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Striatal Hyperintensity in Non-Ketotic Hyperglycemia Induced Hemichorea-Hemiballismus Syndrome: Case Report

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ABSTRACT

Introduction: Hemichorea-Hemiballismus (HC-HB) is usually associated with lesions in the contralateral basal ganglia of the central nervous system and can be caused by infection, neurodegeneration, oncological cases, stroke, and metabolic disorders. HC-HB disorders associated with non-ketotic hyperglycemia are rare and are characterized by involuntary and arrhythmic unilateral movements. This occurrence is more common in cases of women, the elderly and poor control of blood sugar levels.

Case Report: A female patient 46 years old, came with complaints of uncontrolled movements in the right half of the body since July 2021. Then it spread to the legs also 1 month later. Prior to this complaint, it was triggered by a stressor. History of non-routine use of drugs. On general physical examination and vital signs within normal limits. Neurological examination found right hemichorea with arrhythmic characteristics, 1-5 Hz, medium-large amplitude, stereotypy, right arm and leg, suppressibility (+), worsens when stressed. A CT-Scan of the head without contrast showed no abnormalities, but an MRI revealed a non-ketotic hyperglycemic image. The patient then received haloperidol 0.75 mg orally every 12 hours and clobazam 5 mg every 12 hours orally and insulin therapy.

Discussion: In this case report, it was found that female patients was poor in control her blood glucose and causing complications. This condition also known as diabetic striatopathy (DS). Several mechanisms to explain the pathogenesis of DS include metabolic disorders, ischemic injury resulting in ion deposition, hemorrhagic injury, dopaminergic and estrogenic imbalances, autoimmune inflammatory responses, and neurodegeneration. This is supported by MRI imaging that supports the presence of hyperintense on T1.

Keywords: Hyperglycemia, Involuntary Movement, Striatopathy

INTRODUCTION

Diabetes mellitus is a disease that is often encountered every day. Even though diabetes diagnosis and routine therapy are familiar to medical personnel, symptoms sometimes appear with unusual clinical manifestations, including Hemichorea-Hemiballism (HC-HB) Syndrome.[1,2]

Hemichorea-Hemiballism is a disorder of hyperkinetic movements that are continuous and not rhythmic in the proximal or distal parts of the body. Various causes of HC-HB include cerebrovascular disorders, inflammation, infections, metabolic or toxin disorders, neurodegenerative disorders, and neoplasms. Autoimmune diseases and infections are often the cause of hemichorea-hemiballismus in young people. At the same time, cerebrovascular disorders with a background of hypertension and diabetes mellitus often occur in old age.[3]

Hemichorea-Hemiballism is a rare condition. Chorea is an abnormal involuntary movement characterized by

short, sudden, and irregular movements. In more severe cases, ballism can occur, namely sudden and strong movements of the extremities, neck, and trunk, affecting one side of the body or involving one extremity (monoballism). A lesion in one of the cerebral hemispheres causes movement disorders on the contralateral side. Hyperkinetic movements involve the face, arms, and legs alike. The subthalamic nucleus connects with the rubber nucleus, substantia nigra, and globus pallidus of the basal ganglia. Its function is not clearly understood, but lesions in the subthalamic nucleus may produce more severe dyskinesia such as HC-HB.[2]

Non-ketotic hyperglycemia in patients with diabetes mellitus is often the cause of the clinical HC-HB syndrome, which occurs with a typical radiological picture.[2] In the emergency unit, these conditions are often undiagnosed and generally treated early as a stroke. Radiology finding in these cases, plays an important role in determining the proper diagnosis and initial management.[4]

Hemichorea associated with non-ketotic hyperglycemia is a rare complication of diabetes mellitus, frequently seen in elderly diabetic women with poor blood glucose control. This situation can also be called Diabetic Striatopathy (DS).[5] Based on a meta-analysis study, the median age was 71 years, with a male-female ratio of 1-1.8. Its prevalence is very rare, with 1 in 100,000 cases worldwide, but the possibility of misdiagnosis and inconsistency in diagnosis can also cause this. In general, dance movements usually occur on the ipsilateral upper and lower limbs, rarely on both sides of the body, which are characterized by fast, non-static, and irregular limb movements, partially involving the muscles of the face and neck, and accompanied by extrusion eyebrows, mouth skimming, tongue extension and other symptoms. Chorea becomes evident when the person is in an emotional mood and may disappear during sleep.[6]

CASE REPORT

A 46-year-old female came to the polyclinic with main complaints of uncontrolled movements in the right half of the body. The patient initially experienced complaints of movements such as dancing in the right arm in July 2021. After one month, the movements spread to the legs as well. Movement of the proximal arms and legs, such as dancing, can not be controlled. The patient is difficult to move. The movement complaints disappear when the patient sleeps. Complaints get worse if the patient is stressed. One week before, the patient said he had received quite a lot of stress because his husband hit a small child and the patient's older brother died. He denied other complaints, such as headaches, pursed lips, and slurred speech. Before the complaint appeared in July, Past medical history, patient with type II diabetes mellitus for 15 years and hypertension. The patient's blood sugar was found to be high when hospitalized in November, at 412 mg/dl, and fluctuated every day until it reached 600 mg/dl. The blood pressure still fluctuated up and down, around 130-170 systolic. Patient used insulin but not routinely, because the patient had to be assisted by her husband to inject the drug. Similar complaints in the patient's family were denied. The patient is a housewife and denies a history of smoking and drinking alcohol.

At the physical examination during hospitalized, the patient was compost mentis with vital signs and general physical examination within normal limits. Neurological examination found hemichorea dextra with characteristics of involuntary arrhythmic movements, frequency 1-3 hz, medium-large amplitude, stereotyped, distributed on the right hand and foot, cannot be suppressed, gets worse when stressed, disappears during sleep. Head CT-Scan without contrast showed no abnormalities (Figure 1). Head MRI showed hyperintensity on T1-weighted MR images (Figure 2). The patient then received haloperidol therapy of 0.5 mg every 12 hours orally and routine blood glucose

therapy (Insulin 10 units at each meal, 22 units at night). After her blood glucose is within normal limit, her complaints have been improved.

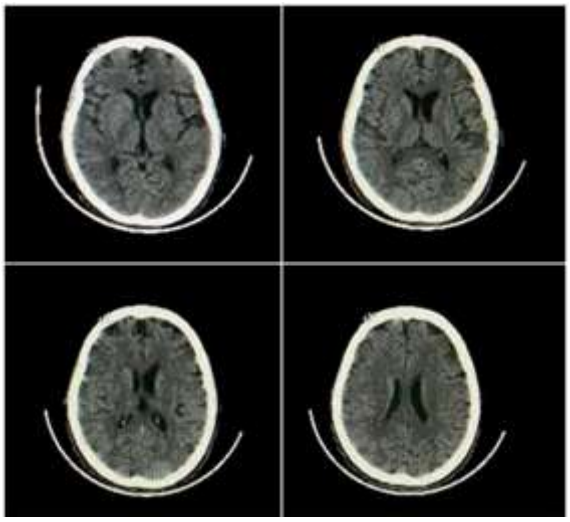

Figure 1. Head CT-Scan without contrast

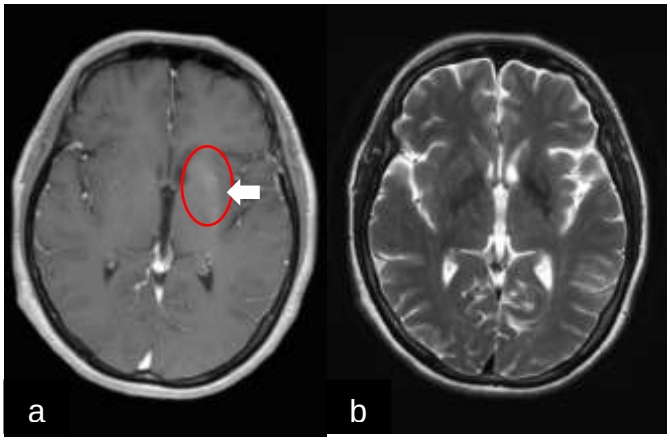

Figure 2. Head MRI without Contras (a. Sequence T1 hyperintensity in the left striatal; b. Sequence T2)

DISCUSSION

In this case, a patient with a history of DM for 15 years with uncontrolled blood glucose where the patient's HbA1c was 12.9%. Patient with main complaint uncontrolled movements in the right half of the body. Currently, the specific mechanism of HC-HB associated hyper glycemia remains unclear. Several theories emerged, including cerebral vascular insufficiency, neuronal dysfunction due to hyperosmolarity caused by hyper glycemia, increased gemistocytes (swollen, reactive astrocytes), petechial bleeding, hyper viscosity, and decreased levels of Gamma-Aminobutyric Acid (GABA) and acetylcholine due to metabolic processes that lead to basal ganglia dysfunction.[7,8] In an autopsy report that was performed, it appears that there is a hyperintensity lesion on the putamen caused by the large number of gemistocytes along the axon, which is thought to have lasted for years. Other studies have also concluded a decrease in blood flow in the putamen due to a decrease in GABA production, which triggers excess dopamine activity. Another theory that emerged recently is the mechanism of petechial hemorrhage due to damage to the blood-brain barrier induced by hyper glycemia, causing transient ischemia in striatal neurons.[8,9]

Clinical manifestations of HC-HB in patients with DS mostly involve the

extremities unilaterally because only the contralateral basal ganglia experience mineral deposition, which causes subthalamic disinhibition. However, it is still unknown why only one side of the basal ganglia is experiencing this deposition. Usually, the initial symptoms experienced by patients vary, but most often, they start from the upper extremities first and then progress to the lower extremities.[10] The mechanism that most likely causes the neurological deficit is hyper glycemia which causes hyperosmolarity and decreased GABA levels, which results in focal ischemia. A study reported that in non-ketotic hyper glycemia conditions, the Krebs cycle was inhibited, and GABA metabolism increased until the level decreased, thereby reducing the threshold for seizure activity. Another hypothesis states a decrease in the seizure threshold due to metabolic disturbances. Hyperosmolarity and dehydration induced by hyper glycemia precipitate focal seizures, producing neurologic deficits in some patients. Some research has also suggested that preformed local ischemic lesions may result in seizures in conditions of metabolic changes due to hyper glycemia. Thus, preformed focal ischemia becomes irreversible due to decreased blood flow to the cerebral area. In addition, most patients with chorea worsen during nervousness and disappear after sleep. The clinical manifestations are similar to those in case-reported patients.[9,11]

Imaging examinations used to detect DS are generally head CT scans and MRIs. The typical neuroimaging features of the contralateral striatum are hyper density on CT scans and hyperintensity on T1-weighted MR images.[12] Distinguishes this imaging from bleeding, namely that there is no mass effect picture and the internal capsule area is not involved. Until now, there have been four hypotheses to explain the pathogenesis of the anomaly in the basal ganglia. The radiographic findings include petechial hemorrhages, mineral deposition (i.e., calcium or magnesium), destruction of myelin, and infarction with astrocytosis. Ischemic injury to the striatal area leads to astrocyte proliferation and the expression of "zinc-friendly" metalloproteins.[13] However, there is still much discussion regarding the mechanism of changes in radiological appearance. At first, it was suspected that the radiological changes were caused by a calcification process. However, these radiological changes may originate from capillary bleeding. Pathological findings based on biopsy examination of brain tissue found the presence of gemistocytes in large numbers, which are suspected to be hyperdense lesions caused by decreased hydration in the cytoplasm of swollen gemistocytes.[2]

The main management of DS is controlling hyper glycemia with proper hydration to correct metabolic disturbances. Although active blood glucose control can successfully overcome hemichorea, the majority of cases require additional anti-chorea drugs to control symptoms.[2] Studies show high levels of effectiveness of anti-chorea drugs for relieving chorea after failure of hyper glycemia control to resolve symptoms. There are four main categories of anti-chorea drugs: antipsychotics, GABA receptor agonists, selective serotonin reuptake inhibitors, and dopamine receptor antagonists. The current study indicates that haloperidol is the most common monotherapy agent against chorea associated with DS, followed by tetrabenazine, risperidone, and clonazepam. Combination regimens may also be considered for patients with refractory chorea. It is important to note that because some patients can experience side effects such as tremors from taking dopamine receptor antagonists, one should start with a small dose and slowly increase.[8] Although the majority of DS patients have a good prognosis, the risk of recurrent chorea after resolution of basal ganglia anomalies is quite high; the recurrence rate is close to 20% in all cases. So that glucose control adherence must be maintained in the treatment process to avoid recurrence after this remission of symptoms.[12]

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