Hemichorea-hemiballismus as the presenting manifestation of nonketotic hyperglycemia in an adolescent with undiagnosed type 2 diabetes mellitus

Sir,

Chorea, hemichorea-hemiballismus (HC-HB) and severe partial seizures may be the presenting features of nonketotic hyperglycemia (NKH) in older adults with type 2 diabetes, but cases in children with diabetes are rare. Here, we present a teenage boy who presented with hyperglycemia induced HC-HB.17-year-old boy presented with a history of sudden onset paroxysm of involuntary movement initially of the right hand and then the right leg and subsequently involving entire right side of the body, increasing during activity and ceasing during sleep, of 2–4 days duration. The patient described this as the feeling of stretching leg and cramps. There was no history of fever, headache, chronic neurological illness. He did not receive or intake of medication. He was a non-smoker and nonalcoholic. He had no family H/O of diabetes or seizure disorder. Clinical examination revealed a lean , dehydrated boy with pulse rate 88/min, respiratory rate 22/min, blood pressure 110/70 mm of Hg. and body mass index 18.5 kg/m^2. Neurological examination revealed normal mental status, normal speech, memory and hearing. Motor and sensory systems were almost normal except reduced tone in the right side with normal coordination and gait. The initial clinical diagnosis HC-HB was made.

This clinical symptom of spontaneous hemichorea was initially considered secondary to a post-streptococcal neurological disease, Syndenham's chorea.However, it was not associated with other clinical features of rheumatic fever (carditis, arthritis, erythema rheumatic nodule and neuropsychological feature (dysarthria and emotional disorder). Also, the choreiform movements are continuous in chorea of Syndenham, rather than paroxysmal as observed in the above case. The serum antistreptolysin O titer was <200 IU. The possibility of continuous focal seizure (epilepsia partialis continua) causing unilateral movement and CNS involvement (the contralateral basal ganglion, the thalamus and subcortical areas) was considered. As a result, electroencephalography and computed tomography (CT) were scheduled.

Ischemic and hemorrhagic strokes are the cause of chorea in most elderly patients and these etiologies were excluded in our patient based on the neuroimaging studies. Other differential diagnoses (systemic lupus erythematosus, Wilson's disease, thyroid disease, basal ganglia calcification) were ruled out. Antinuclear antibodies were negative, and serum ceruloplasmin and thyroxin levels were within normal range. Keyser Fleischer ring was absent on slit-lamp examination. CT of the head did not reveal any lesion in basal ganglia. EEG was normal even during the ictal recording excluding the possibility of epilepsy [Figures 1 and 2].

Other laboratory tests revealed Hb 13 g/dl, total lymphocyte count (TLC) 8000/mm^3 and normal liver function. His blood urea was 42.2 mg/dl and serum creatinine was 1.4 mg/dl. Urine exam showed glycosuria without ketones. Blood glucose concentration was 777 mg/dl and glycosylated hemoglobin A1c level was 13.3%. Estimated blood osmolality was 318 mosm/l and ketones were absent.

His serum sodium showed a level of 130 mEq/l and level of potassium was 5.5 mEq/l. His lipid profile was abnormal: cholesterol = 310 mg/dl, triglycerides = 372 mg/dl, low density lipoprotein (LDL) = 191.3 mg/dl.

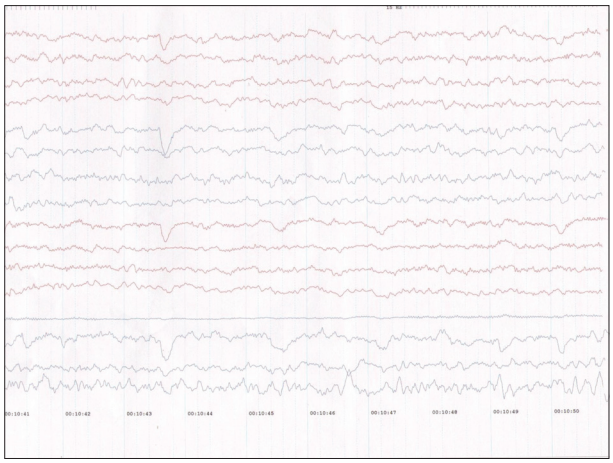

Figure 1: EEG recording during awake

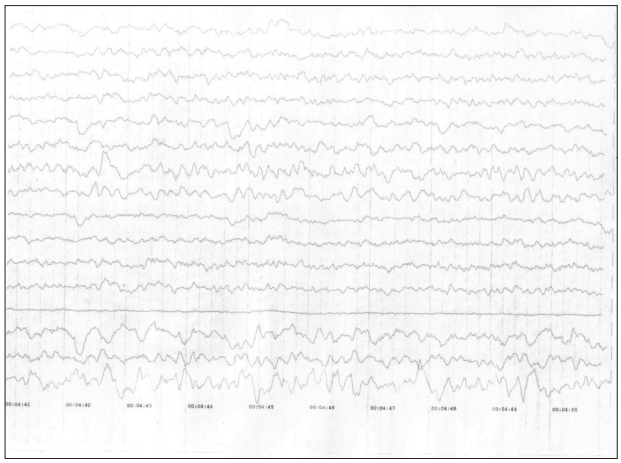

Figure 2: EEG recording during ictal

A diagnosis of diabetes was made and the patient was started on regular insulin 10 units IV stat, followed by 50 units in drip infusion, and normal saline for correction of dehydration and hyponatremia. By 24 hours, his blood glucose was 170 mg/dl and involuntary movements had completely disappeared. He remained asymptomatic after euglycemia was achieved and was discharged on premixed insulin 30:70 subcutaneously, which he tolerated well for a few months. After three months, he experienced recurrent episodes of hypoglycemia, and was shifted to glimepride 1 mg twice per day, which is continuing at time of writing.

Insulin antibody level by chemiluminescence-ADVIA CENTAUR and insulin C-peptide level were within normal limit (56 nmol/l), and thus, we diagnosed type 2 diabetes mellitus, presenting in NKH, severe enough to cause HC-HB.. HC-HB is a rare complication of nonketotic hyperglycemia: only 53 case reports of this particular condition were published between 1985 and 2001.[1] Most of cases were over 60 years of age and represented type 2 diabetes and nonketotic hyperglycemia. The HC-HB can be caused by degenerative changes, metabolic or focal lesions, such as ischemic or hemorrhagic stroke, infections, drugs, toxins, epilepsy and neoplasm as well as diffuse systemic process including systemic lupus erythematosus, Wilson's disease and thyrotoxicosis.[2]

The underlying pathophysiology leading to basal ganglia dysfunction is multifactorial. Many hypotheses have been reported for the development of diabetic HC-HB, such as local gamma aminobutyric acid starvation, disinhibition of dopaminergic neurons, local microhemorrhage, microinfarction and brain edema. During hyperglycemic crisis, the activity of tircarboxylic acid cycle (Krebs cycle) and glucose utilization are depressed in the brain, so cerebral metabolism shifts to alternative pathways. In ketoacidosis, ketones are used as an energy source and gamma-aminobutyric acid (GABA) can be resynthesized. As a result, HC-HB or partial seizures rarely occur with diabetic ketoacidosis. In NKH, the brain metabolizes GABA into succinic acid via the succinic acid semialdehyde pathway and thus depletes GABA rapidly.[3] Advanced imaging analysis reveals reduced cerebral glucose metabolism on positron emission tomography (PET) scans with concomitant hyperperfusion in affected basal ganglia seen on single photon emission computed tomography (SPECT).[4] In some cases, the basal ganglia in diabetic HC-HB are hyperdense without mass effect on CT scans and hyperintense on T_1-weighted magnetic resonance imaging (MRI), but these features reverse after therapy.[1] This evidence supports the idea that basal ganglia, generally weak in hyperglycemia stress, might induce involuntary movement and neurotransmitting functional disorder.

Severe hyperglycemia without ketosis at the clinical onset of diabetes mellitus type 1 has been reported in children and adolescents, but nonketotic hyperglycemia is an unusual cause of chorea ballismus in children, and also, chorea ballismus is a rare manifestation of primary diabetes.

On reviewing the literature,[5] and studying our patient who had no detectable neurological lesion in basal ganglia, it can be seen that the involuntary movements completely disappeared within 24 hours of insulin therapy. So, we suggest that the pathogenesis of hemichorea or hemiballismus was most probably due to hyperglycemia causing striatal neuronal dysfunction.

Since chorea ballismus can be life threatening, and chorea ballismus caused by hyperglycemia itself is a treatable disorder with good prognosis, recognition of this order is important..

Ibraheem Khan, Chandra Kant, Anil Sanwaria, Alok Goyal

Department of Medicine, Jawarhar Lal Nehru Medical College, Ajmer, Rajasthan, India

Corresponding Author: Dr. Ibraheem Khan, VPO-Samraya, Weir, Dist. Bharatpur, Rajasthan – 321 408, India. E-mail: imedicine2008@gmail.com

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