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Review Article

Moyamoya Disease -Factors and Prevalence for Perivascular Spaces in Post-Neurological Adult Patients: Neurosurgeon's Perspective

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Abstract

Moyamoya disease is a blood vessel disorder that occurs in the brain on the internal carotid arteries and its branches. The blood vessel in the carotid artery is narrowed and blocked, reducing and preventing blood flow to the brain. The condition results in the narrowing of the vessels distributing oxygen-rich blood to the brain. This report explores the results and effects of the disease, its pathophysiological, biochemical, genetics and scientific analysis of the disease. This also includes the clinical implementations and unanswered questions concerning the moyamoya disease.

Keywords: Enlarged perivascular space; Moyamoya disease; Vascular disease

Introduction

The Moyamoya disease was named from a Japanese meaning 'puff of smoke', usually resulting from blocked arteries at the brains base. It often appears as tiny blood vessels that form to compensate for the blocked artery. As the blood vessel becomes more blocked, individuals are more likely to suffer from stroke. The condition results in the narrowing of the vessels distributing oxygen-rich blood to the brain [1]. The primary characteristic of moyamoya disease is chronic progressive stenosis which affects the internal carotid artery [2]. A vascular network with abnormality is formed at the base of brain by the stenosis, which also reduces cerebral blood flow and perfusion pressure [3]. Moyamoya disease patients have an increased risk of developing EPVs (enlarged perivascular spaces), which mainly affect female adult patients who have high flow of voids and basal ganglia, high resonance angiography scores detecting coronary stenosis regarding calcium scores in the body, white matter lesions, and ivy signs including swelling, redness, itching, blisters, and difficulties breathing [4]. Flow of voids refer to the signal loss of blood and other fluids with a combination of spin-phase and time-of-flight effects. EPVs are detected through a T2-weighted MRI and they indicate the presence of small vessel disease. The rare disease is progressive, with an appearance of a puff of smoke in the brain area [5]. It is unfortunate that moyamoya is not curable [6]. However, experts suggest that a cerebral revascularization and vascular bypass surgery are considered effective as they help in diverting blood to nearby muscles from the blood vessel, with oxygen finally reaching the oxygen stared brain areas, also reducing the risks of stroke and long-term drastic outcomes [7]. The purpose of this study will be to explore the moyamoya disease through data collection, exploration of the results, pathophysiology, biochemical, genetics, and scientific analysis, later analyzing its clinical implications among other unaddressed issues concerning the disease.

Discussion

Pathophysiology of disease

Moyamoya disease mainly affects the children and the elderly. Research has identified that the Chinese, Korean, and Japanese are at higher risk of getting the disorder [8]. The disorder is also progressive and cannot stop without treatment. Again, it is not curable, but brain surgery is considered the only option to guarantee blood flow back to the brain [9]. Mainly, the pathophysiology of moyamoya disease is unclear. However, it is usually an abnormal cerebrovascular condition whereby patients experience abnormal perforation of blood vessels guaranteeing collateral circulation [10]. The chronic progressive disease has been reported given the increased use of advanced diagnostic radiology systems and medical check-ups [4]. The pathogenesis and etiology of the disease remain unclear. Evidence suggests that the underlying cause of the moyamoya disease is vascular anomalies within the endothelial progenitor cells [11]. Currently, it affects about 15% of patients through autosomal dominant inheritance patterns [12]. Genome-wide studies and genetic analysis on moyamoya disease proves promising to identify the exact pathophysiology of the condition [13].

Biochemistry of disease

Moyamoya disease (MMD) involves the major stenosis of vessels of the circle of Willis [3]. The major proteins involved play active roles during the pathogenesis of the disease including basic fibroblast, vascular endothelial, Beta 1, and hepatocyte growth factor [14]. Further studies have also linked reduced angiogenic activity and with moyamoya disease among patients. Most believe that RNF213 and R4810K prove the susceptibility of mutations with the disease [15].

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Genetics of the disease

Recent research showed low penetrance polygenic mode or autosomal dominance transmissions involving the chromosome 3,6,12, 17, and 8 familial with moyamoya disease [16]. Scholars argue that this way, the disease can be passed through genes from families over hereditary periods. Moyamoya has also been associated with RNF213 [7]. Other studies argue that an excess of proteins at these cells stimulate blood vessels to grow, hence the characteristics leading to the [12]. The symptomatic of these processes include insufficient blood supply in the body cells, hence the rupture of collaterals causing stroke and death to the patient. Genetic approaches hence locate susceptibility of loci in affected families to guarantee a successful understanding of the disease [17].

Scientific Analysis

An accurate analysis of moyamoya disease often involves diagnosis in the clinical setting. The diagnosis process involves MRI imaging to depict the reduced blood flow at the anterior and middle cerebral arteries and internal carotid artery [3]. The imaging shows increased collateral blood, hence confirming the disease [7]. This process also requires an angiogram. Abnormal vascular networks often develop on the stenotic lesions or occlusive in the arterial phase. MR imaging shows the occlusion or stenosis of the middle cerebral artery (MCA) or anterior cerebral ACA [18]. From the beginning, it is evident that moyamoya disease did not depict stenotic changes at the ICA terminal region [19]. The second option integrated is the Suzuki angiographic staging, with is being effective in understanding its pathology among each patient. The staging method also shows the severity of the disease in the Internal carotid artery (ICA) region and subsequent vessels [1]. Thirdly, supportive findings through genetic evaluations and histopathological analysis help understand the susceptibility of the gene among populations [15]. Histological analysis of specimens at surgery and autopsy supports the diagnosis of the diseases, with genes RNF213 and 17q25-ter proving high susceptibility for moyamoya disease [2].

The Clinical Implication of Disease

Firstly, moyamoya disease is associated with diseases such as Graves disease among other chronic illnesses among patients [20]. Others experience cerebrovascular ischemic effects that may result in thyrotoxicosis, including an excessive demand for oxygen given the deficit at the cerebral blood flow [21]. The disease has multiple clinical features, whereby the doctor must be informed in case they appear. Most patients have two peaks at the age of 5 and 40 years, with pediatric patients posing a higher likelihood of having ischemic attacks and intracranial bleeding [10,20]. Down syndrome and thyroid dysfunction have been strongly linked and interrelated to each other [22]. High levels of thyroid risk causing vascular changes and altering the sympathetic stimuli sensitivity [23]. If the graves' disease is not treated, it increases the stiffness of carotid arteries as it is the case with ultrasonography [21]. The most common symptoms of the disease include seizures at an early age, headache, cognitive decline, developmental delays, visual disturbances, weakness and numbness in the body and joints [24].

The Unanswered Questions

Most of the unanswered questions about moyamoya disease include the lack of an exact biological, genetic, biochemical, pathophysiology, and genetics of the disease [15]. This means that more research must be conducted to guarantee that clinicians find the best way to prevent the disease in the future [25].

Conclusion

This study confirmed an increase in the prevalence rate of EPVs among moyamoya disease patients. The presence of other health condition such as hypertension, basal ganglia, and flow voids also increased the risk factor among female patients. Reduced pulsation of the arterial is the major contributor of the increased rate as it inhibits interstitial fluid flow [26,27]. Following the systematic studies, moyamoya disease proved to continually affect many individuals across the states. It is the role of the government and healthcare institutions to ensure that they find more research and analysis on moyamoya disease to identify the probable causes of the disease and ways to guarantee patients protection and prevention from the disorder. All patients with the disorder are required to visit the doctor immediately, ensuring that they are treated to prevent the severity of their illness. It is also important to conduct further research on patients with moyamoya disease to create an understanding of the pathophysiological importance of EPVs and how they can be prevented after performing a surgery on the patients [28,29].

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