J HK Coll Radiol. 2010;12:182-5

CASE REPORT

Unilateral Arteriovenous Malformation Associated with Moyamoya Disease

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ABSTRACT

The presence of moyamoya disease with a brain arteriovenous malformation is extremely rare. The high incidence of morbidity in such cases suggests that this combination of features confers an ominous outlook, whilst the association between these two entities remains unclear. We present a case of acute intracerebral haemorrhage in a patient with unilateral moyamoya and arteriovenous malformation. We also discuss the possible pathogenesis of these two different vascular diseases occurring in combination.

Key Words: *Arterial occlusive diseases; Carotid artery diseases; Intracranial arteriovenous malformations; Moyamoya disease*

中文摘要

單側動靜脈畸形合併Moyamoya病

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Moyamoya病患者同時出現腦動靜脈畸形的情況非常罕見。直至現時為止仍然未清楚這兩種病之間的關係。患者的罹病率很高，出現的病徵組合令患者康復機會不大。本文報告一名患有單側Moyamoya病與腦動靜脈畸形的病人出現急性腦內出血，並討論這兩種血管病的可能致病機理。

INTRODUCTION

Although there appears to be an association between moyamoya disease and arterial anomalies like arteriovenous malformations (AVMs) and aneurysms, the coexistence of both entities is relatively rare. There are at least 16 cases reported in the current literature, 13 of which had bilateral moyamoya disease.[1-4] Apart from our case, the 3 others had unilateral moyamoya disease with an AVM (2 in adults and 1 in a child). The high incidence of reported morbidity in these cases suggests that the presence of moyamoya with a brain AVM is very ominous. Although the association between these two entities is still unclear, we nevertheless discuss the possible pathogenesis of these two different vascular diseases occuring in combination.

CASE REPORT

A 52-year-old man presented with a 5-day history of sudden-onset headache in June 2007. He had a history of hypertension and was taking anti-hypertensive medication. Physical and neurological examination yielded no abnormal results. Plain computed tomography of the brain (Figure 1) revealed a small left basal ganglia acute intraparenchymal bleed extending into the ventricles. There was mild midline shift towards the right with early hydrocephalus. Foci of calcification were also noted in the left lentiform nucleus. Magnetic resonance imaging of the brain (Figure 2) and magnetic resonance

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Submitted: 30 Jul 2009; Accepted: 16 Nov 2009.

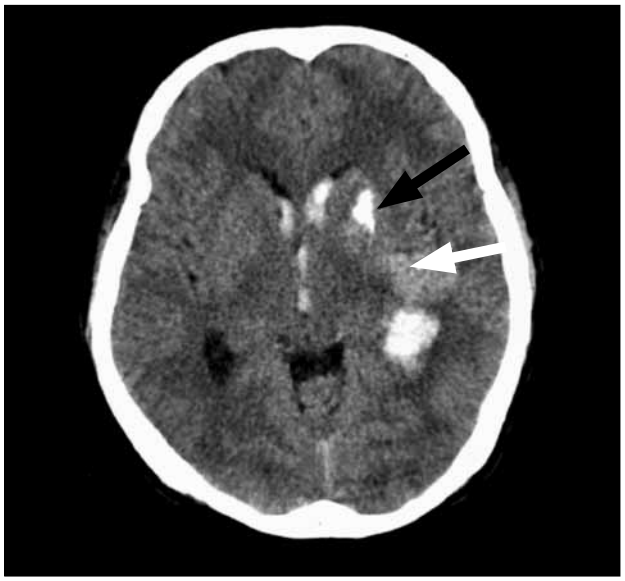

Figure 1. Plain computed tomographic scan of the brain showing small left basal ganglia (black arrow) and intraventricular haemorrhage with foci of calcifications in the left lentiform nucleus (white arrow).

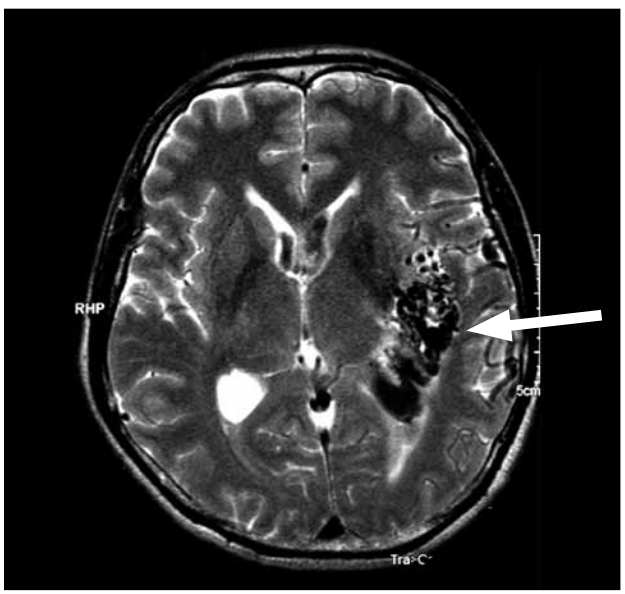

Figure 2. T2-weighted magnetic resonance image (spin-echo, TR 4700 ms, TE 100 ms) showing serpinginous tangle of vessels in the left basal ganglia (arrow) in keeping with a large arteriovenous malformation.

angiography (MRA) revealed a large deep-seated AVM in the region of the left insula extending into the left basal ganglia. MRA revealed a stenosis at the mid M1 segment of the left middle cerebral artery.

Cerebral angiogram (Figures 3 and 4) performed a few days later confirmed a large AVM on the left side. The feeding arteries were from the branches of the left posterior cerebral artery and the proximal branches of the left middle cerebral artery. There were multiple large veins seen to be draining into the deep venous system. There was occlusion of the mid M1 segment of the

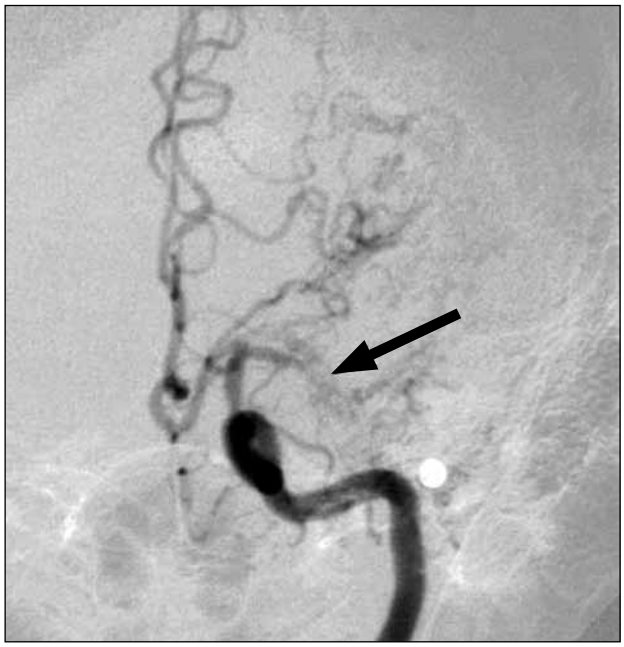

Figure 3. Cerebral angiogram of the left carotid artery demonstrating occlusion of the mid M1 segment of the left middle cerebral artery with multiple tiny collaterals giving a 'puff of smoke' appearance (arrow).

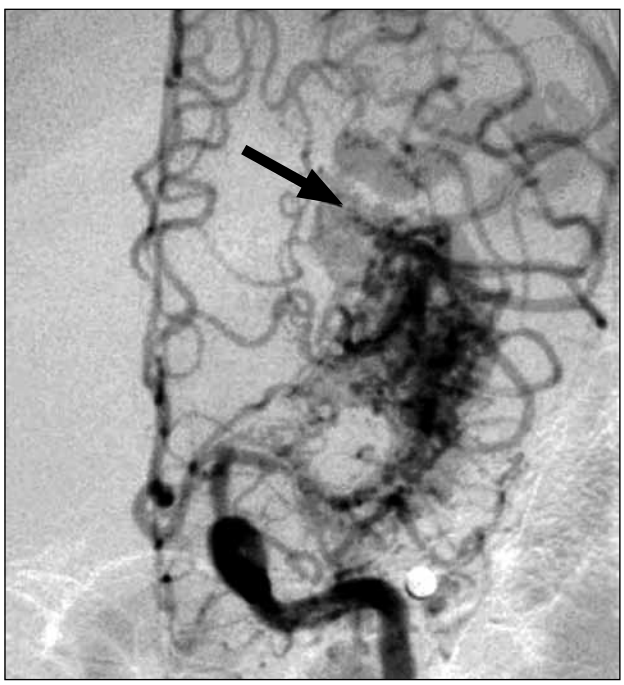

Figure 4. More delayed images show an arteriovenous malformation (arrow) involving the left basal ganglia, fed by the moyamoya vessels and draining into the cortical veins.

left middle cerebral artery and multiple tiny collaterals giving a 'puff of smoke' appearance. The right-sided cerebral angiogram was normal.

The patient was managed conservatively in the ward with antiepileptic medication (phenytoin) and analgesics. Various treatment options involving surgical excision as well as radiosurgery (to shrink the AVM) before surgery were discussed with the patient.

DISCUSSION

Moyamoya is a progressive occlusive vasculopathy mainly affecting the end-arterial vascular bed. It is the Japanese term for 'cloud of smoke drifting in the air', and was first described 50 years ago by Kudo et al.[5] The disease typically involved the clinoid portions of both internal carotid arteries, with formation of collateral vascular networks, known as 'moyamoya' vessels. The latter give a characteristic 'puff of smoke' angiographic appearance.

Moyamoya vessels are thought to represent collateral pathways resulting from an impaired flow in the circle of Willis.[5] As regards causative factors, moyamoya disease may be divided into 2 groups. In the first group that may be termed secondary, there are clear causative factors, such as basal or tuberculous meningitis, irradiation, or atherosclerosis. In the second group termed primary moyamoya, the pathogenetic factors are unknown.

Cerebral AVM is a congenital abnormality consisting of a nidus of abnormal dilated tortuous arteries and veins with a tangle of pathological vessels resulting in shunting of blood from the arterial to venous side, without an intermediary capillary bed. It is accompanied by a variety of dysplasias involving the feeding arteries and draining veins. Flow-related arterial aneurysms as well as venous occlusions or stenoses confer an increased risk of haemorrhage.[5]

The combined presence of moyamoya disease with an AVM is extremely rare. To date, only 16 such cases have been reported in the literature.[1-4] Eleven of the patients presented with symptoms of cerebral ischaemia and remaining 5 with basal ganglia bleed. Owing to the high incidence of subsequent morbidity, we propose that the presence of moyamoya in a brain with an AVM be regarded as very ominous. This view is contrary to that of Mawad and colleagues[1] who suggested that an arterial stenosis proximal to the AVM serves as a built-in mechanism against cerebral haemorrhage from it by reducing blood flow to the nidus.

In 13 out of the 16 patients with moyamoya disease, cerebral angiography demonstrated bilateral involvement of the internal carotid arteries.[1-4] The association between moyamoya disease and AVM remains unclear, there being no satisfactory explanation as to why it is so rare. It is also possible that when both abnormalities manifest in one patient they could be independent vascular entities coincidentally co-existing, as some of the AVMs have been reported not to be supplied by moyamoya collateral vessels but by normal cerebral arteries.[1]

Opinions as to the aetiological relationship between the formation of AVMs and moyamoya vessels are divergent. Although these 2 diseases may co-exist in one patient by chance, some authors suggest a causal relationship. Enam and Malik[6] reviewed 500 patients with AVMs and found that 7 patients had occlusion of one or more major arteries feeding the nidus. The authors postulated that high-velocity turbulence flow caused arterial obstruction and that a large AVM tended to create more turbulence in the feeding vessels. Some authors suggested that in the presence of AVM, there might be increased blood flow and turbulence at the internal carotid bifurcation producing intimal hyperplasia of the vessels and occlusion. Apart from the haemodynamic stress, the authors also suggested that angiogenesis was responsible for proliferation of new capillaries seen with moyamoya phenomenon.[1] The angiogenesis might be induced by the release of angiogenic substances such as transforming growth factors released by AVMs.

Another theory proposed by Lichtor and Mullan[2] was that the angiogenesis process in moyamoya disease is the cause of AVMs. They suggested that in response to the ischaemic changes ensuing in moyamoya disease, perforating vessels and end capillaries become distended causing increased flow into normal draining veins, thus producing the appearance of an AVM. In support of the theory that AVMs appear secondary to moyamoya, Schmit et al[4] described a patient with moyamoya not having surgery and demonstrated the development of an AVM by repeated angiograms performed over a 9-year period.

Our case was unusual not only because of the co-existence of moyamoya disease and an AVM, but also because the moyamoya disease was unilateral. There were no other regions of stenosis to suggest arterioscle-

rosis, and the patient had no history of hyperlipidaemia. There have been a few reports of unilateral moyamoya disease in the literature, both in adults and children. Infrequently there was progression to the typical bilateral form. Young children, however, have a higher tendency to develop contralateral involvement over the years.[7] To our knowledge, there have been 3 previous cases of unilateral moyamoya disease and AVM reported in the literature.[1-4] Suggested causes included: tuberculous meningitis, arteriosclerosis or irradiation,[8] none of which were present in our patient. In most cases, the cause remains undetermined.

There is controversy as to how patients with AVMs occurring with moyamoya disease should be managed. Surgical resection of the AVM risks interruption of delicate collateral channels to adjacent ischaemic brain tissue. Radiosurgery (gamma knife) has been successfully used in the treatment for AVM with moyamoya disease.[3] In our case, the patient opted for radiosurgery and we plan to repeat the cerebral angiogram a year after radiosurgery.

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