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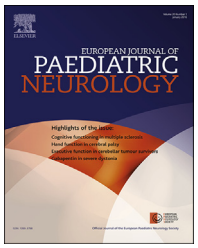

Original Article

Prevalence and clinical implications of renal artery stenosis in pediatric moyamoya disease

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ABSTRACT

Purpose: Until recently, extracranial involvement of moyamoya disease (MMD) had not been fully elucidated. The purpose of this study was to determine the prevalence and clinical implications of renal artery stenosis in pediatric MMD patients.

Materials & Methods: This retrospective study included 101 pediatric (<18 years old) MMD patients who visited our hospital from July 2008 to May 2013. All patients had legible abdominal or renal angiography. Baseline characteristics, including hypertension (HT) and angiographic findings, were retrospectively evaluated.

Result: The median age was 8 (range 2–16) years. Six patients (5.9%) had HT. Renal artery stenosis was identified in 8 patients (7.9%). Five of 8 renal artery stenosis patients had HT. Statistical analysis showed that advanced stages of MMD and HT were associated with renal artery stenosis ($p < 0.05$).

Conclusion: Renal artery stenosis was not uncommon in pediatric MMD. Renal artery evaluation could help determine the cause of HT in advanced pediatric MMD cases. Further prospective and large-scale studies may be helpful in elucidating the extracranial manifestation of MMD.

1. Introduction

Moyamoya disease (MMD) is characterized as an idiopathic, chronic and progressive steno-occlusive cerebrovascular disease involving the terminal portions of the internal carotid arteries (ICA).[1] Due to the paucity of cases, many aspects of this disease remain to be elucidated. In general, MMD is an intracranial vascular stenoocclusive disease, but several previous reports have described extracranial vascular stenoocclusive changes.[2,3] Among these changes, the renal artery is the most commonly reported site of extracranial vascular

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involvement of MMD.[3–9] Most of the previous reports were case reports, with only a few case series available regarding the extracranial involvement of MMD. According to Yamada et al., the prevalence of renal artery involvement of MMD was 8% (7 in 86 patients).[2] Similarly, Togao et al. reported a 5% (4 in 73 patients) prevalence of renal artery involvement of MMD.[10] Both studies included both pediatric and adult patients. In general, the most common cause of renal artery stenosis was atherosclerosis, while the second most common cause was fibromuscular dysplasia.[11] Consequently, it is difficult to conclude that the renal artery stenosis was due solely to the extracranial vascular involvement of adult MMD.

In our study, we only included patients under the age of 18. Furthermore, we retrospectively reviewed the abdominal aortography or selective renal and cerebral angiographies in 101 patients with MMD to determine the prevalence and clinical implications of renal artery lesions in patients with MMD.

2. Materials and methods

This study included pediatric patients with MMD who visited our hospital from July 2008 to May 2013. During the given period, 165 consecutive pediatric MMD patients visited our hospital. Among them, 57 patients without angiographic evaluation of the renal artery and 7 patients with poor quality images were excluded. Therefore, a total of 101 patients under the age of 18 who underwent cerebral angiography with legible abdominal aortography and/or selective renal angiography were included.

Baseline characteristic, including age, gender, history of hypertension (HT), were analyzed by retrospective chart review. We retrospectively reviewed the cerebral angiography and abdominal aortography and/or selective renal angiography results from these 101 patients. To evaluate the cerebral angiographic findings, we classified the stenoocclusive lesions of the intracranial internal carotid artery into the six angiographic stages defined by Suzuki and Takaku and Suzuki and Kodama.[1,12]

Among the 101 patients, abdominal aortography with or without selective renal angiography was performed in 58 patients, and selective renal angiography alone was performed in the remaining 43 patients. We evaluated the results for the presence, location and degree of stenosis in the bilateral renal arteries. The degree of maximal arterial stenosis was graded on a 5-point ordinal scale: no stenosis (<25%), mild stenosis (25 ≤ stenosis degree <50%), moderate stenosis (50 ≤ stenosis degree <75%), severe stenosis (≥75%), and occlusion.

Statistical analysis of age, gender, HT, and type and bilaterality of MMD between patients with or without renal artery stenosis were performed using χ^2 tests, Mann–Whitney U tests and Fisher's exact tests. Additionally, to assess whether increasing cerebral angiographic staging values were associated with renal artery stenosis, we performed trend analysis using linear by linear association in a two-sided χ^2 test. When the cerebral angiographic staging of the two sides was different, the higher staging was used. A P value of 0.05 was considered to indicate a statistically significant difference.

3. Results

Of the 101 patients with MMD, 58 patients were female and 43 patients were male. The median age was 8 years old (range 2–16). The cerebral angiographic staging for all 101 patients was as follows: 15 patients, grade 0; 38 patients, grade 1; 10 patients, grade 2; 49 patients, grade 3; 47 patients, grade 4; 40 patients, grade 5; and 3 patients, grade 6 (mean ± SD, 3.59 ± 1.33). Baseline characteristics of the patients are detailed in Table 1.

Renal artery stenosis was identified in 8 patients (7.9%, 4 male, 4 female). Four patients had unilateral renal artery stenosis and 4 patients had bilateral renal artery stenosis. Therefore, 12 renal artery stenosis cases were found (7 in the right, 5 in the left). Among these case, 6 lesions were classified as mild degree stenosis (50%, 3 in the right, 3 in the left), 2 lesions were moderate (16.7%, 1 in the right, 1 in the left), 1 lesion was severe (8.3%, right) and 3 lesions were occlusions (25%, 2 in the right, 1 in the left). Nine lesions (75%) were located in the proximal third of the main renal artery. The remaining 3 lesions (25%, 2 in the right, 1 in the left) were occluded from the origin of the renal artery (Table 2).

Six of the 101 patients (6%) with MMD had HT. Five of these 6 patients had renal artery stenosis (83%). One patient who had no renal artery stenosis with HT was diagnosed as having essential HT, and blood pressure controlled with medication. The remaining 5 patients with moderate to occlusion grade renal artery stenosis were diagnosed as having renovascular HT with clinical features. Four of these 5 patients underwent renal artery balloon angioplasty and had relatively well

Table 1 – Baseline characteristics of the study population.

	Negative for renal artery stenosis (n = 93)	Positive for renal artery stenosis (n = 8)	P-value
Age (Median yrs. (range))	8 (2–16)	6.5 (4–13)	0.14[b]
Gender (M/F)	39/54	4/4	0.196[b]
Type of MMD			0.708[a]
Ischemic	69	5	
Hemorrhagic	1	0	
Asymptomatic	0	0	
Atypical	23	3	
Suzuki Stage	(186 hemisphere)	(16 hemisphere)	0.02[a]
0	13	2	
1	35	3	
2	8	2	
3	48	1	
4	40	7	
5	40	0	
6	2	1	
Hypertension (n = 6)	1	5	<0.001[c]
Unilateral MMD (n = 15)	13	2	0.339[c]

MMD = moyamoya disease.
[a] χ^2 test.
[b] Mann–Whitney U test.
[c] Fisher's exact test.