Hemichorea-Hemiballismus syndrome in Ketotic hyperglycemia

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ABSTRACT

Diabetes mellitus is a common disorder in Asian people occasionally presenting with unusual manifestations such as hemichorea-hemiballismus (HC-HB) is a rare manifestation of primary diabetes mellitus. Since chorea-ballismus can be life threatening, recognition of this disorder is important, because HC-HB caused by hyperglycemia is a treatable disorder with a good prognosis. Usually Hemichorea-Hemiballismus is induced by non-ketotic hyperglycemia; here we are reporting two cases of elderly women who presented as HC-HB induced by hyperglycemia with positive urine ketone bodies. We also described Computed Tomography (CT), Magnetic Resonance Imaging (MRI) findings in these two patients.

Keywords: Hemichorea-Hemiballismus, basal ganglia, CT, MRI, ketone bodies

INTRODUCTION

The term chorea is derived from Greek choreia, means “a dance”. This hyperkinetic movement disorder consists of irregular, unpredictable, brief, jerky movements that develop from one part of the body to another. Chorea results due to hereditary neurodegenerative diseases, structural damage to deep brain structures, metabolic derangement, certain drugs and hormones. Caudate nucleus, putamen, subthalamic nucleus and thalamus are frequently involved in generation of choreiform movements with a specific neuroradiological findings in brain Computed Tomography (CT) and Magnetic Resonance Imaging (MRI)1,2.

CASE REPORTS

Case 1: A 58 years old female patient came to OPD with complaints of involuntary movements of left upper limb and lower limb, sudden in onset and progressively worsening. These movements are subsiding during sleep. On admission patient was conscious, irritable. Neurological examination showed that patient had involuntary movements, continuous flexion-extension and rotational movements of the left upper limb and lower limb and rabbit mouth movements of the face. No evidence of weakness or sensory disturbance. Deep tendon reflexes were normal and plantar responses were negative. No bowel and bladder incontinence. Her blood glucose level at admission is 441 mg/dl, HbA1C-11.5. Urine for ketone bodies positive(++) . Arterial Blood Gas analysis(ABG) showed pO2:7.1, HCO3-: 11 mmol/L. All other laboratory tests were normal. Initial CT brain showed hyper densities in bilateral basal ganglia (Fig 1). MRI showed hyperintensities on T1weighted (T1W) images (Fig 2), hypointensities on T2 weighted(T2W) images (Fig 3).

After correction of hyperglycemia involuntary movements improved markedly in 5days. These symptoms disappeared completely after 2months with blood sugar control and low dose Tetrabenazine.

Fig 1: CT Brain (p) showing hyper densities in bilateral basal ganglia.

Fig 2: MRI shows hyper intensities at bilateral basal ganglia on T1W images.
Case 2: A 65 years old female patient presented to OPD with complaints of involuntary choreiform movements of both upper limbs and lower limbs. Patient was conscious and irritable. No evidence of weakness or sensory disturbance. Deep tendon reflexes were normal and plantar responses were negative. No bowel and bladder incontinence. Her blood glucose level at admission is 449 mg/dl, HbA1C-13.8. Urine for ketone bodies positive (+++). ABG showed pH: 7.23, HCO₃⁻: 9.6 mmol/L. All other laboratory tests were normal. Initial CT Brain showed minimal diffuse hyper densities noted bilaterally in the lentiform nucleus (Fig 4). MRI Brain showed hyper intensities on T₁W images at basal ganglia (Fig 5). Patient sugars were controlled by intravenous insulin infusion and low dose tetrabenazine (25mg TID) was added. After controlling sugars her movements were improved.

DISCUSSION

Hyperglycemia induced HC-HB is more common in women with a mean age of 72 years. The possible mechanism for hyperglycemia associated HC-HB is poor glycemic control and ketoacidosis. Imbalance between direct and indirect pathways in the basal ganglia leads to increased dopaminergic activity. GABA is the main inhibitory neurotransmitter responsible for indirect and direct pathway in the basal ganglion. Due to changes in striatum and ischemic excitotoxicity of GABAergic neurons results in increased inhibition of subthalamic nucleus (STN). Increased inhibition of STN results in its excitatory action on the internal segments of the globus pallidus (GPI), which leads to decreased GPI neuron inhibitory action on the thalamus. Decreased inhibition of thalamus causes increased excitatory action on cortex results in development of chorea⁴. Patients with positive urine ketones have higher acetoacetate levels. Synthesis of GABA results from increased levels of acetoacetate in ketotic hyperglycemia.

Typically they present as hyper density in striate area in CT and hyper intensity in putamen and/or caudate nucleus in T₁W MRI. Lesions can be differentiated from hemorrhage as there will be no mass effect, edema, volume loss. The clinical presentation can be either unilateral or bilateral. If unilateral, the imaging findings are present at the contralateral basal ganglia. Management consists of strict glycemic control. In refractory cases post synaptic dopamine depleting agents (haloperidol, risperidone), pre synaptic dopamine depleting agents (tetrabenazine) can be used⁵.

CONCLUSION

Here we are reporting two elderly female HC-HB patients caused by hyperglycemia with positive urine ketones. Movement disorders like chorea may be the clinical presentation of the hyperglycemia which could completely recover on early detection and correction of hyperglycemia⁶.
REFERENCES


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