

Diabetic Striatopathy: A Rare Movement Disorder as First Clinical Manifestation of Type 1 Diabetes Mellitus

¹Nirmal Kumar Sharma, ²Jaiprakash Kuldeep, ³Mohammed Shamnas

¹Senior Professor, Department Of General Medicine, GMC, Kota

²Post Graduate Resident, Department Of General Medicine, GMC, Kota

³Post Graduate Resident, Department Of General Medicine, GMC, Kota

Corresponding Author: Jaiprakash Kuldeep, Post Graduate Resident, Department Of General Medicine, GMC, Kota.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Diabetic striatopathy is uncommon movement disorder which is described by characteristic clinical (hemichorea and hemiballismus) and neuroimaging finding (hyper intensity in T1W with hypointensity in T2W and FLAIR in basal ganglia of contralateral side) usually occurring as a late neurological complication of type 2 diabetes mellitus in non ketotic hyperglycemic patients . In this case report we discussed about this rare entity as first clinical manifestation of type 1 diabetes mellitus in a young male patient with its clinical presentation, related pathophysiology and management. We also highlighted the significance of early diagnosis of this reversible condition.

Keywords: Diabetic striatopathy, Hemichorea, Hemiballismus, Non ketotic hyperglycemia, Type 1 diabetes.

Introduction

Diabetes mellitus refers to a group of common disorders that share the phenotype of hyperglycemia .The common complications of diabetes mellitus that involving the nervous system are cranial mono neuropathies and peripheral polyneuropathy .Diabetic striatopathy (chorea, hyperglycemia , basal ganglia syndrome) , Is rare movement disorder of poor metabolic control of DM which may be present at beginning or during course of the

disease occurring as hemichorea (involuntary, continuous, rapid and abrupt contractions that affect half of the body) or more severe form as hemiballismus on contralateral side of the lesion with the neuroimaging abnormalities⁶⁻⁹ in form of striatal hyper intensity on T1-weighted magnetic resonance images(MRI) and hypointensity on T2W/FLAIR.After the vascular causes,nonketotic hyperglycemia is second most common cause of hemichorea and hemiballismus^{10,11}.Most of patients of diabetic striatopathy were having DM type 2 and some case were reported as chorea or ballismus as initial clinical presentation of type 2 diabetes mellitus¹², But here we report a case , presented with symptoms of sudden involuntary movements (hemichorea, hemiballismus) as initial presentation and also diagnosed with DM type 1 simultaneously.

Case Report

A 22 year old male patient with no previous medical history presented in our hospital with non rhythmic involuntary choreiform movement of sudden onset in the right upper and lower extremity for last 10 days. There were also complain of increase frequency of micturition and increase thirst.On physical examination the patient had normal higher mental function with normal power and sensation in the right upper and lower extremities, with no

additional focal neurologic deficit. Deep tendon reflex were normal in both upper and lower limb with plantar bilaterally flexor along with presence of choreiform and ballistic movement in right upper and lower limbs. Non contrast CT was performed and demonstrated hyper attenuation of the left basal ganglia predominantly in caudate nucleus and putamen, shown in fig. 1.

Non contrast MR imaging of the brain demonstrated T1 hyperintensity in the left basal ganglia with hypo intensity in T2 and FLAIR images corresponding to the area of hyper density seen on the preceding CT scan, without generating vasogenic oedema or mass effect as shown in figure 2,3,4.

Figure 1: Axial non contrast CT demonstrates increased density within the LEFT basal ganglia, involving predominantly the putamen and caudate nucleus and the unilateral lesion would be less likely for other metabolic diseases, or prior toxic/global anoxic injury.



Figure 2,3,4: Axial T1 weighted MRI suggested of hyper intensity in left basal ganglia and hypointensity in T2/FLAIR MRI in left Basal ganglia corresponding to the area of hyper density seen on the preceding CT scan.

Figure 2



Figure 3

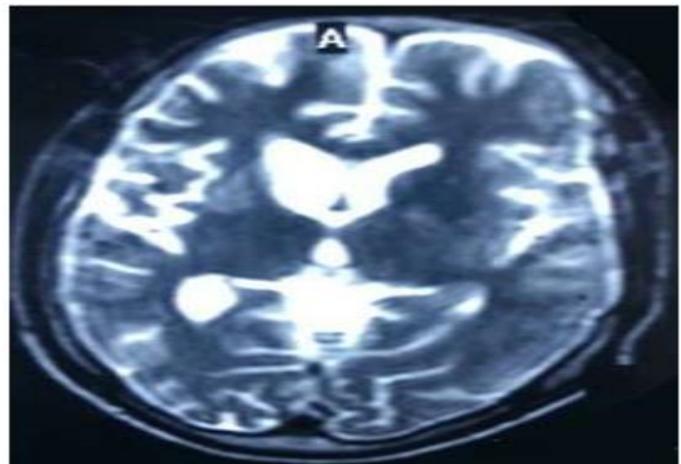


Figure 4



On admission, lab investigations were random blood sugar-419mg/dl, urine ketones were negative.HbA1c level was significantly elevated at 11.7% with low C-peptide level.

Patient was given insulin and symptomatically improved after blood sugar control and he was discharged within 12 days after admission with subsiding of the movements.

Discussion

Diabetic striatopathy typically presents as Hemichorea ,Hemiballismus which are unilateral movement disorder occurred by uncontrolled Diabetes mellitus in the setting of nonketotic hyperglycemia in which lesion affecting the contralateral Basal ganglia (striatum)^{4,13}. Symptoms may be bilateral. Nonketotic hyperglycemia is second most common cause after haemorrhagic or ischemic stroke.¹⁴

Typically cases of diabetic striatopathy are Asian elderly women mostly in type 2 diabetes mellitus, predominantly in developed countries¹⁵⁻¹⁸but rarely cases have been also reported in type1 Diabetes mellitus ^{19,20}.The Pathogenesis suggested for striatopathy is hyper viscosity induced by hyperglycemia leads to decrease cerebral perfusion with increased anaerobic metabolism and reduced level of GABA (inhibitory neurotransmitter) in basal nuclei neuron.

Neuroimaging are characteristic in diabetic striatopathy involving the basal ganglia.The CT can be normal or demonstrate hyper intensity in the striatum but characteristic MRI T1 hyper intensity with variability of signal in T2 and FLAIR in basal nuclei²¹.Some proposed reason for neuroimaging finding are protein denaturation or petechial haemorrhage, mineral deposition²².Also compromised BBB and myelinolysis in diabetics.^{14,23}

The differential diagnosis of T1 hyperintensinty in basal ganglia are Wilson disease which is differentiated by hyperintensity in T2,others are hepatic encephalopathy, magnesium toxicity, carbon monoxide

poisoning²⁴.History and clinical presentation and laboratory results are generally sufficient to differentiate these diagnosis

Conclusion

Diabetic striatopathy which is a rare syndrome with characteristic clinical (chorieform and ballistic movements) and neuroimaging findings T1 hyperintensity in Basal ganglia of contralateral side of movements seen in setting of nonketotic hyperglycemia mostly in elderly women but it is advisable to measure of blood sugar in patients presenting with these type of movement disorder even in early age group as it is reversible condition with normalisation of blood sugar level.

References

1. Harrison principles of internal medicine ,volume2,20th edition.
2. Charnogursky G, Emanuele NV, Emanuele MA. Neurologic Complications of Diabetes. *Curr Neurol Neurosci Rep.* 2014;14:457.
3. Lai PH, Tien RD, Chang MH, Teng MM, Yang CF, Pan HB, Chen C, Lirng JF, Kong KW (1996) Chorea-ballismus with nonketotic hyperglycemia in primary diabetes mellitus. *AJNR Am J Neuroradiol* 17(6): 1057–1064
4. Bizet J, Cooper CJ, Quansah R, Rodriguez E, TelebM, Hernandez GT (2014) Chorea, hyperglycemia, basal ganglia syndrome (C-H-BG) in an uncontrolled diabetic patient with normal glucose levels on presentation.*Am J Case Rep* 15:143–146. doi:10.12659/AJCR.890179
5. Mittal P. Hemichorea Hemiballism Syndrome: The First Presentation of Type 2 Diabetes Mellitus as a Rare Cause of Chorea. *Iran J Radiol.* 2011;8(1):47–49.

6. Rector WG Jr, Herlong HF, Moses H 3rd. Nonketotic hyperglycemia appearing as choreoathetosis or ballism. *Arch Internal Med* 142: 154-155, 1982
7. Sanfield JA, Finkel J, Lewis S, Rosen SG. Alternating choreoathetosis associated with uncontrolled diabetes mellitus and basal ganglia calcification. *Diabetes Care* 9: 100-101, 1986.
8. Altafullah I, Pascual-Leone A, Duvall K, Anderson DC, Taylor S. Putaminal hemorrhage accompanied by hemichorea-hemiballism. *Stroke* 21: 1093-1094, 1990.
9. Nakata K, Takeshima M, Itoshima T, et al. Hemiballism in a diabetic patient. A case report. *Journal of Okayama Saiseikai General Hospital* 22: 47-52, 1990 (in Japanese).
10. Verma R, Praharaj H. Hemichorea-hemiballism as the presenting manifestation of diabetes mellitus. *BMJ Case Rep.* 2013;15:1-2.
11. B, Choi J, Ko H. Globus Pallidus Internus Deep Brain Stimulation for Disabling Diabetic Hemiballism/Hemichorea. *Case Reports in Neurological Medicine.* 2017;5:2165905.
12. CJ Lin, P Huang. Delayed onset diabetic striatopathy: Hemichorea-hemiballism one month after a hyperglycemic episode. *Am J Emerg Med.* 2017;35(7):1036.e3-1036.e4.
13. Abe Y, Yamamoto T, Soeda T (2009) Diabetic Striatal Disease: Clinical Presentation, Neuroimaging, and Pathology. *Inter Med* 48: 1135-1141
14. Bathla G, Policeni B, Agarwal A (2014) Neuroimaging in patients with abnormal blood glucose levels. *AJNR Am J Neuroradiol* 35(5): 833-840. doi:10.3174/ajnr.A3486
15. Bathla G, Policeni B, Agarwal A (2014) Neuroimaging in patients with abnormal blood glucose levels. *AJNR Am J Neuroradiol* 35(5): 833-840. doi:10.3174/ajnr.A3486
16. Oh SH, Lee KY, Im JH, Lee MS (2002) Chorea associated with nonketotic hyperglycemia and hyperintensity basal ganglia lesion on T1-weighted brain MRI study: a metaanalysis of 53 cases including four present cases. *J Neurol Sci* 200(1-2):57-62
17. Kranick SM, Price RS, Prasad S, et al. Clinical reasoning: a 52-year-old woman with subacute hemichorea. *Neurology.* 2008;71(20):e59-e62.
18. SF Sung, CH Lu. Focal neurological symptoms as the presenting manifestations of nonketotic hyperglycemia: Report of two cases. *J Intern Med Taiwan.* 2007;18:206-211.
19. Aquino JH, Spitz M, Pereira JS. Hemichorea-hemiballismus Neurographics 2018 November/December;8(6):424-427; www.neurographics.org as the first sign of type 1b diabetes during adolescence and its recurrence in the setting of infection. *J Child Neurol* 2015;30: 1362-65. 10.1177/0883073814553972
20. Altafullah I, Pascual-Leone A, Duvall K, et al. Putaminal hemorrhage accompanied by hemichorea-hemiballism. *Stroke* 1990;21:1093-94. 10.1161/01.STR.21.7.1093
21. Yu F, Steven A, Birnbaum L, et al. T2*-based MR imaging of hyperglycemia-induced hemichorea hemiballism. *J Neuroradiol.* 2017;44(1):24-30.
22. Atay M, Yetis H, Kurtcan S, Aralasmak A, Alkan A (2014) Susceptibility weighted imaging features of nonketotic hyperglycemia: unusual cause of hemichorea-hemiballismus. *J Neuroimaging.* doi:10.1111/jon.12084
23. Wintermark M, Fischbein NJ, Mukherjee P (2004) Unilateral Putaminal CT, MR, and Diffusion

Abnormalities Secondary to Nonketotic
Hyperglycemia in the Setting of Acute Neurologic
Symptoms Mimicking Stroke. *AJNR Am J
Neuroradiol* 25:975–976

24. Lai PH, Chen C, Liang HL, Pan HB (1999)
Hyperintense basal ganglia on T1-weighted MR
imaging. *AJR Am J Roentgenol* 172:1109-15