Asymptomatic Moyamoya Disease

J.Sales-Llopis

Neurosurgery Department, University General Hospital of Alicante, Foundation for the Promotion of Health and Biomedical Research in the Valencian Region (FISABIO), Alicante, Spain

In asymptomatic Moyamoya Disease, the patient should previously have no ischemic or hemorrhagic episode and be neurologically free.

They who had previously experienced any episode suggestive of TIA, cerebral infarction, intracranial hemorrhage, seizure, or involuntary movement caused by moyamoya disease should be excluded. Careful medical interview should be performed to distinguish moyamoya disease-related headache from non-specific headache such as tension-type headache [1)].

Epidemiology

The prevalence and incidence of asymptomatic moyamoya disease are still unclear. Previously, asymptomatic patients with moyamoya disease have rarely been sporadically reported. Screening of family members with moyamoya disease has also identified small number of asymptomatic patients. Therefore, the incidence of asymptomatic moyamoya disease had been believed very low. In fact, Yamada et al. (2005) reported the results of a nation-wide questionnaire conducted in 1994 and identified 33 asymptomatic patients (1.5%) out of a total of 2,193 patients [2)].

Diagnosis

Recent development of a non-invasive magnetic resonance examination has increased the opportunity to identify asymptomatic patients with moyamoya disease who have experienced no cerebrovascular events. However, their clinical features, prognosis, and treatment strategy are still unclear because of small number of subjects and short follow-up periods.

Survey

The first multi-center, nation-wide survey focused on asymptomatic patients with moyamoya disease was conducted between 2003 and 2006 in Japan. as a result, totally 40 patients were enrolled from 12 hospitals. Their mean age was 41.4 years ranging from 13 years to 67 years. The female-to-male ratio was 2.1. Clues to the diagnosis were tension-type headache in 14 patients, dizziness in 5, and head trauma in 4. Five patients were incidentally diag- nosed on MrI and Mra performed for a brain health check-up. Five diagnoses were made on MrI and Mra performed for screening, because a member of their family had moyamoya disease diagnosed. They were siblings in two and offspring in three. The remaining seven cases were diagnosed on MrI and Mra performed because of an unrelated disease in other organs. Therefore, the prevalence and incidence may be much higher than considered before. The female-to-male ratio and mean age of the patients in these studies were very similar to those of moyamoya disease as a whole [3)].

Registry

Therefore, Kuroda et al. have designed the Asymptomatic Moyamoya Registry (AMORE) study in Japan. The objectives of this nation-wide, multi-center prospective study are to clarify long-term prognosis of asymptomatic patients with moyamoya disease and to determine the risk factors that cause ischemic and hemorrhagic stroke in them [4].

Treatment

Asymptomatic moyamoya disease is not a “silent” disorder and readily progress to cause ischemic and hemorrhagic stroke. It would also be essential to repeat MRI and MRA at regular intervals when asymptomatic patients are conservatively followed up to detect disease progression before any cerebrovascular events occur [5].

[1], [3]
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