Case Report

Cerebral Infarction after Acetazolamide-challenged Single-photon Emission Computed Tomography in a Patient with Adult-onset Moyamoya Disease Accompanied by Several Risk Factors

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Abstract

A 46-year-old woman presented with dysarthria and an uncomfortable pharyngeal sensation. Her medical history included transient ischemic attacks at childbirth 20 years earlier and untreated hypertension for four years. She had been using oral contraceptives to treat vaginal bleeding for 9 years. Diffusion-weighted magnetic resonance imaging (MRI) revealed cerebral infarcts in the bilateral frontal lobes. Angiographic study showed bilateral sten-occlusive changes at the terminal portions of the internal carotid arteries and moyamoya vessels. Her symptoms disappeared spontaneously within 24 h. When acetazolamide-challenged single-photon emission computed tomography (ACZ-SPECT) was performed 7 weeks later, her arterial blood pressure, hemoglobin level, and hematocrit were 155/80 mmHg, 7.9 g/dL, and 31.5%, respectively, because of severe vaginal bleeding. She again complained of dysarthria and the uncomfortable pharyngeal sensation. And she manifested left facial palsy. Diffusion-weighted MRI revealed new cerebral infarcts at different sites in the right frontal lobe. The patient, who suffered from cerebral infarction after ACZ-SPECT, was diagnosed with adult-onset moyamoya disease accompanied by several risk factors such as severe anemia, prolonged hypertension, and oral contraceptive use for 9 years. This case indicates that some patients with moyamoya disease and certain risk factors may experience adverse effects after intravenous ACZ administration.

Key words: acetazolamide, adverse effect, cerebral infarction, moyamoya disease

1. Introduction

Basal and acetazolamide-challenged (ACZ) brain perfusion single-photon emission computed tomography (SPECT) can reveal regions of decreased cerebral perfusion and cerebrovascular reserve in patients with steno-occlusive cerebrovascular diseases. However, intravenous administration of ACZ often elicits transient symp-

toms in such patients. Here, we present an adult-onset moyamoya disease patient with several risk factors such as severe anemia, prolonged hypertension, and long-term oral contraceptive use who suffered from cerebral infarction after ACZ-SPECT.

2. Case Report

A 46-year-old woman presented with dysarthria and an uncomfortable sensation in her pharynx. Her medical history included transient ischemic attacks at childbirth 20 years earlier and untreated hypertension for four years. She had been using oral contraceptives to treat vaginal bleeding for 9 years. Magnetic resonance imaging (MRI) at her first admission revealed cerebral infarcts...
in the bilateral frontal lobes (Fig. 1). Her symptoms disappeared spontaneously within 24 h. Antiplatelet therapy was not administered because adult patients with moyamoya disease sometimes develop intracerebral hemorrhage. Angiographic study showed bilateral steno-occlusive changes at the terminal portions of the internal carotid arteries (ICAs) and an abnormal vascular network at the base of the brain (Fig. 2). The patient was placed under observation and discharged 2 weeks later without neurological deficits.

Basal SPECT with 167 MBq of $[^{123}]$I-isopropyl-$p$-iodoamphetamine (1.5 mL) and ACZ (17 mg/kg)-SPECT were performed 4 and 7 weeks after the infarction, respectively. At the time of the ACZ-SPECT study, the patient’s arterial blood pressure, hemoglobin level, and hematocrit were 155/80 mmHg, 7.9 g/dL, and 31.5%, respectively, because of severe vaginal bleeding; her body temperature was 36.4°C. Other blood studies yielded the following results: plasma total cholesterol 89 mg/dL, LDL cholesterol 31 mg/dL, HDL cholesterol 52 mg/dL, fasting blood glucose 91 mg/dL and hemoglobin A1c 4.4%. She again suffered dysarthria and an uncomfortable pharyngeal sensation. She also manifested left facial palsy. Diffusion-weighted MRI revealed new cerebral infarcts at different sites in the right frontal lobe (Fig. 3). Basal SPECT (Fig. 4A) and ACZ-SPECT (Fig. 4B) confirmed a decrease in cerebral perfusion and vascular reserve in the right hemisphere.

A right superficial temporal artery–middle cerebral artery bypass was performed 2 months after the second cerebral infarction. The patient has not experienced cerebral ischemia for 1 year since this infarction.
Saito, et al. reported that 63% of their patients with major cerebral artery steno-occlusive diseases developed transient symptoms after ACZ-SPECT. However, no patient with moyamoya disease suffered from cerebral infarction after intravenous ACZ administration. ACZ inhibits carbonic anhydrase by converting \( \text{CO}_2 + \text{H}_2\text{O} \) to \( \text{H}^+ + \text{HCO}_3^- \) in red blood cells and induces vasodilation in normal brain tissue. In the cerebral hemisphere on the arterial steno-occlusive side, drug delivery may be decreased and glucose metabolism may be lower. This may result in reduced vasodilation compared with the contralateral unaffected hemisphere and in the manifestation of the blood steal phenomenon.

The risk factors for cerebral ischemia in patients with moyamoya disease are hypercapnia or hypocapnia, hypotension, hypovolemia, and decreased or increased body temperature. Although anemia has not been documented as a risk factor, Sato, et al. suggested that the hematocrit should be maintained above 30% during anes-
thesis in patients with moyamoya disease because of their low cerebral perfusion and the consequent reduction in the delivery of oxygen to the brain.\(^4\)

Impaired cerebral vasoregulation persisted for more than 2 months post-onset in a patient with a small infarct.\(^5\) A previous report described 2 cases of cerebral infarct enlargement after intravenous ACZ administration, but the new infarction occurred at 9 and 19 days after the first infarction.\(^6\) In our patient, the second infarction occurred 7 weeks after the first. This finding may reflect impaired cerebral vasoregulation. Her first infarction may have been attributable to moyamoya disease with decreased cerebral perfusion and cerebrovascular reserve accompanied by severe anemia, prolonged hypertension,\(^7\) and long-term use of oral contraceptives.\(^8\) The second infarction may have been related to impaired cerebral vasoregulation and reduced cerebral perfusion due to the steal phenomenon by ACZ administration.

This case indicates that in patients with moyamoya disease and certain risk factors, the possible adverse effects of intravenous ACZ administration must be considered.

References