

# **Adult-Onset Moyamoya Disease With Repetitive Ischemic Attacks Successfully Treated by Superficial Temporal-Middle Cerebral Artery Bypass**

## **—Case Report—**

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### **Abstract**

**A 78-year-old Japanese woman presented with adult-onset moyamoya disease manifesting as repetitive ischemic attacks over a period of 26 years. Initial ischemic attack occurred at age 52 years. Neurological status deteriorated, associated with worsening in angiographical and single photon emission computed tomography findings over the last 10 years. She suffered frequent transient attacks of aphasia and right hemiparesis. Superficial temporal-middle cerebral artery anastomosis was performed. Postoperative angiography and single photon emission computed tomography revealed a marked increase in cerebral blood flow and symptoms showed considerable improvement. Direct revascularization may be the treatment of choice even for aged patients with moyamoya disease.**

Key words: moyamoya disease, adult, angiography, single photon emission computed tomography, anastomosis

### **Introduction**

Moyamoya disease is a type of cerebral angiopathy characterized by spontaneous occlusive lesions of the internal carotid artery (ICA) and compensatory development of thin, fragile, and net-like arteries called moyamoya vessels at the base of the brain.<sup>2,3,7)</sup> The clinical picture of this disease significantly differs between children and adults. Pediatric-onset cases generally present with symptoms caused by slowly progressive cerebral hypoperfusion and repetitive ischemic symptoms, whereas adult-onset cases tend to suffer hemorrhagic events. However, recent studies on large series of moyamoya disease patients have shown that more than one-third of adult-onset cases did present with ischemic onset.<sup>1,3,5)</sup> Occlusive angiopathy and ischemia seen in adult-onset moyamoya disease rarely show symptomatic progression.<sup>2,7,13)</sup> Only a few case reports have demonstrated repetitive ischemic attacks in such patients.

Here we present a case of adult-onset moyamoya

disease manifesting as repetitive ischemic symptoms over a period of 26 years which was successfully treated by direct revascularization.

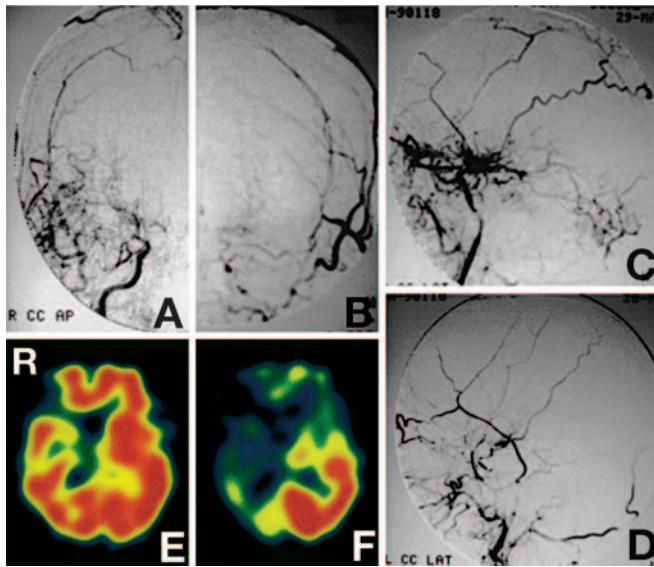
### **Case Report**

A 78-year-old Japanese woman apparently suffered an initial ischemic event at the age of 52 years, when she developed moderate left hemiparesis. Unfortunately no medical records or radiological studies were available on this initial episode. She made a good recovery with minimal residual left hemiparesis, but could live independently.

Her left hemiparesis suddenly deteriorated at the age of 68 years. Computed tomography showed an old infarction in the right frontal lobe. Common carotid angiography revealed occlusion of the bilateral ICAs and development of basal moyamoya vessels (Fig. 1A–D). The posterior circulation was free of arteriosclerotic changes. She did not have hypertension, hyperlipidemia, or diabetes mellitus. The diagnosis was adult-onset moyamoya disease.

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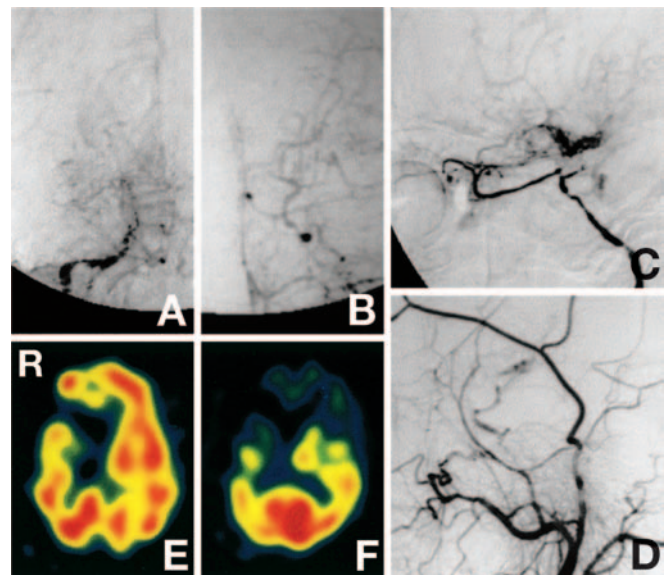
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**Fig. 1** A–D: Carotid angiograms taken at age 68 years showing occlusion of the bilateral internal carotid arteries and development of basal moyamoya vessels. A, B: anteroposterior views; C, D: lateral views of the right (C) and left (D) common carotid arteries. E: Single photon emission computed tomography with iodine-123 iodoamphetamine ( $^{123}\text{I}$ -IMP SPECT) image showing decreased cerebral blood flow corresponding to the old infarction in the right frontal lobe. F:  $^{123}\text{I}$ -IMP SPECT image after acetazolamide administration revealing preservation of vascular reserve only in the left temporo-occipital region.

Single photon emission computed tomography with iodine-123 iodoamphetamine ( $^{123}\text{I}$ -IMP SPECT) demonstrated preserved cerebral blood flow (CBF) and significant decrease of vascular reserve (Fig. 1E, F). The hypoperfusion area in the right frontal lobe was consistent with the old infarction. Surgical treatment was not recommended because the hypoperfusion area demonstrated by SPECT was almost limited to the area of infarction.

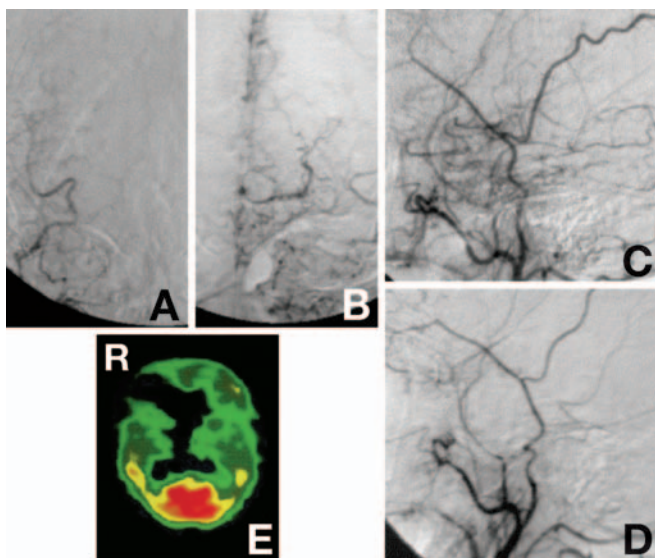
She developed a chronic subdural hematoma, and underwent irrigation and drainage at age 74 years. The hematoma was completely resolved. Common carotid angiography showed a significant reduction in the basal moyamoya vessels and progression of occlusive angiopathy in the bilateral ICAs (Fig. 2A–D). Despite the angiographical deterioration, no surgical treatment was performed because  $^{123}\text{I}$ -IMP SPECT indicated that CBF was preserved except in the old infarct area in the right frontal lobe and only subtle changes in vascular reserve had occurred over 6 years (Fig. 2E, F).



**Fig. 2** A–D: Carotid angiograms taken at age 74 years revealing reduction in the basal moyamoya vessels. A, B: anteroposterior views; C, D: lateral views of the right (C) and left (D) common carotid arteries. E, F: Single photon emission computed tomography with iodine-123 iodoamphetamine images before (E) and after acetazolamide administration (F) showing preservation of the cerebral blood flow and vascular reserve since the first admission.

She was readmitted to our hospital for newly developing aphasia and right hemiparesis at age 78 years. Her symptoms markedly fluctuated due to frequent transient ischemic attacks (TIAs) affecting the left cerebral hemisphere. Angiography showed complete occlusion of the bilateral ICAs at the cervical bifurcation and further decreases in the moyamoya vessels at the base (Fig. 3A–D). SPECT with technetium-99m ethyl-cysteinate dimer demonstrated marked reduction in CBF in the bilateral cerebral hemispheres (Fig. 3E). The frequent TIAs were so severe that SPECT with administration of acetazolamide could not be performed. Right vertebral angiography showed few indications of arteriosclerotic change in the posterior circulation (Fig. 4). Laboratory blood analysis revealed normal levels of serum glucose, total cholesterol, and triglyceride.

Left superficial temporal-middle cerebral artery (STA-MCA) anastomosis was performed to improve the perfusion of the left hemisphere. The TIAs disappeared completely after the operation. Postoperative radiological imaging performed one month later showed improvement of vascularization and CBF

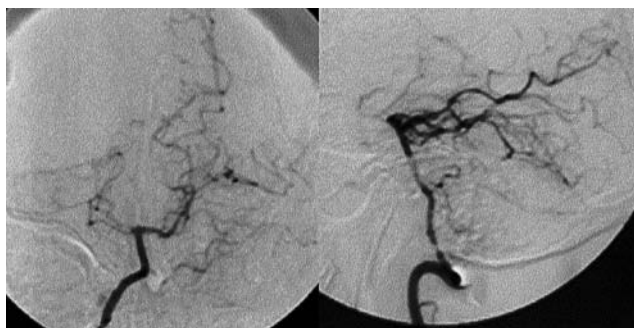


**Fig. 3** A–D: Carotid angiograms taken at age 78 years demonstrating complete occlusion of the bilateral internal carotid arteries and severe decrease in the moyamoya vessels at the base. A, B: anteroposterior views; C, D: lateral views of the right (C) and left (D) common carotid arteries. E: Single photon emission computed tomography with technetium-99m ethyl-cysteinate dimer image revealing extremely reduced cerebral blood flow in the bilateral frontal, parietal, and temporal lobes.

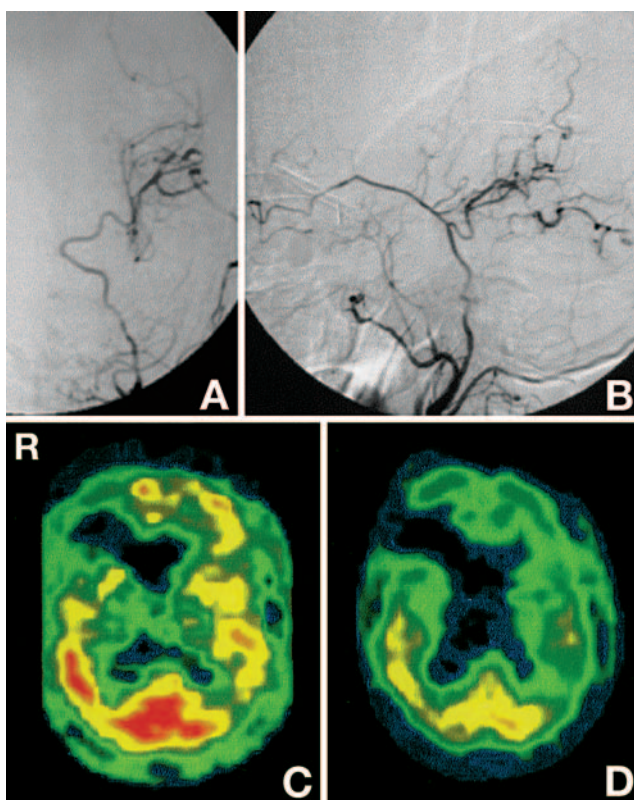
via the anastomosis (Fig. 5). However, the diamox test showed little vascular reactivity in spite of the clinical improvement. She has suffered no TIA or new infarction for a year after the surgery.

## Discussion

Pediatric-onset moyamoya disease often manifests as ischemia and is usually progressive if not treated. Surgical procedures are effective to increase collateral circulation, such as STA-MCA anastomosis,<sup>11,15</sup> encephalo-duro-arterio-synangiosis, and encephalo-myo-synangiosis.<sup>8</sup> On the other hand, adult-onset moyamoya disease with ischemic onset is rarely progressive or shows repeated ischemic symptoms. However, surgical reconstruction is recommended for such cases under the current therapeutic guidelines<sup>3,4</sup> based on the observation that surgery reduces the basal moyamoya vessels and improves some hemodynamic parameters.<sup>6,9,10</sup> Although no randomized study has been carried out to assess the effectiveness of surgical reconstruction for adult-onset ischemic moyamoya disease, surgical reconstruction could improve the clinical symptoms



**Fig. 4** Right vertebral angiograms showing the relatively normal basilar artery and bilateral posterior cerebral arteries.



**Fig. 5** A, B: Postoperative carotid angiograms showing vascular flow in the middle cerebral arteries via the anastomosis. A: anteroposterior view, B: lateral view. C: Single photon emission computed tomography with technetium-99m ethyl-cysteinate dimer (<sup>99m</sup>Tc-ECD SPECT) image showing increased cerebral blood flow in the left cerebral hemisphere. D: <sup>99m</sup>Tc-ECD SPECT image after acetazolamide administration revealing the vascular reserve was still impaired.



as seen in our case.

Three other cases of adult-onset moyamoya disease have presented with the progressive occlusive angiopathy.<sup>12,14,16)</sup> In one case, the progression of occlusive angiopathy was angiographically verified, but the symptomatic recurrence was caused by a hemorrhagic event. However, a recent study from the United States looking at the recurrence pattern in patients with adult-onset moyamoya disease showed that a relatively high number of patients suffered ischemic recurrence.<sup>1)</sup> If patients with adult ischemic moyamoya disease suffer repetitive ischemic attacks, then direct vascular reconstruction could be the treatment of choice.

In our patient, the CBF in the left hemisphere was significantly improved, but the vascular reserve showed little augmentation after the operation, although the TIAs disappeared completely. Radiological improvement might become apparent after a longer period. Her cognitive function was only slightly impaired before the operation, and showed minor improvement after the operation, although not evaluated quantitatively because of the repetitive ischemic attacks before the operation. In some cases of adult ischemic moyamoya disease, the hypoperfusion is so critical that vascular reconstruction might be considered imperative as seen in pediatric-onset moyamoya disease. Bypass surgery should be considered as an effective therapy even for aged patients with moyamoya disease.

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