



Case Report

The Ivy Sign on Fluid Attenuated Inversion Recovery Images Related to Single-Photon Emission Computed Tomography Cerebral Blood Flow in Moyamoya Disease: A Case Report

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ABSTRACT

Moyamoya disease is an idiopathic progressive cerebrovascular steno-occlusive disorder characterized by the formation of numerous collaterals called Moyamoya vessels. Accurate evaluation of vascular status and cerebral blood flow (CBF) is needed for prompt treatment to prevent ischemic and/or hemorrhagic events. The pathogenesis of the ivy sign on fluid attenuated inversion recovery (FLAIR) images of Moyamoya disease patients is unclear. We report a Moyamoya disease case wherein the ivy sign changed in relation to single-photon emission computed tomography (SPECT)-measured CBF during progression and following treatment. A 49-year-old female presented with slight aphasia and right hemiparesis. Magnetic resonance imaging (MRI) diffusion-weighted image revealed cerebral infarction in the left frontal lobe. Cerebral angiography images showed bilateral distal internal carotid artery stenosis and Moyamoya vessels. FLAIR images exhibited the ivy sign. We performed superficial temporal artery–middle cerebral artery (STA–MCA) bypass surgery with encephalogaleosynangiosis (EGS) and encephalomyosynangiosis (EMS) on the left side 6 months after first presentation. After operation, left-side resting CBF gradually improved on SPECT and the ivy sign decreased. On the other hand, right-side CBF gradually deteriorated at rest, and the ivy sign increased. Therefore, we performed STA–MCA bypass with EGS and EMS on the right side 4 years after first presentation. After the operation, the resting CBF increased and the ivy sign decreased. The FLAIR ivy sign may be a useful indicator of both deterioration and improvement of CBF status without the need for CBF imaging using contrast material.

KEYWORDS: Moyamoya disease, FLAIR, Ivy sign, SPECT

ABBREVIATIONS: CBF: Cerebral blood flow, EGS: Encephalogaleosynangiosis, EMS: Encephalomyosynangiosis, FLAIR: Fluid attenuated inversion recovery, ICA: Internal carotid artery, MCA: Middle cerebral artery, MRA: Magnetic resonance angiography, MRI: Magnetic resonance imaging, SPECT: Single-photon emission computed tomography, STA: Superficial temporal artery, PET: Positron emission tomography

INTRODUCTION

Moyamoya disease is an idiopathic progressive cerebrovascular steno-occlusive disorder characterized by the formation of numerous collaterals called “Moyamoya vessels” (17). Cerebral angiography and mag-

netic resonance imaging (MRI) are indicated for the diagnosis and grading of Moyamoya disease (6), and single-photon emission computed tomography (SPECT) has also been used to evaluate cerebral blood flow (CBF)(3). The ivy sign, observed on post-contrast magnetic resonance images (MRI) and fluid attenuated inversion recovery (FLAIR) images, is

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speculated that the slow retrograde flow of engorged pial collateral arteries, the thickening of leptomeninges because of congestion, and maximally dilated pial vessels (14). Several studies have reported an improved ivy sign on MRI after bypass surgery (12,13). The authors reported increased ivy sign with decreased quantitative CBF on N-isopropyl- p-[(123)I] iodoamphetamine (123)I-IMP SPECT and decreased ivy sign with increased CBF following STA-MCA anastomosis over the course of adult Moyamoya disease.

However, the details of changes in the ivy signs are unclear. Here, we report a Moyamoya case with repeated reciprocal ivy sign changes relative to SPECT-measured CBF.

■ CASE REPORT

A 49-year-old female presented with slight aphasia and right hemiparesis. Blood pressure was 134/72 mm Hg and pulse was 66 beats/minute with normal rhythm. The patient had a history of hypertension, but she neither smoked nor drank alcohol. Her mother had a history of cerebral infarction due to Moyamoya disease.

MRI diffusion-weighted image (DWI) revealed cerebral infarction in the left frontal lobe (Figure 1A). Magnetic resonance angiography (MRA) and cerebral angiography images showed bilateral distal internal carotid artery (ICA) stenosis and Moyamoya vessels on both sides of the ICA (Figure 1C, D). FLAIR images exhibited the ivy sign (Figure 1B).

The patient was administered conservative therapy for cerebral infarction and underwent rehabilitation for aphasia and hemiparesis. After rehabilitation, her deficits were improved. We performed superficial temporal artery (STA)-middle cerebral artery (MCA) bypass surgery with encephalogaleosynangiosis (EGS) and encephalomyosynangiosis (EMS) on the left side 6 months after first presentation.

After the left-side operation, CBF gradually improved at rest SPECT CBF and the ivy sign decreased (Figure 2A, B). On the other hand, right-side CBF gradually deteriorated at rest, and SPECT CBF and the ivy sign increased. Therefore, we performed STA-MCA bypass with EGS and EMS on the right side 4 years after first presentation. After operation, she had no neurological deficits, and SPECT CBF increased at rest and

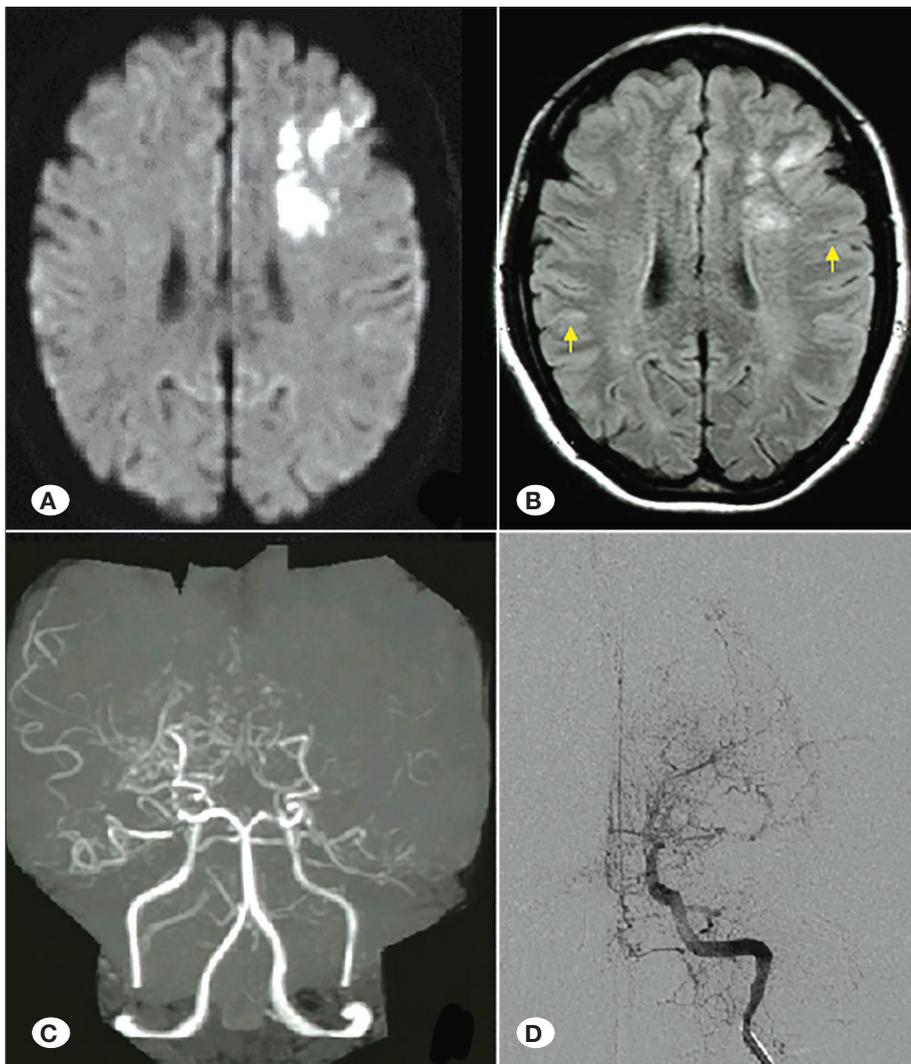


Figure 1: A) MRI-DWI revealed fresh cerebral infarction in left frontal lobe. B) MRI FLAIR images showed the ivy sign. C) MRA revealed bilateral ICA stenosis. D) Typical net-like Moyamoya vessels observed by angiography.

ivy sign decreased (Figure 3). No cerebral ischemic episodes were observed during the following year after two times of operation.

DISCUSSION

In the present study, the FLAIR ivy sign is a useful indicator for the observation of the state of CBF in Moyamoya disease because it is related to SPECT CBF.

Moyamoya disease, first reported in Japan in 1963 (17), is a rare cause of ischemic and hemorrhagic stroke with approximately 10% of cases due to inheritance (7). It is a progressive cerebrovascular disorder of the distal ICA accompanied by formation of net-like collateral vessels at the base of the brain. Disease progression can be divided into six stages (18,19). There are currently no effective drug therapies for Moyamoya disease, and therefore, surgical revascularization by STA-MCA anastomosis with or without an indirect bypass is the standard surgical treatment. As the increase in CBF achieved by indirect revascularization is often unsatisfactory, and direct revascularization is usually feasible, particularly in adults

(1), direct revascularization is recommended by Japanese guidelines for the treatment of Moyamoya disease (6). However, accurate evaluation of vascular status and CBF is needed for prompt treatment to prevent ischemic and/or hemorrhagic events. For evaluating the stages of Moyamoya disease, cerebral angiography is still the gold standard; however, with the continued development of MRI techniques, diagnosis of Moyamoya disease with a > 1.5 Tesla magnet has become possible (4,5,9,21). SPECT and positron emission tomography (PET) have also been used to evaluate CBF (3,15). Moreover, leptomeningeal high-signal intensity on post-contrast T1-weighted MR imaging, the ivy sign is assumed to indicate decreased cerebral perfusion, dilated pial vasculature, and slow flow in developed leptomeningeal collaterals. The ivy sign is also observed on unenhanced FLAIR images (14). In addition, the ivy sign correlated with neurological deficit and previous brain old lesions, and reflected the progression of Moyamoya disease (2,20).

Kaku et al. reported a positive correlation between ivy sign proliferation and cerebral blood volume (CBV) in Moyamoya disease, and a more obvious negative correlation was found

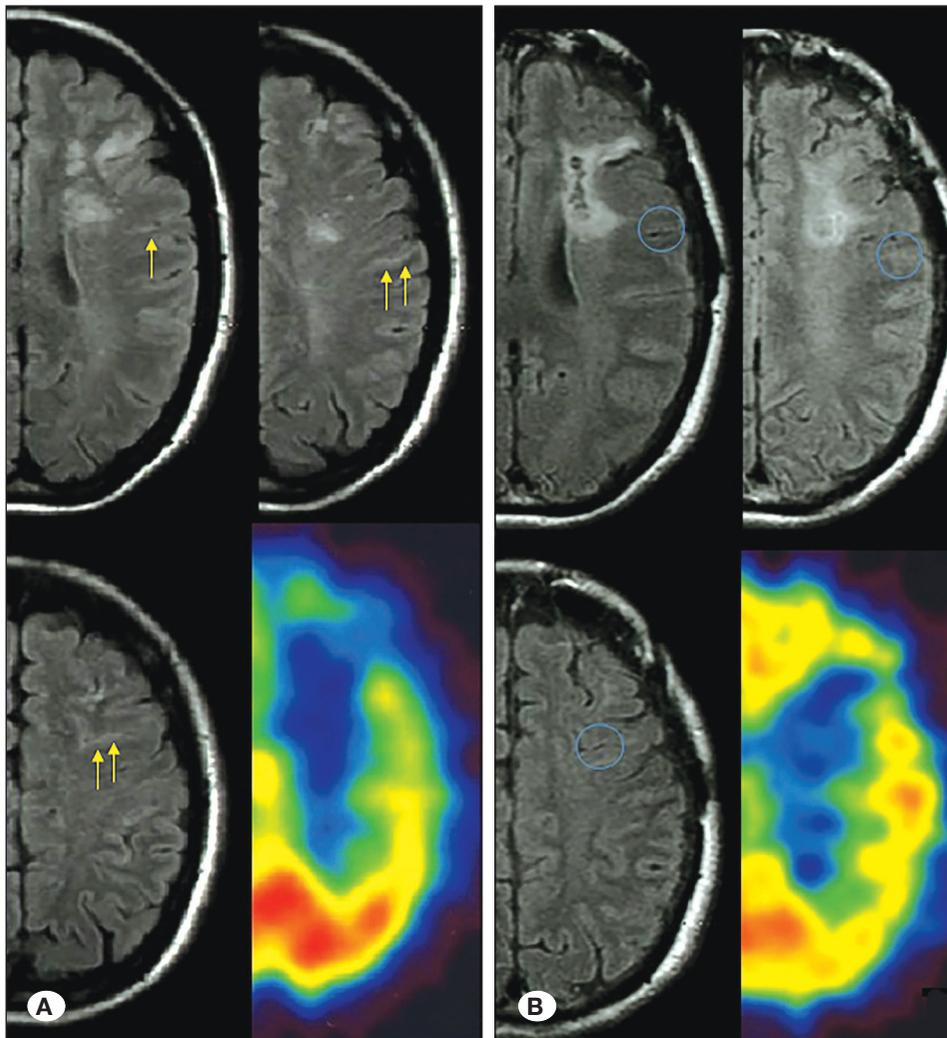


Figure 2:
A) Pre-operative FLAIR images showing the ivy sign and SPECT showing hypoperfusion in the left frontal lobe.
B) Post-operative FLAIR images showing decreased ivy sign in left frontal lobe, and SPECT revealed increased CBF in left frontal lobe.

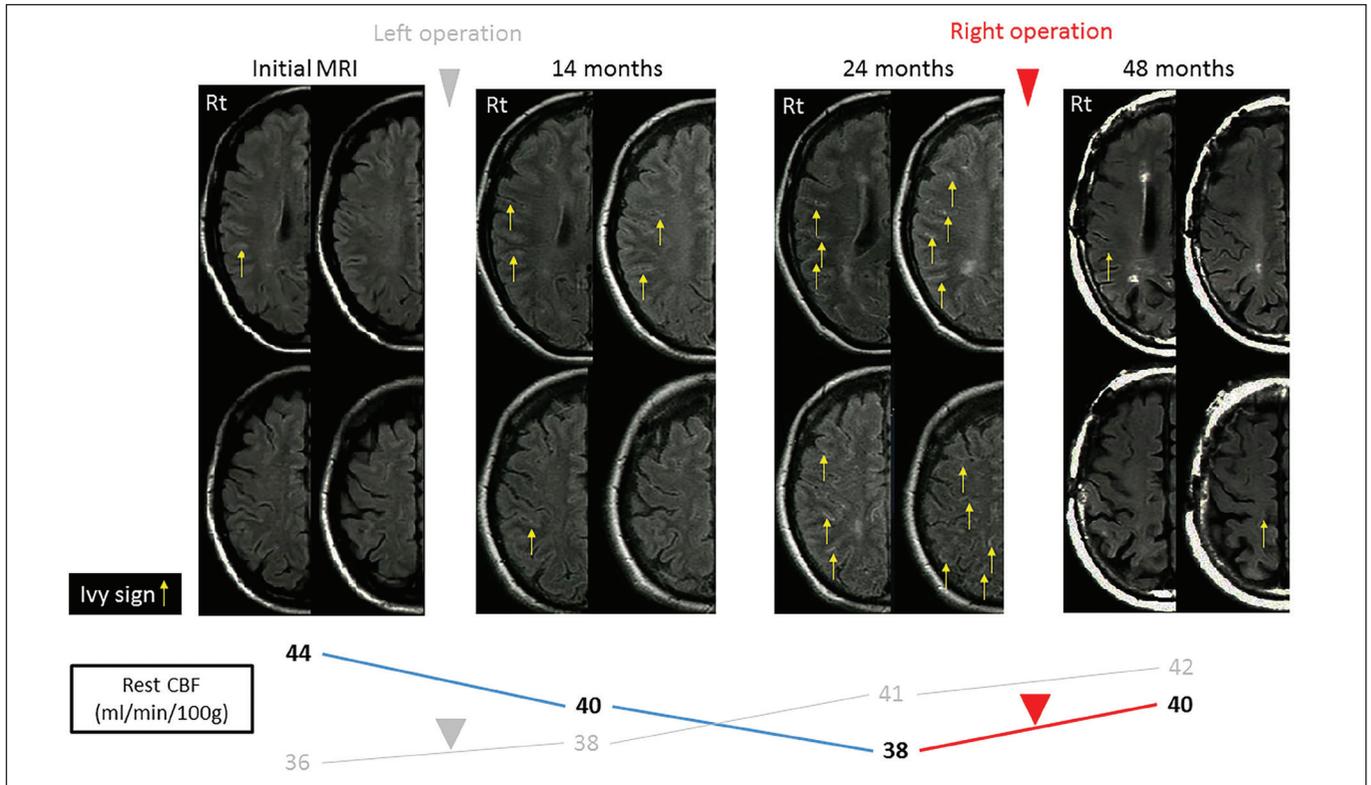


Figure 3: Follow-up ivy sign and relation to SPECT CBF.

between ivy sign proliferation and CBF/CBV (11). A past report found that disappearance of the ivy sign indicated hemodynamic improvement after revascularization surgery (10). Further, Lee et al. reported that the ivy sign on FLAIR is a useful indicator of brain hemodynamic changes following surgery in adult Moyamoya disease (13). However, there are no reports of a FLAIR ivy sign increase during follow-up related to a decrease in quantitative CBF.

In Moyamoya disease, the main aim of surgery is to prevent the brain from ischemic and hemorrhagic events. Thus, careful and regular postoperative hemodynamic evaluation is necessary. Therefore, simple evaluation modalities without contrast material, such as SPECT or PET, could be advantageous. In the present case, the FLAIR ivy sign was inversely related to CBF as measured by SPECT. We suggest that the FLAIR ivy sign is useful for follow-up imaging and helpful for decisions related to surgical timing of Moyamoya disease because it obviates risks associated with catheterization and contrast agents.

In addition, when Moyamoya disease patients are admitted to emergency department with headache or transient disturbance of consciousness, FLAIR images may be a useful indicator to evaluate the hemodynamic condition such as cerebrovascular reactivity and degree of ivy sign can suggest the need for more aggressive medical treatment or revascularization surgery (16).

Horie et al. reported that de novo ivy sign could indicate cerebral hyperperfusion after revascularization procedure, and the ivy sign is therefore a useful tool for risk management postoperatively (8).

Additional cases are required to confirm this relationship between the FLAIR ivy sign and SPECT-measured CBF.

CONCLUSION

Ivy sign on FLAIR images is related to SPECT CBF. Therefore, the FLAIR ivy sign is a useful indicator for follow-up imaging in Moyamoya disease.

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