



**ORIGINAL RESEARCH PAPER**

**Radiology**

**NON-KETOTIC HYPERGLYCEMIC HEMICHOREA HEMIBALISMUS SYNDROME**

**KEY WORDS:**

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**CLINICAL HISTORY**

- A 58-year-old woman presented to our emergency department with complaint of left sided weakness , deviation of face since 15 days and headache.
- Relevant medical history included hypertension and insulin dependent Type-2 diabetes mellitus.
- Headaches were intermittent with loss of coordination in the left upper extremity. Review of systems was positive for mild dysmetria.
- Patient was advised Non Enhanced Computed Tomography (NECT) brain and MRI of brain.

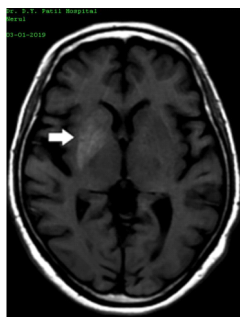
**LABORATORY INVESTIGATION**

- Blood sugar levels were elevated (560 mg/dl).
- Secondary laboratory findings including urine examination were negative for diabetic ketoacidosis.
- Rest of the lab values were within normal limits.

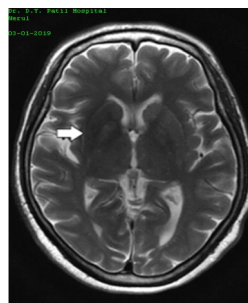
**FINDINGS**



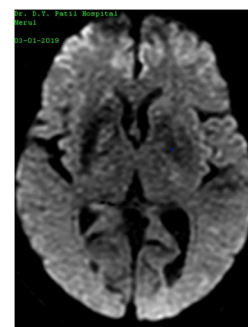
**Axial non contrast CT image of brain demonstrates increased density within the right GCR, involving predominantly the putamen (black arrow) and caudate nucleus (white arrow)**



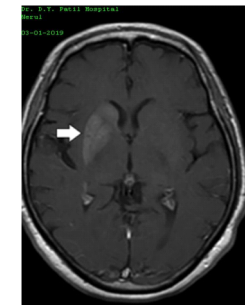
**MRI T1 sequence reveals , increased signal intensity within the right basal ganglia corresponding to the area of hyperdensity seen on CT**



**MRI T2-weighted image demonstrates very faint decreased signal within the right putamen (white arrow) predominantly along the posterior aspect**



**Axial Diffusion-weighted image demonstrates, no evidence of restriction.**



**Post contrast T1-weighted FSE image demonstrates , minimal enhancement within the right basal ganglia (white arro)**

**DIAGNOSIS**

Based on the clinico-radiological and laboratory findings, the diagnosis was: Non-ketotic hyperglycaemic hemichorea (NHH), also known as diabetic striatopathy or chorea, hyperglycemia, basal ganglia (C-H-BG) syndrome

**TREATMENT**

On prompt correction of hyperglycemia, the movement disorder usually resolves within few days- weeks and may not require symptomatic therapy.

Additionally, typical neuroleptic drugs and sometimes benzodiazepines are useful in the management of choreic movements.

A follow-up brain MRI after 6 months usually shows disappearance of the symptoms.

**DIFFERENTIAL DIAGNOSIS**

- Acute ischemic/hemorrhagic stroke
- Chronic hepatic encephalopathy
- Methemoglobin in intracranial hemorrhage
- Calcification
- Manganese toxicity during long-term parenteral nutrition .
- Fahr disease, Wilson disease, and carbon monoxide poisoning

**DISCUSSION**

Hemiballism-hemichorea associated with hyperglycemia was first reported by Bedwell in 1960.

It is a rare cause of T1 bright basal ganglia and one of the neurological complications of non ketotic hyperglycemia.

Estimated prevalence is less than 1 in 1,00,000.

Commonly seen in elderly female patients, typically Asian, with Type 2 diabetes. Female:male ratio is 2: 1 It is a rare but dramatic complication of nonketotic hyperglycemia in patient with uncontrolled diabetes.

Thought the most common cause of hemichorea-hemiballism in adults is vascular lesion in the basal ganglia.

Rarely, it can also be the first clinical manifestation of non-ketotic hyperglycemia.

The etiology for hemichorea-hemiballism occurring in non-ketotic hyperglycemia is poorly understood.

Asian population is particularly more susceptible, which suggests a possible genetic predisposition to the disorder .

**CONCLUSION**

- Clinical features of this condition vary in severity, and overlap considerably with other differential diagnosis.
- Familiarity with the imaging features allows the radiologist to suggest the correct diagnosis and thereby guide its timely management.
- NNH has a good prognosis and can be reversed once hyperglycemia is corrected.

**TAKE HOME MESSAGE**

T1 hyperintensity involving the basal ganglia is a typical feature of hemichorea hemiballismus and should be correlated with the clinical history of the patient to reach the diagnosis

**REFERENCES**

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