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Case Report

C-H-BG Syndrome in a Child – A Rare Case Report

Authors
Geetha M, MD[1], Saminathan D, MD[2]
[1]Resident, [2]Professor and Head
Department of Pediatrics, KAPV Government Medical College and Mahatma Gandhi Memorial Government Hospital, Trichy, Tamil Nadu, India
Corresponding Author
Geetha M
Contact: Ph: 9686320957, Email: *gee_festoon@yahoo.co.in*

Abstract
C-H -BG (chorea, hyperglycemia, basal ganglia) syndrome is a described entity with good prognosis, with less than ten reported cases in pediatrics all in adolescence.
We present this case of a eight year old boy, with hemichorea and non ketotic hyperglycemia.
Keywords: *Chorea, Hyperglycemia, Pediatrics, Chbg Syndrome, Diabetes.*

Introduction

Chorea is a movement disorder characterized by rapid quasi purposive involuntary movement due to a lesion in the basal ganglia. It can be due to metabolic diseases, hypoxic-ischemic events, vascular disorders, structural abnormalities, trauma, drugs and toxins, infections and inflammatory immunological diseases. Chorea involving one side of the body referred as hemi chorea, a rare etiology may be hyperglycaemia necessitating early recognition and appropriate treatment. We are presenting this case with less than ten reported in pediatrics and the first below adolescent age group.

Case Report

A seven year old boy presented with involuntary left upper limb movements since two days. He did not have fever, headache, seizures, altered sensorium, drug intake and pre-existing neurological illnesses. He was the only child born to non consangineous parents and was developmentally and nutritionally normal. General examination, vitals, systemic examination was normal. Left upper limb showed positive milk maid sign and pronator sign consistent with chorea. There was no neurocutaneous marker and abnormal odour. Keyser Fleischer ring was absent in slit lamp examination.

Significant positive investigations were blood sugar of 406mg/dl, urine sugar positive 4+, Urine and plasma ketones were negative. Complete blood counts, ESR, arterial blood gas analysis, renal functions, serum electrolytes, liver functions and were normal.

Computed tomography of brain showed asymmetric calcification of bilateral caudate nucleus and striatum, more on the left side.

C peptide level was 0.14mg/dl confirming type I diabetes mellitus. Thyroid Profile, peripheral smear, Serum calcium, phosphorus, 25 OH Vitamin D, Parathormone, Anti steptolysin O titre Anti nuclear antibodies and Serum Ceruloplasmin were normal.

Patient treated with insulin therapy and neuroleptics haloperidol, euglycemia attained and chorea resolved. Child was discharged with mixed insulin regimen and haloperidol, without any sequelae. Haloperidol was stopped after three months. At follow up of three years, child is symptom free with no recurrent episode and euglycemic on insulin therapy.

Discussion

Hyperosmolar hyperglycemic non-ketotic (HHNK) syndrome is a clinical syndrome of severe hyperglycemia, hyperosmolarity, and intracellular dehydration without ketoacidosis[(1)] This is a rare disease that deserves attention because it is reversible with correction of hyperglycemia. Thus, prompt recognition and treatment is essential to avoid adverse outcomes[(2)]. HHNK is a rare cause and presentation for chorea movements. Hemichorea-hemiballism (HCHB) is a spectrum of involuntary, continuous non-patterned movement involving one side of the body. Hemiballism often evolves into hemichorea. These results from a lesion in the contralateral subthalamic nucleus and adjacent structures. This is now termed as C-H -BG (chorea, hyperglycemia, basal ganglia) syndrome.

In literature, most cases have been described in individuals of Asian descent, females, and the elderly. There are less than ten cases reported in pediatric age group and all are in adolescence. This is the first case reported in young children.

Though the pathogenesis of C-H-BG syndrome is not fully understood, numerous theories abound. These revolve around uncontrolled hyperglyc-emia, GABA depletion, Acetylcholine synthesis, hyperviscosity, disruption of blood brain barrier, vascular insufficiency and incomplete transient dysfunction of the striatum[(2),(3)].

Histological findings in patients with C-H-BG syndrome have reported selective neuronal loss, gliosis, and reactive astrocytes, without evidence of hemorrhage or infarction at the striatal areas[(4)].

Neuro-radiological imaging results are characteristic in patients with hemichorea-hemiballism non-ketotic hyperglycemia. Computed tomography (CT) demonstrates hyperattenuation in the striatum contralateral to the affected side. Magnetic resonance imagining (MRI) shows T1-weighted hyperintensities in striatum and globus pallidus, with restricted diffusion on diffusion-weighted imaging[(5)].

The onset of the disorder usually coincides with severe hyperglycemia and follows a temporal relationship between restoration of blood glycemic levels and improvement of the chorea[(6)]

Several case reports have documented that the hemiballism/hemichorea can occur a few weeks after the blood glucose levels have been controlled and are actively being treated[(7)]. This suggests a delayed reaction to the severe hyperglycemia.

The mainstay of treatment is aggressive glycemic control with either partial or complete resolution of hemichorea-hemiballism[(8)]

Conclusion

CHBG Syndrome, a rare entity hardly reported in children, for abnormal involuntary movements caused by hyperglycemia is a treatable disorder with a good prognosis[(9)].

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