Letter to the Editor

Since the first recognition of moyamoya disease (MD) in 1960s, more than 40 years have passed. It is well known now that the incidence of MD is highest in Japan. Accordingly, many Japanese neurosurgeons believe that MD is an isolated clinical entity and they are most familiar with this disease as evidenced by their enormous experience and a large number of publications from Japan, describing clinical characteristics and surgical techniques for MD.

However, there is profound confusion or definitely conflicting ideas on this disease in the neuroradiological and neurosurgical literature. The Japanese diagnostic criteria of MD (1: steno-occlusive changes at the distal internal carotid arteries, 2: enlarged arteries at the skull base known as moyamoya vessels, and 3: bilateral involvement) excludes the apparently associated diseases such as atherosclerosis, trauma, neurofibromatosis type I, Down syndrome, angiitis, etc. The latter excluded conditions are often referred to as akin-MD or pseudo-MD. The term "moyamoya phenomenon or syndrome," often used in the English literature, is blamed in Japan because this term is interchangeably used both for (true) MD and akin MD, ignoring the discrimination, if any, between the two. MD without any revealing factors today will be proved as akin-MD with certain triggering factors in the near future. This simply implies our ignorance of vascular pathophysiology in MD.

Any diseases that causes steno-occlusive changes at the distal internal carotid arteries can induce reactive angiogenesis (remodeling) of the lenticulostriate arteries as well as the dural (meningeal) arteries. In MD, the most susceptible segment is the distal internal carotid artery, as is the case with post-varicella infection angiopathy. This different susceptibility, thus reactivity, of the intracranial vessels is elegantly, but hypothetically explained by segmental ideas (susceptibility and vulnerability) of the cerebral vasculature [1]. In fact, the concept that angiographic stenoses in the distal internal carotid artery and superficial temporal artery in MD can be compared was denied [3]. Each segment has different phylogenetic, embryological, and hemodynamic backgrounds, as is the case in MD [2]. Many Japanese researches on MD are oriented to the surgical techniques (direct/indirect anastomoses, additional bypass, or extensive craniotomy), ignoring these backgrounds of diverse etiologies of MD. I believe that the Japanese exclusion criteria on MD are misleading and must be more comprehensive for the better understanding of this clinical syndrome.

References

1 Lasjaunias PL: Segmental identity and vulnerability in cerebral arteries. Interventional Neuroradiology 6: 113-124, 2000.
2 Komiyama M: Moyamoya disease is a progressive occlusive arteriopathy of the primitive internal carotid artery. Interventional Neuroradiology 9: 39-45, 2003.
3 Komiyama M, et Al: Steno-occlusive changes in the external carotid system in moyamoya disease. Acta Neurochir (Wien) 142: 421-424, 2000.

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