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Nonketotic Hyperglycemic Hemichorea in a Patient with Uncontrolled Diabetes: A Case Report

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Running Title: *Nonketotic Hyperglycemic Hemichorea*

Abstract: ***Introduction: Non-Ketotic Hyperglycemic Hemichorea (NKHH) is a rare neurological complication of non-ketotic hyperglycemia. Disease usually presents with hemichorea- hemiballismus syndrome seen in Asian elderly female with type 2 diabetes. Symptoms usually resolve on normalization of glucose levels. Case: A 61 year old female Amratben presented to medical emergency room with complaints of involuntary movement of left upper and lower limbs since 1 month. Routine blood tests were normal including thyroid profile. MRI Brain plain study suggestive of a well defined altered signal intensity area involving right basal ganglia (non-ketotic hyperglycemic hemichorea). Patient was treated with Insulin, Metformin and other supportive drugs with which blood sugar was controlled and movement was decreasing in nature.***

Keywords: Non-Ketotic Hyperglycemic Hemichorea Diabetic striatopathy Hyperglyacemia Caudate nucleus and Putamen

1. Introduction

Nonketotic hyperglycemic hemichorea (NHH), also known as diabetic striatopathy or chorea, hyperglycemia, basal ganglia (C-H-BG) syndrome, is a rare neurological complication of non-ketotic hyperglycemia, which causes hemichorea-hemiballismus syndrome.[1]

Epidemiology
Nonketotic hyperglycemic hemichorea is most frequently seen in asian elderly female patients, who have type 2 diabetes mellitus.

Clinical presentation
Chorea and ballismus develop rapidly and can be either unilateral or bilateral and is seen during episodes of non-ketotic hyperglycemia. Symptoms usually resolve upon normalization of glucose levels.

Pathology
Mechanisms causing NKHH include:

1) Hyperviscosity secondary to hyperglycemia, leading to regional blood-brain barrier disruption and metabolic damage,
2) The augmented sensitivity of dopaminergic receptors in a postmenopausal period,
3) Decreased gamma-aminobutyric acid (GABA) availability in the striatum secondary to the nonketotic state.

Radiographic features

CT
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, hemi choreic movements.

MRI
MRI of the brain is the modality of choice for assessing possible nonketotic 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.

T1: hyperintense,
T2/FLAIR: variable but generally hypointense,
SWI: increased susceptibility,
DWI: high diffusion signal[2].

Overall, the T1 hyperintensity is the most consistent finding of the disease. The increase in T1 hypersensitivity is hypothesized to be due to increased protein hydration within gemistocytes. 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.

Treatment and prognosis
Symptoms and imaging findings usually resolve on normalization of glucose levels. Treatment is usually targeted to control Diabetes via Insulin and Oral Hypoglycemic Drugs.

Prognosis of NKH Chorea is usually excellent with resolution of symptoms only with control of hyperglycemia.

Differential diagnosis
Causes of Increased T1 signal in the basal ganglia on MRI

1) Wilson disease and
2) A striatocapsular infarct.

Causes of striatal hyperdensity on CT includes:

1) Hypertensive hemorrhages

2) Basal ganglia calcifications
3) Tay Sachs disease
4) Tuberous sclerosis

2. Case Report

History

A 61 year old female Amratben presented to the medical emergency room with chief complaints of Involuntary purposeless random irregular movement of left upper and lower limbs since last 1 month. Initially choreic movement involved distal part of the left arm below elbow joint which then progressed to involve the whole left upper limb then progress to involve lower limb. There were no complaints of headache, loss of consciousness, nausea, vomiting, any limb weakness, any behavioral problem.

Patient was a known case of Diabetes mellitus type 2 since 7 years for which the patient was not on any treatment since detected. There was no history of similar illness, trauma, Hypertension, stroke and Ischemic heart disease in the past.

Clinical Examination

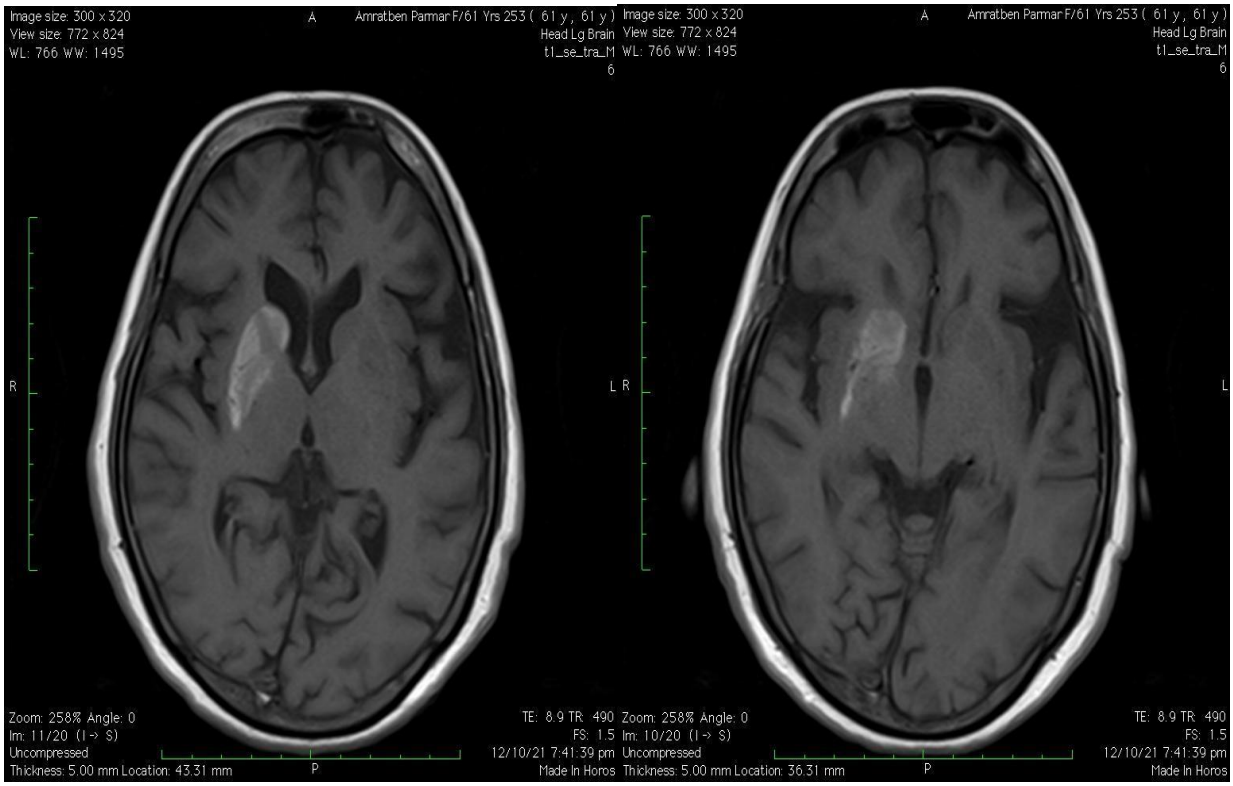
- Vitals were Temperature normal by palpation; pulse 94/min, regular; Blood pressure 150/80 mmHg in Right Brachial artery; sugar 659 mg/dL.
- CNS Examination was Normal Higher function, motor examination revealed normal tone and power (5/5) in all 4 limbs, Planter both side flexor and Deep tendon reflexes absent in right half of body and not elicitable on left half of body. Pupil bilateral reactive to light and Cerebellar examination was normal. Sensory examination was normal.

Laboratory Investigation

In routine examinations, complete blood count, renal and liver function tests were normal. HbA1c 14.6 %, serum acetone absent, TSH was 3.19 IU, urinalysis suggested glycosuria(+++) with no urinary protein or blood loss and urine ketone bodies were negative.

Radiological Imaging

MRI Brain plain study suggestive of a well defined altered signal intensity area (hypointense on T2w, Hyperintense on T1w, blooming on SWI, patchy diffusion restriction on Diffusion weighted images) involving right basal ganglia, possibly of nonketotic hyperglycemic hemichorea.



3. Treatment

Patient was initially treated with insulin with tetrabenazine 50 mg bd, trihexyphenidyl 2 mg tds and other supportive treatment with which the patient was improved and the patient was discharged following which the patient did not come for follow up.

References

[1] Cressman S, Rheinboldt M, Lin D, Blasé J. Nonketotic hyperglycemia-induced hemichorea-hemiballism. Appl Radiol. 2018;47(6):24-26.
[2] Gaillard F.; Foster T. Non-ketotic hyperglycemic hemichorea. Reference article, Radiopaedia.org. (accessed on 08 Nov 2021) https://doi.org/10.53347/rID-21924