Case Report

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Moyamoya Disease

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Abstract

Moya Moya disease is a rare disorder of the carotid arteries - the arteries that supply blood to the brain. Moya Moya disease (MMD) is a slowly progressive bilateral stenocclusive process of the distal internal carotid and proximal portions of the anterior and middle cerebral arteries and the formation of an abnormal vascular network at the base of the brain. All the symptoms of Moyamoya disease arise from reduced blood supply to the brain and/or rupture of the 'Moyamoya vessels'. Reduced blood supply may cause stroke and rupture of the unhealthy Moyamoya vessels causes bleeding within the brain. The diagnosis of moyamoya is suggested by CT, MRI, or angiogram results. There is no cure for this disease. Drugs such as antiplatelet agents (including aspirin) are usually given to prevent clots, but surgery is usually recommended is Burr hole surgery.

Keywords: Moya Moya Disease; Symptoms; Prevention.

Introduction

Moya Moya Disease was first described in Japan by Takeuchi and Shimizu in 1957. Moya moya disease is commonly seen in the Japanese population but it can also occur in other populations. It can present as a pediatric or an adult form, each with varied clinical presentations. Children present with repeated ischemic events and adults present with hemorrhagic events.

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The disease causes progressive occlusion of the circle of Willis, with abnormal dilated collateral vessels on cerebral angiography, which look like "a puff of cigarette smoke" (Moya Moya).

Case Report

Master Ahmad Shaikh, 9 year boy presented to Bai Jerbai Wadia Hospital with the complaints of seizures, recurrent fall while walking, slurred speech and involuntary movements, weakness at left upper limb and lower limb.

MRI report is there is diffuse severe narrowing of supraclinoide segments of bilateral ICA's extending and involving lateral MCA, which are diffusely thines out and narrowed, along with other MR angio findings multiple inyracranial collaterals as describes suggestive of Moyamoya disease.

Cerebral angiogram finding suggestive of Moyamoya disease with high grade narrowing of both supraclinoide ICA's with collateral circulation.

In hospital he has diagnose with Moyamoya Disease. He was treated with anticoagulant and antiplatelate therapy and he had undergone multiple Burrhole (partial, temporal and paritel) indirect revascularization surgery under general aneasthesia.

Prevalence

The prevalence of the disease ranges from 3.2 to 10.5 per 100,000 populations. In general, the disease has been found to be more prevalent among Asians and people of Asian origin. The exact cause of this disease is not known yet. About 57% of the affected patients are Asian and 71% are female. Although the disease may be seen in any age group, it is more common in people from 5-15 years and 30-40 years of age. Family history is present in about 10%-15% of the patients.

In Japan the overall incidence is higher (0.35 per 100,000). In North America, women in the third or fourth decade of life are most often affected, but the condition may also occur during infancy or childhood.

These women frequently experience transient ischemic attacks (TIA), cerebral hemorrhage, or may not experience any symptoms at all. They have a higher risk of recurrent stroke and may be experiencing a distinct underlying pathophysiology compared to patients from Japan.

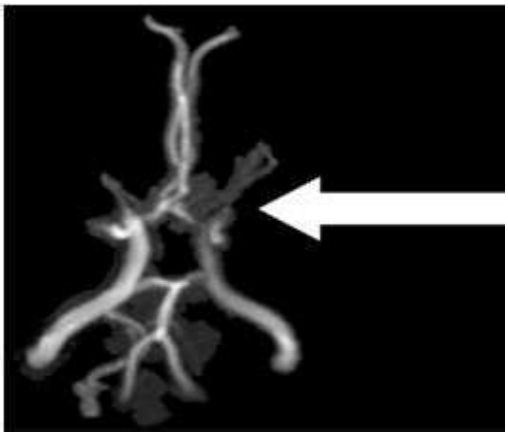

Fig. 1:

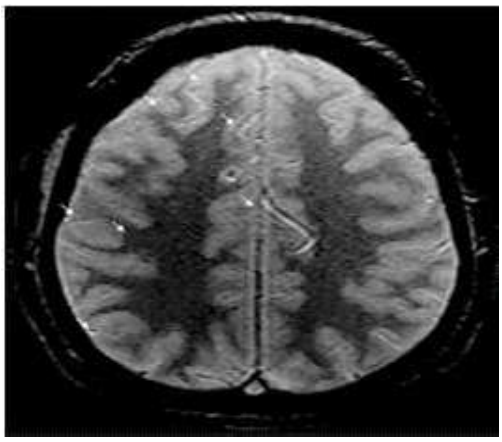

Fig. 2:

Causes

About 10% of cases of Moya Moya disease are familial and some cases result from specific genetic mutations. Susceptibility to Moya Moya disease-2 (MYMY2; 607151) is caused by variation in the RNF213 gene (613768) on chromosome 17q25. Moyamoya disease-5 (MYMY5; 614042) is caused by mutation in the ACTA2 gene (102620) on chromosome 10q23.3; and Moya Moya disease-6 with achalasia (MYMY6; 615750) is caused by mutation in the GUCY1A3 gene (139396) on chromosome 4q32. Loci for the disorder have been mapped to chromosome 3p (MYMY1) and chromosome 8q23 (MYMY3; 608796).

See also MYMY4 (300845), an X-linked recessive syndromic disorder characterized by Moya Moya disease, short stature, hypergonadotropic hypogonadism and facial dimorphism and linked to q 25.3, on chromosome 17".

Moya Moya disease can be either congenital or acquired. Patients with Down syndrome, sickle cell anemia, neurofibromatosis type 1, congenital heart disease, fibromuscular dysplasia, activated protein C resistance or head trauma can develop Moya Moya malformations. It is more common in women than in men, although about a third of those affected are male.

Clinical Manifestation

All the symptoms of Moyamoya disease arise from reduced blood supply to the brain and/or rupture of the 'Moyamoya vessels'. Reduced blood supply may cause stroke and rupture of the unhealthy Moyamoya vessels causes bleeding within the brain. Adults experience hemorrhage more commonly; cerebral ischemic strokes from reduced blood supply are more common in children. Children may have weakness or numbness of an arm or leg, hemiparesis, monoparesis, involuntary movements, headaches, dizziness, or seizures. Mental retardation or persistent neurologic deficits may be present. Adults may have symptoms and signs similar to those in children, but hemorrhage (bleeding) of sudden onset is more common in adults.

Diagnosis

The diagnosis of moyamoya is suggested by CT, MRI, or angiogram results. Contrast-enhanced T1-weighted images are better than FLAIR images for depicting the leptomeningealivy sign in

Moyamoya disease. MRI and MRA should be performed for the diagnosis and follow-up of Moyamoya disease. Diffusion-weighted imaging can also be used for following the clinical course of children with Moyamoya disease, in whom new focal deficits are highly suspicious of new infarcts.

Proliferation of smooth muscle cells in the walls of the Moyamoya affected arteries has been found to be representative of the disease. A study of six autopsies of six patients who died from Moyamoya disease lead to the finding that there is evidence that supports the theory that there is a thickening, or proliferation, of the innermost layer of the vessels affected by Moyamoya. These vessels are the ACA (anterior cerebral artery), MCA (middle cerebral artery), and ICA (internal carotid artery). The occlusion of the ICA results in concomitant diminution of the "puff-of-smoke" collateral's, as they are supplied by the ICA.

Often nuclear medicine studies such as SPECT (single photon emission computerized tomography) are used to demonstrate the decreased blood and oxygen supply to areas of the brain involved with Moyamoya disease. Conventional angiography provided the conclusive diagnosis of Moyamoya disease in most cases and should be performed before any surgical considerations.

Associated Conditions

Many conditions are seen more commonly with Moyamoya like condition, although the exact cause-effect relationship has not been demonstrated. Some of the common conditions are

- Radiotherapy to head and neck
- Down's Syndrome
- Neurofibromatosis type 1
- Sickle cell disease
- Congenital Heart Disease

Treatment

There is no cure for this disease. Drugs such as antiplatelet agents (including aspirin) are usually given to prevent clots, but surgery is usually recommended. Since moyamoya tends to affect only the internal carotid artery and nearby sections of the adjacent anterior and middle cerebral arteries, surgeons can direct other arteries, such as the external carotid artery or the superficial temporal artery to replace its circulation. The arteries are either sewn directly into the brain circulation, or placed on the surface of the brain to reestablish new circulation after a few weeks.

There are many operations that have been developed for the condition, but currently the most favored are the in-direct procedures EDAS, EMS, and multiple burr holes and the direct procedure STA-MCA. Direct superficial temporal artery (STA) to middle cerebral artery (MCA) bypass is considered the treatment of choice, although its efficacy, particularly for hemorrhagic disease, remains uncertain. Multiple burr holes have been used in frontal and parietal lobes with good neovascularisation achieved.

The **EDAS** (encephaloduroarteriosynangiosis) procedure is a synangiosis procedure that requires dissection of a scalp artery over a course of several centimeters and then making a small temporary opening in the skull directly beneath the artery. The artery is then sutured to a branch of the middle cerebral artery on the surface of the brain and the bone is replaced.

In the **EMS** (encephalo-myo-synangiosis) procedure, the temporalis muscle, which is in the temple region of the forehead, is dissected and through an opening in the skull placed onto the surface of the brain.

In the *multiple burr holes* procedure, multiple small holes (burr holes) are placed in the skull to allow for growth of new vessels into the brain from the scalp.

In the STA-MCA procedure, the scalp artery (superficial temporal artery or STA) is directly sutured to an artery on the surface of the brain (middle cerebral artery or MCA). This procedure is also commonly referred to as an EC-IC (External Carotid-Internal Carotid) bypass.

All of these operations have in common the concept of a blood and oxygen "starved" brain reaching out to grasp and develop new and more efficient means of bringing blood to the brain and bypassing the areas of blockage. The modified direct anastomosis and encephalo-myo-arterio-synangiosis play a role in this improvement by increasing cerebral blood flow (CBF) after the operation.

A significant correlation is found between the postoperative effect and the stages of preoperative angiograms. It is crucial for surgery that the anesthesiologist have experience in managing children being treated for moyamoya, as the type of anesthesia they require is very different from the standard anesthetic children get for almost any other type of neurosurgical procedure.

Tips for Family and Caregivers of Patients with Moyamoya Disease

- Educating and supporting the child and family plays a key role. Not surprisingly, parents are shocked and frightened when they learn their children have had strokes, because they do not realize children can have strokes.
- Parents should be instructed to inform surgeons and anesthesiologists to avoid hyperventilation. Parents and children need to identify everyday events that may precipitate mini-strokes (also called Transient ischemic attacks). For example, one patient had a stroke while crying or singing a long note during practice. The precipitation of mini-strokes by everyday events is particularly stressful for parents, who may be reluctant to discipline their child for fear of causing a mini-stroke if the child cries. Some sports such as cricket and soccer that lead to hyperventilation have a high risk of causing mini strokes, and it may not occur to parents to keep their children out of these sports. Schools should be informed about the diagnosis and any restrictions on physical activity.
- Educating the child and family about seizures is an important part of the care of Moyamoya patients. As with other seizure patients, families should be told that brief seizures lasting a few minutes are not thought to be harmful, but medical help should be sought for longer seizures. Emotional support and appropriate advice on pre and post operative care of the patient is an important part of treatment to alleviate the fear, anxiety and uncertainity experienced by the family.

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