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原　　著

Intracerebral Hemorrhage with Moyamoya Disease: Source of Hemorrhage in Three Patients

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Summary

Cerebral aneurysms may be found in patients with Moyamoya Disease. They are classified into two groups, (1) a major artery aneurysm involving the circle of Willis, and (2) a peripheral artery aneurysm. Most peripheral artery aneurysms are usually considered to be pseudo-aneurysms. At angiography, we have found aneurysmal structures in the peripheral cerebral arteries in three patients with intracerebral hemorrhage associated with Moyamoya disease. In one patient, the aneurysmal outpouching could not be confirmed at autopsy. In the second patient, a true peripheral artery aneurysm was found to be a source of hemorrhage. In the remaining one patient, the angiographically seen aneurysmal structure was found to represent a conglomerate of tortuous, dilated collateral vessels.

Cerebral aneurysms may be seen occasionally in patients with intracranial hemorrhage associated with Moyamoya disease, and they are often considered to be a source of hemorrhage[3,8,9,13,15,19]. These aneurysms are often classified into two groups; a major artery aneurysm involving the circle of Willis, and a peripheral artery aneurysm, arising from the Moyamoya vessels or other small, distal arteries[3]. In most previous cases, the peripheral artery aneurysm seen in patients with Moyamoya disease has been considered to be a pseudo-aneurysm secondary to a rupture of the fragile collateral vessel.

In the past 4 years covering April 1979 through March 1983, we have seen three patients of Moyamoya disease associated with a massive intraventricular hemorrhage. In each of these patients, an aneurysmal outpouching was discovered at angiography. All three "aneurysms" were of peripheral type, and they were thought to be a source of hemorrhage, on the basis of its close proximity to the site of hematoma seen on CT, as well as the operative and/or autopsy findings. In the same period, aneurysms were found in the circle of Willis in a few other patients

Key Words: Cerebral aneurysm, Cerebral hemorrhage, Moyamoya disease, Pseudoaneurysm.
索引語：脳動脈瘤，脳内出血，モヤモヤ病，仮性動脈瘤.
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with cerebrovascular occlusive disease with more or less prominent development of collateral vessels. These latter cases were excluded from the present series, as their angiographic features were by no means typical of Moyamoya disease.

Report of cases

Case 1: A 38-year-old man was admitted elsewhere with complaints of sudden, severe headaches and vomiting, followed by a loss of consciousness. A spinal puncture yielded bloody cerebrospinal fluid, and the patient was immediately referred to us for further examination and treatment.

On admission, he was deeply comatose, responding to painful stimuli by decerebrate posturing. A right carotid angiogram showed an occlusion of the terminal portion of the right internal carotid artery and Moyamoya vessels in the base of the brain. The right anterior choroidal artery was hypertrophic and an aneurysmal shadow was seen in its peripheral portion (Figure 1). A left carotid angiogram also showed an occlusion of the terminal left internal carotid artery associated with extensive Moyamoya vessels.

A right ventricular drainage was performed, but the condition of the patient failed to improve and he expired next day. An autopsy confirmed a diagnosis of Moyamoya disease associated with a massive intraventricular hematoma. The right anterior choroidal artery was markedly hypertrophic. This artery was carefully dissected under an operating microscope. Unfortunately, however, the distal portion of the right anterior choroidal artery including the presumed site of the aneurysmal structure was embedded in the firm clot, and the aneurysmal formation per se could not be identified.

Case 2: A 51-year-old woman suddenly experienced severe headaches accompanied by vomiting.

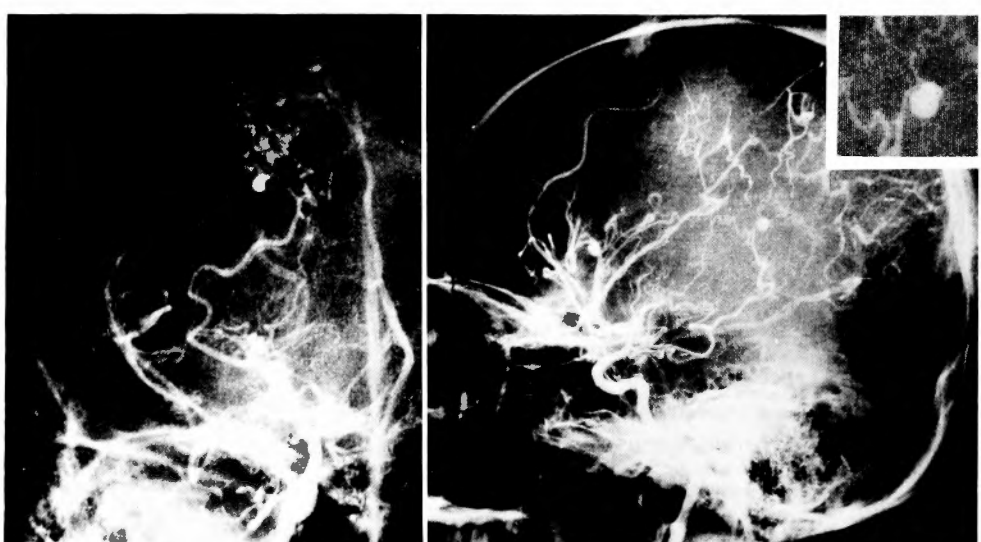
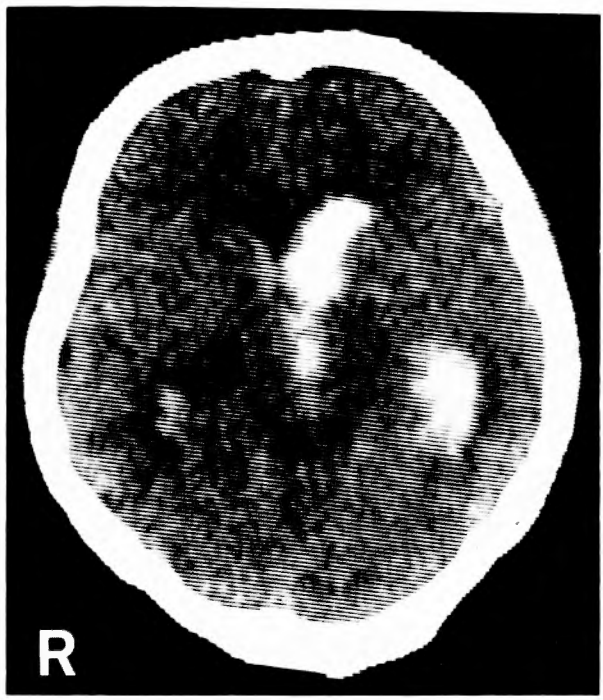


Fig. 1. Case 1. A–P projection (Left) and lateral projection (Right) of right carotid angiogram, showing an aneurysm of the distal anterior choroidal artery (Inset).



F5g. 2. Case 2. CT scan without contrast injection, showing an intraventricular hemorrhage, more marked on the left side. *R* denotes patient's right.

A few minutes later, she had generalized tonic convulsions. She was admitted to a local hospital, and the computed tomography (CT) scanning showed an intraventricular hemorrhage, mainly involving the left lateral ventricle (Figure 2). She was sent to us 18 days later.

On admission, she was disoriented to time, place and person. A right hemiparesis, right homonymous hemianopsia, and Gerstmann's syndrome were present. A left carotid angiography showed an occlusion of the proximal portion of the left middle cerebral artery with Moyamoya vessels in the base of the brain. A peripheral artery aneurysm was found in the left temporal horn (Figure 3). The detailed analysis of angiograms indicated that the parent artery of the aneurysm was the anterior branch of the lateral choroidal artery. A repeat CT scan showed a small enhancing lesion in the medial wall of the left temporal horn, which was thought to have corresponded to the aneurysm (Figure 4).

As the repeat angiography four weeks later showed a definite enlargement of the aneurysm, a left temporal craniotomy was performed and the left temporal horn was entered via a small cortical incision in the middle temporal gyrus. The aneurysm covered by a thin, organizing clot was found to have attached to the medial wall of the temporal horn near the collateral fissure. The aneurysm had no relation with the choroid plexus. It was resected in toto, together with the short proximal and distal segments of its parent artery. Histologically, it was a true saccular aneurysm (Figuer 5).

Case 3: A 50-year-old man was admitted to a local hospital with complaints of sudden, severe

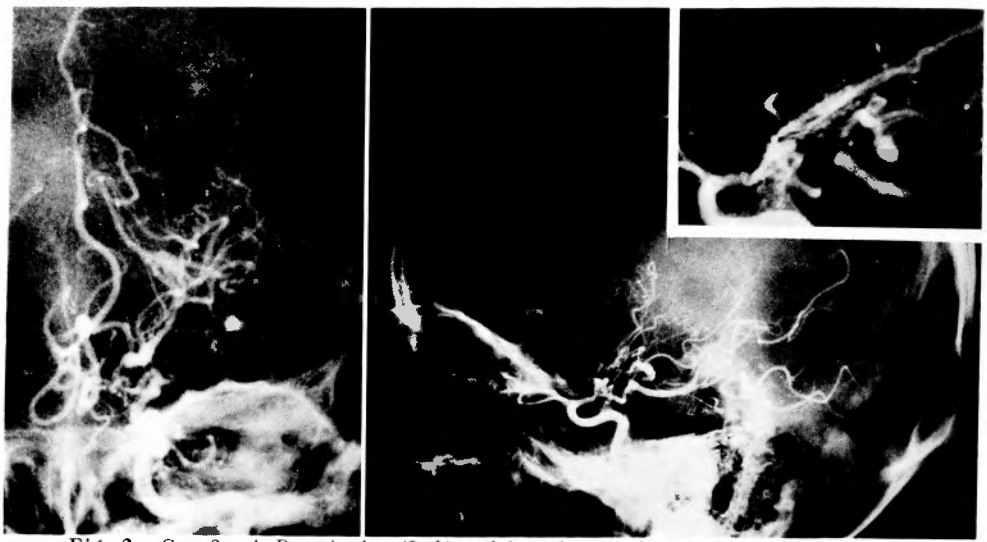


Fig. 3. Case 2. A–P projection (Left) and lateral projection (Right) of left carotid angiogram, showing an occlusion of the middle cerebral artery with Moyamoya vessels in the base of the brain. An aneurysm of the lateral choroidal artery (Inset) is seen.

headaches, nausea and vomiting. CT scan taken on the day of onset revealed an intraventricular hemorrhage, more marked in the right lateral ventricle than in the left (Figure 6).

When he was sent to our hospital 9 days later, he was mildly confused, and nuchal rigidity was moderately severe. Otherwise, no gross neurological abnormalities were found. A right carotid angiography showed a severe stenosis of the terminal portion of the right internal carotid artery, associated with Moyamoya vessels in the base of the brain. Both in the carotid and

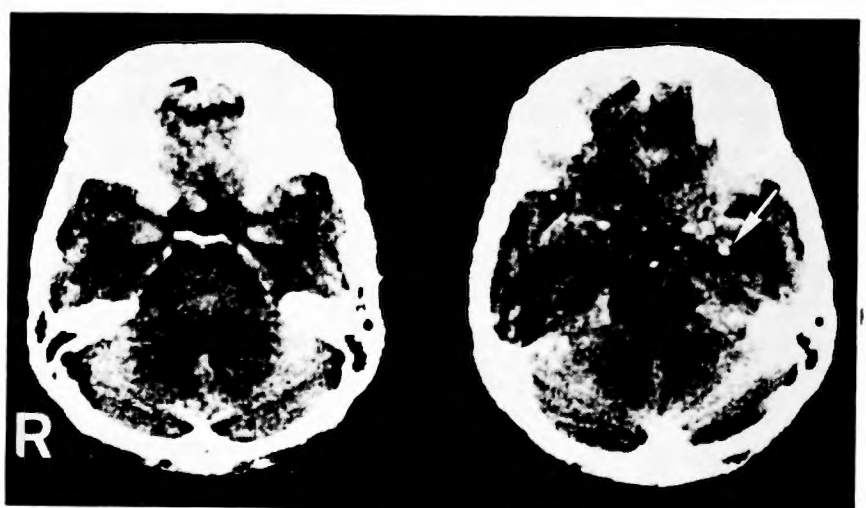


Fig. 4. Case 2. CT scan without (Left) and with (Right) contrast injection. Note a small enhancing lesion in the left temporal horn (Arrow), that corresponds to the aneurysm. *R* denotes patient's right.

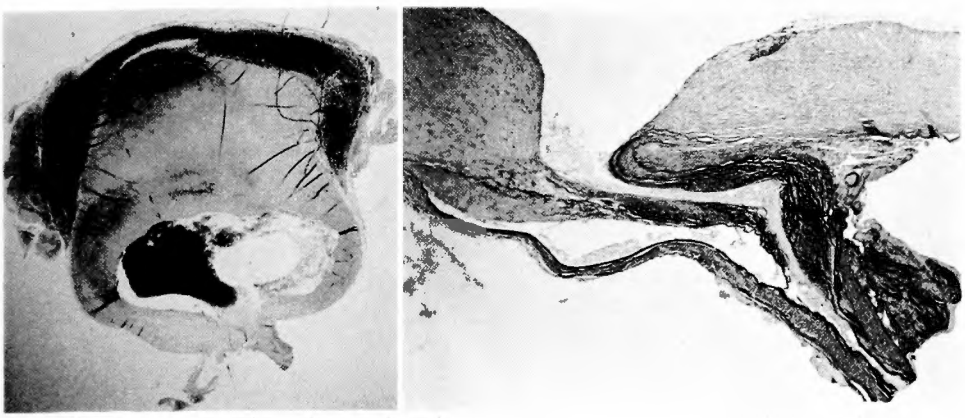


Fig. 5. Case 2. Photomicrograph of the resected aneurysm, H.E., ×14 (Right), and van Gieson, ×128 (Left).

vertebral angiograms, the right lateral posterior choroidal artery was hypertrophic, and an aneurysmal shadow was found in its peripheral portion (Figure 7). A left carotid angiogram also showed an occlusion of the terminal portion of the left internal carotid artery with extensive collateral fine vascular networks in the base of the brain.

Severe headaches recurred on day 24, and he became stuporous and a left hemiparesis appeared. Repeat angiogram and CT scanning showed no remarkable changes. A few days later, he was again fully alert, but a mild motor weakness of right upper extremity was noted in

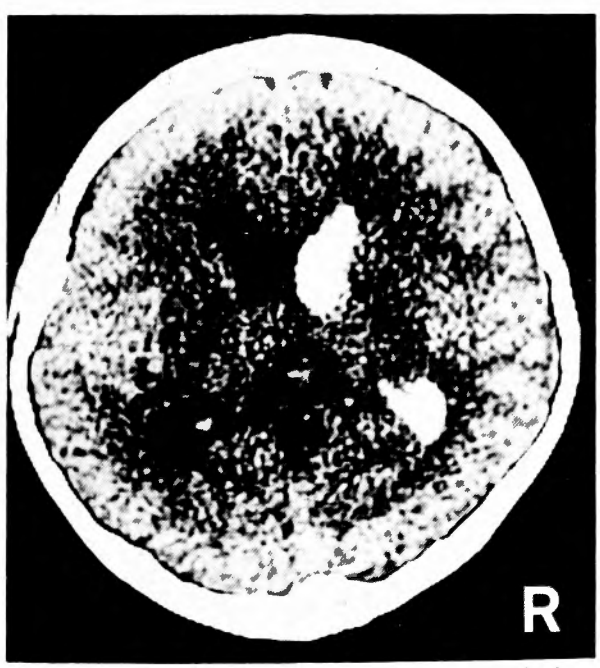


Fig. 6. CT scan without contrast injection, showing an intraventricular hemorrhage, more marked on the right side. *R* denotes patient's right.

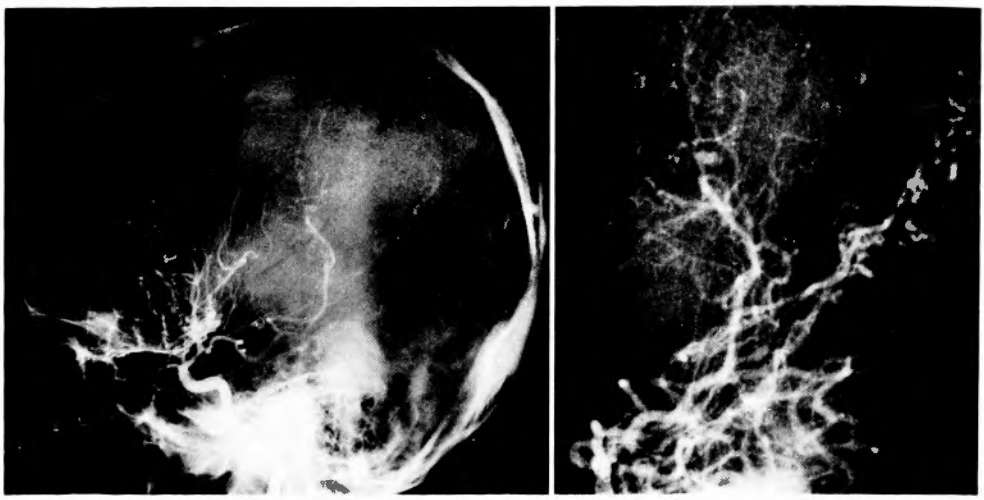


Fig. 7. Right carotid (Right) and vertebral (Left) angiogram, lateral projection, showing a stenosis of the terminal internal carotid artery, Moyamoya vessels in the base of the brain, and an aneurysmal structure (Arrow) in the distal portion of the posterior choroidal artery.

addition to persisting left hemiparesis.

Four weeks after the onset, a right parietotemporal craniotomy was carried out, and the right trigone was entered. Ventricular cavity was filled with the aged amorphous hematoma, which was found to have extended from a small paraventricular intracerebral hematoma above the trigone. The choroid plexus of the left ventricle and the lateral posterior choroidal artery

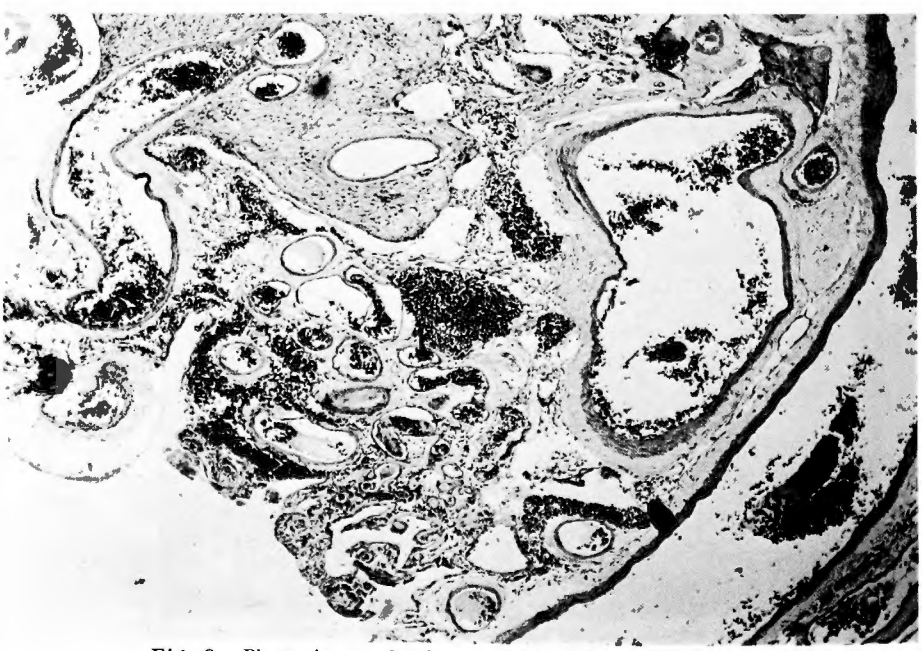


Fig. 8. Photomicrograph of the excised specimen, H. E., ×128.

were identified. No definite aneurysm was recognized, but a conglomerate of small vessels was found in the intracerebral clot. This seemed to correspond topographically to the aneurysmal shadow seen in the previous angiogram. This was resected and the intracerebral and intraventricular clots were evacuated. Histologically, the excised specimen consisted of small tortuous vessels, most likely representing a tangle of small collateral blood vessels (Figure 8).

Discussion

Moyamoya disease in the adult often presents with symptoms of an intracranial hemorrhage. Initially, a rupture of the Moyamoya vessel was presumed to be responsible for such a hemorrhage. Recently, however, a significance of an associated aneurysm as a possible source of hemorrhage has become increasingly known.

Aneurysms seen in patients with Moyamoya disease are classified into two groups; (1) aneurysms of Moyamoya vessels proper or those arising from other small collateral arteries (peripheral type), and (2) aneurysms of larger arteries of the circle of Willis (major artery type)[3]. In the previous papers, most aneurysms of peripheral type have been considered to represent pseudoaneurysms. However, reports of such aneurysms studied histologically are quite few.

In 1978, KODAMA and SUZUKI reported five patients of Moyamoya disease associated with cerebral aneurysms. Two patients had aneurysm at the bifurcation of the basilar artery[9]. In the remaining three patients, an aneurysm of the distal posterior choroidal artery was found at angiography. In each of the latter three cases with aneurysms of peripheral type, the aneurysm was found to have disappeared at repeat angiography obtained 2 to 11 months later. They assumed, therefore, that these peripheral aneurysms are not true aneurysms but pseudoaneurysms. Each patient recovered without an operation, so that the pathological confirmation of the nature of the aneurysm is lacking.

In 1982, YUASA et al. reported a case of an aneurysm of peripheral type associated with Moyamoya disease[19]. The aneurysm was histologically studied and it turned out to be a pseudoaneurysm. In the same year, FURUSE et al. also reported a similar case[3]. The aneurysm of the distal anterior choroidal artery was found in a patient with Moyamoya disease, who developed a massive intraventricular hemorrhage. The aneurysm was resected. Histologically, it was a pseudoaneurysm.

Reviewing the literature, FURUSE et al. collected 35 cases of cerebral aneurysm associated with Moyamoya disease, with or without a previous history of intracranial hemorrhage[3]. Forty-four aneurysms were found in 36 patients, including one of their own. In eight of 19 patients with an aneurysm of major artery type, the aneurysm was confirmed either at operation or at autopsy. All these eight aneurysms were saccular in shape, and they seemed to be true aneurysms. Eighteen aneurysms of peripheral type were found in 17 patients. However, only two of them were removed at operation and two others were studied at autopsy. One was a true saccular aneurysm and the second was a pseudoaneurysm (case of FURUSE et al.[3]). In the remaining two, pathological nature of the aneurysm remained unknown. In 9 other patients including 3 cases reported by KODAMA and SUZUKI[9], aneurysms of peripheral type seen at

angiography were shown to have disappeared at repeat angiography. Such a disappearance of aneurysm at repeat angiography seems to have been accepted by several authors as a strong evidence supporting a diagnosis of pseudoaneurysm, however, such an assumption may not always be true as an untreated, congenital saccular aneurysm also may disappear at repeat angiography[11,17].

In the present series, the aneurysm in the first case was clearly seen as a sharply delineated round lesion in multiple projections including a magnified view. Unfortunately, however, the aneurysm failed to be identified at autopsy, so that its pathological nature could not be confirmed.

In the second case, the aneurysm was found to be a true saccular aneurysm, although the site of the lesion in the wall of the ventricle was unusual. The parent artery of this aneurysm had definitely served as an important collateral route. It had hypertrophied and it seemed to have been subjected to an increased hemodynamic stresses. This case therefore seems to emphasize a role of hemodynamic factor in the development of a saccular aneurysm.

A role of hemodynamic stress in the formation of congenital saccular aneurysms in general has been stressed by various authors[1,2,4,5,6,7,10,12,14,16,18,20]. In patients with saccular aneurysms associated with an occlusion of the major cerebral artery in particular, aneurysms are found often in the sites at which the hemodynamic forces are presumed to be markedly increased as a result of an arterial occlusion. A distribution of such aneurysms therefore differs significantly from that of aneurysms without associated arterial occlusion. An incidence of aneurysms in the peripheral artery, or in the posterior circulation, is unusually high in patients with associated carotid occlusion. This is also the case in patients with Moyamoya disease, where bilateral carotid forks are primarily involved by stenotic processes that are in most cases steadily progressive. The vessels developed as collateral routes are thus prone to sustain progressively increasing hemodynamic forces, and an aneurysm may develop in these collateral vessels. In patients with Moyamoya disease, approximately 40% of associated aneurysms are of peripheral type and seen in the vessels serving as a collateral route, and the aneurysms of major artery type are commonly seen in the posterior circulation (46%)[3].

In the third case in the present series, an aneurysmal outpouching was seen at angiography in the lateral posterior choroidal artery that had markedly hypertrophied. Circumstantial evidence seemd to have substantiated that this vascular lesion might be a source of hemorrhage. Pathologically, however, it was neither a true aneurysm nor a pseudoaneurysm; it was rather tangled, dilated collateral channels. It seems possible that a part of previous cases of distal artery "aneurysm" seen at angiography might have been nothing but such a lesion.

In conclusion, an aneurysm of peripheral type seen angiographically in patient with Moyamoya disease may represent a pseudoaneurysm as a result of a rupture of the fragile collateral vessel. However, a true saccular aneurysm does also develop in the distal artery serving as a collateral route, and an increased hemodynamic stress secondary to the associated arterial occlusion seems to play an important role in the formation, development and rupture of such an aneurysm. Tangled collateral vessels may also present an angiographic image that is hardly differentiated from an aneurysm, either a true aneurysm or a pseudoaneurysm.

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和文抄録

モヤモヤ病の脳内出血—3例における出血源について

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挾間　章忠

モヤモヤ病に脳内出血を合併した例で，脳血管撮影上脳動脈瘤が発見されることがあり，出血源として注目されている．この脳動脈瘤には (1) ウイルス動脈輪にみとめられるものと，(2) 末梢部脳動脈にみとめられるもの，とがあり，後者はふつう仮性動脈瘤と考えられている．

われわれは脳内出血を合併したモヤモヤ病の3例で，脳血管撮影上脳動脈瘤様陰影を末梢部脳動脈に発見し，これらはいずれも，CT，手術，あいは剖検所見から出血源と考えられた．1例では剖検でこの動脈瘤様構造を発見できず，1例は真性動脈瘤が確認され，残り1例ではこの陰影は副血行路を形成する血管の集簇であった．