Hemichorea Secondary to Non-Ketotic Hyperglycemia as the Presenting Manifestation of Diabetes Mellitus

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Abstract
Nonketotic hyperglycemia is an unusual and rare cause of hemichorea. Hemichorea though rare may be the initial manifestation of diabetes mellitus. Correction of the hyperglycemia usually results in total resolution of the signs and symptoms. We present the case of a 71 yr old female, who presented with subacute onset of choreiform movement of left upper and lower extremities over 8 days. Her serum glucose level was 416 mg/dl and urine ketone bodies were absent. Computed tomography of brain showed right caudate nucleus and right lentiform nucleus hyperdensity suggesting hyperglycemia related hemichorea syndrome. Restoration of euglycemia along with treatment with haloperidol and tetrabenazine led to eventual resolution of all symptoms. So, nonketotic hyperglycemia should be kept as a differential diagnosis in a patient with hemichorea.

Introduction
Chorea is characterised by involuntary randomly appearing irregular movements that are not rhythmic or repetitive. Hemichorea is a hyperkinetic disorder involving one side of body, which is caused by lesions of the contralateral striatum. Various conditions such as cerebrovascular insufficiency, neurodegenerative diseases, neoplastic diseases, immunological diseases, infectious diseases, and metabolic diseases are known as secondary causes of this rare disorder. Hyperglycemia is the most common metabolic cause of chorea-ballism. NKH chorea is more common in elderly patients especially females. Hemichorea as the initial presentation of diabetes mellitus is rare. Here we describe hemichorea as the presenting manifestation of diabetes mellitus in an elderly lady.

Case Report
A 71 year female presented with involuntary purposeless nonrepetitive movement of left upper and lower extremities involving both proximal and distal parts for last 8 days. These movements were interfering with her daily activities prompting her to come to our hospital. She did not have abnormal behaviour or altered sensorium or weakness of any side or sensory disturbances. She was not a known hypertensive or diabetic. No past history of similar illness or stroke was elicited.

On examination, she was conscious, alert and oriented. Pulse rate was 84/min regular and blood pressure was 120/80 mm Hg. Cranial nerve examination was normal. Motor examination revealed normal power but hypotonia, hyporeflexia and intermittent non-repetitive involuntary choreiform movements involving left upper and lower extremities. Plantar reflexes were flexor bilaterally. The sensations were intact. Complete blood count, liver and renal function tests were normal. Her serum glucose was 416 mg/dl, HbA1c level was 9.2% and urine ketone bodies were negative. Computed tomography (CT) of brain revealed hyperdensity in right caudate nucleus and right lentiform nucleus (Figure 1).

The patient was started on insulin, haloperidol 2 mg/day and tetrabenazine 50 mg/day. After 5 days, her symptoms improved with marked decrease in involuntary movements. Two weeks later, the abnormal movements disappeared completely and patient was discharged. The patient refused to have a follow up computed tomography (CT) brain.

Discussion
Chorea secondary to hyperglycemia was first reported in 1960. Similar cases have been reported worldwide; most of them are secondary non-ketotic hyperglycemia in type 2 diabetic patients, although rare cases were reported in ketotic hyperglycaemic type1 diabetic patients. The average age of onset is 71 years old, mostly in Asian females, which increases the possibilities of genetic or environmental predisposing factors.

Non-ketotic hyperglycemic chorea might be the first presentation of hyperglycemia, or it might be secondary to poorly controlled diabetes mellitus. Typically in patients with
non-ketotic chorea, CT of the brain may show high density in basal ganglia but at times CT brain may be normal where Magnetic resonance imaging (MRI) brain is useful. Characteristic MRI feature is hyperintensity at contralateral putamen of basal ganglia on T1W images and restricted diffusion in DWI.

Various hypotheses have been proposed to explain the pathophysiology of hyperglycemic chorea, but the exact mechanism is still unknown.3-6 Cerebral vascular insufficiency, petechial haemorrhage,10 hyperviscosity,11 and depletion of both gamma-aminobutyric acid (GABA) and acetylcholine secondary to metabolic changes12 have been suggested as possible mechanisms of non-ketotic hyperglycemic chorea. Acute putaminal dysfunction, secondary to hyperglycemic or hyperosmolar insult, associated with some degree of Wallerian degeneration of the internal white matter of the putamen has been also considered to play a pathogenic role in non-ketotic hyperglycemic chorea.

The prognosis of NKH chorea has been reported to be excellent, with rare exception.8 Prognosis depends on the prompt identification of undiagnosed diabetes or the proper control of the blood sugar in the previously diagnosed patients. The mainstay of treatment is aggressive glycemic control. Haloperidol, tetrabenazine and sometimes benzodiazepines are useful in management of choreic movements.8

**Conclusion**

Hemichorea can be a manifestation and rarely, a presenting sign of diabetes mellitus as in our patient. Non-ketotic hyperglycemia is an unusual but important differential diagnosis in patients with hemichorea as prompt diagnosis and treatment of hyperglycemia has an excellent prognosis.

**References**