

"A CASE REPORT OF MOYAMOYA DISEASE"

By S. T. Khoo,* G. Lim† and H. Y. Lee*

SYNOPSIS

The radiological features of 'Moyamoya Disease' are described with reference to a Chinese lady who presented with subarachnoid haemorrhage.

INTRODUCTION

In the majority of cases, subarachnoid haemorrhage (SAH) is caused by bleeding from an arteriovenous malformation or an aneurysm, other causes being rare. We wish to report a case of SAH in a Chinese patient associated with the rare and little known entity of 'Moyamoya Disease'.

CASE REPORT

The patient, a 54 year old Chinese lady, presented with an abrupt attack of dizziness followed by loss of consciousness lasting about 1 hour on the day of, and just prior to, admission. Physical examination showed terminal neck rigidity, early papilloedema, a pulse rate of 92 beats per minute and a blood pressure of 230/100 mm.Hg. There were no other significant neurological or systemic findings. Lumbar puncture revealed evenly blood-stained cerebrospinal fluid consistent with SAH.

Total cerebral angiography was carried out. The right carotid angiogram (Figs. 1 and 2) showed stenosis of the terminal portion of the internal carotid artery and the adjacent portions of its anterior and middle cerebral branches ("carotid fork" stenosis); this was associated with an abnormal network of fine vessels in the region of the basal ganglia. The left carotid and the vertebral angiograms were normal. The above findings were felt to be compatible with a diagnosis of 'Moyamoya disease'.

The patient was treated conservatively and her recovery from the episode of SAH was

uneventful. The papilloedema resolved and the blood pressure settled without hypotensive treatment to a basal level of 130/80 mm.Hg.

DISCUSSION

The diagnosis of Moyamoya disease, like that of other causes of SAH, is made by cerebral angiography and is based on the findings of carotid fork stenosis and 'moyamoya' vessels in the basal ganglia (Nishimoto and Takeuchi 1972). The word 'moyamoya' is of Japanese origin and is descriptive of something hazy such as a puff of smoke (Suzuki, Takaku and Fukusawa, 1966).

This disease occurs predominantly in the Japanese race (Nishimoto and Takeuchi 1968, Kudo 1968) although in recent years, similar cases have been reported in other races (Taveras 1969, Solomon *et al*, 1970). Nishimoto and Takeuchi (1972) collected together 111 cases from the world literature and summarised the salient features of this disease. The first manifestations appear most frequently during childhood and early adult life and there is a female sex preponderance. Younger patients tend to present with monoplegia or hemiplegia whilst older ones more commonly suffer from SAH. The long-term outlook is poor even if recovery from the initial episode is complete (as in our patient); the tendency is for the attacks to recur.

The aetiology of this disease and the reason for its peculiar racial bias remain obscure. Two theories have been put forward to explain the angiographic appearances. One school of thought believes that the 'moyamoya' vessels represent a true congenital vascular malformation whilst the other postulates that they merely represent collateral vessels consequent to stenosis of the carotid arteries. Evidence in support of one or other theory have been reviewed by Nishimoto and Takeuchi (1972).

*Medical Unit I, Outram Road General Hospital, Singapore 3.

†Department of Radiology, Tan Tock Seng Hospital, Singapore 11.

*S.T. KHOO,

†G. LIM,

*H.Y. LEE, (Present address: Department of Medicine, University of Manchester, U.K.)



Fig. 1a.

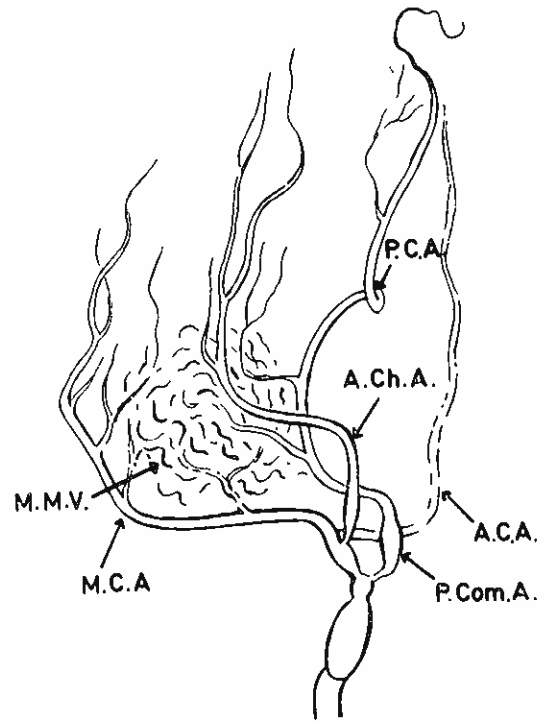


Fig. 1b.

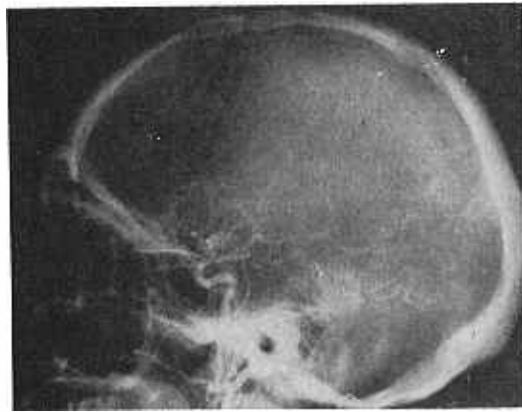


Fig. 2a.

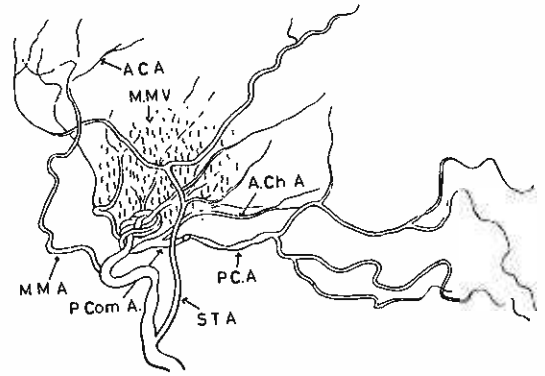


Fig. 2b.

Figures 1 and 2. Right carotid angiogram, frontal and lateral views respectively, showing carotid fork stenosis and 'Moyamoya' vessels in the region of the basal ganglia.

A.C.A. = Anterior cerebral artery
 A.Ch.A. = Anterior Chorioidal artery
 M.C.A. = Middle cerebral artery
 M.M.A. = Middle meningeal artery

M.M.V. = 'Moyamoya' vessels
 P.C.A. = Posterior cerebral artery
 P.Com.A. = Posterior communicating artery
 S.T.A. = Superficial temporal artery.

ACKNOWLEDGEMENTS

We are grateful to Mr. H.Y. Heng for help with the illustrations and to Mr. S.L. Fung for photographic assistance. We thank Prof. P.K. Wong for permission to report this case.

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