

MOYAMOYA DISEASE

Literature review

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ABSTRACT: The term moyamoya (Japanese for 'puff of smoke') refers to the appearance on angiography abnormal vascular collateral networks that develop adjacent to the stenotic vessels. The disease primarily affects children, but it can also occur in adult. In children, the first symptom of moyamoya disease is often stroke or recurrent transient ischemic attack, commonly referred to as mini stroke. Adult most often experience a hemorrhagic stroke due to recurring blood clots in the affected brain vessels. Individuals with this disorder may have disturbed consciousness, speech deficit, sensory and cognitive impairments, involuntary movements and vision problem.

KEY WORDS: Moyamoya , circle of Willis, transient ischemic attack, muscular weakness, consciousness, cognitive impairments, hereditary, stroke.

DEFINITION:

Moyamoya disease is a progressive and occlusive disease of the cerebral vasculature with particular involvement of the circle of Willis and the arteries that feed it.

Moyamoya disease is a disorder caused by blocked arteries at the base of the brain. The name 'moyamoya' describes the appearance of the tangle of tiny vessels that forms to compensate for the blockage.

INCIDENCE:

The incidence of moyamoya disease is highest in Japan. The prevalence and incidence of the disorder, there has been reported to be 3.16 cases and 0.35 cases per 100000 peoples, respectively. Moyamoya disease occurs primarily in Asians, but can also occur in whites, blacks, Haitians and Hispanics. The female to male ratio is 1.8:1.

ETIOLOGY:

The cause of moyamoya disease is not known. The disease is believed to be hereditary. Fukui reported a family history in 10% of patients with the disorder. Mineharu suggested that familial moyamoya disease is autosomal dominant with incomplete penetrance that depends on age and genomic imprinting factors.

PATHOPHYSIOLOGY:

Moyamoya disease is characterized by intimal thickening in the walls of the terminal portions of the internal carotid vessels bilaterally. The proliferating intima may contain lipid deposits. The anterior, middle and posterior cerebral arteries that emanate from the circle of Willis may show varying degree of stenosis or occlusion. This is associated with fibro cellular thickening of the intima, waving of the internal elastic lamina and thickening of the media. Numerous small vascular channels can be seen around the circle of Willis. The pia mater may also have reticular conglomerates of small vessels.

CLINICAL MANIFESTATION:

- Ischemic stroke or haemorrhagic stroke
- Weakness and numbness in arms or legs
- Difficulty in speaking
- Paralysis affecting one side of the body
- Seizures
- Cognitive or learning impairments
- Headache

STAGES OF MOYAMOYA:

The progression of moyamoya follows a typical course and can be classified into stages based on angiography findings.

STAGE I: Narrowing of internal carotid arteries.

STAGE II: Development of moyamoya vessels at the base of the brain.

STAGE III: Intensification of moyamoya vessels and internal carotid artery narrowing (most cases diagnosed at this stage)

STAGE IV: Minimization of moyamoya vessels and increased collateral vessels from the scalp

STAGE V: Reduction of moyamoya vessels and significant internal carotid artery narrowing

STAGE VI: Disappearance of moyamoya vessels, complete blockage of internal carotid arteries and significant collateral vessels from the scalp

DIAGNOSTIC EVALUATION:

- History collection
- Physical examination
- Magnetic resonance imaging
- Angiogram
- Computed tomography perfusion
- Cerebral blood flow studies

MANAGEMENT:

Pharmacologic therapy for moyamoya disease is disappointing. Therapy is primarily directed at complication of the disease. If intracerebral haemorrhage has occurred then management of hypertension is imperative.

In case of severe stroke, intensive care monitoring is indicated, until the patient condition is stabilizes.

If the patient had has ischemic stroke consider anticoagulant or antiplatelet agents.

MEDICATIONS:**HEPARIN:**

Heparin is administered intravenously; the target dose is aimed at maintaining an activated partial thromboplastin time (APTT) of 1.5-2 times control.

WARFARIN:

Which is administered orally, is used if long term anticoagulant is needed? The international normalized ratio (INR) is followed, with a target range of 2-3.

SURGICAL MANAGEMENT:

EDAS Procedure:

The EDAS procedure is a diagnosis procedure that requires dissection of a scalp artery over a course of several inches and then making a small temporary opening in the skull directly beneath the artery. The artery is then sutured to the surface of the brain and the bone replaced.

EMS Procedure:

In the EMS procedure, the temporalis muscle, which is in the temple region of the forehead, is dissected and through an opening in the skull placed on the surface of the brain.

Multiple Burr holes procedure:

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

STA-MCA procedure:

In this procedure, the scalp artery is directly sutured to an artery on the surface of the brain. This procedure is also commonly referred to as an external carotid and internal carotid (EC-IC) bypass.

COMPLICATIONS:

- CSF leaks
- Pseudomeningoceles
- Subdural hygromas
- Myocardial infarction
- Pneumonia
- Deep venous thrombosis
- Pulmonary emboli

REHABILITATION:

Rehabilitation with physical therapy, occupational therapy and speech therapy should be considered, depending on their neurological impairment.

The extent of therapy can range from bedside treatment to full, comprehensive inpatient rehabilitation.

The later would include physical, occupational, speech and cognitive therapy. The condition of the patient, including active comorbidities, dictates his or her involvement in rehabilitation therapy.

CONCLUSION:

In children with this disease, cerebral ischemic events, including stroke occur. In adults, the fragile abnormal vessels can rupture and cause intracerebral haemorrhage. The mortality rate for adult is higher than that for children.

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