

## A case of Diabetes Striatopathy in an Elderly Female

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### ABSTRACT

Diabetes Striatopathy, also known as Non-ketotic hyperglycemic hemichorea or Chorea Hyperglycemia Basal Ganglia Syndrome is rare complication of uncontrolled diabetes. This syndrome has been reported more frequently in elderly Asian women and symptom resolution is typically associated with serum glucose levels and plasma osmolarity normalization.

Herein, we report a case of 75 years old female with CAD-TVD, Hypertension & Type -2 Diabetes Mellitus under oral hypoglycemic drugs presented with intermittent involuntary movement of face and upper limb who was managed for diabetic striatopathy and showed symptomatic improvement in third day of admission.

**Keywords:** Corpus straitum; Diabetes Striatopathy.

### INTRODUCTION

Diabetes Striatopathy (DS), also known as Non-ketotic hyperglycemic hemichorea (NNH) or Chorea Hyperglycemia Basal Ganglia (C-H-BG) Syndrome is rare complication of uncontrolled diabetes. In general, the term "Diabetic Striatopathy" means hyperintense signal changes on MRI confined to the corpus straitum with contralateral hyperkinetic movement disorder. This condition is mainly reported in the elderly Asian population with a female preponderance. It is a cause of hemichorea-hemiballismus syndrome.

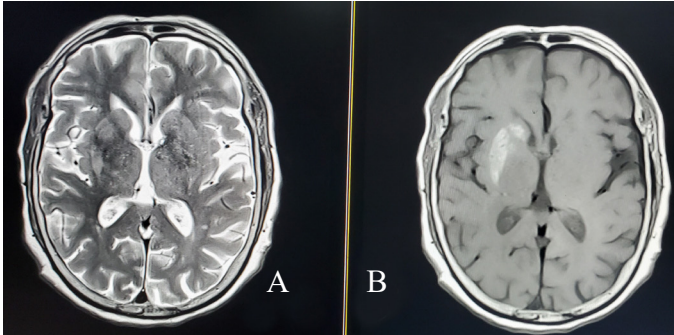
The exact underlying pathophysiology remains unknown but, assumed to be caused due to hyperviscosity secondary to increased blood sugar level, leading to regional blood-brain barrier disruption and metabolic damage. Moreover, the decrease in striatal blood flow causes depletion of gamma-aminobutyric acid (GABA) with resultant dyskinesia.<sup>1</sup>

### CASE REPORT

A 75 years old women, known case of CAD-TVD, HTN and Type 2 DM, presented with complain of intermittent abnormal involuntary movement of face and the left upper limb for 1 month, which has increased in severity for 3 days and is continuous. No history of any altered sensorium or any motor weakness. No history of loss of consciousness. Her other systemic examination was unremarkable. Patient is diabetic for 12 years under oral hypoglycemic drugs.

Investigations revealed initial plasma glucose level of 572.5 mg/dl, with HbA1c of 17.2 %, pH 7.38, urine for acetone was trace initially, and next day repeat urine for acetone was negative. CT head was done 1 month back revealed "well defined homogenous hyperdensity in the right caudate nucleus and lentiform nucleus. MRI brain was done, showed hyperintense signal in T1w, T2w and FLAIR sequences in the right lentiform nucleus and head of right caudate nucleus. On admission, her plasma sugar

was managed with insulin along with oral hypoglycemic drugs, also with GABAergic drug clonazepam 0.25 mg BD and atypical neuroleptics, olanzepine 5 mg HS. Patient showed marked improvement symptomatically within 3 days of admission. Patient is doing well and under follow up with aplan to repeat MRI brain after 3 months



**Figure 1: Plain MRI head showing hyperintensity signal in T2w (A) and faintly hyperintense signal in T1w (B) in the right lentiform nucleus and head of right caudate nucleus.**

## DISCUSSION

Diabetic striatopathy is an uncommon complication of diabetes, commonly seen in elderly Asian females, usually presenting as sudden onset of involuntary hyperkinetic movements (chorea-ballism) and also with specific neuroimaging findings; hyperintense signal changes on T1w and T2w confined to striatum.<sup>2</sup> DS typically happened in patients with long-standing poor control of DM which was further confirmed by remarkable elevation of blood glucose and glycated hemoglobin level, and it could even occur after correction of hyperglycemia.<sup>3</sup> Though DS is well known complication of diabetes in adults, it is a rare entity in childhood with easy-to-treat neurologic manifestation.<sup>4</sup>

CT of the brain initially is normal, but later it can demonstrate subtle hyperdensity in the striatal region (caudate nuclei and putamen). Findings tend to be contralateral to the body side affected by hemiballistic, hemichoreic movements. MRI of the brain is the modality of choice for assessing possible non-ketotic hyperglycemic hemichorea and typically demonstrates signal changes particularly in the putamen and/or caudate. If the changes are unilateral, then they are contralateral to the symptomatic side.<sup>5</sup> MRI reveals hyperintense signal changes in T1, variable but generally hypointense in T2/FLAIR, high diffusion signal in DWI. Overall, the T1 hyperintensity is the most consistent finding of the disease. Other associated findings do not present to the same frequency and tend to vary. Imaging findings gradually resolve after hyperglycemia correction. However, they tend to return to baseline more slowly

than the clinical findings. During hyperglycemic crisis, the Krebs's cycle activity is suppressed in the brain, so cerebral metabolism shifts to alternative anaerobic pathways. In HHS, this shift causes the brain to metabolize gamma amino butyric acid (GABA) into succinic acid via the semialdehyde pathway, leading to rapid depletion of GABA levels, which is an inhibitory neurotransmitter. Thus, its depletion causes disinhibition of the thalamus by the medial globus pallidus, resulting in hyperkinetic movements.<sup>6</sup> Beside DS, other etiologies of chorea/ballism include cerebrovascular, autoimmune, toxic, malignant and infectious illness. In most patients, rapid glucose lowering therapy and hydration quickly resolves the symptoms along with antichorea medications. Treatment usually involves control of hyperglycemia with proper hydration for correction of metabolic derangements and also might require anti-chorea medications such as antipsychotics, GABAergic drugs, selective serotonin reuptake inhibitors or dopamine-depleting agents.<sup>7</sup>

The time of resolution varies from 2 to 28 days. In a large meta analysis of 53 cases, two third of the patients had complete resolution of the symptoms during follow up after 3 months, one third of them only with control of hyperglycemia.<sup>8</sup>

## CONCLUSION

Diabetic striatopathy is a most underdiagnosed complication of diabetes, more frequently encountered in T2DM. It is a disorder of neurological abnormalities defined by characteristic dyskinesia of chorea-ballism and striatal abnormalities on neuroimaging. Thus, early recognition and treatment are rewarding.

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