

Case Report

Hemichorea - hemiballismus in undiagnosed diabetes mellitus asian female patient associated with non ketotic hyperglycemia: putamenal and caudate nucleus lesions on magnetic resonance imaging

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ABSTRACT

Hemichorea-Hemiballismus (HCHB) with corresponding putamenal T1 hyperintensity on MRI has occasionally been reported in diabetes mellitus (DM) type 2 with non ketotic hyperglycemia, which is believed to be pathogenetically related to HCHB. A 65 year female presented with fever, diarrhea, and involuntary movements on all 4 limbs in known case of diabetes mellitus type 2, hypertension, chronic heart failure and chronic renal failure. We report occurrence of HCHB in a female patient with DM type 2, Hypertension and acute renal failure.

Keywords: Hemichorea-Hemiballismus, Hyperglycemia, MRI

INTRODUCTION

Hemichorea - Hemiballismus involves involuntary and random movements of all 4 limbs and includes proximal as well as distal muscles. It is believed to be associated with increased glucose level, occlusive or hemorrhagic vascular lesion, tumor/plaques in multiple sclerosis. Hyperglycemia, though less common, but important cause of HCHB.¹ Diagnosis can be made clinically and can be confirmed by MRI. T1 weighted magnetic resonance imaging are important to determine lesions on the putamen and caudate nucleus which can detect early changes characterized by hyperintensity. Undiagnosed diabetes mellitus should always be suspected in those patients who presents with HCHB if not diagnosed earlier. Treatment is usually symptomatic and prognosis is usually excellent.²

CASE REPORT

A 65 year old female was presented in the outdoor patient medicine department with complains of fever, diarrhea, involuntary movements in all 4 limbs for 15 days with sudden onset and intensity had aggravated since last 2 days. She was complaining of weakness since last 10 days. There was no prior history of any such involuntary movements, pyramidal tract signs, seizures, altered level of consciousness. Her past medical history includes hypertension managed with amlodipine and chronic renal failure managed with protein restricted diet. There were non contributory past medical and family history.

At admission, vitals were normal (Temperature-38 C, Pulse 80/min, Respiratory rate-16/min and regular, Blood pressure-138/88 mmHg on right brachial artery). On physical examination, reflexes, sensation and pulses were normal and equal on all 4 limbs. Muscle tone (1+) was

decreased on all 4 limbs. Abdominal, respiratory, general examination was grossly normal.

Investigations

Routine blood examination: Hemoglobin - 11.5 g% (normal), total white count of 7200cells/mm³ (normal), s.urea 72mg/dL (elevated), s. creatinine 2.72 (elevated), s.Na+-136 mmol/L(normal),s.K+-4.9 mmol/L(normal), total bilirubin-0.6 mg/dL (normal), direct bilirubin- 0.2 mg/dL, indirect bilirubin- 0.4mg/dL, s. calcium total-8.4 mg/dL (normal), s. phosphorus- 3.8 mg/dL, uric acid-14.3 mg/dL (elevated), ESR- 82 after 1st hour(elevated), s.glucose-175 mg/dL (poorly controlled hyperglycemia), HbA1c- 11.0%(undiagnosed diabetes mellitus).

Chest X-ray-PA view, liver function test, ultrasonography of abdomen, blood and urine culture, ECG were grossly normal.

On MRI plain plus contrast with gadolinium, unilateral T1W hyperintensity seen involving bilateral caudate nucleus and lentiform nucleus without any significant signal changes in rest of the sequences or any mass effect/perifocal edema (Figure 1, 2). Few focal hypointensity on T2 and FLAIR MRI images are seen in bilateral caudate nucleus and lentiform nucleus (Figure 3).

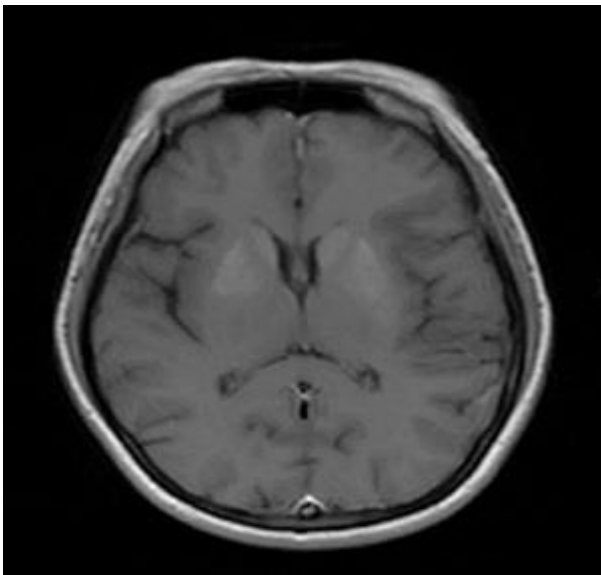


Figure 1: Axial T1 weighted image at the level of basal ganglia shows hyperintensity in bilateral caudate nucleus and bilateral lentiform nuclei.

On CSF examination, proteins-35.3mg/dL (normal), glucose-96 mg/dL (elevated). Physical and microscopic examination was grossly normal. No organism on gram stain and AFB not seen on Ziehl neelsen stain.

On echocardiography, dilated cardiomyopathy with global LV hypokinesia, left ventricular ejection fraction-

30%(chronic heart failure), severe LV systolic dysfunction, mild pulmonary hypertension without any calcification seen.

Treatment

Patient was managed symptomatically. She was given tablet aspirin (81mg)-OD, tablet clopidogrel (75mg)-OD, tablet atorvastatin (40mg)-HS, tablet valproate (200mg)-TDS, tablet metformin (500mg)-BD, tablet famotidine (20mg)-BD. Dietary modification for chronic renal failure, hypertension and diabetes were advised on admission as well as on discharge.

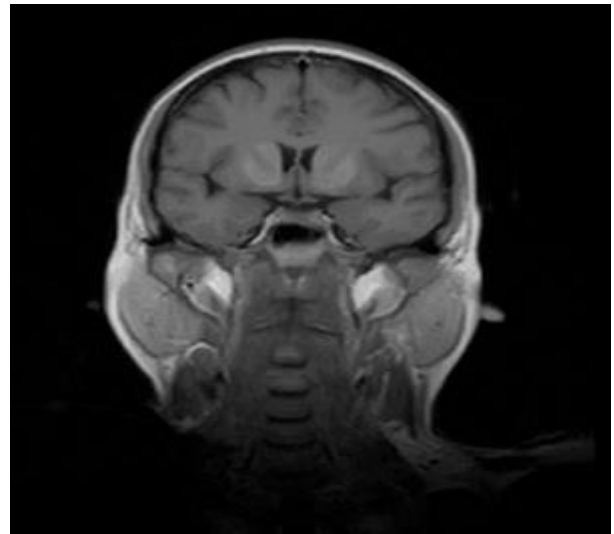


Figure 2: Coronal T1 weighted image at the level of basal ganglia shows hyperintensity in bilateral caudate nucleus and bilateral lentiform nuclei.

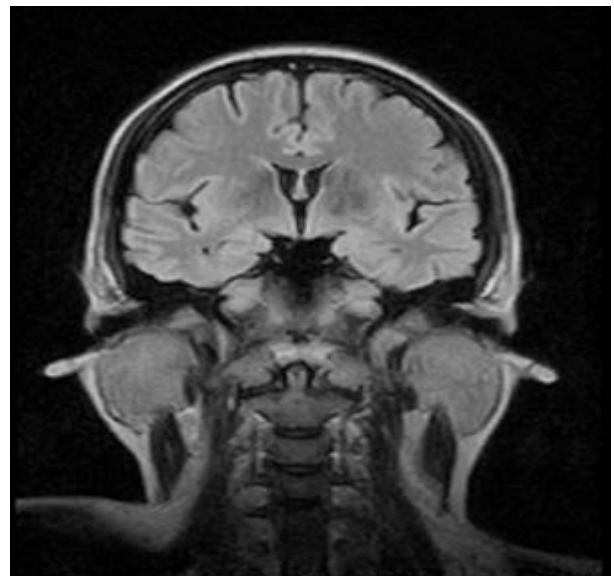


Figure 3: Coronal T2 FLAIR image at the level of basal ganglia shows hypointensity in bilateral caudate nucleus and bilateral lentiform nuclei.

Outcome and follow-up

After 2 months, follow-up of the patient- no present symptoms at that time. s. glucose level-135 mg/dL, s. creatinine-1.62mg/dL, blood pressure 140/82 mmHg. Patient did not notice any involuntary movement anytime. We continued her management for diabetes, hypertension, congestive heart failure and chronic renal failure.

DISCUSSION

HCHB in Asian diabetic women is common. Nonketotic hyperglycemia is one of the unrecognized cause of hemichorea mostly due to lack of awareness and reversibility in euglycemic state.^{3,4} T1 hyperintensity and T2 hypointensity lesions on MRI has been reported sometimes in patient with non ketotic hyperglycemia like in our case.⁵ The pathophysiologic mechanism underlying the imaging finding remain debatable. High signal intensity on T1-weighted images is most likely related to petechial hemorrhage.⁶ Low signal intensity on T1-weighted images is associated with micro calcifications or other metabolic minerals depositions.⁷ Hypointensity on T2 and diffusion weighted imaging(DWI) may be associated with edema.⁸

There is no single mechanism available which can explain the pathophysiology of hyperglycemic chorea.^{4,9} Hyperglycemia causes regional blood flow impairment in the absence of ketoacidosis which leads to attenuation of Kerb's cycle in brain and may increase GABA depletion via the succinct semialdehyde pathway.¹⁰ Disinhibition of dopamine pathways in basal ganglia causes dopaminergic hyperactivity. Older women are inclined because of postmenopausal estrogen deficiency causing dopamine hypersensitivity.^{10,11} Petechial hemorrhage due to hyperglycemia and cerebral ischemia as well as diabetic microangiopathy may contribute to it.¹⁰ Secondary to hyperglycemia, acute putaminal dysfunction may be associated with Wallerian degeneration of the internal white matter of the putamen has been also considered one of the mechanism of nonketotic hemichorea.¹²

Nonketotic HCHB might be the presenting complaint of hyperglycemia as in our case or it can be secondary to poorly controlled diabetes.^{9,13,14} Rapid correction of hyperglycemia can lead to chorea in some case.⁵ The mainstay of treatment is to control of glucose with either partial/complete resolution of HCHB.¹⁵ Clinical and radiological signs may take more time up to 6 months to resolve once we correct the blood glucose level.¹⁵ If it is proved that hyperglycemia was causing hemichorea, most patients have progressively recovered from their hyperkinesia once we control glucose level in their blood.¹⁶

Generally, the prognosis of nonketotic hemichorea has been excellent with some exception.¹⁴ As in our patient, after 2 months of follow up, she was doing fine without

any hyperkinetic activity. In our case, the patients undiagnosed diabetes mellitus might be the reason that she developed non ketotic hyperglycemic HCHB at this age. This was first case in our hospital noted with having HCHB in a patient who has undiagnosed diabetes mellitus.

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