Hemichorea in Hyperglycemia Associated with Increased Blood Flow in the Contralateral Striatum and Thalamus

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We studied a patient with hyperglycemia who developed choreic involuntary movements in the right extremities using single photon emission computed tomography (SPECT) with 123I-N-isopropyl-p-iodoamphetamine. SPECT revealed an increased blood flow in the left striatum and thalamus. Through the control of blood glucose and the administration of haloperidol, the hemichorea was resolved, and the increased blood flow in the striatum and thalamus disappeared. These findings suggest that the increased blood flow, which probably indicates increased neuron activity in the striatum and thalamus, is an underlying pathophysiological state in hemichorea.

Key words: diabetes mellitus, IMP-SPECT, MRI

Introduction

Although chorea is a common hyperkinetic movement disorder, the specific location of its pathologic process is still under debate. We report here a patient with hemichorea associated with increased blood flow on single photon emission computed tomography (SPECT) with 123I-N-isopropyl-p-iodoamphetamine (123I-IMP) in the contralateral striatum and thalamus, and discuss the pathophysiological mechanisms.

Case Report

On August 10, 1992, a 78-year-old woman, known to have diabetes mellitus, presented with a 4-day history of involuntary movements in the right extremities. She had no family history of movement disorders. On examination, she was alert and well oriented. Her intelligence was normal. In the right upper and lower extremities, choreic involuntary movements were present during the resting state. The movements were decreased by activity and disappeared during sleep. Cerebellar ataxia did not exist and gross muscle strength was normal in the four extremities. The deep tendon reflexes showed no laterality and there was no Babinski’s sign. Sensation was intact. The following laboratory data were notable: fasting blood glucose 401 mg/dl, HbA1c 15.1%, 24-hour urine glucose 48.1 g, ketones 1566 \( \mu \text{MOL/L} \) (normal <130 \( \mu \text{MOL/L} \) ) and serum creatine phosphokinase 1701U/L. Normal laboratory investigations included bilirubin, ammonia, calcium, phosphorus, other electrolytes, and parathyroid and thyroid functions. Her serum osmolarity was 291 mOsm/L and blood pH was 7.42. The electroencephalogram revealed a slightly increased amplitude of basic activities in the left frontal region but no paroxysmal activities. An X-ray computed tomography (CT) scan showed a high density area in the left putamen (Fig. 1a). On magnetic resonance imaging (MRI) study, this area was of high signal intensity on T1-weighted and proton-density images (Fig. 1b, 1c). On a T2-weighted image, a small low-intensity area was observed in the left putamen (Fig. 1d). The SPECT study with 123I-IMP, performed on August 21, showed increased blood flow in the left striatum and thalamus (Fig. 2a). Oral administration of haloperidol (1.5–2.25 mg/day) and subcutaneous injection of biphasic isophane insulin (10–14 U/day) were started on August 18. The involuntary movements gradually decreased and disappeared on August 30 (Table 1). Also, the fasting blood glucose level was normalized and the 24-hour urine glucose was reduced to 0.2 g. The SPECT, performed on September 10, showed a slightly decreased blood flow in the left striatum when compared with that in the right. The increased blood flow in the left thalamus disappeared (Fig. 2b).

Discussion

Although hemichorea has been reported in patients with nonketotic hyperglycemia (1, 2), the underlying mechanism is unknown. The present case is notable because ketotic hyperglycemia also caused the choreic involuntary movements. Since the insulin therapy and the administration of haloperidol were started simultaneously, the involvement of the nigrostriatal...
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Fig. 1. a) Non-contrast-enhanced X-ray computed tomographic scan at the level of the basal ganglia, showing a high density area in the left putamen. b) T1-weighted image of magnetic resonance imaging (MRI) at the level of the basal ganglia, showing a high-intensity area in the left putamen. c) Proton-density image of MRI at the same level as in b, showing a high-intensity area in the left putamen. d) T2-weighted image of MRI at the level of the caudal basal ganglia, showing a small low-intensity area in the left putamen.

dopaminergic system in the present case of hemichorea is uncertain. However, some pathological changes in the striatum were considered to be responsible for the choreic involuntary movements, because the abnormal density and abnormal $^{123}$I-IMP uptake were detected on MRI and SPECT, respectively. Albin and colleagues (3) and DeLong (4) have argued that the hyperkinetic movement disorders result from the decreased activity of the subthalamic nucleus (STN). The decreased
activity of the STN, either because of STN destruction or because of selective dysfunction of a subpopulation of the striatal neurons projecting to the lateral globus pallidus, decreases the spontaneous activities of the medial globus pallidus neurons and results in the disinhibition of thalamocortical projections. Decreased blood flow or metabolism in the striatum has been reported in patients with Huntington's disease and benign hereditary chorea (5, 6). However, in the present patient, an abnormally increased blood flow was observed in the contralateral striatum and thalamus on SPECT. Although an increased blood flow is observed in reperfusion after cerebral infarction, this is unlikely in the present patient, because cerebral infarction selectively involving the striatum and thalamus is unusual and the MRI did not show any ischemic lesion. Therefore, the increased blood flow observed in the striatum and thalamus suggests hypermetabolism, or the increased ac-

Fig. 2. a) Single photon emission computed tomography (SPECT) with $^{123}$I-N-isopropyl-p-iodoamphetamine ($^{123}$I-IMP) at the level of the basal ganglia, performed on August 21, showing the increased blood flow in the left striatum and thalamus. b) SPECT with $^{123}$I-IMP at the same level as in a, performed on September 10, showing the slightly decreased blood flow in the left striatum. The blood flow in the left thalamus was not increased.
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Table 1. Involuntary Movements in Relation to Insulin Therapy and the Fasting Blood Glucose Level

<table>
<thead>
<tr>
<th>Date</th>
<th>Isophane insulin (unit/day)</th>
<th>FBG (mg/dl)</th>
<th>Involuntary movements</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aug. 17</td>
<td>-</td>
<td>401</td>
<td>+++       +++</td>
</tr>
<tr>
<td>Aug. 20</td>
<td>10</td>
<td>267</td>
<td>++        ++</td>
</tr>
<tr>
<td>Aug. 23</td>
<td>12</td>
<td>211</td>
<td>+         +</td>
</tr>
<tr>
<td>Aug. 24</td>
<td>14</td>
<td>183</td>
<td>-         +</td>
</tr>
<tr>
<td>Aug. 30</td>
<td>14</td>
<td>148</td>
<td>-         -</td>
</tr>
</tbody>
</table>

FBG: fasting blood glucose, UE: upper extremity, LE: lower extremity.

...tivity of neurons. Meanwhile, the SPECT, performed after the involuntary movements disappeared, showed the slightly decreased blood flow in the striatum of the present patient. A latent degenerative or microvascular lesion might have existed in the striatum. Also, hyperglycemia might trigger the malfunction of neurons in the striatum followed by the thalamic disinhibition, which was represented by the increased blood flow on SPECT.

In the present patient, the high-intensity area on T1-weighted images and the low-intensity area on a T2-weighted image were observed in the putamen. Also, the X-ray CT scan showed the high density area. Recently, the same abnormal findings on MRI and X-ray CT have been reported in diabetic patients with hyperkinetic movement disorders. Altafullah and colleagues reported a hyperglycemic patient with hemichorea of the left extremities, in whom a T1-weighted image demonstrated an increased signal in the right putamen (7). Although they described the case as putaminal hemorrhage or hemorrhagic infarct, the X-ray CT performed 4 days after the onset showed a slightly high attenuation area in the putamen, not surrounded by edema. The mass effect was not observed. These findings are atypical for putaminal hemorrhage or hemorrhagic infarct. Nakata and colleagues have reported hemiballism in a diabetic patient, in whom MRI showed high-intensity on T1-weighted images and the low-intensity area on a T2-weighted image in the contralateral putamen (8). These abnormal signals disappeared on the MRI performed six months after the onset. They have interpreted the putaminal lesion as a hemorrhage, but a hematoma usually shows marked hypointensity on T2-weighted images in the chronic stage (9). Nakamura and colleagues have also reported a diabetic patient with hemiballism, who had high-intensity areas on T1-weighted images in the putamen (10). They carried out a brain biopsy, and the specimen obtained from the putamen showed astrocytosis and vacuolization without deposition of hemosiderin. Therefore, a hemorrhagic event may not explain the putaminal abnormal intensity on MRI in the diabetic patient who has hemichorea or hemiballism. Yahikozawa and colleagues have reported three diabetic patients with hemiballism, in whom MRI showed striatal hyperintensity on T1-weighted images (11). They have presumed that the striatal lesion was caused by mild ischemia and have referred to postanoxic calcification of the basal ganglia. The abnormal intensity in the putamen of the reported cases and the present patient seems to be compatible with calcification of the brain (12–14). In the cases of Yahikozawa as well as in the case of Nakata, the putaminal hyperintensity on T1-weighted images faded or disappeared when the involuntary movements had been ameliorated. This finding suggests that the putaminal hyperintensity indicates a histochemical change, such as reversible mineral deposition, resulting from an abnormal metabolic state accompanied by hemichorea or hemiballism. Further morphological and metabolic studies will be necessary to clarify the underlying mechanism of hyperkinetic movement disorders in patients with hyperglycemia.

References