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# Moyamoya Disease in Malaysia : Two Documented Cases

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## Summary

Moyamoya disease is a rare cause of young strokes. The definitive diagnosis of moyamoya disease is made by cerebral angiography. We report two cases of moyamoya disease in Malaysia

*Key Words:* Moyamoya disease

## Introduction

Moyamoya disease (MD) was first described by Takenchi and Shimazu in Japan in 1957. The word moyamoya is a Japanese adjective for a "puff of smoke"<sup>1</sup>. MD is a condition of unknown etiology characterised by typical angiographic findings. Initially, this condition was believed to be peculiar to Japan but now has been reported worldwide. We describe two cases of MD presenting as recurrent cerebrovascular accidents in adolescent

Chinese males and review the epidemiology of MD in East Asia.

## Case Report

### Case 1

A 13-year-old Chinese boy presented with expressive dysphasia and inability to recognise his family members. He was born at term after an eventful pregnancy. At one month of age he had generalized

tonic clonic seizures. He continued to have normal physical development but was cognitively slow. He had poor grades at school. At 10 years of age, he was admitted to University Hospital, Kuala Lumpur for a left hemiparesis. A CT scan done showed an infarct in the right middle cerebral artery territory. Three months before that he had choreiform movement of the left hand. The family subsequently defaulted follow-up.

On examination, he had moderate cognitive deficit and a residual left hemiparesis with a dystonic left hand. The new physical signs were Wernicke's dysphasia and cortical blindness. The examination of the other systems were normal.

The results of routine blood tests including the full blood count, erythrocyte sedimentation rate, arterial blood gas, creatine phosphokinase and electrolytes were normal. A muscle biopsy showed no evidence of mitochondrial cytopathy. Screen for collagen vascular

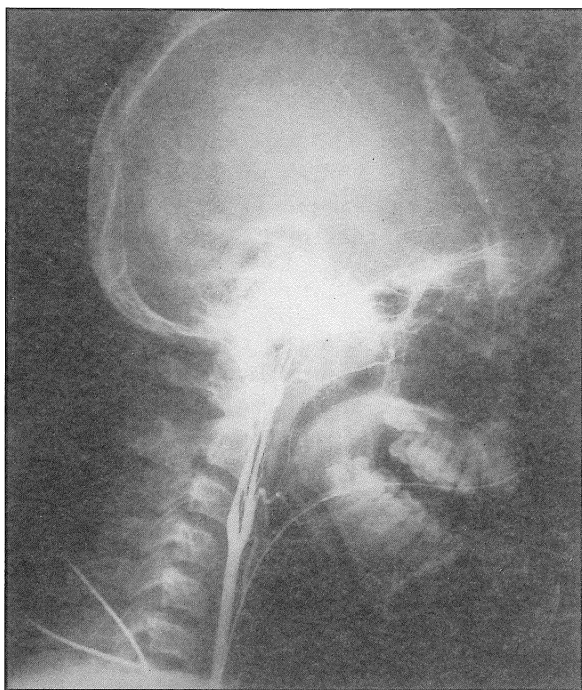
disease was negative. The EEG was consistent with partial epilepsy with secondary generalisation.

The computed tomography of the brain showed an old right parietal – occipital infarct and a recent left parietal – occipital haemorrhagic infarct. The carotid angiogram showed bilateral narrowing and tapering beyond the carotid siphon. Leptomeningeal blush of vessels suggested collateral circulation in the region of the attenuated vessels. These angiographic findings were consistent with a late stage of MD. (Fig. 1)

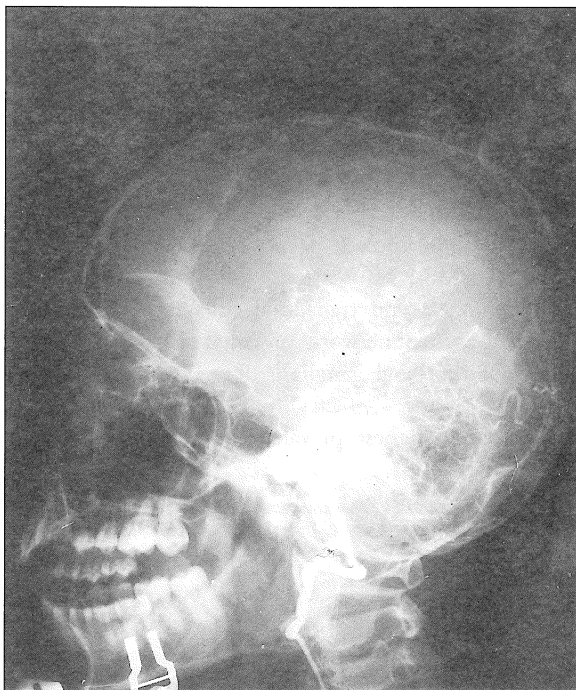
This patient was managed symptomatically with anticonvulsants.

#### Case 2

A 14-year-old boy who was previously well presented with recurrent transient ischaemic attacks (TIA) over a period of three months in both middle cerebral artery territories. These consisted of transient alternating hemiparesis associated with dysphasia if the



**Fig. 1:** Lateral view of the carotid angiogram showing angiographic features of moyamoya disease



**Fig. 2:** Lateral view of a vertebral angiogram showing the "puff of smoke" of moyamoya disease

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dominant side was involved. Hyperventilation when the child consumed spicy food often provoked an attack. The family history was not significant.

On examination, he had a left homonymous hemianopia and left sensory inattention. He was normotensive and the examination of the other systems were unremarkable. Routine blood investigations for a cause of thrombophilia and a hypercoagulable state were negative. The computed tomography of the brain showed a right parieto-occipital infarct with haemorrhagic transformation. The magnetic resonance imaging of the brain showed multiple, small, tortuous, low intensity areas in the basal ganglia representing an abnormal network of parenchymal vessels.

The cerebral angiogram demonstrated bilateral total occlusion of the distal internal carotid arteries and the typical blush of leptomeningeal network of collateral vessels resembling "a puff of smoke". These angiographic findings were diagnostic of MD. (Fig. 2)

This patient was given an antiplatelet drug for the TIA's and is being planned for encephalomyosynangiosis in Japan.

### Discussion

The antemortem diagnosis of MD is made by the presence of pathognomonic angiographic findings. These features include stenosis or occlusion of the internal carotid bifurcation, abnormal parenchymal, leptomeningeal and transdural collateral accompanied by an abnormal network of collateral vessels in the region of the basal ganglia<sup>3</sup>.

MD has the highest prevalence in Japan with 3,300

reported cases up to 1990. Outside of Japan a majority of the cases of MD occurred in East Asia<sup>1</sup>.

We believe that our patients are the third and fourth documented cases in Malaysia. It has been reported that the annual incidence of MD in Japan is 0.08 per 100,000 population<sup>2</sup>. MD has been reported in other continents. There is a marked preponderance of MD in the Chinese, who share some ethnic similarities to the Japanese. In Singapore which is a multiracial country, 38 of 40 cases were Chinese<sup>3</sup>.

The etiology of MD is unknown. The ongoing debate is whether the etiology of the disease is congenital or acquired. The familial incidence of 7-12% in Japan suggest a genetic predisposition to develop MD. Human leucocyte antigen (HLA) studies show that HLA - B40 and HLA - B54 are associated with an increased risk of developing MD in children and adults respectively<sup>1</sup>. The two cases of MD described here were of Chinese background consistent with a known genetic predisposition to MD.

The etiology of MD is most likely acquired in a genetically predisposed individual. Most young patients with unexplained strokes in Malaysia undergo a cerebral angiogram. The wide variation of incidence among different countries in East Asia suggest a possible environmental role in the etiology of MD since most countries in East Asia have ethnic similarities.

In summary, MD is a rare cerebrovascular disease causing young strokes in Malaysia. The two reported cases support the need for a cerebral angiogram in the definitive diagnosis of young strokes.

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