

## Moyamoya Disease and Intracerebral Hematoma

### Clinical Pathological Report

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**Summary.** This report concerns a hypertensive woman who suffered a stroke at the age of 51 and recovered from left hemiparesis after 3 to 4 months. During the subsequent 24 years she had four seizures which involved the left arm and face and became generalized, ending with left hemiparesis, from which she recovered after 4 to 5 days. Carotid angiography was performed in 1967, 1973 and 1974 and the characteristic picture of moyamoya disease was demonstrated. She died at the age of 77 with extensive vascular disease. The literature concerning 215 cases of moyamoya disease, in which there were 14 intracerebral hematomas, is reviewed and discussed.

**Key words:** Moyamoya disease – Intracerebral hematoma

### Case Report

A woman, aged 51, suddenly experienced headache, nausea and vomiting. On examination she was comatose and had a left hemiplegia. The past medical history included alcoholism, syphilis treated at the age of 27, and arterial hypertension discovered at 45 years. The patient slowly recovered in 3 to 4 months.

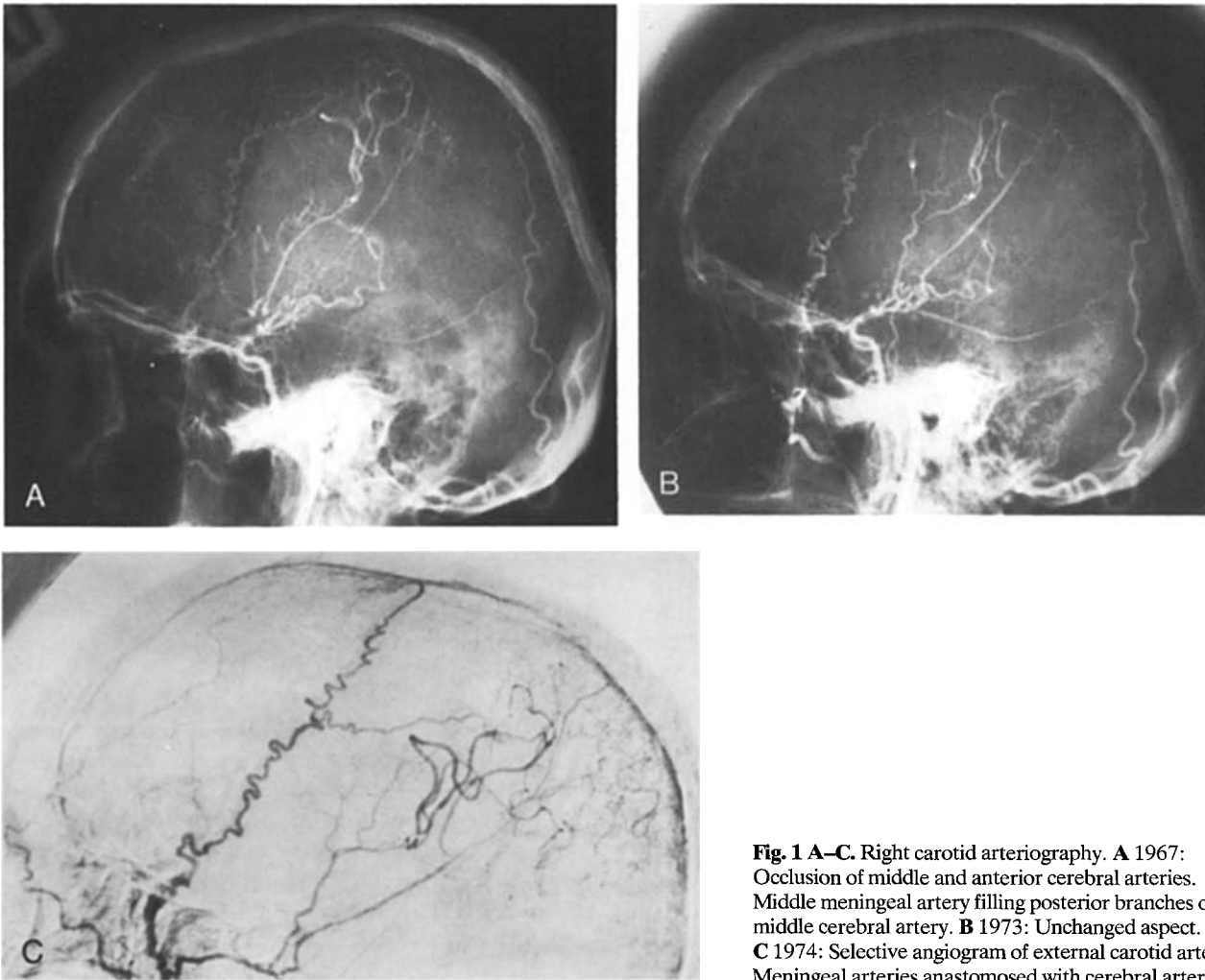
Over the following 24 years she had four Jacksonian episodes beginning in the left arm and face and followed by a generalized seizure with residual left hemiplegia which would completely resolve after 4-5 days. The blood pressure was 200/130 (right) and 170/100 (left).

Examination carried on after recovery disclosed brisk stretch reflexes on the left and left visual extinction. VDRL was negative. The EEG showed paroxysmal slow waves over the right parietotemporal area. From 76 years on, she had various neurological

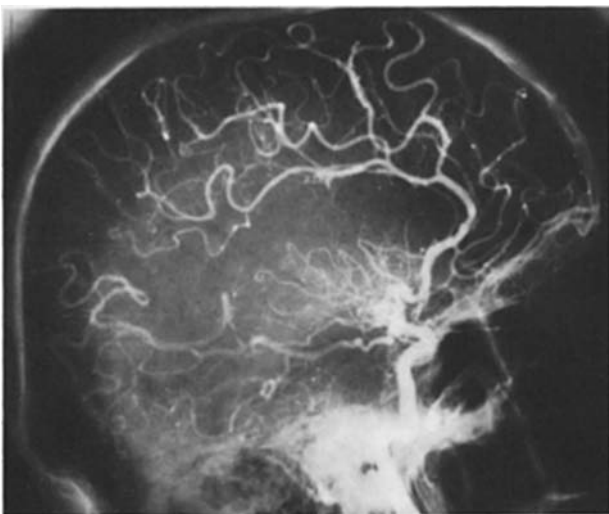
accidents resulting in permanent signs: right motor impairment with bilateral pyramidal signs; left sensory loss and choreoathetotic movements; left hemianopia; partial left third nerve palsy. Swallowing difficulties led to her death in 1975, aged 77, after 25 years of illness.

Serial carotid angiograms were obtained in 1967, 1973 and 1974 (Fig. 1). The initial aspect never changed. On the right side the internal carotid artery was hypoplastic and had a further narrowing beyond the origin of the ophthalmic artery. Only an enlarged anterior choroid artery was filled from the carotid. A large collateral arterial network was seen at the base of the brain. The middle meningeal artery was large and tortuous and filled several rolandic and parietal branches of the middle cerebral artery. An additional selective examination of the external carotid artery was performed in 1974 (Fig. 1) and confirmed the retrograde filling of the middle cerebral artery from a very large middle meningeal artery. The left internal carotid artery had the same hypoplastic but otherwise normal aspect. It filled a very large and tortuous anterior cerebral artery and an extensive basal arterial network (Fig. 2). The posterior cerebral artery had an eccentric, distal narrowing of its lumen.

At autopsy, except for severe bronchopneumonia, the pertinent gross findings were limited to the brain and intracranial arteries. The brain weighed 900 g. After formaldehyde fixation, the cerebrum was examined by coronal sectioning and the cerebellum, pons and midbrain by transverse sectioning. An old cystic hematoma was found in the right external capsule (Fig. 3), extending laterally and caudally into the white matter of the superior temporal gyrus and inferior parietal gyri. The territory of the right posterior cerebral artery was destroyed by a pale softening. On the right side, gross examination of the cerebral vessels (Fig. 3) revealed that the supraclinoid



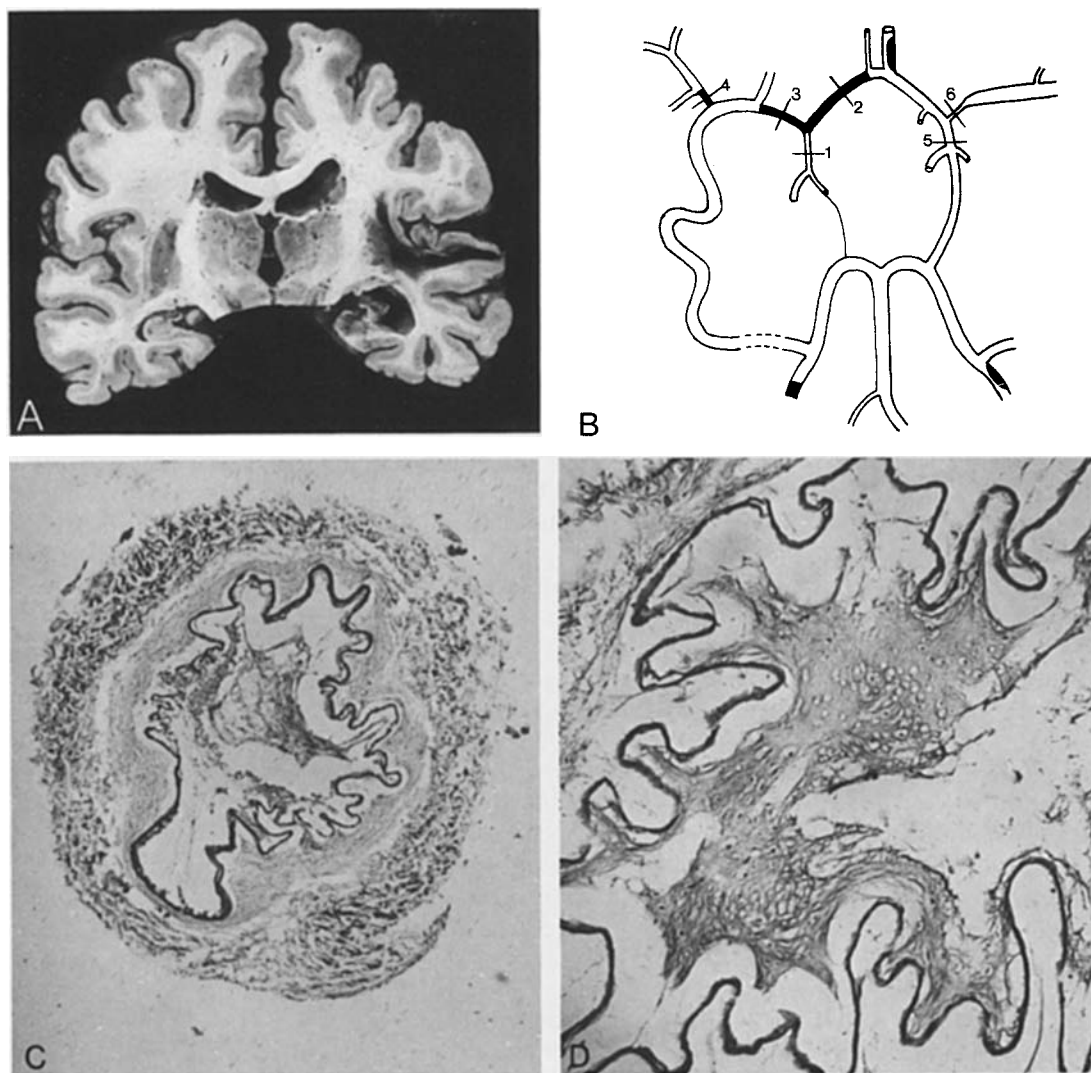
**Fig. 1 A-C.** Right carotid arteriography. **A** 1967: Occlusion of middle and anterior cerebral arteries. Middle meningeal artery filling posterior branches of middle cerebral artery. **B** 1973: Unchanged aspect. **C** 1974: Selective angiogram of external carotid artery. Meningeal arteries anastomosed with cerebral arteries



**Fig. 2.** Left carotid arteriography (1973). Occlusion of the middle cerebral artery and basal arterial network

portion of the carotid artery, the proximal portions of the anterior cerebral artery and of the middle cerebral artery were narrowed and occluded. The middle cerebral artery reached its normal size 1 cm beyond its origin and joined the right posterior cerebral artery by an enlarged temporal branch on the ventral aspect of the temporal lobe.

The supraclinoid portion of the left carotid artery and the proximal left middle cerebral artery were narrowed, but patent; the left anterior cerebral artery was normal. The anterior communicating artery was slightly widened. The vertebral, basilar and posterior cerebral arteries were of normal size and had severe atheromatosis. The right posterior cerebral artery was occluded distal to the widened temporal branch already mentioned. On cross-section the lumen was occluded by a thrombus on the surface of an atheromatous plaque. In the right parietal area the internal aspect of the dura had an unusual network of



**Fig. 3.** **A** Coronal section through red nuclei: intracerebral hematoma of right external capsule extending into white matter of T1. Old pale softening in distribution of right posterior cerebral artery destroying Ammon's horn. **B** Vascular lesions. 1. Right internal carotid artery; 2. Right anterior cerebral artery; 3. Right middle cerebral artery; 4. Posterior branches of the right middle cerebral artery; 5. Left internal carotid artery; 6. Left middle cerebral artery. **C** Right middle cerebral artery  $\times 35$  Masson trichrome. Occlusion by intimal proliferation. Abnormal internal elastic lamina. Medial and adventitial atrophy. **D** Right anterior cerebral artery  $\times 250$  trichrome Masson. Hyalin aspect of intimal proliferation. Abnormally split or interrupted internal elastic lamina

arterial vessels related to the anastomosis between the right middle meningeal artery and the distal branches of the right middle cerebral artery.

### Microscopic Examination

Samples of the vessels were embedded in paraffin, cut and stained with orcein, hematoxylin and eosin, Masson trichrome, periodic acid Schiff. The vertebral, basilar and posterior cerebral arteries had severe atheromatous lesions. The distal internal

carotid arteries, the proximal middle cerebral arteries and the right anterior cerebral artery showed quite different lesions (Fig. 3). Their external diameter and lumen were reduced in size. All layers of the arterial wall were involved. A marked intimal hyalin proliferation narrowed or occluded the vascular lumen. The internal elastic lamina was altered, split and broken up; some sections did not stain with orcein. The media had lost its muscular fibers, the adventitia was very thin. There were neither inflammatory signs, nor atheromata of these vessels.

**Table 1**

	Reference	Age	Diagnosed by
Higashi	[2]	28	Surgical evacuation
Hori	[4]	unknown	Surgical evacuation
Iilvainen	[5]	28	carotid angiography
Lee	[7]	33	Surgical evacuation
		35	Surgical evacuation
Mauersberger	[8]	40	Pneumoencephalography
Nishimoto	[9]	10	Pathological report
Picard	[12]	47	Pathological report
		45	–
Smith	[13]	53	Pathological report
Suzuki	[14]	child	Pathological report
		child	–
		child	–
Vuia	[16]	44	Pathological report

## Discussion

Our patient fulfilled all the clinical and arteriographic criteria of moyamoya disease in an adult. After her initial spontaneous cerebral hemorrhage, she had several epileptic seizures and regressive neurological deficits. This condition allowed normal survival over 23 years in spite of severe arterial hypertension. Angiography disclosed bilateral (although asymmetrical) involvement, i. e., stenosis of the supraclinoid internal carotid arteries; occlusion of the proximal right and left middle cerebral arteries and of the right anterior cerebral artery; an enlarged network of vessels in the region of the basal ganglia; a collateral circulation between the external carotid and leptomeningeal branches of the internal carotid across the arachnoid and the dura.

The pathology of moyamoya disease has no widely accepted features. However, we would like to point out that the abnormal vessels displayed neither inflammatory disease, nor advanced atherosclerosis, at least in the anterior half of the circle of Willis. Some particular points prompted us to report this case: the long survival of this patient 25 years after the first sign, in spite of arterial hypertension; the repeated unchanging arteriograms over 7 years (1967–1974); the few cases of intracerebral hematoma mentioned in earlier publications; the rare and inconclusive autopsy reports about moyamoya disease; the coexistence of hypoplastic arteries in the territory of the carotid artery and of definitely atheromatous arteries in the branches of the basilar artery.

The reported intracerebral hematomas in moyamoya disease are unexpectedly scarce, considering that the fragile thin walled vessels in this condition

might be expected to rupture very easily. Subarachnoid hemorrhage has been found in many of 215 cases reviewed, with only 14 intracerebral hematomas (Table 1). Subdural hematomas were not included.

There are three pathological reports concerning adults. Picard [12] found a hematoma of the right centrum semiovale ruptured into the lateral ventricle. He pointed out the disruption of the internal elastica and connective tissue replacing the media in the anterior choroidal artery.

Smith [13] described a freshly clotted hematoma  $5 \times 4 \times 3$  cm destroying the right centrum semiovale, thalamus and part of the lentiform nucleus, as well as part of the internal and external capsules and claustrum. The abnormal internal carotid arteries had a thickened intima and segmentally located atheroma, while anterior and middle cerebral arteries were small in cross-section but otherwise unremarkable.

An unusual feature of moyamoya disease is narrowing of the internal carotid artery 2 to 3 cm above its origin. Smith considered his findings to be related to congenital hypoplasia of both carotids.

Vuia [16] reported a right frontal hemorrhage ruptured into the lateral ventricle and abnormal intracranial vessels. The supraclinoid carotid arteries were characterized by intimal proliferation, thin media and well developed elastica interna. Proximal anterior and middle cerebral arteries contained marked hypoplasia and intimal proliferation. Small leptomeningeal vessels exhibited poor elastica interna. The patient's condition had not permitted arteriography.

The reports are not all dealing with definite moyamoya disease in the adult. Nishimoto [9] reported a fourth case concerning a girl aged 10, with posttraumatic intracerebral hematoma and sequellae of tuberculous meningitis distorting the anatomical findings.

Under the diagnosis of multiple progressive intracranial occlusions, Taveras [15] described a similar condition but only one of his 10 cases had intracerebral hematoma. In a recent review Nishimoto [10] claims the congenital origin of the disease. Several other publications [1, 3, 11] emphasize the possible lack of specificity of the arterial lesions leading to a moyamoya pattern of collateral vessels.

It is debatable whether the anatomical abnormalities of our patient's vessels had some specificity. The lesions observed can be found in several distinct conditions and particularity in the aged [6]. However, in our patient, atheromatosis was found in vessels of a size equal to those obstructed or narrowed by the

moyamoya process. Only further anatomical reports will make it possible to assess the specificity or the heterogenous nature of this disease.

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