Acta Neurochir (Wien) (2000) 142: 421–424

Acta Neurochirurgica

Steno-Occlusive Changes in the External Carotid System in Moyamoya Disease

M. Komiyama[1], M. Nishikawa[1], T. Yasui[1], S. Kitano[2], H. Sakamoto[2], and Y. Fu[3]

[1]Department of Neurosurgery, Osaka City General Hospital, Osaka, Japan
[2]Department of Paediatric Neurosurgery, Osaka City General Hospital, Osaka, Japan
[3]Department of Neurosurgery, Tsukazaki Hospital, Hyogo, Japan

Summary

To evaluate the steno-occlusive changes in the external carotid system in moyamoya disease, cerebral angiograms of 39 moyamoya patients were retrospectively reviewed. There were 26 females and 13 males, age ranged from 4 to 62 years with a mean of 26 years. Initial symptoms were ischaemia in 27 patients, haemorrhage in 9, and none in 3. Stenosis, occlusion, and dilatation in the external carotid system were analysed angiographically. No stenosis or occlusion of the superficial temporal artery, middle meningeal artery, or occipital artery was observed in either preoperative or postoperative follow-up angiograms in any patients. Steno-occlusive changes do not occur in the external carotid system, but are confined in the internal carotid system in moyamoya disease.

Keywords: Angiography; external carotid system; moyamoya disease.

Introduction

Moyamoya disease is characterized by the steno-occlusive changes of both distal internal carotid arteries with arterial collaterals at the base of the brain known as "moyamoya" vessels [8, 12, 14]. There have been a few reports on the external carotid system in moyamoya disease because it is believed that the steno-occlusive changes occur exclusively in the internal carotid system. Angiographic stenosis in the external carotid system is, however, reported in about 20% of moyamoya patients [3]. If the steno-occlusive changes do occur frequently, spontaneously or surgically established extracranial to intracranial anastomoses might occlude, resulting in further ischaemic events. Steno-occlusive changes in the external carotid system and ischaemic stroke due to these changes are not a common occurrence in our clinical experience. Thus, we undertook the retrospective study to review angiograms of moyamoya disease to clarify the steno-occlusive changes in the external carotid system.

Methods

There are 39 moyamoya patients (26 females and 13 males), and their ages ranged from 4 years old to 62 years old with a mean of 26 years. Figure 1. Angiographic techniques were film screen magnification angiography in the early 12 patients and digital subtraction angiography in the more recent 26 patients. In one patient, preoperative study was performed by film screen angiography and postoperatively by digital subtraction angiography. Bilateral carotid and vertebral angiograms were obtained in all the patients. Selective external and internal carotid and stereoscopic angiograms were obtained in most cases. Their cerebral angiograms were reviewed with special reference to stenosis, occlusion, and dilatation in the external carotid system.

Initial symptoms were ischaemia in 27 patients (transient ischaemic attack in 16 and infarction in 11), haemorrhage in 9, and none in 3. Mean ages of the patients with ischaemic onset and haemorrhagic onset were 14 and 42 years-old at the initial presentation, respectively. Cerebral angiography at the initial diagnosis was available in 25 patients and that at the follow-up examination after bypass surgery was available in 25 patients. In 11 patients, angiography both at the initial and follow-up examinations was available. The angiographic follow-up periods after surgery ranged from 1 month to 16 years with a mean of 6 years. Bypass surgery was performed in 23

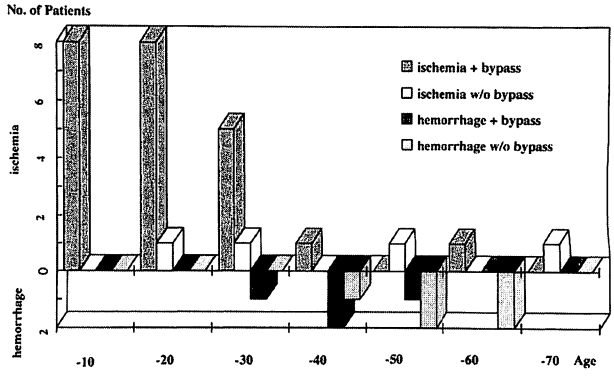
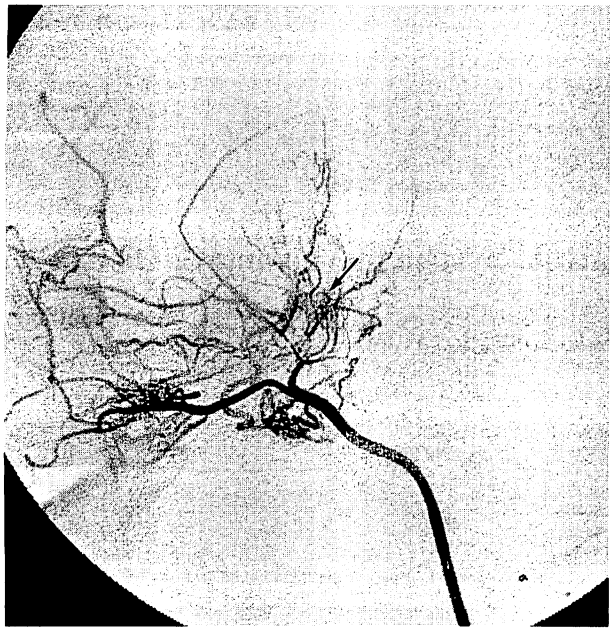
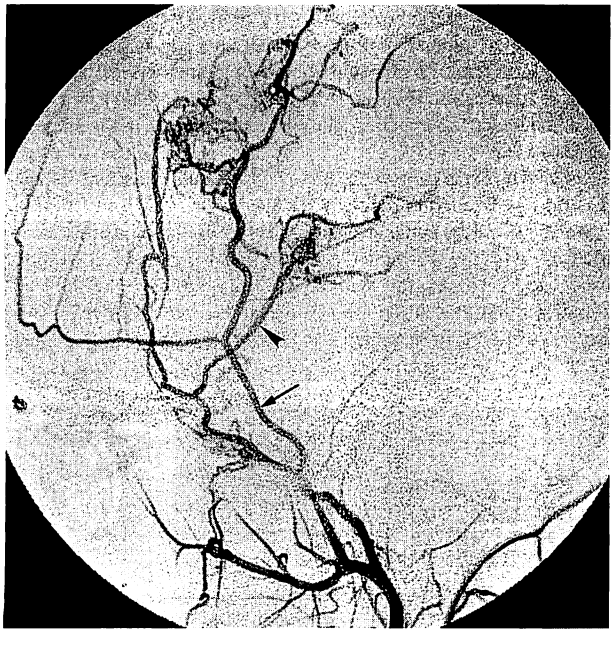


Fig. 1. Patients' data. Age distribution at the time of angiography, initial symptoms, and history of bypass surgery. Three asymptomatic patients are not included



a



b

Fig. 2. A 13-year-old girl. This patient started transient ischaemic attacks at the age of 3 years, and bilateral encephalo-duro-arterio-synangioses were performed at the age of 5. Angiography was carried out at the age of 13. (a) Right internal carotid injection (lateral view) shows occlusion of the internal carotid artery at the level of the posterior communicating artery and development of basal moyamoya vessels (arrow). (b) Right external carotid injection (lateral view) shows typical postoperative anastomoses from the hypertrophied superficial temporal artery (arrow) and middle meningeal artery (arrowhead) to the branches of the middle cerebral artery. No steno-occlusive changes are observed

patients with ischaemic onset, in 4 patients with haemorrhagic onset, and in one asymptomatic patient. Principal surgical techniques included bilateral superficial temporal artery (STA)-MCA anastomoses in 12 patients, bilateral encephalo-duro-arterio-synangioses (EDAS) in 11, unilateral STA-MCA anastomoses and contralateral EDAS in 2, and unilateral EDAS in 3. No bypass surgery was carried out in 11 patients.

Results

No stenosis or occlusion in the external carotid system was observed in any patients. (Fig. 2). Although in some patients, vessels projected on angiograms near the margin of the cranial vault, bony sutures, or skull base seemed to be having mild stenosis, careful observation in other projections confirmed that there was no stenosis in these vessels. Most STAs and/or middle meningeal arteries were hypertrophied after bypass surgery. In a 12-year-old girl, stenosis at the site of STA-MCA anastomosis was observed. (Fig. 3). A segmental, fusiform dilatation of the STA was observed in a 41-year-old female patient. (Fig. 4).

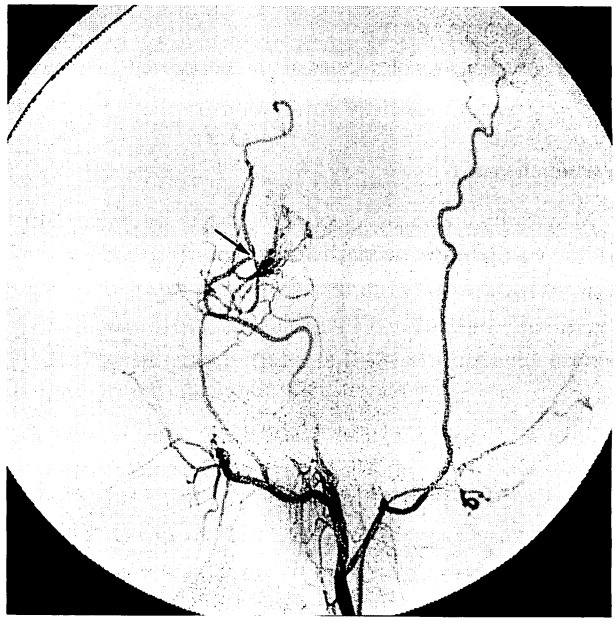


Fig. 3. A 12-year-old girl. This patient started transient ischaemic attacks at the age of 8 years. She underwent superficial temporal artery-middle cerebral artery (*STA-MCA*) anastomoses on the left side at the age of 10 years and on the right side at the age of 12 years. Angiography was performed 2 months after the second operation. Left external carotid angiogram (lateral view) shows a stenosis at the site of STA-MCA anastomosis which was established about 2 years ago (arrow).

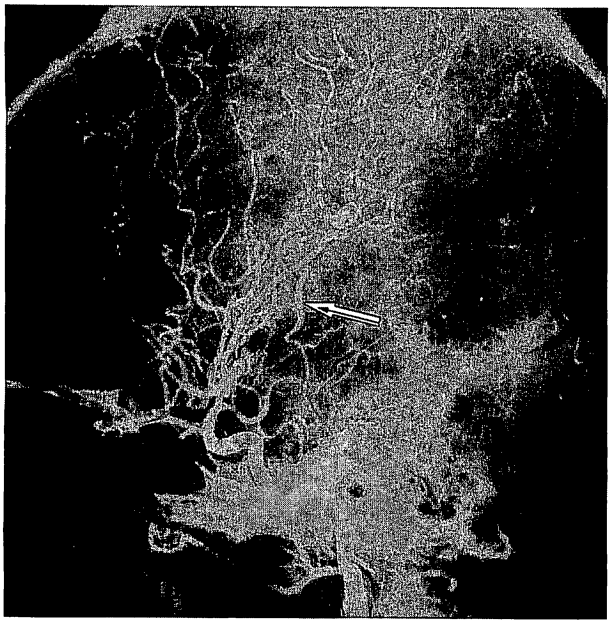


Fig. 4. A 41-year-old female. This patient developed intraventricular haemorrhage. Left common carotid injection (lateral view) shows that the left superficial temporal artery has segmental, fusiform dilatation (arrow), which is repeatedly demonstrated in the follow-up angiograms 1 year later

Discussion

Histopathological changes of the distal internal carotid artery, and the proximal anterior cerebral arteries and MCAs in moyamoya disease are fibrous intimal thickening with laminated elastic fibers [5, 11]. There are few lipid depositions or infiltration of inflammatory cells. Similar histopathological changes may occur in the pulmonary artery, coronary artery, renal artery, or pancreatic artery in moyamoya disease [2, 6, 16], but these changes do not occur in all patients. Like the steno-occlusive change in the internal carotid arteries, stenosis of the renal arteries and coronary artery does not occur in the entire segments, but occurs in the limited segments [16]. This fact suggested that moyamoya disease has both systemic and focal aetiological factors affecting the intracranial as well as extracranial vessels.

Histopathological changes of the STA and middle meningeal artery are proliferation of smooth muscle cells and thickening of the intima, and degeneration and destruction of the smooth muscle cells, which are similar to changes in the distal portion of the internal carotid arteries [9]. Contrary to this, some histological reports did not find any abnormality in the biopsied STA [1, 10]. Our personal experience conforms to these latter reports.

Hoshimaru and Kikuchi [3] report that about 20% (13 of 66 patients) of the external carotid arteries showed stenotic lesions on angiograms, where stenosis of the STA was noted in four of these 13 patients (6.0% of all patients). Although they did not indicate predilection of the sites of such stenotic changes, figures in their report show that the stenoses are limited to the short segments in the distal STAs, middle meningeal arteries, and occipital arteries. Suzui *et al.* [13] report that one of the 12 patients (8.3%) showed stenosis of the STA. To our knowledge, no patient has been reported to have vascular occlusion in the external carotid system. Since there are not enough data on the patients' age distribution, clinical presentation, time of angiographic examination, and history of bypass surgery in the report by Hoshimaru and Kikuchi [3], we can not compare the results of our and their studies precisely. However, the discrepancy between the two studies may be interpreted that stenotic changes in the external carotid system may occur infrequently in limited short segments, and that occlusion in the external carotid system does not occur.

Basic fibroblast growth factor (FGF) is one of the angiogenic factors and a potent mitogen for fibroblasts, endothelial cells and smooth muscle cells [7]. Several reports emphasized the role of basic FGF in angiogenesis in moyamoya disease [4, 13]. High concentration of basic FGF was observed in the cerebrospinal fluid (CSF) [15] and in the STA [4]. A high concentration of the basic FGF receptors in the STA was also reported [15]. Suzui *et al.* [13] speculate that the molecular mechanism responsible for intimal thickening of the intracranial arterial trunk may work for the external carotid system due to increased basic FGF and its receptors in the STA. Although strong staining of basic FGF was observed in the intima and media of the STA and dural vessels in patients with moyamoya disease [4]. This is not disease-specific. Staining of basic FGF was not observed in the STA of two of four moyamoya patients, while less intense staining of basic FGF was observed in the STA of the atherosclerotic patients [4]. Thus, we postulate that the role of basic FGF is not directly related to the angiogenesis in the external carotid system in moyamoya disease, but there may be some different response to basic FGF in the internal and external carotid system, respectively.

Although there are several pathological and radio-

logical studies concluding that moyamoya disease involves not only the internal carotid system but also the external carotid system, our study showed that steno-occlusive changes do not occur in the external carotid system. This suggests that histopathological changes may occur in both the internal carotid and external carotid systems, but susceptibility to cause the steno-occlusive changes is different between them. The distal internal carotid artery is most susceptible, while the external carotid system is less susceptible to the steno-occlusive changes due to unknown pathogenesis. Stenosis at the site of STA-MCA anastomosis in one patient in this series may be attributable to the surgical procedure. Although there was a segmental dilatation of the STA in another patient, we could not find any systemic or topical predisposing factors to develop such a dilatation in this patient.

In conclusion, steno-occlusive changes do not occur in the external carotid system, but are confined to the internal carotid system in moyamoya disease. It is postulated that susceptibility to cause such steno-occlusive changes in the internal carotid system may be different from that in the external carotid system.

References

1. Boone SC, Sampson DS (1978) Observation on moyamoya disease: a case treated with superficial temporal-middle cerebral artery anastomosis. Surg Neurol 9: 189–193
2. Hanakita J, Kondo A, Koyama T, Ishikawa J, Hazama F (1982) An autopsy case of "moya moya" disease. No Shinkei Geka 10: 531–539
3. Hoshimaru M, Kikuchi H (1992) Involvement of the external carotid arteries in moyamoya disease: neuroradiological evaluation of 66 patients. Neurosurgery 31: 398–400
4. Hoshimaru M, Takahashi JA, Kikuchi H, Nagata I, Hatanaka M (1991) Possible roles of basic fibroblast growth factor in the pathogenesis of moyamoya disease: an immunohistochemical study. J Neurosurg 75: 267–270
5. Hosoda Y (1976) A pathological study of so-called "spontaneous occlusion of the circle of Willis" ("cerebrovascular moyamoya disease"). Folia Angiol 14: 85–86
6. Ikeda E (1991) Systemic vascular changes in spontaneous occlusion of the circle of Willis. Stroke 22: 1358–1362
7. Klagsbrun M, Edelman ER (1989) Biological and biochemical properties of fibroblast growth factors. Implications for the pathogenesis of atherosclerosis. Arteriosclerosis 9: 269–278
8. Kudo T (1968) Spontaneous occlusion of the circle of Willis. A disease apparently confined to Japanese. Neurol 18: 485–496
9. Li B, Wang C-C, Zhao Z-Z, Hu Y, Aihara K, Ghazizadeh M, Sasaki Y, Yang S-Y, Pan J (1991) A histological, ultrastructural and immunohistochemical study of superficial temporal arteries and middle meningeal arteries in moyamoya disease. Acta Pathol Jpn 41: 521–530
10. Mastri AR, Silverstein PM, Gold L, Eselius EP (1973) Multiple progressive intracranial arterial occlusions. Stroke 4: 380–386
11. Oka K, Yamashita M, Sadoshima S, Tanaka K (1981) Cerebral haemorrhage in moyamoya disease at autopsy. Virchows Arch 392: 247–261
12. The Research Committee on Spontaneous Occlusion of the Circle of Willis ("Moyamoya" Disease) to the Ministry of Health and Welfare, Japan (1990) Diagnostic criteria for spontaneous occlusion of the circle of Willis ("Moyamoya" disease), In: Annual report (1989) of the Research Committee on Spontaneous Occlusion of the Circle of Willis ("Moyamoya" Disease) to the Ministry of Health and Welfare, Japan, pp 184
13. Suzui H, Hoshimaru M, Takahashi JA, Kikuchi H, Fukumoto M, Ohta M, Itoh N, Hatanaka M (1994) Immunochemical reactions for fibroblast growth factor receptor in arteries of patients with moyamoya disease. Neurosurgery 35: 20–25
14. Suzuki J, Takaku A (1969) Cerebrovascular "moyamoya" disease. Disease showing abnormal net-like vessels in base of brain. Arch Neurol 20: 288–299
15. Takahashi A, Sawamura Y, Houkin K, Kamiyama H, Abe H (1993) The cerebrospinal fluid in patients with moyamoya disease (spontaneous occlusion of the circle of Willis) contains high level of basic fibroblast growth factor. Neurosci Lett 160: 214–216
16. Yamashita M, Oka K, Tanaka K (1983) Histopathology of the brain vascular network in moyamoya disease. Stroke 14: 50–58
17. Yamashita M, Tanaka K, Kishikawa T, Yokota K (1984) Moyamoya disease associated with renovascular hypertension. Hum Pathol 15: 191–193

Correspondence: Dr. Masaki Komiyama, M.D., Department of Neurosurgery, Osaka City General Hospital, 2-13-22, Miyakojima-Hondouri, Miyakojima, Osaka 534-0021 Japan.