Diabetes and neurology: hemichorea–hemiballism in hyperglycaemia

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Introduction

Diabetes affects a large proportion of the population, with the incidence rising. It also has a wide range of complications. We describe here an uncommon neurological complication of poor diabetic control. Prompt recognition of this condition is essential to improve the outcome.

Case presentation

A 71-year-old Chinese man with longstanding diabetes presented with a 4-day history of progressively worsening uncontrollable continuous non-rhythmic right upper limb movements which resolved during sleep. One week prior, his general practitioner noted poor glycaemic control and started him on sitagliptin. Two days later, he reported right shoulder tingling, gradually spreading down the right arm. This was followed, a few days later, by right hand involuntary movements, gradually affecting his entire right upper limb. The patient visited a Chinese medicine practitioner who performed acupuncture and cupping, eventually presenting to hospital after no improvement.

His past medical history included hypertension, hypothyroidism and type 2 diabetes mellitus with poor control, partially due to refusal of insulin because of needle phobia. His regular medications included metformin 500 mg bd, sitagliptin 25 mg od, amlodipine and levothyroxine.

Neurological examination showed right upper and lower limb involuntary movements consistent with hemichorea–hemiballism (HCHB). Blood tests including full blood count, coagulation, liver and kidney function, bone profile and C-reactive protein (CRP) were normal. His blood sugar was 42.4 mmol/L (range 3.0–7.7) with ketones of 0.8 mmol/L (0.6–1.5) and serum osmolality of 297 mOsm/kg (275–295) with Osm gap 12.0 mOsm/kg (<10). His HbA_{1c} was 141 mmol/mol (20–41).

Computed tomography (CT) of the head showed unilateral left striatum faint hyperdensity, with sparing of the internal capsule and with no mass effect, suggestive of HCHB due to hyperglycaemia. Subsequent magnetic resonance imaging (MRI) of the brain was normal.

Intensive glycaemic control was started with insulin. He was treated with clonazepam 500 µg three times a day (tds), to be uptitrated to 1 mg tds. He improved and was discharged, with diabetic clinic follow-up and plan to start risperidone.

Discussion

HCHB presents as continuous involuntary high-amplitude movements affecting one side of the body. HCHB is a rare syndrome with a prevalence of less than 1/100,000; the majority of individuals affected are Asian women in their 7th decade.[1] The increased incidence in Asian populations suggests a genetic predisposition.[2] Ischaemic/haemorrhagic stroke is the most common cause of HCHB, followed by non-ketotic hyperglycaemia. Other aetiologies are shown in Table 1. [3]

The pathophysiology of HCHB due to hyperglycaemia (also known as C-H-BG: chorea, hyperglycaemia, basal ganglia syndrome) is still uncertain. One suggested mechanism is disruption of the blood–brain barrier (BBB) and transient ischaemia of vulnerable neurons caused by hyperviscosity-related hyperglycaemia. Additionally, hyperglycaemia may also impair cerebral autoregulation, causing anaerobic metabolism activation and depletion of gamma-aminobutyric acid (GABA), the main striatal inhibitory neurotransmitter.[2] Interestingly, HBHC may present a few weeks after blood glucose levels are controlled, suggesting a delayed reaction to severe hyperglycaemia.

The majority of described cases have a good prognosis. In conclusion, a low threshold in screening for hyperglycaemia in HCHB, even when there is no known history of diabetes,[4] is essential, as prompt diagnosis and management may significantly improve the outcome. ■

Table 1. Less common causes of hemichorea–hemiballism

Primary/inherited	Secondary/acquired
Wilson's disease	Vitamin B12 deficiency
Multiple sclerosis	Vascular
Huntington's disease	Autoimmune
Motor neurone disease	Endocrine
Demyelinating disease	Drugs
	Brain trauma or tumour
	Toxins

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Conflicts of interest

None declared.

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