An Autopsy Case of Arteriovenous Aneurysm of the Brain

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The direct communication between the arterial and venous systems exists either as an acquired lesion chiefly due to trauma or as a congential malformations. Abnormal arteriovenous communications of the brain have been variously designated by many authors, but the term "arteriovenous aneurysm" seems to be commonly used nowadays.

The arteriovenous aneurysms of the brain are comparatively rare lesions, but they are clinically important racemose malformations of the blood vessels of the brain. In case of deeply situated aneurysms or of remarkably large ones, complete excision of the lesion is impossible and not uncommonly terminated to fatal hemorrhage or extensive damage to the brain.

We recently experienced a case of large arteriovenous aneurysm of the brain, in which the tangled mass of the blood vessels completely replaced the basal ganglia and resulted in fatal subarachnoid hemorrhage.

REPORT OF A CASE

Clinical History:

A 40 year old man (farmer) was admitted to the 2nd medicine of Yamaguchi University Hospital on February 24, 1965, with the chief complaint of right hemiplegia and occasional epistaxis.

The patient was well until the age of twelve, when he gradually developed right hemiplegia. About three years later, right hemiplegia spontaneously disappeared. But when he was 16 year old, he began to have ocassional epistaxis and right hemiplegia developed again and gradually increased in its severity. About six months prior to the admission, he noticed painless soft pulsating swelling at the forehead and nasal dorsum, and he also began to suffer from diminution of vision.

No other members of the family are similarly affected.

Physical Examination:

The patient was well-nourished man who complained of right hemiplegia with disturbances in gait. His blood pressure was 137/7o. The head was normal in size and shape. On the left side of the forehead, pea-sized pulsating mass was noticed. The region of the dorsum nosi was also swollen. Along the left eye brow, dilatation of the blood vessel was seen. A characteristic bruit was audible over the head and neck.

The ocular movements were normal, but there was right homonyme hemianopsia. Fundoscopic examination showed very white discs on both sides and moderate sclerotic changes of the retinal arteries.

Examination of the nose revealed deviation of the nasal septum to the right and bleeding from the nasal mucous membrane of the left nasal cavity.

The lung was clear. The heart was slightly enlarged to the left. The liver was palpable three finger-breadth below the costal margin and was non-tender. The muscle of the right upper and lower extremities were moderately atrophic.

On neurological examination, right hemiplegia with hyperactive knee reflex and positive Babinski reflex on the right side was revealed. His mental state and intelligence were almost normal.

Laboratory Examination:

Studies of the blood disclosed 326×10^4 red cells and 2800 white cells. Hemorrhagic studies were within normal limits. Examination of the blood chemistry revealed hemoglobin 6.0g/dl, hematocrit 19.8%, serum iron 32γ/dl, serum protain 7.1g/dl with albumin/globulin ratio of 1.09, icteric index 5, blood sugar 67 mg/dl, CCFT 2, cholinesterase $0.5_{\triangle}$ph, alk. phosphatase 2.0u, cholesterol 87 mg/dl, NPN 25 mg/dl, urea N 13 mg/dl and GPT 2 u. Examination of the sternal bone marrow was also diagnostic of iron deficiency anemia. The Wasserman reaction of the blood was negative. Urinalysis was normal.

The electrocardiogram revealed left ventricular hypertrophy.

The chest film was normal except for enlargement of the heart silhouette.

Rentogenograms of the skull revealed small, faintly spotted area of calcification (Fig. 1).

Cerebral angiography with injection of 13 ml. of urographine in the left common carotid artery demonstrated the tangled mass of the blood vessels located above the sella trucica (Fig. 2 and 3). The findings of angiography were characteristic of the arteriovenous aneurysm. The left ophthalmic artery was remarkably dilatated and the left orbital region was highly vascular.

Hospital Course: During hospitalization, he occasionally had epistaxis, which continued 30 to 60 minutes and subsided spontaneously. For the treatment of iron deficiency anemia, hematinic drugs were administered. He was occasionally

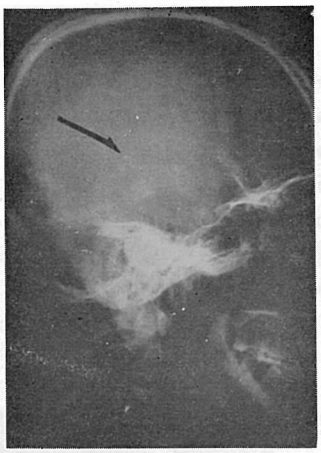

Fig. 1. Lateral rentogenogram of the skull showing fainily spotted area of calcification. (arrow).

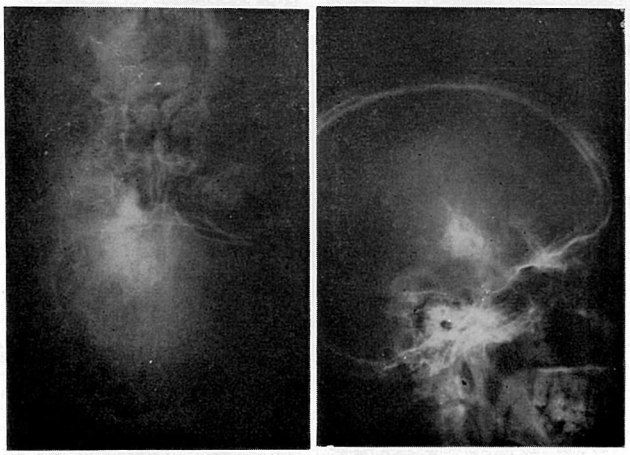

Fig. 2 and 3. Anteroposterior and lateral carebral angiography by injection of urographine in the left common carotid artery. Note the tangled mass of the blood vessels and the markedly dilated ophthalmic artery.

injected hemostatic agent to prevent epistaxis.

On the 24th hospital day, he suddenly became comatose and developed convulsion of the extremities. At that time his blood pressure was 210/120. Pathological reflexes were evident. He increasingly became deep comatose and died within an hour, showing the clinical symptoms of cerebral bleeding.

Postmortem Examination:

Postmortem examination was performed four hours after the death and was

limited to the brain.

The skull was normal in size and shape. Remarkable subarachnoid hemorrhage at the basis of the brain was disclosed and massive coagulated blood was revealed in the ventricles. A typical arteriovenous anerysm, which consisted of a tangled mass of the blood vessels, both arteries and venins, was found in the middle of the left cerebral hemisphere (Fig. 4). The aneurysm was remarkably large and completely replaced the left thalamus, basal ganglia and a part of the basal portion of the left temporal lobe. Because of extensive subarachnoid hemorrhage, careful examination failed to demonstrate the circle of Willis exactly. But the arterial side of aneurysm appeared to have developed from the anterior portion of circle of Willis in relation to the left internal carotid artery. The ophthalmic arteries

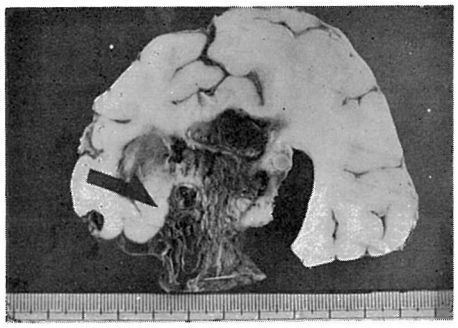

Fig. 4. A large mass consisted of the blood vessels (arrow) completery replaces the basel ganglia and diencephalon, protruding into the lateral ventricle. The ventricular cavity is filled with coagulated blood.

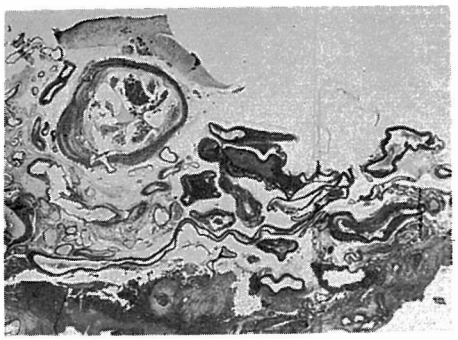

Fig. 5. The vessels in the lesion vary remarkably in their sise and thickness. In the lower field, area of hemorrhage is seen. (Weigert stain)

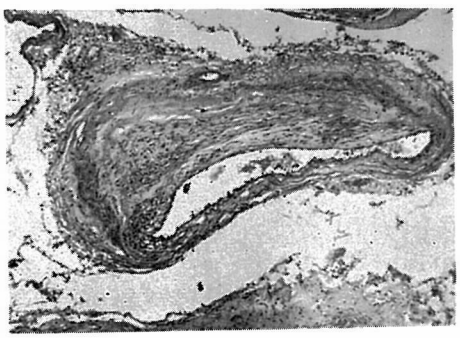

Fig. 6. A single artery in the lesion. The tickness of the wall is markedly irregular even in the same vessel because of partial hyperplasia of the intima. (H.E. ×32)

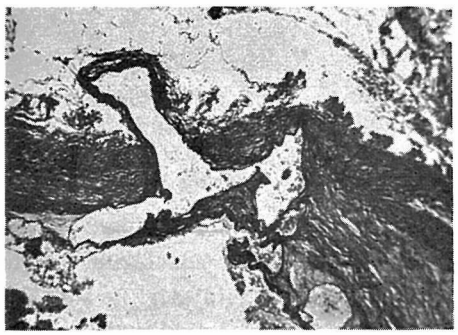

Fig. 7. Irregularity of the blood vessel showing characteristic angulation. (weigert stain. ×32)

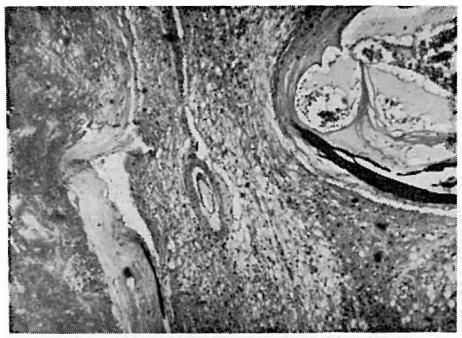

Fig. 8. Calcium deposit in the wall and formation of thrombus (at the left). Between two large vessels, degenerated brain tissue is recognized. (H. E. ×32)

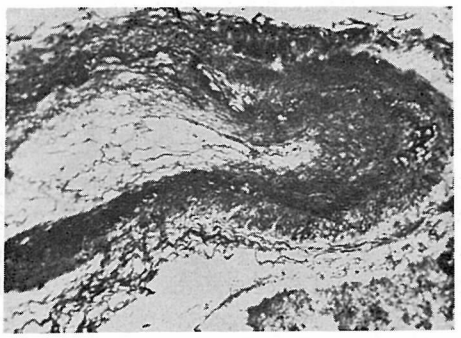

Fig. 9. The elastic fibers are moderately thick and wavy. (Weigert stain. ×32)

were also enlarged in size and the optic nerves were atrophic on both sides. The soft mass on the forehead, though not so evident at the time of autopsy, was consisted of the dilated blood vessels.

Microscopically, the vessels which consisted of the aneurysm varied greatly in size and thickness (Fig. 5). Most of the wall of the vessels revealed hyperplastic intima and marked hyalinization with occasional formation of thrombus, and furthermore the thickness of the wall was remarkably varied even in the same vessel (Fig. 6). Corresponding to the tortuosity, there was a characteristic angulation and infolding of the vessel walls, occasionally resulting in an almost papillomatous appearence (Fig. 7). Calcium deposit was found in the vessel walls and in the supporting connective tissue adjuscent to the vessels (Fig. 8). In the arteries, the elastic fibers were moderately thick and wavy, sometimes showing fragmentation (Fig. 9). The mass on the forehead was also consisted of irregularly dilated artery, the wall of which showed almost the same findings as was seen in the aneurysm. The optic nerves were atrophic and revealed degenerative changes. Petechial hemorrhage was recognized in the mid-brain.

COMMENT

Because of the great variability of the appearence and structure of the lesions, there are numerous terminology as to the angiomatous malformation of the brain. Olivecrona and his associates[1] divided them into two main groups, the cavernous and the racemouse angiomas. Cushing and Baily divided them into the venouse and arterial groups. Dandy[2] prefered to call all of these lesions simply arteriovenous aneurysms. This term seems to be used more commonly nowadays and to leave no confusion about the nature of the lesion.

The arteriovenous aneurysms of the brain have the possibility to result from in a similar manner to that of traumatic aneurysm. In the extremities, such an acquired type of arteiovenous aneurysm are common, but in the brain most of them arise as congenital origin. Abnormal communications between intracranial vessels external to the brain occur chiefly in the internal carotid artery and the cavernous sinuses. In a mojority of reported cases, the vascular anormalies are most common in the territory of the middle cerebral artery, but may occur in arteries anywhere in the brain. A large unusual arteriovenous aneurysm of the great cerebral vein and the arteries of the circle of Willis was reported by B. J. Alpers and F. M. Forster.[3] On the other hand, C. A. Papatheodorou and others[4] reported eight cases of unusually small arteriovenous aneurysm, with special radiographic investigation and method of treatment. In this case, the lesion was enormously larger than those reported by many authors, and it almost completelly destructed the left diencephalon and basal ganglia. Because of extensive hemorrhage in the subarachnoid space and in the lesion, careful examination failed to demonstrate the exact origin of the malformation. However, from the findings of arteriograms and austopsy, the arterial side of the lesion appeared to have originated from the anterior part of the circle of Willis, probably in relation with the internal carotid bifurcation. The ophalmic artery, one of the main branches of the internal carotid artery, was also involved and remarkably enlarged, comressing the optic nerves. Futhermore, the left supraorbital and dorsal nasal artery, which are terminal branches of the opthalmic artery, were involved and clinically showed pulsating swelling at the forehead and dorsum nosi respectivelly. In some cases, moderately distant vessels are also involved and show enlargement and tortuosity. This is a manifestation of the anastomosis between the external and internal arterial systems, especially through the orbit and meninges. As to this problem, Ray[5] reported in detail.

The first clinical symptoms of the cerebral arteriovenous aneurysm usually appear at an early age, and the cardinal signs are epileptic fits, subarachnoid hemorrhage and hemiplegia. As was pointed out by Olivecrona and Riives[1], the symptoms at first occur at long intervals, during which the patient has good health, until years later the initial symptoms reccurs or a new one appears. Gradually, as illness progresses, tendency to increasing frequency and intensity of symptoms become apparent. In this case, the first symptom, hemiplegia, appeared when the patient was 12 year old and a few years later hemiplegia subsided temporarily, but gradually reccured and increased in its severity. Hemiplegia in this patient was probably due to destruction of the basal ganglia and the diencephalon by growing aneurysm. In most of the fatal cases, the direct cause of death is hemorrhage in the subarachnoid space and cerebral tissue, as was reported in this case.

A history of epileptic seizures or hemiplegia, the presence of a bruit are

contributory to the diagnosis. An audible bruit has been heard in relatively few reported cases. But when present, it is clear proof of a large arteriovenous communication. However, it should be borne in mind that a bruit is occasionally observed with saccular aneurysms or with very vascular meningiomas.[1] Cerebral angiography is regarded to be most useful to disclose arteriovenous aneurysms. This shows characteristic figures and indicates the extent and the location of the lesion exactly. C. Loed and E. Favale[6] reported the electroencephalographic findings in case of intracranial aneurysms and indicated the reliability of EEG to express hemodynamic changes in the cerebrum.

About the treatment of the cerebral arteriovenous aneurysms, B. S. Ray,[5] C. Pilcher,[7] Olivecrona[1] J. L. Pool[8] and R. A. Hayne[9] described in detail, and they reported some cases in which the lesions were successfully removed. Ligation of the afferent arteries and radiation therapy have been also tried in some cases in which the total excision is impossible, but these treatments are not regarded to be effective.

SUMMARY

An autopsy case of congenital arteriovenous aneurysm of the brain is reported. The patient suffered from hemiplegia for a long time and died of extensive subarachnoid hemorrhage. Postmortem examination revealed large arteriovenous aneurysm, which arised adjacent to the circle of Willis and completely replaced the basal ganglia protruding into the ventricular cavity. Clinical symptoms and cerebral angiogram were characteristic of arteriovenous aneurysm of the brain.

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