCO_3)- 22.4 mmol/L), and the Electrocardiogram (ECG) showed normal sinus rhythm.

During the neurological examination, the patient's pupils were bilaterally 3 mm in size and reactive to light. Involuntary non-rhythmic movements were observed in the left upper and lower limbs. Assessing power in the left limbs was difficult due to the involuntary movements, but power in the right upper and lower limbs was normal (5/5). The bilateral lower limb plantar reflexes were flexor, and the cranial nerve examination was normal. Sensations were intact. Gait assessment and coordination could not be performed due to the involuntary movements. Respiratory examination

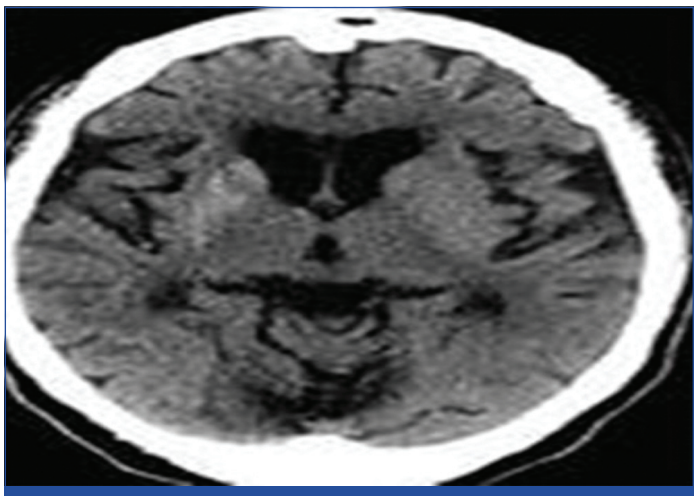
revealed normal vesicular breath sounds, the abdomen was soft and non-tender, and the cardiovascular system examination showed normal heart sounds with no murmurs. Other blood tests were within normal limits, except for a Glycated Haemoglobin (HbA1C) level of 17.1% [Table/Fig-1].

Parameters	Results	Parameters	Results
Haemoglobin	13.4 g/dL	Total bilirubin	0.58 mg/dL
Total count	10.74×10^3/uL	AST	18 U/L
Platelets	275×10^3/uL	ALT	18 U/L
Urea	17.9 mg/dL	ALP	92 U/L
Creatinine	0.73 mg/dL	Calcium	7.9 mg/dL
Na/K/Cl	139/3.8/107 (mEq/L)	Magnesium	1.51 mg/dL
TSH	2.68 mIU/mL	PT/INR	2.7/1.06
HbA1C	17.1%	APTT	25.7

[Table/Fig-1]: Investigations.
TSH: Thyroid stimulating hormone

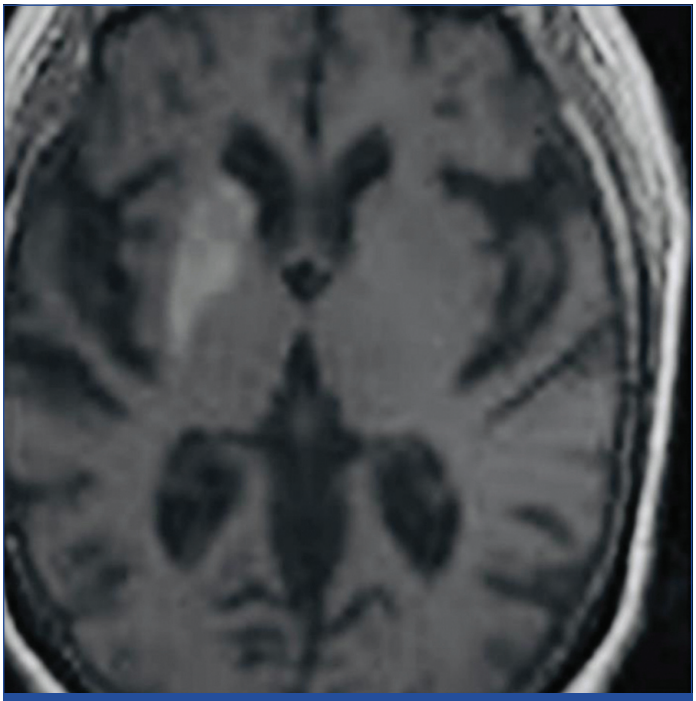
Imaging

Imaging studies were conducted, including a contrast CT scan of the brain, which revealed hyperdensity in the right basal ganglia [Table/Fig-2]. An MRI of the brain showed T1 hyperintensity in the right basal ganglia [Table/Fig-3]. In the ED, the patient received treatment with intravenous fluids (0.9% normal saline) and insulin for high blood sugar levels. Initially, an intravenous infusion of Inj. Actrapid 10 U was administered, followed by a 5 mL/hr infusion. The GRBS was monitored hourly. After 45 minutes of insulin infusion, the involuntary movements resolved. The intravenous infusion of Inj. Actrapid was then switched to a fixed-dose insulin



[Table/Fig-2]: Hyperdensity in right basal ganglia.

regimen, including Inj. Actrapid and Inj. Deglude. The patient was asymptomatic and was discharged with a prescription for Inj. Actrapid 8-8-8 subcutaneously and Inj. Deglude 12U subcutaneously at 10 PM. She was advised to follow-up in the Outpatient Department after two weeks.



[Table/Fig-3]: Hyperintense lesion in right basal ganglia in T1MRI.

DISCUSSION

The DS, also known as non-ketotic hyperglycaemic hemichorea/hemiballismus, is a hyperglycaemic condition associated with chorea or ballism and striatal changes in imaging [1]. It constitutes a rare hyperkinetic movement disorder, with ballism being more proximal and having a larger amplitude than chorea. The reported prevalence is 1 in 100,000 [2], which is likely an underestimation due to misdiagnosis as stroke or unfamiliarity with the clinical condition. Hemichorea can be caused by stroke, neoplasm, infection, demyelination, vasculitis, or metabolic causes. While common in Type 2 Diabetes Mellitus (T2DM), it can also occur in type 1 diabetes mellitus [3]. Hemichorea associated with NKH is a rare complication of diabetes mellitus, first described in 1960 by Bedwell SF [4]. The triad of NKH hemichorea consists of involuntary movements, high blood sugars with normal ketones, and basal ganglia hyperintensity on imaging [5].

A study by Chua CB et al., reviewed 176 patients with DS from 1992 to 2018. The study revealed that the condition was more common in women than men, with a mean age of onset of 67.6 years, an average blood glucose of 414 mg/dL, and glycated haemoglobin of 13.1%. Hemichorea (90.3%) was more commonly seen than chorea (9.7%). Neuroimaging commonly showed isolated putamen and combined putamen-caudate nucleus abnormalities. Successful treatment with glucose control was observed in only 25.7% of cases, while additional antichorea medications were required in 76.2% [1]. Chorea/ballism is due to dysfunction of the basal ganglia and subthalamus. Causes include DS, cerebrovascular, infectious, toxic, autoimmune, and malignancy [5]. The pathophysiology in NKH states is due to a shift in brain metabolism to the anaerobic pathway and inhibition of the Krebs cycle, leading to rapid depletion of Gamma-Aminobutyric Acid (GABA) and reduced inhibitory signals to the striatum and basal ganglia, resulting in hyperkinetic movements [1,6].

In patients with ketosis, Gamma Aminobutyric Acid (GABA) can be resynthesised by using acetoacetate produced in the liver, thereby reducing the incidence of DS in patients with diabetic ketosis or diabetes ketoacidosis [7]. The condition is more prevalent in females of perimenopausal age groups. The exact mechanism of this occurrence is unclear and less proven, although it is hypothesised to be due to a reduction in oestrogen receptors in this age group, causing increased sensitivity of the nigrostriatal dopamine receptors and resulting in hyperkinetic movements [1,8].

Imaging studies are characteristic of DS, as the name implies. Isolated involvement of the putamen and combined involvement of the putamen and caudate nucleus were the most common findings on neuroimaging studies [9]. MRI is considered the most sensitive imaging technique for DS. CT scans show high attenuation in the contralateral striatum, while MRI shows high T1 signals in the same areas [1,3]. Imaging abnormalities may be caused by petechial haemorrhages, mineral deposits (such as calcium or magnesium), myelin destruction, or infarction with astrocytosis [1,10]. Neuroimaging findings may return to normal or persist for up to six months after clinical recovery [11].

The case study revealed that hemichorea could be successfully treated with glucose control alone. In most other studies, additional antichorea medications were required for symptom control. The main types of antichorea medications used are antipsychotics, GABA-receptor agonists, selective serotonin reuptake inhibitors, and dopamine-depleting agents [7]. Haloperidol was the most frequently used medication for chorea treatment [3,12]. Case reports on NKH hemichorea/chorea are described in [Table/Fig-4] [3,11-13].

S. No.	Study (Year)	Age	Clinical presentation	Imaging	Treatment
1	Homaida M et al., (2014) [3]	71-year-old woman	Involuntary movements of the right upper limb and right lower limb	High signal in the left basal ganglia on MRI	Glycaemic control with insulin and haloperidol.
2	Ray S et al., (2015) [11]	61-year-old man	Involuntary movements of the left upper limb and left lower limb	T1 hyperintensity in the right putamen on MRI	Glycaemic control with insulin.
3	Kaeley N et al., (2021) [12]	55-year-old woman	Involuntary bilateral upper limb and lower limb movements	T1 hyperintense lesions in bilateral basal ganglia	Glycaemic control with insulin and haloperidol.
4	Huang X et al., (2022) [13]	71-year-old man	Involuntary movements of the right upper and right lower limb	Infarct and features of Diabetic Striatopathy (DS) in left striatum	Glycaemic control with insulin and clonazepam.

[Table/Fig-4]: Literature review [3,11-13].

CONCLUSION(S)

The DS is a poorly understood and unusual condition, predominantly seen in perimenopausal age groups with uncontrolled blood glucose. It usually presents with hyperkinetic movement. This presentation, along with the imaging findings in the striatum, is pathognomonic of the disease. This condition has a good prognosis and is quickly reversible with adequate glucose control.

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