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HEMICHOREA ASSOCIATED WITH TYPE 2 DIABETES: A RARE NEUROLOGICAL COMPLICATION.

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ABSTRACT:

Diabetic hemichorea/hemiballism is a spectrum of hyper kinetic, involuntary, irregular, purposeless, non-rhythmic, rapid and unsustained movements flowing from one part of the body to another. It involves contra lateral basal-ganglia and often striatum of the brain. Here we are reporting an un-usual case of choreiform movement disorder which was sudden in onset. It was accompanied with abnormally high values of blood glucose. Our patient had a complete remission of symptoms after an adequate control of blood glucose was achieved. This case illustrates the importance and rarity of hyperglycemia as a rare cause of hemichorea. It recovers rapidly and has a good prognosis. Screening for hyperglycemia even in those patients without a prior history of diabetes is very important, once they present with an involuntary movement disorder. Recognition and early treatment is beneficial to prevent adverse outcomes. Today, in the medical literature it is often referred to as C-H-BG (chorea, hyperglycemia, basal ganglia) syndrome.

KEY WORDS: diabetic hemichorea, choreiform movement disorder, C-H-BG syndrome

INTRODUCTION:

Hyper osmolar non ketotic syndrome is a clinical syndrome of hyperglycemia, hyperosmolarity and dehydration without ketoacidosis . Hemichorea is an involuntary, irregular and non rhythmic movement involving one side of the body due to lesions found on the contralateral striatum [1]. Ballism is non rhythmic, varying, large-amplitude involuntary movement predominantly of the proximal extremities. Hemiballism is involuntary flailing type of movements of one side of the body resulting from lesions on the contra lateral sub-thalamic nucleus. Therefore, hemichorea-hemiballism (HCHB) is a clinical spectrum of involuntary movements that involve one side of the body. According to a meta-analysis of 53 such cases mean age was found to be 71.1 years (range = 22 –92 years) and all patients developed C-H-BG over the age of 40. The mean of time interval between the onset of chorea and admission was 18.7 days (range = 0– 180 days). Interestingly, 48 patients (91%) were Asians with female predominance [1]. Also,the mean serum glucose level upon admission was 481.5 and abnormally high serum HbA1c levels with mean of 14.4 (range = 9.9 –19.2%) [1].

The possible causes consist of ischemic / hemorrhagic stroke, CNS neoplasm, systemic lupus erythematosus (SLE), HONK, Wilson's disease and thyrotoxicosis [2]. Recent epidemiology suggests that elderly Asian females are more affected which implicates an underlying genetic predisposition [3]. Females being more affected could be related to post menopausal estrogen induced alterations of mainly two neurotransmitters: GABA or dopamine [4].

The pathophysiology is not fully understood. One of the main neurotransmitter in the basal gangli is called GABA. Hyperglycemia hampers cerebral auto regulation resulting in anaerobic metabolism leading to depletion of GABA transmitters in the basal gangli [5,6]. Hyperviscosity due to hyperglycemia disrupts blood-brain-barrier which leads to vascular compromise of the striatal- neurons [7]. This produces a synergistic effect of uncontrolled hyperglycemia and vascular destitution causing dysfunction of the striatum leading to irregular movements [8]. Histopathology findings in such patients has shown gliosis and neuronal loss without evidence of infarction or hemorrhage at striatum [8].

CASE PRESENTATION:

A man in his late 50's with type 2 diabetes for more than 10 years presented with two days history of involuntary and sudden onset hyperkinetic, irregular, purposeless, non-rhythmic, rapid and unsustained movements of left side of the body. Previously he didn't have any history of CVA or trauma. He had no significant drug addiction or family history of any disease. Huntington's Disease was not taken into account since his age didn't fit the usual age of the disease and there was no family history either. He was conscious and alert with normal higher mental functions. Cranial nerves were normal. His pupils were 3mm bilaterally reactive to light. No motor or sensory deficits were found. No loss of power, muscle strength or tremors were observed. Babinski sign was negative. Cerebellar signs were intact.

His vital signs were normal. Initial bio-chemistry showed a random serum glucose of 544mg/dL, a venous pH of 7.38. Urinary ketones were negative. Serum ketones were 0.4mmol/L (range 0.1 - 0.6mmol/L). Serum potassium 3.6mg/dL while serum sodium was 138mg/dL. Patient's HbA1c on last OPD visit was 12.1%. Hepatic profile was normal. Anti dsDNA antibodies were 2 IU/ml (range 104 IU/ml) ANA and ENA profiles were negative. Serum TSH was 1.9 ng/dL (range 1.8-3.0 ng/dL) while FT4 was 1.2 ng/dL (range 1.0–1.53 ng/dL). Echocardiogram showed no valvular abnormality with an ejection fraction of 65%. Extended work up for Wilson's disease was done. Slit-lamp examination of the eyes showed no Kayser–Fleischer rings. Serum ceruloplasmin levels were 28 mg/dL (range 20-35 mg/dL). 24 hour urinary free copper levels were 15 ug/24 hours (range 10-30 ug). Computed tomography (CT) of the brain revealed no evidence of any acute intra cranial pathology (Figure A). Stroke was excluded as there was no focal neurological deficit except involuntary movements. MRI Brain showed a T1 hyper intensity (asymmetric) of the right putamen (Figure B).During hospitalization Strict blood glucose monitoring was initiated. Insulin was started on sliding scale and levels were maintained between 80 to 120 mg/dL. Oral administration of Haloperidol 0.5mg twice daily was started which showed no response. Dose was gradually increased to 1mg twice a day. By the 8th day, his un-controlled and in-voluntary movements were alleviated. He was discharged on 10th hospital day on haloperidol 1mg twice daily with a strict insulin regimen. During his follow up after 2 months his chorei-form movements had completely resolved. His HbA1c of 8.5% showed an adequate diabetic control.

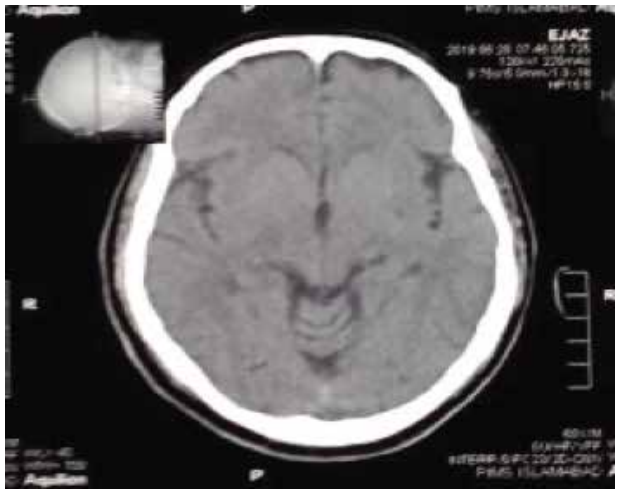

Figure A: Normal CT Brain

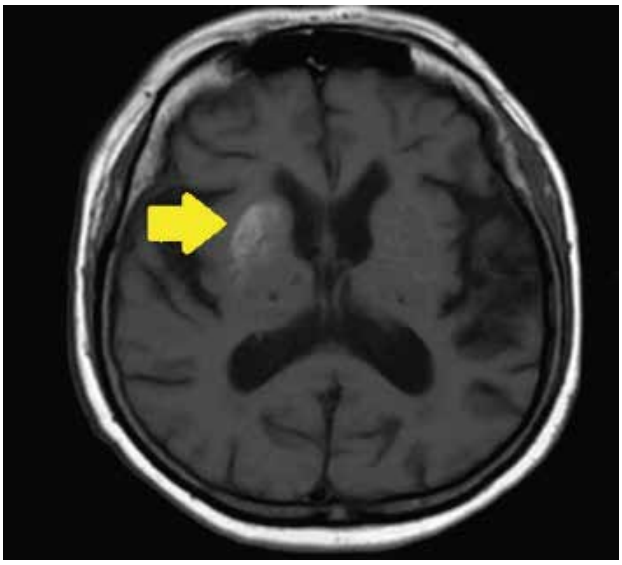

Figure B: MRI brain showing T1 hyperintensity of right putamen

DISCUSSION:

Hemichorea/hemiballismus may occur in patients already known to be diabetics [9] or it may be the first manifestation of diabetes [10]. Neuroradiological findings are consistent with hemi chorea and a nonketotic hyperglycemic state. A T1 weighted hyper-intensity is seen in the striatum/globus pallidus on MRI brain. Restricted diffusion maybe seen on diffusion weighted image (DWI) [11]. Magnetic resonance spectroscopy may reveal low levels of N-acetyl-aspartate to the ratio of creatinine. There is a high choline ratio and often showing lactate peaks [12].

Hemi chorea resolves once the serum glucose levels get under control and can be treated actively [13]. Literature does not describe a constant temporal association between the two, since chorea may or may not resolve after control of blood glucose. It may also appear after hypoglycemia, or even after rapid correction of hyperglycemia in some[14]. The mainstay of treatment is aggressive glucose control for resolution of abnormal movements [15]. There is an improvement in radiological signs on MRI brain that usually takes 6 months to go away once glycemic levels are controlled [16]. In a few refractory cases an additional medication like haloperidol, tetrabenazine or resperidone have been documented to be fruitful [17]. This rare phenomena deserves attention because it is rapidly improved with a good prognosis. Screening for hyperglycemia even in those patients without a prior history of diabetes is very important, once they present with an involuntary movement disorder. Recognition and treatment is beneficial to prevent adverse outcomes.

REFERENCES:

1. Oh SH, Lee KY, Im JH, Lee MS. Chorea associated with non-ketotic hyperglycemia and hyperintensity basal ganglia lesion on T1-weighted brain MRI study: a meta-analysis of 53 cases including four present cases. J Neurol Sci. 2002;200:57–62.

2. Padmanabhan S, Zagami AS, Poynten AM. A case of hemichorea-hemiballis¬mus due to nonketotic hyperglycemia. Diabetes Care. 2013;36: e55–e56.

3. Mittal P: Hemichorea-hemiballism syndrome: a look through susceptibili¬ty weighted imaging. Ann Indian Acad Neurol. 2011;14:124–26.

4. Battisi C, Forte F, Rubenni E et al: Two cases of hemichorea-hemiballism with nonketotic hyperglycemia. a new point of view. Neurol Sci. 2009;30:179–83.

5. Nath J, Jambherkar K, Rao C, Armitano E: Radio logical and pathological changes in hemibal lism-hemichorea with striatal hyperintensity. J Magn Reson Imaging. 2006;23: 564–68.

6. Cheema H, Federman D, Kam A: Hemicho rea-hemiballismus in non-ketot¬ic hyperglycae mia. J Clin Neurosci. 2011;18:293–94.

7. Slabu H, Savedia-Cayabyab S, Senior P, Arnason T: Permanent haemicho¬rea associated with transient hyperglycemia. BMJ Case Rep. 2011; 2011.

8. Lin JJ: Ipsilateral putamen hyperintensity on T1-weighted MRI in non-ketotic hyperglycemia with hemiballism-hemichorea: a case report. Parkinsonism Relat Disord. 2001;7:319–21.

9. hyperglycaemic chorea in diabetic patients. Diabet Med. 2004;21:196–198.

10. Shan DE, Ho DM, Chang C, Pan HC, Teng MM. Hemichorea-hemiballism: an explanation for MR signal changes. Am J Neuroradiol. 1998;19:863–870.

11. Zaitout Z: CT and MRI findings in the basal ganglia in non-ketotic hyperglycaemia associated hemichorea and hemi-ballismus (HC-HB). Neuroradiol. 2012;54:1119–20.

12. Narayanan S: Hyperglycemia-induced hemiballis mus hemichorea: a case report and brief review of the literature. J Emerg Med. 2012;43:442–44.

13. Taboada GF, Lima GA, Castro JE, Liberato B: Dyskinesia associated with hy¬perglycemia and basal ganglia hyperintensity: report of a rare diabetic com¬plication. Metab Brain Dis. 2013;28:107–10.

14. Lucassen EB, Delfyett WT, Stahl MC. Persistent hemichorea and caudate atrophy in untreated diabetic striatopathy: A case report. Case Rep Neurol. 2017;9:299–303.

15. Hwang KJ, Hong IK, Ahn TB et al: Cortical hemichorea-hemiballism. J Neurol. 2013; 260: 2986–92.

16. Shan DE, Ho DM, Chang C, Pan HC, Teng MM. Hemichorea-hemiballism: an explanation for MR signal changes. Am J Neuroradiol. 1998;19:863–870.

17. Battisti C, Forte F, Rubenni E, et al. Two cases of hemichorea–hemiballism with nonketotic hypergly caemia: a new point of view. Neurol Sci. 2009;30:179–183

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Muhammad Hassan; concept, data collection, data analysis, manuscript writing, manuscript review
Haris Majid Rajput; data analysis, manuscript writing, manuscript review
Mazhar Badshah; data analysis, manuscript review