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Case Report

A rare case of hemichorea in the setting of non-ketotic hyperglycaemia with subtle radiological changes: a case report

Liam Mahoney Clifford *and Ammar Wakil

Department of General Medicine, Wyong Hospital, Wyong, Wyong, New South Wales 2259, Australia
*Correspondence address. Wyong Hospital, Pacific Highway, Wyong, New South Wales 2259, Australia, Tel: (02) 4394 8000; Fax: (02) 4394 4844; E-mail: liamclifford@live.co.uk

Abstract

Movement disorders have been associated with hyperglycaemia including chorea, hemiballismus and choreoathetosis. In almost all documented cases, there is an association between clinical and radiological findings. We report a case of an 82-year-old man with hemichorea in the setting of hyperglycaemia and poorly controlled type 2 diabetes. He had subtle striatal changes on his radiology, and with intravenous fluids and insulin, his involuntary movements resolved on day 4, which correlated with improvement in glycaemic control. He was followed up through our local insulin stabilization programme after discharge.

INTRODUCTION

Neurological complications due to hyperglycaemia encompass stroke, peripheral neuropathies, movement disorders and changes in behaviour and consciousness. Diabetic striatopathy is a combination of radiological changes and movement disorders including chorea, ballism and choreoathetosis [1, 2]. Predominantly, this occurs in older women and in Asian populations with type two diabetes mellitus (T2DM) [3]. Usually, correction of a hyperglycaemic state results in partial to complete resolution of the movement abnormalities, and associated radiological changes [2]. Some cases require pharmacological management if the movement disorder persists despite improving glycaemic control. This case report demonstrates the rare neurological manifestations of hyperglycaemia and poorly controlled diabetes.

CASE REPORT

This 82-year-old gentleman had a medical history of T2DM, ischaemic heart disease, hypertension, bilateral total knee replacements, left hip osteoarthritis, macular degeneration and chronic lower back pain (managed with amitriptyline 25-mg nocte). He was a heavy drinker (currently drinks two standards a day) and is a non-smoker who presented to the Emergency Department following a fall with no signs of alcohol withdrawal or relapse, on a background of 2 weeks of right-sided involuntary movements. He had changes in his behaviour, notably, short temper and irritability. Physical examination demonstrated a normal neurological examination except for writhing, dancing-like movements in the right side of the body affecting his face, arm and leg, consistent with hemichorea with no other signs of Parkinsonism. These movements worsened with activity and disappeared with sleep. However, the movements were not incapacitating. His diabetes was poorly controlled with diabetic retinopathy and nephropathy managed with metformin, and prior to admission reported no recent hypoglycaemic episodes. Biochemistry revealed a non-ketotic hyperglycaemic state with a capillary blood glucose level of 25 mmol/L, serum osmolality of 300 mOsm/kg (reference range (RR) 280–300) and a calculated serum osmolality level of 291. His urea, electrolytes, liver and thyroid function tests as well as corrected calcium and magnesium levels were normal. Serum-glycated haemoglobin was 14.6% (136 mmol/mol, RR 20–42). He underwent a non-contrast computed tomography (CT) brain scan which demonstrated abnormal attenuation in the basal ganglia bilaterally, however, slightly more prominent on the left, and no other intracranial abnormalities (Fig. 1). His magnetic resonance imaging (MRI) brain scan (Fig. 2) showed moderate microangiopathy and subtle striatal abnormalities bilaterally.

Expert neurological opinion found no evidence of a causative medication, structural lesion, neurodegenerative disorder, infection, inflammatory process or paraneoplastic syndrome underlying the hemichorea. Initial management involved intravenous regular insulin Actrapid 4 units/hour and then was switched to Ryzodeg 70/30 degludec/aspart co-formulation insulin 30 units, given subcutaneously pre-breakfast. He received 4 days of treatment with subcutaneous insulin Ryzodeg and the movements completely resolved, with the blood glucose levels decreasing initially to 11–24 mmol/L during intravenous infusion and then to 6–12 mmol/L on subcutaneous insulin. After discharge, his insulin doses were titrated according to home self-monitoring

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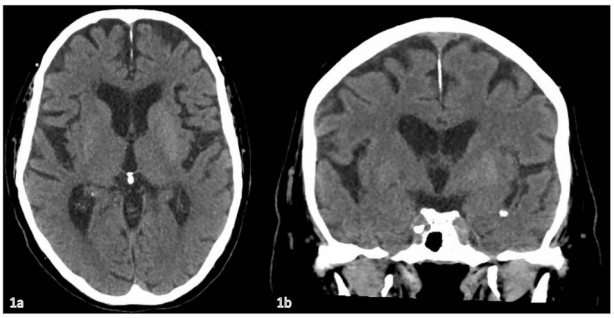

Figure 1. CT brain scan. (1a) Axial view and (1b) coronal view. Mild hyperdensity in both basal ganglia was present, slightly more prominent on the left.

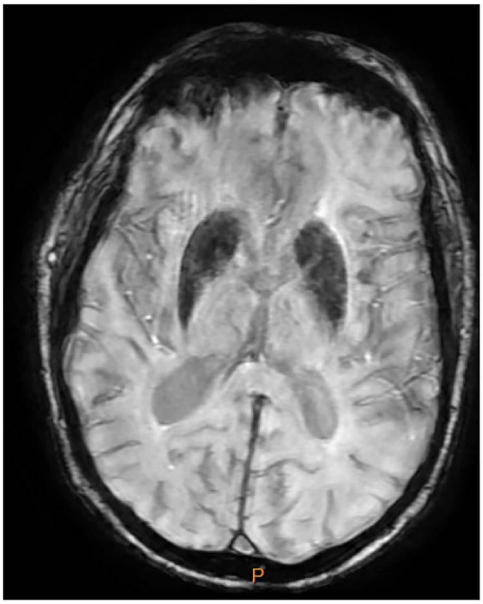

Figure 2. MRI at the level of the basal ganglia: SWI sequencing with signs of mineralization.

blood glucose levels through the insulin stabilisation programme to ensure good diabetes control and prevent relapse of the hemichorea. It was concluded that the hemichorea was secondary to hyperglycaemia and poorly controlled T2DM.

DISCUSSION

Involuntary movements in hyperglycaemic syndromes typically involve the limbs; however, they less commonly affect muscles of the jaw and tongue, and usually are unilateral [4]. Generally, a significant positive correlation exists between the clinical and imaging findings, with radiological changes occurring usually contralateral to the affected limbs [3]. A lack of corresponding radiological changes during the acute phase of hyperglycaemic chorea is rare and may be misleading [2, 5, 6]. Patients with positive findings can be misdiagnosed as an acute intracerebral haemorrhage. The differential diagnosis for T1-weighted hyperintensities in the basal ganglia includes hypoxia, chronic liver disease, manganese toxicity, disorders of calcium metabolism and rarer diseases such as lupus. Other causes are alcohol and methanol toxicity, carbon monoxide poisoning, hypoxaemia and deep cerebral venous sinus thrombosis [2, 3, 7]. An important cause of bilateral hyperdense basal ganglia is calcification, which commonly occurs due to age and can mask pathologies such as striatopathy.

In non-ketotic hyperglycaemia, brain metabolism shifts to the alternative anaerobic pathway with rapid depletion of gamma-aminobutyric acid (GABA), resulting in hyperkinetic movements. In the presence of ketosis, GABA can be resynthesised by using acetoacetate thereby explaining the less common occurrence of striatopathy in diabetic ketoacidosis [2]. Hemichorea related to diabetes slowly improves following restoration of glycaemic control [3, 8]. It is postulated the quick resolution is due to rapidly reversing changes in the brain parenchyma [2, 3, 8]. Importantly, an investigation for associated paraneoplastic conditions requires consideration in this age group if symptoms persist or recur.

The mainstay is treatment of hyperglycaemia, usually with insulin and intravenous fluids to enable rapid but safe correction of the glucose levels [1–3]. Additional therapies include anticonvulsants, GABAergic agents, selective serotonin reuptake inhibitors, benzodiazepines, and dopamine-depleting inhibitors. In this case there was rapid spontaneous resolution following glycaemic control and hydration, thus evading the need for symptomatic control [9]. A rapid decrease in intravascular osmolality results in water movement into brain cells, causing cerebral oedema [10]. The current postulated pathophysiology underpinning diabetic striatopathy includes astrocyte infarction, petechial haemorrhages, mineral deposition, methaemoglobin deposition, gliosis, atrophy, myelinolysis and myelin destruction [2, 6]. This case supports the phenomenon of petechial haemorrhages, given the subtle abnormalities on imaging, which is consistent with previous literature, particularly where patients were monitored, and their lesions resolved after correction of hyperglycaemia [1, 3, 5, 9].

CONCLUSION

There are very few cases documented of hyperglycaemia with hemichorea or hemiballismus and unremarkable imaging. The mechanism is unclear but microhaemorrhages in the brain parenchyma that resolve with resolution of the hyperglycaemia are postulated. This case highlights the subtlety and novelty in detecting radiological abnormalities, the importance of recognising hyperglycaemia as a cause of involuntary movements, and to avoid rapid correction of very high hyperglycaemia due to the risk of cerebral oedema. We present this case to help educate fellow clinicians regarding the difficulties detecting changes with diabetic striatopathy, and the importance of clinical examination in these cases.

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CONFLICT OF INTEREST STATEMENT

None declared.

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DATA AVAILABILITY STATEMENT

Data sharing is not applicable to this article as no datasets were generated or analysed during the current study.

ETHICAL APPROVAL

No ethical approval was required for this case report.

CONSENT

The authors obtained written informed consent from the patient for this published case.

GUARANTOR

Ammar Wakil is the guarantor of this publication.

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