Renal Artery Lesions in Patients With Moyamoya Disease

Angiographic Findings

Ichiro Yamada, MD; Yoshiro Himeno, MD; Yoshiharu Matsushima, MD; Hitoshi Shibuya, MD

Background and Purpose—Renal artery lesions in moyamoya disease have been described sporadically in several case reports. The purpose of this study is to evaluate the angiographic findings of renal artery lesions in moyamoya disease and to determine the prevalence of renal artery lesions in patients with moyamoya disease.

Methods—Eighty-six consecutive patients with idiopathic moyamoya disease were prospectively examined with both cerebral angiography and abdominal aortography. The findings of abdominal aortography were reviewed for the presence and appearance of renal artery lesions and compared with the clinical data and cerebral angiographic findings.

Results—Of 86 patients with idiopathic moyamoya disease, 7 patients (8%) were found to have renal artery lesions. Six patients (7%) had stenosis in the renal artery, and 1 patient (1%) had a small saccular aneurysm in the renal artery. Two patients (2%) with a marked renal artery stenosis presented with renovascular hypertension, which resulted in an intraventricular hemorrhage in 1 patient. Furthermore, the renal artery stenosis in the 2 patients with renovascular hypertension was successfully treated with percutaneous transluminal angioplasty. There was no significant correlation between the presence of renal artery lesions and cerebral angiographic findings.

Conclusions—Seven (8%) of 86 patients with moyamoya disease showed renal artery lesions, including 6 stenoses (7%) and 1 aneurysm (1%). Renal artery lesions are a clinically relevant systemic manifestation in patients with moyamoya disease. (Stroke. 2000;31:733-737.)

Key Words: angiography • moyamoya disease • renal artery

Although moyamoya disease, a rare cerebrovascular occlusive disease of unknown etiology, is predominantly seen in Japan, cases have also been reported elsewhere.1–4 The cerebral angiographic features of this disease are as follows: (1) bilateral stenosis or occlusion of the supraclinoid portion of the internal carotid artery (ICA) that extends to the proximal portions of the anterior cerebral artery (ACA) and the middle cerebral artery (MCA) and (2) parenchymal, leptomeningeal, and transdural collateral vessels that supply the ischemic brain.5,6

Recently, renal artery lesions in moyamoya disease have been described sporadically in several case reports.7–15 However, to our knowledge, no report has yet determined the prevalence of renal artery lesions in a large-scale study of patients with moyamoya disease. In this study we prospectively performed both cerebral angiography and abdominal aortography in 86 patients with moyamoya disease to evaluate the angiographic findings of renal artery lesions and to determine the prevalence of renal artery lesions in patients with moyamoya disease.

Subjects and Methods

Patients
From April 1986 to December 1998, 86 consecutive patients, confirmed to have idiopathic moyamoya disease by means of cerebral angiography, were studied at our departments of radiology and neurosurgery. Fifty-eight patients were female, and 28 patients were male. Their ages ranged from 2 to 48 years (mean ± SD age, 16 ± 12 years). None of the 86 patients had an underlying disease, thereby confirming the diagnosis of idiopathic moyamoya disease. The study protocol was approved by the institutional review board, and informed consent was obtained from all patients.

Imaging Examinations
All 86 patients underwent cerebral angiography that included bilateral internal and external carotid arteriography and unilateral or bilateral vertebral arteriography, with the use of the transfemoral catheterization technique for both carotid and vertebral arteriography. All 86 patients also underwent abdominal aortography to examine the renal artery.

Image Analysis
On the basis of the angiographic findings, we classified steno-occlusive lesions of the ICA bifurcation into 5 different stages according to the degree of narrowing in the supraclinoid portion of the ICA and the proximal portions of the ACA and MCA: stage 1, mild to moderate stenosis of the ICA bifurcation (≤80% reduction in diameter); stage 2, severe stenosis of the ICA bifurcation (>80% reduction in diameter); stage 3, occlusion of either the ACA or MCA; stage 4, occlusion of the ICA bifurcation, with partial retention of the ACA or MCA main trunk; and stage 5, occlusion of the ICA bifurcation, with no detectable ACA or MCA main trunk.6 Stenotic or occlusive lesions in the posterior cerebral artery (PCA) were also identified. Thus, the PCA was graded as normal, stenotic, or occlusive.
or occluded. Furthermore, we also classified the basal cerebral moyamoya vessels on the basis of their presence and appearance into 4 grades: none, slight, moderate, or marked. The findings of abdominal aortography were reviewed for the presence and appearance of lesions in the abdominal aorta and its major branches, including the bilateral renal artery. Renal artery lesions included stenosis and aneurysm. Furthermore, the degree of maximal renal artery stenosis was graded with a 5-point ordinal scale: 0, no stenosis; 1, <50% stenosis (slight stenosis); 2, 50% to 75% stenosis (moderate stenosis); 3, >75% stenosis (marked stenosis); and 4, occlusion.

Finally, a statistical analysis of the comparison between groups was performed with the $\chi^2$ test, Fisher exact test, unpaired Student’s $t$ test, Mann-Whitney $U$ test, or Spearman rank correlation test. A $P$ value < 0.05 was considered to indicate a statistically significant difference.

## Results

### Renal Artery Lesions

Of 86 patients with idiopathic moyamoya disease, 7 patients (8%) were found to have renal artery lesions (Table 1). Six patients (7%) had stenosis in the renal artery, and 1 patient (1%) had a saccular aneurysm in the renal artery. The grades of renal artery stenosis were as follows: slight, 2 patients; moderate, 2 patients; and marked, 2 patients (Figure 1).

The 2 patients (2%) with a marked stenosis of the renal artery presented with renovascular hypertension (Table 1). In 1 (case 1) of the 2 patients, renovascular hypertension resulted in an intraventricular hemorrhage, which was the first symptom of this patient with moyamoya disease (Figure 1).

Furthermore, the renal artery stenosis in the 2 patients with renovascular hypertension could be successfully treated with percutaneous transluminal angioplasty (PTA) (Figure 1). In case 1 blood pressure was 210/110 mm Hg, and after the PTA procedure the blood pressure was reduced to 120/80 mm Hg. In case 2 blood pressure was 170/110 mm Hg, and after the PTA procedure the blood pressure was reduced to 110/70 mm Hg. Thus, by means of

### TABLE 1. Renal Artery Lesions in Patients With Moyamoya Disease

<table>
<thead>
<tr>
<th>Patient/ Age, y/Sex</th>
<th>Renal Artery Lesion</th>
<th>Grade of Stenosis</th>
<th>Renovascular Hypertension</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/25/F</td>
<td>Stenosis</td>
<td>Marked</td>
<td>Present</td>
</tr>
<tr>
<td>2/10/F</td>
<td>Stenosis</td>
<td>Marked</td>
<td>Present</td>
</tr>
<tr>
<td>3/22/F</td>
<td>Stenosis</td>
<td>Moderate</td>
<td>Absent</td>
</tr>
<tr>
<td>4/18/M</td>
<td>Stenosis</td>
<td>Moderate</td>
<td>Absent</td>
</tr>
<tr>
<td>5/23/F</td>
<td>Stenosis</td>
<td>Slight</td>
<td>Absent</td>
</tr>
<tr>
<td>6/5/F</td>
<td>Stenosis</td>
<td>Slight</td>
<td>Absent</td>
</tr>
<tr>
<td>7/24/F</td>
<td>Aneurysm</td>
<td>Not applicable</td>
<td>Absent</td>
</tr>
</tbody>
</table>

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**Figure 1.** Images of a 25-year-old woman with moyamoya disease manifesting renal artery stenosis who presented with renovascular hypertension and intraventricular hemorrhage (case 1). A, Left internal carotid arteriogram shows that the left ICA is occluded in the distal supraclinioid portion and that the ACA and MCA are also occluded in the proximal portions. Moderate moyamoya vessels are seen in the basal cerebral region. B, Right internal carotid arteriogram shows that the right ACA is occluded in the proximal portion. Slight moyamoya vessels are seen in the basal cerebral region. C, Unenhanced CT scan shows an intraventricular hemorrhage in the third and both lateral ventricles. D, Abdominal aortogram shows a marked stenosis in the proximal portion of the left renal artery. E, The PTA procedure using a balloon catheter has dilated the stenosis in the left renal artery. F, The PTA procedure shows the dilatation of the arterial stenosis.
the PTA procedure, we could control hypertension in the 2 patients.

With regard to renal artery aneurysm, a saccular aneurysm was found in 1 (1%) of the 86 patients with moyamoya disease. This case involved a 24-year-old woman, and a small saccular aneurysm was seen in the distal part of the renal artery main trunk (Figure 2).

The renal artery lesions (n=7) in patients with moyamoya disease were more frequent in females (n=6) than in males (n=1), although this difference was not statistically significant (P>0.05, Fisher exact test). The mean±SD age for 7 patients (18±8 years) with moyamoya disease manifesting renal artery lesions was slightly higher than that for the other 79 patients (15±12 years) with moyamoya disease who had no renal artery lesions, but this difference was not statistically significant (P>0.05, unpaired Student’s t test).

Cerebral Angiographic Findings

The cerebral angiographic findings in the 7 patients with moyamoya disease manifesting renal artery lesions are summarized in Table 2. In comparison with the 79 patients with moyamoya disease who had no renal artery lesions, there was no statistically significant correlation between the presence of renal artery lesions and cerebral angiographic findings.

Steno-Occlusive Lesions

In all 7 patients, stenosis or occlusion of the supraclinoid portion of the ICA and of the proximal portions of the ACA and MCA was detected bilaterally (Figure 1). The stages of stenotic or occlusive lesions in the 14 ICA bifurcations were as follows: stage 3, 5 arteries; stage 4, 8 arteries; and stage 5, 1 artery (Table 2). Of 14 ICAs, 5 arteries (36%) were stenotic and 8 arteries (57%) were occluded; of 14 ACAs, 2 arteries (14%) were stenotic and 12 arteries (86%) were occluded; and of 14 MCAs, 1 artery (7%) was stenotic and 11 arteries (79%) were occluded. Furthermore, of the 14 PCAs, 2 arteries were found to be stenotic and 6 arteries occluded (Table 2). Thus, a total of 8 PCAs (57%) in 6 patients manifested stenotic or occlusive lesions.

Collateral Vessels

In all 7 patients, basal cerebral moyamoya vessels were detected bilaterally in the cerebral hemispheres (Figure 1). The grades of the moyamoya vessels in the 14 cerebral hemispheres were as follows: slight, 4 hemispheres; moderate, 9 hemispheres; and marked, 1 hemisphere (Table 2). Furthermore, leptomeningeal collateral vessels from the PCA to the anterior circulation were detected in 6 hemispheres (43%) (Table 2).

<table>
<thead>
<tr>
<th>Patient/ Age, y/Sex</th>
<th>Side</th>
<th>Stage of ICA Bifurcation</th>
<th>PCA Lesion</th>
<th>Steno-occlusive Lesion</th>
<th>Collateral Vessels</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Moyamoya Vessels</td>
</tr>
<tr>
<td>1/25/F</td>
<td>Right</td>
<td>3</td>
<td>Normal</td>
<td>Slight</td>
<td>Present</td>
</tr>
<tr>
<td></td>
<td>Left</td>
<td>4</td>
<td>Normal</td>
<td>Moderate</td>
<td>Present</td>
</tr>
<tr>
<td>2/10/F</td>
<td>Right</td>
<td>4</td>
<td>Stenotic</td>
<td>Marked</td>
<td>Absent</td>
</tr>
<tr>
<td></td>
<td>Left</td>
<td>3</td>
<td>Normal</td>
<td>Slight</td>
<td>Present</td>
</tr>
<tr>
<td>3/22/F</td>
<td>Right</td>
<td>4</td>
<td>Normal</td>
<td>Slight</td>
<td>Present</td>
</tr>
<tr>
<td></td>
<td>Left</td>
<td>4</td>
<td>Occluded</td>
<td>Moderate</td>
<td>Absent</td>
</tr>
<tr>
<td>4/18/M</td>
<td>Right</td>
<td>4</td>
<td>Occluded</td>
<td>Moderate</td>
<td>Absent</td>
</tr>
<tr>
<td></td>
<td>Left</td>
<td>5</td>
<td>Occluded</td>
<td>Slight</td>
<td>Absent</td>
</tr>
<tr>
<td>5/23/F</td>
<td>Right</td>
<td>3</td>
<td>Normal</td>
<td>Moderate</td>
<td>Present</td>
</tr>
<tr>
<td></td>
<td>Left</td>
<td>4</td>
<td>Stenotic</td>
<td>Moderate</td>
<td>Absent</td>
</tr>
<tr>
<td>6/5/F</td>
<td>Right</td>
<td>3</td>
<td>Occluded</td>
<td>Moderate</td>
<td>Absent</td>
</tr>
<tr>
<td></td>
<td>Left</td>
<td>3</td>
<td>Occluded</td>
<td>Moderate</td>
<td>Absent</td>
</tr>
<tr>
<td>7/24/F</td>
<td>Right</td>
<td>4</td>
<td>Normal</td>
<td>Moderate</td>
<td>Present</td>
</tr>
<tr>
<td></td>
<td>Left</td>
<td>4</td>
<td>Occluded</td>
<td>Moderate</td>
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</tr>
</tbody>
</table>
Discussion

Our results have revealed that of 86 patients with idiopathic moyamoya disease, 7 patients (8%) were found to have renal artery lesions. Six of the 7 patients (7% of the 86 patients) had stenosis in the renal artery, and 1 patient (1% of the 86 patients) had a saccular aneurysm in the renal artery. Previous case reports7–15 have sporadically described renal artery lesions in moyamoya disease. However, the present report is the first one that demonstrates the prevalence of renal artery lesions in a large-scale study of patients with moyamoya disease.

Although the prevalence of renal artery disease in the general population is unknown, the prevalence of renal artery stenosis was 4.3% in an autopsy study.6 The autopsy study included a number of older patients with arteriosclerotic renal artery stenosis, but patients with moyamoya disease are much younger than patients in the autopsy study. Thus, the prevalence (8%) of renal artery lesions in patients with moyamoya disease appears to be considerably higher than that in the corresponding general population. We compared moyamoya patients manifesting renal artery lesions with moyamoya patients who had no renal artery lesions on the basis of the classification of the patients, cerebral angiographic findings, and renal angiographic findings. However, there was no statistically significant correlation between the presence of renal artery lesions and the sex of the patient, age of the patient, or cerebral angiographic findings.

In our series, 2 patients (2%) with a marked stenosis of the renal artery presented with renovascular hypertension for their initial symptoms in moyamoya disease. The hypertension in patients with moyamoya disease manifesting renal artery stenosis was also reported in the previous case reports.7–15 Thus, the presence of renovascular hypertension in young patients may suggest the possibility of moyamoya disease in the differential diagnosis of hypertension.

One of the 2 patients with renovascular hypertension suffered from an intraventricular hemorrhage, which was the onset of moyamoya disease in this patient. In general, children with moyamoya disease tend to present with cerebral ischemic symptoms, and adults with moyamoya disease tend to manifest symptoms due to cerebral hemorrhage.6,17,18 In our series also, the patient who had an intraventricular hemorrhage was a 25-year-old adult woman. The presence of cerebral hemorrhage in patients with moyamoya disease may indicate renal artery stenosis that causes renovascular hypertension.

By means of the PTA procedure, we were able to successfully treat the renal artery stenosis in the 2 patients with moyamoya disease who had renovascular hypertension. Thus, we were able to control the hypertension in the 2 patients with moyamoya disease. We believe that the PTA procedure is the first choice of treatment for renal artery stenosis in patients with moyamoya disease. In previous reports,6,10,11,15 renal artery stenosis in moyamoya disease has been treated with renal autotransplantation or PTA.

With regard to renal artery aneurysm, a small saccular aneurysm was found in 1 (1%) of the 86 patients with moyamoya disease. This case involved a 24-year-old woman, and the aneurysm was seen in the distal part of the renal artery main trunk. There has been only 1 case of renal artery aneurysm reported in the literature.15 However, cerebral aneurysms associated with moyamoya disease have been discussed in several reports.6–17 Yamada et al6 reported that of 76 patients with moyamoya disease, cerebral aneurysms were found in 2 patients (3%), and the prevalence of aneurysms was higher in adults with moyamoya disease than in children with moyamoya disease. Thus, aneurysms in the renal artery also may be associated with the occurrence of cerebral aneurysms in patients with moyamoya disease.

By using histopathologic examination and morphometric analysis, Ikeda19 has demonstrated that moyamoya disease involves not only the intracranial vessels but also the extracranial vessels and that there are systemic etiologic factors that cause intimal thickening in the systemic vessels. Moyamoya disease involves focal etiologic factors that act around the terminal portions of the ICA. In addition to focal etiologic factors, however, systemic etiologic factors that lead to intimal fibrous thickening in both the intracranial and extracranial vessels appear to be involved in moyamoya disease.19 Our data show that some patients with moyamoya disease have renal artery stenosis and no hypertension, but stenotic lesions in moyamoya disease are usually progressive. Thus, early detection of renal artery lesions may be valuable in the management of patients with moyamoya disease. The renal artery lesions are a clinically relevant systemic manifestation in the management of patients with moyamoya disease.

In conclusion, 7 (8%) of 86 patients with idiopathic moyamoya disease showed renal artery lesions, including 6 stenoses (7%) and 1 aneurysm (1%). Two patients (2%) with a marked renal artery stenosis presented with renovascular hypertension, which resulted in an intraventricular hemorrhage in 1 patient. Renal artery stenosis with renovascular hypertension was successfully treated with the PTA procedure. Renal artery lesions are a clinically relevant systemic manifestation in the management of patients with moyamoya disease.

References


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