Putaminal Changes before the Onset of Clinical Symptoms in Diabetic Hemichorea-hemiballism

Nobuhito Nakajima1, Masayuki Ueda1, Hiroshi Nagayama2 and Yasuo Katayama2

Abstract

An 81-year-old woman with poorly controlled diabetes mellitus was hospitalized due to hemichorea-hemiballism (DHC-HB). A radiological examination revealed typical putaminal changes of diabetic hemichorea-hemiballism (DHC-HB). Interestingly, brain computed tomography, performed before symptom onset, disclosed a hyperdense lesion in the left basal ganglia, indicating persistent basal ganglia impairment, even before the onset of symptoms, under sustained hyperglycemia. Additionally, an increase in the cerebrospinal fluid level of homovanillic acid was related to the symptom appearance of DHC-HB. Pronounced potential basal ganglia impairment under hyperglycemia and central dopaminergic hyperactivity was important for the development of DHC-HB in this patient.

Key words: diabetes mellitus, hyperglycemia, diabetic hemichorea-hemiballism, homovanillic acid, involuntary movement

(Intern Med 53: 489-491, 2014)
(DOI: 10.2169/internalmedicine.53.1359)

Introduction

Diabetic hemichorea-hemiballism (DHC-HB) is a disorder characterized by unilateral abnormal involuntary movements. It is most often found in patients with diabetes and extreme hyperglycemia. The major radiological features of DHC-HB are contralateral putaminal abnormalities that typically include hyperintensity on T1-weighted magnetic resonance imaging (T1WI) and hyperdensity on computed tomography (CT) (1, 2). Whether putaminal abnormalities occur before the onset of DHC-HB, however, remains unclear. We herein report a case of DHC-HB with the early onset of putaminal abnormalities.

Case Report

An 81-year-old woman with poorly controlled diabetes mellitus was hospitalized due to abnormal involuntary movements in her right limbs that persisted for one week. Blood tests showed marked hyperglycemia (535 mg/dL) with an elevated HbA1c level (11.8%). A neurological examination performed on admission revealed choreiform movements with irregular twitching in the right limbs, including balistiform movements in the right leg. Brain CT performed seven days after symptom onset revealed slight left putaminal hyperdensity, and T1WI revealed left putaminal hyperintensity (Figure A, B). The patient had complained of a headache 40 days before disease onset, and brain CT was performed by chance at that time. The CT images were retrospectively examined and found to exhibit putaminal abnormalities similar to those observed after symptom onset (Figure C). The presence of hyperglycemia remained unclear at that time because blood tests were not performed.

Based on these findings, the patient was diagnosed with DHC-HB. She was treated with intensive insulin therapy and haloperidol (3 mg/day). The DHC-HB symptoms disappeared four weeks after onset. After switching the insulin therapy to oral antidiabetic drugs, her blood glucose level surged, and DHC-HB relapsed on the right side 78 days after the initial onset. However, no left putaminal abnormalities were observed on follow-up CT performed 97 days after the initial onset (Figure D). Because the DHC-HB reignited despite an improvement in the radiological findings, further examinations were performed. A cerebrospinal fluid (CSF)
examination showed that the CSF level of homovanillic acid (HVA) was 92.9 ng/mL at relapse. After achieving intensive diabetic control, the DHC-HB disappeared again 119 days after the initial onset. The CSF-HVA level dropped to 51.9 ng/mL at 132 days after the initial onset, regardless of the administration of a stable daily dose of haloperidol.

**Discussion**

The patient displayed a putaminal CT abnormality prior to the onset of DHC-HB. To our knowledge, putaminal changes preceding symptom onset in cases of DHC-HB have only been previously reported in one patient who exhibited hyperdensity in the striatum 14 days before symptom onset (3). The precise pathogenesis underlying the occurrence of putaminal abnormalities on CT and T1WI remains unclear. These radiological changes are primarily considered to involve increased reactive astrocytes in the basal ganglia, and it is also known that the lesions occasionally contain small areas of hemorrhage and/or infarction (4, 5). In the present patient, DHC-HB relapsed even after the disappearance of the CT abnormality, which was observed prior to symptom onset. A case of DHC-HB without putaminal abnormalities has also been reported (6). These findings indicate that functional impairments rather than visible radiological changes in the basal ganglia are important for the development of DHC-HB, even though the radiological findings of DHC-HB may vary among the examinations. Furthermore, hyperglycemia may exacerbate potential basal ganglia impairments, as intensive diabetic control is effective against DHC-HB.

Hemichorea and hemiballism are considered to be associated with subthalamic inactivation (7). DHC-HB is also related to insufficient inhibition from the internal segment of the globus pallidus to the thalamus, reportedly due to the depletion of gamma-aminobutyric acid (8). However, other
studies have shown that dopaminergic hyperactivity is involved in hemichorea and hemiballism of other etiologies (9). Indeed, the CSF-HVA level was decreased in the present patient when the DHC-HB subsided. The elevated CSF-HVA level together with the pharmacological effects of neuroleptics suggests an association between dopaminergic hyperactivity and DHC-HB in this patient. However, the CSF-HVA data should be interpreted cautiously because the significance of CSF-HVA has not been established, even in Parkinson’s disease. Further studies involving the characteristics of CSF-HVA for DHC-HB are needed to clarify this possibility.

In conclusion, our observations in this patient indicated persistent basal ganglia impairment, even before the onset of symptoms under sustained hyperglycemia. Additionally, the presence of pronounced potential basal ganglia impairment under hyperglycemia and central dopaminergic hyperactivity is important for the development of DHC-HB.

The authors state that they have no Conflict of Interest (COI).

References