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Case Report

Hemichorea in a diabetes mellitus patient following acute ischemic stroke with changes in regional cerebral blood flow

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Abstract

It is not unusual to observe hemichorea in patients with diabetes mellitus, with origins attributable to recent ischemia. Our patient was a 66-year-old female with diabetes mellitus who suddenly developed right hemichorea, mild muscle weakness of the right upper extremity, ideational apraxia, and acalculia. Her blood glucose was 600 mg/dL, and HbA1c was 13.3%. After the patient underwent head magnetic resonance imaging (MRI), a new cerebral infarction was observed in the left frontal lobe, and treatment was started with edaravone and cilostazol. At the same time, insulin treatment was also started for hyperglycemia. The acalculia and ideational apraxia improved approximately 1 week after treatment initiated, and the hemichorea also decreased. ECD-SPECT was performed on admission, and it was observed that blood flow was decreased in the left frontal lobe and striatum, but increased in the thalamus; two weeks later on follow-up ECD-SPECT, blood flow had increased slightly in the left forebrain and striatum, while it had decreased slightly in the thalamus. This suggests that the cause of hemichorea was related to ischemia. When the activity of the pallidum is impaired, it is presumed that the inhibitory activity towards the thalamus weakens and the thalamic cells become over-excited, causing chorea.

Keywords: diabetes mellitus; hemichorea; cerebral infarction; regional cerebral blood flow; ECD-SPECT

1. Introduction

It is well-known that involuntary movements can accompany diabetes mellitus (DM)[1,2] and metabolic disorders of the subthalamic nucleus and/or striatum. Ischemia is generally considered responsible for this phenomenon.[3] However, to the best of our knowledge, there have been no previous reports of diabetic patients exhibiting chorea accompanied by changes in regional cerebral blood flow (rCBF). We herein describe a case of DM in which hemichorea was triggered by cerebral infarction and remission was achieved with treatment. This patient's course of disease and treatment supported the notion that ischemia was involved. In the present case, performance of single photon emission computed tomography using the ^{99m}Tc-ECD Patlak Plot method (ECD-SPECT)[4] in the acute phase revealed decreased blood flow in the left striatum and increased blood flow in the left thalamus; a repeat ECD-SPECT study performed 2 weeks later revealed that both involved areas were smaller than they were on the first ECD-SPECT study. To the best of our knowledge, this is the first report of a diabetic patient exhibiting chorea with changes in rCBF in the acute and postacute phases.

2. Case report

The patient was a 66-year-old female who had no history of cigarette smoking or alcohol drinking. Her past medical history included hypertension, hyperlipidemia, and DM discovered several years before this presentation, all of which the

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patient had ignored and which had therefore remained untreated. On September 11, 2010, abnormal behaviors were noted, including taking a bath with her undergarments on and becoming taciturn, and development of involuntary movements of the right hand and foot. A family member observed the aberrant behavior for a few days, but the symptoms did not improve. The patient and her husband consulted members of our department and she was hospitalized on September 14, 2010 (on day 4).

On physical examination, the patient was slightly obese, with a height of 154 cm and a weight of 66.2 kg. Her temperature was 35.8°C, blood pressure was 208/108 mmHg, and her pulse rate was 94/minute and regular. No abnormalities were noted on general physical examination. Her consciousness was clear, but detailed neurological examination revealed ideational apraxia and acalculia. Slight muscle weakness of the right upper extremity was noted. There were no abnormalities of tendon reflexes, coordinated movements, the sensory system, or ambulation. Hemichorea involving the right upper and lower extremities was observed. Blood cell counts were within the normal range. Total cholesterol was 231 mg/dL, the triglyceride level was 168 mg/dL, blood glucose was 600 mg/dL, and the HbA1c was 13.3%. Urinalysis results included protein (−), glucose (+++), and ketones (−). On head magnetic resonance imaging (MRI) on admission (on day 4), a recent cerebral infarction was observed in the left frontal lobe. A high signal intensity lesion was observed in the left frontal lobe on T2-weighted imaging (T2WI) (Fig. 1B), fluid-attenuated inversion recovery (FLAIR) (Fig. 1C), and diffusion-weighted imaging (Fig. 1D); a low signal intensity lesion was seen on apparent diffusion coefficient (ADC)-map imaging (Fig. 1F). Moreover, the left striatum showed a high signal intensity on T1-weighted imaging (T1WI) (Fig. 1A) and a low signal intensity on T2WI (Fig. 1B), and there was a mild low signal intensity region on T2* (Fig. 1E). On magnetic resonance angiography (MRA) performed on day 4, an oozing-like intensity area from the left middle cerebral artery was observed. This was a region of medium intensity having an irregular border (Fig. 2). ECD-SPECT was performed on day 4 and showed decreased cerebral blood flow not only in the left frontal lobe but also in the left striatum (Fig. 3). Based on these results, a diagnosis of acute-phase cerebral infarction was made, and treatment with edaravone (60 mg per day, intravenous, from day 4 till day 14) and cilostazol (200 mg per day, oral administration) was started. At the same time, insulin treatment for hyperglycemia was also started. The acalculia and ideational apraxia improved approximately 1 week after treatment had begun, and the hemichorea also decreased. Two weeks after onset of symptoms (on day 18), when the hemichorea had almost disappeared, a second ECD-SPECT was performed; this revealed increased blood flow in the forebrain and striatum, as well as reduced blood flow in the thalamus as compared with the initial examination (Fig. 3).

3. Discussion

Suggested causes of chorea in DM patients include metabolic disorders caused by hyperglycemia,[2] as well as ischemia[5,6] and bleeding[7] in the striatum. There are several reports mentioning the involvement of cerebral ischemia.

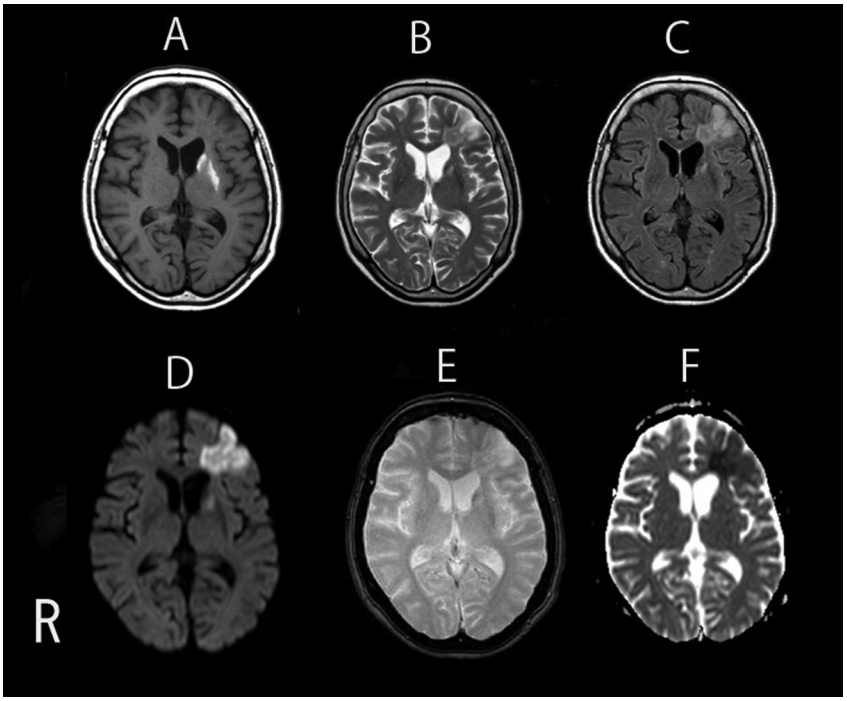

Fig. 1. Admission head MRI: (A) T1-weighted imaging (T1WI); (B) T2-weighted imaging (T2WI); (C) fluid-attenuated inversion recovery (FLAIR); (D) diffusion-weighted imaging; (E) T2* imaging; (F) apparent diffusion coefficient (ADC)-map imaging. The MRI findings are interpreted as: (a) old left striatal lesion with negative pressure effect; (b) hyperglycemic effect on the left striatum (high signal intensity in T1WI, nearly normal signal change in T2WI) (Figs. A and B); (c) acute infarction in the left frontal lobe (Figs. D and F).

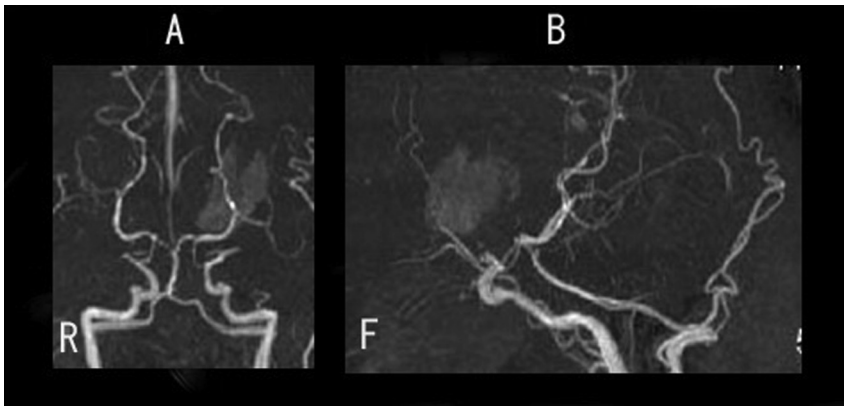

Fig. 2. Admission head MRA: (A) frontal view; (B) lateral view. An oozing-like intensity area from the left middle cerebral artery is observed; this is a region of medium intensity having an irregular border.

Ohara et al. conducted autopsies on DM patients exhibiting chorea and reported the presence of recent cerebral infarctions.[5] Pantano et al. reported the case of an 80-year-old man who developed right-sided hemichorea after a lacunar stroke of the left internal capsule, and described increased blood flow in the left thalamus in the postacute phase (on day 18 after clinical onset) when hemichorea was still present.[8] Kim et al. reported four cases of hemichorea with acute cerebral infarction. The sites of cerebral infarctions were the putamen, globus pallidus, caudate nucleus, and subthalamic nucleus. In these patients, ECD-SPECT was performed in the acute phase (on days 3–7) and showed decreased blood flow in the basal nucleus and increased blood flow in the thalamus on the contralateral side.[9] However, to the best of our knowledge, there have been no reports of diabetic patients exhibiting chorea with changes in rCBF in the acute and postacute phases. This is the first report of rCBF in both the acute and postacute phases of a diabetic patient exhibiting chorea triggered by an acute stroke. In this case, hemichorea almost disappeared in the postacute phase. ECD-SPECT is very useful for detecting[9–11] and evaluating[10,12] changes in neurological diseases. In this patient, after ECD-SPECT was

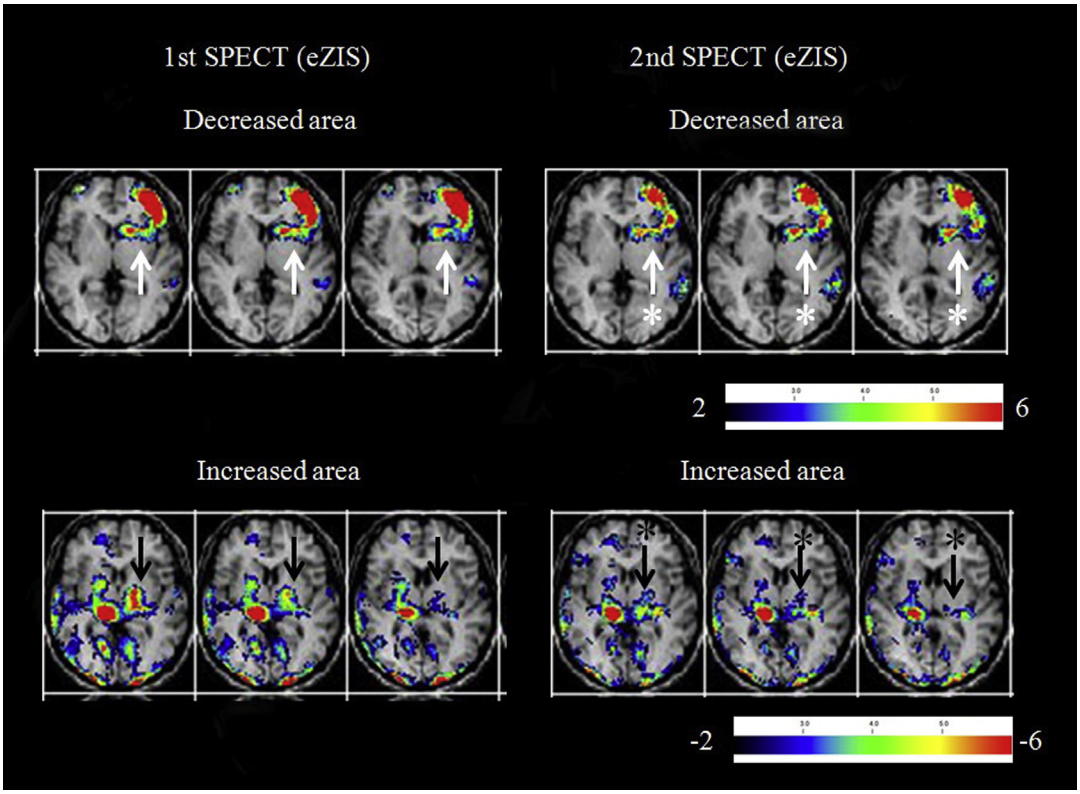

Fig. 3. ECD-SPECT using easy Z-score imaging system (eZIS).[4] A positive Z-score indicates an area of decreased regional cerebral blood flow (rCBF) (upper column), and a negative Z-score indicates an area of increased rCBF (lower column) in the patient compared with the control database. Focusing on the left hemisphere, the first ECD-SPECT shows a relative decrease in the left striatum (⟹) and an increase in the left thalamus (➡). The second ECD-SPECT shows that these areas in the left striatum (*⟹) and in the left thalamus (*➡) are smaller than those seen on the admission ECD-SPECT.

performed, the easy Z score imaging system (eZIS)[4] was applied for quantitative analysis of brain perfusion. With this method, affected areas with decreased or increased rCBF can be detected easily, precisely, and objectively.

In the current case, because high signal intensity was seen in T1WI, and nearly normal signal change in T2WI (Figures A and B), it was speculated that right hemichorea was primarily caused by a hyperglycemic effect on the left striatum. Furthermore, decreased blood flow in the left striatum in the acute phase could be a trigger which caused this right hemichorea. On the 1st ECD-SPECT, blood flow in the left frontal lobe and the left striatum decreased, while blood flow in the thalamus increased. In addition, the reduced blood flow in the left forebrain and striatum had increased on ECD-SPECT 2 weeks later, while blood flow in the thalamus had decreased. According to these results, though acute infarction of the left striatum was not found, a penumbra could have been present in this area in the acute phase. Therefore, blood flow in the left striatum had increased in the postacute phase. This suggests that the cause of hemichorea in the present case was related to ischemia. When the activity of the striatum is impaired, it is presumed that inhibitory input to the thalamus weakens and the thalamic cells become over-excited, causing chorea. In the postacute phase, recovery of the reduced blood flow in the striatum might improve the hemichorea. On MRA, an oozing-like intensity area from surrounding blood vessels was observed; this was a region of medium intensity having an irregular border. There are reports that describe this as a microhemorrhage, but the precise details regarding the cause of this phenomenon remain unclear. There are also previous reports that chorea is caused when gradual bleeding occurs in the putamen.[7] In the present case, the left striatum may have been injured by a slow microhemorrhage before the onset of cerebral infarction; this may have precipitated the onset of hemichorea.

In conclusion, regarding our reported case of a diabetic patient exhibiting chorea triggered by an acute stroke with changes in rCBF in the acute and postacute phases, the cause of hemichorea appeared to be related to ischemia.

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