Diabetic striatopathy: a rare condition and diagnostic dilemma

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DESCRIPTION
A 71-year-old woman presented with a 2-week history of right arm and leg choreiform movements following a sudden weakness in the right arm and leg. The patient had a medical history of type 2 diabetes mellitus (DM), essential hypertension, bipolar disorder and hypothyroidism. She did not have a history of diabetic retinopathy. She was admitted 3 months ago with COVID-19 infection, during which her antidiabetic medication was reduced following recurrent episodes of hypoglycaemia. Her diabetes was poorly controlled on current admission with Glycated haemoglobin (HbA1c) levels measured at 106 mmol/mol compared with 60 mmol/mol on earlier admission, while her blood sugar ranged between 18 and 20 mmol/L. Gliclazide was introduced as an addition to her antidiabetic management of metformin, (1 g twice a day), on her admission day. Owing to typical choreiform movements in the patient’s right arm, shoulder and leg, the assessment of power was difficult. The patient was hypertensive on admission (182/76) and was suffering from delirium, but there were no other positive neurological findings. Unfortunately, the patient did not have lactate or venous gas levels measured during the current admission. The patient did not have any changes to suggest diabetic retinopathy.

A CT scan showed high attenuation in the left basal ganglia, initially thought to represent stroke with luxury perfusion and/or petechial haemorrhages (figure 1). The patient was initially managed as stroke and was also administered haloperidol to limit her involuntary movements, without significant improvement. The patient was also given aspirin and atorvastatin. Subsequent MRI scans showed high T1 signal in the left basal ganglia, with no changes on other sequences (figure 2). The radiological changes were thought to represent diabetic striatopathy and due to continued poor diabetic control (blood sugar 14.6–20.2 mmol/L), the patient was administered humulin M3, and the gliclazide was stopped. This change in medication resulted in a complete resolution of the patient’s haemichorea after 1 hour following the initiation of insulin therapy. Haloperidol was successfully withdrawn without recurrence of symptoms. There was also a noticeable resolution of the patient’s delirium as her glycaemic control improved.

Interestingly, the patient was admitted 2 years prior to the current admission with a 10-day history of involuntary movements of the left arm associated with left arm weakness. The patient had an HbA1c level at 61 mmol/mol. A CT scan and MRI of the head were carried out, which did not indicate organic pathology; therefore, the patient was discharged with a diagnosis of functional movement disorder.

Diabetic striatopathy is a rare disorder that usually presents with a variety of hyperkinetic movement disorders, and is associated with poor glycaemic control. It is more prevalent in elderly diabetic women with poor glycaemic control (mostly type 2 DM but can occur in type 1 DM).

About 90% of cases of diabetic striatopathy occur in members of Asian ethnic groups. We believe that our case report is unique as the patient is Caucasian; however, there have been an increasing number of reports from Europe, north and Latin America more recently. The exact causative mechanism of this condition is unclear, although it is thought to be due to a decline in oestrogen receptors in postmenopausal women, which causes increased sensitivity of the nigrostriatal dopamine system receptors, resulting in chorea. Cerebrovascular insufficiency, petechial haemorrhage, hyperviscosity and depletion of both gamma-aminobutyric acid and acetylcholine secondary to metabolic changes have been suggested as possible mechanisms.

CT and MRI findings are characteristic; CT shows high attenuation in contralateral striatum (caudate nucleus and putamen), and MRI shows high T1 signal in the same areas as the most consistent feature, other changes being variable, while MRI is regarded as more sensitive. The exact cause of these findings has not been clearly identified in the literature. At least four hypotheses have been presented for these findings, including petechial haemorrhages, mineral deposits, myelin destruction and infarction with astrocytosis.

Petechial haemorrhage would explain the CT findings, but they are not seen consistently on MRI scans (on susceptibility images). Calcification could explain the CT scan findings, although this is unlikely due to reversible changes. Myelin destruction can show a high T1 signal on MRI; however, the presence of this damage is not supported by pathology reports. Reactive astrocytosis and an abundance of gemistocytes could explain the high T1 signal on the MRI; protein desiccation during Wallerian degeneration could explain CT findings.

We believe that the two latter processes combined could potentially explain all the imaging findings. On the susceptibility-weighted imaging sequence shown in figure 2F, we wondered about a couple of petechial haemorrhages or hyperdense vessels, although difficult to confirm since these areas also show mineralisation. If present, these may represent
neurovascular uncoupling, a syndrome usually associated with diabetic microangiopathy. The impaired regulation of cerebral blood flow in relation to energy crises may lead to vasogenic oedema and striatal cytotoxicity, and blood extravasation.6

Only a few cases are reported in the literature where patients have no radiological changes.7 Our patient is, therefore, relatively unique as she also did not have any imaging findings on the previous admission where she displayed similar symptoms. The patient’s HbA1c levels were relatively lower on the previous admission, however, suggesting that imaging changes could be related to glucose levels. Some reports suggest that COVID-19 can affect insulin resistance and challenge efforts to control blood sugar levels.8 9 The reduction of antidiabetic medication during the last admission may also have contributed to higher blood glucose levels. Evidence exists that the presence of diabetic retinopathy is an indicator of worse prognosis in patients with hyperkinetic disorders related to diabetes.7 In our patient’s case, however, the absence of evidence of retinopathy was associated with better prognosis and disappearance of the involuntary movements with tight glycaemic control, and we feel this supports the above hypothesis.

The prognosis of the condition was found to be excellent (one case series showed 74% of patients have complete resolution of the chorea over a period ranging from 1 hour to 10 months) with management of hyperglycaemia.10 Patients may sometimes require dopamine antagonists to alleviate their symptoms. Imaging changes usually resolve on follow-up imaging at 6 months.

This patient also tested positive for COVID-19 on the previous admission, although the effect of this infection on the patient’s current symptoms is not clear; however, some reports indicate that COVID-19 infection increases insulin resistance and worsens diabetic control.

In conclusion, we have presented a rare case of diabetic striatopathy, which remains a poorly understood condition. Our patient is unique in having negative imaging on a previous admission with the same features but had typical imaging findings on current admission. This case report is intended to serve as a reminder of this condition. The clinical and imaging findings are reversible, with an excellent prognosis, if the cause is correctly identified and managed with consistent glycaemic control.

Learning points

► Diabetic striatopathy is a poorly understood and lesser known condition, associated mainly with elderly females who have poorly controlled diabetes. The condition can cause haemichorea and hemiballismus, which are resistant to treatment with usual therapy.
► Diabetic striatopathy can present with a variety of kinetic disorders, and imaging findings are highly suggestive of the disease.
► This condition resolves quickly and has a good prognosis in most cases.
► Screening of diabetic retinopathy in a patient with diabetic striatopathy can give an indication of the likely prognosis of the hyperkinetic movement disorders associated with the disease.

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