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Letter to the Editor

Nonketotic, hyperglycaemic hemichorea – a diagnostic challenge for the treating physician

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To the editor,

Non-ketotic hyperglycaemic hemichorea (NHH), 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, especially of east Asian descent [2]. The exact mechanisms underlying NHH are unclear although there are several explanations involving GABA pathways. NHH is often missed or misdiagnosed as a haemorrhagic stroke, completely altering the management strategy. We report a case of a 63-year-old lady who presented with choreiform movements with characteristic non-contrast computed tomography (NCCT) changes of NHH, who was successfully treated with glycaemic control.

A 63-year-old female presented to Teaching Hospital, Anuradhapura with uncontrolled rhythmic movements of the upper and lower limbs on the left side of one week's duration. The patient had a history of type 2 diabetes mellitus for more than 5 years with poor control of blood sugar. The choreiform limb movements were impeding her daily activities and causing significant psychological distress. NCCT scan of the brain revealed abnormal hyperdensity in the right putamen and globus pallidus (Figure 1).

At presentation, her random blood sugar level was 348 mg/dl with an HbA_1C value of 14.2%. Arterial blood gases were 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 a short course of resperidone. She made a complete recovery within one week with complete resolution of CT findings (Figure2).

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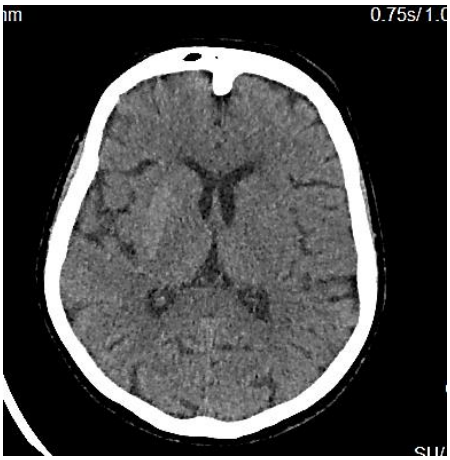

Figure 1: NCCT showing right side basal ganglia hyper density

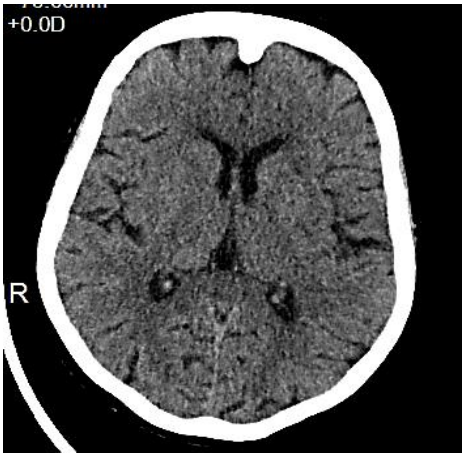

Figure 2: Repeat NCCT brain

NHH is a rare complication of poorly controlled diabetes [1]. The patient with NHH typically shows a triad of non-ketotic hyperglycemia, hemichorea and high signal or high density basal ganglia on MRI T1or CT scan [3].

The exact mechanisms underlying NHH are unclear. However, there are several postulated hypotheses. One theory suggests that it is due to a reduction in gamma ammino butaric acid (GABA), the main inhibitory neurotransmitter in the basal ganglia, due to cerebral hypo perfusion along with hyperviscosity due to hyperglycaemia. A direct metabolic effect of increased plasma glucose and an augmented response to dopamine due to receptor hypersensitivity may also play a role [2, 4]. Glycaemic control with normalization of blood sugar and the use of neuroleptic drugs are the mainstay of management.

NHH has a characteristic appearance on NCCT, where the basal ganglia show hyperdensity [5].The diagnosis of NHH can be missed in the clinical setting as the hyperdensity may not be clearly visible and the clinician may attribute the movement disorder to a lacunar infarct in the background of diabetes mellitus. (5) Even when it is visible it may be mistaken for a basal ganglia haemorrhage [4, 6]. These diagnostic errors lead to erroneous management of this condition.

Increased awareness of NHH could cut down extensive testing and speed up treatment in a patient presenting with the typical clinical picture of this functionally debilitating but treatable condition.

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