Pulseless disease associated with multiple intracranial aneurysms

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Summary. A case is presented of a 64-year-old woman with pulseless disease associated with multiple intracranial aneurysms which were located in the internal carotid circulation at the left internal carotid-ophthalmic artery and the left internal carotid-posterior communicating junction, and in the basilar-vertebral circulation, at the left basilo-anterior inferior cerebellar artery junction and at the top of the basilar artery. Autopsy verified that her death was due to rupture of the aneurysm at the top of the basilar artery. From the cerebral angiographic and autopsy findings, it can be presumed that overloaded intracranial hemodynamic forces played some role in the development of the aneurysms. The present case is discussed together with our three previously reported cases.

Key words: Pulseless disease – Aortitis syndrome – Takayasu’s arteritis – Intracranial aneurysm – Aneurysmal rupture – Multiple aneurysms

Only a few reports are available concerning ruptured intracranial aneurysms during the course of pulseless disease. Eight cases have been reported [1–6], including three from our previous reports [5, 6]. In this paper we present another such rare case, encountered in a 64-year-old woman who showed the initial symptom of subarachnoid hemorrhage, and discuss some mechanisms of the development of intracranial saccular aneurysm, based on our own four cases.

Case report

A 64-year-old woman had been diagnosed as having hypertension and cardiac hypertrophy seven years previously, but did not receive any medical treatment.

On August 6, 1984, while bathing, she experienced the sudden onset of severe headache together with motor weakness of a few minutes’ duration in both legs. The episode was accompanied by frequent nausea and vomiting, but she remained conscious. She was immediately taken to an emergency hospital, where a computed tomographic scan (CT scan) revealed high-density areas in both basal and sylvian cistern and in the subarachnoid space over the brain surface. The subarachnoid hemorrhage was confirmed by lumbar puncture. On the following day, the patient was transferred to our clinic for further evaluation.

On admission (August 7), she complained of severe headache with nuchal rigidity, and responded to simple questions without disorientation or memory disturbance. The pulsation of the left radial artery was weak (100/min, regular rhythm), but the blood pressure was 170/90 mmHg in the right upper extremity.

Examination of the blood disclosed hemoglobin 13.1%, hematocrit 40.4%, red blood cells 4.76 million/cu mm and white blood cells 15,300/cu mm. The erythrocyte sedimentation rate was 85 mm/hr, and the serum C-reactive protein was 3+. STS was negative in the blood and in the cerebrospinal fluid.

Roentgenography of the skull was normal. A plain CT scan at the emergency hospital showed high-density areas in the basal cisterns and the superficial subarachnoid space (August 6) (Fig. 1a and b), and another CT scan taken on the occasion of aneurysmal re-rupture (September 9) (Fig. 2a and b) presented high-density areas in the basal cisterns and the ventricular system caused by reflux of the subarachnoid hemorrhage through the fourth ventricle, resulting in the formation of a hydrocephalic state. A ⁹⁹ᵐTc-MAA lung perfusion scan demonstrated lack of radionuclide activity in the right middle and upper lobe. A plain chest CT scan indicated a saccu-
Fig. 1a and b. Plain CT scan at the first attack of subarachnoid hemorrhage (on August 6). CT scan shows high density in the basal cistern and subarachnoid space.

Fig. 2a and b. Plain CT scan on the occasion of aneurysmal re-rupture (on September 9). CT scan shows high density in the basal cistern and in the ventricular system, and the formation of hydrocephalus.

Fig. 3. An oblique subtraction view of an aortogram of a 64-year-old woman with pulseless disease. The right common carotid artery and the left subclavian artery are completely occluded. The left common carotid artery (big arrows) are dilated. The right vertebral artery (arrow heads) and the right subclavian artery (small arrows) are patent.

Thoracic aortography (Fig. 3)

Figure 3 shows complete occlusion of the right common carotid artery and the left subclavian artery at their origin. The occlusion of the latter precluded visualization of the left vertebral and internal thoracic artery. Cerebral blood supply was maintained only through the left internal common carotid and the right vertebral arteries. The margin of the thoracic aorta was irregular with exuberant calcification. At the origin of the bronchial artery, an irregular, saccular shaped aneurysm was opacified.

Left carotid arteriography (Fig. 4a, b)

The right anterior and middle cerebral arteries were opacified from the left internal carotid artery through the anterior communicating artery, which was the major blood supply route to the right hemisphere. In the left internal carotid arteriography, three aneurysms of the saccular type were visualized: at the origins of the posterior communicating artery, of the left ophthalmic artery and at the top of the basilar artery which was opacified via the dilated left posterior communicating artery. It could not be determined, however, which of these aneurysms had ruptured.

The left external carotid arteries were well developed with some tortuosity. In the late arterial phase, the left vertebral artery was opacified through the collateral circulation: the occipito-vertebro-basilar anastomosis.

Following aneurysmal re-rupture (on September 9), the patient's condition deteriorated, and she died on September 13. Autopsy was performed.

Autopsy

The right common carotid artery was completely obstructed at its origin, but the right vertebral artery was patent. The left common carotid artery was di-