Moyamoya disease associated with aneurysm

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Five cases of moyamoya disease associated with aneurysm are reported. In three cases, the aneurysms were located at the peripheral portion of the posterior choroidal artery, and in two at the basilar artery. Based on these cases, the symptoms and mechanisms of formation of aneurysms in moyamoya disease are discussed.

KEY WORDS • moyamoya disease • aneurysm • pseudo-aneurysm • posterior choroidal artery • basilar aneurysm • ventricular hemorrhage • subarachnoid hemorrhage

There have been few reported cases of moyamoya disease, and fewer still associated with aneurysms. Up to now, we have experienced five such cases; in three patients the aneurysms arose from the posterior choroidal artery, and in two from the basilar artery. Based on these five cases, we discuss the formation process of these aneurysms and their symptomatic occurrence in moyamoya disease.

Case Reports

Case 1

This 16-year-old girl suddenly commenced vomiting on December 24, 1968, followed by loss of consciousness. On admission, the patient was comatose, and had right hemiparesis and a stiff neck. The lateral view of a left carotid angiography, performed on December 28, demonstrated the narrowing of the C1, A1, and M1 portion. The anterior, middle, and posterior cerebral arteries were not visible. Moyamoya vessels at the base of the brain were visualized as well as an aneurysm shadow at the peripheral portion of the left posterior choroidal artery. The anteroposterior (AP) view confirmed an aneurysm shadow approximately 3 mm in diameter located near the intersecting point of the lateral and upper walls of the lateral ventricle.

On the 12th day after the onset of symptoms, the patient became almost alert, and by the 35th day she was able to walk independently. A repeat left carotid angiogram, performed 42 days after admission, showed the disappearance of the aneurysm shadow in both the lateral and AP views.

Case 2

This 39-year-old man was admitted to a local hospital on May 5, 1973, with complaints of sudden headache and vomiting. Neurological examination revealed stiff neck and Kernig's sign. The cerebrospinal fluid (CSF) was bloody.

A left carotid angiogram was performed on the 6th day after the onset of symptoms. On the 13th day, a right carotid angiogram showed, in the lateral view, severe stenosis at the carotid fork together with a small moyamoya network at the base of the brain.
Neither the anterior nor the middle cerebral arteries were seen. On the other hand, the posterior communicating artery was dilated and the posterior cerebral artery was clearly filled. From the findings, a diagnosis of moyamoya disease was made, and conservative therapy was selected. On the 28th day after the initial onset, the patient experienced a recurrence of severe headache, but he remained alert. Neurological findings revealed stiff neck, but no motor disturbance was found. The CSF was stained pink. The patient was transferred to our hospital on July 6.

Left carotid angiography was performed on the 43rd day after the second attack. On the 47th day, a second right carotid angiogram showed, in the lateral view, a round aneurysm shadow approximately 3 mm in diameter at the peripheral portion of the right posterior choroidal artery (Fig. 1 left). This finding had not been seen on the earlier angiograms. This shadow was confirmed on the AP view and remained visible even on the capillary phase. It was located 29 mm from the midline, near the intersecting point of the lateral and upper walls of the lateral ventricle.

After discharge, a third right carotid angiogram was performed, about 11 months after the second attack. The lateral and AP views showed the disappearance of the aneurysm shadow (Fig. 1 right).

Case 3

This 48-year-old woman had suffered a cerebral stroke at the age of 32 years. Fortunately consciousness was recovered within a week and she was discharged without recurrence. On October 15, 1974, she developed sudden frontal headache and vomiting. On admission, she was alert, but had a right hemiparesis. The CSF was xanthochromic.

Left carotid angiography was performed on the 15th day after the onset of symptoms, and right carotid angiography on the 17th day. On the lateral view of the left carotid angiogram, stenotic changes were found at the peripheral portion of the internal carotid artery, and a small moyamoya network was revealed at the base of the brain. A round aneurysm shadow, 2 mm in diameter, was...
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Fig. 3. Case 4. Anteroposterior views of the left vertebral angiogram. Left: Four weeks after onset. Arrow indicates the aneurysm shadow at the basilar bifurcation. Right: Fifteen months after onset. Arrow indicates the aneurysm shadow, which is slightly larger than before.

found at the peripheral portion of the left posterior choroidal artery. This shadow was still visible on the venous phase taken 6 seconds later. On the anteroposterior view, the same aneurysm shadow, measuring 3 mm in diameter, was confirmed 28 mm from the midline. On January 30, 1975, a left carotid angiogram showed the aneurysm shadow had disappeared completely.

Case 4

This 35-year-old man developed severe headache and vomiting without motor disturbance or loss of consciousness. On February 19, 1974, the 4th day after the onset of symptoms, a left carotid angiogram, on the lateral view (Fig. 2), showed narrowing of the internal carotid artery at the carotid fork and a small moyamoya network at the base of the brain. Main cerebral arteries were not revealed at all. Left vertebral angiography was performed on March 5, and showed on the lateral view an aneurysm shadow of $5 \times 7$ mm at the basilar bifurcation. The AP view revealed the aneurysm presenting 1.5 mm to the right and above the basilar bifurcation (Fig. 3 left).

Diagnosis of moyamoya disease associated with basilar aneurysm was established from these findings. Conservative therapy was chosen with close observation of the course of the aneurysm. Repeat left vertebral angiography was performed on April 15, 1975, 14 months after the onset. The AP view disclosed that the aneurysm shadow had become slightly larger and more rounded (Fig. 3 right).

Case 5

This 42-year-old man suddenly lost consciousness and was admitted to our department. A diagnosis of subarachnoid hemorrhage was made, based on a finding of bloody CSF. The lateral views of left and right carotid angiograms revealed moyamoya vessels at the base of the brain. The lateral view of a left vertebral angiogram revealed an aneurysm shadow at the bifurcation of the basilar artery. The shadow looked like two
aneurysms projecting in opposite directions — one upward and the other downward.

The diagnosis of moyamoya disease associated with basilar aneurysm was made, and a craniotomy was performed on May 30, 1975. Two aneurysms were found; one, $6 \times 8 \times 9$ mm in size, at the bifurcation of the basilar artery, and the other, $5 \times 6 \times 7$ mm in size, at the right superior cerebellar artery junction. Yellow-brown pigmentation and adhesion were observed around the aneurysms, indicating ruptures of the aneurysms. The aneurysms were treated by ligation and muscle wrapping. Vertebral angiograms on April 24, 1977, revealed no aneurysm shadow.

Discussion

Previous reports state the low incidence of moyamoya disease associated with aneurysms.\(^2,5,6\) In our clinic, however, five cases with aneurysms were found in 56 moyamoya cases (21 children and 35 adults). Since all five cases were among the 35 adult patients, the incidence in adults is 14%.

In three of our five cases, the aneurysm shadows were seen at the peripheral portion of the posterior choroidal artery. Each aneurysm was located in the brain tissue around the upper lateral edge of the lateral ventricle. These aneurysm shadows look different from the ordinary saccular aneurysms arising from the circle of Willis. Furthermore, four of them disappeared completely on the follow-up angiograms.

From these facts, we assume that these aneurysm shadows are not true aneurysms but pseudo-aneurysms, which indicate the bleeding point in the brain tissue. The course of these pseudo-aneurysms may perhaps be explained in the following way: initially a small artery may rupture near the lateral ventricle. The blood penetrates into it, but the small blood clot, or, in other words, the pseudo-aneurysm, remains. The blood, which has penetrated into the lateral ventricle, circulates into the subarachnoid space. Therefore, the symptoms and signs may be misunderstood as those of subarachnoid hemorrhage. Later, the blood clot vanishes by complete absorption.

On the other hand, the two basilar aneurysms are regarded as saccular aneurysms. The aneurysm in Case 4, contrary to the pseudo-aneurysms, grew slightly instead of vanishing. In Case 5, the aneurysms were confirmed at the craniotomy to be saccular and ruptured aneurysms.

In moyamoya disease, as the chronic progressive stenosis or occlusion advances at the carotid fork bilaterally, blood flow gradually decreases in the internal carotid arterial system. On the contrary, the blood flow in the vertebrobasilar system increases, causing high pressure on the vessel wall. When a medial defect of the basilar arterial wall exists, there is a strong probability that the aneurysm may develop at that point.

Cases may present in which an aneurysm has already been formed before moyamoya disease occurs. In such cases, the aneurysm growth is accelerated by the progress of moyamoya disease. Hence, it is concluded that increased blood flow of the vertebrobasilar arterial system greatly effects aneurysm growth, whether the formation is before or after the occurrence of moyamoya disease.

It has been generally believed that the symptomatic recognition of moyamoya disease in adults is due to subarachnoid hemorrhage, and the cause of the hemorrhage has been regarded as the rupture of the dilated moyamoya vessels in the subarachnoid space. There is no clear supporting evidence for this assumption. Our first three cases suggest that one of the mechanisms of symptomatic occurrence may be the ventricular hemorrhage following the bleeding in the brain tissue. Our proposition is supported by the fact that there have been several patients with intracerebral hematoma and ventricular hemorrhage among the autopsied moyamoya cases.\(^1,11\) From the other two cases associated with basilar aneurysms, we strongly suspect another mechanism, that is, subarachnoid hemorrhage due to the rupture of the basilar aneurysm.

References


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