

HEMICHOREA-HEMIBALLISM SYNDROME WITH NON KETOTIC HYPERGLYCEMIA

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ABSTRACT

Background: Hemichorea hemiballism can result from multiple disease including non ketotic hyperglycemia. Focal neurological symptoms may be the initial clue for non ketotic hyperglycemia. **Objective:** To assess the frequency of Hemichorea hemiballism associated with non ketotic hyperglycemia. **Methodology:** In this cross sectional study, convenient sampling technique was used. It was conducted in Pathology and Radiology departments, Bahawal Victoria Hospital (BVH) from 1st January 2016 to 31st May 2017. During this period a total of 20 patients, who presented with acute hemichorea and hemiballism were selected. A detailed history and examination was done in all patients. Blood sugar, serum electrolytes, serum calcium, Hb A1C, liver function tests, renal function tests, lipid profile, urinary ketones and MRI were done in these patients. Hemichorea hemiballism was labeled when patients have hypoglycemia and showed high signal intensity in basal ganglia on T1 weighted MRI and no other cause was present. **Results:** Out of total 20 patients, 6 (30%) has hyperglycemia and showed high signal intensity in basal ganglia on T1 weighted MRI scans, which is typical for hyperglycemia associated hemichorea and hemiballism. Other patients had other causes including hemorrhagic infarct and vascular malformations. **Conclusion:** This study showed that many patients have hemichorea hemiballism and highlights the importance of distinguishing hemichorea hemiballism syndrome from other pathologies as it is resistant to anticonvulsant therapy and a mere glycemic control can result in resolving the symptoms and saving the patients from unnecessary medications.

Key words: Hemichorea- hemiballism, Non-ketotic hyperglycemia, Seizures, Movement disorders.

INTRODUCTION

Diabetes mellitus patients may have non-ketotic hyperglycemia and may result in hemichorea-hemiballism (HC-HB) associated with unique radiological features.¹ Human brain needs a continuous supply of glucose for its function and stable plasma glucose levels are essentially maintained through hormones and signaling mechanisms.² Hyperglycemic patients can present with HC-HB, hypotonia, and/or seizures.^{3,4} HC-HB is an involuntary and non-rhythmic movement disorder involving one side of the body, that results from a vascular insult.^{5,6,7} Neoplasm, traumatic brain injury, vascular malformations, tuberculomas or non-ketotic hyperglycemia being other causes.⁸ Association of HC-HB with hyperglycemia is reported in literature among elderly patients with diabetes mellitus and in Asians.^{6,7,9} Besides HC-HB, T1 shortening of the basal ganglia can occur in liver disease, neurofibromatosis type 1, and calcifications.¹⁰ These symptoms can be misdiagnosed as stroke or epilepsy and with the help of laboratory tests and imaging, such cases can be correctly diagnosed and treated. Delayed diagnosis in such cases affects the morbidity and mortality.¹¹⁻¹³ This study highlights the significance of identifying hemichorea hemiballism syndrome from other diseases as it is resistant to anticonvulsant

therapy¹¹ and a mere glycemic control can result in resolving the symptoms and saving the patients from unnecessary medications. The objective of this study was to assess the frequency of hemichorea-hemiballism associated with non ketotic hyperglycemia.

METHODOLOGY

In this cross sectional study, convenient sampling technique was used. It was conducted in Pathology and Radiology departments of Bahawal Victoria Hospital, Bahawalpur (BVH) from January 1st 2016 to May 31st 2017.

During this period a total of 20 patients, who presented with acute hemichorea and hemiballism were selected. A detailed history and examination was done in all patients. The patients who had a family or personal history of epilepsy, history of head injury or any kind of seizures were excluded from the study. Blood sugar, serum electrolytes, serum calcium, Hb A1C, liver function tests, renal function tests and lipid profile were done on fully automated chemistry analyzer (Mindray BS 400, Merck Diagnostics) and found in normal range. Urinary ketones were done by urine strips which was negative in all studied samples and MRI were done in the department of Radiology. Hemichorea hemiballism was labeled when patients have hyperglycemia and showed high level of signal

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Received: 12-03-2018

Accepted: 21-08-2018

intensity in basal ganglia on T1 weighted MRI and no other cause was presented.

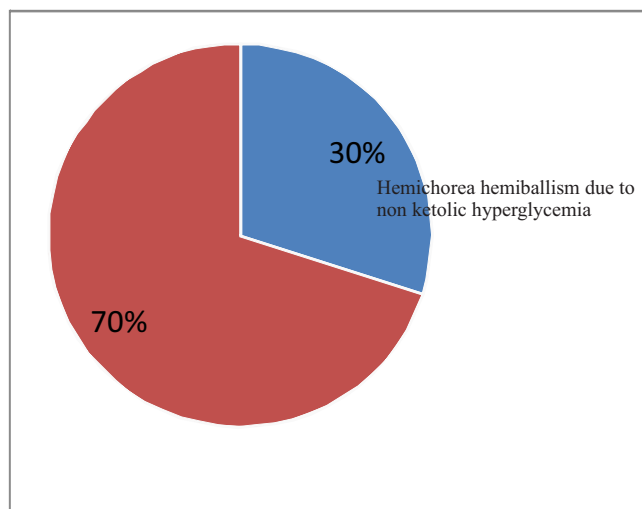
SPSS 23 was used for entering data and calculating mean, frequency and standard deviation of variables. Descriptive statistics were used to show patients' base-line characteristics. Ethical approval was sought from ethical committee of hospital.

RESULTS

Out of the twenty patients, six patients (30%) has hyperglycemia and showed high signal intensity on T1 weighted MRI scans which is typical for hyperglycemia associated hemichorea and hemiballism. (Figure I) Seven patients (35%) has infarction, four (20%) has vascular malformations and three (15%) has other causes for their movement disorders.

The mean age of the patients with hemichorea and hemiballism was 64 ± 3 years. Range of duration of diabetes mellitus was 5-20 years. Among these six patients having hemichorea hemiballism mean plasma glucose level was 15.5 ± 1.5 mmol/L, mean serum osmolality was 298 ± 4 mosm/L and mean HbA1c was 11.91 ± 0.5 . Ketones in urine was negative in all six patients and among these six patients abnormal movements improved after correction of hyperglycemia.

Figure I: Frequency of hemichorea hemiballism due to non ketotic hyperglycemia.



DISCUSSION

Nonketotic hyperglycemia is hyperglycemia in the absence of ketoacidosis. In hemiballismus/hemichorea there are involuntary rapid and jerky movements and occasional uncoordinated swings

mostly unilateral limb. Uncontrolled diabetes mellitus may present with this disorder after years of poor glycemic control.¹² The pathogenesis underlying this is believed to be a because of hyperglycemia-induced changes in brain resulting in inhibition of the subthalamic nuclei and excitatory cortical output.¹³

In our study, male to female ratio was 2 to 1 where as Younes et al report that more females patients present with this disorder.¹⁴ The male to female ratio, however, is controversial.¹⁵ The mean age of the patients in this study was 64 years which is consistent with other studies.¹⁶⁻¹⁸ Glucose levels varied from 12.5 to 18.5mmol/L and HbA1c from 11 to 13.1%. Osmolality was between 290 to 306 mosmol/L. Other studies and case reports showed similar findings.^{14,15} Vascular lesions are the most common cause of HB-HC¹⁶ and our study showed consistent results however Roy et al reported non-ketotic hyperglycemia as the commonest cause.⁸

More and more studies are showing increased prevalence of this condition in people of Asian origin^{17,18} so this study is important as it highlights the importance of distinguishing hemichorea hemiballism syndrome from other pathologies as it is resistant to anticonvulsant therapy and a mere glycemic control can result in resolving the symptoms.

CONCLUSION

This study showed that many patients could have hemichorea hemiballism and highlights the importance of distinguishing hemichorea-hemiballism syndrome from other pathologies as it is resistant to anticonvulsant therapy and simply glycemic control can result in resolving the symptoms and saving the patients from unnecessary medications.

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Article Citation: Sarfraz L, Nisar S, Hussain ST. Hemichorea-Hemiballism Syndrome With Non Ketotic Hyperglycemia . *JSZMC* 2018;9(3): 1473-75.