Magnetic resonance imaging volumetric analysis for diabetic striatopathy with two episodes of hemichorea-hemiballism syndrome

A case report

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Abstract

Rationale: Diabetic striatopathy (DS) is an uncommon movement disorder among diabetic patients characterized by clinical hemichorea-hemiballism with neuroimage change of the striatum. Here, we report a case of DS with relapsed hemichorea-hemiballism attacks even during euglycemic period, and the MRI changes by volumetric analysis.

Patient concerns: A 69-year-old diabetic female suffered from a relapsed episode of hemichorea-hemiballism during her euglycemic period after the treatment of hyperglycemia.

Diagnoses: To investigate the serial MRI changes in a case with diabetic striatopathy who had clinical hemichorea-hemiballism syndrome.

Interventions: Semi-quantitative volumetric analyses from T1 images of these brain MRIs were obtained during the disease course.

Outcomes: Besides, the negative finding of the first brain MRI during her first hospital admission, three afterward MRI examinations disclosed a waxing-and-waning mode of volume change from high-signal T1 images in left striatum. The clinical symptoms paralleled with the neuroimage changes in striatum. The MR signal volume changes were valuable for the clinical course of the hemichorea-hemiballism caused by diabetic striatopathy

Lessons: Serial MR images for the diabetic striatopathy presented as a key pathognomonic relationship with the clinical hemichorea-hemiballism syndrome, assessed by our simplified volumetric analysis. Clinical involuntary movements may relapse and persist even with euglycemic condition as our case.

Abbreviations: DS = diabetic striatopathy, HHS = hyperosmolar hyperglycemic state, MRI = magnetic resonance imaging.

Keywords: diabetic striatopathy, MRI volumetric analysis, recurrent hemichorea-hemiballism syndrome

1. Introduction

Limb hemiballistic or hemichoreiform movements are uncommon manifestations of diabetes mellitus that occur during the hyperglycemic or euglycemic period\textsuperscript{[1,2]} and also during hypoglycemic episodes,\textsuperscript{[3]} with delayed onset\textsuperscript{[3]} and a persistent course.\textsuperscript{[4]} Diabetic striatopathy (DS) is characterized by unilateral hyperkinesia with contralateral basal ganglion changes, which are visualized using T1-weighted magnetic resonance imaging (MRI).\textsuperscript{[5]} Herein, after getting informed consent we report a case of DS with 2 clinical episodes of hemichorea-hemiballism even when blood sugar levels were under control. We also recorded the hyperintense volumetric changes of DS from serial MRI images.

2. Case report

A woman aged 69 years presented with a 15-year history of type 2 diabetes mellitus. She had poor adherence to the anti-diabetic medicines during the past year and her blood sugar levels fluctuated. She experienced general weakness, appetite loss, and right lower limb tremors 1 week prior to her first hospital stay. On admission, she had hyperglycemia (blood glucose level, 718 mg/dl), complicated by a hyperosmolar hyperglycemic state (HHS). Right hemichorea-hemiballism occurred on the second day after admission. Electroencephalography did not record epileptiform discharges. One week later, the patient recovered from the hemichorea-hemiballism and was able to walk without assistance. She discharged without clinical sequelae after 16 days from the initial admission. However, 2 weeks later, the patient’s
right hemichorea-hemiballism relapsed and she returned to our ward. Her brain MRI (Fig. 1A) revealed a prominent high-intensity lesion of the left basal ganglion on T1-weighted images. The patient initially received valproic acid and incremental doses of haloperidol according to her symptoms, with the later addition of topiramate and quetiapine since high-doses haloperidol did not elicit satisfactory responses. She discharged 26 days after her 2nd hospital stay, although mild hemichoreiform symptoms persisted. She visited our OPD clinics regularly, and full recovery from the hemichoreiform symptoms was reported 3 months after the second hospital admission. Two subsequent MRI examinations for DS revealed smaller and fainter signal changes in the left basal ganglion (Fig. 1B and C).

3. MRI volumetric analyses
This patient took the brain MRI series for her DS by using a 1.5T MRI Scanner, MAGNETOM Aera, Siemens, Erlangen Germany. Using semi-quantitative analyses, we recorded at least 3 image sections wherein a signal change on the T1-serial images from the left basal ganglion was recorded. We subsequently added up each area and multiplied this value by the thickness of the sections to obtain the calculated volume data with signal changes.

Three serial brain MRI images (T1 FLAIR) taken on Day 3, 40 and in Month 3.5 after the initial hemichorea-hemiballism event were subjected to the analyses. The average calculated volumes of the coronal and horizontal images fluctuated in magnitude and were as follows: 5356, 13042, and 4069 mm³. The estimated highest volume with hyperintensity T1 signal changes occurred about at 40 days after her initial admission, within the period of the recurrence of the hemichoreiform-hemiballistic event.

4. Discussion
DS is a rare manifestation in diabetic patients with HHS,[6] with a clinical manifestation of contralateral hemichorea-hemiballism. Bilateral limb movements occur with bilateral striatal lesions.[7] Our patient’s movement disorder lasted for more than 3 months, even when her blood sugar levels were under control using insulin and anti-diabetic agents. These symptoms may be attributable to the change in DS revealed by the brain MRI. We reported this case due to the following 3 clinical interesting issues: relapsed course of hemichorea-hemiballism, euglycemic condition throughout the course of relapse, and chronological MRI signal changes by semi-quantitative volumetric analyses. Clinical movement disorder in this patient occurred with pathognomonic contralateral DS lesion in the basal ganglion, sparing the caudate nucleus. However, the evolution of MRI changes does not explain why there was a symptom-free period between two attacks of hemichorea-hemiballism. It is well-known that hemichorea-hemiballism occurs not only during HHS, but even in cases where hyperglycemia is improved[1,3,8] or during hypoglycemic conditions.[2]

The cause of the DS lesions is not well known. Ischemic vasculopathy may be the first possibility, but some cases with clinical choreiform movement disorder did not reveal MRI changes.[6] One recent case revealed small lacunar infarcts with reactive astrocytosis and neuropeptide Y immune-reactive neurons in the putamen on autopsy.[3] Biopsy of another case disclosed patchy necrosis, thickened arteriole wall and narrowing of the vessel lumen.[9] In our case, we did not have tissue information for her DS, but the serial MRI signal changes, along with the semi-quantitative volumetric analysis, indicated a clinical improvement.

Pharmacologic management by dopamine depletion agents, such as haloperidol or other major tranquilizers, are the first choice to control the hyperkinetic movement disorder associated with DS. We attempted to control the hyperkinesia exhibited by our patient with the administration of anti-epileptic agents on the initial impression of epilepsia partialis continua (EPC) which may mimic this involuntary movement disorder present in diabetic
patients. EPC may present cortical epileptiform discharges resulting in myoclonic jerks in the contralateral limbs. On the other way, non-pharmacological treatments for severe diabetic hemichorea-hemiballism syndrome, such as repetitive transcranial magnetic stimulation and deep brain stimulation have been reported.

DS is one of the consequences of the CNS involvement of diabetes mellitus and is a key clinical issue associated with movement disorder. A neurochemical hypothesis has been discussed. Further clinical investigation by using molecular imaging or pathological examination may be necessary for a large-scale study.

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