Case Report

DOI: 10.7860/JCDR/2017/27266.9939

Hemi-Chorea in a Patient with Ketotic Hyperglycemia: An Unusual Presentation

Internal Medicine Section

PENDELA VENKATA SATISH[1], KUDARAVALLI PUJITHA[2], NEHA AGRAWAL[3], THOMAS MATHEW[4], SUDHA VIDYASAGAR[5]

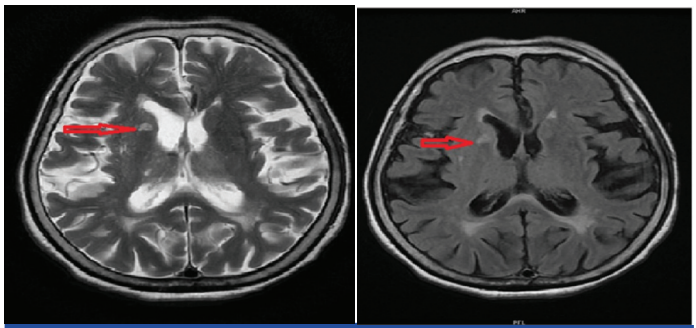
ABSTRACT

Chorea has often been associated with lesions in the basal ganglia and in the sub thalamic nucleus. It is possible for a patient with chorea-ballismus to have hyperglycemia at the initial presentation. We hereby present a case of an 81-year-old female, who was on treatment for type 2 diabetes mellitus and presented to us with sub acute onset of abnormal movements of right side of the body. She had semi purposeful, rapid and jerky movements of right upper limb and lower limb along with abnormal tongue movements. Laboratory data showed very high blood glucose levels, urine ketones were positive and pH of arterial blood was normal. MRI brain showed hyperintensities in right basal ganglia. So, hyperglycemia induced hemichorea was considered as a possibility and she was treated with insulin. These abnormal movements decreased subsequently with treatment and patient is doing better in the follow-up visits. This presentation is extremely rare, as review of literature showed similar presentations in patients with non ketotic hyperglycemia but not reported so far in diabetic ketosis.

Keywords: Choreo-athetoid movements, Diabetic ketosis, Type 2 diabetes mellitus

CASE REPORT

An 81-year-old female, who was a diagnosed case of type 2 diabetes mellitus presented to the OPD with history of involuntary movements of right upper and lower limbs since one month which had increased in severity over the past one week. She was on irregular oral anti diabetic medication (tablet metformin 500 mg BD, tablet glimepiride 2 mg OD). On examination her vitals were stable. Involuntary semi purposeful, rapid and jerky movements were noticed in right upper and lower limbs [Video-1]. Jack in box movements were noticed in relation to the tongue. Tone, power, reflexes, sensory system were normal. Cardiovascular, respiratory systems and abdominal examination did not reveal any abnormalities. Our potential differentials based on the presentation (subacute onset hemichorea in an elderly lady) included senile chorea, Huntington's chorea, late onset Wilson's disease, lacunar stroke/haemorrhage, hypo/hyperthyroid state, Sydenham's chorea, drug induced movement disorder, metabolic disorders (hyperglycemia, uremia, hypoparathyroidism). Her routine haemogram showed normal haemoglobin (13 gm/dl), platelet count (1.77 lakh/µl) and total leukocyte count (7200/µl). Random blood sugar was 400 mg/dl with HbA1c value of 17.4%. Urine ketones were 40 mg/dl. Arterial blood pH was 7.39 and electrolytes were normal (potassium: 4.3 mmol/l, sodium: 135 mmol/l, calcium: 9 mmol/l). Serum urea and creatinine were normal. Prothrombin time of 13.4 secs and aPTT 26 secs were noted. TSH was within normal limits (3.5 uU/ml) and MRI brain showed hyperintensities in right basal ganglia [Table/Fig-1,2]). Based on the examination findings and laboratory values the diagnosis were narrowed down to hyperglycemia induced hemichorea. She was initially treated with intravenous insulin infusion till her blood sugar was near normalised and then switched over to subcutaneous insulin with which she improved. Patient improved with the treatment dramatically and she was discharged with subcutaneous insulin regimen. This relief of symptoms further confirmed about the diagnosis of hyperglycemia induced hemichorea. During subsequent follow up visit after one month, minimal choreo-athetoid movements were noticed for which she was started on low dose haloperidol (2 mg OD). Movements decreased by next visit and haloperidol was tapered and stopped.



[Table/Fig-1]: T2 MRI brain axial section showing hyperintensity in right basal ganglia;
[Table/Fig-2]: T1 MRI brain axial section showing hyperintensity in right basal ganglia.

DISCUSSION

Chorea may occur due to causes affecting the basal ganglia like hypoxia, metabolic abnormalities, immune mediated disorders, infections, drugs etc. Rarer causes of chorea include essential thrombocythemia, uncontrolled diabetes mellitus, pregnancy, etc. Hyperglycemia can be a cause for the first presentation of hemichorea-ballismus [1,2]. It may also be associated with poorly controlled diabetes and old age [3]. Cases have been reported where patients presented with choreo-athetotic movements, were found to have non ketotic hyperglycemia [4]. Most of the patients present with insidious onset, involuntary movements which slowly progress over few days to weeks with uncontrolled blood sugars, increased serum osmolality with absent ketones in serum and urine. In few patients chorea starts few days after uncontrolled sugars are noted and even after achieving adequate control. The exact mechanism of how this uncontrolled sugars cause involuntary movements is still an unsolved mystery. According to one hypothesis Gamma Aminobutyric Acid (GABA) which is the inhibitory neurotransmitter in the central nervous system is depleted as it is used up by the brain as an energy source because of lack of insulin [5]. The lack of GABA may result in hemichorea and hemiballismus. Another hypothesis is transient focal cerebral ischemia due to hyperglycemia and hypoperfusion [6]. Biopsy reports demonstrate hypertrophic astrocytes to be the main abnormal finding within the basal ganglia [7]. The characteristic radiographic finding in non ketotic hyperglycemia patients with chorea is a high signal intensity basal ganglia lesion

on the T1-weighted brain MRI [8]. These MRI changes may be attributed to the protein desiccation and wallerian degeneration occurring in the basal ganglia neurons due to hypoperfusion. Thus, hyperglycemia in type 2 diabetes mellitus should be considered as one of the important causes of involuntary movements (hemichorea or hemiballismus) in elderly females with poor glycemic control [9-11]. It is better to get a detailed imaging of brain with T1, T2, diffusion weighted and Apparent Diffusion Coefficient (ADC) series MRI, to look for ischemia or perfusion defect or any bleeding, before concluding on the pathology as treatment options may vary accordingly. Literature has shown similar movements in patients with non ketotic hyperglycemia [2,6], but, so far no case has been reported in a patient with ketotic hyperglycemia. Why these movements persist in some patients despite adequate glycemic control is still unclear. Neuronal loss from ischemia and putaminal micro hemorrhage are possible causes.

CONCLUSION

Ketotic hyperglycemia can also result in hemichorea in diabetic patients. The abnormal movements resolved completely with control of the blood glucose level. The hypotheses explaining the pathophysiology of chorea in hyperglycemia have not involved ketone bodies and the occurrence of chorea in both ketotic and non ketotic hyperglycemia lends support to this.

Video-1: The pattern of involuntary movements was mainly continuous, arrhythmic, purposeless, and dancing like movements that were confined to right upper limb(mainly) and lower limb. The involuntary movements attenuated when she was relaxed, and it disappeared during her sleep.

REFERENCES

[1] Ifergane G, Masalha R, Herishanu YO. Transient hemichorea/hemiballismus associated with new onset hyperglycemia. Can J Neurol Sci. 2001;28(4):365-68.
[2] Milburn-McNulty P, Michael BD, Woodford HJ, Nicolson A. Hyperosmolar non-ketotic hyperglycaemia: an important and reversible cause of acute bilateral ballismus. BMJ Case Rep. 2012; 2012: bcr1120115084.
[3] Suto Y, Mori M, Kagimoto H, Saito J. A case of hemichorea with hyperglycemia presenting with low signal intensity in the striatum on T2*-weighted gradient-echo magnetic resonance imaging. RinshoShinkeigaku. 2004;44(2):86-90.
[4] William G. Rector Jr, H. Franklin Herlong, Hamilton Moses III. Nonketotic hyperglycemia appearing as choreoathetosis or ballism. Arch Intern Med. 1982;142:154-55.
[5] SF Sung, CH Lu. Focal neurological symptoms as the presenting manifestations of nonketotic hyperglycemia: report of two cases. J Intern Med Taiwan. 2007;18:206-11.
[6] Duckrow RB, Beard DC, Brennan RW. Regional cerebral blood flow decreases during hyperglycemia. Ann Neurol. 1985;17(3):267-72.
[7] Shan DE, Ho DM, Chang C, Pan HC, Teng MM. Hemichorea-hemiballism: an explanation for MR signal changes. AJNR Am J Neuroradiol. 1998;19:863-70.
[8] Nagai C, Kato T, Katagiri T, Sasaki H. Hyperintense putamen on T1-weighted MR images in a case of chorea with hyperglycemia. AJNR Am J Neuroradiol. 1995;16(6):1243-46.
[9] Chang CV, Felicio AC, Godeiro Cde O Jr, Matsubara LS, Duarte DR, Ferraz HB, et al. Chorea-ballism as a manifestation of decompensated type 2 diabetes mellitus. Am J Med Sci. 2007;333(3):175-77.
[10] Felicio AC, Chang CV, Godeiro-Junior C, Okoshi MP, Ferraz HB. Hemichorea-hemiballism as the first presentation of type 2 diabetes mellitus. Arquivos de Neuro-Psiquiatria A. 2008;66(2):249-50.
[11] Lee SH, Shin JA, Kim JH, Son JW, Lee KW, Ko SH, et al. Chorea-ballism associated with nonketotic hyperglycaemia or diabetic ketoacidosis: characteristics of 25 patients in Korea. Diabetes Res Clin Pract. 2011;93(2):e80-83.

PARTICULARS OF CONTRIBUTORS:
1. Junior Resident, Department of Medicine, K.M.C., Manipal, Karnataka, India.
2. House Surgeon, Department of Medicine, K.M.C., Manipal, Karnataka, India.
3. Junior Resident, Department of Medicine, K.M.C., Manipal, Karnataka, India.
4. Junior Resident, Department of Medicine, K.M.C., Manipal, Karnataka, India.
5. Professor and Head, Department of Medicine, K.M.C., Manipal, Karnataka, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Pendela Venkata Satish,
Ajay Arcade, Manipal-576104, Karnataka, India.
E-mail: pvsatish.135@gmail.com

FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Feb 07, 2017**
Date of Peer Review: **Feb 22, 2017**
Date of Acceptance: **Mar 30, 2017**
Date of Publishing: **May 01, 2017**