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# An Uncommon Hyperosmolar Nonketotic Condition

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## Abstract

**Introduction:** Hyperglycemia may present with neurological symptoms. This rare condition is called non-ketotic hemichorea-hemiballismus (HCHB). When investigating the causes of neurological symptoms in these cases, magnetic resonance imaging (MRI) imaging typically has a hyperintense appearance in T1 sections and a variable intense appearance in T2.

**Case report:** A 68-year-old female patient was admitted to the emergency department with speech disorder. Blood sugar elevation and cranial tomography revealed hyperdense appearance in the left caudate nucleus and lentiform nucleus. The patient was hospitalized with a preliminary diagnosis of HCHB. Magnetic resonance in hyperintense T1W sequences in the nucleus caudatus head and corpus on the left, putamen, globus pallidus. Significant hypointense signal changes were observed in T2W sequences. The patient was admitted to the hospital for treatment and his blood sugar regulation was restored.

**Conclusion:** HCHB is a rare condition in emergency. After the regulation of blood sugar, neurological picture regression is observed. Blood sugar levels should be checked and appropriate treatment should be given in patients presenting with neurological symptoms. **Keywords:** Hyperglycemia, chorea, hemiballismus.

## Introduction

The hyperosmolar hyperglycemic state (HHS) is the most serious acute hyperglycemic emergency. It is mostly seen within type 2 diabetes. It is a syndrome that presence hyperglycemia, hyperosmolarity, and dehydration in the absence of ketoacidosis. The mortality rate is high. It is a life-threatening emergency. Patients may present polydipsia, polyuria, nausea, vomiting, lethargy. A wide variety of acute focal and global neurologic changes may be present delirium, coma, hemiparesis, and sensory deficits<sup>1-2</sup>. Hyperglycemia has been reported to cause intracranial pathology with intracranial acidosis, extracellular glutamate accumulation, brain edema, blood-brain barrier impairment<sup>3</sup>. The appearance of choreiform movements with hyperglycemia is called hyperglycemic nonketotic hemichorea-hemiballismus (HCHB). Especially in elderly, Asian, female patients, irregular, involuntary, unilateral abnormal movements accompanied by high blood glucose levels were observed<sup>4-6</sup>. In this study, we aimed to present a patient with speech disorder who accompanied hyperglycemic nonketotic conditions without loss of consciousness and choreiform movements.

## Case report

A 68-year-old female patient presented with an urgent speech impairment. She answered only the questions asked, "I am

fine." She had a history of hypertension, coronary artery disease, and diabetes. Light reflex was bilateral positive, pupils were isochoric, facial asymmetry and motor deficits were not present, deep tendon reflexes were bilateral flexor.

Her blood glucose level was 533 mg/dl, blood pressure was 150/89 mmHg, pulse rate was 78/minute, respiratory rate was 14/minute, fever was 36°C. Hemoglobin 12.8 g/dl, leukocyte count 4.4 thousand/ $\mu$ l, platelet count 132 thousand/ $\mu$ l, aspartate aminotransferase (AST) 15 (U/L), Alanine aminotransferase (ALT) 9 (U/L), urea 22 (mg/dL), creatinine 0.74 (mg/dL), blood glucose level 553 mg/dl, serum osmolality 304 mOsm/kg, pH 7.45 and urine ketone was negative. Cranial tomography (CT) revealed hyperdense appearance in the left caudate nucleus and lentiform nucleus. In the emergency follow-up and treatment of the patient, his blood glucose level decreased to 273 mg / dL, and his control CT showed a marked decrease in hyperdensity of the left caudate nucleus and lentiform nucleus (Figure 1). The patient was admitted to the internal medicine department for further examination. During the follow-up in this department, the patient had status epilepticus seizure and she was intubated and taken to the intensive care unit. Midazolam and phenytoin were started for seizures. The patient was found to have a high fever during the follow-up in the intensive care unit. Cranial magnetic resonance imaging (MRI) performed at the time of hospitalization revealed hyperintense

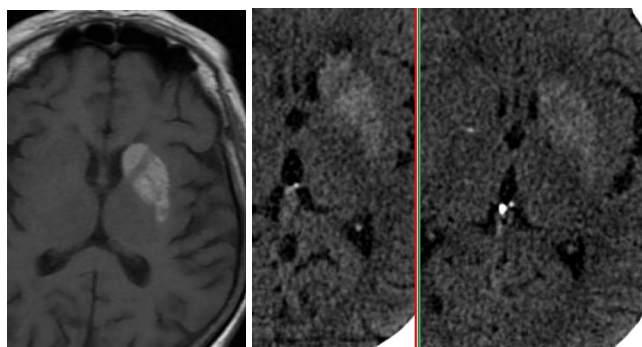


Figure 1

Figure 2

T1W sequences in the nucleus caudatus head and corpus on the left, putamen, globus pallidus (Figure 2). Significant hypointense signal changes were observed in T2W sequences. The hyperintense areas identified in the GE sequence were not compatible with blood. Radiological findings of the patient with a pre-diagnosis of HCHB were interpreted as supporting the pre-diagnosis. The control MRI taken 10 days later revealed similar findings to the previous one. Electroencephalography showed a slight diffuse deceleration of bioelectric activity. The patient was extubated on the 5th day of intubation and followed up in the intensive care unit and then transferred to the ward. The patient whose general condition improved during the follow-up visits was discharged.

## Discussion

HCHB is a rare condition<sup>7</sup>. It is usually unilateral involuntary, continuous and irregular movements, sometimes mimicking central nervous system diseases such as consciousness disorder. The mechanism is not clear. It has been reported that hyperglycemia changes the metabolic activity in the brain by causing hypoperfusion and abnormal movements are the result of this condition<sup>4</sup>. Basal ganglia have metabolically high activity and are affected by metabolic diseases, toxic substances, and neurodegenerative diseases<sup>5</sup>. MRI findings may also be seen due to calcium accumulation in neurons<sup>8</sup>. In MRI, hyperintense appearance in basal ganglia in T1 sections is attributed to secondary swelling of reactive astrocytes to hyperglycemia<sup>9</sup>. Hyperglycemia is associated with an improvement in these cases when treated with hyperglycemia. In these cases, cranial tomography shows hyperdense appearance in the basal ganglia, putamen and/or caudate nucleus on the opposite side of the lesion<sup>9</sup>. MRI typically has a hyperintense appearance in T1 sections and a variable intense appearance in T2<sup>3-6</sup>. It has been shown that petechial hemorrhages or calcifications may also cause these radiological features<sup>7-8</sup>. In the case reported by Hansford et al., it was reported that classical images were detected on CT and MRI without typical movements<sup>10</sup>. In our case, typical lesions were detected on CT and MRI without choreiform movements. These radiological findings may be

confused with a stroke<sup>11</sup>. Measuring blood glucose levels in these patients is life-saving. HCHB, which can be seen in a wide range of symptoms, has a good prognosis with appropriate treatment<sup>5</sup>. In patients with known diabetes, irregular blood glucose level regulation, or newly detected high blood sugar levels, the emergency physician should consider the diagnosis of HCHB in abnormal choreiform movements.

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