Case Report

A Case Report on Diabetic Striatopathy; A Diagnostic Dilemma

Mark SHK[1], Ekta SA[2], Alam I[3], Ahmed KGU[4]

Abstract

Diabetic striatopathy (DS) is an acute hyperkinetic movement disorder characterized by hemiballismus-hemichorea (HBHC) due to nonketotic hyperglycemia. Diabetic striatopathy occurs due to striatal (putamen, caudate nucleus, globus pallidus) pathology. The exact pathogenesis and mechanism remain unclear but may involve hyperviscosity, ischemia, and alterations in basal ganglia neurotransmitters. We present here a case of HBHC syndrome with right-sided neuroimaging findings and contralateral chorea due to uncontrolled type 2 diabetes mellitus. She had a history of poorly controlled type 2 diabetes and presented with involuntary movements of her left limb suggestive of chorea. Laboratory tests confirmed hyperglycemia, with an elevated hemoglobin A1c level. Neuroimaging revealed T1-hyperintensity in the right putamen. The patient was diagnosed with diabetic striatopathy and responded well to insulin therapy and haloperidol with a rapid resolution of symptoms. The striking features on imaging such as computed axial tomography (CT) scan of the brain and T1-weighted magnetic resonance imaging (MRI) of the brain can mislead the clinician to an erroneous diagnosis of a cerebral hemorrhage and/or ischemic infarct, especially in an acute setting. With careful and thoughtful analysis, an accurate diagnosis can spare the patient unnecessary anxiety and medical costs.

Keywords: *hyperkinetic movement disorder, hemichorea, hemiballismus, diabetes mellitus, diabetic striatopathy.*

DOI: https://doi.org/10.3329/jdmc.v33i1.83496
J Dhaka Med Coll. 2024; 33(1) : 42-46

Introduction

Diabetic striatopathy (DS) is a condition occurring in individuals with type II diabetes mellitus (T2DM). It is an acute hyperkinetic movement disorder secondary to nonketotic hyperglycemia. DS is a rare disorder with an estimated prevalence of 1 in 100,000, which is likely an underestimation due to unfamiliarity and missed diagnosis.[1] This syndrome was first described by Bedwell in 1960.[2] It is more prevalent in females, and the greatest risk factor is old age.[3] It is presented as development of hemiballismus-hemichorea (HBHC) associated with hyperintensity on T1-weighted magnetic resonance imaging (MRI) of the putamen, caudate nucleus, and globus pallidus in various combinations, with the putamen being most involved.[4,5] Imaging usually shows an absence of mass effect or contrast enhancement, indicating an intact blood-brain barrier. The internal capsule is normally spared.[6] The most likely pathology of DS involves myelin destruction caused by swollen reactive astrocytes called gemistocytes.[7] This hyperkinetic movement disorder is usually improved with judicious glucose control, sometimes necessitating treatment with anti-chorea agents such as dopamine antagonists (haloperidol), vesicular monoamine uptake (VMAT-2) inhibitors (tetrabenazine), or gamma-

1. Dr. Sayeef Hossain Khan Mark, Registrar, Department of Neurology, Dhaka Medical College Hospital, Dhaka
2. Dr. Shams Afroze Ekta, Phase B Resident, Department of Neurology, Dhaka Medical College Hospital, Dhaka
3. Dr. Iftikher Alam, Associate Professor, Department of Neurology, Dhaka Medical College Hospital, Dhaka
4. Prof. Kazi Gias Uddin Ahmed, Professor & Head, Department of Neurology, Dhaka Medical College Hospital, Dhaka

Correspondence : Dr. Sayeef Hossain Khan Mark, Registrar, Department of Neurology, Dhaka Medical College Hospital, Dhaka, Mobile: 01632168642, Email: mark.dmck63@gmail.com

Received: *11.10.2023* ***Accepted:*** *08.01.2024*

aminobutyric acid (GABA) agonists (clonazepam).[8]

Case Presentation

We present the case of 60-year-old woman with poorly controlled diabetes mellitus and one month history of sudden, involuntary, rapid, jerky, irregular movement of the left upper and lower extremity, sparing the tongue, face, and the neck which got worsened over 2 weeks. These movements did not abate with sleep. Her speech was unaffected, but her gait was interrupted by the motion. Past medical history showed 10-year history of diabetes mellitus with non-compliance to anti-diabetic medication. She had no previous history of trauma, no history of similar symptoms. There was no family history of chorea, and she did not smoke cigarettes or drink alcohol. She only used to take metformin for diabetes but irregularly. There was no significant occupational or environmental exposure to toxins. On examination, her blood pressure was 130 mmHg systolic and 80 mmHg diastolic with a pulse of 76 beats per minute. On initial inspection, examination was notable for high amplitude involuntary dyskinesia of the axis (trunk), left upper extremity, and left lower extremity. The movement was rapid, involuntary, jerky, random, irregular, large amplitude, purposeless, flinging, sometimes forceful. On gait examination, the patient's left foot and lower leg jerked during the swing phase, making her gait irregular and unsteady. Her speech was normal. Her extraocular muscles were intact with normal-amplitude eye motion in all directions without nystagmus. The rest of the cranial nerve examination was normal. Blinking frequency was also normal. Kayser-Fleischer rings were not detected. There were no stigmata of connective tissue disease or metabolic or nutritional disorders.

Motor strength was preserved in the upper and lower extremities. Coordination examination with left finger-to-nose test and left heel-to-shin test demonstrated sinuous movements due to the choreoathetosis, but the target was reached. Patient's deep tendon reflexes were normal. Sensory examination was generally preserved to small and large fiber modalities in the fingers and toes except for a diminished vibration sense bilaterally. Plantar responses were flexor. The patient's random glucose level was 301 mg/dL (normal range: 70-115 mg/dL), and his hemoglobin A1c was 12% (normal range: 3.8-5.6%). The basic metabolic panel was otherwise normal.

Table -I

Investigations.

Investigations.			
Full Blood Count	White Blood Cells (× 10^9 /L)	8.4	4.5 – 11
	Haemoglobin (g/dL)	11.5	11-13
	Platelets (x 10^9/L)	239	150 – 400
Liver Biochemistry	Alanine Transaminase (U/L)	17	< 40
	Aspartate Transaminase (U/L)	22	< 40
	Serum Albumin (g/L)	39	35 - 50
Renal Profile	Serum Creatinine (mg/dl)	89	20 - 275
	Blood Urea (mg/dl)	16	6 - 24
Electrolytes	Serum Sodium (mmol/L)	139	135 – 145
	Serum Potassium (mmol/L)	3.9	3.5 – 5.5
	Serum Calcium (mmol/L)	2.4	2.2 to 2.7
	Serum Magnesium (mg/dl)	1.7	1.6 – 2.5
Thyroid Function Test	Thyroid Stimulating Hormone (mIU/L)	3.95	0.5 to 5.0
Urine	Urinary Ketone	Absent	
Other Investigations	Serum Ceruloplasmin (mg/dl)	16	14 - 20

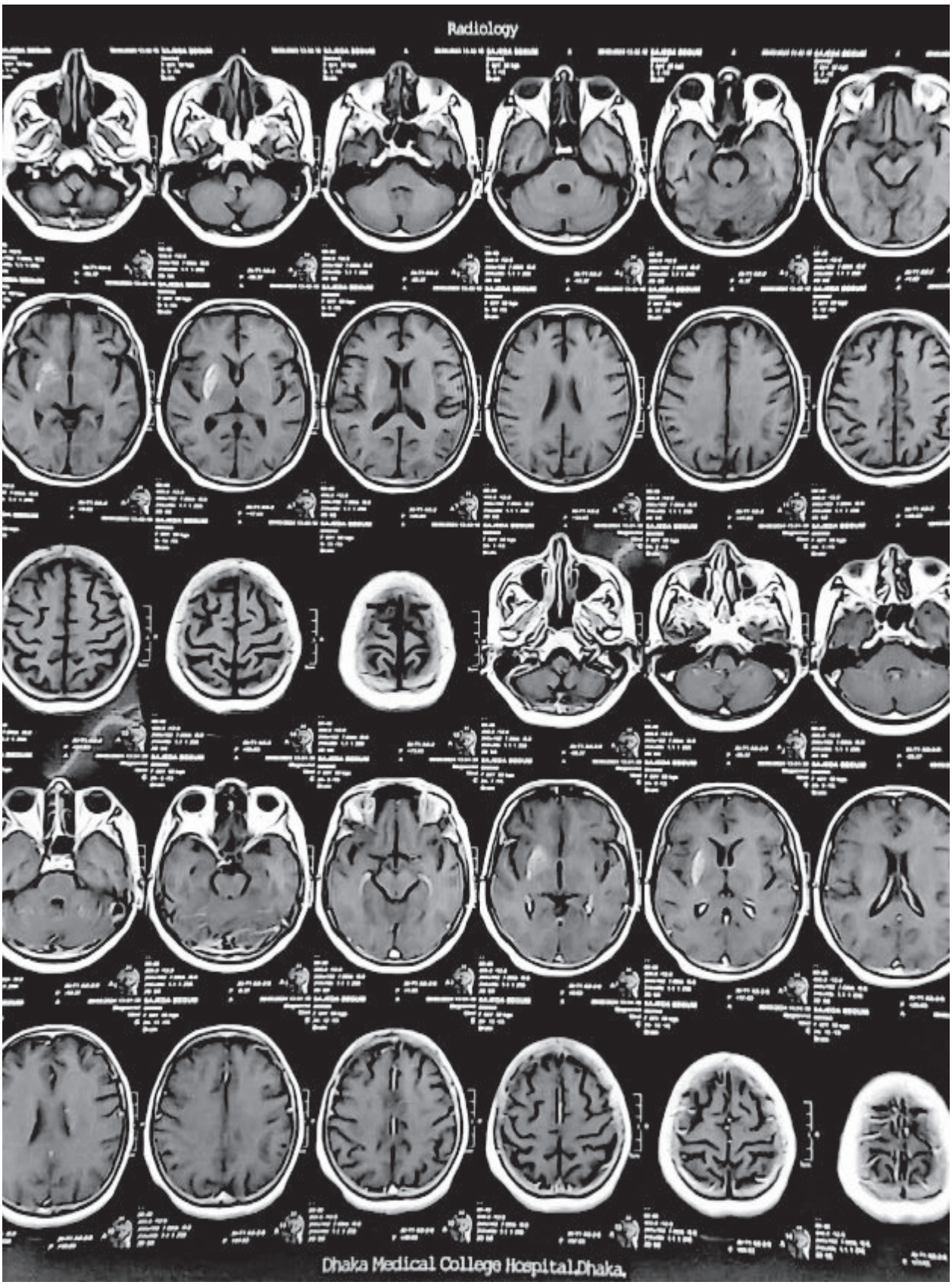

Fig.-1:*T1-weighted MRI of the brain showing hyperintensity in the right putamen*

Based upon the absence of a family history of chorea, the characteristic clinical history of chronic poorly controlled diabetes, and the characteristic MRI brain findings, probable diagnosis of Diabetic Striatopathy was made. The patient was treated with insulin for hyperglycemia and a trial of haloperidol daily was prescribed to reduce the amplitude and frequency of the disabling choreoathetosis. The patient demonstrated substantial improvement of the movement disorder following the treatment within a week.

Discussion

Acute HBHC is a defining clinical characteristic of DS. However, diagnosing DS is not always straightforward as there are many differential diagnoses of HBHC. Common clinical presentations of DS include HBHC involving unilateral limbs, progression from upper to lower extremities with suppression of HBHC during sleep.[8] In our case, the patient's acute onset of unremitting characteristic unilateral A1c HBHC and the hemoglobin 12% raised suspicion for DS. Based on the principles of T1-weighted MRI sequences, T1-shortening resulting in hyperintensities can be caused by various factors, including melanin, subacute haemorrhage with methaemoglobin, fat, slow-velocity blood flow, high protein content, and the presence of paramagnetic transition metals such as manganese, iron, zinc, and copper, which have unpaired electrons.[14] The clinical features observed in this patient can exclude certain causes of T1 hyperintensity. For example, disorders related to copper deposition (i.e. Wilson disease) and other similar conditions typically present as chronic diseases with a gradual onset.[15] Early subacute blood on an MRI brain demonstrates hypodensities on both susceptibility-weighted imaging (SWI) and apparent diffusion coefficient (ADC) sequences, and late subacute blood shows hyperintensity on diffusion-weighted imaging (DWI) sequences.[11,12] An ischemic infarct would show high signal intensity on DWI sequence and low signal intensity on ADC sequence.[13]

According to a meta-analysis published in 2020, 96.6% of the patients reported had type 2 diabetes, and the average hemoglobin A1c was 13.1%. In total, 97.7% of patients presented with chorea/hemichorea, and the striatal area mostly involved (78.6%) was putamen. It was also noted that MRI had a greater sensitivity (95.33%) of detecting abnormalities associated with DS, compared with that of CT (78.9%)[14]. In this case, MRI showed the typical feature in putamen which is the most common site. Greater successful treatment rate (76.2%) was noted in patients treated with combined glycemic control and antichorea medications, compared with 25.6% in those treated only with glycemic control. In this case, insulin and haloperidol together helped to make rapid recovery within a week.

There also have been multiple meta-analyses to understand the phenomenon. However, the limited number of case data was a major limitation to explain the pathogenesis and phenomenon[15]. We believe our case report will contribute to current available data in the literature.

DS can mimic other movement disorders and strokes, which can be challenging. This report creates awareness to physicians as well as mid-level providers that DS is a good differential diagnosis to consider when confronted with a movement disorders, especially in patients with uncontrolled diabetes. Acute-onset chorea in poorly controlled hyperglycemia in type 2 diabetes should alert the providers to screen with MRI for DS.

Conclusion

In conclusion, physicians need to be alert of possibility of DS, as its timely-bound condition and prognosis partially depend on the time window of detection and management. Nevertheless, the radiologist needs to pay attention to the distinct features of this condition and should not dismiss other possibilities, such as haemorrhage in patients with atypical presentation. Further study of its pathophysiological mechanisms is needed to guide better management.

References:

1. Ondo WG. Hyperglycemic nonketotic states and other metabolic imbalances. Handb Clin Neurol. 2011;100:287–91

2. Bedwell SF. Some observations on hemiballismus. Neurology. 1960;10:619–22. Available from: https://doi.org/10.1212/wnl.10.6.619

3. 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.

4. Abe Y, Yamamoto T, Soeda T, et al. Diabetic striatal disease: clinical presentation, neuroimaging, and pathology. Intern Med. 2009;48:1135–41.

5. Cosentino C, Torres L, Nuñez Y, Suarez R, Velez M, Flores M. Hemichorea/hemiballism associated with hyperglycemia: report of 20 cases. Tremor Other Hyperkinet Mov (N Y). 2016;6:402.

6. Sitburana O, Ondo WG. Tetrabenazine for hyperglycemic-induced hemichorea-hemiballismus. Mov Disord. 2006;21:2023–5.

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. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC8337567/

8. Chua CB, Sun CK, Hsu CW, Tai YC, Liang CY, Tsai IT. "Diabetic striatopathy": clinical presentations, controversy, pathogenesis, treatments, and outcomes. Sci Rep. 2020;10:1594.

9. Heit JJ, Iv M, Wintermark M. Imaging of intracranial hemorrhage. J Stroke. 2017;19:11–7.

10. Khedr SA, Kassem HM, Hazzou AM, Awad E, Fouad MM. MRI diffusion-weighted imaging in intracranial hemorrhage (ICH). Egypt J Radiol Nucl Med. 2013;44:625–34.

11. Allen LM, Hasso AN, Handwerker J, Farid H. Sequence-specific MR imaging findings that are useful in dating ischemic stroke. Radiographics. 2012;32:1285–97.

12. Ginat DT, Meyers SP. Intracranial lesions with high signal intensity on T1-weighted MR images: differential diagnosis. Radiographics. 2012;32:499–516.

13. Chang X, Zhang J, Jiang Y, Wang J, Wu Y. Natural history and genotype-phenotype correlation of pantothenate kinase-associated neurodegeneration. CNS Neurosci Ther. 2020;26:754–61.

14. Lin YT, Chen SC, Yip PK, et al. Magnetic resonance imaging volumetric analysis for diabetic striatopathy with two episodes of hemichorea-hemiballism syndrome: a case report. Medicine (Baltimore). 2019;98:e17249. Available from: https://doi.org/10.1097/MD.0000000000017249

15. Xu Y, Shi Q, Yue Y, et al. Clinical and imaging features of diabetic striatopathy: report of 6 cases and literature review. Neurol Sci. 2022;43:6067–77. Available from: https://doi.org/10.1007/s10072-022-06342-y

.