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A Case of Moyamoya Disease Associated with Basilar Aneurysm*)

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ABSTRACT

A case of Moyamoya disease associated with basilar aneurysm was reported. This case was rare in that it developed from subarachnoid hemorrhage attributable to rupture of basilar aneurysm. The pathological findings of the intracranial blood vessels were almost identical to the findings presented in previous reports. In the vertebral basilar artery system, a remarkable intimal thickening of the intimal cushion type was observed on the side opposite to the origin of the aneurysm. This finding in light of its position was possibly implicated in the formation of cerebral aneurysm in view of the irregularity in blood flow at that site. In the study of the blood vessels of the entire body, slight localized thickening of the intima was observed in the coronary artery, aorta, common carotid artery, renal artery, and common iliac artery, suggesting a possible relation with systemic disease.

INTRODUCTION

It is known that the common symptom development pattern of adult forms of Moyamoya disease is intracranial hemorrhage. Of these, intracerebral hemorrhage is predominant. It is reported that subarachnoid hemorrhage in Moyamoya disease is attributable largely to intracerebral hematoma which penetrates through the cerebral surface and/or ventricle to flow into the subarachnoid space[9,18].

We recently experienced a case of Moyamoya disease which developed from subarachnoid hemorrhage due to rupture of basilar aneursm. This case will be presented with some review of the literature.

CASE

Patient: 35 year old male.

Past history:

There is no past history of chronic inflammation above the cervical region. There is further nothing remarkable to add.

Present illness:

On December 1, 1981 while driving a truck, he became unconscious. Being discovered in this state, he was admitted to a hospital for emergency care. Immediate conduct of CT and bilateral CAG led to a diagnosis of subarachnoid hemorrhage attributable to Moyamoya disease. On December 3, he was transferred and admitmitted to the Department of Neurosurgery of Hiroshima Prefectural Hiroshima Hospital for detailed examination and treatment.

Neurological findings:

Akinetic mutism with paralysis of the left side was noted. Even with application of such stimulation as severe pain he could only move his right upper extremity slightly.

CT findings on day of onset (Fig. 1):

On plain CT, subarachnoid hemorrhage was observed in the basal cistern, bilateral Sylvian fissures, interhemispheric fissure, and superior cerebellar fissure. Intracerebral hematoma and hemorrhage into the ventricle were not noted.

Carotid angiography (Fig. 2):

*) 野村雅之，北岡　保，吉原高志，福原敏行，門前徹夫：脳底動脈瘤を伴うモヤモヤ病の一例

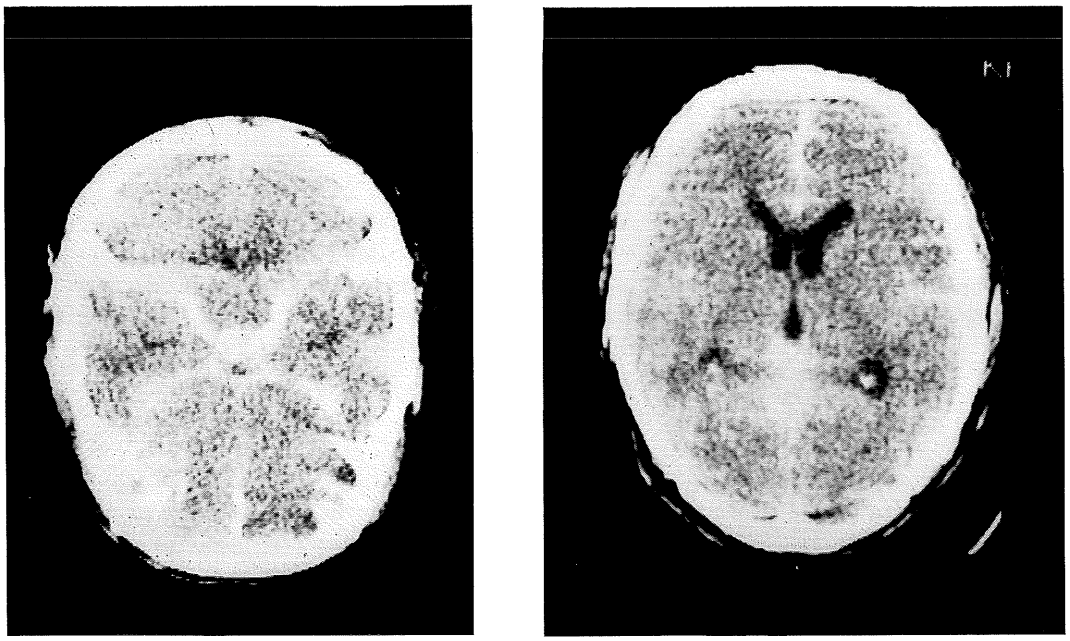


Fig. 1. Computerized tomography scans showed subarachnoid hemorrhage inthe cisterns on day of onset.

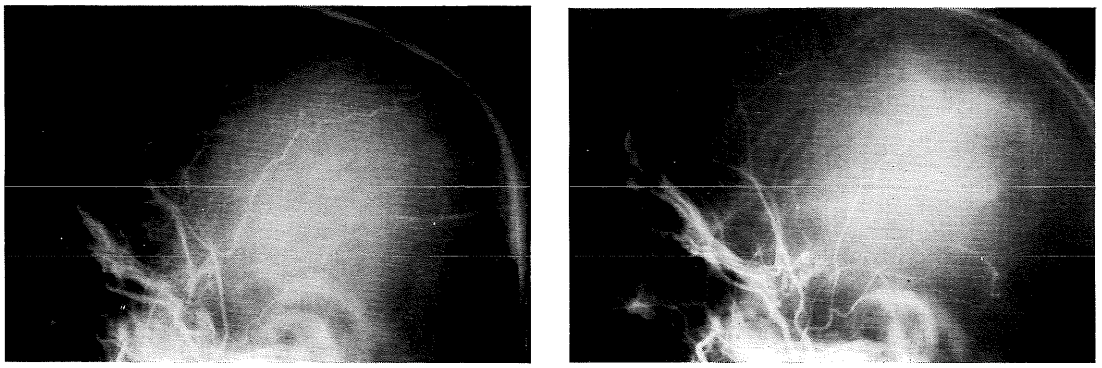


Fig. 2. Left; Left carotid angiography. Right: Right carotid angiography. Carotid angiography revealed Moyamoya vessels in the basal ganglia and occlusion of the middle cerebral artery.

The bilateral internal carotid arteries were generally narrow with thread-like stenosis immediately after branching of the enlarged-ophthalmic artery. The bilateral middle cerebral arteries were occluded at their origin and could not be visualized at all. The basilar Moyamoya vessels could be seen, but the mumber on the right side was smaller than on the left side. The bilateral anterior cerebral arteries received blood flow from the narrowed internal carotid arteries and via the transdural cortical anastomosis from the ophthalmic artery to the anterior falx artery. However, their visualization was poor and the development of the right anterior cerebral artery was found to be better than that of the left side. On the left side, the region of the posterior cerebral artery could be partially visualized via the medial meningeal artery. Vertebral angiography was not performed.

On December 5, the 5th day of hospital admission, the patient suddenly developed apnea. As rebleeding was suspected, the second CT scan was immediately made.

Second CT findings (Fig. 3):

On plain CT, subarachnoid hemorrhage was

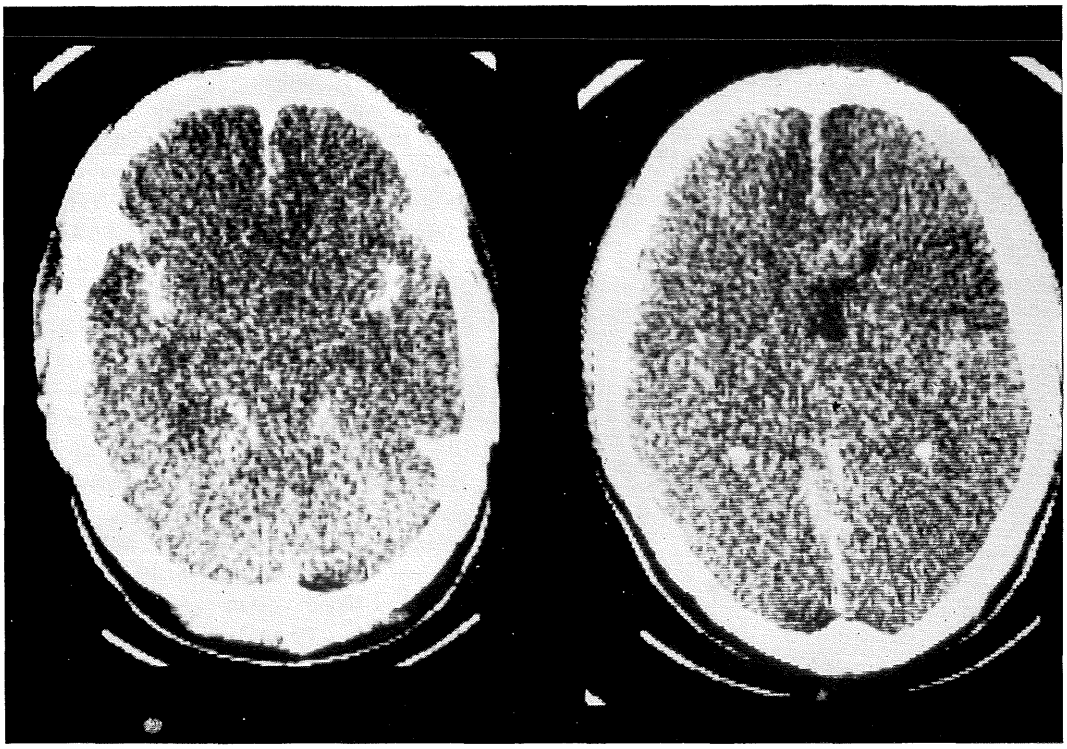


Fig. 3. Computerized tomography scans demonstrated low density areas in the bilateral frontal lobes in the 5th day after onset (arrows).

observed in the bilateral Sylvian fissures, basal cistern and interhemispheric fissure. The findings were hardly different from those observed at time of onset. However, a low density area suggestive of infarct was present on the falx side of the bilateral frontal lobes and from the right frontal lobe to the parietal lobe. There was no dilatation of both lateral ventricles, 3rd and 4th ventricles.

The patient did not recover thereafter and he expired on the same day.

AUTOPSY FINDINGS

1. Gross findings of the circle of Willis (Fig. 4)

The bilateral internal carotid arteries and the right middle cerebral artery were remarkably narrow. The left posterior communicating artery was large, but the right side was of trace size. The basilar artery and bilateral posterior cerebral arteries were well developed. The bilateral anterior choroidal arteries were enlarged and well developed until they branched. From the internal carotid arteries and the circle of Willis, enlarged and tortuous Moyamoya vessels were seen. At the branching point of the left superior cerebellar artery, cerebral aneurysm with a hematoma massfirmly attached thereto was seen. It was of the succular type and measured 5 mm × 5 mm × 7 mm in size.

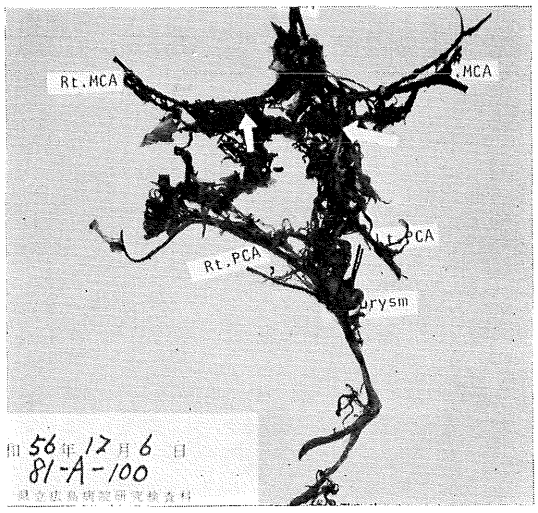


Fig. 4. Photograph of the circle of the Willis. The bilateral internal carotid arteries were remarkably narrow (arrows).

2. Histological findings of the major intracra-

nial vessels

In the left terminal region of the internal carotid arteries (Fig. 5), there was a partial

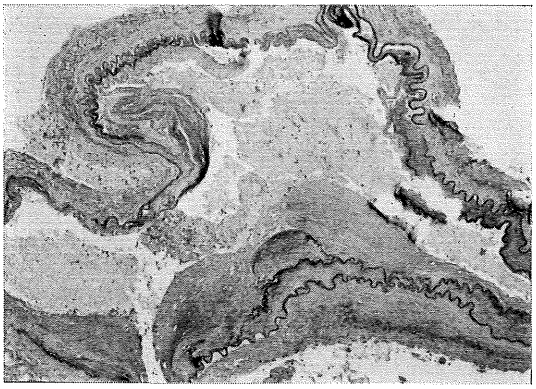


Fig. 5. Photomicrograph of the terminal portion of the left internal carotid artery showing intimal thickening, disrupted elastic fivers and rarefacted muscle layer. (Orcein stain)

protrusive thickening of the intima with stratification, diastasis, and abnormal curvature of the elastic fibers. Almost extensive rarefaction of the muscle layer was seen. The major changes at the right terminal site were remarkable rarefaction of the muscle layer and abnormal curvature, diastasis, and fragmentation of the elastic fibers. The hypertrophic changes of the intima were extremely mild.

Anterior and middle cerebral arteries: The changes in these arteries were generally mild. Local thickening of the intima was observed. In addition, such changes as slight rarefaction of the muscle layer and diastasis and fragmentation of the elastic fibers were observed.

Basilar artery and posterior cerebral arteries: Slight intimal thickening was observed at some sites, but such changes were extremely rare and mild in degree. Changes of the elastic fibers, such as diastasis and stratification, could be relatively well observed. However, the media of these blood vessels were all well maintained without any evidence of rarefaction.

Moyamoya vessels (Fig. 6): Most of the Moyamoya vessels showed hardly any change, but enlarged blood vessels with thin walls were scatteringly seen. Some showed partial intimal thickening together with stratification and diastasis of the elastic fibers.

Cerebral aneursm: Most of the sites with aneurysm-like protrusion had organized thrombus and at sites with partial rarefaction dieresis accompanied with inflammatory cell reaction

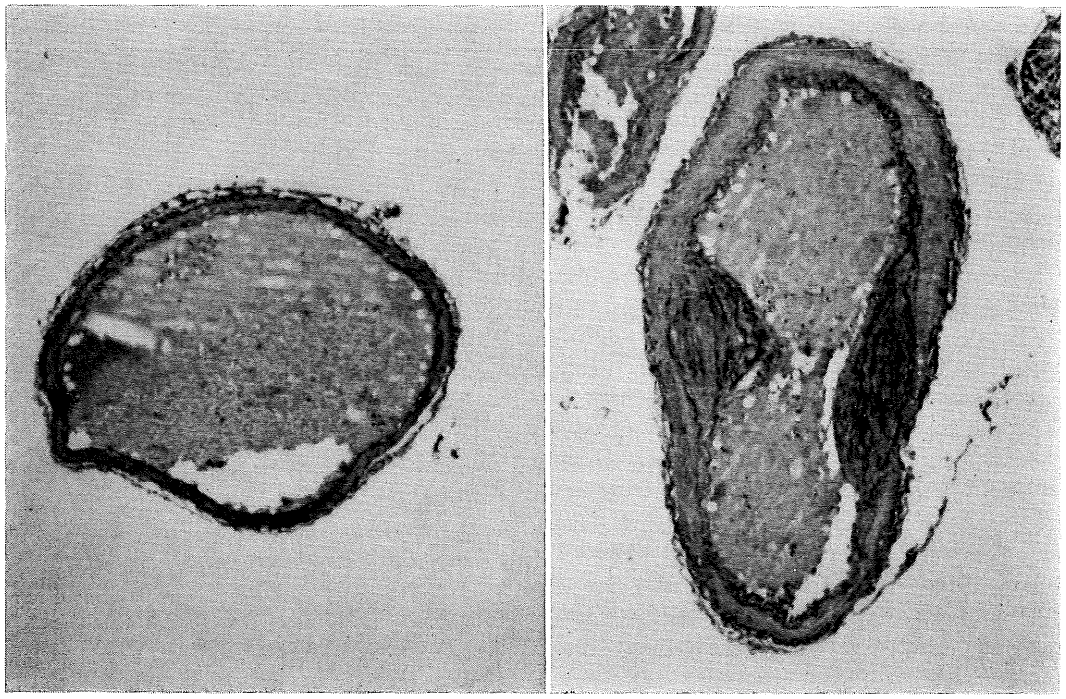
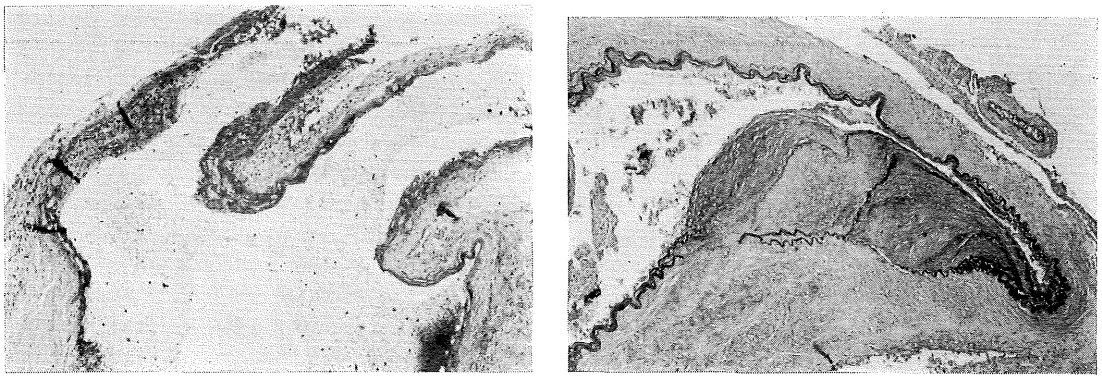


Fig. 6. Photomicrographs of the Moyamoya vessels showing dilatation in diameter with thinning of the wall. (Orcein stain)



(a) (b)

Fig. 7-a. Photomicrograph of the Basilar aneurysm with rupture. (Orcein stain)
Fig. 7-b. Photomicrograph of the basilar artery. A remarkable intimal thickening was obserbed. (Orcein stain)

was seen (Fig. 7-a). A remarkable intimal thickening accompanied by stratification of the elastic fibers could be observed in part in the vascular wall on the side opposite to the origin of the cerebral aneurysm (Fig. 7-b).

3. Histological findings of the blood vessels of the entire body excluding the cranial vessels (Fig. 8)

Partial intimal thickening and route abnormality of the elastic fibers could be seen in the coronary artery, aorta, common carotid artery, renal artery, and common iliac artery. Very slight atherosclerotic changes were observed in the aorta, but such changes were not found at other sites.

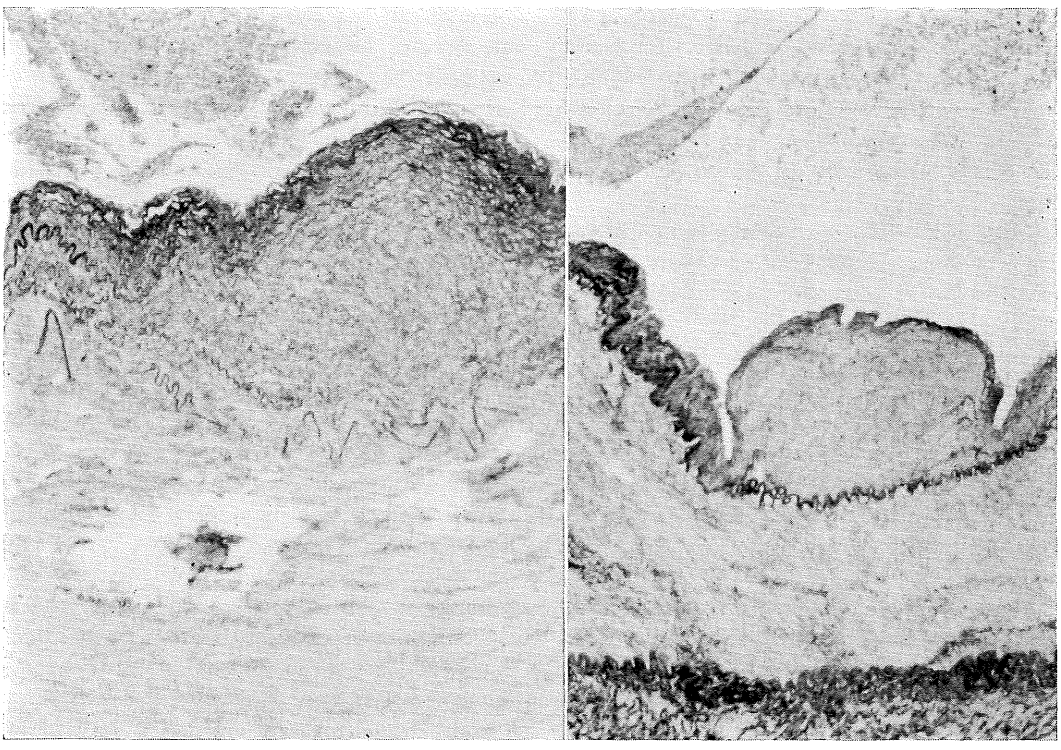


Fig. 8. Left; Common iliac artery. Right; renal artery. Partial intimal thickening and abnormal shape of the elastic fibers were obserbed. (Orcein stain)

DISCUSSION

Thirty three cases of Moyamoya disease associated with cerebral aneurysm have been reported in the literature. Aneurysms can be classified according to site of development into aneurysms within the Moyamoya vessels and aneurysms without the Moyamoya vessels. Nineteen cases[2,5,9,11,13,17,19,21,22,24,25] are aneurysms of the former category (anterior choroidal artery, posterior choroidal artery, and penetrating artery) and 14 cases are those of the latter category.

In classifying aneurysms developing without the Moyamoya vessels, there are 6 cases of basilar artery bifurcations[1,2,7,10,14], 2 cases of posterior communicating artery[1,4], case of anterior communicating artery[26], 1 case of the peripheral portion of the posterior cerebral artery[22], one case of distal anterior cerebral artery[26], and 4 cases of multiple aneurysms[1,3,10,15]. The 3 cases of multiple aneurysms[1,10,15] include basilar aneurysm and in comparison with the general development site of cerebral aneurysm, cerebral aneurysms in Moyamoya disease are often observed in the posterior half of the circle of Willis. As for the reasons why it is common in the posterior half of the circle of Willis, it is reported that due to blood flow disturbance in the anterior half of the circle of Willis in Moyamoya disease, there is an increase in the hemodynamic load on the vertebrobasilar system[10].

The age of the 15 cases of complication of aneurysms without the Moyamoya vessels ranged from 18 to 45. These belong to the adult type of Moyamoya disease.

There are four reported cases of rupture of the cerebral aneurysm[1,10,22]. Cases of rupture were confirmed in three cases by surgery[1,10,22] and on autopsy in one case[1]. All of these cases were aneurysm of the posterior half of the circle of Willis. There are three cases of cerebral aneurysm which were followed over time[1,10] and in both of these cases it was located in the posterior half of the circle of Willis. In two cases, it enlarged in size[1,10] and in one case the size has not increased even after one year following bypass procedure and clipping was performed.

Direct operation was conducted on associated aneurysm; that is on 3 cases of ruptured cerebral aneurysm and on 4 cases of non-ruptured cerebral aneurysm[1,4,26]. In direct surgery attention should be directed to the following two points. Because of the presence of anastomosis between cranial blood vessels and the meningeal artery of the external carotid artery system and superficial temporal artery of the scalp, attention should be made not to cut these anastomoses during skin incision and craniotomy. Next, there are instances where it is impossible to reach the aneurysm being blocked by the basilar Moyamoya vessels[15].

The major pathohistological findings of the cerebral trunk arteries in Moyamoya disease include deviated laminated thickening of the intima, flexion, tortuosity, and stratification of the elastic plate, and atrophy of the tunica media[16]. These changes are slight in the vertebral basilar artery system. Nagamine et al.[14] have reported such findings as arterial dilatation and thickening of the muscle layer of the tunica media due to exposure to localized hypertension. Furthermore, according to Tanaka et al.[23] thickening of the tunica media characteristic of Moyamoya disease (called intimal cushion) is minimal in degree and such findings are also observed in the base of the posterior cerebral artery and basilar artery. In our case, the vascular findings of the anterior half of the circle of Willis are in agreement with those given in earlier literature. In the vertebral basilar artery, remarkable intimal thickening was observed in the vascular wall on the side opposite the origin of the cerebral aneurysm, but such findings could not be observed in other sites. This finding is considered to be intimal cushion type change described by Tanaka et al. but as it is located on the side opposite the origin of the aneurysm, the disturbance in blood flow which developed at this site may have influenced the formation of aneurysm.

The pathohistological findings of the Moyamoya vessels show a mixture of dilative changes and stenotic and occlusive changes. As stenotic findings, Tanaka et al.[23] have reported such findings as elastofibrotic thickening of the intima and tunica media, edematous intimal thickening, and organized thrombi. Matsuo et al.[12] have observed near the origin of Moyamoya vessels intimal changes associated with intimal thickening of the circle of Willis and a picture of occlusion and desquamation among these

perforating branches and have suggested that an occlusive mechanism similar to that of the circle of Willis might also be involved in Moyamoya vessels. The major finding of our case is dilatation. Slight intimal thickening, stratification of the elastic fibers, and diastasis were observed in part, but these findings may be findings of thickening in response to increased blood flow.

In the study of the blood vessels of the entire body, it has been reported that in many of the cases intimal changes similar to intracranial changes were observed in the renal artery, common carotid artery, aorta, coronary artery, and pulmonary artery[14,23]. Similar findings were also observed in our case, which support the assumption that this disease is a cerebral type of disease bringing rise to arterial lesions in the entire body.

This cause of death of this case is considered to be attributable to rapid progress of cerebral ischemia due to cerebral vascular spasm inasmuch as there was massive subarachnoid hemorrhage in the interhemispheric fissure and basal cistern and in the CT conducted on the 5th day after onset there was an appearance of low density area almost consistent with the area of anterior cerebral artery confirmed by cerebral angiography.

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