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A LITRATURE REVIEW ON: MOYAMOYA DISEASE

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ABSTRACT

Moyamoya disease, a known cause of pediatric stroke is an unremitting is an unremitting cerebrovascular occlusive syndrome of unidentified etiology that can lead to devastating permanent neurological disability if left untreated. Cerebrovascular events are the primary presenting symptoms and are related both to stenosis and occlusion of the ices and their main branches. On cerebral angiography the formation of collateral vessels has the appearance of a puff of smoke (moyamoya in Japanese) which became conspicuous with the refinement with the modern imaging technique. This chronic cerebral antipathy is observed in children and adults. It mainly leads to brain ischemic events in children, and to ischemic and hemorrhagic events in adults. This is a rare condition, with a marked prevalence gradient between Asian countries and western countries. Identification of the genes involved in moyamoya disease and several monogenic moyamoya syndromes unraveled different pathways involved in the development of this angiopathy. Studying genes and pathways involved in monogenic moyamoya syndromes may help to give insights into pathophysiological models and discover potential candidates for medical treatment strategies.

Keywords: Moyamoya diseases.

INTRODUCTION

The syndrome was pronounced in Japan 1957 by “Takeushi and Shimizu” and was termed moya “ Suzuki and Takaku” in 1969.[14] Moyamoya means “a puff of smoke” in Japanese. The reason why the name was given is due to the typical appearance of Collateral blood vessels that develop at the base of the brain and resemble a puff of smoke between western Countries, MMD is exceptionally rare in African Americans; it presumably remains a misdiagnosed cause of ischemic and/or hemorrhagic stroke. MMD has two age peaks: in childhood and in young adults. Worldwide studies showed women are more frequently affected than men.[3,9,16.]

Moyamoya disease, a known cause of pediatric stroke, is an unremitting cerebrovascular occlusive disorder of unknown etiology that can lead to devastating, permanent neurological disability if left untreated.[1] It is associated with increased mortality; smoking and hypertension are modifiable risk factors. About 20% of MA patients, including pediatric and younger MA patients, may exhibit headaches of vascular origin with migraines features; decrease in cerebral blood flow or cerebrovascular reserve and spreading cortical depression are possible pathophysiological mechanisms for these features. The initial efforts to elucidate the genetic makers of MA reflected this complexity, with a wide range of genes, chromosomes and hereditary disease reported to the potential makers.[2]

Unlike what is seen in conditions such as M1 stenosis, in which the lesions affect only one vessels, the hypo perfusion in MMD involves adjacent vascular territories, causing more significant ischemia, which chronically induces the formation of collateral vessels of arterial branches of the ICA and the posterior circulation. The collateral arise mainly from the choroid arteries, although Trans cranial and Trans Dural collaterals are also commonly seen in the external carotid arteries, the ophthalmic artery, and the meningeal artery.[4]

Another important risk factor is previous exposure, in predisposed Person, to cranial radiotherapy to treat brain tumors. In our reality, this disease has a rare incidence, according national studies, mainly in the Non- Asian population.[5] Moyamoya is an intriguing disease and little is known about its pathogenesis.[6] Puff of smoke term we apply to a peculiar angiographic picture consisting of abnormal net-like vessels at the base of the brain.

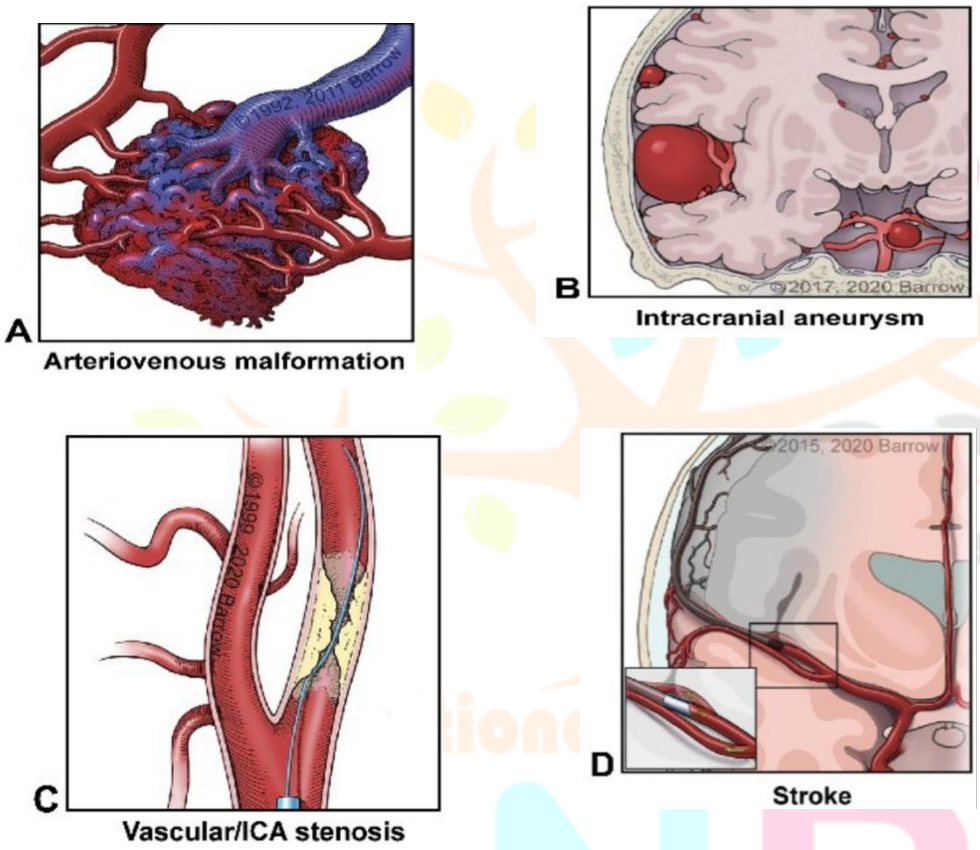
In 1963 we suggested that this pattern constituted a new disease entity and since then, many similar cases have been reported not only in Japan, but from all over the world.[8] The specific treatment to prevent such complications to usually revascularization surgery, associated or not with therapeutics relating to the underlying disease.[11] With the development of noninvasive diagnostic tools such as magnetic resonance angiography (MRA) and other investigations, Moyamoya disease has been increasingly reported from many other regions of the world.[12] Suzuki and Tataku also sub classified the chronological stages of the disease:[13]

1. Narrowing of the carotid siphon
2. Initiation
3. Intensification
4. Minimization
5. Reduction and
6. Damage of Moyamoya Veins.

However, majority of the cases are reported in Asia and other non- Caucasian region. Moyamoya disease has remained rather unexplored in Pakistan. So far only one case series of four patients has been described, while a few case reports exist.[10] It is an intrinsic pathological disease condition where the complete etiology is not

well understood. Because of the contraction of artery, the patient experiences less amount of blood movement to the cerebrum. A smaller amount of blood circulation leads to ischemia, which is not as much of supply of oxygen. The development of fixed-dose combinations (FDCs) is becoming increasingly high either improving compliance or to benefit from the Added effects of the two or more active drugs given together. They are being used in the treatment of a wide range of conditions and are Particularly useful in the management of chronic conditions.[15,16]

PATHOPHYSIOLOGY:



In Moyamoya disease stenotic differences initially look in the ICAs distally at the level of their bifurcation stenotic artery reveal endothelial hyperplasia affected vessel lake inflammatory variations. It is too responsible for vascular tumor etiology of Moyamoya disease. Genetic and environmental factors possible play significant roles in disease process.[1] Different pathogenic mechanism may underline pathologic vessel changes MA and mounting evidence shows that MA and acquired proliferative disease of the itima. Previous studies demonstrate various association of MA with atherosclerotic and autoimmunue disease.[2,8] Their disease are various stages for example: Arterio venous malformation, intracranial aneurysm ,vascular/ICA stenosis, stroke.

The first episode of stroke with areas of restricted diffusion inactive of acute ischemia the right cerebral hemisphere. The second episode with progressive bilateral ischemic changes.[3] The pathogenesis is not clear their vessels appear predisposed to rupture and interior hemorrhage. Commonly seen in adults there are both cerebral and systematic vascular abnormalities.[4] Chronic state of cerebral ischemia .this fact determines the development of extensive network of collateral fraglie vessels that involve the base of brain.[5] The common of pediatric cerebro muscular disease in East Asian countries. The disease are higher among individual especially those of Japanese, Korean and Chinese descent.[6] The development of moyamoya pathology, intimal thickening and media thinning.by affecting vascular endothelial or smooth cell proliferation or migration.[7] Protein cells, genes and signalling pathways related to cerebral angioppathy characteristic.[8]

In MA associated disorders[8]:

1. Arteriovenous Malformation

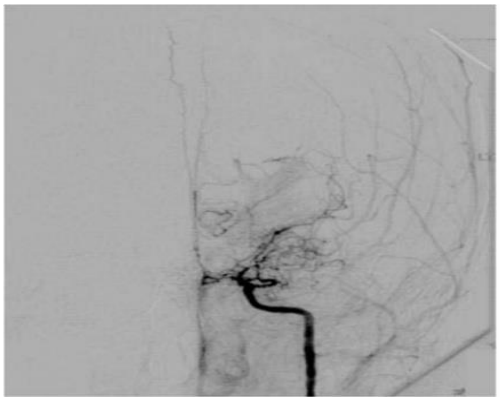
2. Intracranial aneurysm

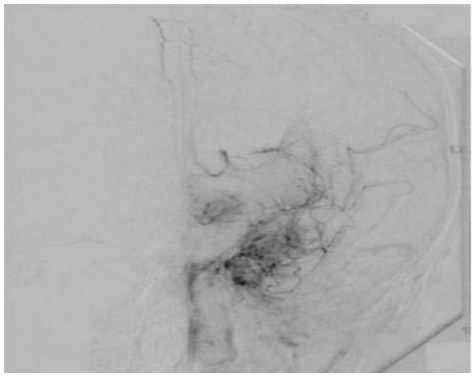
3. Vascular/ internal carotid artery stenosis and

4. Stroke

Fibroblast growth factor has been proposed possible mediator of the neovascular response. A factor involved in angiogenesis and expression of connective tissue genes, was also shown to be elevated in the disease. An unknown CSF protein has been detected in some patients with Moyamoya.[9] Angiogenesis and vasculogenesis- related inflammatory and autoimmune factors seen to be contributing to be pathophysiology of MMD.[10] The most common examples of disease of that can cause moya moya syndrome are sickle cell disease, neuro-fibromatosis type 1, cranial irradiation and down syndrome. Moyamoya disease is bilateral disease.[11]

A linkage study of moyamoya disease using markers on chromosome. Which the HLA gene positioned. Where they causative gene is reported to be located of chromosome.[1,2.] Thickened itima in the intracranial blood vessel. They attributed this pathological development the thrombotic components, such as platelets and plasma constituents.[13] Magnetic response angiogram showing bilateral stenosis suggestive of moyamoya disease. Angiogram showing stenosis of the supraclinoid ICA and MCA consistent with moyamaya disease. Cerebral angiogram showing the “puff of smoke” that comprises the abnormal collaterals.[14]





One such mechanism is that the presence of pro inflammatory cytokines in addition to muted RNF 213 leads to increased angiogenesis. Detected and increased level of hepatocyte growth factor (HGF) in cerebro spinal fluid (CSF) and diseased arteries in patient with MMD.[15] Intake hypothalamus is perfused by reversed flow through these cholesterols. Therefore this defect can result in hypothalamic vascular in sufficiency.[16]

SYMPTOMS:

Symptoms/clinical presentation:-

1. Children by Moyamoya disease classically present with sign of cerebral ischemia.
2. Cerebral infraction
3. Extremity paralysis
4. Crying, coughing and blowing
5. Vasoconstrication of normal blood vessels.
6. Cardiac illness and renal artery stenosis.
7. Infection as tuberculos meningitic and leptospirosis atherosclerosis and fibromuscular dysplasia.[1]

Common-

1. Ischemia
2. Hemorrhage
3. Ischemic attack
4. Stroke

Fewer common-

1. Seizure
2. Headache

Rare-

1. Crying, coughing and even during execution of wind instrument.
2. Clude epileptiform
3. Headache
4. Visual disturbance
5. Paresis
6. Movement disorder progressive cognitive deterioration and psychaitric symptom.
7. Affect the basal ganglia and thalmly[5:]

1. Crebel ischaemia
2. Intracranial hemorrhage
3. Ischemic attack
4. Hemorrhage headache
5. Seizure
6. Cognitive[4]
7. Psudo psychiatric symptom
8. Dehydration
9. Fever
10. Obesity
11. Migraine like headache is also common symptom in MMD
12. Chronic hypoxemia[11]
13. Dysmorphic syndrome

TREATMENT

1. Early diagnosis prompted appropriates the surgical management of at most important.
2. They can main treatment is Aspirin and a calcium channel blocker.
3. Easy bruising bleeding and gastrointestinal irritation aspirin dose adjusted.
4. Calcium channel blocker are to be more effective reducing frequency of severity.[1]

Treatment of MMD depends upon patient clinical presentation and stage of disease. Treatment option includes observation and monitoring medical treatment and surgical treatment.[5]

1. Multiple surgical procedures designed to argument CBF disate. Can treat carotid artery have been successful treating Moyamoya disease.
2. TI As rapidly decreases or disappears and stroke rarely recur.
3. Ischemic symptom recommending surgery.
4. Patient cans the angiographical criteria.

5. SPECT with acetazolamide challenged done to look reduce cerebral prefusion.[1]
6. Direct anastomotic bypass.
7. Although there is currently no specific a therapeutic strategy effective or reversing background vascular abnormality MMD.
8. Used in stroke prophylaxis portably changed in history of a disease.
9. The cornerstone of clinical approach to MMD is prophyleptic use of a antiplatelet drug, anticonvulsant drug.
10. Few direct bypass treatment can the used.
11. As well as a multiple burr hole surgery.[14]

CONCLUSION:

Moyamoya diseases is comparatively rare, is a widely recognized cause of strock in children. The better understanding of the pathophysiology, symptoms and the treatment of the disesaes.

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