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Medicine

CASE SERIES: CHOREA HYPERGLYCEMIA BASAL GANGLIA SYNDROME

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ABSTRACT Chorea Hyperglycemia Basal Ganglia (CHBG) syndrome is a rare condition that manifest within the setting of uncontrolled diabetes mellitus. The objective of this case series is to present patients with this syndrome and provide information about case work up and treatment. We are also presenting review about current understanding of the pathophysiology and treatment of this syndrome and how it was applied to our patients. This case series involves 3 cases, two females and one male with history of poorly controlled diabetes mellitus who presented to our emergency with complaints of uncontrolled involuntary movements in limbs. All three patients have high blood glucose levels on presentation and on further work up all have Hb1AC more than 11%. There CT head demonstrated abnormal increased intensity within lenticular nucleus and caudate head. Their MRI brain demonstrated non specific T1 and T2 hyper intensity and corpus striatum predominantly in putaman and globus pallidus. These findings were consistent with the movement pattern the patients were displaying and with the diagnosis of CHBG. Gradual control of blood sugar levels over 48 hours led to resolution of chorieform movements. Early recognition and gradual treatment of elevated blood sugar levels appears to lead to total resolution choreiform symptoms.

KEYWORDS : Chorea Hyperglycemia Basal GangliaSyndrome,Putamen,Globus pallidus

INTRODUCTION

Chorea Hyperglycemia Basal Ganglia syndrome is a rare condition, which manifests in setting of uncontrolled nonketotic diabetes mellitus. It is best characterised by the manifestation of hemichorea – hemiballism with uncontrolled blood sugar levels. Not much is known currently regarding pathogenesis of this condition, but a detailed literature review revealed various proposed theories. Most of these theories are regarding concept that hyperglycaemia may impair the cerebral auto regulatory mechanism of the CNS. This can lead to hypoperfusion and then leading to activation of anaerobic metabolism[1].This will lead to the creation of GABA within basal ganglia neurons[1]. GABA and Acetate depletes rapidly in nonketotic hyperglycemia which leads to reduction in acetylcholine synthesis[2]. The hyperviscosity caused by hyperglycemia then causes disruption of blood brain barrier and result into transient ischemia of striatal neurons[3]. The synergistic effect of uncontrolled hyperglycemia and vascular insufficiency are believed to cause incomplete stratum neuronal dysfunction which leads to hemichorea- hemiballism[4]. Extensive literature review about this syndrome also revealed that few histologically findings have also been reported in cases of CHBG syndrome, these are characterised by selective neuronal loss, gliosis and reactive astrocytes without any evidence of haemorrhage or infarction[4]. There are limited number of case reports regarding CHBG. Few report suggests that there are patients who present with acute onset of chorea in setting of well controlled sugar with elevated Hb1AC. We present a case series of three patients who presented in our casualty with acute onset chorea with history of poorly controlled diabetes mellitus due to non-compliance to medications which was eventually determined to be CHBG.

CASE:1

A 52-year-old normotensive diabetic male presented with two days history of involuntary movements in both upper limbs. Initially he noticed that his both upper limbs being fidgety which gradually progresses in intensity to wide flinging movements. Patient was known case of type 2 diabetes mellitus for past 12 years and he was non compliant to his medications since last two months. reviewing his previous records, they were suggestive of various random blood sugar readings ranging from 380 to 460.His BMI was 28.2 kg/m^2, his prescriptions were suggestive of regimen of insulin lantus with short acting insulin daily patient denied any recent changes in his medications or any illness or headache. On day of presentation, patients random blood sugar was 542mg/dl. On day off admission his sample for Hb1AC was sent which came out to be 11.2%, his vitals were normal upon presentation and on physical examination patient was having choreiform movement in both upper limbs. Patient was fully conscious and oriented to time place and person, with no other signicant abnormality in neurological examination. urine for ketones was negative and ABG was also normal. our differential diagnosis included neoplastic disorder (metastatic), ischaemic/hemorrhagic stroke, trauma, et cetera. Patients NCCT head was done in emergency which was suggestive of hyperintensity in bilateral lenticular nucleus and caudate head(figure:1) subsequent MRI brain was done on next day of admission which was suggestive of non-specific T1 and T2 hyperintensity in right corpus striatum predominantly in putamen and globus pallidus (figure:2). All of these changes were consistent with a diagnosis of CHBG. Complete hemogram, serum electrolytes including serum sodium, potassium, calcium, KFT, LFT were normal, viral serology for HIV hepatitis B hepatitis C, liver function test, lipid profile, thyroid function function test were also normal. Patient was started on short acting insulin infusion to control blood sugars along with the tablet haloperidol to reduce the symptoms of patient. After 48 hours of admission with reduction of blood sugar levels, patient symptoms were also controlled

CASE: 2

96-year-old normotensive woman, Known case of type 2 diabetes mellitus for past 18 years, presented with history of involuntary movement and right arm. As described by patient and her son, her right arm began jerking uncontrollably While she was at home. Prior to this development her son revealed that she's diabetic and not taking any medications for past 20 to 25 days and she has been irregular in taking her medication since past three to four months. The patient had no history of weakness of the limbs, altered sensorium, fever or trauma and these involuntary movements resolved during her sleep. Her random blood sugar level was 432 mg/dl. Her ABG was done there was no acidosis/alkalosis. A sample was sent for blood investigations including complete hemogram with various electrolytes LFT and KFT and urine sample was sent for ketone bodies and all reports were within normal limits her Hb1AC was 12.2%. Her BMI was 24.4 kg/m^2. Patient's NCCT was done and it was also suggestive of bilateral hyper intensities in lenticular nucleus and caudate head following which next day MRI brain was done which was also suggestive of non specific T1 and T2 hyper intensities in B/L corpus striatum and predominantly in putaman and corpus striatum. All these changes were consistent with the diagnosis of CHBG. Other conditions that have been known to

cause hyper intensity emerging abnormality on MRI includes carbon monoxide toxicity, methanol toxicity and cyanide toxicity. Hyperammonemia from chronic liver disease and leih disease are also known to cause this. But our patient did not have any history that would relate to these possibilities. This patient was also started on short acting insulin infusion to control their blood sugar levels and along with Tablet haloperidol to reduce symptoms of patient after 48 hours of admission patients blood sugar levels were controlled and her symptoms were also controlled.

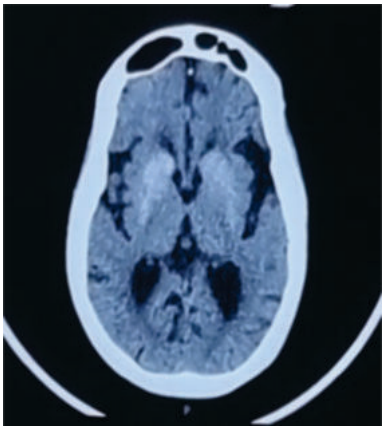
CASE:3

A 64-year-old diabetic lady with history of hypertension presented with one week history of involuntary movements in left arm and leg. Initially she noticed her left upper limb having involuntary movements which gradually progressed and intensified. Patient was known case of type 2 diabetes mellitus for past eight years Patient was on oral hypoglycemic agents regiment along with long acting insulin Lentus. Patient was also a known case of hypertension for past five years and was on oral drugs on presentation her blood pressure was 126/76mm of hg. Her random blood sugar level was 536mg/dl, her previous records were also suggestive of high random blood sugars ranging from 316 to 520mg/dl. On examination patient's vitals were normal and she was having chorieform movements in left upper limb and lower limb but patient was fully conscious and oriented to time place and person. The patient had normal sensorium and her higher mental functions were normal. complete cranial nerve examination was performed and found to be normal. patient also observed to have normal muscle tone, strength and deep tendon reflexes in all four limbs our sensory system examination an babinski sign were also normal.

Initial biochemistry revealed a random blood sugar of 536mg/dl, normal arterial pH with negative urine ketones. Complete blood counts and serum electrolytes along with KFT and LFT were also normal, patients viral serologies for HIV with hepatitis B and C along with lipid profile and thyroid function tests were also normal. patients NCCT head was also suggestive of hyper intensity in bilateral lenticular nucleus and caudate head. Next day her MRI brain was done which was suggestive of non specificT1 and T2 hyper intensity in bilateral basal ganglia. These lesions were characteristics of CHBG. Subsequently the patient was started on regular human insulin infusion along with tablet haloperidol patient symptoms were controlled within 36 hours with control of the patients blood sugar.

DISCUSSION AND CONCLUSION

CHBG is a rare entity and deserves awareness in light of fact that it is remaining underdiagnosed. Most of the patients who present with CHBG are of older age, female sex and Asian origin[5]. CHBG has also been described to more commonly involve upper limbs as compared to lower limbs[6]. Based on literature available patient with CHBG syndrome have benign clinical course that can be managed medically. The onset usually coincides with hyperglycaemia with relation between improvement of symptoms with control of blood sugar[6]. Thus mainstay of treatment appears to be gradual glycemic control leading to either partial or complete resolution of hemichorea/hemiballism[7]. As per literature available there is no clear data on how quick blood control should be achieved in CHBG. A gradual improvement in blood sugar seems to be appropriate and leads to improvement of symptoms. Clinical and radiological signs have been reported to take around six months to resolve after correction of hyperglycemia[7].



M.Figure:1

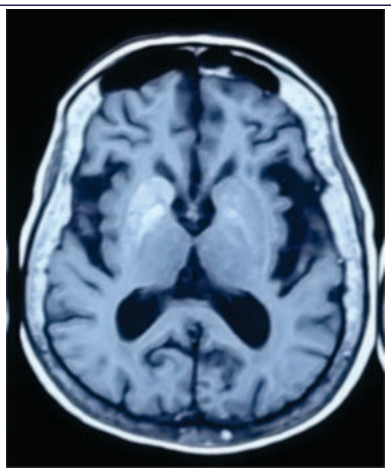


Figure :2

REFERENCES

1. J. Nath, K. Jambhekar, C. Rao, and E. Armitano, "Radiological and pathological changes in hemiballism-hemichorea with striatal hyperintensity," Journal of Magnetic Resonance Imaging, vol. 23, no. 4, pp. 564–568, 2006.
2. H. Cheema, D. Federman, and A. Kam, "Hemichoreahemiballismus in non-ketotic hyperglycaemia," Journal of Clinical Neuroscience, vol. 18, no. 2, pp. 293-294, 2011.
3. H. Slabu, S. Savedia-Cayabyab, P. Senior, and T. Arnason, "Permanent haemichorea associated with transient hyperglycemia," BMJ Case Reports, vol. 2011, 2011.
4. J. J. Lin, "Ipsilateral putamen hyperintensity on T1-weighted MRI in non-ketotic hyperglycemia with hemiballism– hemichorea: a case report," Parkinsonism & Related Disorders, vol. 7, no. 4, pp. 319–321, 2001.
5. G. P. Paraskevas, G. S. Vlachos, S. Vassilopoulou, E. Anagnostou, K. Spengos, and V. Zis, "Hypoglycemiainduced hemichorea in a patient with Fahr's syndrome," Neurological Sciences, vol. 33, no. 6, pp. 1397–1399, 2012.
6. J. Bizet, C. J. Cooper, R. Quansah, E. Rodriguez, M. Teleb, and G. T. Hernandez, "Chorea, hyperglycemia, basal ganglia syndrome (C-H-BG) in an uncontrolled diabetic patient with normal glucose levels on presentation," American Journal of Case Reports, vol. 15, pp. 143–146, 2014.
7. K. J. Hwang, I. K. Hong, T. B. Ahn, S. H. Yi, D. Lee, and D. Y. Kim, "Cortical hemichorea-hemiballism," Journal of Neurology, vol. 260, no. 12, pp. 2986–2992, 2013.