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Childhood Moyamoya Disease: A Clinical and Angiographic Study from Eastern India

Moyamoya disease (MMD) is a rare cerebrovascular disease of childhood with majority of cases from Japan. This is a case series of 14 children diagnosed on the basis of characteristic angiographic findings. Various clinical features and chief angiographic findings were analyzed.

Moya moya disease is a rare cerebrovascular disease of unknown etiology. We herein present a case series of 14 children (7 males and 7 females, mean age: 6.89; age range: 2-14 years) identified among 241 diagnostic cerebral angiographies performed over a period of two years. Majority ($n=11$) presented with ischemic stroke; and only one with hemorrhagic stroke. Ten patients had recurrent TIA; weakness of 1 or more limbs was seen in 7; headache in 6 and seizures in 4 patients. Cerebral ischemic symptoms including cognitive defect, speech and sensory disturbances were present in 2, 3, and 3 children, respectively. The only child who presented with hemorrhagic stroke had thalassemia major, and the history of antecedent head trauma was present in one child. None had positive family history of the disorder. Ancillary laboratory tests including blood glucose, serum electrolytes, complete hemogram with peripheral blood smear for sickle cells, and serum lactate levels were normal in all study subjects. The patient with thalassemia major showed a hypercoagulable state.

Cerebral angiographies showed obstruction or stenosis of the supraclinoid portion of the ICA and the proximal portions of anterior and middle cerebral artery with a typical fine network of vessels at the base of brain with hazy, puff-of-smoke appearance, and development of transdural and leptomeningeal anastomoses in all children. Bilateral abnormalities were present in majority (11/14). Two angiographies showed stenosis in posterior circulation also.

Hemorrhagic stroke in pediatric Moyamoya disease is reportedly uncommon. In the present case, it is believed to be as a result of hypertension which occurs due to vasopressive substances provided by multiple transfusions [1]. Apart from stroke, headache was another consistent feature in our series which is presumed to be closely related to cerebral hypoperfusion [2].

Cognitive decline as initial manifestation signifies the need to consider Moyamoya disease in children presenting with such symptoms in absence of typical findings. The unilateral disease can progress to bilateral disease [3] thus requiring long term follow-up later. Involvement of posterior circulation indicating rapid progression of disease, is in accordance with other recent studies from India [4,5].

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