Clinical features and treatment outcomes in childhood moyamoya: A case series from a tertiary hospital in Thailand

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Background: Moyamoya is a rare, chronic progressive steno-occlusive disease and has been reported as a cause of childhood stroke worldwide. In children has a more progressive clinical and radiological process than adult. The prevalence of moyamoya is high in East Asian countries compared to the western countries. Surgical revascularization has been increasing used in moyamoya to improve cerebral blood flow and restore reserve capacity and in turn preventing further cerebral infarction. There were few studies of childhood moyamoya in Thailand. This study was done to describe the clinical presenting symptoms, radiographic features, treatment and outcomes

Objective: To describe clinical features, treatment and outcomes of children with moyamoya from a tertiary hospital in Thailand

Materials and Method: We reviewed medical records of children moyamoya in patients aged less than 18 years between January 2005 and June 2020. Demographic data, clinical presentations, radiological findings, treatment and clinical outcomes were reviewed.

Result: Thirty-seven children, 24 girls, with a median age 9.3 years, were identified. The most common presenting symptoms were hemiparesis (89.2%), cranial nerve palsies (21.6%) and speech abnormalities (16.2%). All presented with ischemic events, 22 (59.5%) with arterial ischemic attack (AIS) and 15 (40.5%) with transient ischemic attack (TIA). Nineteen patients (51.4%) were found to have associated diseases and thalassemia were the most common. Twenty-four patients (64.9%) underwent surgical revascularization. Nine patients (26.5%) experienced recurrence of ischemic stroke, and all were in surgical group (5 during immediate post-surgery). At the last follow-up time, 27 (79.4%) were classified as good outcome with a mRS score ≤ 2. No significant difference between surgical and medical group. Patients in medical group had significantly more progressive vasculopathy or new infarction by neuroimaging studies than surgical group, $p= 0.001$. Patients who had AIS were at risk of poor outcome.

Conclusion: Moyamoya is progressive vasculopathy with multiple recurrent ischemic events. Although nearly 80% had a good outcome, the rest 20% required assistance in their activity of daily living. Although the overall outcome assessed by mRS is not different among surgical and medical group, a progressive of vasculopathy or new asymptomatic ischemic events on follow-up neuroimaging studies were significantly more often in medical group. This finding should be taken into account to stratify treatment option in medical group. A further study in a randomized controlled study is needed.

Key words: hemiparesis, moyamoya, moyamoya syndrome, Thailand