Case report

Nonketotic hyperglycemic hemichorea – a diagnostic challenge for the treating physician

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Abstract

Non-ketotic hyperglycemic hemichorea (NHH) is a rare phenomenon occurring in patients with type II diabetes mellitus. The exact mechanisms for NHH is unclear. There are several proposed mechanisms involving GABA pathways. We report a case of a 63-year-old female who presented with Choreiform movements with characteristic CT changes of NHH, whose abnormal movements rapidly resolved with good glycaemic control.

Key words: chorea, non-ketotic hyperglycemic hemichorea, diabetes, basal ganglia hyperdensity

Introduction

Non-ketotic hyperglycemic hemichorea also known as diabetic striatopathy or chorea-hyperglycaemia basal ganglia (C-H-BG) syndrome is a rare phenomenon occurring in patients with type II diabetes mellitus[1]. It is more common among females, specially of east Asian descent[2]. It is often missed or misdiagnosed as a haemorrhagic stroke completely diverting the management strategy. We report a case of NHH, which was successfully treated.

Case report

A 63-year-old female presented to Teaching Hospital Anuradhapura with uncontrolled rhythmic movements of left side upper and lower limbs of one-week duration. The patient gave a history of type 2 diabetes mellitus for more than 5 years but the control of blood sugar was poor.

Her choreiform limb movements were disturbing her daily activities and causing significant psychological distress. A non-contrast CT scan of the brain revealed an abnormal hyperdensity in the right putamen and globus pallidus (Figure 1).

At presentation her random blood sugar was 348 mg/dl with HbA1C value of 14.2%. Arterial blood gas was normal and urine ketone bodies were negative. Her full blood count, liver function tests, ECG, 2D echo and carotid Doppler were normal. She was started on sub cutaneous insulin and risperidone. She made a complete recovery in one week with complete resolution of CT findings (Figure 2).

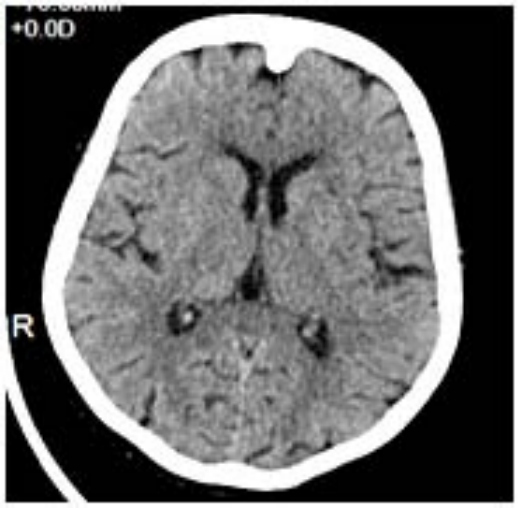

Figure 1. NCCT showing right side basal ganglia hyper density.

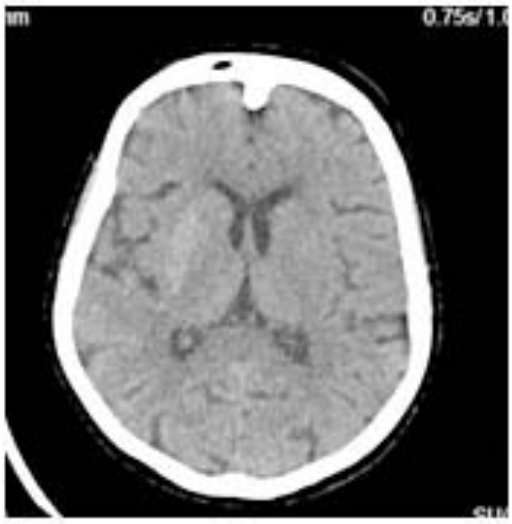

Figure 2. Repeat NCCT brain.

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Discussion

NHH is a rare complication of poorly controlled diabetes[1]. Typical patient with NHH chorea will have the triad: nonketotic hyperglycemia, hemichorea, and basal ganglia increased signal in T2MRI or high density in CT scan[5].

The exact mechanisms for NHH is unclear, however there are several postulated hypotheses. One theory suggests reduced activity of Gamma Ammino Butayric Acid (GABA), the main inhibitory neurotransmitter in the basal ganglia, due to cerebral hypo perfusion along with hyper viscosity due to hyper glycaemia. Direct metabolic effect of increased plasma glucose and augmented response to dopamine due to receptor hypersensitivity is also suggested to play a role[2,3].

NHH has a characteristic appearance in NCCT where basal ganglia show hyper density[4]. The diagnosis of NHH can be missed in clinical setting as the hyper density may not be clearly visible and clinician may attribute the movement disorder to a lacunar infarct in the background of diabetes mellitus[5]. Even when its visible it may be mistaken for a basal ganglia hemorrhage unless you are aware of NHH[3,6]. These diagnostic errors lead to erroneous management of this condition. Proper glycaemic control with normalization of blood sugar is the mainstay of management.

Conclusion

Awareness of the NHH could cut down extensive testing on a typical presentation of this functionally debilitating but treatable condition. The general treatment measures include improving control of blood glucose and the use of neuroleptic drugs if necessary.

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