

Hemichorea-Hemiballism in Hyperosmolar Hyperglycemic State: A Rare Presentation

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Abstract: *Hemichorea-hemiballism is a syndrome of involuntary movement in one side of the body because of a process in basal ganglia. This condition considered as a rare presentation in hyperglycemic hyperosmolar state. Knowledge about this syndrome can lead to better management of the disease. We present a case of a 52 year old woman with the complaint of involuntary movement on her right upper and lower extremities, since four hours before admission. The patient was diagnosed with diabetes mellitus type 2 on oral anti diabetic but didn't take her medicine in the last 2 months. Blood glucose control achieved after 11 days of hospitalization. The involuntary movement ceased on the 17th day. This case shows hyperglycemic hyperosmolar state with hemichorea-hemiballismus as one of the complication. The involuntary movement stopped few days after good blood glucose control. The fluctuation of the blood glucose is to be expected because of the complicated nature of the diagnosis and comorbidity in our patient.*

Keywords: hemichorea, hemiballism, hyperosmolar hyperglycemic state, diabetes mellitus

1. Introduction

Hemichorea-hemiballism (HCHB) syndrome is characterized by unilateral involuntary movements proximal to the body, caused by vascular disruption in contralateral striatum of basal ganglia.^[1] Hemichorea characterized as random involuntary movement, while hemiballism is sudden and swinging movements limited to one side of the body.^[2] This syndrome usually appear as one-sided chorea/ballism in patient with intracranial or metabolic diseases. Intracranial disease such as ischemic or hemorrhagic stroke, neoplasm, and encephalitis were reported to have HCHB syndrome as one of their manifestation. The same manifestation occur in the patient with metabolic disease: Wilson's disease, thyrotoxicosis, systemic lupus erythematosus, and non-ketotic hyperglycemic hyperosmolar state.^[1,3]

Several literature described HCHB in hyperglycemic hyperosmolar occurs mostly in elderly female with Asian descent, which had history of uncontrolled diabetes for a long time.^[2] HCHB related with hyperglycemia hyperosmolar state was first reported 60 years ago and they were quite rare, with the estimated prevalence worldwide is less than 1 in 100,000. The female to male ratio is 1.8 : 1. Reported case mostly came from Asia, but there was a report from South America.^[4]

In HCHB, treatment of underlying disease is important. This is due to no anti-choreic medication is particularly effective, and adverse effects may worsen function disability, hence the importance in recognizing this condition.^[5] Due to rarity and importance of prompt recognition and treatment, we decided to make this case report. The aim of this case report is to present a case of HCHB as an unusual presentation for hyperglycemic hyperosmolar state as glycemic control is the key to resolution of such condition.

2. Case Report

A 52-year old woman came to the Emergency Room (ER) with a complaint of involuntary movement in her right lower arm and right lower leg since 4 hours prior. The involuntary movement happened for less than one minute and kept repeating in random time interval. Patient had a history of cerebrovascular thromboembolic non hemorrhagic stroke two years ago that caused right hemiparesis. Patient had a history of diabetes mellitus type 2 since 5 years ago, was prescribed with metformin 3 times a day but stopped taking the medicine since 2 months prior. She had glucometer at home and often time her blood sugar was between 300-400 mg/dL. Since one month ago, patient has a wound in her right foot, started with a painless blister ±3 cm in diameter. The wound got worse and expanded from her sole to her ankle.

On admission, patient was conscious with normal vital sign (blood pressure of 100/70 mmHg, pulse rate 90 bpm, respiratory rate of 25x/ minutes, and axillar temperature of 37°C). Rales were found on both lungs. The wound in her right foot was ±15 cm with necrotic tissue and pus.

Laboratory finding on admission shows blood glucose is 771 mg/dL. Other laboratory findings shows leukocyte count of 29,550/μl, hemoglobin of 8,8 g/dL, thrombocyte count 580.000/μl, blood urea nitrogen of 113 mg/dL, creatinine serum of 4.2 mg/dL, sodium of 123mmol/L, potassium 5.1 mmol/L, and chloride 86 mmol/L. Blood gas analysis is done and the blood pH is 7.27, pCO₂ 33 mmHg, pO₂ 93 mmHg, HCO₃ 15 mmol/L, ABE (Actual Base Excess) -12 mmol/L, SBC (Standard Bicarbonat) 16 mmol/L, and O₂ saturation 95%. The urinalysis result shows proteinuria +2, glucose +3, blood +3, keton +1, leukocyte esterase 500 Leu/μL, the sediment of erythrocyte 5-6/HPF (high power field), leukocyte of 7-8/HPF, bacteria ++, crystal +. Chest radiograph shows infiltrate in both lungs. Patient was scheduled for head Computed Tomography-scan (CT-scan), but because of the limited resources, the condition of the patient, and the current pandemic of Corona Virus Disease-

19 (COVID-19), we decide to delay sending her to another hospital to do head CT-scan. Her blood culture was not examined due to limited resources.

Patient was treated with sodium chloride 0.9% 1000 ml in the first hour of admission, then 1500 mL per 24 hours, ceftazidime 2x1 gram IV, levofloxacin 1x750 mg IV, insulin 8 unit bolus IV, then continued with insulin drip 4 unit/ hour IV, esomeprazole 1x40 mg IV, phenytoin 3x100 mg PO, folic acid 2x1 mg PO and clobazam 2x0.5 mg PO. Blood glucose were monitored every 1 hour. Patient was transfused with packed red cell with target hemoglobin 10 g/dL.

During hospitalization, blood glucose was difficult to control. Phenytoin and clobazam was discontinued and the patient was given haloperidol 2x5mg PO and valproic acid 2x150 mg PO on the 10th day of hospitalization due to minimal response of the treatment. On the 17th day of hospitalization, blood glucose control was achieved, and the involuntary movement decreased and stopped.

3. Discussion

The HCHB in hyperglycemic hyperosmolar state is an unusual presentation and often misdiagnosed. It is believed that HCHB pathogenesis is related to hyperglycemia and hyperviscosity. Hyperglycemia disturbs cerebral perfusion, activating anaerobic metabolism and decreasing inhibitory transmitter (GABA) in the basal ganglia. Hyperviscosity is also related to hyperglycemia. Temporary ischemia of striatal neurons is caused by hyperviscosity damaging the blood brain barrier. HCHB is the product of uncontrolled hyperglycemia and vascular disturbance that causes striatum dysfunction. Under microscope, the striatal area shows there were signs of neuronal loss, gliosis, reactive astrocytes, and no infarction or bleeding found.^[2,6]

In acute setting, acquired causes of HCHB is most likely. In examination, confirming the presence of HCHB is the first priority. It is best to examine out of the corner of the eye, for example fidgeting foot while patient is speaking, especially in subtle case. Investigation of hemichorea should be directed to most likely cases, with blood test and neuroimaging.^[5] In HCHB caused by hyperglycemic hyperosmolar state, head CT scan will show hyperdensity lesion in the putamen (basal ganglia) region.^[7] Case series by Misra et al. shows that Magnetic Resonance Imaging (MRI) imaging in patient with HCHB can varies. One patient showed normal MRI imaging, but other patient showed hyper-intensities in basal ganglia.^[8] It shows heterogeneous nature of this entity.^[9] In settings where facility is available, imaging such as head CT scan and MRI should be performed to help rule out other possible causes and support the diagnosis on HCHB. But if there is no available facilities, diagnosis could be made by clinical presentation and hyperglycemia.^[7] From the beginning of admission, our patient was alert and has involuntary movement in her right lower arm and leg with high blood sugar hence the diagnosis of HCHB in hyperglycemic hyperosmolar state was made.

Patient's poorly controlled blood sugar was worsen due to infection on her right foot, pneumonia and urinary tract infection. The diagnosis were made by following the criteria of pneumonia, urinary tract infection, and sepsis.^[11-16] Pneumonia was made as diagnosis based on the rales found in both her lungs, and the bilateral infiltrate on both lungs on chest radiograph. Urinary tract infection can be diagnosed with unspecific symptoms or condition with appearance of bacteriuria, leukocyte esterase, pyuria, and microscopic hematuria in patient's urinalysis. Because of this this three sources of infection, leukocytosis, systolic blood pressure (100mmHg) and tachypnea, our patient fulfilled the diagnostic criteria of sepsis, which contributes to her metabolic acidosis state and difficulty of achieving target blood glucose.

Hemichorea-hemiballismus happens due to severe hyperglycemia, thus, control of blood sugar will usually resolves it partially or completely, but radiological resolution takes around 6 months.^[10] In chorea, treatment of underlying disease is more important. Because, no anti-choreic medication is particularly effective, and adverse effects may worsen functional disability. For functionally disabling chorea, sulpiride, olanzapine and risperidone at low dose is useful.^[5] In several case reported before, the HCHB in hyperglycemic patient resolved partially or completely few days until few weeks after the normal blood glucose level maintained.^[1,2,3] The involuntary movement in our patient ceased after 17 day of hospitalization and 6 days after blood glucose was controlled. This emphasized the importance in blood sugar control in this patient, as medication did not give significant improvement.

4. Conclusion

Hemichorea-hemiballismus is an unusual presentation for hyperglycemic hyperosmolar syndrome. It should always be considered in poorly controlled diabetes mellitus patient with prompt recognition and glycemic control is the key to resolution.

5. Author Contribution

All authors contributed equally.

6. Conflict of Interest

There is no conflict of interest in this case report.

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