

# Hemichorea Associated with Nonketotic Hyperglycemia

Calica Aghata Laksana<sup>1</sup>, Ni Nyoman Ayu Suciari<sup>2</sup>, Anak Agung Istri Kumala Dewi<sup>3</sup>

<sup>1,2</sup>General Practitioner, Wangaya General Hospital, Denpasar, Bali, Indonesia

<sup>3</sup>Internist, Wangaya General Hospital, Denpasar, Bali, Indonesia

**Abstract:** *Hyperosmolarity hyperglycemic non-ketotic (HHNK) syndrome is a clinical syndrome of severe hyperglycemia, hyperosmolarity, and Intracellular dehydration without ketoacidosis. This reports described a 66 year old female with hemichorea associated with hyperglycemia. She had history of type 2 diabetes mellitus and poor glycemic control. She was treated with insulin, trihexyphenidil, and haloperidol.*

**Keywords:** Hemichorea, Nonketotic, Hyperglycemia

## 1. Background

Hyperosmolarity hyperglycemic non-ketotic (HHNK) syndrome is a clinical syndrome of severe hyperglycemia, hyperosmolarity, and Intracellular dehydration without ketoacidosis. Hemichorea associated with non-ketotic hyperglycemia is a syndrome characterized by the sudden occurrence of hemichorea or its more severe expression, hemiballism. It typically affects older adults, especially females. Not much is currently known regarding the pathogenesis of this condition.

Most of these theories revolve around the recognition that hyperglycemia may impair the cerebral autoregulatory mechanism of the central nervous system, causing hypoperfusion and the activation of anaerobic metabolism and depletion of gammaaminobutyric acid (GABA) in basal ganglia neuron. GABA is the main inhibitory neurotransmitter in the basal ganglia and depleted rapidly in non ketotic hyperglycemia causing a reduction in acetylcholine synthesis. Hyperglycemia also induced hyperviscosity and causes a disruption of the blood brain barrier and transient ischaemia of the neurons and leads to hemichorea in these patients.

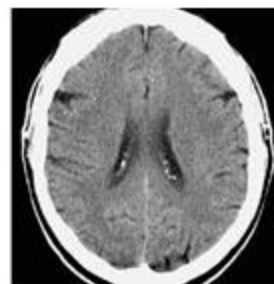
We present the case of elderly woman that presented to emergency unit with acute onset of hemichorea in the setting of poorly controlled diabetes mellitus which was eventually determined to be Non Ketotic Hyperglycemia induced hemichorea.

## 2. Case

An 66-year old woman came to emergency unit with acute involuntary movements about three weeks. As explained by the patient, the involuntary movement initially started in left legs then progressed to involved left arms. These involuntary movement is resolved during sleep. She had history of type 2 diabetes mellitus for about 3 years with no medication and poor glycemic control.

Physical examination findings were significant for left upper and lower extremity hemichorea-like movements. Facial expression was normal. All cranial nerves were assessed and

were normal on examination. Her blood glucose level on admission was 370 mg/dL and urine keton (-). Glycated hemoglobin was 13.9%.



A computed tomography (CT) scan of the head (Fig. 1A) showed no abnormal findings.

She was diagnosed with chorea associated with NKH. The treatments included improving blood glucose with insulin and symptomatic treatment of chorea with trihexyphenidil twice a day and haloperidol 3 times a day. Strict blood glucose control was initiated with insulin glargine 12 units every nights and scheduled dose of 10 units of insulin glulisine before meals. The blood glucose levels were maintained from 370 – 120 mg/dL during hospitalization. The results of the CT scan of the head were normal.

On the third day of admission, patient's involuntary movements were improved. Blood sugars level becomes normalized within few days. By the 6<sup>th</sup> hospital days, she was discharged as the clinical improvement of her condition was better and her blood sugar were normal.

## 3. Discussion

Movement disorders related with hyperglycemia can be in the form of chorea. This happened most often in people between the ages of 50 and 80 years in poorly controlled blood sugar level.

The molecular pathogenesis underlying hyperglycemic hemichorea remains unclear, and 2 major hypotheses have been proposed. First, GABA was used as an alternate energy

substrate during hyperglycemic crisis. GABA depletion leads to thalamic disinhibition and hyperkinesia. Second, vasculopathy was hypothesized to be one of the underlying pathogenesis. Long term hyperglycemia causes hyperviscosity of blood, which leads to latent ischemia of the striatum and subsequent hemichorea. The absence of insulin leads to hyperglycemia and the release of free fatty acids from the adipose tissue, which are metabolized to ketone bodies. It can be used as a GABA substitute to temporarily compensate for the hyperglycemia-induced GABA depletion. Thus, it reflects the rarity of hemichorea in patient with diabetic ketoacidosis.

While cranial CT imaging is normal in some patients, hyperdense lesions in the basal ganglion region which is the known radiological finding of hyperglycemia-related chorea is observed in some other patients.

As observed in the patients reported here, regulation of blood glucose is the most efficient factor in improvement of involuntary movements and abnormal disappear in hours. However, hemichorea may continue for more than three months in 20% of the patients.

Although the findings in these patients are generally reversible, the picture may get worse in conditions accompanied by hyperosmolarity/ hyperviscosity and even mortality may be observed. In this patient, the complaints regressed 4 days after the blood glucose was reduced and the patient was followed up with haloperidol treatment.

#### 4. Conclusion

In conclusions, although hemichorea is rarely caused by a dysfunction of glucose metabolism, we advise not to overlook the chance to check blood glucose whenever patients with this type of hemichorea are encountered. In a patient with history of uncontrolled diabetes, Hemichorea Hyperglycemia Non Ketotic Syndrome should be considered as one of the differential diagnosis that causes Hemichorea/Hemiballism. This is a rare disease that can be reversible with correction of hyperglycemia. Thus, prompt recognition and treatment is essential to avoid adverse outcomes.

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