Cerebrovascular moyamoya disease associated with an intracranial pseudoaneurysm

Case report

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A 51-year-old woman became unconscious 19 hours after the onset of a headache. Computerized tomography disclosed an intracerebral hematoma in the left temporal lobe, with ventricular penetration. Angiography demonstrated the characteristic appearance of cerebrovascular moyamoya disease as well as an aneurysm-like shadow in the left temporal lobe, which proved on histological examination to be a pseudoaneurysm.

KEY WORDS • cerebrovascular moyamoya disease • intracerebral hematoma • pseudoaneurysm

Occasional cases of cerebrovascular moyamoya disease associated with an intracranial aneurysm have been reported. The aneurysms have been described as saccular, fusiform, or pseudoaneurysms. To our knowledge, this is the first report of a pseudoaneurysm verified histologically in a patient with moyamoya disease.

Case Report

This previously healthy 51-year-old woman experienced a sudden attack of headache on July 11, 1979. Nineteen hours later, while working as a caddie, she became unconscious. She was taken to another hospital, where a spinal tap yielded bloody cerebrospinal fluid. She was immediately transferred to Kagoshima Municipal Hospital.

Examination. On admission, her vital signs were normal, and general physical examination was unremarkable. She was unconscious but responded to painful stimuli. She had a right hemiparesis and a stiff neck.

Computerized tomography showed an intracerebral hematoma in the left temporal lobe with ventricular penetration (Fig. 1). Left carotid angiography showed that the internal carotid artery was occluded just above the origin of the anterior choroidal artery. The posterior communicating and cerebral arteries were large. Many fine vessels were noted at the base of the
Sylvian fissure was opened and the intracerebral hematoma was reached through a corticotomy at the medial aspect of the temporal lobe. A small, firm, dark red aneurysm-like body was discovered within the hematoma, and was extirpated when no parent artery was identifiable. After removal of the hematoma, external ventricular drainage was instituted, and the bone flap was discarded for external decompression.

**Postoperative Course.** The patient's neurological status remained unchanged. Sixteen days after the operation a right ventriculoperitoneal shunt was inserted for treatment of progressive hydrocephalus, complicated by meningitis. Her condition progressively deteriorated and she died on October 29, 1979. An autopsy was not performed.

**Pathological Examination.** Histological examination showed that the wall of the lesion was composed of concentric layers of fibrin with red blood cells between them (Fig. 5). An elastica-Masson stain demonstrated no elastic fibers. The pathological diagnosis was a hematoma.

**Discussion**

Subarachnoid hemorrhage (SAH) is the most common initial symptom in adult patients with cerebrovascular moyamoya disease. The generally accepted hypothesis as to the mechanism causing SAH is that it arises from rupture of the moyamoya vessels in the subarachnoid space. Lee and Cheung\(^5\) pointed out that the mortality rate associated with SAH in cases of cerebrovascular moyamoya disease was unusually high.
Moyamoya disease and pseudoaneurysm

low compared with that associated with bleeding from berry aneurysms. They ascribed the difference to the small size of the moyamoya vessels. Kodama and Suzuki\(^4\) reported three cases of cerebrovascular moyamoya disease associated with an aneurysmal shadow in the peripheral portion of the posterior choroidal artery. They presumed it to be a pseudoaneurysm because it was no longer visible on the follow-up angiogram. They suggested that the SAH might be due to penetration of the intracerebral hematoma into the ventricle. Okuma, \textit{et al.},\(^9\) described a case of cerebrovascular moyamoya disease in which an aneurysm shadow was visualized at the peripheral portion of Heubner’s artery. The lesion ruptured during angiography and resulted in SAH. They postulated that it was a pseudoaneurysm formed at the site of the rupture of Heubner’s artery, and that the initial SAH was caused by penetration of the intracerebral hematoma from the rupture of the artery into the third ventricle. In 1967, Cole and Yates\(^2\) described two types of small masses found within, or close to, a massive intracerebral hemorrhage: Type I consisted of perivascular blood clot, with the outer surface formed by a smooth layer of fibrin; and Type II appeared as a beaded distension by blood of the perivascular space. These authors suggested that both types might be the result, and not the cause, of the severe hemorrhage. Our case seemed to be consistent with their Type I.

There are a few cases in which a ruptured saccular aneurysm was identified at surgery or at autopsy in a patient with cerebrovascular moyamoya disease.\(^1,4,8\) Adams, \textit{et al.},\(^1\) recommended that all patients with

![Fig. 4. Lateral view of a right retrograde brachial arteriogram with an appearance similar to Fig. 3. Slight narrowing of the distal portion of the basilar artery can be seen.](image)

SAH and moyamoya disease should be carefully investigated for a possible aneurysm, and believed these patients should be treated as if the aneurysm were the source of bleeding.

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\textbf{References}


![Fig. 5. Photomicrograph of the aneurysm-like body demonstrating the wall to be composed of concentric layers of fibrin with red blood cells between them. H & E, × 3.25.](image)
rupture during cerebral angiography and spontaneous regression of the aneurysm. No Shinkei Geka 8: 181-185, 1980 (Jpn)

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