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Cerebral Haemorrhage in Moyamoya Disease at Autopsy*

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Summary. Nineteen fatal cases of occlusion of the circle of Willis, so-called Moyamoya disease, were examined clinicopathologically.

Fresh and massive cerebral haemorrhage was confirmed in 14 and cerebral infarcts of 4 of 19 patients. Among these 14 patiens, massive haemorrhage was found in the basal ganglia, thalamus and hypothalamus of 9, and in the thalamus, cerebral peduncle and midbrain of 5. Pathologically, fibrosing stenoses or occlusions involved the circle of Willis and its major branches in all cases. In 13 of 17 patients numerous collateral channels, muscular in type, paralleled the circle, bypassing the occluded natural passages. Rupture of dilated small muscular collateral arteries was demonstrated in fresh and old haemorrhagic lesions in 3 of the 14 patients. Saccular aneurysm of cerebral arteries in the subarachnoid spaces was present in two of the 19. No rupture involved the perforating arteries in the subarachnoid space.

These findings strongly suggest that in patients with Moyamoya disease rupture of overgrown perforating arteries as collaterals in brain may be main cause of single or repeated cerebral haemorrhage. Stenoses or occlusions of these perforators are presumably an important factor in the occurrence of cerebral infarcts.

Key words: Moyamoya disease – Cerebral haemorrhage – Occlusion of the circle of Willis – Perforating artery – Cerebral infarct

Introduction

In 1957 Takeuchi and Shimizu described a patient with occlusion of the circle of Willis, so-called Moyamoya disease. Since then other instances have been

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reported from hospitals elsewhere in the world (Weidner et al. 1965; Vuia et al. 1970; Serdau et al. 1979) but especially Japan (Maki and Nakata 1965; Nishimoto et al. 1965; Nomura 1965; Sano 1965; Suzuki et al. 1965; Suzuki and Takaku 1969; Suzuki et al. 1976; Kudo 1966, 1967; Kodama et al. 1976). Moyamoya disease is recognized as a clinical disease entity with angiographic findings of bilateral but asymmetrical stenosis or occlusion of the distal ends of internal carotid arteries and an unusual vascular network in the region of the basal ganglia. Similar angiographic findings in various disease entities (Mathew et al. 1970; Rosengren 1974; Painter et al. 1975; Numaguchi et al. 1976) including neurocutaneous syndrome, meningitis, sickle cell disease, tetralogy of Fallot and radiation angiitis must be excluded. The pathogenesis and prognosis of this disease remain uncertain. In general, patients under 16 years of age experience sudden onset of unconsciousness and convulsion as their initial symptoms; patients older than 16 years typically present with the signs and symptoms of subarachnoid haemorrhage or cerebral haemorrhage. There is a paucity of reports (Vuia et al. 1970; Suzuki et al. 1976; Kodama et al. 1976; Serdau et al.

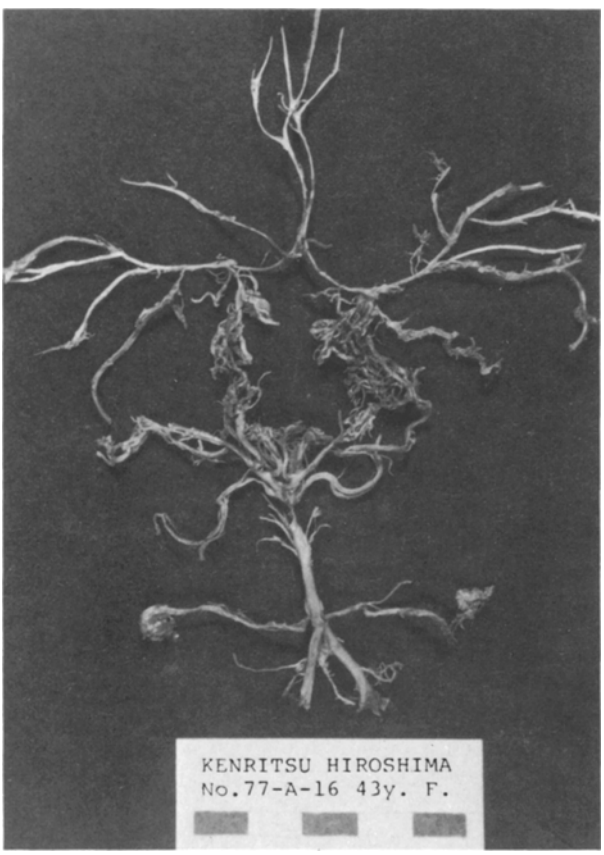

Fig. 1. Overgrown and dilated arteries, branching from the circle of Willis (No. 11). Narrowing of the anterior and middle cerebral arteries is found. The basilar artery just proximal to the superior cerebellar arteries is tapered where a short segment of occlusion is present

1979) of the intracranial lesions, except for the pathological changes in the intracranial extracerebral blood vessels.

The frequency, distribution and cause of cerebral lesions of 19 autopsy cases with Moyamoya disease are reported herein.

Materials and Methods

Nineteen cases diagnosed clinically as Moyamoya disease and autopsied during the 10 years from 1970 to 1979 were examined pathologically. For each patient the pathological protocols, clinical

Table 1. Autopsy cases with occlusion of the circle of Willis

Nos.	Age (years)	Sex	Initial symptoms	Episodes	Course	Treatment
1	7	F	Headache, nausea, vomiting	Frequent	5 years (Y)	Conservative
2	8	F	Numbness in rt. lower extremity	3	7 months (M)	Conservative
3	17	M	General weakness	Frequent	12Y	Conservative
4	18	F	Headache, vomiting, unconsciousness	2	2Y & 8M	Conservative
5	27	F	Seizure, vomiting, rt. hemiplegia	1	5Days(D)	Conservative
6	30	F	Headache, vomiting	3	3Y & 4M	Conservative
7	38	F	Headache, vomiting	2	4M	Conservative
8	41	M	Aphasia, gait disturbance, rt. homonymous hemianopsia	3	6Y & 2M	Conservative
9	41	F	Unconsciousness	2	2Y & 4M	Removal of hematoma Ventricular drainage
10	42	F	Unconsciousness	4	14Y	Ventricular drainage
11	43	F	Rt.hemiplegia	3	7Y	Conservative
12	45	M	Weakness in rt. lower extremity	1	10Y & 9M	Conservative
13	46	F	Transient unconsciousness, dysphasia	1	Several Months	Conservative
14	46	M	Unconsciousness	3	14Y	Conservative
15	48	M	Headache, vomiting, unconsciousness	1	1M	Removal of hematoma Ventricular drainage
16	52	F	Vomiting, unconsciousness, urinary incontinence	1	14D	Ventricular drainage
17	54	F	Unconsciousness	1	2D	Conservative
18	63	M	Headache, vomiting, unconsciousness	1	11D	Conservative
19	64	F	Unconsciousness	1	8D	Conservative

Table 2. Distribution of intracranial haemorrhage and cerebral infarcts in autopsy cases with occlusion of the circle of Willis

Nos.	Sites of massive haemorrhage	Sites of infarcts
1.	Subarachnoid haemorrhage due to rupture of post. cerebral-post. communicating aneurysm	
2.	Rt. basal ganglia, frontal lobe with intraventricular extension	
3.	Midbrain, pons, medulla oblongata (petechiae)	Bilat. caudate nuclei, thalami, bilat. cingulate gyri, rt. frontal lobe
4.	Rt. cerebral peduncle with intraventricular extension	
5.	Lt. cerebral peduncle	
6.	Bilat. basal ganglia, hypothalami with intraventricular extension. Rt. parietal lobe, lt. occipital lobe	
7.	Lt. basal ganglia, thalamus with intraventricular extension	Rt. internal capsule
8.		Multiple lesions in cerebral hemispheres
9.	Lt. basal ganglia, thalamus, with intraventricular extension	
10.	Lt. basal ganglia, thalamus, hypothalamus with intraventricular extension	
11.	Lt. thalamus, hypothalamus, cerebral peduncle, midbrain with intraventricular extension	
12.		Rt. frontal, parietal and temporal lobes, lt. temporal lobe
13.	Rt. basal ganglia with intraventricular extension	
14.	Rt. thalamus, midbrain, pons	
15.	Thalamus, midbrain, pons (small foci of haemorrhage)	Bilat. temporal lobes, multiple lesions around third ventricle
16.	Lt. basal ganglia, thalamus, temporal lobe with intraventricular extension	
17.	Rt. basal ganglia, thalamus, hypothalamus, white matter of frontal, parietal and temporal lobes with intraventricular extension	
18.	Rt. basal ganglia, thalamus, hypothalamus with intraventricular extension	
19.	Pons, cerebral peduncle, tuber cinereum, optic chiasm with intraventricular extension	

records, organs including brain, microscopic slides and paraffin blocks of tissue were available for study. Arteries at the base of brain in the 4 cases were removed by the authors as shown in Fig. 1; sections were obtained from narrowed, occluded portions and from bifurcation of the circle of Willis. The brain was examined macroscopically by cutting serially in the frontal plane. Histological sections of the brain and the blood vessels in subarachnoid space were routinely stained with haematoxylin and eosin, elastic van Gieson and Mallory's phosphotungstic acid haematoxylin.

Three patients (Nos. 4, 5 and 10) have already been reported by Mabuchi et al. (1973), Handa et al. (1969) and Ando et al. (1967).

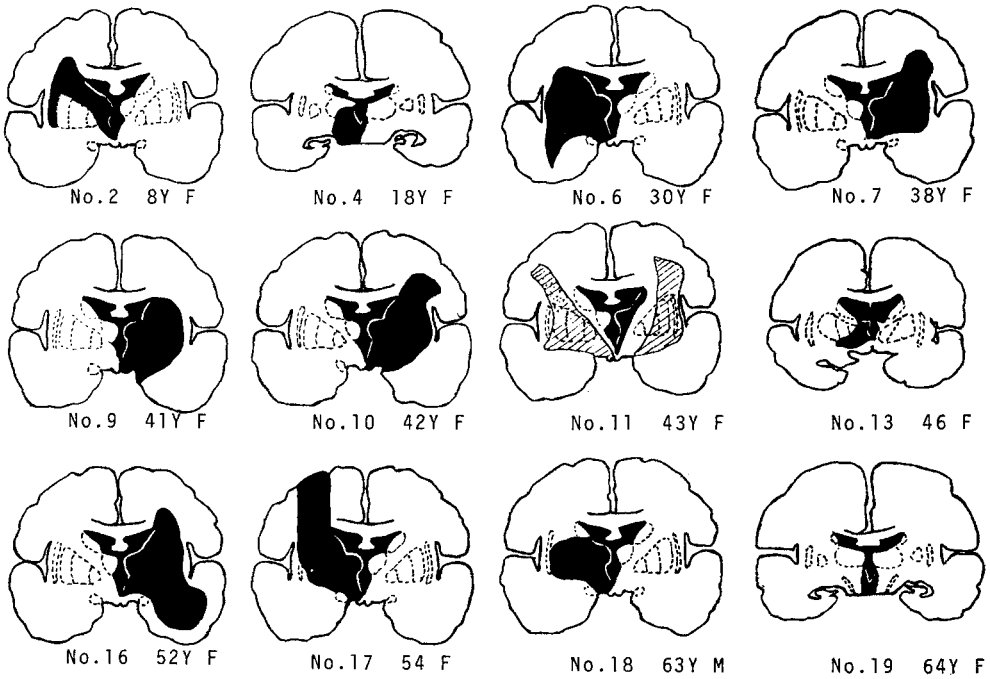

Fig. 2. Schematic illustration of the distribution of cerebral haemorrhage in patients with Moyamoya disease. Fresh haemorrhage ■; Old haemorrhage ▨

Results

The main clinical features of our 19 patients are presented in Table 1. These patients ranged in age from 7 to 64 years, with a female to male ratio of 13 to 6. Thirteen patients presented with typical haemorrhagic strokes and of them, 6 (Nos. 4, 15, 16, 17, 18 and 19) died in the first episode. Including these patients, death occurred from 2 days to 14 years after the initial symptoms.

The distribution of cerebral haemorrhage and infarcts is summarized in Table 2. Fourteen of the 19 patients revealed fresh massive cerebral haemorrhage, and in 12 of them, there was intraventricular extension. The localization and distribution of haemorrhage is illustrated schematically in Fig. 2. Observing Mutlu's classification of massive cerebral haemorrhage (1963), 4 were of the medial type (the lateral type of old haemorrhage was also present in one of the patients) and 8 were of the combined type. The distribution pattern of haemorrhage was divided into two groups. One group (9 patients) involved the basal ganglia (the caudate nucleus, lenticular nucleus, claustrum and amygdaloid nucleus), thalamus and hypothalamus. The other (5 patients) involved the thalamus, cerebral peduncle and midbrain. Massive haemorrhage in the former extended into the island of Reil, the temporal, frontal and parietal lobes.

In two patients (Nos. 11 and 17) massive intraparenchymal haemorrhage with intraventricular involvement broke into the subarachnoid space on the convexity. Patient No. 11 who died with fresh, massive, unilateral cerebral haemorrhage with intraventricular extension had had two cerebrovascular accidents,

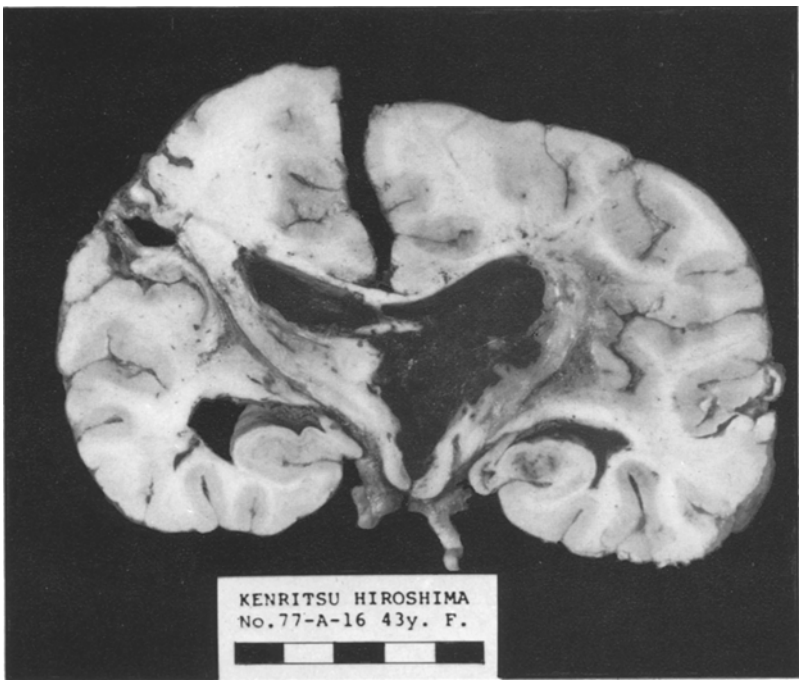

Fig. 3. Coronal section of the cerebral hemispheres (No. 11) at the level of the mamillary bodies. This shows fresh haemorrhage in the left thalamus and hypothalamus. Old haemorrhage is noted in the bilateral basal ganglia

7 years and 7 months previously, accounting for bilateral old haemorrhages (Table 2, Fig. 2) at autopsy. Proliferation of small- to medium-sized muscular arteries showing dilatation was found within the fresh haemorrhagic lesion in the left thalamus and hypothalamus (Figs. 3, 4). One of these arteries was ruptured but showed no inflammatory reaction, fibrinoid necrosis or atheromatous changes. This artery was 0.4×1.6 mm in outer diameter (Fig. 5) and had a duplicated internal elastic lamina and slight fibrous intimal thickening. In the right hemisphere, proliferated small muscular arteries were dilated, but unruptured. In old haemorrhagic foci of the right basal ganglia (Fig. 6a) and the right midbrain (Fig. 6b), disrupted small arteries were tortuous, shrunken and occluded by organization. The arteries, depicted in Fig. 1 and schematically in Fig. 8 showed complete occlusion of the left internal carotid (C1 portion), anterior cerebral (A1 portion) and middle cerebral (M1 portion) arteries and severe stenosis of the right internal carotid (C1 portion) and anterior cerebral (A1 portion) arteries. Impressive numbers of small tortuous collateral channels branched from the circle of Willis to bypass occluded segments (Fig. 1). The basilar artery, just proximal to the branching site of the superior cerebellar arteries, tapered to a small fraction of its normal caliber where a short segment of occlusion was noted. It appears, therefore, that circulation in the circle of Willis was mainly derived from the internal carotid arteries, and that collater-

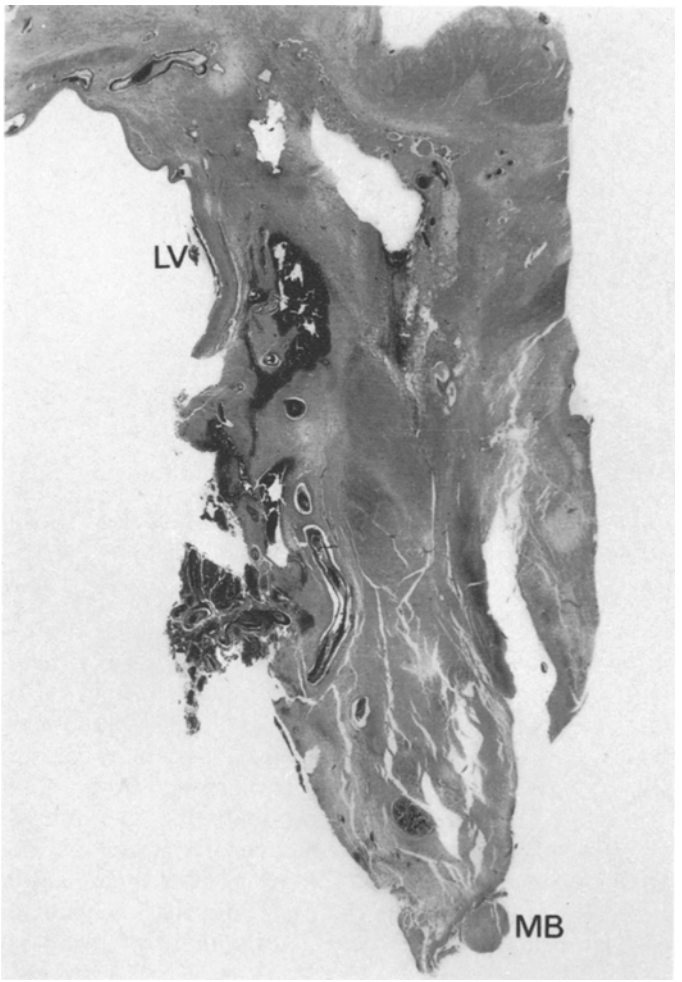

Fig. 4. Close-up view of the coronal section shown in Fig. 3. A group of the muscular-type arteries found in the left thalamus. *MB*, Mamillary body, *LV*, Lateral ventricle. Hematoxylin and eosin stain. ×3

als derived from the circle of Willis supplied the cerebral hemispheres. One of these collateral arteries ruptured, resulting in cerebral haemorrhage with intraventricular extension.

Patient No. 9 had two episodes of unconsciousness at two years and at four months prior to her terminal cerebral accident. At that time she was diagnosed as having Moyamoya disease by carotid angiography. An attack of unconsciousness recurred, followed by aphasia and right hemiplegia. She died shortly after the removal of an intracerebral haematoma and bilateral ventricular drainage. At autopsy, massive haemorrhage (Fig. 2) was found in the left cerebral hemisphere. As in the preceding case, a medium-sized muscular artery (0.8 × 3.7 mm in outer diameter) in the peripheral area of massive haemor-

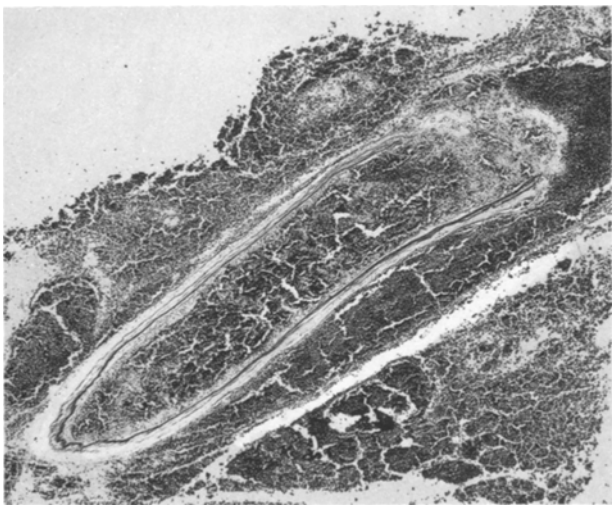

Fig. 5. Microscopic findings of the haemorrhagic lesion shown in Fig. 4. Rupture of the arterial wall (0.4 × 1.6 mm in outer diameter) is found. White thrombus is formed at the ruptured site. Elastic van Gieson's stain. × 35

rhage, displayed a rupture underlying an intraluminal fibrin thrombus. There was no inflammatory reaction, fibrinoid necrosis or atheromatous change.

Patient No. 13 had transient ischaemic attacks (TIA) and died after several months. At that time she was diagnosed as having Moyamoya disease by carotid angiography. At autopsy, massive cerebral haemorrhage was found in the right basal ganglia with extension to the third and lateral ventricles. A ruptured medium-sized muscular artery (0.5 × 2.0 mm in outer diameter) was found in the peripheral area of massive haemorrhage. This artery revealed no inflammatory reaction, fibrinoid necrosis or atheromatous change. Hyperplastic collateral arterial channels, derived from the circle of Willis, were found at the base of the brain.

Collateral arteries (Fig. 7), branching off the circle of Willis, were identified in 13 of 17 patients. These were medium- or small-sized muscular arteries; the majority revealed no pathological changes except for dilatation. Some of them were occluded by loose fibrous connective tissue with wavy internal elastic lamina or by organized thrombus or were stenosed by intimal elastofibrosis. These findings varied from patient to patient; none presented fibrinoid necrosis, granulomatous reaction or inflammatory cellular infiltration.

The stenoses and occlusions of the circle of Willis and its major branches are illustrated schematically in Fig. 8. Histologically, stenoses or occlusions resulted from intraluminal proliferation of loose fibrous connective tissue in associated with variable intimal elastofibrosis (Figs. 9, 10). The conspicuously wavy internal elastic lamina reflected narrowing of the artery.

The anterior cerebral arteries (A1 portion) were bilaterally stenotic in 11 patients, bilaterally occluded in 4 and unilaterally stenotic or occluded in 2. The middle cerebral arteries (M1 portion) were bilaterally stenotic in 8 patients,

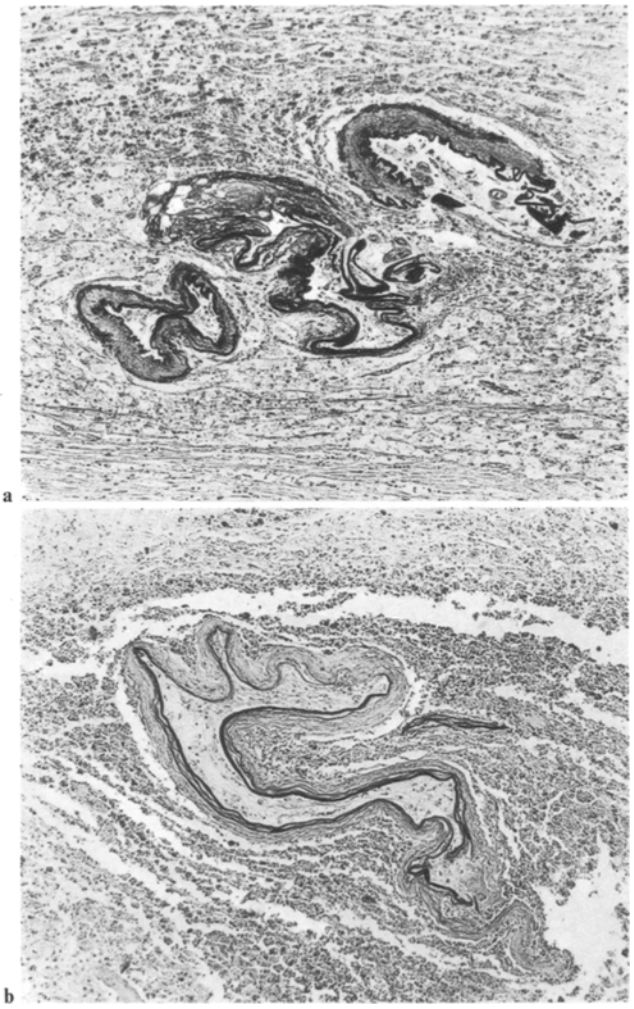

Fig. 6a, b. Microscopic section of the ruptured arteries with organization of the arterial lumen in the old haemorrhagic foci (No. 11). **a** Right basal ganglia, **b** Right side of midbrain. Elastic van Gieson's stain. ×78

bilaterally occluded in 7 and unilaterally occluded in one. Four patients without massive cerebral haemorrhage presented bilateral occlusions of the middle cerebral arteries (M1 portion) in 3, bilateral stenoses in one (M1 portion), and bilateral stenoses of A1 portion in 4. Patients with stenoses of A1 or M1 were more likely to have fresh, massive cerebral haemorrhage than those with complete occlusions of these channels.

Two patients presented saccular aneurysms of the circle of Willis. One (No. 1) died from subarachnoid haemorrhage when a posterior cerebral-posterior communicating artery aneurysm ruptured (Fig. 11). The second (No. 19) had an unruptured saccular aneurysm of a perforating artery.

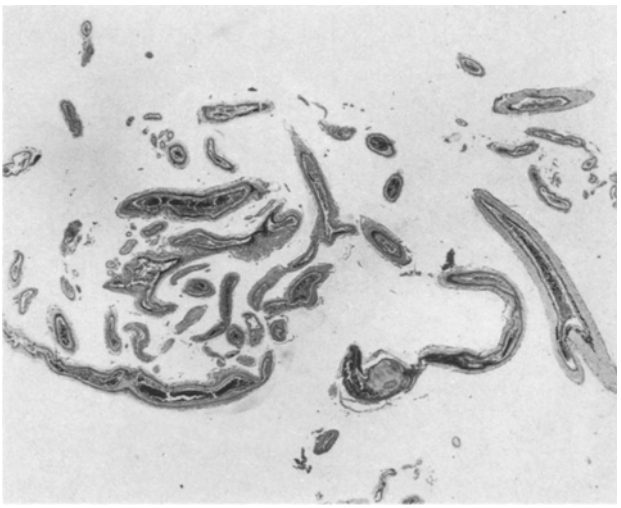

Fig. 7. Microscopic section of the hyperplastic perforating arteries (No. 17). These arteries are small- and medium-sized muscular in type. Some arteries show intimal elastofibrosis or intimal fibro-cellular thickening. Elastic van Gieson's stain. ×10

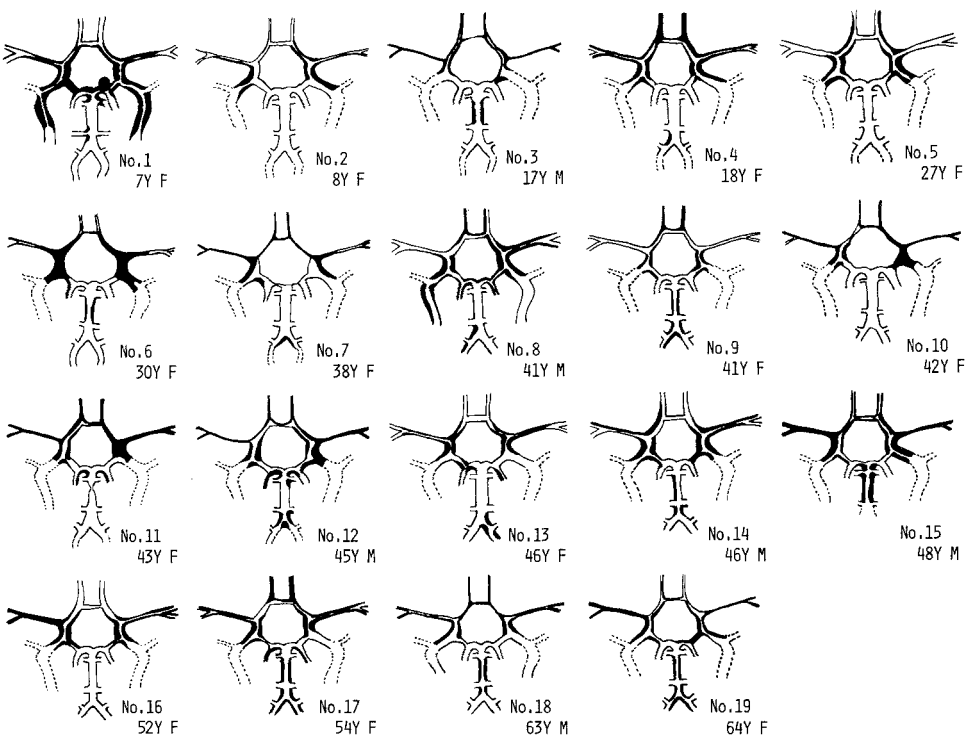

Fig. 8. Schematic illustration of the arterial occlusion in patients with Moyamoya disease. ▬ Occlusion, ═ Stenosis

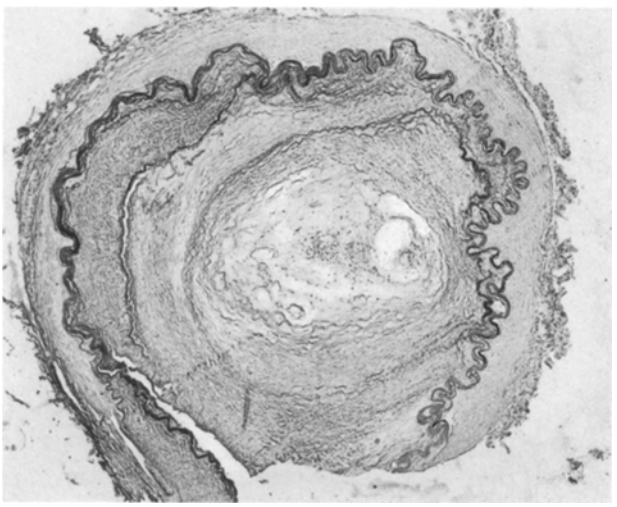

Fig. 9. Microscopic section of the left internal carotid artery (C1 portion) with complete occlusion by laminated fibrocellular thickening of the intima (No. 14). Elastic van Gieson's stain. ×42

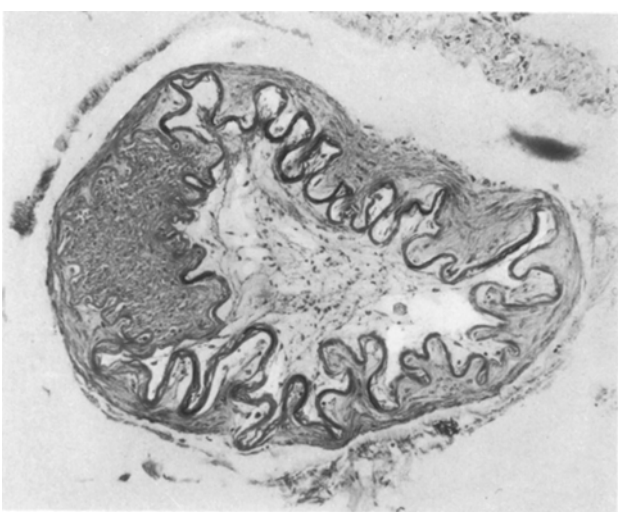

Fig. 10. Microscopic section of the right anterior cerebral artery (A1 portion) with complete occlusion by loose connective tissue and wavy internal elastic lamina (No. 4). Focal proliferation of smooth muscle cells is noted. Marked attenuation of the media is found. There is no inflammatory reaction or necrosis. Elastic van Gieson's stain. ×130

Cerebral infarcts were found in 4 patients. In two patients the infarcts (Fig. 12) involved the distribution of the anterior and/or middle cerebral arteries. The remaining 2 patients presented multiple infarcts about the third ventricle corresponding with the territory of the perforating arteries derived from the circle of Willis and middle cerebral artery. Complete occlusion (Fig. 8) of both

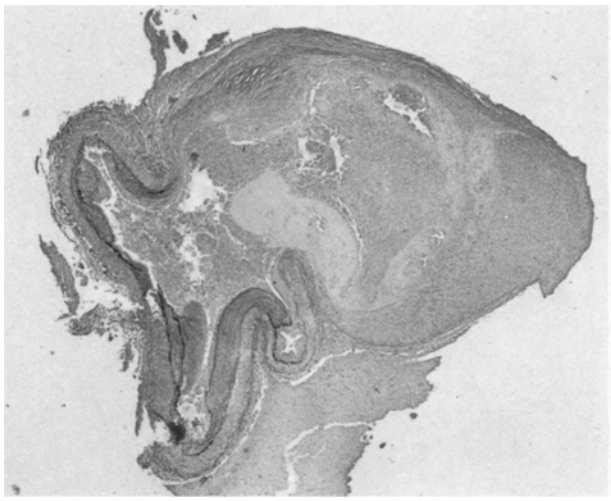

Fig. 11. Microscopic section of ruptured aneurysm of the right posterior cerebral-posterior communicating artery. Laminated recent thrombus is found at the ruptured site. Intimal fibro-cellular thickening is also found. Elastic van Gieson's stain. ×20

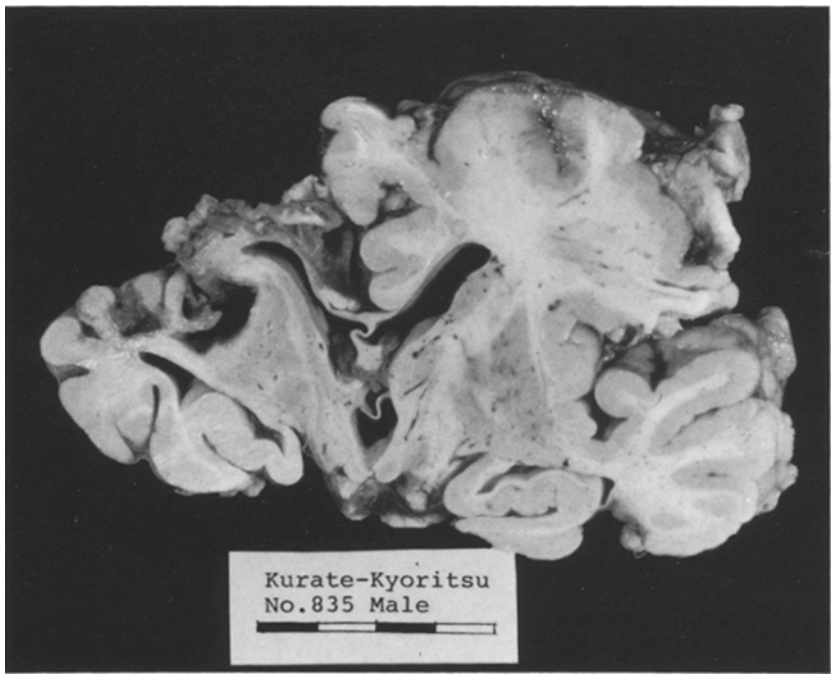

Fig. 12. Coronal section of the cerebral hemispheres at the level of the mamillary bodies (No. 12). The old infarcts in the perfusion areas of the right anterior and middle cerebral arteries are noted

middle cerebral arteries and of the anterior communicating artery was found in patient No. 3, and complete occlusion of the right anterior cerebral artery was observed in patient No. 8. Both of the anterior and middle cerebral arteries of patient No. 12, and both of the middle cerebral arteries of patient No. 15 were completely occluded.

Discussion

Despite numerous reports, little is known about the incidence, distribution, and cause of complicating intracranial haemorrhage in Moyamoya disease. It is said that most adult patients fall victim to sudden subarachnoid haemorrhage; as in the cases in children, however, a few adults presented with transient ischaemic attacks (TIA). The prevalent concept is that subarachnoid haemorrhage results from the rupture of Moyamoya vessels (abnormal net-like mesh of arterial vessels at the base of brain) in the subarachnoid space.

Suzuki et al. (1976) and Kodama et al. (1976) have given enlightening descriptions of the intracranial haemorrhage in this disease. In each of 3 patients, carotid angiograms demonstrated small aneurysms of the distal portion of the posterior choroidal arteries. In each instance, the aneurysmal shadow lay adjacent to the upper lateral edge of the lateral ventricle. As they had disappeared completely on follow-up study angiographically, they were considered to be pseudo-aneurysms indicative of a focus of bleeding to the brain tissue. When haemorrhage occurred, blood could readily penetrate into the adjacent lateral ventricle. It was suggested, therefore, that the sudden clinical onset represent intraventricular haemorrhage.

In the present study of Moyamoya disease, the distribution and cause of the intracranial lesions were investigated at autopsy; 14 of 19 patients showed fresh cerebral haemorrhage. Cerebral haemorrhage developed in the thalamus, hypothalamus and basal ganglia and intraventricular extension was found in 12 patients. In 2 patients the haemorrhagic foci extended through the white matter of the frontal, parietal and temporal lobes, reaching the cerebral cortex. There were no cases of primary subarachnoid haemorrhage due to rupture of Moyamoya vessels. Our findings would indicate that intracranial haemorrhage in Moyamoya disease is primarily intracerebral and that the secondary subarachnoid haemorrhage follows intraventricular extension or direct penetration of the cortex.

The arterial supply of the thalamus, hypothalamus, basal ganglia, midbrain and cerebral peduncle is derived from the premamillary arteries, the perforators from the posterior communicating and posterior cerebral arteries, the thalamoperforating arteries, the anterior choroidal arteries and the lateral striate arteries; all of these arteries branch from the circle of Willis (McCormick and Schochet 1976). In 13 patients, a meshwork of overgrown and dilated small- to medium-sized muscular arteries, branching from the circle of Willis, were demonstrated. From their location, these collateral arteries are clearly derived from the perforating branches. as previously suggested by Kodama (1971).

Disrupted medium-sized muscular arteries were found within the massive haemorrhagic foci in 3 patients. We could not exclude the possibility that these

arteries were overgrown perforators developed as collaterals. Furthermore, disrupted and organized small arteries in an old haemorrhagic focus in one patient suggested that collaterally developed perforating arteries might rupture repeatedly. To our knowledge, these changes in the overgrown perforating arteries have not been previously reported.

Massive cerebral haemorrhage of hypertensive origin usually arises from small arteries (100–200 μ) in the basal ganglia, thalamus and cerebral gray matter which have undergone fibrinoid necrosis (plasmatic angionecrosis) and developed microaneurysms (Ooneda 1974). In our patients, the ruptured arteries, devoid of necrosis, were larger in size than those in hypertensives, and showed only slight fibrous intimal thickening. This evidence indicates that the mechanism of cerebral haemorrhage in Moyamoya disease is different from that in systemic hypertension. It is possible, though not established, that rupture of the overgrown and dilated collateral channels results from rapid fluctuation of intracerebral arterial pressure.

In the present study, only one patient showed subarachnoid haemorrhage, due to a ruptured saccular aneurysm of the circle of Willis, but no rupture was present in the overgrown collateral channels in the subarachnoid space. Previously, Kodama et al. (1977) had observed haemorrhage from a ruptured aneurysm of the basilar artery in 35 year-old-man, and Pool et al. (1967) reported an unruptured aneurysm of the right middle cerebral artery in a seven month-old-girl with Moyamoya disease.

Cerebral infarcts were found in 4 patients. Their location in Moyamoya disease was not always in the territory of the anterior or middle cerebral arteries but also involved foci about the third ventricle. The numerous collateral dilated perforators supplied these areas. Their occlusion would produce cerebral infarcts not only in the territory of the anterior or middle cerebral arteries but also in the original territory perfused by the perforators. Our observations support the view that hyperplastic collateral perforators play an important role in the development of cerebral haemorrhage or cerebral infarcts in Moyamoya disease and result from slowly growing stenosis and occlusion of the distal ends of the internal carotid arteries and proximal portions of the anterior and middle cerebral arteries.

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