

Chorea hyperglycemia basal ganglia syndrome: A case report from Nepal

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Abstract

A rare case of chorea hyperglycemic basal ganglia syndrome in a 56-year-old woman who presented with left-sided hemichorea in the setting of uncontrolled, non-ketotic, type II diabetes mellitus is reported. Early blood glucose control could lead to complete resolution of symptoms. Despite an excellent prognosis, delayed recognition and management can lead to prolong disability due to movement disorder.

Keywords

Chorea, chorea hyperglycemic basal ganglia syndrome, movement disorder, non-ketotic hyperglycemia

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Introduction

Chorea is characterized by excessive, involuntary, irregular, and non-rhythmic semi purposeful movements. It can be caused by vascular, degenerative, genetic, autoimmune, neoplastic, infectious or metabolic pathologies.¹ Diabetes mellitus can have different neurological manifestations, commonest being stroke and peripheral neuropathy. However, in some settings, it can also manifest as chorea hyperglycemic basal ganglia syndrome (CHBG). CHBG syndrome is a rare metabolic cause of chorea seen in the setting of uncontrolled non-ketotic diabetes mellitus. Awareness and prompt recognition of CHBG syndrome is crucial since correction of hyperglycemia can lead to clinical and radiological improvement.² We present a case of CHBG in an elderly female which completely remitted following adequate blood glucose control.¹

Case presentation

A 56-year-old female presented with 1-week history of involuntary, irregular, non-rhythmic, jerky movements of left side of the body. The symptoms started insidiously in her left arm. Then, it gradually progressed to the left leg and left-sided perioral area within a few days. Involvement of distal limbs was more pronounced than proximal, and upper limb was more prominent than lower limb. The movement was continuous, aggravated when she tried to reach for objects

and hold them. She had stumbled upon her foot few times so she needed to walk very cautiously and slowly. The choreatic movements decreased when she was relaxed or with rest, and disappeared with sleep. There was no history of headache, fever, loss of consciousness, weakness, seizure, and vomiting. Her past medical history was significant for type II diabetes mellitus, which was diagnosed 2 months back. She was being treated with lifestyle modification and metformin. However, she had been non-compliant with the therapy. She did not have other co-morbidities or family history of similar illness or history of sore throat in the preceding few weeks to months. There was no history of intake of neuroleptics, anti-depressants, antihistamines, and other medications likely to cause chorea as their side effects.

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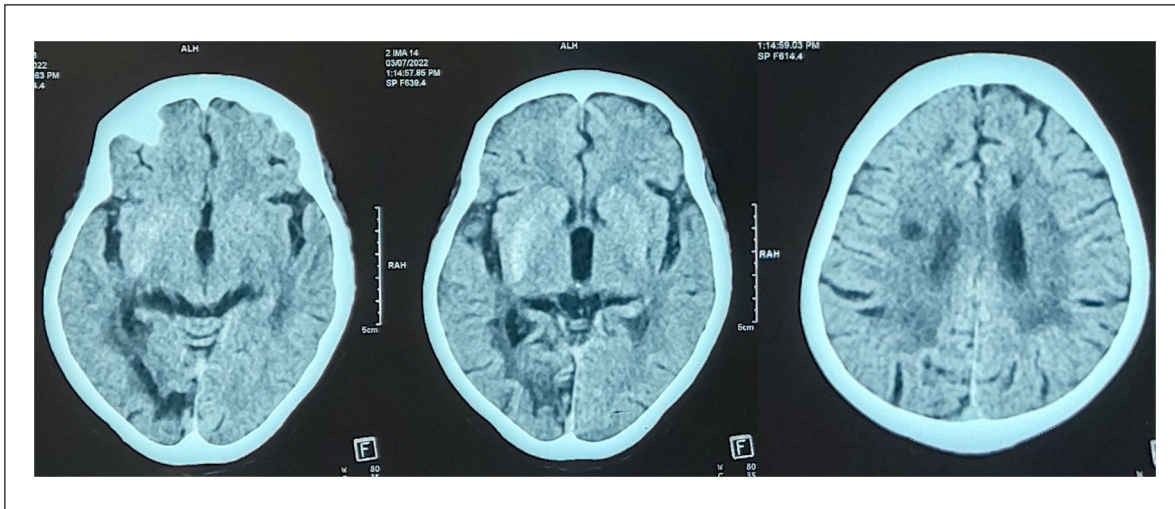


Figure 1. Computerized tomography scan of the head shows areas of prominent high density at the tail of the right putamen and caudate nucleus, and mild hyper density in left putamen. Areas of hypodensity and encephalomalacia were noted in right occipital region and hypodense lesion in bilateral corona radiata.

On examination, the vital signs were stable. The Glasgow Coma Scale was E4V5M6, and pupils were bilaterally equal and reactive to light.³ The fundus examination was normal. Physical examination of all the systems was normal except for the choreiform movements on the left upper and lower extremity. The movement did not cease or reduce with cognitive or motor distraction maneuver. There was no entrainment of the movements. There was no variability in chorea distribution, but frequency and amplitude decreased at rest and increased in response to movement against gravity and carrying objects. Both milkmaid sign and whack-a-mole sign were positive. The examination of higher mental function, cranial nerves, and the sensory system was normal. The power, reflexes, and tone of the right half of the body was normal. However, the tone in left upper and lower limb was reduced and hand grip was impersistent.

The thyroid function test, C-reactive protein, serum electrolytes, and vitamin B12 levels were within normal limits. Tests for anti-nuclear antibody, anti-streptolysin O antibody, and anti-N-methyl-D-aspartate (NMDA) receptor antibody were negative. Her random blood glucose level was 412 mg/dL and HbA1C was 9.9% on the day of her admission. The fasting blood glucose level on the next day was 340 mg/dL while the post prandial glucose level was 426 mg/dL. Chest X-ray and Mantoux tests were normal. Urinary ketones were absent. Peripheral blood smear was normal. Computerized tomography (CT) scan of the head showed areas of high density at the tail of the right lenticular nucleus and right caudate nucleus. Areas of hypodensity were noted in right occipital region with encephalomalacia, possibly, previous asymptomatic ischemic stroke. There were other multiple small hypodense lesions, likely to be silent old lacunar infarcts (Figure 1). Magnetic resonance imaging (MRI) of the brain was refused by the patient due to financial reason.

Based on the history, laboratory findings, and radiologic findings, the diagnosis of CHBG syndrome was made. She was started on basal-bolus regimen of insulin along with tetrabenazine and sodium valproate. The patient's symptoms eventually resolved with control of blood sugar levels. The symptoms became intermittent after 78 h of therapy and completely resolved after 10 days. There has been no further recurrence of choreiform movements during 3 months of follow-up.

Discussion

CHBG syndrome is a rare condition that manifests as hemichorea and/or hemiballism in the setting of uncontrolled diabetes mellitus. It is more commonly seen in elderly females of Asian origin, which is the case in our patient as well.¹ The exact pathogenesis of this condition is unknown. However, there are multiple theories on pathogenesis of CHBG, which involves neurotransmitter imbalance between dopamine and Gamma-aminobutyric acid, ischemic insult, hemorrhagic insult, osmotic injury, and autoimmune insult secondary to anti-glutamate dehydrogenase.²

The differential diagnosis in this case included ischemic or hemorrhagic stroke involving basal ganglia, Sydenham's chorea secondary to rheumatic heart disease, central nervous system (CNS) neoplasms, lupus encephalitis, CHBG syndrome, autoimmune encephalitis, Huntington's disease, chorea secondary to acanthocytosis, hyperthyroidism, vitamin B12 deficiency, and drug toxicity. The reasons for exclusion of each of these diagnoses have been summarized in Table 1.

The clinical features of this condition involve unilateral or bilateral chorea with or without facial involvement, pyramidal tract signs, dystonia, and transient muscle

Table 1. Reasons for exclusion of the differential diagnosis.

Diagnosis	Reason for exclusion
Stroke	Patient had insidious onset and gradual progression of symptoms.
CNS neoplasms	There are no clinical features of elevated intracranial pressure or radiological findings in CT scan.
Sydenham's chorea	There was no history of sore throat in the past. Anti-streptolysin antibodies were not elevated.
Huntington's chorea	There was no family history of similar illness.
Hyperthyroidism	The thyroid function tests were normal.
Autoimmune encephalitis	The higher mental function was unaffected and anti-N-methyl-D-aspartate receptor was absent.
SLE encephalitis	ANA titers were negative.
Chorea secondary to acanthocytosis	The peripheral blood smear was normal.
Vitamin B12 deficiency	The vitamin B12 level was normal.
Drug toxicity	There was no history of intake of drugs other than metformin.

ANA: Antinuclear antibody; CNS: central nervous system; CT: computerized tomography; SLE: Systemic Lupus Erythematosus.

weakness.^{4,6} The symptoms typically resolve during sleep. Elevated glucose levels and HbA1c levels are seen at the presentation.^{4,5} Arm-leg involvement is the most frequent followed by arm-leg-face involvement.⁵ Majority of the patients have a history of uncontrolled type 2 diabetes mellitus and are hyperglycemic but non-ketotic at presentation.⁵ This was the case in our patient as well.

The characteristic findings on a T1 weighted MRI are high signal intensity in the contralateral putamen or caudate nucleus.⁶ This may be due to reactive proliferation of gemistocytes.⁷ CT scan also reveals hyperdensity in contralateral basal ganglia.^{8,9} However, the involvement of basal ganglia can be bilateral as well.⁵ Recent systematic review showed that MRI was more sensitive than CT scan in detecting basal ganglia changes. Although, MRI could not be done in this case due to refusal by the patient, with detailed history, examination, lab tests, and CT imaging, our patient was timely diagnosed and treated for CHBG syndrome. In our patient, head CT showed occipital infarction with encephalomalacia and periventricular multiple lacunar infarctions suggestive of old silent strokes. These lesion sites are very less likely etiology for acute chorea. However, hyperintensity in right putamen and improvement of chorea after control of blood sugar level suggest chorea in our patient.

The management of this condition is blood glucose control. In addition, drugs like haloperidol, tetrabenazine, and valproate can be given as well. A recent meta-analysis found significantly shorter recovery time when glucose control solely was used in comparison to addition of the aforementioned drugs.⁵ However, there was no difference in recurrence rates in between two groups. This difference was hypothesized to be due to less severe disease in the group receiving only glucose control versus the group receiving additional drugs.

The prognosis of this condition is good with majority of cases resolving completely. However, recurrence is seen in 18.2% patients.⁵ As diabetic retinopathy correlates with striatal angiopathy, absence of retinopathy in our patient may be one of the factors for the good prognosis.¹⁰ Small minority of

intractable cases can be treated with surgical procedures like pallidotomy or transcranial magnetic stimulation.^{11,12}

Conclusion

Clinicians should suspect CHBG syndrome if chorea occurs in a setting of uncontrolled diabetes mellitus which cannot be explained by other pathologies. Although, it has an excellent prognosis, delayed management can lead to permanent atrophic changes in caudate and persistent symptoms. Hence, it is necessary for clinicians to be aware of this rare condition.

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Author contributions

RO found the case and helped with initial editing. SKB wrote the initial draft of the manuscript. HK edited the draft and reshaped it into this manuscript. AS obtained the consent. BPG, RK, RR, and NG helped with the final editing and proofreading of the manuscript. All authors approved the final version of the manuscript and agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Consent for publication

Written informed consent was obtained from the patient and her daughter for publication of this case report and any accompanying images.

Declaration of conflicting interests

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Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed consent

Written informed consent was obtained from the patient (bilateral thumb impression) as well as her daughter (signature) for their anonymized information to be published in this article

Study place and time

Tribhuvan University Teaching Hospital, March 2022 to May 2022.

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