Case Report

Hemichorea - hemiballismus due to non-ketotic hyperglycemia: a case report

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Abstract

Non-ketotic hyperglycemia induced Hemichorea-Hemisballismus is a reversible condition found in elderly diabetics with poor glycemic control. The patients present with acute onset of involuntary movements and neuroimaging is invariably done to rule out stroke. T1 hyperintense signal in the basal ganglia [characteristically involving the putamen] and absence of stroke like imaging features helps in making the diagnosis of Non-ketotic hyperglycemia induced Hemichorea-Hemisballismus and rule out stroke. Correction of hyperglycemia results in marked and immediate improvement of the symptoms.

Keywords: Hemichorea-Hemisballismus [HC-HB], non-ketotic hyperglycemia [NKH], Type 2 diabetes Mellitus, basal ganglia T1 hyper intensity.

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INTRODUCTION

Hemichorea-Hemisballismus [HC-HB] is an unusual involuntary non-rhythmic, hyperkinetic movement disorder involving one side of the body[1,2]. The most common cause of HC-HB is vascular insult to the contralateral basal ganglia[1-3]. Other conditions causing HC-HB include non-ketotic hyperglycemia [NKH], human immunodeficiency virus infection, vasculitis, mass lesion and drug induced [antiepileptics, oral contraceptives][1,2,4]. Association of hyperglycemia and HC-HB is well recognised[5]. It is seen to be more prevalent in elderly patients with Type 2 diabetes mellitus and in the asian population[2, 3,6].

CASE REPORT

A 60-year-old woman presented to the outpatient department with a seven-day history of sudden onset of involuntary movements of the left upper and lower limb. Symptoms were minimal at first and progressively worsened over the past one week. Patient was a known case of type 2 diabetes mellitus for the past 3 years on treatment. No history of neurological disorders or other significant illness was elicited. On examination, power was normal in all 4 limbs. Milkman grip was positive in the left upper limb and the pronator sign was also positive. Involuntary movements were more in the left upper limb and the left lower limb. Laboratory investigations showed mean plasma glucose level of 321 mg/dl under hemoglobin A1c level of 12.8. Urine sample was negative for ketones. MRI of the brain was requested to rule out stroke. MRI showed T1 hyperintense signal in the left caudate nucleus with no associated corresponding abnormality in T2-weighted, diffusion weighted and T2 weighted gradient sequences [figure 1 (a-d)]. With the Background clinical history and Laboratory investigation results, diagnosis of Nonketotic hyperglycemia induced Hemichorea-Hemisballismus was made. Patient was managed with T. Revocon 25 mg, Inj. magnesium sulfate, T. Oleanz and insulin for glycemic control. Patient improved symptomatically and was hence discharged.

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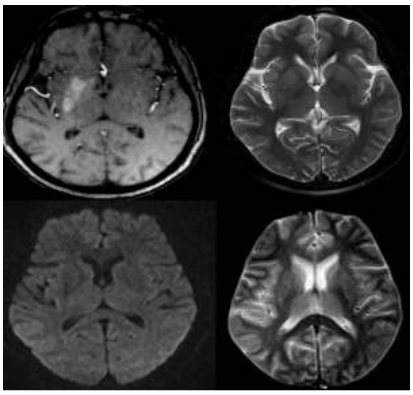

Figure 1 (a-d): MRI sections of the brain show T1 hyperintense signal in the left caudate nucleus with no associated corresponding abnormality inT2-weighted, diffusion weighted and T2 weighted gradient sequences

DISCUSSION

Bedwell, in the year 1960, first reported the association of Hemichorea-Hemisballismus with hyperglycemia[7,8]. Subsequent studies supported the theory of Bedwell[5,9,10]. The pathophysiology of nonketotic hyperglycemia induced Hemichorea-Hemisballismus is poorly understood. It is controversial and confusing to the various studies providing Variable histopathological results[1,5]. Initially, the signal changes were proposed to be due to focal hemorrhage and calcification. However, histopathology studies and follow up imaging do not favor the above-mentioned theories[1,5]. Recent histopathological studies have shown gliosis, gemistocyte accumulation, and selective loss of neurons with no evidence of hemorrhage or infarction[11-13]. The role of hyperglycemia causing endothelial dysfunction and increased oxidative stress is also a possibility, but yet to be established[14]. MR imaging reveals abnormal signal in the putamen with or without the involvement of the globus pallidus and caudate nucleus. the onset of Hemichorea-Hemisballismus is commonly caused by abnormal signal in the Contralateral basal ganglia; however, Couple of cases of Hemichorea-Hemisballismus with involvement of the ipsilateral basal ganglia have been reported[7]. A Other common causes of T1 hyperintense signal of the basal ganglia are hepatic Encephalopathy[10], manganese toxicity due to parenteral nutrition, mild ischemia/hypoxia, neurofibromatosis, Wilson's disease and carbon monoxide poisoning[16]. The above conditions usually affect the bilateral basal ganglia in contrast to nonketotic hyperglycemia induced Hemichorea-Hemisballismus. A few cases of bilateral basal ganglia involvement in nonketotic hyperglycemia induced Hemichorea-Hemisballismus have also been reported. However, nonketotic hyperglycemia induced Hemichorea-Hemisballismus patients have a slightly different presentation compared to the other patients. Signal changes in the globus pallidus are seen in patients with hepatocerebral disease and manganese toxicity, in contrast to putamen involvement in nonketotic hyperglycemia induced Hemichorea-Hemisballismus. Patients improve immediately once the euglycemic treatment is instituted. The imaging findings are usually reversible, but occasionally the signal changes may persist for More than 6 months. in patients in whom the imaging findings resolved, the resolution of the signal changes lag behind clinical improvement.

CONCLUSION

Nonketotic hyperglycemia induced Hemichorea-Hemisballismus Is an important differential diagnoses in patients with Hemichorea-Hemisballismus. Recognition of the characteristic clinicoradiological signs help in institution of early and appropriate therapy.

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