CASE REPORT

DOI: 10.4274/globecc.galenos.2022.70299
Glob Emerg Crit Care 2023;2(1):30-32

Managing Hemiballismus Hemichorea Caused by Hyperglycemia in an Emergency Department

Sarper Yılmaz[1], Süleyman Alpar[2]

[1]University of Health Sciences, Kartal Dr. Lütfi Kırdar City Hospital, Department of Emergency Medicine, İstanbul, Turkey
[2]Beykent University Faculty of Medicine, Department of Emergency Medicine, İstanbul, Turkey

Abstract

Chorea, which is a common kind of uncontrollable movement disease, chorea is caused by dysfunctional neural networks that connect the basal ganglia with the frontal cortex. If a patient complains of chorea, a complete medical history is required to rule out other possible causes, including infection with group A beta-hemolytic streptococcus, a history of drug use, as well as the patient's age at start and course (acute or insidious). Static chorea may be caused by structural or chemical injury to the basal ganglia or by benign hereditary chorea, both of which are associated with neurodegenerative illnesses. It is also necessary to conduct a neurological examination that includes an evaluation of the distribution and features of chorea. Non-ketotic hyperglycemia is a common cause of acute chorea. In addition to hemichorea, hyperglycemia may also produce chorea that is widespread. Patients with hyperglycemia that fit the normal description have been shown to have an unusual symptom of uncontrolled high hyperglycemia: hyperglycemia-induced hemiballismus hemichorea (HIHH). Even if all patients present with abnormal, hyperglycemic non-ketotic chorea, the differential diagnosis should be considered. Dopamine blockers, tetrabenazine, and topiramate are used to treat HIHH patients who do not respond to hyperglycemia treatment.

Keywords: Hemiballismus hemichorea, movement disorders, hyperglycemic states

Introduction

Chorea is a prominent involuntary movement disorder that is caused by malfunctioning neural networks linking the basal ganglia and frontal cortical motor regions. The condition is defined by a continuous stream of random, short, involuntary muscular contractions that may be caused by a number of factors. Chorea is produced by the caudate nucleus, putamen, subthalamic nucleus, and thalamus, as well as their interconnected connections [1]. According to Piccolo et al. [2] the most common etiologies of sporadic chorea admitted to different neurology departments during a five-year period were vascular (40%), drug-induced (14%), Huntington's disease (10%), and acquired immunodeficiency syndrome (10%), with the diagnosis remaining uncertain in 6%. Hereditary choreas often improve gradually and are symmetrical; but acquired choreas are more likely to be acute or subacute in nature and might be asymmetrical or unilateral.

Chorea often affects the distal limbs and face, but it may also impair breathing and phonation, resulting in slurred speech or involuntary vocalizations. Patients may conceal chorea by blending it into voluntary motions (parakinesia). Patients are often oblivious of their strange motions, which family members can often see. Chorea is often present during rest and may be exacerbated by distracting motions, although it vanishes during sleep [3].

Neurologic examination is also critical and must include an assessment of chorea distribution and related characteristics. Acute chorea is often attributed to non-ketotic hyperglycemia. Hyperglycemia may also cause hemichorea or widespread chorea [4]. On T1-weighted images of afflicted patients, an

Address for Correspondence: Assoc. Prof. Sarper Yılmaz MD, University of Health Sciences, Kartal Dr. Lütfi Kırdar City Hospital, Department of Emergency Medicine; Health Management in Emergencies and Disasters, Yeditepe University, İstanbul, Turkey
Phone: +90 507 280 11 64 **E-mail:** sarperyilmaz08@gmail.com **ORCID-ID:** orcid.org/0000-0001-8166-659X
Received: 25.05.2022 **Accepted:** 01.08.2022

magnetic resonance imaging often reveals a high signal intensity in the contralateral striatum. Once glycemic control is attained, chorea should gradually resolve itself, although aberrant movements may continue for more than a year.

The doctor should make every effort to rule out a secondary etiology during therapy. A simple laboratory workup may aid in elucidating the triggering event. Secondary chorea often improves or resolves when the primary cause is appropriately treated or the offending drug is withdrawn. Regrettably, no medication has been shown to significantly reduce or stop the course of inherited choreas, with the exception of copper-reducing therapy in Wilson disease.

Hemichorea/hemiballism associated with non-ketotic hyperglycemia is a well-recognized syndrome in the emergency department (ED), but few case series have been reported in the literature.

Case Report

A 58-year-old woman was taken to an ED a week ago with a left upper-limb movement that she could not control. Her heart failure was complicated by a history of high blood pressure, coronary artery disease, and type 2 diabetes mellitus (DM). While awake, the subject participated in these activities, which ceased after the individual fell asleep. The five-year anniversary of her diagnosis with type 2 DM had not helped her regulate her blood sugar levels.

When the patient was hospitalized, the following were her vital signs and blood glucose levels: body temperature: 37.3 degrees Celsius; pulse rate: 90 beats per minute; breathing rate: 15 beats per minute; and heart rate: 140/72 mmHg.

According to the findings of a neurological evaluation, the patient had normal verbal capacity and showed involuntary atetoid motions of her left upper limb (Figure 1).

It is normal for her to have hypotonia and weak muscles at her age. A basic medical assessment found nothing further concerning. A random blood sugar reading of 515 mg/dL was likewise within the normal range, and the results of inflammatory markers, thyroid function tests, and an electrocardiogram were all normal. Blood and urine tests showed no signs of acidity or ketones. An abnormally dense putamen was seen on computarized tomography images of the brain without contrast (Figure 2).

Ballistic or choreiform movements in the setting of high blood sugar and the absence of ketoacidosis are typical clinical and radiological signs that aid to establish the diagnosis. Low doses of insulin and olanzapine were administered to the patient. Three days later, glycemic control was attained. After two days, the chorea had disappeared.

Discussion

To characterize hyperglycemia-induced hemiballismus hemichorea (HIHH), as HIHH was initially described by Bedwell, groundbreaking in 1960. Many women in their fifties to eighties (mean age seventy-one years) report having

Figure 1. Involuntary atetoid motions of left upper limb

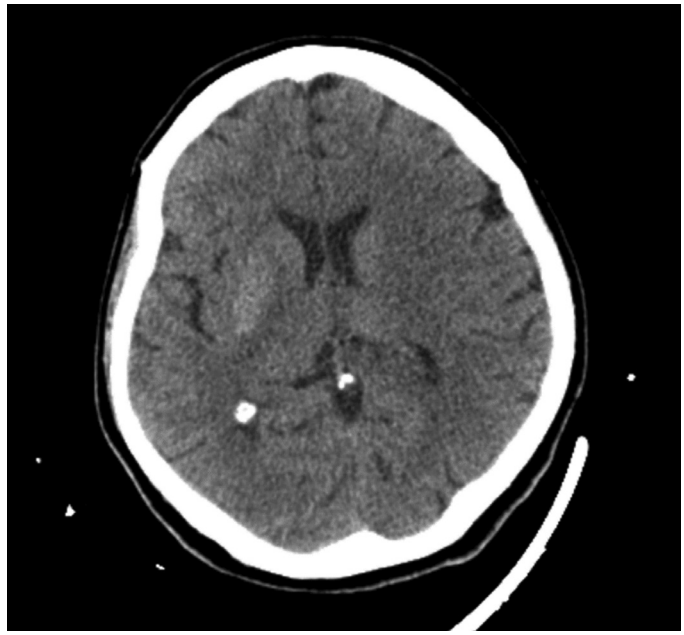

Figure 2. Non-contrast computed tomography demonstrates putaminal hyperdensity on the right hemisphere

choreic and ballistic motions on one side of their bodies. These women do not have a history of DM, but the symptoms arise over a few hours and resolve after 24-48 hours of achieving normal blood sugar levels. The vulnerability of Asian women is unexplained [4]. Many popular drugs, including Hormone Replacement therapy, L-dopa, and others, should be checked out as potential causes. In individuals under the age of fifty, Wilson's disease or Huntington's chorea may be genetic. Human immunodeficiency virus (HIV), hyperthyroidism, the antiphospholipid syndrome, and carbon monoxide poisoning are all possible causes of post-streptococcal Sydenham's chorea.

The pathophysiology of HIHH is shrouded in mystery. The following are a number of the options that have been discussed [5].

1- Blood-brain-barrier disruption may lead to intracellular acidosis and localized metabolic dysfunction.

2- There have been reported instances of petechial hemorrhages, while others have been associated with striatal infarction.

3- It is possible that postmenopausal women's dopamine receptors are more sensitive, which may contribute to hyperkinesis.

4- Non-ketotic individuals lack acetoacetate, a component required for the conversion of gamma-aminobutyric acid (GABA) into dopamine.

This type of hemiballismus is characterized by irregular, involuntary, and unbalanced movement patterns. GABA ergic neurons in the contralateral striatum may be affected by hyperglycemia and ischemia excitotoxicity. Overactivation of the subthalamic nuclei resulting in an increase in excitatory cortical output is a consequence [5]. The disorder is accompanied by hyperintensities in the striatum and globus pallidus, as well as apparent diffusion coefficient mapping without contrast enhancement. Striatal infarction is a rather uncommon occurrence. N-acetylaspartate: creatine ratio and N-acetylaspartate: cholinate ratio may be related with a higher lactate peak [4].

A great majority of these abnormalities vanish after treating hyperglycemia. Anti-nuclear antibody, anti-streptolysin antibodies, and HIV enzyme-linked immunosorbent assays may be required in the correct environment, along with thyroid and liver function testing. Dopamine-blocking medicines, tetrabenazine, or topiramate have been utilized in circumstances when hyperglycemia therapy is inadequate. Dopamine receptor blockers have traditionally been regarded as being the most effective medication for alleviating the intensity of choreic movements, regardless of their etiology. Although first-generation antipsychotic drugs (typical neuroleptics) have a long history of being used to treat chorea, there is less evidence to support their effectiveness, and they are increasingly avoided owing to their increased risk of adverse effects. Second-generation anti-psychotic medications (atypical neuroleptics), such as olanzapine, risperidone, and aripiprazole, have been shown to help alleviate chorea and may have a more favorable side effect profile.

Conclusion

This type of hyperglycemia may show up as an indicator of uncontrolled and severe hyperglycemia: HIHH. Any patient with abnormal glucose levels should have hyperglycemic non-ketotic chorea included in the differential diagnosis, as indicated in this case report. Non-ketotic hyperglycemia, although rare, is an essential differential diagnosis for individuals with hemichorea-hemiballismus because of its positive prognosis when detected and treated early.

Ethics

Informed Consent: Informed consent was obtained from the patients.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: S.Y., S.A., Concept: S.Y., S.A., Design: S.Y., S.A., Analysis or Interpretation: S.Y., S.A., Literature Search: S.Y., Writing: S.Y.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

References

1. Lee MS, Marsden CD. Movement disorders following lesions of the thalamus or subthalamic region. Mov Disord. 1994;9:493-507.
2. Piccolo I, Defanti CA, Soliveri P, Volontè MA, Cislaghi G, Girotti F. Cause and course in a series of patients with sporadic chorea. J Neurol. 2003;250:429-35.
3. Phillips W, Shannon KM, Barker RA. The current clinical management of Huntington's disease. Mov Disord. 2008;23:1491-504.
4. Chang CV, Felicio AC, Godeiro Cde O Jr, Matsubara LS, Duarte DR, Ferraz HB, et al. Chorea-ballism as a manifestation of decompensated type 2 diabetes mellitus. Am J Med Sci. 2007;333:175-7.
5. Lin JJ, Chang MK. Hemiballism-hemichorea and non-ketotic hyperglycaemia. J Neurol Neurosurg Psychiatry. 1994;57:748-50.