# Moyamoya disease presenting as an intraventricular haemorrhage in an adult African man

**E Silber**
*MBBCh, FCP(Neuro)SA*

**P Szkup***
*MD*

**D Saffer**
*FRCP*

*Department of Neurology and *Department of Radiology, Baragwanath Hospital and University of the Witwatersrand.*

## Abstract

Moyamoya disease is a chronic non-inflammatory arteriopathy affecting vessels of the central nervous system, described rarely in non-Oriental populations. We document this condition in a middle-aged African male with typical clinical and angiographic features and review the current literature.

## Case report

A 37 year old African man presented with sudden onset of severe headache, and associated photophobia and neck pain for four days previously. He described a similar event a year previously which had resolved spontaneously. There was no significant family history or history of trauma, hypertension or symptoms suggestive of a vasculitic process.

Examination revealed a normotensive, pyrexial (T=38°C) man with no dysmorphic features, evidence of trauma or sites of infection. He was drowsy, disoriented but rousable, with a Glasgow Coma Score of 14/15. Examination of his cranial nerves, motor and sensory system, cerebellum and fundi revealed no abnormalities. He had significant meningism with positive Kernig and Brudzinski's signs.

Laboratory investigations showed a normal full blood count and differential count, urea, electrolytes and bleeding times. The ESR was 5 mm after one hour. Serology for anti-nuclear factor, rheumatoid factor, syphilis (RPR/TPHA) and HIV ELISA were negative.

Non-contrast enhanced axial CT revealed intraventricular haemorrhage into the lateral, third and fourth ventricles and subarachnoid blood in the basal cisterns (Figure 1). A small amount of intracerebral blood in the area of the left caudate nucleus and genu of the internal capsule was detected.

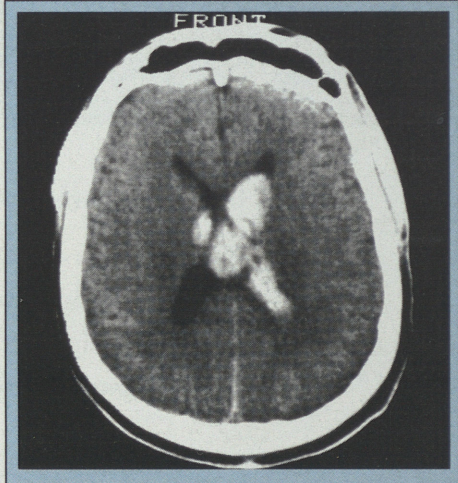


*Figure 1: Noncontrast axial CT showing intraventricular haemorrhage in lateral ventricles.*

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A lumbar puncture was not performed on admission, but one done approximately one month later to exclude a chronic meningitic process was normal.

Management for a subarachnoid haemorrhage was initiated. He was treated with oral Nimodipine, analgesia and bedrest. He made an uneventful recovery and is now asymptomatic.

Four-vessel cerebral angiography revealed bilateral occlusions of the proximal middle (MCA) and left anterior cerebral (ACA) arteries. Multiple leptomeningeal collateral vessels joined the left posterior cerebral artery (PCA) to the left MCA, left ophthalmic artery to the ACA, and the right PCA to the right MCA. Prominent penetrating thalamostriate and lenticulostriate arteries were visualised (Figures 2 and 3).

External carotid angiography showed multiple transdural collateral channels predominantly between the right superficial temporal, right middle meningeal and right middle cerebral arteries (Figure 4). Extracranial angiography revealed a normal aorta, common carotid, vertebral, renal, superior mesenteric and coeliac arteries.

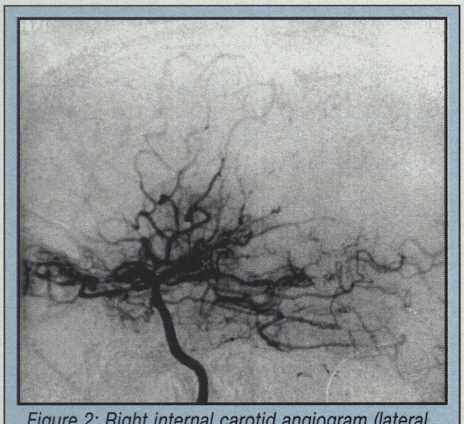

*Figure 2: Right internal carotid angiogram (lateral projection) demonstrating occlusion of the right anterior cerebral artery, partial occlusion of the middle cerebral artery and prominent "moyamoya" thalamostriate perforating branches.*

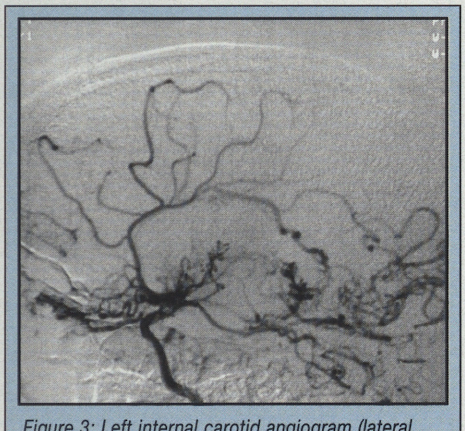

*Figure 3: Left internal carotid angiogram (lateral projection) demonstrating occlusion of the left middle cerebral artery and prominent leptomeningeal collaterals to the posterior circulation.*

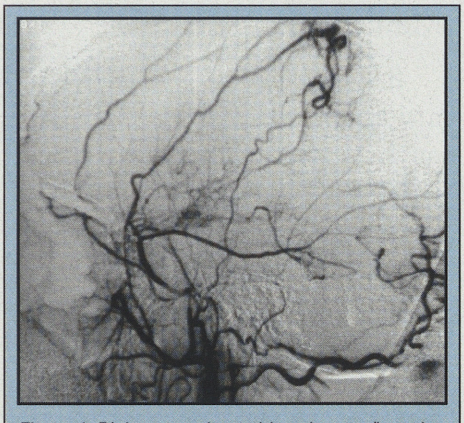

*Figure 4: Right external carotid angiogram (lateral projection) demonstrating dural anastomotic vessels from the middle meningeal and occipital arteries.*

## Discussion

These angiographic findings are typical of Moyamoya disease. This condition was first described in the Japanese literature in 1957.[1] Suzuki and Tahaku[2] first used the term "Moyamoya", the Japanese for "smoky, puffy or vague" to describe the hazy "puff of smoke" appearance of the abnormal capillaries at the base of the skull as seen on cerebral angiography.

### Pathology

The pathological change is a non-inflammatory and non-atheromatous arteriopathy characterised by thickening of the intima with a wavy duplicated elastic lamina. The disease process affects arteries of the Circle of Willis, although renal artery abnormalities have occasionally been described.[3] Stenosis or occlusion occurs bilaterally at the distal internal carotid artery and the proximal anterior and middle cerebral arteries. As a result, numerous small or medium-sized muscular arteries branch from the Circle of Willis forming anastomoses on the cerebral surface and dilated, fragile perforating arteries penetrate the brain parenchyma. These abnormal vascular networks seen in the arterial phase of the angiogram as the "Moyamoya" pattern are thus a compensatory process. Transdural anastomoses from the ophthalmic, external carotid and vertebral arteries are seen pathologically and angiographically. Although posterior circulation occlusion is not included in the diagnostic criteria of Moyamoya disease,[3] a recent report has shown that in a series of 76 patients, 43% of posterior cerebral arteries were either stenosed or occluded.[4]

Angiography is also useful for following disease progression. Early disease stages show progressive vessel occlusion and the development of the network of collaterals. Later in the disease process prominent transdural anastomoses occur with gradual disappearance of the moyamoya network.

CT and MRI may be normal or may show cortical atrophy, ventriculomegaly, white matter low densities or haemorrhage. MRI angiography is gaining ground as a useful screening and follow up modality.[5]

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## Epidemiology

This is an uncommon disease, with an estimated incidence in Japan of 0.08/100 000 population/year. It has a female preponderance of approximately 60%.[3] Its incidence in non-Oriental populations is considerably lower.

We have found three documented cases with classical angiographic findings occurring in patients of African origin: a 37 year old woman of South African origin who presented with repeated left cortical infarctions resulting in an expressive aphasia, right hemiparesis and progressive cognitive decline;[6] a 50 year old Afro-American man presenting as a subarachnoid haemorrhage;[7] and a 16 year old Afro-American woman who presented with seizures and right hemiparesis.[8]

The aetiology is unknown. Genetic factors have been suggested because of a higher familial incidence[9] and a higher incidence in patients with Down's syndrome.[10] A similar angiographic pattern may be seen in patients with neurofibromatosis, arteriosclerosis, meningitis, neoplasia, trauma and following cerebral irradiation.

## Clinical presentations

The disease has two peak ages of onset, in the first and fourth decades, with differing clinical presentations. Young patients usually present with ischaemic complications. These are either recurrent transient ischaemic attacks or completed strokes. They are usually precipitated by straining or hyperventilation and may result in motor, sensory, language or cognitive deficits. Seizures are another common presentation. In the fourth decade, patients usually present with haemorrhage caused by rupture of the fragile penetrating collaterals. Haemorrhage is either intraventricular or intracerebral into the thalamus or basal ganglia, but primary subarachnoid haemorrhage (SAH) is rare.[11] SAH usually occurs from rupture of a co-existing aneurysm present in up to 3% of patients.[4] The prognosis in older patients is worse with an approximate 10% mortality.[1]

## Management

Because the aetiology is unknown, management is symptomatic. No medical therapy, including vasodilators, anti-coagulants, steroids, low molecular weight dextran or anti-convulsants have been shown to be of benefit.[3] Surgery attempts to prevent ischaemic complications by establishing a collateral circulation by means of external to internal arterial anastomosis. Commonly performed procedures are superficial temporal to middle cerebral artery anastomosis, encephalomyosynangiosis (suturing temporalis muscle to a dural defect) or encephalodurosynangiosis (suturing superficial temporal artery to a dural defect) in an attempt to develop neovascularisation.[1]

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