

Hemorrhagic moyamoya disease—benefits of revascularization

More and more patients with moyamoya disease are being diagnosed and treated in India, and in this context, this report is an important contribution to the literature on moyamoya disease from India. Management of hemorrhagic moyamoya disease is controversial, and this is a welcome report detailing the various aspects of the management of this entity.

As is quite well known, moyamoya disease has two different manifestations—ischemic and hemorrhagic, and both have different pathophysiology. While the ischemic variety predominates in the pediatric population, the proportion of adults having the hemorrhagic variety differs in various ethnicities. According to the most estimates, around 30–60% of adults present with hemorrhage in the Japanese population, while only 14–22% of patients outside Japan have hemorrhagic presentation.^[1] While a number of publications have asserted the central role surgical revascularization for the ischemic variety of the disease, the management of the hemorrhagic variety remains uncertain.^[2] This is important, because the prognosis of the hemorrhagic moyamoya disease is very poor. Only 45% patients of patients suffering from hemorrhage due to moyamoya disease make complete recovery, and the mortality from the first bleed is 6–8%. Moreover, the rebleeding rate has been estimated at 7%/person/year, and the proportion of patients with rebleed having a favorable outcome reduces to 20–25%.^[3] Hence prevention of hemorrhage is of paramount importance in patients with hemorrhagic moyamoya disease.

There would be many approaches to the prevention of hemorrhage in a patient of moyamoya disease. One would be primary prevention of hemorrhage in patients with moyamoya disease. It is well known that patients who undergo surgical revascularization for their moyamoya disease in the childhood have a very small risk of hemorrhage in their adult period. Would revascularization in their childhood even for their asymptomatic moyamoya disease prevent them

from hemorrhage in their adulthood? However, this is an impractical approach, as majority of patients with hemorrhagic MMD are identified at the time of their first hemorrhage.

The other approach would be to identify the population most vulnerable for rebleed. It has been shown that microbleed on MRI is a marker for microvascular vulnerability, and their presence in a patient with hemorrhagic MMD confers a higher risk of rebleed. Dilatation and abnormal branching of anterior choroidal and posterior communicating arteries are risk factors for further hemorrhage. Presence of aneurysms on the abnormal moyamoya vessels is rare and confers high risk of hemorrhage. These aneurysms must be treated with microsurgery or endovascular treatment before attempting any revascularization, because they have a high propensity for rebleed.

The final and most commonly used approach, as the authors describe, is surgical revascularization. Unfortunately, although it is unequivocally shown in large case series and meta-analysis that revascularization is beneficial in reducing ischemic symptoms in MMD, it is not the case with the hemorrhagic variety. Indeed, there is conflicting evidence on the effect of revascularization on the prevention of hemorrhage in MMD. In a large Japanese survey, there was no difference between the rebleed rate in patients undergoing bypass surgery versus those who did not (16.2% vs 16.8%).^[4] However there are other authors, as has been pointed out in this article, who have shown that the patients undergoing bypass have a lower rebleed rate.^[5] The reason behind the discrepancy is not known, but it maybe partly explained by the pathophysiology of the repeat hemorrhage in patients with hemorrhagic MMD. There are two kinds of repeat hemorrhage in MMD. The first hemorrhage usually presents within the first 2 months of original hemorrhage and occurs at the same site, and it is because of the small aneurysms or pseudoaneurysms of the feeding vessels. The second hemorrhage is in a location distant to the presenting hemorrhage and occurs few months to many years following the first hemorrhage, and it is postulated to be due to abnormal hemodynamic stress on the abnormal collateral vessels. Whether surgical revascularization reduces both these factors is unclear from the existing evidence. There are many authors who believe that direct bypass reduces the hemodynamic stress on the collaterals, and there

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is an improvement over the natural history of the disease. A clearer answer would probably be available after the completion of Japanese Adult Moyamoya Trial, which randomizes patients with hemorrhagic MMD to best medical management, or best medical management with direct bypass, with some form of indirect bypass at the surgeons discretion.^[6] At the same time, it would be advisable to undertake studies in the local populations worldwide, because there are definite differences in the presentations of adult with MMD in different populations.

Finally, it is of immense importance that there is a systematic collection of data for patients with moyamoya disease from major institutions in India, so that there are data regarding Indian population. In our experience, there is a strong regional preponderance in patients with MMD, with majority being from the eastern part of India. There is a preponderance of ischemic variety of MMD, and majority of adults present with ischemic symptoms as well. A comprehensive prospective database of the disease from major institutions in India will answer many questions specific to our population, without extrapolating the Japanese or Western data to the Indian setting.

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