Moyamoya Syndrome Associated with Neurofibromatosis Type 1 in Children: Perioperative and Long-Term Outcome After Pial Synangiosis

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Introduction

Children with neurofibromatosis type 1 (NF1) are at greater risk to develop a progressive arteriopathy consistent with moyamoya. This study aims to evaluate the clinical and radiographic characteristics, surgical course and outcome of surgically treated children with NF1 and moyamoya.

Methods

Records of all patients seen in a pediatric NF1 clinic were reviewed to identify those with both moyamoya syndrome and NF1. Of these children, a subset underwent a standardized cerebral revascularization surgery, pial synangiosis, as part of a consecutive series of 412 moyamoya patients treated at a single institution between 1988-2010.

Results

Thirty-five patients had both NF1 and moyamoya syndrome, with 29 undergoing pial synangiosis. Preoperatively, 21/29 patients (72%) were symptomatic prior to surgery, with the majority (19/21 symptomatic patients, 90%) experiencing ischemic symptoms. No patients presented with hemorrhage. On imaging, 18/29 (62%) had radiographic evidence of prior stroke at time of diagnosis of moyamoya. Ten patients had been treated with cranial irradiation and all (100%) presented with bilateral disease at initial diagnosis. Average age at surgery was 8.1 years (range 1.3-15.5). Perioperative complications included 1 stroke and 1 infection. Average follow-up was 75.5 months (range 9.4 – 253.3), with 21/22 patients (95%) demonstrating stable or improved neurologic status (using the modified Rankin scale) and radiographic data indicating a 13-fold reduction in stroke rate. Patients with previous cranial radiation had higher stroke risk.

Conclusions

To our knowledge, this is the largest reported series of children with NF1 and moyamoya. Other than a relatively high prevalence of asymptomatic patients, the clinical and radiographic features of moyamoya in this population are comparable to primary moyamoya disease. Prior cranial irradiation is associated with greater extent and severity of disease at presentation. Surgical revascularization in this population is safe, with a substantial (13-fold) and durable reduction in stroke risk.

Learning Objectives

By the conclusion of this session, participants should be able to:

1)Describe the importance of recognizing the potential for moyamoya to exist in NF1 patients

2)Discuss, in small groups, the means by which to evaluate these patients preoperatively

3)Identify effective treatments including pial synangiosis and provide a proper review of operative risks for relevant cases.