Development of a de novo arteriovenous malformation after bilateral revascularization surgery in a child with moyamoya disease

Case report

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The development of a de novo arteriovenous malformation (AVM) in patients with moyamoya disease is extremely rare. A 14-year-old girl developed an AVM in the right occipital lobe during the 4-year postoperative period following successful bilateral revascularization surgeries. She suffered a transient ischemic attack with hemodynamic compromise of the bilateral hemispheres at the age of 10 years. Results of an initial examination by 1.5-T MRI and MR angiography satisfied the diagnostic criteria of moyamoya disease but failed to detect any vascular malformation. Bilateral direct and indirect revascularization surgeries in the anterior circulation relieved her symptoms, and she underwent MRI and MR angiography follow-up every year after surgery. Serial T2-weighted MRI revealed the gradual appearance of flow voids in the right occipital lobe during the follow-up period. Magnetic resonance angiography ultimately indicated the development of an AVM 4 years after these surgeries when catheter angiography confirmed the diagnosis of an AVM in the right occipital lobe. The AVM remained asymptomatic, and the patient remained free of cerebrovascular events during the time she was observed by the authors. Acquired AVM in moyamoya disease is extremely rare, with only 3 pediatric cases including the present case being reported in the literature. The development of a de novo AVM in a postoperative patient with moyamoya disease appears to be unique, and this case may provide insight into the dynamic pathology of AVMs.

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Key Words • arteriovenous malformation • moyamoya disease • de novo development • vascular disorders

Arteriovenous malformation (AVM) is a congenital cerebrovascular disease that is believed to appear at the early embryonic stage of 3 weeks; however, evidence suggesting its dynamic nature of rapid growth, recurrence after total removal, and de novo development, despite its “congenital” nature, is increasing.1,2 Moyamoya disease is a chronic occlusive cerebrovascular disease with unknown etiology characterized by bilateral steno-occlusive changes in the terminal portion of the internal carotid artery and an abnormal vascular network at the base of the brain.3 Although the rare association of moyamoya disease with an AVM has been documented in detail, the development of de novo AVMs in patients with moyamoya disease is extremely rare; it has been reported in only 2 pediatric cases.3,4 Here, we describe a 10-year-old girl with ischemic-onset moyamoya disease who underwent successful bilateral revascularization surgeries and developed acquired AVM in the occipital lobe during the 4-year follow-up period.

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A 10-year-old girl had repeated weakness in her bilateral upper extremities, was diagnosed with moyamoya disease, and was subsequently transferred to our hospital. A neurological examination showed no abnormalities, and 1.5-T MR angiography revealed steno-occlusive changes at the terminal portion of the bilateral internal carotid arteries (Fig. 1A). No abnormality was found in the posterior circulation, including the right posterior ce-

Abbreviations used in this paper: AVM = arteriovenous malformation; PCA = posterior cerebral artery.
rebral artery (PCA) territory on MR angiography (Fig. 1A). Axial T2-weighted MRI showed several flow voids at the bilateral basal ganglia (asterisks in B), but none in the occipital lobe (C). Flow voids were evident in the right occipital lobe 3 (arrow in E) and 4 (arrow in F) years after surgeries, while it was equivocal 1 year after the surgeries (D). G: Magnetic resonance angiogram obtained 4 years after the surgeries, demonstrating apparent visualization of the nidus in the right occipital lobe (arrow). H: Right anterior oblique view of the right vertebral angiogram revealing a right occipital lobe AVM (arrow).

**Fig. 1.** A: Initial MR angiogram demonstrating steno-occlusive changes at the terminal portions of the bilateral internal carotid arteries (arrows). B–F: Temporal profile of T2-weighted MR imaging before surgery (B and C) and 1 year (D), 3 years (E), and 4 years (F) after bilateral direct/indirect revascularization surgeries. Preoperative MR images demonstrated several flow voids at the bilateral basal ganglia (asterisks in B), without any abnormality in the right occipital lobe (C). Flow voids were evident in the right occipital lobe 3 (arrow in E) and 4 (arrow in F) years after surgeries, while it was equivocal 1 year after the surgeries (D).

We demonstrated the temporal profile of MRI findings for the development of AVM during a 4-year postoperative period after bilateral revascularization surgeries for moyamoya disease in a 14-year-old girl. The exact mechanism by which an acquired AVM developed in the vascular territory supplied by the PCA without steno-occlusive changes is unknown. In light of the absence of hemodynamic compromise in the occipital lobe, as shown by SPECT pre- and postoperatively, cerebral ischemia was not likely to be the major trigger for the development of the acquired AVM. Because we did not perform extended indirect pial synan-

**Discussion**

We demonstrated the temporal profile of MRI findings for the development of AVM during a 4-year postoperative period after bilateral revascularization surgeries for moyamoya disease in a 14-year-old girl. The exact mechanism by which an acquired AVM developed in the vascular territory supplied by the PCA without steno-occlusive changes is unknown. In light of the absence of hemodynamic compromise in the occipital lobe, as shown by SPECT pre- and postoperatively, cerebral ischemia was not likely to be the major trigger for the development of the acquired AVM. Because we did not perform extended indirect pial synan-
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giosis and/or an additional bur hole technique in the posterior circulation, the initiation of AVM development could not have been due to the iatrogenic arteriovenous fistula at the initial surgery for moyamoya disease. Alternatively, based on the absence of catheter angiography at the initial onset of moyamoya disease as well as on the classic observation that AVM is a congenital disease, we do not completely rule out the possibility that the patient initially had a micro-VM or small arteriovenous fistula before bilateral revascularization, which was not evident on MRI. From the viewpoint of basic pathology, moyamoya disease and an AVM are known to have similar biological backgrounds of the increased expression of angiogenic factors such as vascular endothelial growth factor, as well as inflammatory molecules, including tumor necrosis factor-α, interleukin-6, and matrix metalloproteinases.4,7 The expression of these molecules in moyamoya disease could have caused the acquired AVM to develop in the present case. Alternatively, the development of the de novo AVM in the present case may provide an insight into elucidating the dynamic pathology of AVMs.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Fujimura. Acquisition of data: Fujimura, Kimura, Ezura, Niizuma. Analysis and interpretation of data: Fujimura. Drafting the article: Fujimura. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Fujimura. Administrative/technical/material support: Kimura, Niizuma. Study supervision: Tominaga.

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