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Acute hemichorea in a young type 1 diabetic

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Abstract

Purpose/Aim: Acute movement disorder is an uncommon presenting symptom in patients with diabetes mellitus. We report a 20-year-old lady with poorly controlled type 1 diabetes, who presented with acute hemichorea and was found to have two rare diabetes-related central nervous complications of diabetic striatopathy and severe moyamoya disease (MMD).

Materials and methods: She was treated with aggressive glycemic control; clonazepam and tetrabenazine as well as aspirin stroke prophylaxis for her MMD with resolution of her chorea 3 months later. She subsequently underwent cerebral revascularization surgery for her MMD.

Results: This case highlights the possible differentials of acute chorea in diabetic patients and explores the pathophysiological mechanisms that may underlie both conditions in patients with type 1 diabetes. **Conclusion:** We recommend performing both magnetic resonance imaging (MRI) and magnetic resonance angiogram (MRA) brain for comprehensive evaluation of diabetic patients with new onset chorea. Prompt and accurate diagnosis is crucial as it guides prognostication and treatment strategies.

Key words: diabetic striatopathy; moyamoya disease; movement disorder; chorea

Introduction

Diabetes mellitus (DM) is associated with a variety of disturbances in the peripheral and central nervous system, as a result of hyperglycemia, hypoglycemia, or secondary micro- and macrovascular complications^[1]. Acute movement disorder is an uncommon presenting symptom in diabetic patients but can manifest during various time points. Abnormal movements in hyperglycemic patients have been reported to include hemichorea-hemiballism, hemifacial spasm, paroxysmal kinesiogenic dyskinesia and opsoclonus myoclonus^[1]. In patients with hypoglycemia, chorea, ataxia, paroxysmal dyskinesias have also been reported^[1]. More commonly, abnormal movements arise from vascular complications of DM including strokes and mild Parkinsonism syndrome^[1].

We report a rare case of diabetic striatopathy and moyamoya disease (MMD) in a young type 1 diabetic lady who presented with acute hemichorea. We were interested to explore the possible common pathophysiological pathways resulting in these two rare conditions in type 1 diabetic.

Case report

A 20-year-old Malay lady with type 1 diabetes mellitus, vitiligo and hyperlipidemia, presented with a 2-day history of left-sided involuntary, continuous jerky and writhing movements. She was diagnosed with type 1 diabetes at 9 years of age when she presented with diabetic keto-acidosis with positive serum glutamic acid decarboxylase [GAD] autoantibodies. She had poor treatment compliance and disease control with a prior glycated haemoglobin at 14% and existing complications of nephropathy and peripheral neuropathy. At presentation, she was alert with distressing left-sided choreo-athetoid movements of her limbs. Initial investigations revealed severe non-ketotic hyperglycemia.

Diagnosis of diabetic striatopathy was made via magnetic resonance imaging (MRI) brain which showed high T1-weighted signal in the right basal ganglia (caudate nucleus and lentiform nucleus) with low T2-weighted and FLAIR signal. The diffuse weighted imaging (DWI) was normal, excluding acute infarcts (Figure 1a to 1c). Brain magnetic resonance angiography (MRA) done demonstrated severe stenoses in bilateral supraclinoid internal carotid arteries with surrounding prominent collateral vessels, suggestive of moyamoya vasculopathy (Figure 1d) which was thought to be an incidental finding. Workup including anti-phospholipid syndrome, vasculitis, systemic lupus erythematosus and homocysteine, was negative.

We instituted aggressive glycemic control and symptomatic control of her chorea with clonazepam and tetrabenazine. She was given aspirin stroke prophylaxis for her MMD. Three months later, her chorea completely resolved in keeping with diabetic striatopathy. However, the repeat MRI/MRA scans 6 months later showed persistent right basal ganglia signal abnormalities, volume loss and dystrophic calcifications, which is rare in cases of diabetic striatopathy. There were concerns that the basal ganglia changes were contributed by ischemia from her MMD. She subsequently underwent a cerebral revascularization surgery for her MMD involving indirect extracranial-to-intracranial (EC-IC) carotid artery bypass via encephaloduro-arterio-myosanagiosis (EDAMS). There has not been recurrence of the movement disorder.

Being a report on single case report, a formal DSRB approval was not required as per institutional policy. A written consent was obtained from the patient.

Discussion

Chorea is a relatively uncommon presentation in patients with DM. The most recognized cause is diabetic striatopathy which is characterized by a constellation of dyskinesia and radiological striatal abnormalities. Other differentials include basal ganglia strokes, bleed, tumours, encephalitis, neurodegenerative disorders, drugs, autoimmune conditions like systemic lupus erythematosus, vasculopathies like moyamoya disease, metabolic disorders including hypoglycemia etc. Rarely, diabetic striatopathy may be the first presentation of DM thus it should be included in the list of differentials for chorea, especially in elderly patients. A detailed history, clinical examination and evaluation for hyperglycemia or hypoglycemia is usually the first step. Further evaluation with MRI brain scan is required for diagnosis and to exclude other differentials. MRA brain scan is not routinely ordered, but we suggest that it be included in view of the possibility of MMD in DM patients.

Diabetic striatopathy is usually seen in the elderly Asian women with uncontrolled type 2 diabetes^[2]. The classic MRI finding includes contralateral corpus striatum hyperintensity on T1-weighted image, hypo or isotense on T2-weighted image and lack of restrictions on diffusion images^[3]. The MRI abnormalities usually resolves in few months however persistent hyperintensity up to years have been reported^[2,3]. Movement disorder generally resolves within 24-48 hours of normoglycemia however may persist for longer duration. There are a few hypotheses for this condition. Firstly, depletion of gamma-aminobutyric acid (GABA) related to non-ketotic hyperglycemia^[2]. Secondly, cellular acidosis and regional hypometabolism due to disruption of blood brain barrier^[2,3]. Thirdly, transient and incomplete striatal ischemia².

We initially attributed our patient's clinical symptoms to diabetic striatopathy. However, the concomitant finding of persistent basal ganglia radiological abnormalities which is rare in diabetic striatopathy and severe MMD in our patient posed a diagnostic consideration of its role in her acute chorea. MMD, a progressive stenotic angiopathy of the major vessels in the circle of Willis, from a combination of smooth muscle cell hyperplasia and luminal thrombosis, can also rarely cause chorea in 5% of affected patients, presumpitively through basal ganglia ischemia^[4].

Recent studies have challenged the regional metabolic failure and ischemia theory in both diabetic striatopathy and MMD^[3,5]. Instead, striatal hypermetabolism has been found to be a common pathophysiological pathway in both conditions resulting in chorea^[4,5]. Striatal hypermetabolism could represent activation of a direct pathway, an inhibitory signal pathway to the basal ganglia, which negatively regulates thalamocortical projection, resulting in activation of the motor circuit^[4].

The finding of both diabetic striatopathy and MMD, which are rare conditions predominantly in Asian populations, present in our patient with type 1 DM is unlikely to be fortuitous. Genetic predisposition towards basal ganglia dysfunction is highly likely^[6]. In addition, autoimmunity in DM may contribute to the development of both conditions. The autoimmune inflammatory processes may lead to endothelial damage resulting in MMD^[7,8]. The immunological reaction may also cause inflammation in basal ganglia circuits resulting in striatal hypermetabolism^[4,9,10].

Lastly, vascular abnormalities in MMD have been postulated to cause physical disruption of the normal striatal signaling^[4]. Interestingly, our patient had similar angiographic findings

(Figure 1e) as other reported cases of MMD-induced chorea— a dilated and extended lenticulostriate artery at the basal ganglia⁴] which may have disrupted the striatal signaling.

Conclusion

Our case highlights 2 rare causes of acute chorea in type 1 diabetic patients – diabetic striatopathy and MMD. We highly recommend both MRI and MRA brain evaluations for all diabetic patients with acute chorea. Prompt and accurate diagnosis is crucial as there are different treatment strategies such as that of stroke prophylaxis and re-vascularisation surgery in MMD. The relationship of chorea and striatal hypermetabolism, as well as the contribution of genetic, autoimmune and vascular factors, requires more investigations.

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Declaration of interest statement

The authors report no conflicts of interest.

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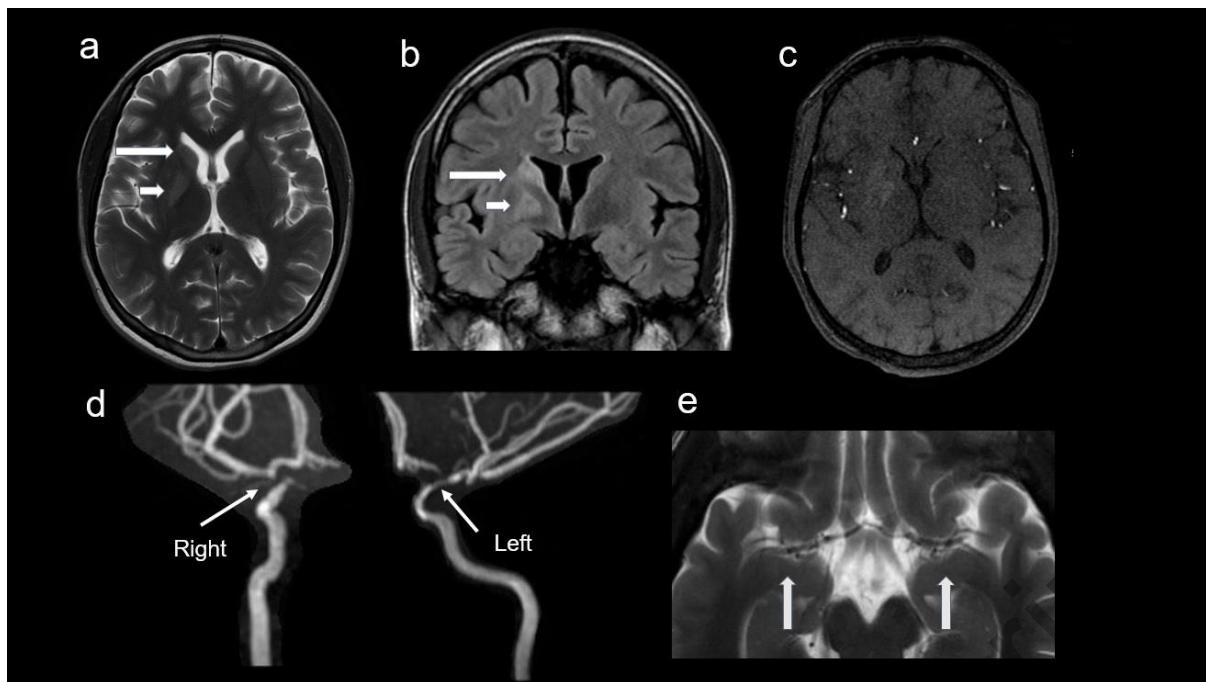


Figure 1: (a) Axial T2-weighted and (b) FLAIR images show hyperintense signal within the right caudate head (long white arrow) and lentiform nucleus (short white arrow). (c) Axial T1-weighted image shows corresponding T1-hyperintense signal, in keeping with diabetic striatopathy. (d) Time-of-flight MR angiography shows steno-occlusive changes at the bilateral terminal internal cerebral arteries (white arrows), extending into the proximal A1 and M1 segments on the left. (e) Of note is also the presence of prominent lenticulostriate vessels along the bilateral M1 segments