Unique Angiographic Appearances of Moyamoya Disease Detected with 3-Dimensional Rotational Digital Subtraction Angiography Imaging Showing the Hemodynamic Status

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Background: The aim of this study was to identify the unique morphological arterial features in patients with moyamoya disease on 3-dimensional rotational digital subtraction angiography. *Materials and Methods:* One hundred seven hemispheres of 58 consecutive patients with moyamoya disease that were analyzed with fused 3-dimensional images of internal carotid angiograms and vertebral angiograms that were marked with different colors were reviewed. Angiographic findings in the posterior watershed area were classified, and the utility of the classification was analyzed by comparing it with clinical presentations and quantitative hemodynamic parameters obtained with positron emission tomography. *Results:* Two unique angiographic appearances were identified. A vacant vessel appearance (no arterial inflow despite absence of cortical infarction) was observed mostly in transient ischemic attack hemispheres. In hemispheres with a vacant vessel appearance, cerebral blood flow was decreased, cerebral blood volume was increased, and mean transit time was prolonged significantly ($P = .00017$, $P = .0061$, and $P = .00026$, respectively). A cocktail vessel appearance (mixture of carotid and vertebral arterial flow) was most commonly observed in asymptomatic cases, as well as in ischemic hemispheres. Cerebral blood volume increased and mean transit time was prolonged significantly ($P = .036$ and $P = .014$, respectively) in hemispheres with a cocktail vessel appearance. The trend of progression in hemodynamic severity in the order of normal appearance, cocktail vessel appearance, and vacant vessel appearance in the watershed area was statistically significant. *Conclusion:* Fused 3-dimensional digital subtraction angiography demonstrated unique angiographic features in the watershed area, and this represented the degree of cerebral hemodynamic impairment in moyamoya disease. **Key Words:** Moyamoya disease—digital subtraction angiography—positron emission tomography—cerebral ischemia—posterior watershed area.

Introduction

Moyamoya disease is characterized as a progressive stenosis or occlusion of the terminal portions of bilateral internal carotid arteries (ICAs), accompanied by the development of collateral networks to compensate for the reduced cerebral perfusion.[1] Clinical presentations vary, and determination of the treatment strategy is a difficult issue in each case.[2] One of the important factors in determining the treatment strategy is cerebral hemodynamic status, and positron emission tomography (PET) is the most reliable method to evaluate it.[3] Moreover, cerebral angiography is also useful for understanding vascular structure and planning the surgical procedure because of its high spatial resolution and temporal information. Therefore, we perform digital subtraction angiography (DSA) before surgical treatment in all cases. Although the classification of moyamoya disease on conventional cerebral angiography is well-known and quite useful to evaluate the stage of progression,[1] we thought that more detailed information may be acquired by 3-dimensional (3D) rotational DSA. Furthermore, we presumed that the visualization of fused 3D rotational DSA, including different arterial circulation systems, might be helpful to understand this unique and complicated disease. In addition, the relationships between PET data and DSA images should be elucidated because these examinations are mutually complementary for evaluating the status of moyamoya disease.

In the present study, the findings of DSA were retrospectively analyzed in 58 patients with moyamoya disease. The characteristic visualization on fused 3D rotational DSA, especially in watershed areas between the anterior and posterior circulations, was identified, and the angiographic features were compared with the clinical presentations and PET data.

Materials and Methods

A total of 58 consecutive patients with moyamoya disease who underwent DSA at our institution between April 2012 and February 2017 were documented. Twenty-six (44.8%) were male, and 32 (55.2%) were female. The mean age was 25.2 years, and it ranged from 2 to 61 years. Thirty-two patients presented with a transient ischemic attack (TIA) defined as motor, sensory, visual, or language deficits that resolved within 24 hours. Fourteen patients presented with infarction, and 5 presented with hemorrhage. Seven patients were diagnosed incidentally (Table 1). Of 58 patients, 107 hemispheres that were evaluated both with 3D rotational internal carotid angiography (ICAG) and with 3D rotational vertebral angiography (VAG) were selected for the present analysis. Based on the clinical presentation, the hemispheres were divided into 4 groups, that is TIA, infarction, hemorrhage, and asymptomatic. The TIA group included 34 hemispheres. The infarction

Table 1. *Baseline characteristics of moyamoya disease patients*

Age, years, mean (range)	25.2 (2-61)
<16, n (%)	22 (37.9%)
≥16, n (%)	36 (62.1%)
Sex, n (%)	
Male	26 (44.8%)
Female	32 (55.2%)
Type of clinical symptom, n (%)	
TIA	32 (55.2%)
Infarction	14 (24.1%)
Hemorrhage	5 (8.6%)
Asymptomatic	7 (12.1%)
Medical comorbidities, n (%)	
Hypertension	10 (17.2%)
Diabetes mellitus	1 (1.7%)
Dyslipidemia	5 (8.6%)
Smoking	10 (17.2%)
Ischemic heart disease	1 (1.7%)

Abbreviation: TIA, transient ischemic attack.

group and the hemorrhage group included 21 hemispheres and 6 hemispheres, respectively. Forty-six hemispheres were asymptomatic, including those recognized as the contralateral neurological manifestation-free side compared with the symptomatic side of moyamoya disease, and incidentally diagnosed cases on magnetic resonance imaging (MRI). Hemispheres that had previously undergone revascularization surgery were excluded. Collateral supplies from the external carotid artery (ECA) system were analyzed in each hemisphere by conventional DSA. During the same period, 10 patients with 15 hemispheres underwent 3D rotational ICAG and VAG for other diseases, such as unruptured cerebral aneurysm, brain arteriovenous malformation, cerebral cavernous angioma, and polyarteritis nodosa. Males and females were equal in number, and the mean age was 57.5 years, ranging from 15 to 82 years. These 15 hemispheres were analyzed as the non-moyamoya disease group.

All study protocols were approved by the ethics committee of Tokyo Medical and Dental University (M2000-1331). All participants gave their written consent after being informed of the details of the study. In patients under 20 years of age, written consent was given by parents or appointed attendants.

DSA and Fusion Technique

DSA was performed by a biplane flat-panel detector angiographic system "Artis zee biplane" (Siemens Healthcare GmbH, Forchheim, Germany) in all cases. The femoral artery was punctured with placement of a sheath introducer, and a 3Fr or 4Fr catheter was inserted via the introducer. The tip of the catheter was placed in the ICA