A Young Type 1 Diabetic with Acute Hemichorea: Rare central nervous system complications

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Background

Chorea is a rare presenting symptom in diabetic patients. Other than ischemic stroke, diabetic striatopathy (hyperglycemia-induced hemiballism-hemichorea) is a top differential in diabetic patients with acute chorea in association with non-ketotic hyperglycemia and characteristic basal ganglia lesion on neuroimaging. The chorea is often transient and reversible with aggressive glycemic control.

It is postulated that chronic hyperglycemia leads to hyperviscosity, in turn causing cerebral hypoperfusion, anaerobic metabolism and reduced γ-aminobutyric acid levels. This in turn leads to increased thalamocortical activity, causing abnormal movements. This is usually seen in elderly female patients of Asian descent with type 2 diabetes mellitus.

In addition, Moyamoya syndrome, an idiopathic chronic progressive angiopathy, can also present with chorea and has been found to be associated with autoimmune conditions such as type 1 diabetes mellitus.

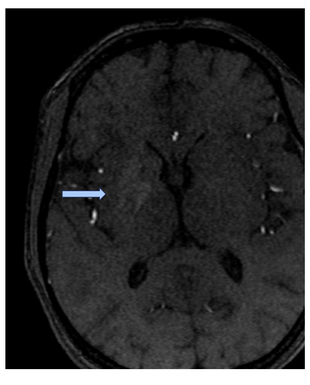
We describe a young adult with poorly controlled type 1 diabetes who presented with hemichorea and was found to have two rare diabetes-related central nervous complications of diabetic striatopathy and severe moyamoya disease (MMD), both of which could result in acute movement disorder.

Presenting History

A 20 year old lady presented acutely with involuntary writhing movements of the left upper and lower limbs for 2 days.

Her medical history is significant for poorly controlled type 1 diabetes mellitus since 9 years old. She was poorly compliant to insulin therapy. Her comorbidities include vitiligo, hyperlipidemia and steatohepatitis.

On examination, she had choreo-athetoid movements of the left upper and lower limbs, with occasional hemiballismus, and mild ipsilateral hemiparesis on her left side.



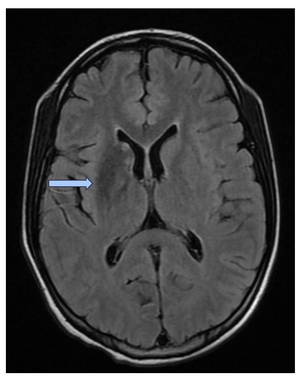


Figure 1: Abnormal signal in the right basal ganglia (involving both caudate nucleus and lentiform nucleus) with high T1W signal (left image) and low T2W/FLAIR signal (right image)

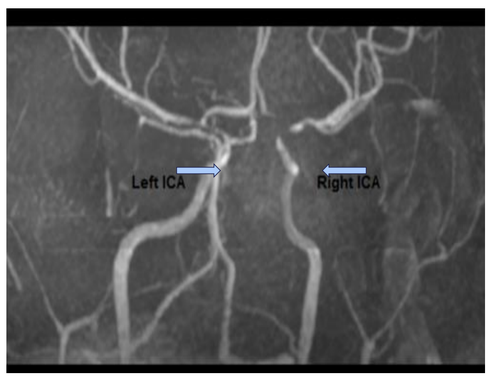


Figure 2: MRA showing severe stenoses in bilateral supraclinoid ICAs extending to the carotid T-junctions, with surrounding prominent collateral/ perforating vessels suggestive of a moyamoya pattern.

Clinical Evaluation

Emergent MRI brain demonstrated abnormal signal in the right caudate nucleus and lentiform nucleus (low T2W/FLAIR signal and high T1W signal) suggestive of diabetic striatopathy. [Figure 1] There is corresponding marked non-ketotic hyperglycemia (venous glucose 36.0 mmol/L) with a glycated haemogloblin (HbA1c) of 12%.

Incidentally, MR angiography brain revealed severe stenoses of both terminal internal carotid arteries with multiple collaterals, suggestive of severe bilateral Moyamoya syndrome. [Figure 2] A hypercapnoea challenge demonstrated exhausted vasodilatory reserves with paradoxical reduction of flow velocities. Extensive investigations for the aetiology of the Moyamoya syndrome that included autoimmune cerebral vasculitis, were unremarkable.

The patient was managed with aggressive glycemic control and symptomatic treatment for her movement disorder, with full resolution of her symptoms after 4 weeks. She was started on aspirin and advised for vascular bypass surgery for her Moyamoya syndrome.

She underwent cerebral revascularization surgery involving indirect extracranial-to-intracranial (EC-IC) carotid artery bypass via encephaloduro-arterio-myosyanagiosis (EDAMS) without any recurrence of chorea thus far.

Discussion

Moyamoya syndrome is a rare condition of which diabetes is a possible aetiology. Chorea is a rare presenting symptom, secondary to ischaemic dysfunction and imbalance in the complex basal ganglia circuitry.

We describe a young adult with poorly controlled type 1 diabetes, hyperlipidemia and smoking, who presented with acute hemichorea. Given the risk factors, our initial differential was that of a thrombotic stroke. She was eventually found to have 2 rare diabetes-related central nervous complications of hyperglycemia-induced striatopathy and severe Moyamoya syndrome, both of which could result in acute movement disorder.

MRI and MRA brain are important investigations to help differentiate the three causes. Aggressive glycemic control often results in resolution of the movement disorder in diabetic striatopathy. However, it is important to ensure there are no concomitant diabetes-related vascular stenoses, that would increase stroke risk and necessitate different management.

References:

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