Perspective

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Fragile Collateral Vessels at the Base of The Brain

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Description

Moyamoya disease is a cerebrovascular condition predisposing affected patients to stroke in association with progressive stenosis of the intracranial internal carotid arteries and their proximal branches. Patients with characteristic moyamoya vasculopathy plus associated conditions are categorized as having moyamoya syndrome. This review describes the demographic characteristics, pathogenesis, evaluation, and treatment of moyamoya disease and syndrome.

Multifactorial Inheritance

Moyamoya disease is a specific chronic cerebrovascular occlusive disease first reported by Japanese surgeons in 1957. The disease is characterized by stenosis or occlusion of the terminal portions of the bilateral internal carotid arteries and abnormal vascular network in the vicinity of the arterial occlusion. It may cause ischemic attacks or cerebral infarction, which is more frequent in children than in adults. In adults, cerebral hemorrhage may occur. The disease is distributed in all age groups, but the highest peak is in childhood at less than 10 years of age. The characteristic histopathology features of the steno-occlusive arteries are fibro cellular thickening of the intima containing proliferated smooth muscle cells and prominently tortuous and often duplicated internal elastic lamina. There is usually no athermanous plaque in the arterial wall. Etiology of the disease is still unknown; however, multifactorial inheritance is considered possible because of a higher incidence of the disease in Japanese and Koreans and approximately 10% of familial occurrence among the Japanese.

Moyamoya disease is an uncommon cerebrovascular disease that is characterised by progressive stenosis of the terminal portion of the internal carotid artery and its main branches. The disease is associated with the development of dilated, fragile collateral vessels at the base of the brain, which are termed moyamoya vessels. The incidence of moyamoya disease is high in east Asia, and familial forms account for about 15% of patients with this disease. Moyamoya disease has several unique clinical features, which include two peaks of age distribution at 5 years and at about 40 years. Most paediatric patients have ischaemic attacks, whereas adult patients can have ischaemic attacks, intracranial bleeding, or both. Extracranial–intracranial arterial bypass, including anastomosis of the superficial temporal artery to the middle cerebral artery and indirect bypass, can help prevent further ischaemic attacks, although the beneficial effect on haemorrhagic stroke is still not clear. In this Review, we summarise the epidemiology, aetiology, clinical features, diagnosis, surgical treatment, and outcomes of moyamoya disease. Recent updates and future perspectives for moyamoya disease will also be discussed.

Moyamoya Disease

Moyamoya, meaning a “hazy puff of smoke” in Japanese, is a chronic, occlusive cerebrovascular disease involving bilateral stenosis or occlusion of the terminal portion of the internal carotid arteries (ICAs) and/or the proximal portions of the anterior cerebral arteries and middle cerebral arteries (MCAs). The Ministry of Health and Welfare of Japan has defined 4 types of Moyamoya Disease (MMD): ischemic, hemorrhagic, epileptic, and “other.” The ischemic type has been shown to predominate in childhood, while the hemorrhagic type is more often observed in the adult population. The highest prevalence of MMD is found in Japan, with a higher female to male ratio. Studies have shown a possible genetic association of MMD linked to chromosome 17 in Japanese cases as well as in cases found in other demographics. During autopsy, intracerebral hematoma is found and most commonly serves as the major cause of death in patients with MMD. Moyamoya vessels at the base of the brain are composed of medium-sized or small muscular arteries emanating from the circle of Willis, mainly the intracranial portions of ICAs, anterior choroidal arteries, and posterior cerebral arteries, forming complex channels that connect with distal positions of the MCAs. Off of these channels are small tortuous and dilated vessels that penetrate into the base of the brain at the site of the thalamoperforate and lenticulostriate arteries. On angiography, there is the characteristic stenosis or occlusion bilaterally at the terminal portion of the ICAs as well as the moyamoya vessels at the base of the brain. Six angiographic stages have been described, from Stage 1, which reveals a narrowing of the carotid forks, to Stage 6, in which the moyamoya vessels disappear and collateral circulation is produced solely from the external carotid arteries. Cases with milder symptoms are usually treated conservatively; however, more severe symptomatic cases are treated using revascularization procedures. Surgical treatments are divided into 3 types: direct, indirect, and combined/other methods.

Direct bypass includes superficial temporal artery-MCA bypass or use of other graft types. Indirect procedures bring in circulation to the intracranial regions by introducing newly developed vasculature from newly approximated tissues. These procedures may not be enough to prevent further ischemia; therefore, a combination of direct and indirect procedures is more suitable. This article will give a review of the epidemiology, natural history, pathology, pathophysiology, and diagnostic criteria, including imaging, and briefly describe the surgical treatment of MMD.