Case report

Non-Ketotic Hyperglycemic Hemichorea as an Initial Manifestation of Type 2 Diabetes Mellitus: Case Report and Review of The Literature

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Abstract

Non-Ketotic Hyperglycemic Hemichorea (NKHCB) is a rare movement disorder associated with uncontrolled diabetes mellitus. It has been reported in type 2 diabetes, and much rarer in type 1 diabetes mellitus. It is characterized by hemichorea-hemiballism that resolves with glycemic control, though some cases may be unremitting. It has hardly been reported as the presenting complaint in a new diagnosis of diabetes mellitus. We hereby discuss a case of NKHCB as a first complaint in a 52-year-old lady not previously known to have diabetes. The CT scan did not reveal any abnormality. The disorder resolved within a month of the diagnosis after proper glycemic control and use of neuroleptic agents. NKHCB is an important differential diagnosis to consider in patients presenting with hemichorea-hemiballism, even if not previously known to be diabetic.

Key words: Hemichorea, Hemiballism, Diabetes, Diagnosis

Introduction

Non-Ketotic Hyperglycemic Hemichorea (NKHCB) is a rare movement disorder associated with uncontrolled diabetes mellitus. It has been reported in type 2 diabetes, and much rare in type 1 diabetes mellitus. It is characterized by hemichorea-hemiballism that resolves with glycemic control, though some cases may be unremitting (1,2). It has hardly been reported as the presenting complaint in a new diagnosis of diabetes mellitus(3,4). It has been commonly reported in females compared to males(3,5). This case illustrates the importance of maintaining a high index of suspicion in patients presenting with hemichorea-hemiballism, because prompt glycemic control leads to resolution of symptoms. We hereby discuss a case of NKHCB as a first complaint in a 52 year old lady not previously known to have diabetes.

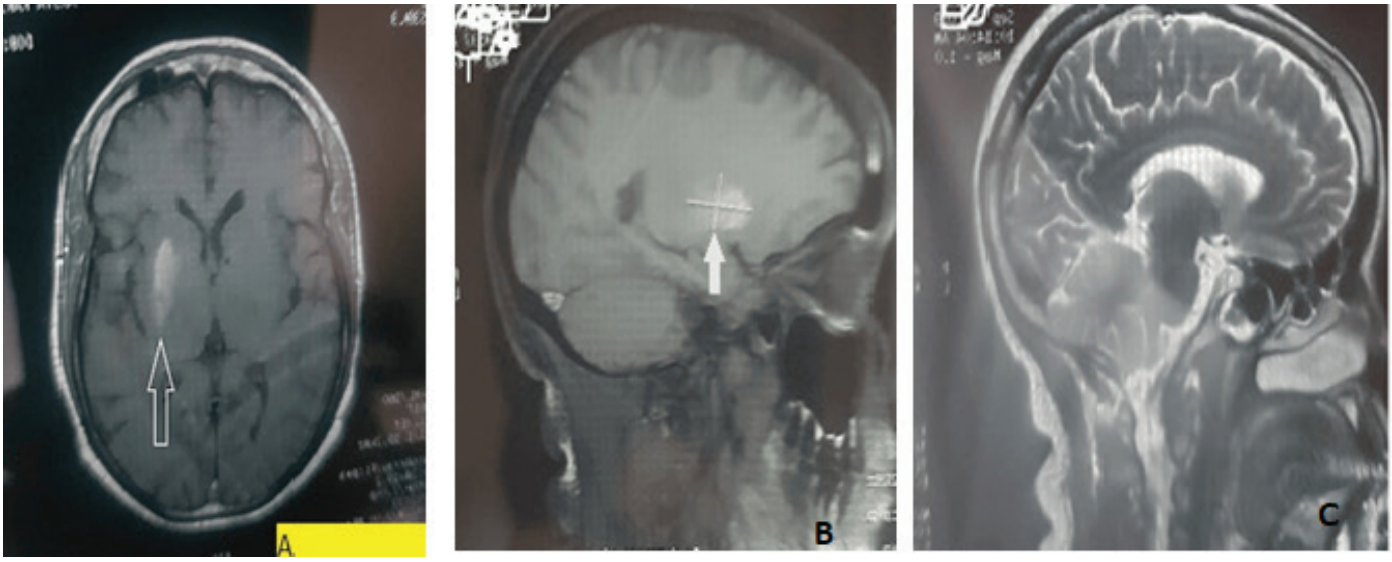
Case report

A 52 year old patient presented to our hospital with one month history of involuntary movements of the right upper and lower limbs. She had no history of trauma, stroke or hypertension, and she was alert. The movements interfered with feeding and ambulation. Her examination revealed right-sided periodic choreiform and ballistic movements, suppressed by sleep. She had normal motor power, reflexes and preserved sensation over all dermatomes.

Chemistry revealed normal sodium of 136mmol/l, urea of 7.0mmol/L, calcium of 2.30mmol/L, and creatinine of 81umol/L. Her random blood glucose was 33.0mmol/L, and HbA1c of 11.0%. Her blood count showed white blood cell of 7.5 with no left shift, haemoglobin of 14.2g/dl and ESR of 14mm/hr. She had normal thyroid and hepatic function panels. CSF studies revealed normal proteins and glucose.

The CT scan did not reveal any abnormality. An MRI showed a T1 hyperintensity on the left lentiform nucleus, with normal attenuation in T2 and DW sequences. This is shown in Figures A-C.

Figure A-C: T1-Weighted axial MRI of the patient showing the hyperintensity in the left lentiform nucleus (arrow). B shows a T1-weighted sagittal MRI of the patient showing the hyper- intense lesion in the lentiform nucleus, while C shows a T2-weighted MRI of the patient with the hyper-intense lesion not visible. Instead, there is normal attenuation on a T2-weighted MRI image



The patient was initiated on 10 units regular insulin thrice with dose titration until euglycemia was achieved. She was also started on haloperidol 5mg twice a day due to the distressing abnormal movements, but this was withdrawn after 3 days because of adverse effects. The symptoms gradually improved with a complete resolution at 6 weeks. Her glycemic control remains excellent on lantus (glargine) insulin and metformin.

Discussion

Glucose derangements have been reported to cause movement disorders, and in rare cases, may present as the first manifestation of diabetes mellitus (4). Elderly women are more predisposed to this condition (6,7). A review of 53 cases of Non-Ketotic Hyperglycemic Hemichorea revealed a mean age of 71.1 years (age range 22-92 years), with a female to male predisposition of 15:1.8. In this series, 88.6% of the patients reported hemichorea, compared to 11.4% who had bilateral chorea, and a small group with facial involvement (8).

The characteristic finding is a T1-weighted brain MRI hyper-intensity (9,10) MR, and Single-Photon Emission CT [SPECT]. In hemichorea, the hyper-intensity is in the contralateral basal ganglia, affecting the caudate, putamen or lentiform nucleus (11,12). The other differentials for T1- hyper-intensity include manganese toxicity, chronic liver failure, calcium metabolism abnormalities with calcification, Wilson's disease, neurofibromatosis and hypoxic brain injury (5).

The pathogenic process is thought to be due to hyperglycemia-induced perfusion changes in the contralateral striatum and ischemic excitotoxicity of GABAergic neurons. This leads to excessive inhibition of the subthalamic nuclei and excitatory cortical output (12–14). It is postulated that a dopamine hypersensitivity or hyperactive dopaminergic state exists in post-menopausal women, hence their predisposition to this condition (15,16).

The mainstay of treatment encompasses aggressive glycemic control. In refractory cases, drugs that block postsynaptic dopamine (D2) receptors such as haloperidol, risperidone and olanzapine can be used. Tetrabenazine, a dopamine depletor has also been used with some success. Topiramate and benzodiazepines also improve symptoms due to GABAergic properties (17).

Resolution of chorea is variable, with one series reporting a range of one day to 10 months. In this series, majority of the patients reached full recovery within 6 months (8). Radiological resolution typically lags behind clinical progress, and repeat MRI should be delayed until several months after resolution of symptoms (8,11,12).

Conclusion

In conclusion, non-ketotic hyperglycemic hemichorea is a rare but important differential to consider in patients presenting with hemichorea-hemiballism. Prompt diagnosis carries an excellent diagnosis. All patients with this presentation should be screened for diabetes mellitus.

References

1. Pinsker JE, Shalileh K, Rooks VJ, Pinsker RW. Hemichorea-hemiballism secondary to non-ketotic hyperglycemia. *J Clin Med Res*. 2015; **7**(9):729–730.
2. Verma R, Praharaj HN. Hemichorea-hemiballism as the presenting manifestation of diabetes mellitus. *BMJ Case Rep*. 2013; **2013**:bcr2013200064.
3. Cosentino C, Torres L, Nuñez Y, Suarez R, Velez M, Flores M. Hemichorea/hemiballism associated with hyperglycemia: Report of 20 cases. *Tremor Hyperkinetic Mov*. 2016; **6**(0):402.
4. Haan J, Kremer H, Padberg G. Paroxysmal choreoathetosis as presenting symptom of diabetes mellitus. *J Neurol Neurosurg Psychiatry*. 1989; **52**:133.
5. Bizet J, Cooper CJ, Quansah R, Rodriguez E, Teleb M, Hernandez GT. Chorea, Hyperglycemia, Basal Ganglia Syndrome (C-H-BG) in an uncontrolled diabetic patient with normal glucose levels on presentation. *Am J Case Rep*. 2014; **15**:143–146.
6. Janavs JL, Aminoff MJ. Dystonia and chorea in acquired systemic disorders. *J Neurol Neurosurg Psychiatry*. 1998; **65**(4):436–445.
7. Lee BC, Hwang SH, Chang GY. Hemiballismus–hemichorea in older diabetic women: A clinical syndrome with MRI correlation. *Neurology*. 1999; **52**(3):646–646.
8. 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. *J Neurol Sci*. 2002; **200**(1–2):57–62.
9. Lai PH, Tien RD, Chang MH, Teng MM, Yang CF, Pan HB, *et al*. Chorea-ballismus with nonketotic hyperglycemia in primary diabetes mellitus. *Am J Neuroradiol*. 1996; **17**(6):1057–64.
10. Nagai C, Kato T, Katagiri T, Sasaki H. Hyperintense putamen on T1-weighted MR images in a case of chorea with hyperglycemia. *Am J Neuroradiol*. 1995; **16**(6):1243–46.
11. Hsu JL, Wang HC, Hsu WC. Hyperglycemia-induced unilateral basal ganglion lesions with and without hemichorea. A PET study. *J Neurol*. 2004; **251**(12):1486–490.
12. Shan DE, Ho DM, Chang C, Pan HC, Teng MM. Hemichorea-hemiballism: an explanation for MR signal changes. *Am J Neuroradiol*. 1998; **19**(5):863–870.
13. Ondo WG. Hyperglycemic nonketotic states and other metabolic imbalances. In: Handbook of Clinical Neurology [Internet]. Elsevier; 2011 [cited 2024 May 23]. p. 287–91. Available from: https://linkinghub.elsevier.com/retrieve/pii/B9780444520142000215
14. Guo Y, Miao Y wei, Ji X fei, Li M, Liu X, Sun X pei. Hemichorea associated with nonketotic hyperglycemia: Clinical and neuroimaging features in 12 patients. *Eur Neurol*. 2014; **71**(5–6):299–304.
15. Dixit A. Nonketotic hyperglycemia hemichorea and hemiballismus: a case report. *J Med Case Reports*. 2024; **18**(1):154.
16. Hansford BG, Albert D, Yang E. Classic neuroimaging findings of nonketotic hyperglycemia on computed tomography and magnetic resonance imaging with absence of typical movement disorder symptoms (hemichorea-hemiballism). *J Radiol Case Rep*. 2013; **7**(8):1–9.
17. Driver-Dunckley E, Evidente VGH. Hemichorea-hemiballismus may respond to topiramate. *Clin Neuropharmacol*. 2005; **28**(3):142–144.