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Case Study

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A CASE REPORT OF NON-KETOTIC HYPERGLYCEMIC HEMICHOREA SYNDROME

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ABSTRACT

Non-ketotic hyperglycemic hemichorea(NHH) also known as diabetic striatopathy is a rare condition associated with chorea/ballism. It is the second most common cause of hemichorea-hemiballismus syndrome, other causes being stroke, demyelination and neoplasms. Chorea/ballism is the involuntary irregular non rhythmic movements of limbs which can be unilateral or bilateral. NHH is mostly seen in elderly patients with type 2 diabetes mellitus. On correction of hyperglycemia, there is reversal of clinical symptoms and imaging findings of basal ganglia.

KEYWORDS: Hemichorea, Diabetic, Non-ketotic, T1 hyperintensity.

INTRODUCTION

Non-ketotic hyperglycemic hemichorea is a rare neurological complication of uncontrolled type 2 diabetes mellitus. The main clinical finding is hemichorea/hemiballism which is a hyperkinetic movement disorder characterised by involuntary irregular non rhythmic movements of the limbs which can be unilateral or bilateral. The classic imaging finding is of striatal hyperdensity on CT and hyperintensity on T1 weighted MRI which is contralateral to the body side affected by hemichorea. An early recognition is of utmost importance as symptoms and imaging findings partially or completely resolve upon normalisation of blood glucose levels and avoid unnecessary treatments.

CASE REPORT

A 70 year old male patient presented to the Emergency Department with chief complaints of involuntary movements of left upper and lower limb since last 1 week which was gradually progressive in nature. The patient was diagnosed with type 2 diabetes mellitus two months ago. He was prescribed oral hypoglycemic agents which he took `infrequently. There was no history of loss of consciousness, headaches, slurred speech, fever, seizure or weakness. There was no history of neuroleptic drug intake.

On physical examination, there was mild hypotonia of left upper limb. His blood pressure was 118/80 mm of Hg and pulse rate was 82 beats per minute.

On laboratory investigation, his random blood sugar (370 mg/dl) and HbA1c (14 NGSP) were raised. Urine was positive for sugar but negative for ketone bodies. His total leukocyte count, platelet count and ECG were within normal limits.

The patient underwent an emergency NCCT brain which revealed hyperdensity involving right caudate nucleus and right lentiform nucleus. There were underlying cerebral atrophic changes and mild white matter ischemic changes.

Subsequently, MRI brain was done which showed T1 hyperintensity in right lentiform and right caudate nucleus. Hypointense signal was seen in right lentiform nucleus on T2 and FLAIR sequences. On diffusion weighted images, there was no diffusion restriction. On susceptibility weighted imaging, no susceptibiity artefacts were noted.

Based on clinical findings of type 2 diabetes mellitus with poor glycemic control, hemichorea and classic imaging findings of T1 hyperintensity in caudate and lentiform nucleus contralateral to the side of hemichorea, the final diagnosis of NHH was made.

The patient was started on insulin and haloperidol. Symptoms of hemichorea improved after 4 days and disappeared by 14th day and the patient was discharged.

Follow up NCCT brain of the patient after 3 months was done which shows complete resolution of basal ganglia hyperdensity.

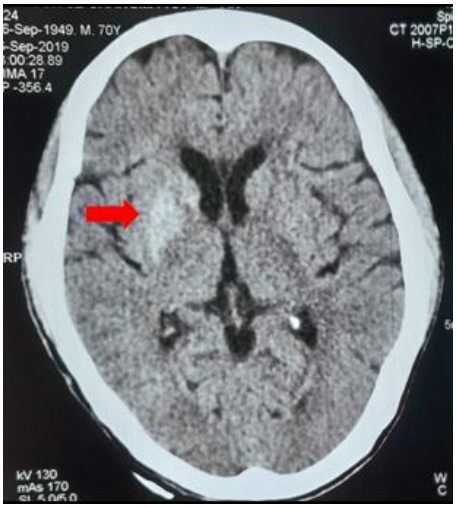

Figure 1: NCCT brain shows hyperdensity in right caudate and right lentiform nucleus.

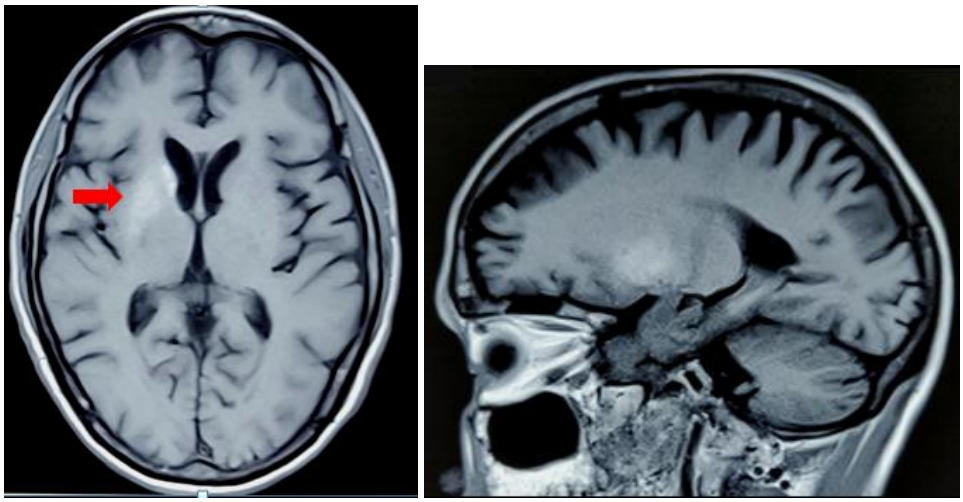

Figure 2(a) T1 weighted axial and 2(b) T1 weighted sagittal MRI brain images show hyperintensity in right caudate and right lentiform nucleus.

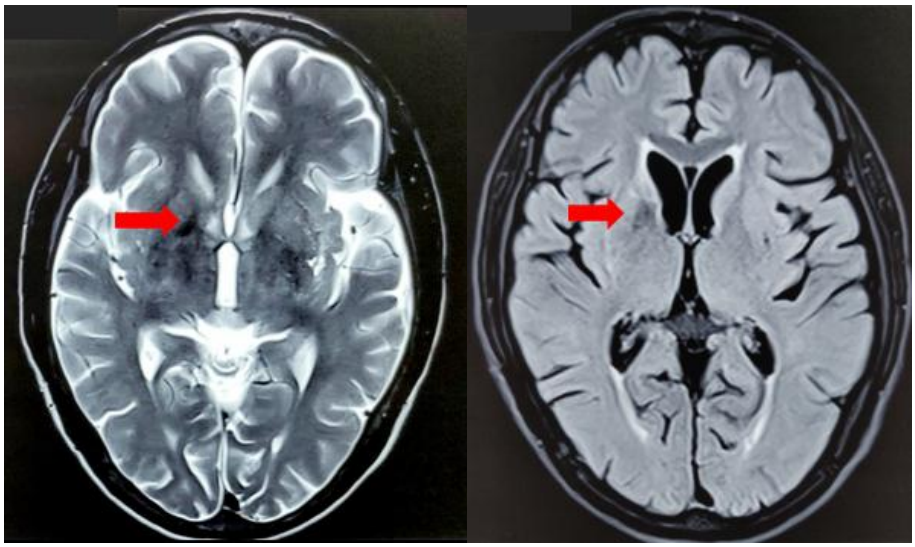

Figure 3 (a) T2 weighted axial and 3(b) FLAIR images show hypointensity in right lentiform nucleus.

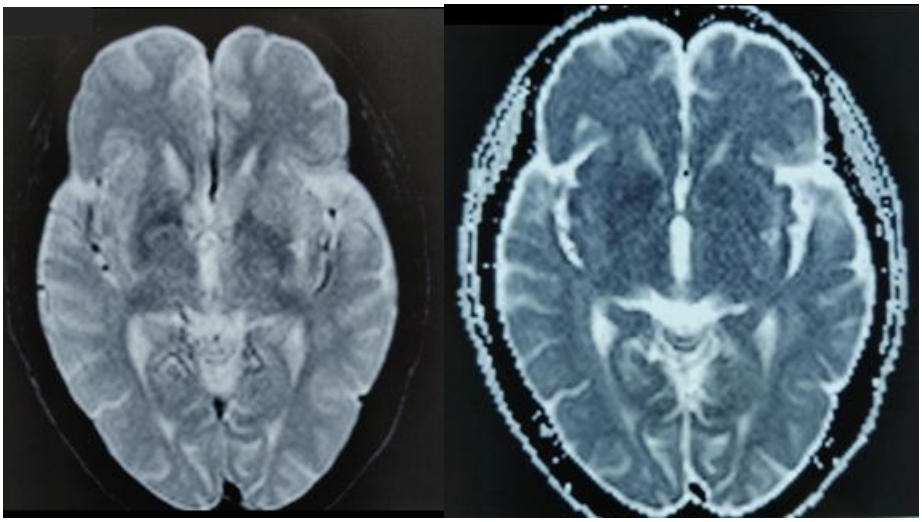

Figure 4(a) Diffusion weighted images and 4(b) ADC maps show no evidence of diffusion restriction.

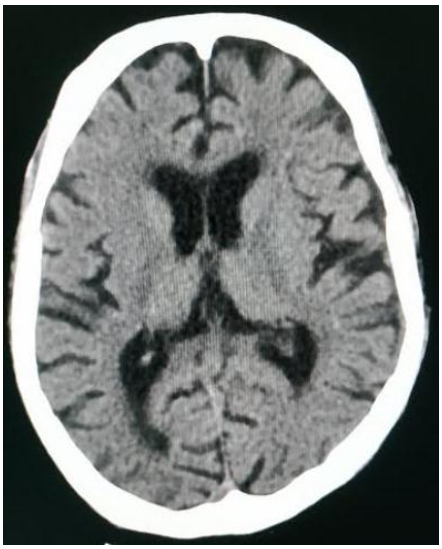

Figure 5: Shows resolution of hyperdensity on follow-up NCCT brain.

DISCUSSION

Hemiballism-hemichorea is a rare hyperkinetic movement disorder characterized by continuous involuntary movements of an entire limb or of multiple limbs on one side of the body. These movements are irregular, non rhythmic, of variable amplitude, and poorly patterned.[1] Ischemic or hemorrhagic stroke represents the most common cause of disease; nonketotic hyperglycemia is the second most common cause.[1-3]

Non-ketotic hyperglycemia affects patients with poorly controlled diabetes mellitus who classically present with the clinical finding of hemibalism-hemichorea.[4] The condition involves mainly Asians, with slight female predominance. This condition is thought to occur most frequently in the elderly, most commonly in the seventh decade.[5]

The pathophysiology of non-ketotic hyperglycemic hemichorea is poorly understood. Some mechanisms include blood hyperviscosity caused by hyperglycemia, leading to blood brain barrier disruption; increased sensitivity of dopaminergic receptors in postmenopausal period, decreased gamma-aminobutyric acid (GABA) in striatum secondary to a nonketotic state.[6]

The most frequently described finding associated with nonketotic hyperglycemia on non-contrast head CT examination is a hyperdense putamen and/or caudate nucleus contralateral to the side of the hemichorea.[4] The MRI findings include hyperintensity in basal ganglia on T1-weighted brain MRI on the side contralateral to patient's symptoms. The putamen and caudate nucleus are most commonly involved. The T2-weighted findings are much more variable with the majority of basal ganglia lesions described as either hypo-intense or iso-intense to background normal basal ganglia.[5]

Differential diagnosis for increased T1 signal within the basal ganglia include early microcalcification, senescent, metabolically derived, or dystrophic from prior deep gray matter anoxic, toxic or hypoglycemic injury, or Fahr's disease. Hyperdensity on CT could also be correspondingly expected dependent upon the extent of

mineralization.[7] Additionally, excessive manganese deposition in conjunction with chronic hepatic encephalopathy or total parenteral nutrition, as well as Wilson's disease, can present with T1 shortening and basal ganglia hyperintensity.[8] A key discriminating factor, however, is the typically bilateral involvement seen with the above listed causes, in contrast to the classically asymmetric and unilateral changes observed in NHH.[9]

The mainstay of NHH treatment is control of hyperglycemia with proper hydration to correct the underlying metabolic imbalance.[10] Imaging findings gradually resolve after hyperglycemia correction. However, they tend to return to baseline more slowly than the clinical findings.[4]

CONCLUSION

Non-ketotic hyperglycemic hemichorea is a rare entity which affects patients with poorly controlled diabetes mellitus and has unique clinical and radiological findings. Although there are many causes of striatal hyperdensity, the classic imaging findings contralateral to the side of hemichorea and resolution of symptoms and imaging findings on normalization of blood glucose is what makes this condition unique.

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