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RESEARCH ARTICLE

CASE REPORT: HEMICHOREA-HEMIBALLISMUS IN NONKETOTIC HYPERGLYCEMIC FEMALE PATIENT: PUTAMINAL HIGH INTENSITY LESION ON MRI T1-WEIGHTED IMAGES

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Abstract

A 65-year-old female known case of diabetes mellitus type 2, hypertension, chronic heart failure and chronic renal failure was admitted to our hospital with fever, diarrhea, and sudden onset of hemichorea-hemiballismus in right upper limb and lower limb. On admission blood glucose level was 175mg/dl, hemoglobin A1c was 11% and the serum osmolality was 292 mOsm/l. Urine analysis was negative for glucose and ketone bodies. After correction of blood glucose to normal level choreiform and ballistic movements disappeared. MRI showed high intensity on T1-weighted images in the left putamen, which disappeared 4 months after onset. The putaminal abnormality was the cause of her involuntary movements. The cause of high intensity in the putamen might be the multiple petechial hemorrhages which can occur in association with nonketotic hyperglycemia. We report an occurrence of hemichorea-hemiballismus in a female patient with diabetes mellitus type 2.

Introduction

Case Presentation

A 65-year-old female presented with complaints of fever, diarrhea and sudden onset of involuntary flinging and jerky movements of right upperand lower limbsfor 15 days with increased intensity since last 2 days. Movement was associated with seizure, and it was decreasing during sleep. There was no history of any such abnormal involuntary movements, focal neurological deficit, convulsions, altered sensorium or altered level of consciousness. Her medical history included poorly controlled diabetes mellitus on metformin, hypertension on amlodipine and chronic kidney disease with protein restricted diet. She did not take any other medications. She was a non-alcoholic and non-smoker.

On admission, vitals appeared normal (Body temperature-37.5^{0}C, Pulse rate-82/min, Respiratory rate-18/min and regular, Blood pressure-136/88 mm of Hg). On neurological examination, choreiform and flinging type of involuntary jerky movements of her right upper and lower limbs were observed. Hypotonia was noted on right side. Higher functions, reflexes, sensation, and rest of motor system examination was normal. Other systemic examination was also normal.

Investigations

Blood examination revealed : Haemoglobin: 11.5 g/dL (slightly low), Total White Blood Cell Count: 7,200 cells/mm^3 (within normal range), Serum Urea: 74 mg/dL (elevated), Serum Creatinine: 2.27 g/dL (elevated), Serum Sodium: 136 mmol/L (normal), Serum Potassium: 4.9 mmol/L (normal), Total Bilirubin: 0.6 mg/dL (normal), Direct

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Bilirubin: 0.2 mg/dL (normal), Indirect Bilirubin: 0.4 mg/dL (normal), Serum Calcium (Total): 8.9 mg/dL (normal), Serum Phosphorus: 3.9 mg/dL (normal), Uric Acid: 14.4 mg/dL (elevated), Erythrocyte Sedimentation Rate (ESR): 84 mm/h (elevated), Serum Glucose: 175 mg/dL and HbA1c: 11.0% (indicating poor glycemic control)

The chest X-ray (PA view), liver function tests, abdominal ultrasonography, blood and urine cultures, and ECG appeared to be within normal limits. Urine analysis was normal and negative for glucose or ketone bodies.

On non contrast MRI of brain(Figure 1), unilateral T1-weighted hyperintensity was seen within the left putamen, however with no significant corresponding signal changes in rest of the sequences. No mass effect or peri-lesional edema was seen. Subtle enhancement in left putaminalregion was seen in contrast enhanced T1-weighted images (Figure 2).Periventricular hyperintensities on T2 weighted and Fluid attenuated inversion recovery (FLAIR) images (Figure 3) werereported in bilateral frontal periventricular white matter, suggestive of deep white matter small vessel ischemic changes. The corresponding MRI images are as depicted below.

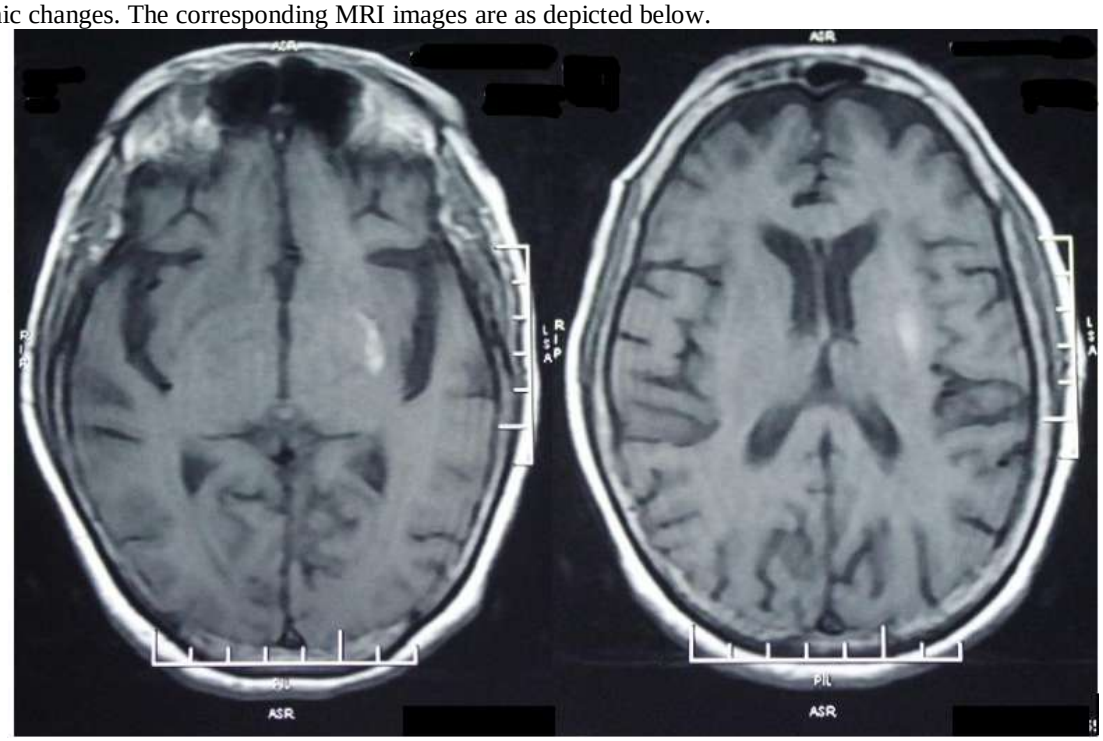

Figure 1:- Axial T1W images show left putaminal discretehyperintensity without any signs of acute surrounding edema.

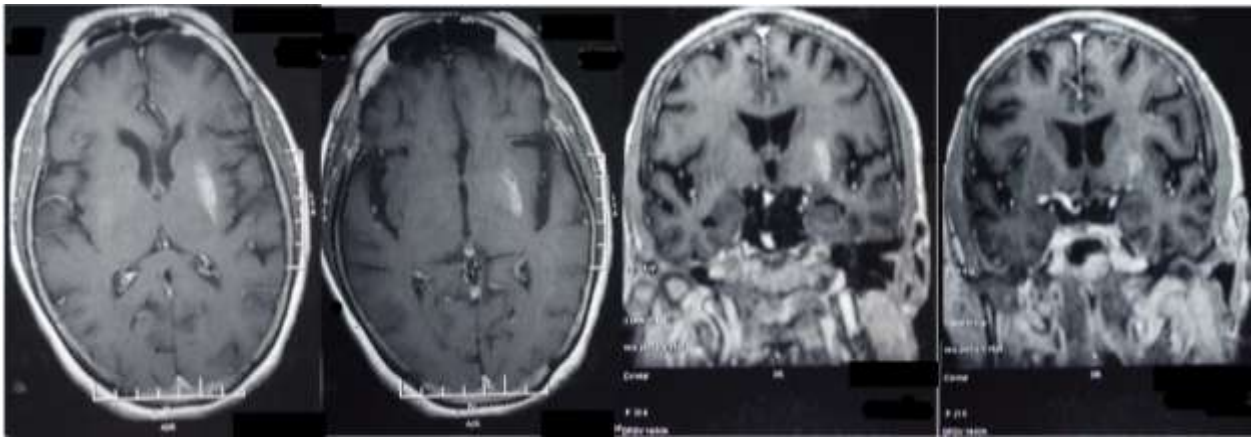

Figure 2:- Axial (*left*) and Coronal (*right*) post contrast T1W images showing mild subtle enhancement of the left putamen region.

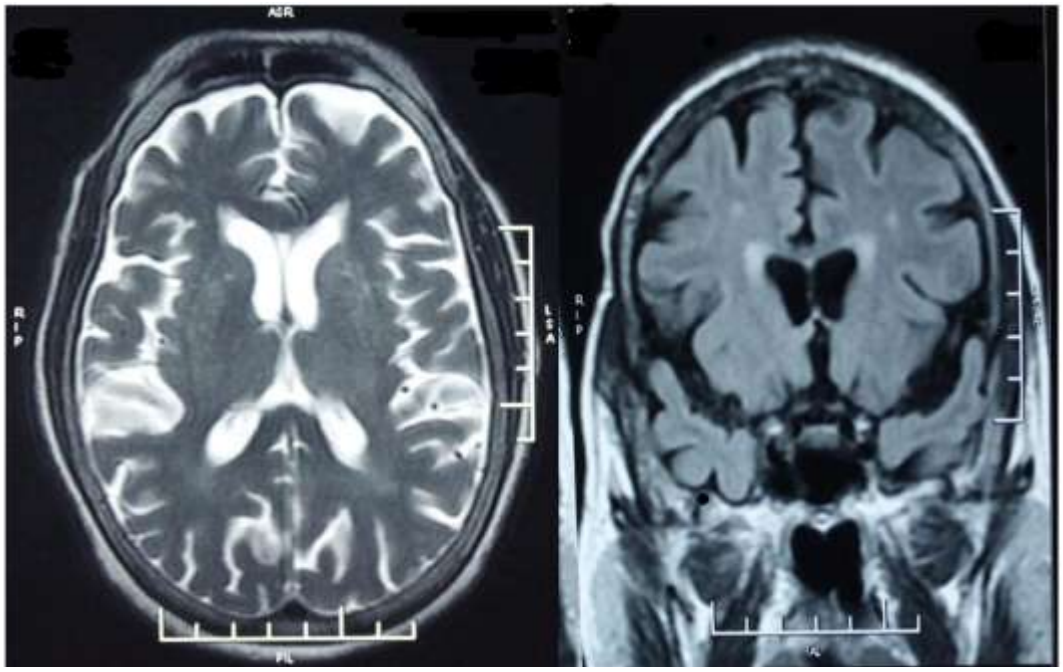

Figure 3:- Axial T2 (*left*) and FLAIR (*right*) coronal images show no corresponding abnormal signal in the left putamen region. Chronic periventricular ischemic changes can be seen on FLAIR images.

Cerebrospinal fluid (CSF) analysis revealed a protein level of 35.3 mg/dL (within normal range) and a glucose level of 96 mg/dL (elevated). Physical and microscopic examinations of the CSF were unremarkable. No organisms were detected on Gram stain, and acid-fast bacilli were not observed on Ziehl-Neelsen staining.

Echocardiography showed findings consistent with dilated cardiomyopathy, characterized by global left ventricular hypokinesis and a reduced left ventricular ejection fraction of 30%, indicating chronic heart failure. There was severe left ventricular systolic dysfunction and mild pulmonary hypertension, but no calcifications were noted.

Treatment

The patient was managed with symptomatic treatment, which included the following medications: metformin (500 mg) twice daily, aspirin (325 mg) once daily, clopidogrel (75 mg) once daily, atorvastatin (40 mg) at bedtime, valproate (200 mg) three times daily, and famotidine (20 mg) twice daily. Dietary modifications were advised to address chronic renal failure, hypertension, and diabetes both at admission and upon discharge.

A follow-up after two months showed no signs of involuntary movements. The patient's glucose level was 136 mg/dL, serum creatinine was 1.60 mg/dL, and her blood pressure was 140/82 mmHg. Her management plan for diabetes, hypertension, congestive heart failure, and chronic renal failure was continued.

Discussion

Hemichorea-hemiballismus is relatively common among Asian diabetic women, with nonketotic hyperglycemia being an often underrecognized cause. This is largely due to a lack of awareness and the fact that the condition can resolve once blood glucose levels are normalized.[1,2] In some cases, as seen in our patient, MRI findings may show T1 hyperintensity and T2 low or high signal intensity, which has been reported in association with nonketotic hyperglycemia.[3] The exact pathophysiological mechanism behind these imaging findings remains unclear. T1 hyperintensity is most likely linked to petechial hemorrhages, while T1 hypo intensity may indicate microcalcifications or the deposition of other metabolic minerals.[4,5] Additionally, hyperintensity on T2 and diffusion-weighted imaging (DWI) is typically associated with edema.[6]

The pathophysiology of hyperglycemic chorea-ballismus is not fully understood, and no single mechanism has been identified to explain it.[2,7] Hyperglycemia, in the absence of ketoacidosis, can impair regional blood flow, which may disrupt the Krebs cycle in the brain and lead to increased depletion of GABA through the succinate

semialdehyde pathway.[8] This metabolic disturbance can result in disinhibition of dopamine pathways in the basal ganglia, causing dopaminergic hyperactivity. Older women, particularly postmenopausal individuals, are more susceptible due to estrogen deficiency, which can increase dopamine hypersensitivity.[8,9] Additionally, petechial hemorrhages from hyperglycemia, cerebral ischemia, and diabetic microangiopathy may contribute to the condition. Another potential mechanism involves acute dysfunction of the putamen, which may lead to Wallerian degeneration of the putaminal white matter, a phenomenon also considered to play a role in nonketotic hemichorea-hemiballismus.[10]

Nonketotic hemichorea-hemiballismus can either present as the initial symptom of hyperglycemia or develop as a complication of poorly controlled diabetes, as seen in our case. [7,11,12]In some instances, rapid correction of hyperglycemia itself can trigger chorea.[3] The primary treatment is to control blood glucose levels, which often leads to partial or complete resolution of hemichorea-hemiballismus.[13]However, both clinical and radiological improvements may take time, sometimes up to six months, even after blood glucose is normalized.[13] When hyperglycemia is confirmed as the cause of hemichorea-hemiballismus, most patients show gradual recovery from their hyperkinetic movements once their blood glucose is properly managed.[14]

Nonketotic hemichorea-hemiballismus generally has an excellent prognosis, though there are some exceptions.[12] In our patient, after a two-month follow-up, she showed no signs of hyperkinetic activity and was doing well. In this case, the patient's noncompliance with her diabetic medications and lack of dietary restrictions may have contributed to the development of nonketotic hemichorea-hemiballismus at her age. This was the first recorded instance of hemichorea-hemiballismus in our hospital in a patient with poorly controlled diabetes.

References

1. Dewey RB Jr, Jankovic J. Hemiballismus-hemichorea. Clinical and pharmacologic findings in 21 patients. Arch Neurol 1989; 46:862-7.
2. S. Lee, J. Shin, J. Kim et al., "Chorea-ballism associated with nonketotic hyperglycemia or diabetic ketoacidosis: characteristics of 25 patients in Korea," Diabetes Research and Clinical Practice, vol. 93, no. 2, pp. e80–e83, 2011.
3. Nagai, C., Kato, T., Katagiri, T. and Sasaki, H. (1995) Hyper intense putamen on T1-weighted MR images in a case of chorea with hyperglycemia. American Journal of Neuroradiology, 16, 1243-1246.
4. Chang, M.H., Chiang, H.T., Lai, P.H., et al. (1997) Putamenal petechial hemorrhage as the cause of chorea: A neuroimaging study. Journal of Neurology, Neurosurgery, and Psychiatry, 63, 300-303. doi:10.1136/jnnp.63.3.300
5. Cherian, A., Thomas, B. and Baheti, N.N., Chemmanam T. and Kesavadas C. (2009) Concepts and controversies in nonketotic hyperglycemia-induced hemichorea: Further evidence from susceptibility-weighted MR imaging. Journal of Magnetic Resonance Imaging, 29, 699-703. doi:10.1002/jmri.21672
6. Chu, K., Kang, D.-W., Kim, D.-E., Park, S.-H. and Roh, J.-K. (2002) Diffusion-weighted and gradient echo magnetic resonance findings of hemichorea-hemiballismus associated with diabetic hyperglycemia: A hyper viscosity syndrome? Archives of Neurology, 59, 448-452. doi:10.1001/archneur.59.3.448
7. C. Song, X. Yang, G. Xing et al."Hemichorea associated with nonketotic hyperglycemia in a female," Neuro Endocrinology Letter, vol. 33, no. 5, pp. 489–492, 2012.
8. Mestre TA, Ferreira JJ, Pimentel J. Putamenal petechial hemorrhage as the cause of nonketotic hyperglycemic chorea: A neuropathological case correlated with MRI findings. J Neurol Neurosurgery Psychiatry 2007; 78:549-50.
9. Chu K, Kang DW, Kim DE, Park SH, Roh JK. Diffusion weighted and gradient echo magnetic resonance findings of hemichorea-hemiballismus associated with diabetic hyperglycemia : A hyper viscosity syndrome? Arch Neurol 2002;59:448-52.
10. M.Wintermark,N.J.Fischbein,P.Mukherjee, E.L.YuhandW. P. Dillon, "Unilateral putamenal CT, MR, and diffusion abnormalities secondary to nonketotic hyperglycemia in the setting of acute neurologic symptoms mimicking stroke," American Journal of Neuroradiology, vol. 25, no. 6, pp. 975–976, 2004.
11. C. V. Chang, A. C. Felicio, C. Godeiroet al., "Chorea-ballism as a manifestation of decompensated type 2 diabetes mellitus," American Journal of the Medical Sciences, vol. 333, no. 3, pp. 175– 177, 2007.
12. D. Branca, O. Gervasio, E. le Piane, C. Russo, and U. Aguglia, "Chorea induced by non-ketotic hyperglycemia: a case report," Neurological Sciences, vol. 26, no. 4, pp. 275–277, 2005.
13. Hwang KJ, Hong IK, Ahn TB et al: Cortical hemichorea-hemiballismus. J Neurol, 2013; 260(12): 2986–92
14. Lin JJ, Chang MK: Hemiballism-hemichorea and non-ketotichyperglycemia. J Neurol Neurosurgery Psychiatry, 1994; 57(6): 748–5015.