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A Case of Non Ketotic Hyperglycemic-Hemichorea Hemiballismus Syndrome

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Abstract: ***Diabetic striatopathy is a rare cause of involuntary movements associated with poorly controlled diabetes. A 50-year-old woman with longstanding type 2 diabetes presented with spontaneous, painful movements that began in the right hand and progressed to the entire right upper limb over three days. Her blood glucose was 401 mg/dL, and glycated hemoglobin was 16.3%, reflecting poor glycemic control. Brain imaging revealed abnormalities in the left putamen and globus pallidus, opposite the affected limb. The clinical presentation and imaging findings were considered consistent with nonketotic hyperglycemic hemichorea-hemiballismus syndrome. Treatment included insulin adjustment and risperidone for movement control. The frequency and intensity of involuntary movements decreased before discharge on the fourth hospital day. At one-month follow-up, symptoms persisted but continued to improve. This case highlights the importance of considering diabetic striatopathy in patients with poorly controlled diabetes who develop acute involuntary movements on one side of the body. Early recognition and appropriate treatment may support motor recovery***

Keywords: Diabetic striatopathy, Nonketotic hyperglycemia, Hemichorea-hemiballismus, Type 2 diabetes, Involuntary movements

1. Case Report

A 50-year-old female patient with a past medical history of poorly controlled type 2 diabetes mellitus presented to the ER with uncontrolled movements of the right upper extremity that started spontaneously three days prior to presentation. These movements started with right hand and progressed to the entire right upper extremity. She had associated right upper extremity pain. she had no such prior episodes of these spontaneous movements, also no recent history of viral illness or trauma. She had no other symptoms and her vitals were within normal limits, on physical examination, the patient was noted to have writhing movement of the entire right upper extremity. The movement could be voluntarily stopped with muscle contraction, but this was causing severe pain from the shoulder radiating to the hand. Light touch Sensation in the bilateral lower extremities from the feet to the knees. Power was in the extremities. No cranial nerve deficits and no gait abnormalities. The patient's initial laboratory testing was serum glucose of 401mg/dL, blood urea nitrogen of 30 mg/dL, and creatinine of 1.52 mg/dL. Due to persistent neuromuscular symptoms, a non-contrast head computed tomography (CT) and head magnetic resonance imaging (MRI). The CT showed a subtle hypodensity in the left putamen and globus pallidus T2-weighted fluid-attenuated inversion recovery (FLAIR) MRI showed hypointensity of the lentiform nucleus on the left side

Investigations
Random mean blood sugar, HbA1c, CT imaging

2. Discussion

Nonketotic hyperglycemic hemichorea- hemiballism , also referred to as diabetic striatopathy, is a well described cause of acute acquired hemichorea in adults. Most cases occur in the setting of poorly controlled, longstanding type 2 diabetes (mean blood glucose 414mg/dl, mean glycated hemoglobin 13.1 percent), although this is also seen in newly diagnosed diabetes or acute hyperglycemia. Older age and female sex are other risk factors. Most patients present with acute hemichorea involving both arm and leg; The characteristic finding on MRI is hyperintense signa. Lesions typically occur in the globus pallidus and putamen, These lesions generally correspond with ballistic and choreiform movements on the contralateral side of the observed imaging findings. The differential diagnosis for non ketotic hyperglycemia hemichorea hemiballismus syndrome may include neoplastic disorders of the brain, Huntington's disease, stroke, trauma, or neurotoxic substance use. The hypothesized mechanism involves a shift from aerobic to anaerobic metabolism during the pathogenesis of non-ketotic hyperglycemia, leading to decreased levels of GABA due to consumption, and subsequent disinhibition of the thalamus and hyperactivation of dopaminergic signaling, causing uncoordinated movements. This is in comparison to diabetic ketoacidosis, where GABA substrates are synthesized, leading to regional metabolic imbalance and basal ganglia dysfunction. Poor glycemic control can lead to vascular complications beyond peripheral vascular disease, increase the risk of cerebrovascular ischemia, and lead to decreased cerebral blood flow.

Hospital Course
The patient's symptoms and imaging findings were consistent with non-ketotic hyperglycemia hemichorea hemiballismus syndrome. The patient was initially treated with 15 units of regular insulin and 6 units of Lispro in casualty. During the hospital course, the patient's insulin regimen was adjusted, and symptoms were also treated with risperidone. Serum glucose remained elevated, ranging from 200 mg/dL to 400 mg/dL. hemoglobin A1C (HbA1C) was 16.3%. The patient was discharged on day4 on a proper insulin regimen and choreiform symptom management with risperidone. At the time of discharge, her uncontrolled arm movement had decreased in frequency and intensity. Patient came for follow up one month later noted that while her symptoms were still present, they were continuing to decrease in severity and frequency.

3. Conclusion

NHHH syndrome also called diabetic striatopathy, a rare cause of hemichorea in adults is generally seen in the setting of poorly controlled long- standing type 2 DM which generally resolves after glycemic control. Early recognition and treatment are important to give the greatest chance of motor recovery. Insulin therapy and other neuroleptic agents to support glycemic control and dopaminergic signaling are the mainstays of care.

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