CASE REPORT

Chorea Hyperglycaemia Basal Ganglia Syndrome in an Uncontrolled Diabetic Patient

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ABSTRACT

Chorea hyperglycaemia basal ganglia (CHBG) syndrome is a rare neurological manifestation that occurs in patients with uncontrolled type 2 diabetes, rarely can be manifested in type 1 diabetes as well. Also called as diabetic striatopathy (DS), it occurs in the background of non ketotic hyperglycaemia. DS predominantly leads to a unilateral reversible injury of the basal ganglia visualised as hyperintensities on MRI brain, resulting in various movement disorders such as hemichorea and hemiballismus. Being a rare yet reversible entity, its recognition and early treatment by the clinicians is important. Here, we report a case of 79 yr. old gentleman with CHBG syndrome, whose symptoms resolved with adequate glycaemic control.

KEY WORDS: Diabetes, Chorea, Hemiballismus.

Introduction

Neurological manifestations of uncontrolled blood sugars in diabetic patients, like peripheral neuropathy and stroke are well-known. However, there is another rare neurological complication of poor glycaemic control termed as "Chorea-hyperglycaemia basal ganglia syndrome".

When a patient presents with symptoms such as chorea and hemiballismus, the differential diagnosis usually include haemorrhagic or ischemic stroke, space occupying lesion, neoplasm, systemic lupus erythematosus, Wilson's disease or thyrotoxicosis[1]. CHBG is usually not a top consideration. However, we need to have a suspicion of CHBG syndrome in an uncontrolled diabetic patient. Brain is susceptible to injury by various metabolic disorders, one of them being uncontrolled blood sugar levels which affect the basal ganglia leading to involuntary movements.[2] Chorea Hyperglycaemia Basal Ganglia syndrome (CHBG) is a rare condition that occurs in uncontrolled Type 2 diabetes mellitus and characterised by hemichorea/hemiballismus with non-ketotic hyperglycaemia[3]. The main importance of identifying this cause is, it is reversible with adequate glycaemic control. The mechanisms known to cause this rare treatable cause are hyperglycemia and hyperviscosity disrupting the blood brain barrier causing metabolic damage and a non ketotic state leading to depletion of neuronal GABA[2].

Hereby, we report a case of a 79yr old gentleman who presented with involuntary movements of left upper limb and lower limb and was diagnosed as CHBG syndrome.

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Doi: 10.46347/jmsh.v9i3.22.542

Case Report

A 79-year-old male, known diabetic presented with complaints of involuntary movements of left upper limb and lower limb since 5 days. Initially, he noticed that his left hand was fussy which gradually progressed in intensity to wide flinging movements. These movements which started in the upper limb, over the next 1-2 days involved the left lower limb with, similar brief and jerky movements. There was no history of fever, joint pain, and no history of cardiac illness. There was no history of altered behaviour or any psychiatric illness.

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The family gave history that patient was not compliant with the diabetic medications.

General physical examination revealed acanthosis nigricans, vitals were within normal limits. Neurological examination revealed normal higher mental functions, normal tone, power of 5/5 in all 4 limbs and normal deep tendon and superficial reflexes. There were brief involuntary jerky movements involving left upper limb and left lower limb. Left sided hemichorea with ballistic component was noticed. Investigations done revealed blood sugar of 379mg/dl with no acidosis or ketosis. HbA1c was 9.9%. Urine routine showed no ketone bodies. Other investigations revealed a normal hemogram, renal and liver parameters, with normal thyroid function tests. HIV testing done was negative. Fundus examination revealed moderate NPDR bilaterally.

MRI Brain T1 image revealed hyperintense abnormality in the right basal ganglia. These findings were consistent with CHBG syndrome. Gradual control of his blood sugar levels was achieved using basal bolus insulin regimen. Over a period of 6 to 7 days, there was significant improvement in his symptoms with a basal bolus insulin regimen using Insulin Glargine 24Units at 10pm daily along with a regular insulin of 14Units thrice daily for postprandial control of blood sugar levels. However, as the movements were still disabling and were not completely controlled despite adequate glycemic control after 1 week of insulin treatment, antichorea medications were prescribed. He was discharged upon improvement of symptoms and adviced to be compliant with medications. On follow up after 1 months, there was no recurrence of his symptoms and antichorea medications were tapered and stopped. After 3 months, HbA1c was 7 %, and no recurrence of symptoms.

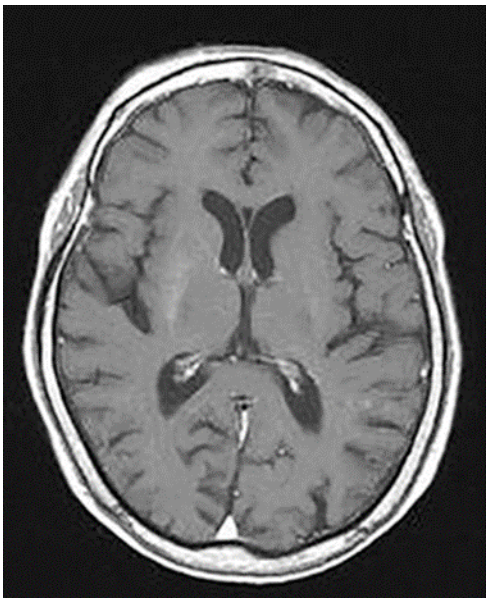

Figure 1: T1 weighted MRI Brain showing hyperintensity in right basal ganglia

Discussion

CHBG warrants awareness as a complication of long standing poorly controlled diabetes mellitus, usually occurring in type 2 diabetes mellitus, with few case reports in type 1 diabetes patients also described[1,2].Most of the available case reports are from Asian population and elderly females which probably indicates a genetic predisposition[3]. It is usually reported in elderly females with hyperglycaemic hyperosmolar state and rare in the setting of diabetic ketoacidosis[4]. The increased sensitivity of dopaminergic neurons in postmenopausal females could explain the increased female preponderance.

Chorea, which is one of the dyskinesia, is characterized by jerky, involuntary and irregular movements. When similar movements occur on one side of the body, it is hemichorea[1].

Hemichorea is a hyperkinetic movement disorder that occurs because of lesions of contralateral striatum. There are various causes like haemorrhagic or ischemic stroke, neoplasm, SLE, Huntington's disease, Sydenham's chorea, HIV, hyperthyroidism, trauma and drug toxicity. CHBG can occur both in patients with long standing uncontrolled diabetes mellitus or as an initial manifestation of hyperglycaemia leading to the diagnosis of diabetes mellitus[3]. Our patient was diagnosed as CHBG after ruling out the above causes.

The exact pathogenic mechanism of CHBG remains unknown, and various hypotheses have been proposed which include deposition of myelin breakdown products, metabolic acidosis, petechial haemorrhage, calcification, or gemistocytosis due to ischemia. There is a hypothesis that in metabolic brain diseases, release of lipids due to myelinolysis may present as abnormal signals on T1- and T2-weighted MRI.[2] Hyperglycaemia is hypothesized to activate anaerobic metabolism during which, the brain metabolizes gamma-aminobutyrate (GABA) into succinic acid, which causes metabolic acidosis. In nonketotic hyperglycaemia, insufficient ketoac-

etate for production of GABA and acetylcholine resynthesis can lead to lack of these neurotransmitters. Depletion of GABA and acetylcholine within the basal ganglia may lead to the onset of chorea[2,3]. Some studies also suggest the possibility that the metabolic disturbances unmask previous established asymptomatic striatum vasculopathy.[5]

The radiological findings in CHBG syndrome include hyperintensities in striatum and Globus-pallidum in T1 weighted MRI. In a study of 20 cases of chorea associated with hyperglycaemia, almost all the patients had striatal abnormalities on neuroimaging which was either contralateral or bilateral in few cases[3].

These patients usually have partial or complete resolution of symptoms with adequate blood sugar control recovery usually occurring within 48hrs, however can take 4-6 weeks for improvement of symptoms. However, when symptoms are too disabling and recovery is slow, use of medications such as tetrabenazine, anticonvulsants, and botulinum injection trials showed mix results. Haloperidol is one of the common medication used, but the outcomes vary, where some patients might not show significant improvement[3,6,7]. In our patient, both blood sugar control with insulin and oral hypoglycaemic agents and use of haloperidol helped in controlling the symptoms.

Unmasking of diabetes mellitus by coronavirus infectious disease of 2019 (COVID-19) is well-established with the duo having bidirectional relationship. However, very few cases of COVID 19 induced diabetic striatopathy has also been established[8].

Our patient improved symptomatically with blood sugar control over 3 months.

This case report though emphasizes on early recognition as well as treatment of CHBG, it alone may not be sufficient enough to deduce an average time between adequate glycaemic control and symptom control. Case series, case control studies and retrospective studies are further required to expand on the pathophysiologic mechanisms and to further identify risk factors.

Conclusion

CHBG, being a complication of uncontrolled non-ketotic diabetes mellitus, clinicians must be aware of it, as early recognition and treatment leads to resolution of the choreiform movements. Early identification helps to cut down the expensive workup and medications which would probably have side effects which in turn add to the discomfort of the patient and frequent hospital visits. Unfamiliarity with these conditions may lead the treating physician to attribute it to psychological or psychiatric disturbances in the absence of other neurological signs. Further research is needed to study the underlying mechanisms, risk factors and newer treatment modalities for refractory cases.

References

1. Sperling M, Bhowansingh R. Chorea Hyperglycemia Basal Ganglia Syndrome in a 63-Year-Old Male. Case Reports in Medicine. 2018;2018:1–4. Available from: https://doi.org/10.1155/2018/9101207.
2. Choi S, Rhee SY, Kim HS. Chorea Hyperglycemia Basal Ganglia Syndrome in a Young Patient with Type 1 Diabetes Mellitus: a Case Report. Brain & Neurorehabilitation. 2020;13(2):1–6. Available from: https://doi.org/10.12786/bn.2020.13.e8.
3. Gurursamy S, Sivamani K, Pitchai S, Chogle AS. Hemichorea Hyperglycaemia Basal Ganglia Syndrome in a Patient with Type 1 Diabetes Mellitus: A Rare Case Report. Journal of Clinical and Diagnostic Research. 2020;14(7):OD01–OD02. Available from: https://www.jcdr.net/articles/PDF/13810/44147_CE[Ra1]_F(KM)_PF1(AJ_KM)_PN(SL).pdf.
4. Das L, Pal R, Dutta P, Bhansali A. "Diabetic striatopathy" and ketoacidosis: Report of two cases and review of literature. Diabetes Research and Clinical Practice. 2017;128:1–5. Available from: https://doi.org/10.1016/j.diabres.2017.03.008.
5. Taboada GF, Lima GAB, Castro JEC, Liberato B. Dyskinesia associated with hyperglycemia and basal ganglia hyperintensity: report of a rare diabetic complication. Metabolic Brain Disease. 2013;28(1):107–110. Available from: https://doi.org/10.1007/s11011-012-9357-z.
6. Tsalta-Mladenov ME, Georgieva DK, Andonova SP. Hyperglycemic hemichorea due to diabetic striatopathy: case-based review. Current Medical Research and Opinion. 2022;38(3):365–369. Available from: https://doi.org/10.1080/03007995.2021.2015159.
7. Jaafar J, Rahman RA, Draman N, Yunus NA. Hemiballismus in Uncontrolled Diabetes Mellitus. Korean Journal of Family Medicine. 2018;39(3):200–203. Available from: https://doi.org/10.4082/kjfm.2018.39.3.200.
8. Ghosh R, Dubey S, Roy D, Ray A, Pandit A, Ray BK, et al. Choreo-ballistic movements heralding COVID-19 induced diabetic ketoacidosis. Diabetes & Metabolic Syndrome: Clinical Research & Reviews. 2021;15(3):913–917. Available from: https://doi.org/10.1016/j.dsx.2021.04.010.

How to cite this article: Krishnamurthy V, Anusha A, Jayaram H. Chorea Hyperglycaemia Basal Ganglia Syndrome in an Uncontrolled Diabetic Patient. J Med Sci Health 2023; 9(3):347-350

Date of submission: 24.12.2022
Date of review: 11.01.2023
Date of acceptance: 02.09.2023
Date of publication: 28.12.2023