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Journal of Neurological Sciences 157 (1998) 100–104

JOURNAL OF THE
NEUROLOGICAL
SCIENCES

An atypical case of adult Moyamoya disease with initial onset of brain stem ischemia

Teruyuki Hirano^{1,*}, Eiichiro Uyama, Kazuhiro Tashima, Shuji Mita, Makoto Uchino

Department of Neurology, Kumamoto University School of Medicine, Kumamoto, Japan

Received 11 August 1997; received in revised form 9 December 1997; accepted 16 December 1997

Abstract

We present an atypical case of adult moyamoya disease whose clinical onset consisted of ischemic symptoms of the brain stem. She initially presented with left hemisensory disturbance caused by a pontine lesion, followed by a myelopathy of the upper cervical spinal cord. Eight months later, she presented with left hemiplegia and disturbed consciousness. Magnetic resonance angiography showed significant narrowing of both horizontal portions of the middle cerebral arteries (M1). Conventional angiography revealed bilateral occlusion of the internal carotid arteries. Her anterior circulation was supplied from the vertebro-basilar system through Moyamoya vessels and leptomeningeal collaterals. The intracranial steal phenomenon was thought to be the reason for the preceding events in the brain stem and upper cervical spinal cord. In addition, transcranial color-coded duplex sonography (TCCS) showed identical findings to conventional angiography with antegrade flow in the proximal M1 and retrograde flow in the distal M1. Thus, TCCS was useful for diagnosing the M1 occlusion in this case of Moyamoya disease. © 1998 Elsevier Science B.V.

Keywords: Moyamoya disease; Brain stem ischemia; Steal phenomenon; Cerebral angiography; Magnetic resonance angiography; Transcranial color-coded sonography (TCCS)

1. Introduction

Moyamoya disease (MIM 252350 [14]) is a clinical entity of unknown etiology, characterised by the angiographic finding of bilateral occlusion of the terminal portion of the internal carotid artery (ICA), together with a vascular network at the base of the brain [9]. The disease occurs mainly in females, particularly of Japanese origin [14]. The age distribution of patients with this disease displays two peaks [9]. The clinical onset in children is usually one of cerebral ischemia, whereas intracerebral hemorrhage is the common manifestation in adults.

The diagnostic criteria for moyamoya disease are based on conventional angiography [9]. Although such recent modalities as magnetic resonance angiography (MRA) [6,17] or transcranial Doppler sonography [15] have been described for the diagnosis of moyamoya disease, the usefulness of modalities other than conventional angiography has not been confirmed. Transcranial color-coded duplex sonography (TCCS) is a new method that allows the simultaneous visualisation of the brain parenchyma in B-mode, and the basal cerebral artery blood flow by using color flow imaging [2,3,11]. TCCS permits the noninvasive assessment of intracranial collaterals through the circle of Willis [12].

We describe here a woman with atypical moyamoya disease whose clinical onset consisted of ischemic symptoms of the brain stem, and discuss the mechanism of her brain stem ischemia. We also evaluated the usefulness of TCCS to depict her hemodynamic state, compared to MRA and conventional angiography.

*Corresponding author. Address for correspondence: Department of Neurology, Austin and Repatriation Medical Center, Studley Road, Heidelberg, Victoria 3084, Australia. Tel.: +61 3 949 65529; fax: +61 3 945 72654; e-mail: terry@austin.unimelb.edu.au

¹Present address: Department of Neurology, Austin and Repatriation Medical Center, Heidelberg, Victoria, Australia.

2. Case report

A 45-year-old right-handed woman with no family history of Moyamoya disease and no history of convulsion after hyperventilation, was admitted in March 1996. In June 1995, the patient noticed dysesthesia in her left face, oral cavity and left extremities. A mid-pontine infarction was diagnosed after MRI brain scan. She recovered gradually and was discharged in September 1995. Three months later, she developed tinnitus, followed by a sensory disturbance in both legs. The sensory disturbance gradually ascended to involve all dermatomes below and including C1, as well as the lower lip and chin suggesting partial involvement of the mandibular division of the trigeminal nerve. An MRI scan showed a new lesion in the upper cervical spinal cord. The patient was diagnosed as having multiple sclerosis, and pulse steroid therapy (1 g of methyl-prednisolone) was instituted. However, there was no neurological improvement. Several weeks later, her symptoms and signs ameliorated. In February 1996, she complained of left hemisensory disturbance. She developed weakness of her left leg in the middle of March, which progressed to complete left hemiparesis over two weeks. She was again treated with pulse steroid therapy without any response, and she developed disturbed consciousness.

On admission, the patient exhibited akinetic mutism and left hemiplegia, involving the face. Her left extremities were hypotonic, but the deep tendon reflexes were markedly increased with positive Babinski's sign. Pinprick and vibration sense were impaired in the left upper extremity. Pinprick was also diminished below the T4 level bilaterally.

She showed no skin lesion such as café-au-lait spots, port wine nevus, hypopigmentation, or adenoma sebaceum. Extensive laboratory studies including complete blood cell count, serum calcium, coagulation factors, fibrinolytic activity, various autoimmune antibodies, echocardiogram, and cerebrospinal fluid analysis were normal. The electroencephalogram did not show any abnormalities, including the so-called rebuild-up phenomenon [10].

2.1. MRI, MRA

A brain MRI (Magnetom; Siemens) on April 9, 1996, showed large ischemic lesions in the bilateral fronto-parietal lobe, predominantly in the right hemisphere. They were located in the anterior watershed and terminal zone of the middle cerebral artery (MCA) territories. There were significant vascular narrowings in the proximal right MCA, the proximal left MCA and left anterior cerebral artery (ACA) A-1 segment on three-dimensional time-of-flight MRA (Fig. 1).

2.2. Conventional angiography

Cerebral angiography done on April 15 revealed bilater-

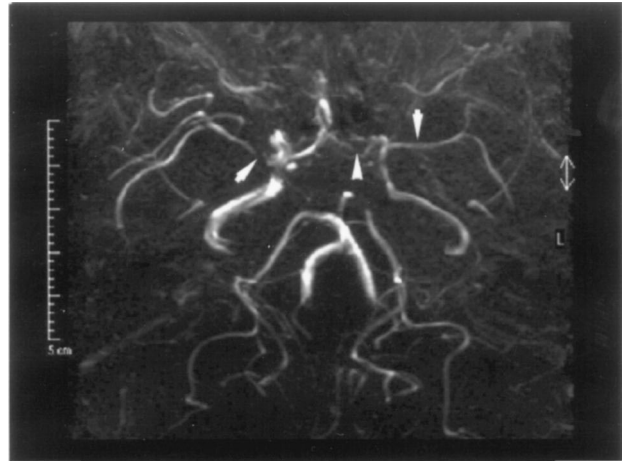


Fig. 1. MR angiography (April 9, 1996). The three-dimensional time-of-flight MRA suggests the presence of significant vascular narrowing in the proximal right middle cerebral artery (MCA), the proximal left MCA (arrows) and left anterior cerebral artery A-1 segment (arrow head).

al occlusion of the ICA after the origins of the ophthalmic arteries (Fig. 2). They terminated with abnormal vascular networks. The vertebral angiogram showed a good collateral flow to the ICA territories (Fig. 3). Although the posterior communicating arteries were not detected, both MCA territories were filled through leptomeningeal collaterals from the posterior cerebral arteries (PCA). This collateral blood flow filled the lateral lenticulostriate arteries as well. Similarly, both ACA territories were supplied from pericallosal arteries through the PCA (Fig. 3). In addition to these retrograde collaterals, the MCA frontal branch was also supplied from small abnormal vessels branched from the PCA. These small vessels and the distal end of both occluded ICA appeared as moyamoya vessels.

2.3. TCCS

TCCS was performed on May 2 using the HDI 3000 (Advanced Technology Laboratories Inc., USA) instrument. About four-fifths of the proximal part of the horizontal MCA (M1) segments showed flow towards the probe, and were displayed in red bilaterally; the distal one-fifth of both M1 segments showed flow away from the probe and were displayed in blue (Fig. 4A–C). Using duplex scanning, these flow directions were confirmed with pulse wave Doppler measurements (Fig. 4D, E). These findings suggested occlusion of the MCA.

2.4. Treatment and biopsy

To prevent further ischemic events in the left hemisphere, a left superficial temporal artery-MCA anastomosis was performed on June 4, 1996. A brain biopsy specimen was also obtained from the right frontal lobe under informed consent. Examination of the tissue sections revealed ischemic necrosis with glial degeneration, mas-

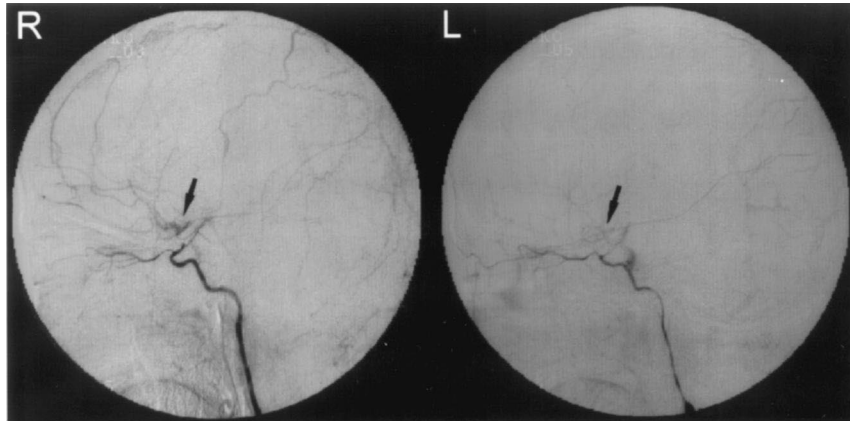


Fig. 2. Bilateral carotid angiography. The lateral view of the carotid angiogram shows the bilateral occlusion of the internal carotid arteries (ICA). Moyamoya vessels (arrows) are shown in the vicinity of the bilateral ICA terminal, but they are not as prominent as in a typical Moyamoya patient.

sive infiltration of foam cells, and occasional reactive inflammatory cells. In addition, there were abnormal networks of small vessels in the pia matter.

3. Discussion

The diagnosis of Moyamoya disease is based on angiography [9]. The criteria for a 'definite' diagnosis comprise: (1) stenosis or occlusion at the terminal portion of the ICA and at the proximal portion of the ACA and MCA, (2) abnormal vascular network in the arterial phase in the vicinity of the arterial occlusion, and (3) bilateral involvement [9]. The present case met these criteria as there was no other known cause such as arteriosclerosis, menin-

gitis, neoplasm, Down's syndrome, von Recklinghausen neurofibromatosis, tuberous sclerosis, incontinentia pigmenti, encephalotrigeminal angiomas, William-Beuren hypercalcemia and supravalvular aortic stenosis, trauma, or irradiation [8,9,19] to account for her illness. However, our patient was an atypical case of adult-onset Moyamoya disease as she presented slowly progressing brain ischemia at the age of 45 years, and a brain stem lesion preceded the supratentorial lesions.

It is thought that in more than one-half of the patients with adult-onset moyamoya disease it causes cerebral hemorrhage [19]. On the other hand, cerebral ischemia is not a common initial manifestation in this group of patients. Although various neurological manifestations have been reported [9], brain stem involvement is very

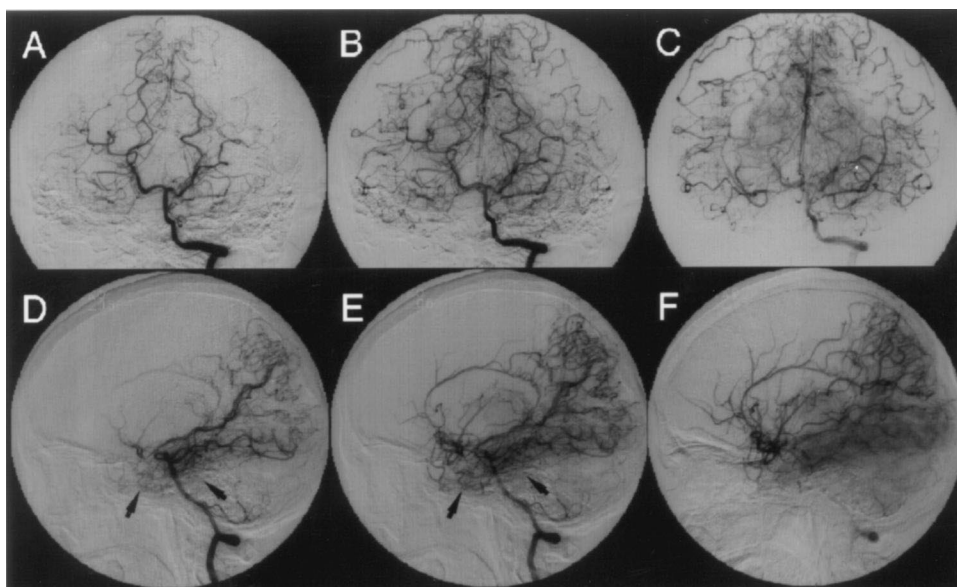


Fig. 3. Left vertebral Angiography. Serial pictures of left vertebral angiogram (AP view: A, B, C; lateral view: D, E, F) show the prominent collateral vessels supplying the anterior and middle cerebral artery territories. The Moyamoya vessels are seen around the origin of the posterior cerebral arteries (arrows). The horizontal segments of the middle cerebral arteries (M1) are bilaterally filled by two collateral pathways. The M1 proximal portion is filled through the Moyamoya vessels and the terminal portion by retrograde flow through leptomeningeal vessels.

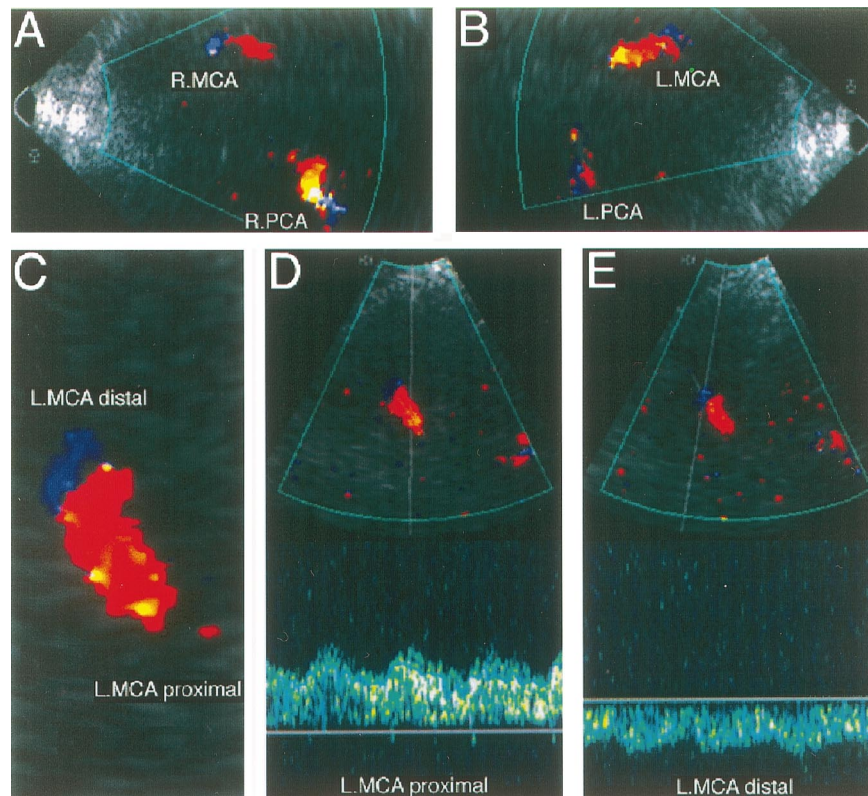


Fig. 4. Transcranial color-coded real-time sonography (TCCS). Color flow imaging from bilateral temporal bone windows (right: A; left: B). An enlargement (C) of the left middle cerebral artery horizontal segment (M1) shows antegrade flow toward the probe (displayed in red color) in the proximal portion of M1. However, the distal portion of M1 flows away from the probe (displayed in blue). Doppler flow measurement of proximal (D) and distal (E) parts of the M1 segment confirm the change in flow direction.

rare. The reason our patient initially presented with a brain stem symptom is unclear. According to the angiographical classification of stage in Moyamoya disease [16], our case seems to be classified as stage 5. While the moyamoya vessel usually reduces in this stage, the collateral circulation from the external carotid artery increases. In the present case, however, the ICA territories were perfused by collateral flow from the vertebrobasilar artery system. It is likely that to maintain the supratentorial blood supply, the intracranial steal phenomenon occurred in the territory of the basilar artery, and may explain the brain stem symptoms.

Pathological characteristics of moyamoya disease in previously presented studies [7,13,18] are eccentric intimal thickening and mural thrombus in the main branches of brain arteries. Similar vascular damage is also seen in perforating arteries. Lipohyalinosis, degeneration of the elastic lamina and microaneurysms have also been reported. Although our patient's vertebral angiogram showed no abnormality, there might have been some vascular fragility in the basilar artery or its small branches. Yamashita et al. found abnormal dilated vessels that were not identified by angiography [18]. On the other hand, Ikeda and Hosoda examined 28 cases of moyamoya disease and found thrombi composed of platelets and fibrin

in a small branch of the basilar artery of one patient [7]. In addition, the steno-occlusive lesion of the anterior circulation may alter the circulatory dynamics in the basilar artery system [1].

MRI and MRA are generally useful for the screening of patients for Moyamoya disease [4–6,17]. Although significant vascular narrowing of bilateral MCA and left ACA were described, occlusion of the ICA was not detected by MRA. The discrepancy between the results of the catheter angiogram and the MRA was thought to be caused by the unique intracranial hemodynamics of this patient, i.e. the retrograde filling of the MCA branches.

By using TCCS, we detected a unique MCA flow pattern in our patient. We found a change in the flow direction between the proximal and distal portions of the same M1 segment. We hypothesize that the proximal M1 portion was filled by antegrade flow from the Moyamoya vessels, and that the distal portion was supplied by retrograde flow from leptomeningeal collateral flow through the PCA. Consequently, we believe that the end of the antegrade flow is the occluded site of the MCA.

In summary, we describe an atypical case of Moyamoya disease who showed slowly progressing symptoms, probably due to a hemodynamic mechanism. The initial brain stem ischemia seemed to be caused by intracranial steal

through Moyamoya vessels. In the present case, TCCS accurately described the unique flow pattern of the occluded MCA.

Acknowledgements

The authors thank Dr Jyun-ichiro Hamada and Prof. Yukitaka Ushio (Department of Neurosurgery, Kumamoto University School of Medicine, Kumamoto, Japan) for performing surgical treatment and obtaining the brain biopsy. We also thank Dr Stephen J. Read (Department of Neurology, Austin and Repatriation Medical Center, Heidelberg, Victoria, Australia) for reviewing our manuscript.

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