

Moyamoya disease in a unique, primarily Caucasian Midwestern U.S. population reveals higher prevalence of autoimmunity

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Introduction

Moyamoya disease is a rare intracranial cerebrovascular occlusive disease of unknown etiology. Moyamoya populations are largely composed of individuals with Asian ethnicity. North American populations reported in the literature have been of a fairly diverse ethnic and racial composition (5-9). Less commonly it can be identified in patients with chromosomal abnormalities (4). In addition to a lack of understanding its etiology, comorbidities and risk factors have not been well studied. Examined here are the characteristics of Moyamoya disease in a unique, primarily Caucasian, Midwestern U.S. population with the goal of providing insight into pathogenesis of this disease.

Methods

After IRB approval, clinical data from patients with angiographic proven Moyamoya disease evaluated at the Mayo Clinic between 1980 and 2011 were collected and statistically analyzed. Prevalence of comorbidities, particularly stroke risk factors and autoimmune diseases, were calculated and compared with that of the general population (1-3). Prevalence of thyroid disease in the Moyamoya population was compared to that among the Mayo Clinic patient population, since this condition varies widely from region to region.



Figure 1. Age distribution. All patients were adults at time of Mayo Clinic presentation, so very few were pediatric at time of Moyamoya presentation.

Results

Data from 94 patients was studied. Percentage of patients who were female was 72.3%, similar to other reports (7-9). Patient ages are shown in Figure 1, and ethnicities are shown in Figure 2. The ethnic composition of this population was primarily Caucasian (87%), with other groups including Asian (7%), African-American (5%) and Native-American (1%). Presentation characteristics are shown in Table 1. Comorbidities were examined to determine whether prevalence was higher among patients with Moyamoya than in the general U.S. population, and this is shown in Tables 2 and 3. A significantly higher prevalence was seen with autoimmune disease, particularly Type 1 diabetes mellitus (8.4%, compared with 0.4% in the general population) and thyroid disease (12.8% hypothyroidism and 2.1% Grave's disease, compared with 5% and 0.4% in the general population, respectively). Cerebrovascular risk factors including smoking, hyperlipidemia, and hypertension were also examined and no significant differences were identified compared with the general population.



Figure 2. Patient race.

Moyamoya Characteristics and Presentation		
Laterality Unilateral Bilateral Unknown	15 (11.1%) 74 (82.2%) 5 (5.3%)	
Location ICA alone ACA alone MCA alone Multiple locations Posterior circulation involvement Unknown	14 (15.9%) 0 9 (10.2%) 61 (69.3%) 10 (10.6%) 10 (10.6%)	
Presentation Ischemic stroke TIA Headache Hemorrhage Cognitive decline Memory Ioss Asymptomatic	54 (58.1%) 32 (34.4%) 20 (21.5%) 15 (18.1%) 6 (6.5%) 5 (5.4%) 3 (3.2%)	

Table 1. Characteristics and presentation of Moyamoya seen among our patient cohort.

Autoimmune disease	Number of patients (%)	Prevalence in general population
Type 1 diabetes mellitus	8 (8.5%)	0.4%*
Thyroid disease (hyper- and hypothyroidism)	16 (17.0%)	6.2%**

Table 2. Autoimmune comorbidities. *Data from U.S. national statistics (2). **Data from Mayo Clinic database, as described in Methods.

Conclusions

This population based study of a unique Midwestern population of Moyamoya patients demonstrates a significantly higher prevalence of autoimmune disease than that observed in the general U.S. population. This would support an underlying autoimmune component to the pathogenesis of Moyamoya, at least in a subgroup of patients with this disease.

Learning Objectives

By the conclusion of this session, participants should be able to: 1) Describe the well-known associations with Moyamoya disease, 2) Describe the characteristics of a unique Midwestern U.S. Moyamoya population, and 3) Describe the new evidence presented here for an underlying autoimmune etiology to Moyamoya, and the implications to managing patients with autoimmune disease and patients with Moyamoya disease.

Cerebrovascular risk factor	Number of patients (%)	Prevalence in general U.S. population
Hyperlipidemia	25 (26.6%)	16.3%1
Hypertension	19 (20.2%)	24.7%3
Smoking history	12 (12.8%)	19.3% ²
Diabetes mellitus type 2	5 (5.3%)	7.9%2
History of radiation to the head	5 (5.3%)	-
History of head trauma	4 (4.3%)	-
History of cerebral inflammatory disease	4 (4.3%)	-
Fibromuscular dysplasia	3 (3.2%)	

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Table 3. Cerebrovascular risk factors. Data for prevalence in general population comes from U.S. national statistics, as referenced. National data excluded for some factors due to low representation among patients.

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