SHORT REPORTS

Chorea Due to Nonketotic Hyperglycemia

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Summary

A 62 year old diabetic and hypertensive male presented with sudden onset generalized chorea. Investigations revealed uncontrolled diabetes with absent ketones and normal serum osmolality. Achievement of euglycemia with insulin therapy abolished the involuntary movements completely within a day. The direct effect of hyperglycemia causing striatal neuronal dysfunction could be the pathogenesis of the chorea in our patient.

Key words: Chorea, Non-ketotic hyperglycemia.

Introduction

Chorea or ballismus can be caused by a wide variety of degenerative, metabolic or vascular disorders affecting the basal ganglia. Nonketotic hyperglycemia is, however, rare and usually presents with delirium, coma, focal or generalized seizures and focal neurological deficits. Hyperkinesia in the form of chorea, ballismus or choreoathetosis has been reported in patients with nonketotic hyperglycemia, with complete response to therapy for hyperglycemia.1,2 These involuntary movements may even be the presenting symptoms of the underlying diabetes mellitus.3

Case Report

A 62 year old man, was brought with history of sudden onset involuntary movements involving both sides of the body and face, increasing during activity and ceasing during sleep, of two days duration. There was no previous history of fever, headache or transient ischemic attacks. He was diabetic for eight years (on glibenclamide, 5 mg per day), hypertensive for five years (on enalapril, 5 mg and amlodipine 10 mg per day). He was non-smoker and non-alcoholic. Clinical examination revealed a well built man with normal memory, speech and orientation, having choreiform movements involving all the four limbs and grimacing movements of the face. There was hypotonia of muscles with normal power and deep tendon reflexes. His blood pressure was 140/86 mm Hg. Kleyser Fleisher ring was absent on slit lamp examination. Investigations revealed hemoglobin of 13 g%, total count of 11,800 /mm³, differential count of N 80 E1 L19; and normal liver function tests. His BUN and serum creatinine were 20 mg% and 0.8 mg% respectively. Blood glucose concentration was 367 mg%. Estimated blood osmolality was 294 mosm/L and ketones were absent. He had serum sodium of 132 mEq/L; potassium 4.2 mEq/L; serum calcium and magnesium levels were normal. Serum antistreptolysin-O titre was < 200 IU. CT of the head did not reveal any lesion in the basal ganglia.

Patient was started on regular insulin subcutaneous, hourly till his blood sugar was brought below 250 mg%, following which insulin – buffered glucose infusion was started and continued for a day. At the end of 24 hours, his blood sugar was 148 mg% and the involuntary movements had completely disappeared. Patient was continued on the antihypertensives mentioned earlier and his blood pressure remained under control. He remained asymptomatic during rest of his hospital stay and was discharged on a combination of regular and intermediate acting zinc insulin (subcutaneous) before breakfast and dinner.

Discussion

The pathogenesis of chorea or ballismus associated with nonketotic hyperglycemia is poorly understood. Several hypotheses put forward to explain these movement disorders associated with nonketotic hyperglycemia include, relative dopaminergic hypersensitivity, undefined effect of hypersomolality, decrease in aminobutyric acid and hypometabolism of striatal cells due to hypoperfusion. In nonketotic hyperglycemia, the shift to anerobic metabolism causes brain to utilise aminobutyric acid which is synthesized from acetoacetate. Unlike in ketoacidosis, acetoacetate is rapidly depleted in nonketotic hyperglycemia causing cellular dysfunction.3,4 In addition, striatum may be directly susceptible to alterations in blood glucose levels as shown by paroxysmal choreoathetosis in a patient with hypoglycemia.5 Chorea had been reported in patients of nonketotic hyperglycemia with high serum osmolality, which however was normal in our case. The CT scan of brain was normal in our case, as in earlier reports.3 Hyperintense lesions in the basal ganglia, on T1WI of MRI have been demonstrated in one study.6 Another study revealed hypoperfusion in

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corresponding areas on SPECT. Chang et al compared six nonketotic hyperglycemic chorea patients with 10 age matched controls, not having any structural lesion by MRI. The SPECT scan showed the presence of hypoperfusion in the striatum contralateral to the symptomatic chorea in five patients and hypoperfusion in both striatum in one. As SPECT scan could not be performed in the present case, hypoperfusion or structural lesion of basal ganglia could not be ruled out. Bilateral chorea at presentation, hyperglycemia with normal serum osmolality and rapid response of chorea to correction of hyperglycemia point towards the direct effect of hyperglycemia as the pathogenesis of chorea in this case.

Chorea should be considered potentially reversible when associated with nonketotic hyperglycemia, as rapid detection and early correction of hyperglycemia could lead to complete recovery of these involuntary movements in some cases.

References