

Vascular Cognitive Impairment in Adult Patients with Moyamoya Disease

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Abstract

Moyamoya disease (MMD) in adult patients is commonly accompanied by vascular cognitive impairment (VCI). The mechanism behind is still unclear and has been related to its hemodynamic disturbances. Surgical revascularization seems to be the only efficacious way to maintain the cognitive outcome, while problems such as the selection of operation indications and timing, and the long-term efficacy still need to be solved. The purpose of this article is to review the related literatures in recent years and to summarize the results in our research on the mechanism and surgical intervention of VCI in patients with MMD.

Keywords: Moyamoya disease; Vascular cognitive impairment; Blood oxygen level-dependent functional magnetic resonance imaging

Introduction

Moyamoya disease (MMD) is a chronic cerebrovascular disease, characterized by the progressive stenosis of the arteries of the circle of Willis, which results in the formation of a collateral network of capillaries at the base of the brain [1]. The distribution of age at onset of MMD has been suggested at 5 years of age as children and at about 40 years of age as adults [2]. Adult patients with MMD frequently suffer from vascular cognitive impairment (VCI), the etiology of which has been associated with hemodynamic disturbances. Simply put, VCI is a syndrome with evidence of subclinical cerebral vascular damage or clinical stroke and cognitive disorder affecting at least 1 cognitive domain: memory, executive function/attention, language and visuospatial functions [3]. And executive function/attention has been proved to be the main cognitive domain that deteriorates in adult MMD [4].

Cerebral hemodynamic failure and executive dysfunction

Executive function, which is reported to be predominantly affected in adult MMD, has been associated with reduced cerebrovascular reserve (CVR) in frontal areas as measured with perfusion MRI and the acetazolamide challenge [5]. However, previous studies of adult MMD revealed that executive dysfunction is the consequence of ischemic damage to dynamic factors such as cerebral hypoperfusion, rather than to cerebral gray matter [6,7]. In addition, previous studies revealed that VCI is a continuous process in adult MMD with the progression of cerebral hypoperfusion [8,9]. Thus, early detection of frontal hypoperfusion may be the first step to save the cognition.

Insights into the mechanism of VCI in adult MMD

Our recent research focused on the correlation between VCI and intrinsic brain activity in adult patients with MMD. First of all, since many popular neuropsychological tests, especially the screening tests such as the Mini-mental state examination (MMSE) and Montreal Cognitive Assessment (MoCA), were developed for MCI and AD, their application in VCI is limited. Thus we examine the applicability of a battery of cognitive assessment scales covering global cognition, executive, memory, language and visuospatial functions in a cohort of consecutive patients with cerebral ischemia. Results showed that the executive subtest of Memory and Executive Screening (MES-EX) and the Trail Making Test-B (TMT-B) were of the most sensitive and specific in detection of VCI [10].

Then we adopted the resting-state blood oxygen level-dependent functional magnetic resonance imaging (BOLD-fMRI) to investigate the spatial pattern of intrinsic brain activity in adult MMD. It has been

demonstrated that brain activity is spatially organized as networks for specific tasks. The central executive network (CEN), comprised of the dorsolateral prefrontal cortex and posterior-parietal cortex, contributes fully to executive function. Also, the self-referential network (SRN), the default mode network (DMN) and many other networks contribute partially to executive function. Our results firstly presented the specific functional pattern of adult MMD, and then indicated that this pattern changes following cognitive impairment.

In addition, we investigated the efficacy of surgical procedures used to treat adult MMD with both neuropsychological tests of high sensitivity and specificity and resting-state BOLD-fMRI. Previous study has implied that surgical revascularization reverses cerebral cortical thinning [11]. Thus we hypothesized that cognitive dysfunction might also be reversed. Our further results will be published to debate the issue.

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