CASE REPORT

Rare case of chorea-hyperglycaemia-basal ganglia (C-H-BG) syndrome

Sajjad Ahmad,¹ Priya Mohan Babu,¹ Lavanya Shenbagaraj,² Lindsay George³¹Department of Diabetes and Endocrine, University Hospital Llandough, Llandough, UK²Department of Gastroenterology, University Hospital Llandough, Llandough, UK³Department of Diabetes and Endocrinology, University Hospital Llandough, Llandough, UK

Correspondence to

Dr Sajjad Ahmad,
drsajjad618@yahoo.com

Accepted 30 March 2018

SUMMARY

An 83-year-old woman presented with acute-onset haemichorea and haemiballism particularly affecting the left side of the body. She was known to have type 2 diabetes, which was poorly controlled with sitagliptin. She was hyperglycaemic but not ketotic or acidotic. After she was started on insulin and good glycaemic control was achieved, her abnormal movements dramatically improved. MRI of the brain showed a T1-weighted hyperintense lesion on the right basal ganglia, which is typical of chorea-hyperglycaemia-basal ganglia syndrome. Other causes of chorea, for example, Huntington's disease, Sydenham chorea, Wilson's disease, malignancy, systemic lupus erythematosus, haemorrhage/infarction, thyroid dysfunction, drug-induced chorea and antiphospholipid syndrome, were excluded or deemed less likely given her rapid response to achieving near euglycaemia.

BACKGROUND

Physicians need to know about this rare but important complication of hyperglycaemia as early diagnosis and management will result in resolution of symptoms and in better patient outcomes. This will also help avoid more expensive and invasive investigations.

CASE PRESENTATION

An 83-year-old woman presented with a 2-week history of continuous, involuntary, non-rhythmic movements predominantly involving the left upper and lower limbs. She denied any headache, and visual or speech disturbances. She reported osmotic symptoms of polyuria and polydipsia. There was no history of falls or head injury prior to her admission.

Her medical history included type 2 diabetes mellitus, ischaemic heart disease, hypertension and atrial fibrillation. She was a non-smoker with no history of alcohol intake.

Medications included sitagliptin, warfarin, bisoprolol, bumetanide, solifenacin, lansoprazole, irbesartan, digoxin, calcium and atorvastatin.

Neurological examination demonstrated left-sided periodic choreiform and ballistic movements, which improved during sleep. There was no dysmetria, ataxia or dysidiadochokinesis. Her pupils were equal and reacted normally to light. No cranial nerve deficit was noted. Deep tendon reflexes were normal with no motor or sensory deficit in any limb.

INVESTIGATIONS

Initial laboratory tests revealed hyperglycaemia of 30.8 mmol/L with hyponatraemia of 129 mmol/L, suggesting pseudohyponatraemia. She had an acute kidney injury with a creatinine of 130 µmol/L presumed to be due to dehydration secondary to glucotoxicity. Her venous pH was 7.41, bicarbonate was 26.9 mEq/L and blood ketones were normal. The remaining blood test results on admission are shown in [table 1](#).

A CT head scan was performed which showed no evidence of acute intracranial pathology. The differential diagnoses included metabolic-related asymmetric chorea, a vascular event, autoimmune causes or a neurodegenerative cause.

Subsequently an MRI of the head showed T1-weighted (T1W) hyperintensity on the right globus pallidus, which was highly suggestive of hyperglycaemia-induced haemichorea-haemiballism syndrome ([figure 1](#)). She was started on Lantus insulin and her glycaemic control improved. This had a dramatic effect on her choreiform movements, which completely resolved. She had normal caeruloplasmin, Anti-streptolysin O (ASO) titre, Anti-neutrophil cytoplasmic antibody (ANCA), anti-GAD (glutamic acid decarboxylase), antivoltage-gated potassium channel, anticardiolipin and glycine receptor antibodies, B₁₂, folate, ferritin, thyroid function tests and erythrocyte sedimentation rate (ESR).

DIFFERENTIAL DIAGNOSES

- ▶ Huntington's disease.
- ▶ Sydenham chorea.
- ▶ Wilson's disease.
- ▶ Malignancy.
- ▶ Systemic lupus erythematosus.
- ▶ Basal ganglia
- ▶ Haemorrhage/infarction.
- ▶ Thyroid dysfunction.
- ▶ Drug-induced chorea.
- ▶ Antiphospholipid syndrome.

OUTCOME AND FOLLOW-UP

This woman had hyperglycaemia-induced haemichorea-haemiballism syndrome, which is a rare but recognised complication of poorly controlled diabetes in the medical literature. She had significant hyperglycaemia on once-daily sitagliptin with a background of type 2 diabetes. Her gliclazide was stopped 3 months prior to this admission when she was admitted with pneumonia. During this admission she had

To cite: Ahmad S, Mohan Babu P, Shenbagaraj L, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-223920

Table 1 Blood results

Biochemical profile		
Sodium	129 mmol/L	133–146
Potassium	4.8 mmol/L	3.5–5.3
Urea	7.6 mmol/L	2.5–7.8
Creatinine	130 µmol/L	46–92
Bilirubin	17 µmol/L	<21
Protein	72 g/L	60–80
Albumin	34 g/L	35–50
Globulin	38 g/L	22–43
Alkaline phosphatase	97 U/L	30–150
Alanine transaminase	31 U/L	<50
Calcium	2.44 mmol/L	2.20–2.60
Phosphate	0.94 mmol/L	0.80–1.50
Haemoglobin	131 g/L	115–165

symptomatic hyperglycaemia with a very high haemoglobin A1c of 132 mmol/mol. After achieving glycaemic control with insulin, her involuntary movements completely resolved. Follow-up MRI was not pursued due to improvement in her symptoms and increased frailty.

DISCUSSION

The exact mechanism of this syndrome is poorly understood, but it is thought to be related to hyperglycaemia-induced alteration in cerebral blood flow autoregulation, causing decreased perfusion with resulting activation of anaerobic metabolism and thus

Figure 1 MRI of the lesion in the basal ganglia.

depletion of the inhibitory neurotransmitter gamma aminobutyric acid in the basal ganglia.^{1,2}

Hyperviscosity associated with hyperglycaemia causes damage to the blood–brain barrier, leading to temporary ischaemia of susceptible striatal neurons.³

This was first described by Bedwell⁴ in 1960 when he reported a case of a 57-year-old woman with haemiballism that resolved as the blood glucose levels were normalised. It was Yahikozawa *et al*⁵ who first reported three patients with diabetes with the unique combination of haemiballism and striatal hyperintensity on T1W MRI as a new syndrome in 1994.

Patients reported in the literature are typically elderly women of Asian origin with known type 2 diabetes, but there are reports of symptoms coinciding with the first presentation of diabetes.

Involuntary movements resolve quickly in most of the cases, but some may take many months to resolve. A literature search reveals that radiological changes take 2–11 months to completely disappear.⁶

Learning points

- Chorea-hyperglycaemia-basal ganglia syndrome is a rare but recognised complication of diabetes.
- This is reversible in most cases with prompt achievement of euglycaemia.
- Dramatic response to glucose-lowering agents and T1-weighted lesions in the basal ganglia on MRI are key to the diagnosis.
- Patients with no history of diabetes should have their blood glucose and haemoglobin A1c checked as this could be the first presentation of diabetes.

Contributors SA: wrote the discussion with references and edited the case history. LS: wrote the case history. PMB: obtained consent and helped in writing the discussion. LG: critical analysis and final review of the case.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent Next of kin consent obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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