



RESEARCH ARTICLE

MOYA MOYA DISEASE

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ABSTRACT

MoyaMoya disease was discovered in 1957 by Japanese surgeon (Takeuchi and Shimizu). It is a rare progressive cerebrovascular disorder that affects the brain blood vessels and the internal carotid arteries become narrowed or blocked. Its most commonly occur in paediatrics and adults. It is a idiopathic vascular disorder of intracranial arteries, the cause is unknown and sometimes it may be due to DNA mutation, the incidence of this disease is high in East Asia and more common in female at birth as compared to male at birth, in children's usually occurs between the age group of 5 to 10 years while in adults occur between the age of 30 to 50 years and with the symptoms of facial asymmetry, seizure, involuntary movement and also experience with the symptoms of ischemic attack and intracranial bleeding. Most common symptoms in paediatric are weakness of extremities, sensory disturbance and headache. It can be diagnosed by MRI, Cerebral angiogram, CT, Tran's cranial Doppler ultrasound and it can be managed with the Revascularization surgery, vasodilator, antiplatelet, antifibrotic and anticonvulsants drugs.

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INTRODUCTION

In Japanese language the name of moyamoya means the "puff of smoke". or (hasu puff smoke) here there are genetic forms of moyamoya of Asian ancestry, but the disease is being diagnosed more and more in patients of all ethnicities. This disease is more common in patients of female at birth as compared to male at birth. It can occur at any age usually in children it occurs between the age of 5 to 10 years while in adults occurs between age of 30 to 50 years. The ministry of health and welfare of Japan has defined 4 types of moyamoyadisease Ischemic -63.4%, Hemorrhagic - 21.6%, Epileptic -7.6%, Others-7.5%. Ischemic type is more predominate in childhood and hemorrhagic type is more in adult's population highest in Japan. Studies have shown a possible genetic association of mmd linked to chromosome 17 Japanese cases as well as found in other demographic. A survey from 2003 report 7700 Japanese were treated for moyamoya disease and almost 100% increases over the 3900 reported in 1994. Studies from 2002 to 2006 states the incidence rate is now up to 0.94 patients per 100000 people with the prevalence of 10.5 patients per 100000. Through this survey the prevalence rate is more in women than men with a female to male ratio of 1.8:1. The highest prevalence for male at the age 10 to 14 years and smaller peak is 35 to 39 years and 55 to 59 years. In female it is highest at 20 to 24 and smaller peak at 52 to 54 years.

DEFINITION

MoyaMoyadisease is a chronic occlusive cerebrovascular disease involving bilateral stenosis or occlusion of the terminal portion of the ICA's and are the proximal portion of the ACAS and MCAS. MoyaMoya disease also termed as bilateral hypoplasia of the ICAS (intracranial arterial stenosis)

CAUSES

The exact cause of disease is unknown, but it's thought to be a combination of genetic factors and other influences. Mutations in the RNF213 gene are linked to some cases, family history, and also associated with neurofibromatosis type 1, sickle cell disease, Graves' disease, atherosclerosis, down syndrome,

CLINICAL FEATURES

In paediatrics the most common symptoms are weakness of extremities, sensory disturbance and headache, cerebral ischemia, IN ADULTS Ischemic attack, intracranial bleeding, Facial asymmetry, Hemiplegia, Difficulty in speaking, Seizure, Involuntary movement. Progressive difficulty in thinking.

DIAGNOSIS

MRI, Cerebralangiogram, Computertomography, Electroencephalogram, Transcranial Doppler ultrasound these are used to detect the picture of brain and blood vessels.

TREATMENT

The MMD can be treated with medically and surgically and anaesthetic management. 77% of cases can be treated with Revascularization surgery, medically can be treated with vasodilator, antiplatelet agents, antifibrotic agents and anticonvulsants drugs.

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