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REVIEW ON MOYAMOYA (PUFF OF SMOKE)- A RAREST DISEASE

J. Kavitha[1], S. Sivakrishnan[2]*

1. *Associate Professor, Department of Periodontia, RMDCH, Annamalai University, Annamalai Nagar, Chidambaram-608002, Tamilnadu, India.*
2. *Assistant Professor, Department of Pharmacy, FEAT, Annamalai University, Annamalai Nagar, Chidambaram-608002, Tamilnadu, India.*

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ABSTRACT

Moyamoya is a progressive disorder that affects the brain's blood vessels (cerebrovascular). It is characterized by the stenosis (narrowing) and/or occlusion (closure) of the carotid artery in the skull, the main artery that carries blood to the brain. At the same time, tiny blood vessels at the base of the brain open up in an apparent effort to carry blood to the brain distal to the blockage. MMD can occur at any ages, though symptoms that usually occur in 30-50-years-old adults and 5-10-years-old children. The first symptom of MMD is usually recurrent transient ischemic attack (TIA) or stroke, particularly in children. Adults may experience these symptoms and also bleeding in the brain (hemorrhagic stroke) from abnormal blood vessels in the brain. Diagnosis may include MRI, CT, Cerebral angiogram, Transcranial Doppler ultrasound, PET, SPECT, and EEG. Medication may be prescribed to help in seizure control or decrease the risk of stroke, including Calcium channel blockers, Blood thinners, Anti-seizure medications, and Revascularization surgery are also performed for MMD. This article evaluates the types, causes, diagnosis, symptoms, and treatment of MMD.

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Introduction

Moyamoya disease [1, 2] is a rare blood vessel (vascular) condition, which blocks or narrows the carotid artery in the skull and restricts blood flow to the brain. Then, tiny blood vessels start up at the skull base to continue to provide oxygen to the skull. Such tiny vessels are the vessels dubbed "moyamoya," after which the disease was named.
Inadequate blood flow instead results in a decreased oxygen supply to the brain and it is this lack of oxygen that triggers symptoms of medium age.[3-5] Within the brain, the disorder can cause a TIA, stroke, or bleeding [6]. It can also influence the functions of the brain and induce cognitive and behavioral disorders or disabilities.

Epidemiology

For unknown reasons, MMD is relatively more prevalent in East Asian countries such as Japan and Korea, than those living in the Western Hemisphere. MMD more often affects infants, although the disorder can occur to adults. This may be attributed to certain hereditary differences in certain communities. Approximately 10 percent of Moyamoya instances have a hereditary source in Asian countries. Even though MMD can occur at any age, there are two peak occurrence rates in children between the ages of five and ten, and in adults between 30 and 50 years. MMD is found worldwide, but is more prevalent in East Asian countries, especially Korea, Japan, and China. This may be attributed to certain hereditary differences in certain communities. MMD usually develops in people below the age of 20 in Japan. The disorder is projected to occur in 1 of every 300,000 people in Japan. While Moyamoya was originally identified in Japanese ethnicity persons, cases have been reported from other parts of Asia as well as from Europe, North and South America, and most series recorded in the western hemisphere have a number of Asian descent patients. Of interest, most patients are acute cases in North America, with current studies indicating that fewer than 4 percent of the cases in this demographic are maternal[7].

Signs & Symptoms

MMD may occur at any age, there are two peak occurrence cycles in children between the ages of 5 and 10 years, and in adults between 30 and 50 years. Children with a medium-sized disease can present with a number of symptoms, but often

Corresponding Author: Dr. S. Sivakrishnan, Assistant Professor, Department of Pharmacy, FEAT, Annamalai University, Annamalai Nagar, Chidambaram-608002, Tamilnadu, India. Email: sivacdm82 @ gmail.com

with those linked to decreased blood flow in the brain, including stroke, TIAs, headaches, epilepsy, involuntary or sometimes progressive developmental delay.

While moyamoya adults have symptoms and signs of brain ischemia [8], they are much more prone to experience intracranial hemorrhage than the infants, possibly attributable to the collapse of the tiny Moyamoya blood vessels in the environment of elevated blood pressure seen in adulthood.

Accompanying symptoms and signs [9, 10] of MMD due to decreased blood supply to the brain include vomiting, hallucinations, eye, arm or leg stiffness, numbness or paralysis, usually on one side of the body, vision problems, difficulty with certain persons communicating or hearing (aphasia), cognitive deficits, and repetitive gestures. These symptoms can be triggered by exercise, crying, coughing, straining, or fever.

Causes

The triggers of the moyamoya are unclear. This is understood, however, whether the condition can manifest as a discrete, predominant disease that can or must exist in combination with a variety of separate underlying disorders, as described above. It is likely that moyamoya phenotypic arteriopathy currently represents a specific end-path of a variety of pathophysiologically distinct processes.

The main common disorder may be inherited genetically as an autosomal recessive condition and accounts for around 10 percent of all Japanese events. Two big mutations have recently been documented to be correlated with different subpopulations of the patients with moyamoya. The first, R179 mutations in the ACTA2 gene, interact with a radiographically distinct subtype of MMD, found in a very limited subset of patients affiliated with a wider population of ACTA2 mutations affecting the heart and aortic disorders [11]. More specifically, mutations in RNF213 are closely correlated with the adulthood-presenting classic East Asian, longitudinal, idiopathic family disorder, which may appear in up to 70 percent of all Eastern Asian moyamoya family instances [12]. Secondary MMD develops in combination with numerous neurological diseases or illnesses, including some central nervous system infections.

- ✓ **Family history:** Family member with MMD, risk of developing the disease is 30 to 40 times higher than that of the general population — a factor that strongly suggests a genetic constituent.
- ✓ **Medical condition:** MMD often exists in conjunction with another illness, namely type 1 neurofibromatosis, sickle cell disease [13] and, among many others, Down syndrome [14].
- ✓ **Being female:** Females have a significantly higher prevalence of the disease with middle age.
- ✓ **Being young.** Although adults may have MMD, children below 15 years of age are most frequently affected.

While the cause of the disease is unclear, some factors can increase your risk of having the infection, including:

- ✓ To be of Asian origin: MMD is found worldwide, but is more prevalent in East Asian countries, especially China, Japan, and Korea. This may be attributed to certain hereditary differences in certain communities. This same higher prevalence has been reported among western Asians.

Complications

Many moyamoya condition symptoms are linked with stroke consequences such as:

- ✓ **Sight issues**: Any individuals with MMD suffer vision problems as a consequence of the stroke.
- ✓ **Movement disorders:** For certain people with MMD, though uncommon, spontaneous activation of some muscles occurs.
- ✓ **Learning or developmental issues.** A child can have mental-processing issues following a stroke, which may impair schoolwork as well as cause social challenges and poor self-esteem.

Diagnosis

The specialist must study the signs, family background, and medical records to determine MMD. A physical examination will be conducted and multiple tests may be required to determine MMD and any symptoms causing it [15]. Moyamoya diagnosis can be rendered in most cases with a thorough evaluation of an MRA and MRI. Cerebral arteriography can validate the diagnosis, determine the exact degree of restriction of blood vessels, show the current patterns of blood flow to various brain areas, and require medical decisions to be made [16] for these purposes, it is the primary diagnostic tool for this disorder. In fact, catheter angiography can help identify critical blood vessels named "transdural collateral" that are active in some situations and can have a significant impact on surgical preparation and prognosis [17].

Tests may include:

- ✓ **Magnetic resonance imaging (MRI):** An MRI utilizes strong magnets and radio waves to create accurate brain pictures. The physician injects a dye into a blood vessel to see veins and arteries and show the movement of the blood (magnetic resonance angiogram). If appropriate, the doctor can prescribe an MRI of the perfusion. This sort of visualization will determine how often blood flows into the vessels.
- ✓ **Computerized tomography (CT) scanning:** CT scan utilizes a sequence of X-rays to create an accurate brain image. The physician injects a dye into a blood vessel to display blood flow in the veins and arteries (CT angiogram).

- ✓ **Cerebral angiogram:** Introduces a large, narrow tube (catheter) into a blood artery in the groin during a cerebral angiogram and directs it to the brain utilizing X-ray images. Then, the physician injects dye into the blood vessels of the brain via the catheter to make them noticeable under X-ray imaging.
- ✓ **Transcranial Doppler ultrasound:** Sound waves are utilized to obtain images of blood vessels of the brain in a transcranial Doppler ultrasound process. The doctor can utilize this test to collect information about brain blood vessels.
- ✓ **Positron emission tomography (PET) scan or single-photon emission computerized tomography (SPECT):** The physician injects a tiny volume of healthy nuclear substance into these experiments and inserts radiation monitors around the brain. PET offers snapshots of brain function. SPECT monitors blood supply to various brain parts.
- ✓ **Electroencephalogram (EEG):** detects electrical stimulation in the brain by collecting scalp-attached electrodes. Children with MMD also suffer from EEG abnormalities.

Treatment

1) Medications

Medicines can be used to minimize the likelihood of stroke or to better prevent epilepsy, including:

- ✓ **Blood thinners:** Aspirin or some other drugs avoid strokes (for preventing or reducing the development of small blood clots within narrowed vessels)

In extreme circumstances, anticoagulants such as Lovenox or coumadin are given in extremely ill patients with repeated symptoms, although they are seldom recommended as long-term treatments owing to the apparent possibility of brain bleeding in this state.

- ✓ **Calcium channel blockers:** Often known as calcium antagonists, this form of drug can enhance pain symptoms and decrease symptoms due to acute ischemic attacks. Furthermore, calcium channel blockers need to be treated cautiously, because they can often lower blood pressure, which can raise the likelihood of stroke.
- ✓ **Anti-epilepsy:** These can be administered as reported to have an underlying epilepsy condition regardless of a patient or infant.

Revascularization surgery

There is no drug available to interrupt the development of cerebral artery narrowing and the condition can tend to worsen in the vast majority of cases irrespective of care.

Surgical techniques are intended to restore blood flow to the brain by diverting scalp blood supply to the brain surface and thereby counteract the gradual leakage of blood volume from the brain hemisphere. There are several suggested surgical techniques for treating moyamoya and they were categorized into so-called "indirect" and "primary" operations [18].

Direct revascularization procedures

During direct revascularization operation, doctors directly suture (thread) the scalp artery into the brain artery (superficial temporal artery to middle cerebral bypass surgery) to rapidly improve blood supply to the brain. Direct bypass surgery in children may be complicated due to the extent of the blood arteries that need to be removed. Direct revascularization surgery, like stroke, has the possibility of complications.

Indirect revascularization procedures

The aim of indirect revascularization is to slowly improve blood supply to the brain over time. Indirect revascularization may be marginally lower than overt revascularization. Types of indirect revascularization procedures include encephaloduroarteriosynangiosis (EDAS) [19] or encephalomyosynangiosis (EMS), or their combination.

The surgeon divides (dissects) a scalp artery for many inches in encephaloduroarteriosynangiosis (EDAS) and conducts a tiny temporary opening in the skull immediately below the artery and connects (sutures) the preserved scalp artery to the brain surface, which enables blood vessels from the artery to expand into the brain over time. The chirurgist then replaces the bone and closes the skull. In encephalomyosynangiosis (EMS), a muscle in the temple area of the forehead is removed (dissects) and positioned on the surface of the brain by a gap in the skull to help maintain blood supply.

In EMS and in EDAS: Dissects a muscle in the temple area of the forehead in this operation, and then positions it on the surface of the brain after connecting the scalp artery to the surface of the brain. The muscle continues to keep the artery in position over time as blood arteries expand through the brain.

Surgeons can extract a portion of a fatty tissue layer from the abdominal area, position it on the brain surface, and add the scalp artery to the fatty layer blood vessel (omental transplant). Such operation will get back blood supply to the brain. Surgeons may make multiple burr holes in the skull to allow the growth of new blood vessels, either in combination with other procedures or as a separate procedure. People with MMD may develop a blood vessel bulge or ballooning in the brain, known as a brain aneurysm. If this occurs, surgery might be required to avoid or treat a ruptured aneurysm in the brain.

Conclusion

MMD is a chronic, occlusive cerebral vasculature condition with specific involvement of the Willis circle and the arteries that support it. Medicines can be recommended to reduce the possibility of a stroke or to better manage seizures. Long-term outcomes after both forms of operation have been very strong, with long-term stroke reduction shown in both pediatrician and adult patient series reported. Importantly, recent data shows that the most important factor predicting a successful surgical outcome is receiving treatment at a center that annually performs a large number of cases with moyamoya. Genetic counseling can be beneficial for patients and their families if they have an inherited form of MMD. Occupational and physical therapy can help to restore some loss of stroke-caused physical function. Cognitive-behavioral therapy may help address emotional issues, such as how to deal with anxiety and uncertainty regarding future stroke.

Acknowledgment

Nil

Conflict of interest

Nil.

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