CASE REPORT HYPERGLYCEMIA: AN UNUSUAL CAUSE FOR HEMICHOREA-HEMIBALLISM

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ABSTRACT

Hemichorea - hemiballism is a rare neurologic disorder due to oxidative stress leading to neurodegeneration of the dentate nuclei and striata. It is rarely observed in diabetes. One such case occuring in an adult female diabetic is described.

Key words: Hyperglycemia hemichorea - hemiballism, diabetes, dentate nuclei, striata.

INTRODUCTION

Diabetes is common all over the world. In Pakistan its prevalence is reported to be about 6.0-6.9% in males and 2.5-3.5% in females. It leads to complications related to every system of the body. Hyperglycemia in diabetic patients is an important cause leading to oxidative stress in central nervous system (CNS)¹. This oxidative stress is an important factor in the pathogenesis of neuron degeneration in CNS^{1,2}. This could lead to hemichorea-hemiballism (HCHB) clinically and hyperperfusion in dentate nuclei and striata leading to striatal hyperintensity on MRI25. The condition was first described in humans by Rector et al in 1982⁶. There has been no report of this condition from Pakistan. We hereby report an interesting case of HCHB due to hyperglycemia with complete remission of symptoms after control of blood sugar level.

CASE REPORT

A 40 year old married lady presented to the emergency department of Civil Hospital Karachi with involuntary movements of left half of the body for 12 days. Movements were sudden in onset and persisted through out the day making it difficult for her to fall asleep. However they disappeared during sleep.

On examination, patient was vitally stable. She was

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having choreiform movements of left upper and lower limb. She could not willfully suppress the movements. Deep tendon reflexes were diminished in all limb. Babiniski sign was negative. Vibration, position sense and sensory system were intact. Examination of all other systems was unremarkable. The complete blood count, serum electrolytes, LFT's, PT, APTT, serum calcium and albumin were within normal limits. The random glucose level was elevated. It was 390 mg/dl at the time of presentation. She was started on Tab. haloperidol 5 mg b.i.d. and combined NPH/Regular insulin 70/30 (inj. Mixtard-30). Her response to insulin was slow and blood sugar fluctuated for few days before finally gradual normalizing with parallel improvement in the involuntary movements. A line graph of her blood sugar levels and involuntary movements on a visual scale of 0-100 are given in Figure 1. As her blood sugar was controlled, her choreiform movements improved remarkably. Her thyroid profile showed normal FT3, FT4 and slightly higher TSH. Serum ceruloplasmin and MRI brain were normal. Haloperidol was stopped after 4 days. A diagnosis of HCHB due to non-ketotic hyperglycemia was made.

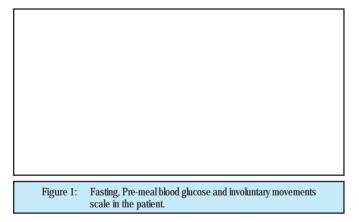
DISCUSSION

Non-ketotic hyperglycemia has been associated with various neurological abnormalities. HCHB is one of them. The characteristic imaging finding in these patients is of striatal hyperintensity²⁻⁵. Despite the characteristic clinical and imaging findings, the underlying mechanism is still unclear. Various mechanisms have been postulated. Some have suggested that it causes ischemic changes in the

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striatum associated with hyperglycemia and hyperviscosity⁷. Examination by [18F]-fluorodeoxyglucose (FDG) positron emission tomography (PET) have documented markedly reduced rates of cerebral glucose metabolism in the corresponding lesions on T1-weighted magnetic resonance images thus providing direct evidence of regional metabolic failure⁸. The metabolic derangements associated with hyperglycemia and vascular insufficiency contribute to regional metabolic failure in patients with poorly controlled diabetes mellitus^{8,9}. These hyperintense lesions have been shown to be reversible on follow up scans after adequate glycemic control¹⁰.

Ohmori et al reported two cases of HCHB in 2005⁴. One of the cases had painful HCHB. Both the cases responded to pimozide and strict glycemic control⁴. The presently reported patient did not have painful HCHB and haloperidol was used instead of pimozide for a short duration of 4 days only. Some researchers have also found topiramate effective in controlling the involuntary movements in such cases¹¹. Ahlskog et al¹² reported in a case series of five cases of HCHB. Four patients did not improve and had persistent involuntary movements despite the glycemic control on five year follow up. One of them did not have any striatal hyperintensity. The cause could not be ascertained of this unresponsiveness¹².



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