Indian Journal of Pharmacy and Pharmacology 2023;10(2):69–72

Content available at: https://www.ipinnovative.com/open-access-journals

Indian Journal of Pharmacy and Pharmacology

Journal homepage: https://www.ijpp.org.in/

Review Article

Moyamoya – A progressive disorder an update and review

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ARTICLE INFO

Article history:
Received 10-05-2023
Accepted 16-06-2023
Available online 04-07-2023

Keywords:
Moyamoya
Cerebrovascular disease
Neurological

ABSTRACT

There is stenosis or narrowing of the internal carotid artery in Moyamoya disease, a relatively rare kind of neurological condition. This may result in illnesses like ischemia, haemorrhage, or even a potentially fatal stroke. It is still unclear what this disease's specific pathogenesis is understood. The Moyamoya disease has a total of six stages. Scientists have not yet developed a drug that can fully treat moyamoya disease because it is such a rare disorder. Surgery is the only approach that works here. But once more, this won't help with finishing the course of treatment. It will only aid in slowing the spread of the disease

1. Introduction

"Moyamoya disease" is a chronic neurological disorder where there is stasis of the internal carotid artery and the circle of Willis. It was first mentioned in the Japanese literature in the year 1957 and then, it was later named by the Japanese scientists Suzuki and Takaku in the hear 1969.[1] It is an intrinsic pathological disease condition where the complete etiology is not well understood. Because of the narrowing of the artery, the patient experiences less amount of blood circulation to the cerebrum. Less blood circulation leads to ischemia, which is less supply of oxygen. By the action of the body's compensatory mechanism, collateral vessels start to develop and smaller newer blood vessels get enlarged. This can be found by performing a diagnostic cerebral angiogram. In the angiogram, a puff-like smoke is observed which is termed moyamoya, which is due to enlarged lenticulostriate and thalamoperforating arteries.[2] The stenosis of the carotid artery is not only due to atherosclerosis or inflammation, but it may also occur because of some other reasons.[3]

2. Pathology

When the specimens are observed, they showed the present hyperplasia of proliferating smooth muscle cells, especially in the tunica media region of the arterial wall.[4] It was impaired by an irregular layering of elastic lamina.[3] There was a micro-aneurysm formation, which leads to a higher risk of haemorrhage mostly in the adult patients suffering from Moyamoya disease.[5] Apoptosis may harm the arterial walls and so it can damage the arterial wall.[6] The formed collateral arteries are very sensitive and so they can lead to aneurysm development.[7] As the arterial wall has to undergo high pressure and so there is increased stress. Because of this, the arterial walls become weak in elasticity and they may lead to haemorrhage. The mitochondria of Endothelial colony-forming cells in the cross-sectional histologic assessment of arteries from affected patients demonstrate an atypically thickened tunica intima and a decreased tunica media layer.[8] Assessing the arteries of the patient suffering from Moyamoya disease by cross-sectional histologic shows a thick tunica intima and reduction in the tunica media layer.[9]

The exact pathology of moyamoya disease is still not properly known.

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https://doi.org/10.18231/j.ijpp.2023.027

2.1. The following types of moyamoya disease (involvement of chromosomes) have been described in the literature:

1. MYMY1-chromosome 3p
2. MYMY2-RNF213 gene on chromosome 17q25
3. MYMY3-chromosome 8q23
4. MYMY4-X-linked recessive condition characterized by MMD, short stature, hyper gonadotropic, hypogonadism, and facial dysmorphism
5. MYMY5-ACTA2 gene on chromosome 10q23
6. MYMY6 with achalasia-GUCY1A3 gene on chromosome 4q32

2.2. Diagnosis[10–12]

1. Diagnosis of the Moyamoya disease is done by observing-
2. Abnormal vessels forming network (Moyamoya vessels)
3. Unilateral lesions
4. Stenosis or narrowing in the internal carotid artery
5. Fibromuscular dystrophies
6. Intracranial haemorrhage
7. Paediatric craniopharyngioma
8. Anterior circulation stroke
9. Basilar artery thrombosis
10. Blood dyscrasias
11. Cavernous sinus syndromes
12. Cerebral aneurysms
13. Dissection syndromes

The above-mentioned parameters can be checked which can be helpful for the diagnosis of moyamoya disease, Stages in Moyamoya disease.[13]

1. *Stage:* It is characterized by the narrowing of the internal carotid artery. This can be viewed by angiographic examination.
2. *Stage:* It is characterized by angiographic examination, where there is initiation and appearance of basal moyamoya. The arteries in the cerebrum get dilated.
3. Stage**:** It is "Intensification of Moyamoya artery.When the angiographic examination is performed, the moyamoya vessels appear as puff-like smoke. The deflection of the anterior cerebral artery (ACA) and middle cerebral arteries (MCA) is noted.
4. *Stage:* It is "minimization of basal moyamoya".Here, the moyamoya vessels start reducing and the transdural collateral is visible. Here, the deflection of the posterior cerebral artery (PCA) is recorded.
5. *Stage:* It is the "reduction of Moyamoya disease". In the angiographic examination, the absence of the cerebral arteries is seen Along with this, an increase in the transdural collateral is also seen.
6. *Stage:* It is the "disappearance of moyamoya".In the angiographic examination, the moyamoya vessels are no more visible. There is an occluded internal carotid artery seen. The blood circulation to the cerebrum is done from the external carotid artery.

3. Evaluation/Examination

3.1. Magnetic resonance imaging (MRI)

It is used to detect small subcortical lesions that cannot be detected by performing Computed Tomography (CT) It is sensitive and non-invasive. It can also help to find out the haemorrhage or brain stroke. They help in the visualization of the occluded or the blocked vessels of the immoral carotid artery The moyamoya vessels are observed as unusual vessels in MRI examination. And if there is any opinion it can be observed bilaterally.[14–16]

3.2. Magnetic resonance angiography (MRA)

Magnetic resonance angiography (MRA) is widely used in a majority of magical cores for the visualization of small vessels and is also used to assess cerebrovascular diseases with higher safety. The transdural collateral blood supply was properly seen using MRA technology (Sacket al., 2000). It is a non-invasive procedure. It is revealed that now the asymptomatic Moyamoya disease is much more serious than Cartier period.[17,18]

3.3. Conventional cereal angiography

It is also one of the methods used for the evaluation of Moyamoya disease in the cerebrum It is used to see the intracranial aneurysm, by using a contrast agent. It shows the opacified blood vessels in the brain. So if the patient is suffering from moyamoya disease, the occluded internal carotid artery can be seen along with the formation of collateral blood vessels.[19,20]

3.4. Transcranial doppler (TCD)

It uses a non-invasive sound method to study the cerebral blood flow velocity and pulsating index. It shows the blood flow in the major intracranial ones, which works the principle of the Doppler effect where the movement of blood flow in the vessels can be demonstrated. It is operator dependent process where skilled professionals are required to operate the device and so it is less used in compared to MRI and MKA.[21,22]

3.5. Electroencephalography (EEG)

It is used to determine and check the electric signal in the brain where an electrode is attached to the scalp. The observation is in the form of wavy lines. It is also very important for the diagnosis of patients suffering from

seizures. Suzuki and Kodoma have given a distinctive ECG finding in around 50% of the patients suffering from a moyamoya disease. This was termed as 'Rebuild-up' phenomenon, which is a reappearance of the slow waves of the higher amplitudes within 20-30 seconds followed by cessation of hyperventilation. [23–25]

3.6. Cerebral perfusion measurement

1. It is defined as the net pressure gradient that supplies oxygen to the brain tissues. It is calculated as the difference between the mean arterial pressure (MAP) and the intracranial pressure (ICP). It is measured in millimeters of mercury (mm Hg). [26] It generally shows.
2. Increase in oxygen fraction extraction.
3. Decreased global cerebral blood flow with posterior cerebral blood flow distribution.
4. Cerebrovascular reactivity to carbon dioxide and acetazolamide in the internal carotid artery region [27]

3.7. Treatment

Up till now, there is no proper treatment approach for the treatment of moyamoya disease. If any individual has any of the symptoms related to Moyamoya disease, then he or she should immediately get it diagnosed. Early diagnosis of the disease and surgery intervention within the specified time may help in preventing the situation from worsening. Surgical interventions will only act as a secondary prevention for the disease condition, it will stop the disease progression.

4. Conservative Management

It helps in maintaining blood circulation to the cerebral tissues and it prevents the risk of stroke, which may eventually lead to Moyamoya disease. Aspirin, a blood thinner, is most commonly used in patients suffering from Moyamoya disease so that stroke chances can be prevented. It is used as a maintenance therapy for reducing the chance of thrombosis due to stenosis of the artery. The normal dose of Aspirin consumed is between 50 to 100 mg. Any other analgesic or anti-epileptic drug can also be used to prevent headaches or chances of epileptic attack. [28]

4.1. Surgical revascularization

It is considered one of the most very important methods for the treatment of Moyamoya disease. It improves the blood flow to the cerebral tissues which eventually helps in prevention or reducing the chances of strokes to a larger extent. Some indication includes apparent cerebral ischemia, decreased cerebral blood flow, etc. This method is most commonly adopted for children as it is highly recommended by paediatrics because in children the rate of the disease progression is very fast.

4.2. Surgical revascularization includes two methods namely

1. Direct revascularization
2. Indirect revascularization
3. Direct revascularization

It is a complicated method of surgical intervention where skilled surgeons are required and the cerebral blood flow is monitored carefully. Here, the superficial temporal artery (scalp artery) is used as the main blood-supplying vessel in direct bypass. In simple terms, the scalp artery is connected to the cerebral artery to increase the blood flow to the cerebral tissues,

4.3. Indirect revascularization

It is a simpler method as compared to direct revascularization. The only drawback of this type of surgical intervention is that the time required for increasing the cerebral blood flow is more. Some of the major techniques performed under this method are encephalon spongiosis (EMS), encephalon duro arterial myo synangiosis (EMAS), etc. By performing these techniques, the blood flow rate in an ischemic brain can be enhanced. [29,30]

5. Conclusion

It was observed that Moyamoya disease is a chronic type of disease that causes stenosis or occlusion of the internal carotid artery. Due to this the blood flow to the cerebral tissues is highly affected. Lesser blood supply to the cerebral tissue may lead to chances of hemorrhagic events, ischemic conditions, stroke, or episodes of an epileptic attack. There is no medicine for the complete treatment of this disease. But the disease progression can be reduced by performing the surgical revascularization technique which includes direct revascularization and indirect revascularization. Several other studies are being carried out to understand the pathophysiology of the Moyamoya disease condition. In the future, we can also see some research work on the medicines that would be beneficial in effectively treating the moyamoya disease condition.

6. Source of Funding

None.

7. Conflict of Interest

None.

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Cite this article: Khan S, Anas Shaikh M, Shaikh A, Wais M. Moyamoya – A progressive disorder an update and review. *Indian J Pharm Pharmacol* 2023;10(2):69-72.