Disappearance and development of cerebral aneurysms in moyamoya disease

Case report

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The authors report a case of multiple cerebral aneurysms associated with moyamoya disease, in which the reduction and disappearance of bilateral internal carotid artery aneurysms and the new development and enlargement of basilar artery aneurysms were demonstrated on sequential cerebral angiograms. Possible mechanisms of the spontaneous disappearance and development of cerebral aneurysms in association with moyamoya disease are discussed.

KEY WORDS  • cerebral aneurysm  • moyamoya disease  • internal carotid artery  • basilar artery  • multiple aneurysms

Cases of cerebral aneurysms associated with moyamoya disease have been reported occasionally. We report another such case, which is unique in that sequential angiographic studies revealed the reduction and disappearance of internal carotid artery (ICA) aneurysms and the development and enlargement of basilar artery (BA) aneurysms in conjunction with the advancement of moyamoya disease.

Case Report

This 44-year-old woman, who had been suffering from hypertension for a long time, visited our service on September 11, 1978, complaining of progressive left hemiparesis. Eight months prior to the visit, she had had an episode of transient left hemiplegia. The patient was alert, with an elevated blood pressure of 180/110 mm Hg. Slight left hemiparesis with hypalgesia was noted, predominantly in the upper limb; there were no other neurological abnormalities.

Course. The patient became transiently aphasic and disoriented on January 20, 1979, and was admitted to our ward for further examination on April 28, 1979. Computerized tomography performed at that time (Fig. 1) revealed massive low-density areas in the right frontal lobe and in the left cerebral hemisphere, with mildly dilated ventricles and cortical atrophy. Small localized high-density nodules were demonstrated at the bilateral edges of the basal cistern. The inside of the nodules enhanced fully on administration of con-
Contrast medium. Four-vessel cerebral angiography disclosed moyamoya disease associated with multiple cerebral aneurysms of the C1 portion of the right ICA, the left ICA-posterior communicating artery (PCoA), and the right basilar artery-superior cerebellar artery (BA-SCA). The patient was treated only conservatively, according to the wish of her family, and discharged. Her symptoms gradually worsened on the following day to include dementia, motor aphasia, and tetraparesis.

The patient underwent follow-up cerebral angiography on October 31, 1980, and again on January 14, 1982. At this latter admission, she was demented, uncooperative, irritable, and had a voracious appetite. She had complete motor aphasia and responded to our commands only by a shout. Severe tetraparesis predominantly in the right side made it impossible for her to walk. Her blood pressure was 110/80 mm Hg. Swallowing disturbance and urinary and fecal incontinence were observed. No abnormalities were found in the laboratory data. Electroencephalography showed low-voltage slow waves diffusely in the left cerebral hemisphere.

**Angiographic Findings.** At the first admission in April, 1979, right carotid angiography (Fig. 2 upper left) revealed a slender cervical ICA, markedly stenosed or occluded above and distal to the origin of the PCoA. An irregularly shaped aneurysm (14 × 13 × 10 mm in size), which had a rather large sac volume in proportion to its orifice, was found on the C1 portion of the ICA. There was a fine network of moyamoya vessels with poor filling in the immediately proximal portion of the anterior and middle cerebral arteries. Left carotid angiography (Fig. 2 upper right) showed marked stenosis above and distal to the origin of the large PCoA, and a saccular ICA-PCoA aneurysm (6 × 6 × 5 mm in size). Moyamoya vessels were faintly demonstrated, with poor filling in the immediately proximal portion of the anterior and middle cerebral arteries. The embryonal posterior cerebral artery was well opacified. Retrograde filling of the

**Fig. 2.** Cerebral angiograms taken on the first admission in April, 1979. *Upper Left:* Right carotid angiogram showing a markedly stenosed or occluded internal carotid artery (ICA) above and distal to the origin of the posterior communicating artery (PCoA). Note the irregularly shaped C1 aneurysm. Moyamoya vessels, with poor filling in the immediately proximal portion of the anterior and middle cerebral arteries, are demonstrated. *Inset:* Enlarged view of the aneurysm. *Upper Right:* Left carotid angiogram showing a markedly stenosed ICA above and distal to the origin of the large PCoA. Note the saccular ICA-PCoA aneurysm. Moyamoya vessels are faintly demonstrated. The embryonal posterior cerebral artery is well opacified. *Lower:* Bilateral retrograde brachial angiogram showing a well developed vertebrobasilar system with a small saccular basilar artery-superior cerebellar artery aneurysm.
Aneurysms associated with moyamoya disease

Fig. 3. Follow-up cerebral angiograms taken on the second admission in October, 1980. Upper Left: The degree of stenosis at the right C1 portion of the internal carotid artery (ICA) has progressed, with complete disappearance of the C1 aneurysm. Upper Right: The degree and extension of stenosis of the left distal ICA has progressed. Note the disappearance of the posterior communicating artery (PCoA) and the reduction of the ICA-PCoA aneurysm. Lower: Enlargement of the basilar artery (BA)-superior cerebellar artery aneurysm is demonstrated, accompanied by a newly developed saccular aneurysm at the top of the BA.

distal portion of the middle and anterior cerebral arteries was observed via the leptomeningeal and rete mirabile anastomoses. Bilateral retrograde brachial angiography (Fig. 2 lower) showed a well developed vertebralbasilar system with a small saccular BA-SCA aneurysm (2 × 1.8 × 1.5 mm in size). Although the left posterior cerebral artery was not opacified, the right one was well opacified. Retrograde filling of the distal portion of the middle and anterior cerebral arteries was also observed.

At the second admission in October, 1980, the degree of stenosis at the right C1 portion of the ICA had progressed, with complete disappearance of the C1 aneurysm (Fig. 3 upper left). The degree and extension of stenosis of the left distal ICA had progressed, with the disappearance of the PCoA and the reduction of the ICA-PCoA aneurysm to 2.5 × 2.5 × 3 mm (Fig. 3 upper right). However, enlargement of the BA-SCA aneurysm to 3.5 × 2.5 × 2 mm was demonstrated, with a newly developed saccular aneurysm (2 × 2 × 2 mm in size) at the top of the BA (Fig. 3 lower).

At the third admission in January, 1982, the right C1 portion of the ICA was slightly opacified, but the C1 aneurysm was still not demonstrated (Fig. 4 upper left). Marked stenosis of the left distal ICA extended just above and distal to the origin of the ophthalmic artery. The ICA-PCoA aneurysm had disappeared completely (Fig. 4 upper right). However, further enlargement of the BA-SCA aneurysm to 4 × 2.5 × 2.5 mm and of the aneurysm at the top of the BA to 3.8 × 3.8 × 3 mm was demonstrated (Fig. 4 lower). Figure 5 is a schematic illustration of the circle of Willis and the associated cerebral aneurysms in this case.

Discussion

Cerebral aneurysms associated with cerebrovascular moyamoya disease have been classified as “peripheral artery aneurysms” and “major artery aneurysms.”4,7,8 The former type are located at the basal ganglia, near or in the abnormal fine network of moyamoya vessels. They are presumed to be pseudoaneurysms, indicating the initial hemorrhagic points of moyamoya vessels, because of their location and transient appearance on the follow-up angiograms.7,10 Recently, some peripheral artery aneurysms have been histologically proven to be pseudoaneurysms.4,11
Major artery aneurysms are ordinary aneurysms arising from the circle of Willis, and are considered to be true aneurysms; they may be malformations, coincidental, or hemodynamically induced. Peripheral artery aneurysms frequently disappear spontaneously, but there has been no report of reduction or disappearance of major artery aneurysms. In our case, the disappearance of the right C1 aneurysm and the reduction and final disappearance of the left ICA-PCoA aneurysm were observed, accompanied by the progression of stenosis of the distal ICA's. The velocity of blood flow within an aneurysmal sac is said to be disproportional to the sac volume. Both the right C1 aneurysm and the left ICA-PCoA aneurysm had a rather large sac volume in proportion to their orifice; thus, the stasis of blood flow within the sac might allow intra-aneurysmal thrombosis. The characteristic pathological changes in the distal ICA's associated with moyamoya disease include eccentric, dense, and fibrous thickening of the intima, combined with a slight splitting of the internal elastic lamina and thinning of the tunica media. Enlargement of these lesions in our patient is presumed to have caused not only stenosis of the parent vessels distal and proximal to these ICA aneurysms, but also the deformation of the aneurysms to form a relatively narrow neck, and this was considered responsible for their disappearance.

The precise relationship of the development of the right C1 aneurysm to the pathological changes in the distal ICA is unknown. In a report on an experimental study of the genesis of moyamoya disease, Kasai et al., noted that, prior to organic stenosis at the carotid fork, an arterial wall injury resulting from arteritis may initially occur. Although in our case the left ICA-PCoA aneurysm might have developed coincidentally, the increased blood flow in the embryonal posterior cerebral artery via the large PCoA, caused by the progressive stenosis of the distal ICA, is considered to have played a significant role in the development of the ICA-PCoA aneurysm.

Even though cerebral aneurysms arise more frequently in the internal carotid system than in the vertebrobasilar system, cerebral aneurysms associated with moyamoya disease occur more frequently in the vertebrobasilar system. Kodama and Suzuki and Adams, et al., considered the effect of hemodynamic stress on the development of BA aneurysms, and speculated that cerebral blood flow might be increased in the vertebrobasilar system instead of the ICA system due to the chronic progressive stenosis or obliteration of the bilateral ICA's. Our case is unique in

Fig. 4. Follow-up cerebral angiograms taken on the third admission in January, 1982. Upper Left: The right C1 portion of the internal carotid artery (ICA) is slightly opacified, although the C1 aneurysm is still not demonstrated. Upper Right: Marked stenosis of the left distal ICA has extended just above and distal to the origin of the ophthalmic artery. Note the complete disappearance of the ICA-posterior communicating artery aneurysm. Lower: Further enlargement of both the basilar artery (BA)-superior cerebellar artery aneurysm and the aneurysm at the top of the BA is demonstrated.
Aneurysms associated with moyamoya disease

![Diagram of the circle of Willis and associated cerebral aneurysms]

**Fig. 5.** Schematic illustration of the circle of Willis and the associated cerebral aneurysms in this case. On the first admission in April, 1979 (left), multiple (right C1 portion of the internal carotid artery (ICA), left ICA-posterior communicating artery (PCoA), and right basilar artery-superior cerebellar artery (BA-SCA)) cerebral aneurysms were associated with marked stenosis of the bilateral distal ICA’s in association with moyamoya disease. On the second admission in October, 1980 (center), the right C1 aneurysm had completely disappeared and the left ICA-PCoA aneurysm was reduced. There was progressive stenosis of the bilateral distal ICA’s. However, the right BA-SCA aneurysm was enlarged, and there was a newly developed aneurysm at the top of the BA. On the third admission in January, 1982 (right), the left ICA-PCoA aneurysm had disappeared completely, and there was further progressive stenosis of the distal ICA. Both the BA-SCA aneurysm and the aneurysm at the top of the BA were enlarged.

that the relationship of hemodynamic stress to the development and enlargement of BA aneurysms in association with moyamoya disease was verified by follow-up angiography (Fig. 4).

**References**


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