



# Progressive Cerebral Arteriopathy – Moyamoya Disease: A Report of Two Cases with Different Clinical Presentation

## *Arteriopatia progressiva cerebral – Doença de moyamoya: Relato de dois casos com apresentações clínicas distintas*

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### Abstract

Moyamoya disease is a chronic and unusual cerebrovascular disorder characterized by progressive stenosis and occlusion of the distal portions of internal carotid arteries and its main branches within the circle of Willis. Posterior circulation (vertebral and basilar arteries) may also be affected; however, this presentation is uncommon. As well as stenosis of the terminal portion of intracranial arteries, it is seen the development of a network of collateral vessels abnormally dilated at the base of the brain with an aspect of a “puff of smoke,” whose term in Japanese is described as “moyamoya.” The present study aims to report two consecutive cases of patients who presented to our service with different clinical manifestations. Further investigation with digital subtraction angiography showed a moyamoya pattern.

### Keywords

- ▶ cerebral arteriopathy
- ▶ progressive vasculopathy
- ▶ moyamoya

### Resumo

A doença de moyamoya (DMM) é uma desordem cerebrovascular crônica de rara incidência, caracterizada pela estenose progressiva das porções terminais das artérias carótidas internas, associada à proliferação de vasos colaterais anormalmente dilatados na base do crânio, cujo aspecto se assemelha a uma “fumaça,” definido pelo termo em japonês “moyamoya.” A circulação posterior (artérias vertebrais e artéria basilar) também pode ser acometida, porém de forma menos frequente. A apresentação clínica é variada. O presente estudo objetiva relatar dois casos de pacientes que apresentaram diagnóstico angiográfico compatível com a DMM e manifestações clínicas distintas.

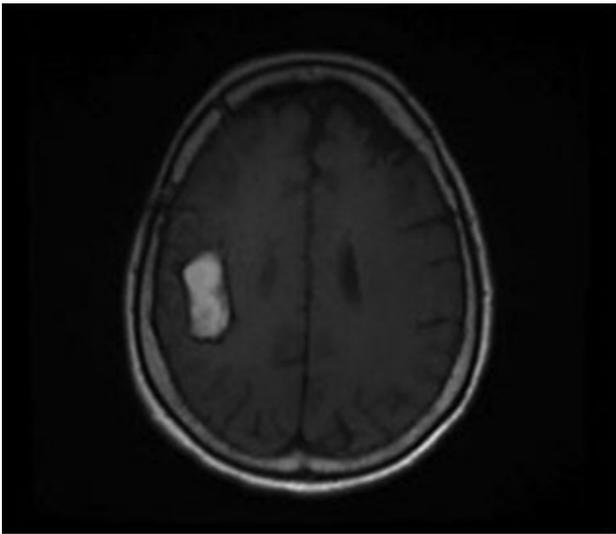
### Palavras-chave

- ▶ arteriopatia cerebral
- ▶ vasculopatia progressiva
- ▶ moyamoya

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**Fig. 1** MRI of the encephalon – FLAIR sequence, axial cut – showing right frontoparietal intraparenchymal hematoma.

### Introduction

Moyamoya disease (MMD) is a cerebrovascular disorder of rare incidence, characterized by stenosis and progressive occlusion of the terminal portions of the internal carotid arteries and their main branches in the circle of Willis.<sup>1,2</sup> The posterior circulation (vertebral arteries and basilar artery) can also be affected, but this occurs with less frequency.<sup>3</sup> In addition to progressive stenosis, it is observed in this pathology the development of a network of abnormally dilated collateral vessels at the base of the skull, which can take on an aspect of smoke, named in Japanese as “Moyamoya.”<sup>1,4</sup> The clinical manifestation is variable, and the patient may be asymptomatic or they may have, among other symptoms, headaches, seizures, focal neurological deficit and even severe cases with ischemia or cerebral hemorrhage.<sup>5–7</sup> The present study aims to report two cases of patients, consecutively treated in our service, who presented angiographic diagnosis compatible with MMD and distinct clinical manifestations.

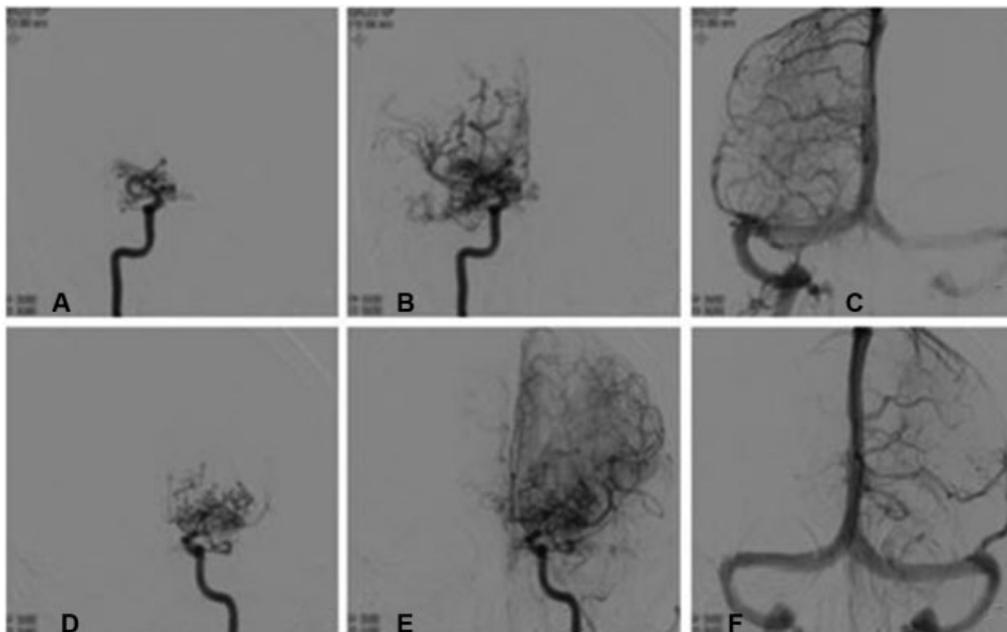


**Fig. 2** Cerebral angiography image – arterial phase, 3D TOF – showing significant reduction of caliber in the terminal portions of the internal carotid arteries (arrows), with dilated collateral vessels at the base of the skull (arrowheads).

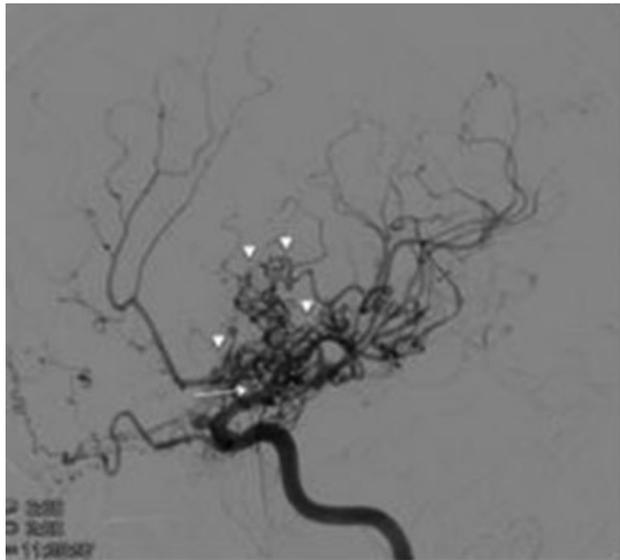
Case 1–Female patient, 71 years old, hypertensive, presented with acute hemiplegia on the left side and lowering level of consciousness. Magnetic resonance imaging (MRI) of the cerebrum showed a right intraparenchymal frontoparietal hematoma (►Fig. 1). Cerebral angiography showed occlusion of the terminal portions of the carotid arteries (►Fig. 2).

### Case Report

Case 1–Female patient, 71 years old, hypertensive, presented with acute hemiplegia on the left side and lowering level of consciousness. Magnetic resonance imaging (MRI) of the cerebrum showed a right intraparenchymal frontoparietal hematoma (►Fig. 1). Cerebral angiography showed occlusion of the terminal portions of the carotid arteries (►Fig. 2).



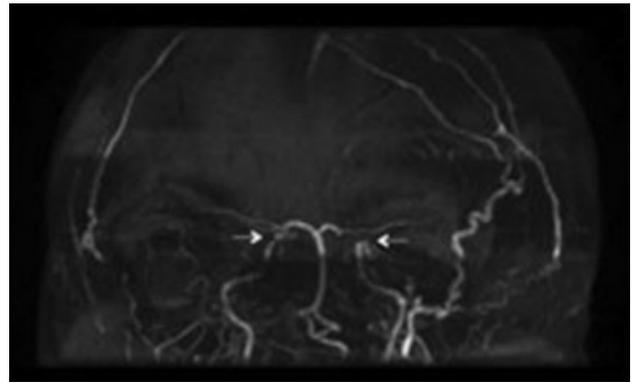
**Fig. 3** Digital angiography image by subtraction – anteroposterior incidence – showing severe stenosis of the supraclinoid segment of the internal carotid arteries and their terminal branches (anterior and middle cerebral arteries), with collateral vessels dilated at the base of the skull (“Moyamoya vases”). The right internal carotid artery; D-F, left internal carotid artery. The venous phase is normal (C and F).



**Fig. 4** Digital angiography image by subtraction – profile incidence – showing severe stenosis of the terminal portion of the left internal carotid artery and its branches (white arrow), with dilated collateral vessels at the base of the skull (“Moyamoya vases” – arrow heads).

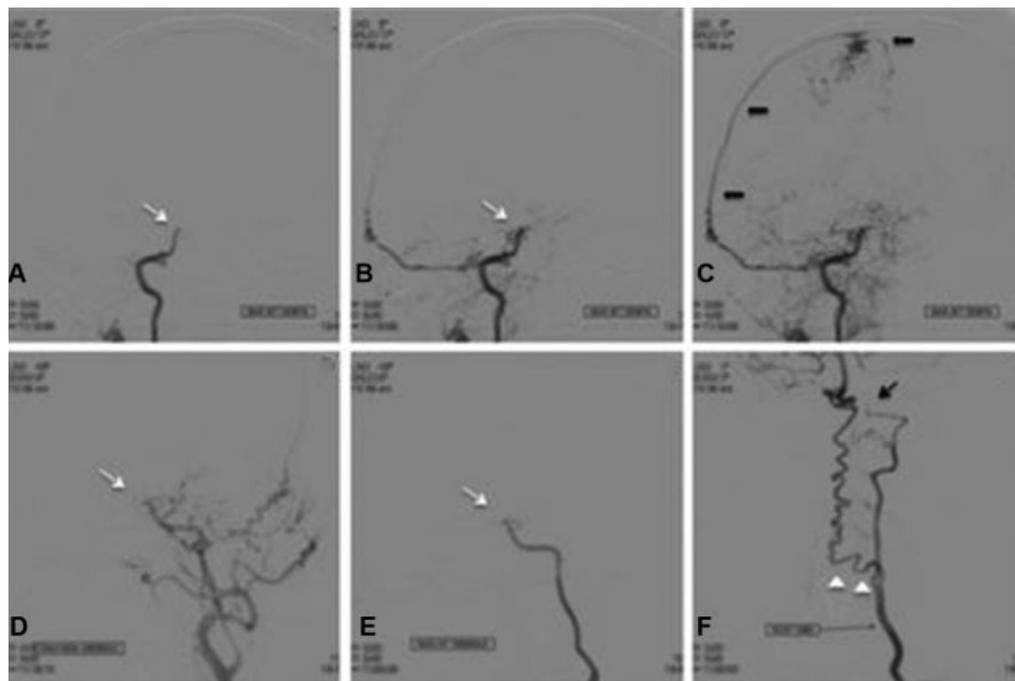
Complementary investigation was then performed with digital angiography by subtraction, which showed severe internal bilateral stenosis of the supraclinoid carotid arteries, with the presence of dilated collateral vessels at the base of the skull (►Figs. 3 and 4). Additional exams showed the presence of a falcemic trait (hemoglobin dosage: S  $\frac{1}{4}$  36%).

Case 2–Female patient, 40 years old, with no previous history of comorbidities, arrived at our service with chronic

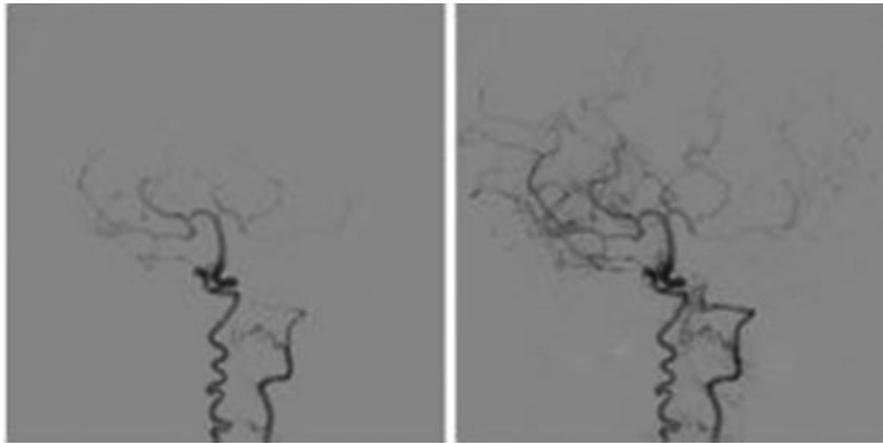


**Fig. 5** Cerebral angioMRI image – arterial phase, 3D TOF – showing blood flow interruption in the intracranial segments of the internal carotid arteries (arrows).

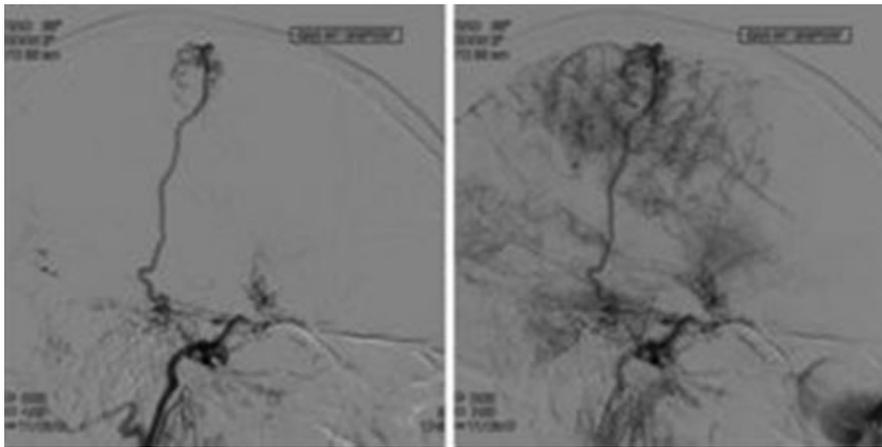
headache refractory to various types of treatment. Neurological examination was normal. Cerebral angioMRI showed occlusion of the supraclinoid carotid arteries and vertebral arteries in their intracranial segments (►Fig. 5). Digital angiography was performed and evidenced subocclusive stenosis of the main intracranial arteries, with the encephalic circulation predominantly nourished by anastomosis between the left vertebral artery (intraforaminal segment – V2) with the anterior spinal artery. This, in turn, vascularized the basilar artery, the posterior cerebral arteries and the vessels of the anterior circulation through the posterior communicating arteries (►Figs. 6 and 7). A pial vascularization was observed through the marginal tentorial artery (also known as the Bernasconi and Cassinari artery), originated in



**Fig. 6** Digital angiography image by subtraction showing: A, B, D, E, subocclusive stenosis of the supraclinoid segment of the internal carotid arteries (white arrows); transdural anastomosis with pial vascularization originated in the intracavernous segment of the right internal carotid artery (black arrows); F, anastomosis between the intraforaminal segment (V2) of the left vertebral artery and the anterior spinal artery (arrowheads), which meets the increased caliber. Occlusion of the intracranial segment of the vertebral artery (black arrow) is observed.



**Fig. 7** Digital angiography image by subtraction with injection in the left vertebral artery – anteroposterior incidence – showing vascularization of the basilar artery and its branches through anastomosis with the anterior spinal artery.



**Fig. 8** Digital angiography image by subtraction – profile incidence – showing pial vascularization through the marginal tentorial artery (Bernasconi and Cassinari artery) originated in the intracavernous segment of the right internal carotid artery.

the intracavernous segment of the right internal carotid artery (–Fig. 8).

## Discussion

Moyamoya disease is a rare pathology, with a reported incidence of 0.086 cases per 100,000 individuals.<sup>8,9</sup> Originally thought to affect predominantly people of Asian origin, it is now observed to afflict people from various ethnic backgrounds around the world. The incidence among females is two times higher than in males.<sup>10,11</sup> It is characterized by the progressive occlusion of the terminal portions of the carotid arteries and their main branches in the circle of Willis (anterior and middle cerebral arteries), with the compensatory development of a network of collateral vessels at the base of the skull (called “Moyamoya vases”). It usually affects the two cerebral hemispheres and has two peaks of presentation: the first, around the age of 5 years old, and the second, after 40 years old. The posterior circulation is affected in a less frequent way.<sup>3</sup> Most children with this pathology manifest symptoms resulting from cerebral ischemia, while adults present, more frequently, with intracranial hemorrhage.<sup>3,9,12</sup> In our study, we report the cases of two adult patients who presented to our service with distinct

clinical manifestations: one of them, with severe and focal neurological deficit due to hemorrhage; the other, with progressive headaches and normal neurological examination. Extensive diagnostic investigation with neuroimaging exams and laboratory tests was performed, including thyroidopathy research, sickle cell anemia and atherosclerotic disease.

## Conclusion

Moyamoya disease is a pathology of rare incidence and difficult diagnosis, with multiple forms of clinical presentation. Its diagnosis should be suspected in the clinical context of patients with neurological alteration and progressive occlusion of the carotid arteries and their intracranial terminal branches.

### Conflicts of Interests

The authors declare that there are no conflicts of interests.

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