Factors associated with moyamoya syndrome in the Kentucky regional population

Catherine Wang, MD[1] • Stephen Grupke, MD[1] • Jessica Lee, MD[2] • Justin Fraser, MD [1]

[1]Neurosurgery, University of Kentucky • [2]Neurology, University of Kentucky

Background: Moyamoya disease is a progressive occlusive disease of the cerebral vasculature, with abnormal collateral networks bypassing stenotic vessels. Though first described as a bilateral phenomenon affecting East Asians, similar angiographic features are evident in patients with other medical conditions, known as moyamoya syndrome. Most conditions in the literature referencing moyamoya syndrome exist only as case reports. This study reports both new and previously identified conditions associated with moyamoya syndrome within a Kentucky regional population.

Purpose: The goals of this study are twofold; first, to investigate comorbid conditions in patients with moyamoya syndrome, in particular looking for the presence of co-existing autoimmune disorders as well as disorders predisposing to hypercoagulability, and second, to determine whether the presence of diabetes as a comorbid condition is related to the severity of moyamoya disease burden upon presentation, or the success of subsequent revascularization.

Methods: Inclusion criteria for this retrospective chart review were patients evaluated at University of Kentucky Medical Center, from June 1, 2011 to June 1, 2015, diagnosed with moyamoya and treated by the investigators. Data collected include patient demographics, presenting manifestation, vessels involved, co-morbid conditions present, abnormal laboratory values, and treatment administered. Data storage and analysis were performed using REDCap hosted at the University of the Kentucky; the protocol was IRB-approved.

Results: Thirty-one patients with moyamoya were enrolled (6 males, 14 females). Of these, 26 presented with ischemic stroke, 2 with hemorrhagic stroke, and 3 with other (TIA, syncope). 68% had involvement of the internal carotid artery, 71% of the middle cerebral artery, and 22% of the anterior cerebral artery, with 45% having multi-vessel involvement. The most common comorbid condition was hypertension in 61% of patients. Co-existing autoimmune conditions were present in 26% of patients, including autoimmune polyglandular syndrome 1, autoimmune hepatitis, Addison's disease, psoriasis, ITP, hypothyroidism, Crohn's disease, rheumatoid arthritis, lupus, and multiple sclerosis. Another 13% had co-existing pro-thrombotic disorders, including protein C and S deficiency, Factor V Leiden, hemoglobin G trait, and prothrombin G20210A mutation. Diabetes mellitus was a comorbid condition in 11 out of the 31 patients. It was not found to have a correlation to Suzuki grade of moyamoya disease at presentation (p=0.37). Twenty-one patients received surgical intervention; two were treated with intracranial stenting and 19 with indirect encephalo-duro-arterio-synangiosis bypass. All fifteen who received angiogram after intervention showed evidence of neovascularization.

Conclusions: Our retrospective study of moyamoya patients at UK Medical Center reveals multiple co-existing autoimmune and pro-thrombotic disorders. Diabetes mellitus was found to be a common comorbid condition, but was not significantly correlated with severity of disease. Our experience with intracranial stenting and indirect bypass demonstrates excellent evidence of re- vascularization within one year of treatment.