

Progression of Arteriovenous Malformation in Moyamoya Syndrome

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Summary

We report a case of moyamoya disease (MMD) associated with arteriovenous malformation (AVM). The 30-year-old female patient presented with left-hemispheric transient ischaemic attacks (TIAs) involving dysphasia and right-sided hemiparesis. CT-scan and lumbar puncture showed no evidence of intracranial haemorrhage. Cerebral angiography revealed typical moyamoya vessels and occlusion of multiple cerebral arteries with consecutive collateral blood supply. Moreover, a left-parietal AVM with a diameter of approximately 2 cm was detected. An extra-intracranial arterial bypass (EIAB) connecting the left superficial temporal artery (STA) with a cortical branch of the left middle cerebral artery (MCA) was performed (STA-MCA anastomosis) and yielded subsequent resolution of the neurological deficit. Nine months post-operatively neurological deficits similar to those of the initial presentation recurred. Repeated angiography suggested comparatively increased AVM blood flow, and successful extirpation of the AVM gradually re-established almost full functional ability. However, deterioration of the neurological condition developed again. We herewith present the first European case of moyamoya disease associated with arteriovenous malformation and report the clinical course under an alternative neurosurgical treatment consisting of STA-MCA anastomosis and delayed extirpation of the AVM.

Keywords: Moyamoya disease; cerebral ischaemia; extra-intracranial arterial bypass; extirpation of arteriovenous malformation.

Introduction

Moyamoya disease is rarely found in conjunction with cerebral AVM, and few cases have been reported in the literature [1, 4, 5, 10, 11, 15]. While Australia [11], the United States [5, 12] and Japan [1, 4, 10, 15] have described one, four and four cases, respectively, no cases have thus far been reported from other countries. In eight of these cases the AVM was managed conservatively [1, 4, 5, 11, 12, 15], whereas the ninth patient underwent simultaneous extirpation of the AVM and prophylactic encephalo-aponeurotic synangiosis [10]. Apart from these cases, there has been a radiological study [8] describing the angio-architec-

ture of cerebral AVM associated with occlusive vascular disease in 13 patients, nine of whom showing collaterals of various types.

The aim of this report is to contribute to the clinical understanding of this rare combined disease and to briefly review pertinent literature.

Case Report

A 30-year-old woman was admitted to our hospital on January 10, 1992 with sudden development of left-occipital cephalgia, nausea and hypaesthesia of the II. through V. finger on the right. Dysaesthesia of the right-sided fingertips and cheek was also present. The past medical history was significant for episodes of left-sided visual disturbances, aphasia, and intermittent paresis of the right arm. Physical and neurological examination was normal except for mild hemiparesis and dysdiadochokinesis on the right.

A plain CT-scan revealed a hypodensity in the area of the left caput nuclei caudatus and narrow basal cisterns but was otherwise unremarkable. While evoked potentials and electroencephalography detected no impressive functional abnormality, transcranial Doppler (TCD) demonstrated severe bilateral flow acceleration which was not clearly attributable to distinct arteries. However, the abnormal signals were felt most likely related to the left MCA and the right posterior cerebral artery (PCA). Increased flow velocity was also present in the basilar and right vertebral artery. Flow accelerations evoked by acetazolamide (ACZ) challenge were abnormally low or absent, and the flow velocity even dropped about 15% in the distribution of the left MCA. On January 20, 1992 detailed cerebral angiography disclosed a large caliber right vertebral artery with remarkably enlarged bilateral PCA and posterior choroid arteries. Anastomosis was present between the vertebro-basilar and supratentorial circulation. Carotid angiography demonstrated an extraordinarily large caliber of both external carotid arteries and their branches as well as proximal occlusion of both anterior cerebral arteries and distal occlusion of both MCA. A cork-screw-like altered vessel convolution mainly originating from the left MCA also showed blood supply from extradurally located anastomosis. Capillary network-like anastomoses involving both lenticulostriate arteries were traceable in the basal ganglia regions bilaterally. Left external carotid angiogram revealed a prominent transdural anastomosis between the left middle meningeal and left anterior cerebral

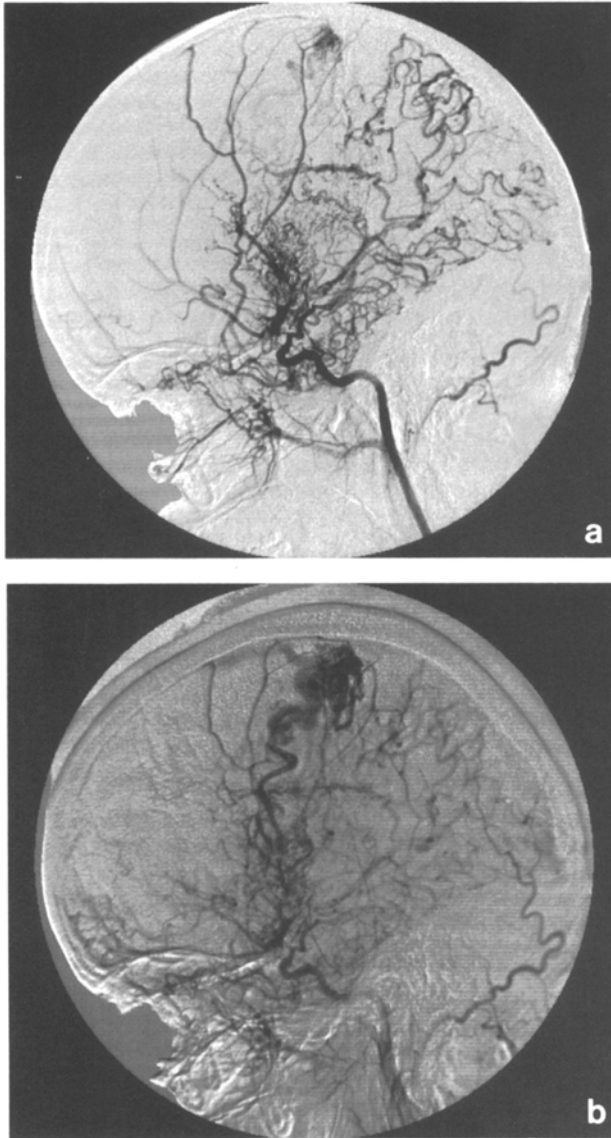


Fig. 1. (a) Left carotid angiogram (January 20, 1992) before left STA-MCA anastomosis demonstrating the AVM in 12 o'clock position and the cork-screw-like altered vessel convolution characteristic for MMD. (b) Left carotid angiogram (January 27, 1993) after EIAB surgery and before extirpation of the AVM suggesting comparatively increased blood flow despite a slight difference between the angiographical phases

artery; an additional transdural anastomosis between the left maxillary and left ophthalmic artery was also demonstrated. Moreover, this study disclosed a left-sided AVM in parieto-paramedio-post-central projection with a diameter of approximately 2 cm and enlarged venous drainers (Fig. 1 a). The site of this lesion was confirmed by cerebral MRI on January 24, 1992 (Fig. 2).

On February 25, 1992 the patient underwent EIAB surgery establishing a left-sided STA-MCA anastomosis. Postoperatively, the neurological deficits gradually resolved, and the patient remained free of complaints over a period of nine months when the

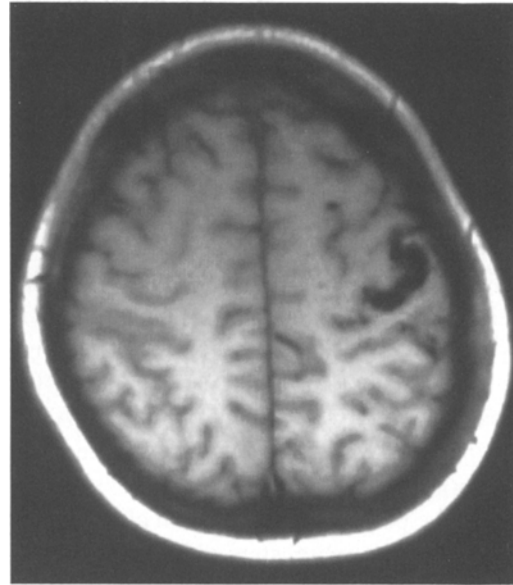


Fig. 2. T1-weighted cerebral MRI (January 24, 1992) showing the left-parietal localization of the AVM

symptoms recurred in a fashion similar to the initial presentation. Repeated cerebral angiography on January 27, 1993 showed the EIAB to be patent and suggested comparatively increased AVM blood flow (Fig. 1 b). The AVM was further examined by dynamic CT-scan study which demonstrated presence of intense steal effect and considerable shunt volume, whereas the diameter of the AVM had remained constant on MRI.

Successful extirpation of the AVM was performed on February 16, 1993. A right-sided hemiparesis occurred postoperatively and was progressively improving when the patient was transferred to a rehabilitation unit on March 23, 1993. However, after a period of approximately two and a half years the neurological deficit worsened anew, and the patient was readmitted to our hospital on October 11, 1995. This time, in addition to recurrence of left-hemispheric TIAs, she had experienced transient left-sided motor weakness. A CT-scan was performed on admission and remained inconclusive. On October 18, 1995 cerebral angiography showed no significant progression of MMD or presence of AVM, and the patient has been treated conservatively.

Discussion

From the nine cases of moyamoya disease associated with arteriovenous malformation previously described in the literature, six initially presented with symptoms of cerebral ischaemia [4, 5, 10–12] while a haemorrhage had taken place in the remaining three [1, 5, 15]. Distribution of gender was essentially equal and age ranged from 11 to 79 years (mean 39 years). In five cases the AVM was located in the frontal region without definite preference of side [1, 4, 5, 10, 15]; the remaining locations were left-parieto-occipital [12], in the corpus callosum and in the dis-

tribution of the right MCA [5]. Particularities were the description of *dural* AVM [15] and, in another case, the occurrence of the cerebrovascular phenomena within the context of *Sneddon's syndrome* [11]. In addition, a case of *de novo* development of a cerebral AVM in the exact location of a previous infarct in a child with MMD was very recently reported [12]. While operations for MMD such as encephaloduroarteriosynangiosis, encephalomyosynangiosis, encephaloponeurotic synangiosis, omentum transplantation, perivascular sympathectomy and superior cervical ganglionectomy were carried out in four cases [1, 4, 5, 10], only one patient underwent resection of the AVM [10]. In that patient, the left MCA was occluded one month after surgery.

In our case, the patient presented with clinical features of an ischaemic cerebral state and corresponding findings on CT-scan. The TCD results on initial admission suggested either compensatorily increased blood flow or long-distance stenosis. Given a constant vessel diameter, the relationship between blood flow velocity and blood flow within a large artery is linear. Acetazolamide is known to cause extracellular acidosis in the brain by reversibly inhibiting carbonic anhydrase; resulting from vasodilation at the arteriolar level, cerebral blood flow (CBF) is subsequently increased as a normal response [14]. The vasomotor reactivity may diminish if the vasodilatory mechanisms are impaired or the already maximally dilated vessels are incapable of further response. In this patient, the paradoxical decrease of flow velocity in the left MCA after ACZ infusion was therefore indicative of a possible intracerebral steal phenomenon.

The patient's neurological condition clearly improved following EIAB bypass but unfortunately deteriorated after nine months, leading to the extirpation of the left-parietal AVM. The fact that the operatively achieved improvement of the haemodynamic condition was lost may either be due to the natural course of MMD with progressive vascular narrowing and insufficient collateral blood supply or to increasing steal effect by the AVM that overtaxed bypass capacity. The latter consideration seemed more likely since on the one hand repeated angiography had suggested comparatively increased AVM blood flow and on the other hand the patient had regained almost full functional ability after extirpation of the AVM. Despite a constant diameter of the AVM on MRI one may assume that a "functional" progression of the AVM on the basis of vessel dilation not influencing the diameter of the AVM had occurred, the mecha-

nism of which remains unclear. Noteworthy, the hypercapnic vasoreactivity in moyamoya syndrome is severely reduced [13], probably due to already maximal vasodilation and/or decreased visco-elastic capacity of the cerebral vessels [2, 3, 9], also indicating highly impaired compensatory potential with regard to haemodynamic steal caused by an AVM. Yet, the recurrence of neurological deficits after extirpation of the AVM proves MMD to be an advancing process, which may be true on the level of microcirculation even if not detectable by angiography.

Several hypotheses have been raised to explain the genesis or coincidence of AVM in this disease [1, 4, 5, 10, 12, 15], and it was pointed out that in the presence of a cerebral AVM there may be an increase of blood flow and turbulence at the internal carotid bifurcation producing focal intimal hyperplasia which may lead to arterial mural thickening and progressing stenosis [8]. In contrast, under ischaemic conditions some cerebral AVM may result from hyperstimulated angiogenesis involving basic fibroblast growth factor, a powerful mitogen for fibroblasts, vascular endothelium, and smooth muscle cells [7].

Surgical mortality and morbidity rates of small AVM (less than 4 cm in diameter) are lower than the projected natural risk [6], so that surgical excision is almost always indicated. On the other hand, extirpation of the AVM does not seem to definitively improve the impaired haemodynamics in MMD as seen in this case.

References

1. Akiyama K, Minakawa T, Tsuji Y, Isayama K (1994) Arteriovenous malformation associated with moyamoya disease: case report. *Surg Neurol* 41: 468–471
2. Bishop CCR, Insall M, Powell S, Rutt D, Browse NL (1986) Effect of internal carotid artery occlusion on middle cerebral artery blood flow at rest and in response to hypercapnia. *Lancet* 1: 710–712
3. Fazekas JF, Alman RW (1964) Maximal vasodilation of cerebral vessels. *Arch Neurol* 11: 303–309
4. Kayama T, Suzuki S, Sakurai Y, Nagayama T, Ogawa A, Yoshimoto T (1986) A case of moyamoya disease accompanied by an arteriovenous malformation. *Neurosurgery* 18: 465–468
5. Lichtor T, Mullan S (1987) Arteriovenous malformation in moyamoya syndrome. Report of three cases. *J Neurosurg* 67: 603–608
6. Luessenhop AJ, Rosa L (1984) Cerebral arteriovenous malformations. Indications for and results of surgery, and the role of intravascular techniques. *J Neurosurg* 60: 14–22
7. Lyons MK, Anderson RE, Meyer FB (1991) Basic fibroblast growth factor promotes in vivo cerebral angiogenesis in chronic forebrain ischemia. *Brain Res* 558: 315–320
8. Mawad ME, Hilal SK, Michelsen WJ, Stein B, Ganti SR

- (1984) Occlusive vascular disease associated with cerebral arteriovenous malformations. *Radiology* 153 (2): 401–408
9. Norrving B, Nilsson B, Risberg J (1982) RCBF in patients with carotid occlusion: resting and hypercapnic flow related to collateral pattern. *Stroke* 13: 155–162
 10. Okada T, Kida Y, Kinomoto T, Sakurai T, Kobayashi T (1990) Arteriovenous malformation associated with moyamoya disease. *Neurol Med Chir (Tokyo)* 30: 945–948
 11. Scott IA, Boyle RS (1986) Sneddon's syndrome. *Aust N Z J Med* 16 (6): 799–802
 12. Schmit BP, Burrows PE, Kuban K, Goumnerova L, Scott RM (1996) Acquired cerebral arteriovenous malformation in a child with moyamoya disease. *J Neurosurg* 84: 677–680
 13. Tatemichi TK, Prohovnik I, Mohr JP, Correll JW, Quest DO, Jarvis L (1988) Reduced hypercapnic vasoreactivity in moyamoya disease. *Neurology* 38: 1575–1581
 14. Vorstrup S, Henriksen L, Paulson OB (1984) Effect of acetazolamide on cerebral blood flow and cerebral metabolic rate for oxygen. *J Clin Invest* 74: 1634–1639
 15. Yamasaki F, Hotta T, Taniguchi E, Eguchi K, Hashizume A, Kodama Y, Yuki K (1995) A case of dural arteriovenous malformation in the anterior fossa associated with an occlusion of

the unilateral middle cerebral artery with moyamoya phenomenon. *No Shinkei Geka* 23 (7): 603–607

Comments

It is the first European report on moyamoya disease associated with arteriovenous malformation. This combination is very rare, because moyamoya disease is thought to be an acquired disease, whereas AVM is a congenital disease.

Y. Yonekawa

Several cases of arteriovenous malformation associated with moyamoya disease have been reported especially from Japan, but the coincidence is rather rare. I think such a case presented in this report is extremely rare in Europe. The photos demonstrate the condition well.

K. Takakura

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