Study of the posterior circulation in moyamoya disease

Clinical and neuroradiological evaluation

SUSUMU MIYAMOTO, M.D., HARUHIKO KIKUCHI, M.D., JUN KARASAWA, M.D.,
IZUMI NAGATA, M.D., TOSHO IKOTA, M.D., and SHIGEKAZU TAKEUCHI, M.D.

Department of Neurosurgery, National Cardio-Vascular Center, Osaka, Japan

Eighty-two cases of cerebrovascular moyamoya disease were studied by cerebral angiography and computerized tomography. Occlusive lesions were demonstrated not only in the anterior circulation but also in the posterior circulation, and they were associated with the development of an abnormal vascular network (moyamoya vessels). Although occlusive lesions do occur in the vertebrobasilar system, the vertebrobasilar system also acts as a source of collateral channels to the anterior circulation in this disease.

KEY WORDS • moyamoya disease • posterior circulation • angiography • computerized tomography • collateral circulation • vertebrobasilar system

More than 20 years have passed since “moyamoya disease” was first described, and in that time several surgical treatments have been reported. Although the etiology of this disease is still debated, its angiographic features have been elucidated in detail. The chief features of this disease are as follows: 1) progressive occlusion of cerebral arteries (especially around the carotid fork); 2) the development of an abnormal vascular network; and 3) the presence of extensive transdural and leptomeningeal collateral vessels. However, previous reports have concentrated mainly upon lesions in the anterior circulation, and a systematic study of the posterior circulation in moyamoya disease has never been made. In this paper, we describe clinical and neuroradiological findings in the posterior circulation in moyamoya disease.

Clinical Material and Methods

Between 1973 and 1983, we treated 90 patients with moyamoya disease. All these patients were under 15 years old. Of these children, 82 had received no surgical treatment before admission to our department. These 82 cases were selected as the clinical subjects for this study.

All 90 patients underwent angiography with transfemoral catheterization and magnification. Clinical assessment included activities of daily life (ADL) and evaluation of visual field defects. In 56 cases, the correlation between the angiographic findings and the computerized tomographic (CT) findings was noted. The low-density area in CT was classified into the following regions: 1) the territory of the posterior cerebral artery (PCA); 2) the territory of the middle cerebral artery (MCA); 3) the territory of the anterior cerebral artery (ACA); 4) the border area between the PCA and MCA territories; 5) the border area between the MCA and ACA territories; and 6) other regions.

Results

Moyamoya disease usually shows progressive occlusive lesions in the cerebral arteries along with the development of an abnormal vascular network providing collateral flow. These characteristics can be found also in the posterior circulation. The abnormal vascular network develops as a result of occlusive lesions in the PCA’s, basilar artery, or vertebral arteries. These networks are composed mainly of posterior choroidal arteries, of thalamogeniculate arteries, and of other thalamoperforating arteries that irrigate the thalamus and the posterior portion of the basal ganglia. When this network is well developed, it develops anastomoses with medullary vessels in the parietal subcortex (Fig. 1).

Among the 82 cases, 10 patients had a normal posterior circulation with no occlusive lesion. In 49 cases, occlusive lesions were demonstrated in the quadrigeminal segment of the PCA’s, and they developed the abnormal vascular network mentioned above. In 13 of
Posterior circulation in moyamoya disease

FIG. 1. Left vertebral angiograms, early arterial phase (left) and late arterial phase (right). The posterior cerebral arteries are occluded at their proximal portion. The abnormal vascular network is well developed, showing anastomoses with the medullary vessels in the parietal subcortex.

FIG. 2. Left vertebral angiograms, frontal (left) and lateral (right) views. Few cortical branches of the posterior cerebral artery are visualized. Both posterior cerebral arteries are occluded at their quadrigeminal segments.

FIG. 3. Right vertebral angiograms, frontal (left) and lateral (right) views. An occlusive lesion extends to the proximal portion of the posterior cerebral arteries. The abnormal vascular network is well developed.

FIG. 4. Left vertebral angiograms, frontal (left) and lateral (right) views, showing occlusive lesions affecting the trunk of the basilar artery. Since the abnormal vascular network is composed mainly of perforating arteries arising from the top of the basilar artery and from the posterior cerebral arteries, this network is small.

FIG. 5. Right vertebral angiograms, frontal (left) and lateral (right) views. The right vertebral artery is occluded at the craniocervical junction. Transdural anastomosis between the posterior meningeal branch of the vertebral artery and the superior cerebellar artery is demonstrated.

In these 49 cases, few cortical branches of the PCA were visualized (Fig. 2). In 23 cases, the occlusive lesion extended to the proximal segment of the PCA's or to the basilar artery (Fig. 3). The abnormal vascular network enlarged as the occlusive lesion progressed; however, when the occlusive lesion reached the trunk of the basilar artery, the network regressed in size, suggesting severe disturbance of the posterior circulation (Fig. 4). Vertebral artery occlusion was demonstrated on four sides in three cases (Fig. 5).

The vertebral angiograms in the 82 cases were classified into the following three groups: 1) Normal or slightly involved group (Fig. 6 left pair). In this type, most of the cortical branches of the PCA fill normally. A small stenotic lesion may exist in the quadrigeminal segment of the PCA. 2) Moderately involved group...
S. Miyamoto, et al.

FIG. 6. Classification of patterns seen on vertebral angiography, frontal views (upper) and lateral views (lower). The normal or slightly involved group is illustrated (left), the moderately involved group (center), and the severely involved group (right).

In this pattern, few cortical branches of the PCA are visualized. The enlarged abnormal vascular network is demonstrated to irrigate the thalamus and the basal ganglia. 3) Severely involved group (Fig. 6 right pair). The occlusive lesion progresses into the trunk of the basilar artery, or into the vertebral arteries. The abnormal vascular network is small.

Internal carotid angiograms were similarly classified into three groups: 1) Normal or slightly involved group (Fig. 7 left pair). In this pattern the cortical branches are mostly filled normally. A small stenotic lesion may exist around the carotid fork. 2) Moderately involved group (Fig. 7 center pair). Few cortical branches of MCA’s are visualized in this type. The enlarged abnormal vascular network (“basal moyamoya”) develops around the carotid fork. 3) Severely involved group (Fig. 7 right pair). The internal carotid arteries are completely occluded. The basal moyamoya network is small.

When these two classifications are applied in the 82 cases, the interrelationship between the anterior circulation and the posterior circulation can be divided into five angiographic stages (Fig. 8). These preoperative angiographic stages showed some correlation with the preoperative clinical assessment of ADL (Fig. 9).

The CT findings related to the angiographic stages are summarized in Fig. 10. No thalamic infarct was demonstrated. In the PCA territory, the incidence of infarct increased in the later angiographic stages. In the border zone between the PCA and MCA territories, infarct was much more frequently found at Stage III, IV, or V than at Stage I or II. This tendency was also detected in the MCA territory. In the ACA territory, or in the border zone between the ACA and MCA territories, no correlation could be found. Homonymous hemianopsia, attributed to the infarct in the visual cortex, was detected in nine cases (11% of the total series) (Fig. 11).

Discussion

Several preceding studies have clarified the pathological nature of moyamoya disease. Progressive oc-
Posterior circulation in moyamoya disease

Fig. 7. Classification of patterns seen on internal carotid angiography, frontal views (upper) and lateral views (lower). The normal or slightly involved group is illustrated (left), the moderately involved group (center), and the severely involved group (right).

Fig. 8. The interrelationship between the anterior circulation and the posterior circulation in moyamoya disease, showing five stages of interaction.

Fig. 9. The interrelationship between the angiographic stages and the preoperative activity of daily living: G = good, F = fair, P = poor.
in the visual cortex. FIG. 11. Computerized tomography scans showing an infarct in the visual cortex.

culation. In this report, we stress that these pathological features can also be found in the posterior circulation. The occlusive lesion in the posterior circulation seemed to start initially from the quadrigeminal segment of the posterior cerebral arteries in most cases, and showed proximal progression. The interrelationship between the anterior circulation and the posterior circulation was classified into five angiographic stages, which showed some correlation with the preoperative ADL. It can be assumed from these facts that occlusive lesions in the posterior circulation show steady progression similar to the pattern seen in the anterior circulation.

In the ACA territory, no correlation could be demonstrated between the incidence of infarct and the angiographic stage. This may be due to collateral vessels supplying this region from the internal maxillary artery or ophthalmic artery. PCA = posterior cerebral artery; MCA = middle cerebral artery; ACA = anterior cerebral artery.

The abnormal vascular network in the posterior circulation is referred to as “posterior basal moyamoya” in the following discussion. Posterior basal moyamoya is composed mainly of posterior choroidal arteries, thalamogeniculate arteries, and other thalamoperforating arteries. No thalamic infarct could be found in our series. This is contrary to the findings in other entities, in which PCA occlusion has been reported in association with thalamic infarct. 

Homonymous hemianopsia attributed to the infarct in the visual cortex was detected in 11% of our series. Because of the young age and mental retardation of the patients, visual field defects may occur more often than recognized. Further study on this aspect is needed.

Although in some cases posterior basal moyamoya showed anastomosis with the medullary vessels in the parietal subcortex, suggesting its potential contribution as collateral channels to the MCA territory, its role in providing collateral supply is somewhat limited. In cases at angiographic Stage I or II, in which the posterior circulation remains almost intact, the incidence of in-
Posterior circulation in moyamoya disease

farct in the MCA territory was much lower than in patients showing Stage III, IV, or V. In other words, the vertebobasilar system supplies collateral channels to the anterior circulation in moyamoya disease, even though there are occlusive lesions in the posterior circulation. However, a normal or slightly affected vertebobasilar system may be the only effective source of collateral supply to the anterior circulation.

References


Manuscript received April 24, 1984.
Address reprint requests to: Susumu Miyamoto, M.D., Department of Neurosurgery, National Cardio-Vascular Center, 5-125, Fujishiro-dai, Suita, Osaka, Japan.