

A Case Series of Hemichorea due to Non Ketotic- Hyperglycemia with Unique MRI Brain Finding

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Abstract

Hemiballismus- hemi chorea (HB-HC) is an uncommon disorder, mostly seen among the people in Asian region. Recent reports have revealed that it is associated with non-ketotic hyperglycemia. MR spectroscopy in the present study showed a low NAA/Cr, suggesting pronounced energy depletion and neuronal dysfunction. Scientists observed that unilateral lesions occurred in elderly diabetic patients in striatum due to hyperglycemia. Our study reveals that, dyskinesia in elderly diabetic patients leads to an initial manifestation of diabetes among the both sexes.

Keywords: Hemichorea; MRI; Non ketotic-hyperglycemia

Introduction

Hemi chorea generally occurs due to a stroke, neoplasm or granuloma in the contra lateral basal ganglia [1]. Metabolic derangements especially hyperglycemia can also cause focal neurological dysfunction like hemiparesis, focal seizure [2,3]. Recently there have been case reports of hemichorea reported due to non-ketotic hyperglycemia with characteristic MRI findings predominantly in the Asian population [4]. The aim of this study is to highlight the important aspects pertaining to the clinical and neuroradiological diagnosis of hemichorea associated with nonketotic hyperglycemia.

Materials and Methods

Patients admitted with acute hemichorea were included for the study. A detailed history and examination was done in all patients. Hemogram, Blood sugar, serum electrolytes, serum calcium, HbA1C, liver function test, renal function test, lipid profile and MRI of the brain were done.

Results

From May 2008 to September 2009 there were 9 patients admitted with hemichorea. 3 patients (33%) were found to have acute striatal infarcts, 2 patients (22%) had granuloma and other four patients (45%) had hyperglycaemia without ketosis.

The median age of these 4 patients who had hyperglycemia was 60 years (58 to 70 yrs), male to female ratio was 1:1. All were right handed persons. 3 patients were known diabetic, their mean duration of diabetes was 12 years. 1 patient was detected to be diabetic at admission. None of these patients had any history of cerebrovascular event in the past.

All these patients presented with an acute history of involuntary jerky movements involving one side of the body, mean duration of symptom was 2 days. All had left sided chorea with normal cognition and motor power in the limbs. All had high blood sugars with mean of 318 mg/dl (252-475 mg/dl) and mean HbA1c of 11.2. None had ketone bodies in the urine. Serum electrolytes, Calcium, blood urea nitrogen, creatinine and Liver function tests were normal.

Neuroimaging of these patients with MRI revealed unilateral hyper intensity in Striatum in T1W images contralateral to the side of hemichorea with mild hypointensity in T2 flair. There was no restriction in diffusion weighted images and MRS showed decreased NAA peak (Figure 1).

Chorea subsided once sugars were controlled, average duration was 4 days.

Discussion

Hemiballismus- hemi chorea (HB-HC) is an uncommon disorder, a vascular lesion being the most common cause [1]. More recently there have been reports of HBHC associated with non-ketotic hyperglycemia amongst people of Asian origin [3]. In our study non-ketotic hyperglycemia was the commonest cause for hemichorea. This could be due to the high prevalence of diabetes mellitus which was 62.4 million in 2011 [5] and estimated to increase to 100 million by 2030 [6].

Dubinsky et al. first reported two patients with non-ketotic hyperglycemia with neuroimages that demonstrated a faint increase in densities over the contralateral basal ganglia [7]. Later, Yahikozawa et al. suggested that a combination of HB-HC and striatal hyperintensity on T1- weighted MRI in diabetic patients may constitute a unique syndrome [8]. A study by Lee et al. reviewed the characteristic clinicoradiologic syndrome and suggested that it was benign [9].

Previous studies [3] revealed that this dyskinesia often occurred in elderly diabetic patients and could also be an initial manifestation of diabetes with equal incidence among both sexes. In our study one patient was newly detected with diabetes on admission. The incidence among both sexes was equal.

In MRI T1 weighted images showed hyperintensity signals in the contralateral striatum. The mechanism of this characteristic signal changes on MRI is still debated. In view of the acute clinical manifestation and the striatal lesion corresponding to the territory of lenticulostriate artery, it has been suggested that the lesion may be secondary to vascular insult. Based on the evolution of clinical manifestation and the findings of the neuroimages, Chang et al. [10] suggested that petechial hemorrhage in the putamen might be the pathological mechanism to cause the dyskinesia. Moreover, studies of

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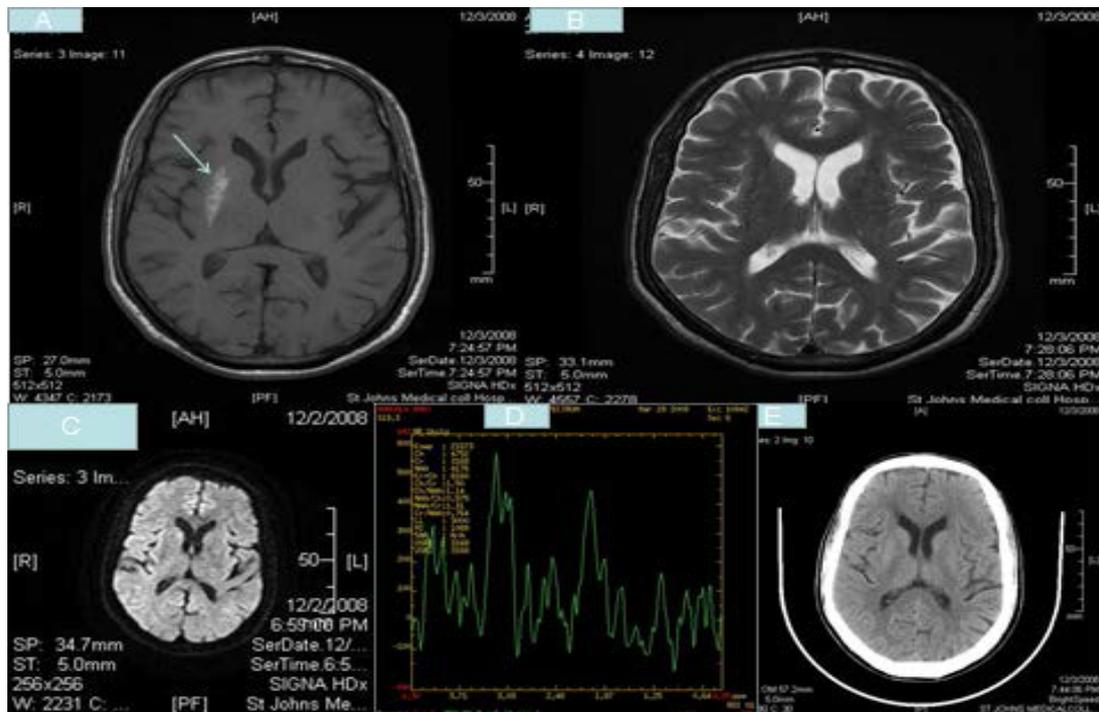


Figure 1: T1W axial image (A) showing striatal hyperintensity (arrow), T2W axial images (B) showing hypointensity, Diffusion weighted images (C) showing no restriction, magnetic resonance spectroscopy (D) showing reduced NAA peak and CT axial images (E) showing hyperdensity in striatum (MRI was done by General Electric HBX, 1.5 Tesla machine, 2007 model with 4.3 version software).

Altafullah et al. and Broderick et al. suggested that the lesions might have resulted from hemorrhagic insult [11,12]. However, a biopsy specimen from the hyperintense putamen revealed only a slight astrocytosis and vacuolization or a fragment of gliotic brain tissue with abundant gemistocytes with no deposition of hemosiderin [13].

MR spectroscopy in the present study showed a low NAA/Cr, suggesting pronounced energy depletion and neuronal dysfunction. This is secondary to neuronal loss or damage either due to hypoperfusion or metabolic failure. Studies by SPECT [14,15] and PET [16] have revealed the reduction of blood flow and metabolism in the contralateral striatum of patients with HB-HC caused by non-ketotic hyperglycemic.

The probable hypothesis as proposed by Juei-Jueng Lin et al. is that the metabolic derangements (hyperglycemia) and cerebral vascular insufficiency both contributed to regional metabolic failure in basal ganglion cells [17].

Proper functioning of basal ganglia has been compromised by chronic ischemia through microangiopathic changes, even though an actual infarct has not yet developed. When a stage of metabolic crisis is achieved, either there is further compromise of circulation and glucose hypometabolism, which precipitates the clinical syndrome of hemichorea [17,18]. This process triggers reactive astrocytosis in the basal ganglia and probably enhances local manganese accumulation and thus produces the typical signal changes seen on MRI [17].

Conclusion

Unilateral lesions can occur in Striatum due to hyperglycemia in elderly diabetic patients, which is hyperintense in T1W images. Recognition of this unique clinicoradiologic manifestation is important because correction of the underlying hyperglycemia will lead to rapid improvement.

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