Moyamoya disease with repeated intraventricular hemorrhage due to aneurysm rupture

Report of two cases

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Two cases of moyamoya disease associated with repeated intraventricular hemorrhage are reported. The origin of bleeding was thought to be a distal aneurysm of the choroidal artery. The aneurysms were confirmed radiologically and histologically. The relationship between moyamoya disease and aneurysms is discussed, and a treatment proposed.

Key Words • cerebral aneurysm • intraventricular hemorrhage • moyamoya disease

The most common clinical manifestation of moyamoya disease in adults is subarachnoid, intra-cerebral, or intraventricular hemorrhage. The incidence of hemorrhage exceeds 60% in adults presenting with moyamoya disease and is 10% in children. Initially, rupture of the affected vessel was thought to be responsible for such a hemorrhage; recently, however, an associated aneurysm has been considered as a possible source of bleeding. To our knowledge, moyamoya disease with repeated intraventricular hemorrhage due to a peripheral aneurysm has not been reported previously. We have recently treated two such cases, which we describe here.

Case Reports

Case 1

This previously healthy 41-year-old woman experienced the sudden onset of headache followed by loss of consciousness. On admission to another hospital, she regained consciousness without focal signs. A computerized tomography (CT) scan showed an intraventricular hemorrhage, mainly involving the left lateral ventricle (Fig. 1 left). Left carotid angiography revealed occlusion of the terminal portion of the right internal carotid artery (ICA) associated with extensive moyamoya vessels, and demonstrated the characteristic appearance of moyamoya disease as well as an aneurysm in the distal portion of the posterior choroidal artery (Fig. 2 left). The patient's condition gradually improved with conservative treatment; however, she again experienced a sudden attack of headache 2 weeks after the initial ictus. A CT scan showed a new intraventricular hemorrhage.

Examination. The patient was admitted to our clinic 3 weeks after the second hemorrhage. A CT scan obtained upon admission revealed a partially thrombosed aneurysm on the left lateral ventricle (Fig. 1 right). Follow-up left carotid angiography showed that the size of the aneurysm was markedly increased (Fig. 2 right).

Operation. Four weeks after the second hemorrhage, direct surgery to the aneurysm was performed via the

![Fig. 1. Case 1. Left: Unenhanced computerized tomography (CT) scan obtained after the first hemorrhage, showing an intraventricular hemorrhage mainly involving the left lateral ventricle. Right: Unenhanced CT scan obtained 3 weeks after the second hemorrhage showing a partially thrombosed aneurysm on the left lateral ventricular wall (arrow).](image-url)
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left parieto-occipital approach. The parent artery was trapped and the aneurysm, which was surrounded by an old clot, was excised.

Postoperative Course. The patient’s postoperative course was complicated by right hemiparesis. She was discharged to a rehabilitation facility. Six months after the initial operation, her hemiparesis improved; hoping to decrease hemodynamic stress in the involved vessels and to ameliorate the cerebral circulation, we performed bilateral superficial temporal artery-middle cerebral artery (STA-MCA) anastomoses. She was discharged without additional neurological deficits.

Pathological Examination. Histological examination of the surgical specimen showed that the wall of the lesion was composed of collagen fibers and fibrin; no elastic fibers were identified. The pathological diagnosis was a pseudoaneurysm (Fig. 3).

Case 2

This previously healthy 48-year-old woman was admitted to a local hospital with complaints of sudden severe headaches, nausea, and vomiting. On admission she was fully conscious with focal signs. Computerized tomography scans obtained on the day of onset revealed an intraventricular hemorrhage, primarily in the left lateral ventricle, and a small infarction in the right frontal lobe (Fig. 4). A right carotid angiogram showed occlusion of the terminal portion of the right ICA associated with moyamoya vessels at the base of the brain. A left carotid angiogram demonstrated typical findings of moyamoya disease and a small aneurysmal shadow at the distal portion of the hypertrophic anterior choroidal artery (Fig. 5 left). She was treated conservatively.

Examination. The patient was admitted to our clinic 30 days after the ictus for further evaluation; her gen-
eral status was good and she was neurologically normal except for a slight headache. Five days after admission, she again experienced a sudden severe headache. A CT scan revealed a new intraventricular hemorrhage and, after contrast enhancement, a small aneurysmal shadow was seen on the left lateral ventricular wall (Fig. 6). A left carotid angiogram demonstrated definite enlargement of the aneurysm (Fig. 5 right).

Operation. Five days after the second hemorrhage, a small left frontal craniotomy was performed to avoid damage to the vital collateral vessels of the STA and middle meningeal artery. The left lateral ventricle was entered via the transcallosal approach; however, the aneurysm itself could not be identified. The parent artery was clipped, after which a left STA-MCA anastomosis was performed.

Postoperative Course. The patient’s postoperative condition was satisfactory except for a transient right hemiparesis. Three weeks after the initial operation, a right STA-MCA anastomosis was carried out without worsening of the neurological manifestation. She was discharged without neurological deficits.

Discussion

Reports on the association of moyamoya disease and aneurysm have increased in recent years. Typically, aneurysms associated with moyamoya disease arise from the vertebrobasilar system and are probably related to the increased flow in the posterior circulation resulting from stenosis of the ICA. This mechanism is probably similar to that involved in the development of aneurysms on the feeding arteries of arteriovenous malformations. The aneurysms are classified into two groups: those involving the circle of Willis (major artery aneurysms) and those located within the collateral or moyamoya vessels (peripheral artery aneurysms). Approximately 40% of associated aneurysms are of the peripheral type and found mainly at the distal portions of the perforating arteries and the anterior and posterior choroidal arteries. In most reported cases, the peripheral artery aneurysm was considered to be a pseudoaneurysm secondary to rupture of the fragile collateral vessel. Some of these aneurysms were no longer present on follow-up angiograms. Few of these aneurysms have been studied histologically because they were located peripherally. Only eight cases in the literature included histological findings (Table 1). In our Case 1, histological examination revealed a pseudoaneurysm. In our Case 2, no pathological confirmation of the nature of the aneurysm was obtained; however, the location
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<td>Mauro, et al., 1980</td>
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* SAH = subarachnoid hemorrhage; ICH = intracerebral hematoma; IVH = intraventricular hemorrhage; ACA = anterior choroidal artery; LSA = lenticulostriate artery; PChA = posterior choroidal artery; PCA = posterior cerebral artery.

and increase in size of the aneurysm were most consistent with a pseudoaneurysm. It is difficult to identify the exact origin of the bleeding in our two patients. However, it is likely that repeated hemorrhage is attributable to the aneurysms because of their anatomical situation and their enlargement after the second hemorrhage. Although it is only speculation to describe a hemodynamic relationship between occlusion of the ICA's and the occurrence of the aneurysm, the abnormal dilatation of the anterior and posterior choroidal arteries in our patients suggests that the wall of these arteries would be burdened by distention stress. Under such a hemodynamic stress, the formation of an aneurysm at a fragile portion of the parent artery is possible.

The clinical course of our patients raises several points with regard to the management of such cases. Patients with moyamoya disease associated with repeated intraventricular hemorrhage should be carefully investigated for a possible peripheral artery aneurysm. Direct surgery should be considered to prevent rupture if the aneurysm is thought to be the source of bleeding. On the other hand, direct surgery of an aneurysm arising from a peripheral artery is difficult because of the danger of damaging vital collateral vessels. We suggest that direct surgery of the aneurysm and cerebral revascularization should be performed at the same time to provide a better outcome in patients where the parent artery serves an important collateral route and preservation of the parent artery is thought to be impossible.

References

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