Quasi Moyamoya disease

Inherited or acquired disorders and conditions may present in conjunction with MMD. This condition is known as quasi-moyamoya disease. To identify the clinical features of quasi-MMD in Japan.

A questionnaire was mailed directly to 241 departments that had acknowledged treating quasi-MMD patients in the primary survey.

A total of 114 departments replied to the questionnaire. The data of 108 patients (66 female and 42 male; female to male ratio 1.57) were registered and analyzed. Mean age was 30.6 years old, with a peak in children. Seven patients (7 %) exhibited familial MMD. The initial clinical manifestation was motor weakness, followed by transient ischemic attack and headache. Their imaging study type included ischemic type in 64 patients (63.4 %), bleeding type in seven (6.9 %), and normal in 27 (26.7 %). Stenoocclusive lesion was seen in the internal carotid artery in more than half of the patients. Development of moyamoya vessels was mild in approximately 40 % of the patients. Almost all cases were accompanied by cerebral hypoperfusion. About half of them were unilateral lesion. Vascular reconstruction was employed for the approximately 60 % patients. The prognosis did not change significantly.

Clinical features of quasi-MMD were revealed in the nationwide study. In quasi-MMD, unilateral lesion is dominant, and the development of moyamoya vessels and intracranial hemorrhage are less dominant [1)].

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Hayashi K, Horie N, Izumo T, Nagata I. Nationwide survey on quasi-moyamoya disease in Japan. Acta Neurochir (Wien). 2014 Feb 6. [Epub ahead of print] PubMed PMID: 24499994.