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DIABETIC STRIATOPATHY: A RARE MANIFESTATION OF UNCONTROLLED HYPERGLYCEMIA.

Neurology

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ABSTRACT

Diabetic striatopathy is a a rare clinical condition seen in patients with non ketotic hyperglycemia(NKH). It can also be first manifestation of diabetes mellitus in certain patients. Prompt recognition of this condition which presents as hemichorea- hemiballismus(HC-HB) is essential as delay in diagnosis may lead to persistent manifestations,which might not improve even after correction of hyperglycemia.

KEYWORDS

Diabetic striatopathy, Non ketotic hyperglycemia, Hemichorea-hemiballismus

INTRODUCTION:

Diabetic striatopathy (DS) is defined as a hyperglycemic state associated with both or either one of the two following conditions: (1) chorea/ballismus (2) striatal hyperdensity on CT or hyperintensity on T1-weighted MRI sequence.[1] Hemichorea is usually associated with a contralateral lesion in the central nervous system. It may be caused by hereditary neurodegenerative diseases, following structural damage to deep brain structures, autoimmune disorders, metabolic derangement, or drug related(levo dopa exposure)[2]. Hemichorea associated with non-ketotic hyperglycemia (HC-NH) is very rare complication of diabetes mellitus[3]. It is commonly seen in elderly diabetic women with poor blood glucose control. Here we describe a case of elderly diabetic female with hemi chorea with characterstic MRI findings but without hyperosmolar hyperglycemic state(HHS).

CASE:

A 65 year old female presented to us with complaints of Sub acute onset involuntary movement of left side which were non patterned mostly involved the distal extremities and it was more in lower limbs than in upper limb. There was no facial and oro-lingual involvement. The movements disappeared during sleep. There was no h/o fever, no family history, history of recent drug intake other than anti diabetics, behavioural abnormalities, self mutilating behaviour. She had a history of type 2 diabetes mellitus from 10 years and she was on metformin[1gm/day], sitagliptin 100mg/day and insulin glargine 10units. She had a history of cataract extraction 1 year back and had diabetic retinopathy.

On examination patient was conscious alert oriented to time place and person. Her blood pressure was 130/80 mm Hg and pulse rate 80/ minute. She was not dehydrated. She had choreiform movements involving left side sparing the face. She had hypotonia on left side wih normal tone on the other. Reflexes were normal. Power was normal. Examination of cranial nerves didn't reveal any abnormality.

Her random blood sugar at the time of presentation was 308mg/dl. Urea was 94mg/dl, creatinine was 2.3 mg/dl. Serum sodium was 140mEq/ L. Serum osmolality was 315milliosmoles/kg. Urine ketones were negative and an arterial blood gas analysis was within normal limits. Her glycated haemoglobin levels were 13.0 which signified poor glycemic control Rest of the investigations including thyroid function tests, parathyroid hormone levels were within normal limit. Anti nuclear antbody (Hep 2) method was negative. Genetic analysis for CAG repeats were also negative. . CT scan of brain was normal. MRI brain showed T1 hyperintensity in right lentiform nucleus[image 1].

Patient was treated with iv fluids and insulin. Even after normalisation of her blood sugar levels, maintaining it below 180mg/dl within 24 hours, her choreiform movements were persisting. So she was put on haloperidol 2.5mg/day gradually increased to 5mg/ day. Within two week her movements disappeared. Her glycemic control was optimised with subcutaneous basal bolus regimen and haloperidol tapered. At the time of her discharge there were no abnormal movements and patient was maintaining blood sugar levels within range of 140-180mg/dl.

DISCUSSION:

The term "diabetic striatopathy" (DS) has been introduced recently to denote a condition in diabetic patients in which there is a combination of striatal hyperintensity on T1-weighted MRI and contralateral movement disorder.[4] DS is seen in patients with long-standing diabetics with poor control, and it could persist after correction of hyperglycemia which was seen in this case. The patient had a very high Hba1c which reflected her poor control.

Diabetic striatopathy which manifests as hemichorea/hemi ballismus is a rarely described entity, typically seen in elderly Asian women with type 2 diabetes. It has also been very rarely reported in type 1 diabetes and also those presenting with diabetic ketoacidosis[5]. It can also be seen in children[6]. A lot of mechanisms has been hypothesized to explain the diabetic striatopathy but most accepted pathophysiological mechanism is depletion of gamma-Aminobutyric acid, and accumulation of manganese-containing gemistocytes in the basal ganglia.The high signal of T1-weighted MRI images in these patients is caused by ischemic injury[7]. This leads to astrocytes proliferation and the expression of zinc-friendly metalloproteins along with accumulation of manganese ions resulting in paramagnetism[8].

A Meta analysis from 72 articles identified 176 patients (male:female = 1:1.7) with mean age 67.6 ± 15.9 (range, 8–92). Among them, 96.6% had type 2 DM with 17% being newly diagnosed. Average blood glucose and glycated hemoglobin concentrations were 414 mg/dL and 13.1%, respectively. Most patients (88.1%) presented with hemichorea/hemiballism. Isolated putamen and combined putamen-caudate nucleus involvements were most common on neuroimaging studies with discrepancies between CT and MRI findings in about one-sixth of patients. Unilateral arm-leg combination was the most frequent with bilateral chorea in 9.7% of patients. Successful treatment rates of chorea with glucose-control-only and additional anti-chorea medications were seen in 25.7% and 76.2 % of cases respectively[9].

The diagnosis of diabetic striatopathy was eastablished with presence of high blood sugar on admission and the characterstic neuroimaging findings. In this case patient was not in a hyperosmolar hyperglycaemic state(HHS). HHS requires serum osmolality to be in the range of 330milliosmoles/kg and above with blood sugars ranging more than 600mg/dl. This patient had poorly controlled diabetes with nephropathy. Other conditions that can contribute to chorea like hyperthyroidism hypocalcemia, hypomagnesemia, lupus were ruled out. There was no family history of chorea and CAG repeats were negative. The neuroimaging ruled out the possibility of vascular

causes like infarction/ haemorrhage. To conclude diabetic striatopathy is a rare complication of poorly controlled diabetes. It can be presenting manifestation of a new onset diabetes also. A thorough knowledge about this condition can prevent misdiagnosis and delay in treatment.

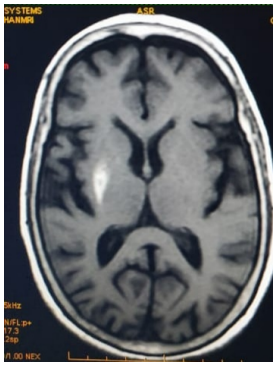

Image 1 MRI image of the patient showing T1 hyperintensity in right lentiform nucleus.

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