

## Case Report

# A case of unilateral moyamoya disease suffered from intracerebral hemorrhage due to the rupture of cerebral aneurysm, which appeared seven years later

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## Abstract

**Background:** Whether unilateral moyamoya disease (MMD), confirmed by steno-occlusive lesion at the terminal portion of internal carotid artery with formation of moyamoya vessels unilaterally and normal or equivocal findings contralaterally, is an early form of definite (bilateral) MMD remains controversial. It is well-known that adult patients with MMD tend to suffer from cerebral hemorrhage, occasionally due to the rupture of aneurysm arising from moyamoya vessel.

**Case Description:** A 61-year-old woman was diagnosed as unilateral MMD incidentally and followed by magnetic resonance imaging annually. Seven years after the diagnosis, cerebral aneurysm appeared on the moyamoya vessel. Before further examination, the aneurysm ruptured and resulted in massive cerebral hemorrhage.

**Conclusion:** Even in the unilateral MMD, cerebral hemorrhage may occur due to the rupture of cerebral aneurysm. Careful follow-up is recommended and early treatment is required once cerebral aneurysm is detected.

**Key Words:** Cerebral aneurysm, Intracerebral hemorrhage, moyamoya disease, Unilateral

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## INTRODUCTION

Moyamoya disease (MMD) is characterized by progressive occlusion of the internal carotid artery (ICA) or its terminal branches, associated with formation of extensive collateral vessels (moyamoya vessels) at the base of the brain. The diagnostic criteria of the Research Committee on Moyamoya Disease (Spontaneous Occlusion of the Circle of Willis) of the Ministry of Health and Welfare of Japan (RCMJ) considers only cases with bilateral

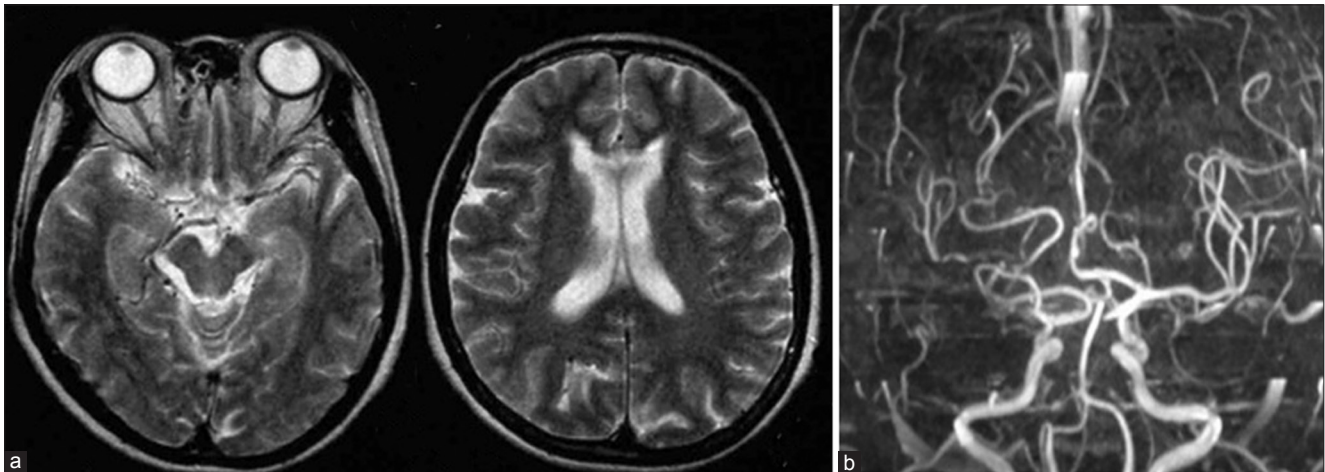
lesions as definite MMD.<sup>[1]</sup> However, some cases with unilateral involvement also show angiographic findings on the affected side similar to those of definite cases, and so are classified as probable MMD. Pediatric patients are well-known to present with ischemic attack, whereas adults tend to suffer from intracranial bleeding among patients with MMD.<sup>[10]</sup> Thus, it is reported that cerebral aneurysm arising from moyamoya vessels can be a cause of hemorrhage.<sup>[6]</sup> We have reported clinical feature of nine cases of unilateral MMD<sup>[3]</sup> and one of the patients deteriorated because of rupture of cerebral

aneurysm during follow-up. Here, we report a case of unilateral MMD suffered from cerebral hemorrhage due to the rupture of aneurysm, which appeared 7 years after initial diagnosis.

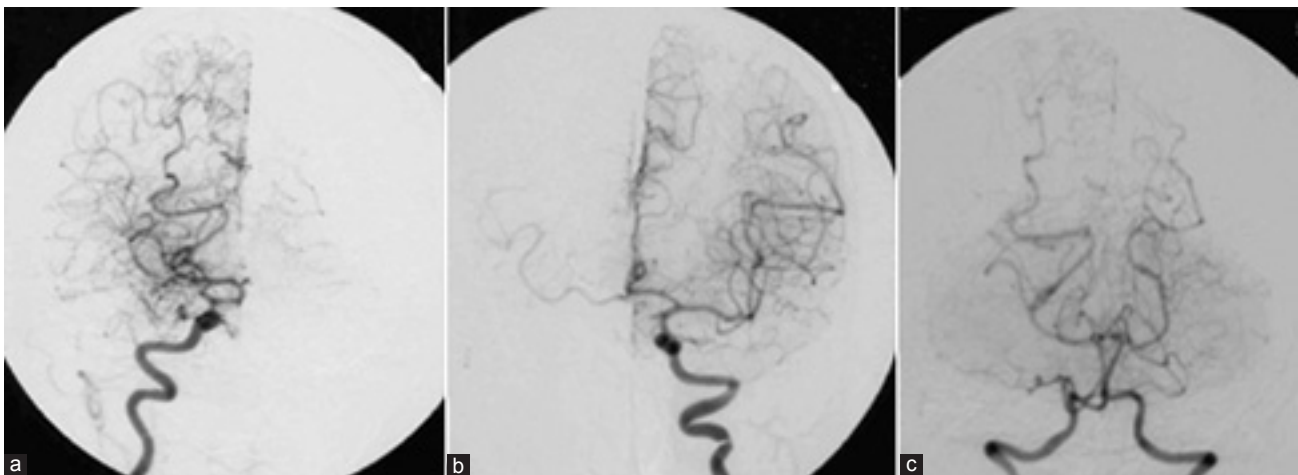
## CASE REPORT

A 61-year-old woman without any risk factor of arteriosclerosis visited a nearby hospital because of mild head injury. Magnetic resonance (MR) imaging demonstrated no apparent traumatic lesion. However, flow-void sign of the right middle cerebral artery was sluggish [Figure 1]. Cerebrovascular disease was suspected and she was referred to our hospital for further examination. She was neurologically intact and had no significant history of weakness or sensory disturbance. Moreover, her family has not experienced cerebrovascular disease. Right carotid angiogram demonstrated stenosis of the terminal portion of the

ICA with moyamoya vessels [Figure 2a]. Left carotid angiogram and left vertebral angiogram showed no apparent abnormality [Figure 2b and c]. Single photon emission computed tomography (SPECT) demonstrated hypoperfusion in the right cerebral hemisphere. The response to the acetazolamide was impaired [Figure 3]. She was diagnosed as unilateral MMD and followed by MR imaging annually. During 6 years, she had been intact and MR imaging had not detected progression of MMD. However, 7 years after the diagnosis, cerebral aneurysm appeared in the right basal cistern, probably from anterior choroidal artery, and conventional angiography was planned [Figure 4]. Before that, she became comatose and brought to our hospital emergently. Computed tomography (CT) showed massive intraventricular hemorrhage [Figure 5a]. Enlargement of the aneurysm was revealed by CT angiography [Figure 5b-d]. Intracerebral hematoma was evacuated and the aneurysm was obliterated by bipolar coagulation. Postoperatively,

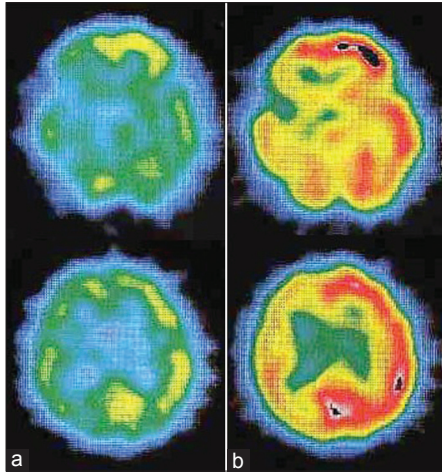


**Figure 1: Magnetic resonance (MR) image. (a) MRI T2-weighted image shows no ischemic lesion. The flow-void signal of the right middle cerebral artery is sluggish, (b) MRA shows steno-occlusive at the terminal portion of the right internal carotid artery**



**Figure 2: Cerebral angiography. (a) Right cerebral angiography reveals severe stenosis at the terminal portion of the internal carotid artery with development of moyamoya vessels, (b) Left carotid angiography shows no apparent abnormality, (c) Left vertebral angiography shows no apparent abnormality. The leptomeningeal anastomosis is developed in the right side**

she was in vegetative state and transferred to the rehabilitation hospital.

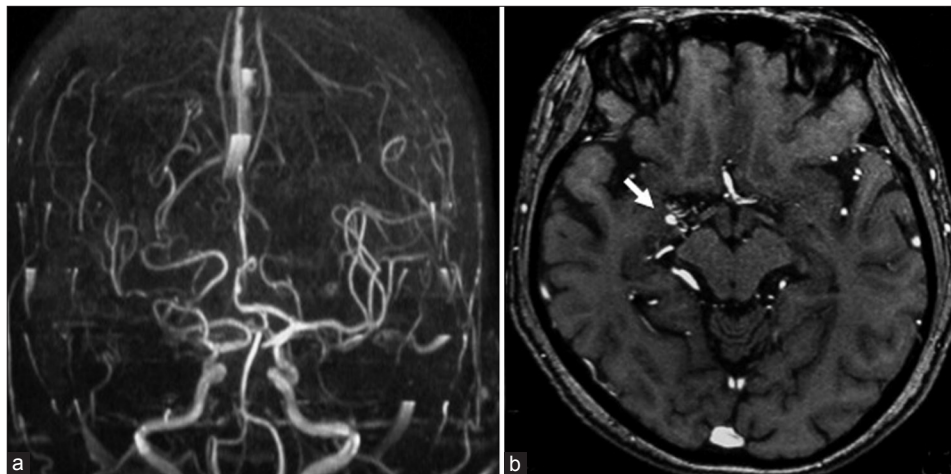


**Figure 3:** Single photon emission tomography. (a) Right hypoperfusion was detected, (b) Response to the acetazolamide was impaired in the right side

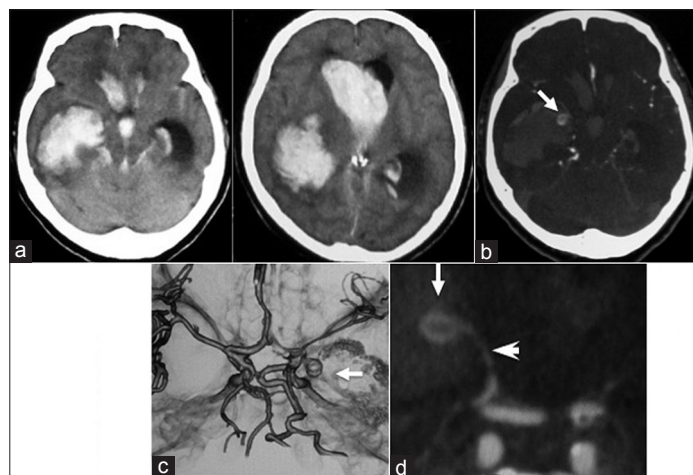
## DISCUSSION

Whether unilateral lesion, confirmed by typical angiographic evidence of MMD unilaterally and normal or equivocal findings contralateral, is an early form of definite (bilateral) MMD remains controversial.<sup>[6]</sup> In a study of 64 patient with unilateral MMD followed up for 1-7 years, progression to bilateral disease was noted in 17 (27%) patients, and such progression to bilateral disease within 5 years was frequent in children with early onset of MMD.<sup>[7]</sup> Family history is occasionally present for patients with unilateral MMD.<sup>[4]</sup> Therefore, it is considered that some of the unilateral cases are etiologically same as definite MMD. In our case, right side showed appearance of MMD, however, left side was normal. Then, diagnosis of unilateral MMD was made.

Cerebral hemorrhage is main clinical manifestation in adult patient with MMD. The fragile moyamoya vessels are considered as a cause of the hemorrhage. Even in unilateral MMD, hemorrhage is a significant clinical



**Figure 4:** MR angiography 7 years after the onset. (a) MIP image shows no significant change, (b) Source image revealed cerebral aneurysm



**Figure 5:** Computed tomography (CT). (a) CT shows massive intraventricular hemorrhage, (b) CT with contrast medium revealed enlargement of the cerebral aneurysm, (c) CT angiography showed cerebral aneurysm arising from right anterior choroidal artery, (d) CT angiography coronal image showed cerebral aneurysm (arrow) arising from right anterior choroidal artery (arrow head)

manifestation. Ikezaki *et al.* found that 58% of the adult unilateral MMD patients suffered from hemorrhagic stroke.<sup>[4]</sup> MMD is frequently associated with intracranial aneurysms, probably as a result of the intrinsic pathology of MMD such as hemodynamic stress and fragile structure of the collateral vessels. These aneurysms have been generally classified into three distinct subtypes: aneurysms at major artery (around the circle of Willis), distal peripheral arteries (such as the anterior or posterior choroidal artery), and those of moyamoya vessels (basal collaterals).<sup>[6]</sup> A review found that 56% of the aneurysms associated with MMD occurred around the circle of Willis, 22% on the peripheral collateral vessels, and 18% in the basal ganglia. Aneurysms around the circle of Willis predominantly developed in the posterior circulation. Kasamo *et al.* reported a case of unilateral MMD associated with multiple aneurysms, which resulted in subarachnoid hemorrhage.<sup>[5]</sup> In our case, the aneurysm appeared at the moyamoya vessel, anterior choroidal artery. The aneurysm appeared 7 years after diagnosis of MMD. Her blood pressure was in normal range and other risk factors, which induce atherosclerosis, was negative. She had been stable neurologically and radiologically. However, once aneurysm appeared, it enlarged rapidly and ruptured. Longer follow-up on ischemic lesion as well as cerebral aneurysm is required even in the stable unilateral lesion.

The inherent difficulty in surgical treatment of these aneurysms, due to deep location, small size, and sometimes unfavorable morphology, is compounded in the setting of MMD by vessel hemodynamic fragility.<sup>[6]</sup> Vascular recanalization is an option since hemodynamic stress reduction may result in disappearance of the aneurysm.<sup>[11]</sup> Recently, improving microvascular techniques and technology may enable more frequent endovascular treatment for these aneurysms.<sup>[2,9]</sup> In this patient, as shown in Figure 5, feeding artery was relatively dilated and endovascular treatment might be possible. If

it failed, bypass surgery was a choice. Anyway, we should treat the aneurysm as soon as possible, once it appears during follow-up.

## CONCLUSION

In conclusion, cerebral aneurysm may appear during follow-up of unilateral MMD and can be a cause of intracerebral hemorrhage. Once aneurysm is detected, further examination and early treatment is required.

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