The Abnormally Tangled Artery Simulating an Intracranial Aneurysm with Complete Defect of the Terminal Portion of the Internal Carotid Artery
—Report of a Case—

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Abstract

A peculiar case of abnormally tangled cerebral artery with complete defect of the terminal portion of the internal carotid artery is reported. The patient suffered from an intraventricular hemorrhage. Preoperative angiography suggested an aneurysm at the junction of the right internal carotid and posterior communicating arteries and also an occlusion of the terminal portion of the right internal carotid artery which was thought to have developed asymptotically prior to the hemorrhage. The right cerebral hemisphere was supplied mainly by an artery arising from the anterior cerebral artery and partly by several cortical arteries of the anterior and posterior cerebral arteries. Abnormal fine arteries originating from the internal carotid artery also supplied the basal part of the hemisphere. Operative findings, however, revealed that the aneurysm-like mass was not a true aneurysm, but rather was abnormally elongated tangled arteries composed of the posterior communicating and posterior cerebral arteries. Furthermore, no vessels were found where the terminal portion of the internal carotid artery and the horizontal portion of the anterior and middle cerebral arteries were considered to exist. This case offers the first example in which the complete defect was localized at the terminal portion of the internal carotid artery. The radiological findings and the pathogenesis of these peculiar lesions are discussed.

Key words: cerebral aneurysm, cerebral arteriosclerosis, cerebral hemorrhage, cerebral thrombosis, megadolichoartery

Introduction

A marked elongation and/or widening of the cerebral arteries is sometimes encountered in elderly patients but rarely in the young. This condition has been variously termed; arteriosclerotic aneurysm, fusiform aneurysm, cerebral arterial ectasia, dolicho-ectatic artery, and megadolichoartery. It is thought to be different from a congenital berry aneurysm either in morphology or in etiology. Arteriosclerosis may be one of the main etiological factors responsible for these lesions.

Agenesis or hypoplasia of the internal carotid artery (ICA) is a very rare anomaly, entirely asymptomatic, and sometimes accompanied by congenital berry aneurysms. This lesion is also quite different from an embolic or a thrombotic occlusion of a cerebral artery, and it is usually easy to clinically differentiate between the two.

This paper delineates a peculiar case of an abnormally elongated tangled artery with complete defect of the terminal portion of the ICA, in which the tangled artery was misinterpreted as an aneurysm and the filling defect of the distal ICA as a usual arteriosclerotic occlusion.

Case Report

This 62-year-old woman had been well until January 9, 1980 when she suddenly suffered from a severe headache and vomiting, followed by loss of con-
sciousness for several minutes. She was admitted to a local hospital, and was found to be hypertensive with systolic pressure of 240 mmHg. Two hours after the attack the patient became unconscious and vomited coffee-like fluid. Sixteen hours after the attack, she regained consciousness and was transferred to our neurosurgical clinic. She had a history of high blood pressure of 10 years' duration and of an ovarian cyst surgically treated 15 years before. She had been taking no antihypertensives for several years prior to this admission.

On admission, the patient was stuporous and confused. The pupils were normal, ocular fundi were invisible because of cataract, and the eyes were slightly deviated to the right. There was moderate weakness of the left extremities with Babinski's sign on the left. Nuchal rigidity and Kernig's sign were remarkable. Blood pressure was 180/70 mmHg, and the pulse rate was 54/min with arrhythmias. No bruit was audible in the head or neck. There were no other physical abnormalities except for a surgical scar in the lower abdomen. Routine blood tests and serologic tests for syphilis found nothing remarkable. Lumbar puncture demonstrated bloody cerebrospinal fluid.

Skull X-ray examination showed a small irregular calcification, which was proven to be part of the arterial wall by subsequent angiography of the suprasellar region. Computed tomography (CT) revealed an intraventricular hemorrhage of the right lateral ventricle (Fig. 1). There was a small dumbbell-shaped high density spot compatible with the calcification in the suprasellar region. Right carotid angiography unmasked an elongated ICA and an aneurysm-like mass at the junction of the ICA and the posterior communicating artery (PCoA) (Fig. 2).

Neither the anterior cerebral artery (ACA) nor the middle cerebral artery (MCA) were visualized, while the right posterior cerebral artery (PCA) was well visualized via the large PCoA. Several fine, tortuous arteries were seen around the mass, but they were considered not to represent Moyamoya disease.

Serial angiography of both carotid and left vertebral arteries was performed to ascertain the precise anatomy of the lesion. Multiplane right carotid angiograms suggested that the aneurysm-like mass was a true aneurysm arising from the junction of the ICA and PCoA, and also suggested an occlusion of the ICA distal to the aneurysm (Fig. 3). Left carotid angiography showed that the left ICA was also elongated and supplied the left MCA and both ACAs. A small artery, thought to be the accessory middle cerebral artery, was seen arising from the ACA and supplying a large area normally supplied by the absent MCA (Fig. 4A). The right cerebral hemisphere was also supplied by several cortical branches of the right ACA and PCA in a retrograde fashion, but the usual main branches of the MCA were never seen, even in the late phase films of the serial angiogram.

Left vertebral angiography disclosed that the left PCA was also elongated and tortuous, while the basilar artery was only slightly so (Fig. 4B). The right PCA was not visualized, probably because it was supplied by the large PCoA on the right side. Based on these clinical and radiological findings, a final diagnosis of intraventricular hemorrhage due to rupture of aneurysm, asymptomatic occlusion of the terminal portion of the ICA prior to the present hemorrhagic episode, and marked elongation of the large cerebral arteries due to arteriosclerosis was made.

On January 30, when the patient was still stuporous

![Fig. 1 Preoperative CT showing an intraventricular hematoma almost exclusively localized in the right lateral ventricle. A dumbbell-shaped high density spot in the right suprasellar cistern gave an X-ray absorption number compatible with calcification (arrow).](image-url)
and hemiplegic, right craniotomy was performed to obliterate the aneurysm and to relieve the patient from increased hydrocephalus detected by the repeated CT.

First, ventricular drainage was done in the right occipital region, and then right fronto-temporal craniotomy was carried out. After the chiasmatic...
cistern was opened, the right ICA, which was involved by marked arteriosclerosis, was followed backward to the origin of the PCoA. At this point we saw the aneurysm-like mass situated in the suprasellar cistern embedded within fibrin debris and the large elongated ACA compressing the right optic nerve medially (Fig. 5A). A small artery, the accessory MCA, was noted arising from the ACA and running toward the Sylvian fissure. Several fine arteries were also noted originating from the ICA just lateral to the aneurysm-like mass, but the terminal portion of the ICA and the horizontal trunk of the ACA and MCA (A₁ and M₁ segments) could not be seen at all.

The aneurysm-like mass was then carefully dissected.

Fig. 4 Left carotid (A) and left vertebral angiography (B). The elongated left internal carotid artery supplies both anterior cerebral arteries. A small artery arising from the anterior cerebral artery supplies a large part of the right cerebral hemisphere (arrowheads). Cortical branches of the anterior cerebral artery are well developed and supply the right cerebral surface. The left posterior cerebral artery is also elongated while the right one is not visualized because it is fed by the right posterior communicating artery.

Fig. 5 A: Operative photograph showing the terminal portion of the ICA and a part of the tangled artery. B: Schematic drawing of the whole lesion confirmed by surgery. The aneurysm-like mass was an abnormally elongated tangled artery composed of the PCoA and the P₂ segment of the PCA. The P₁ segment of the PCA was markedly hypoplastic (arrow). A small artery (arrowheads) and several fine arteries originated from the ACA and ICA, respectively, but the terminal portion of the ICA and the horizontal trunk of the ACA and MCA could not be seen. OcN: oculomotor nerve; OFA: orbito-frontal artery; OpN: optic nerve. For other abbreviations, see text.

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Eventually, it was found to be not a true aneurysm, but an abnormally elongated tangled artery (Fig. 5B). This tangled artery consisted of the PCoA and the P2 segment of the PCA. The lateral part of this artery firmly adhered to the temporal lobe and had a small aneurysm-like outpouch where intraventricular hemorrhage might occur. As the outpouch had rather a thick wall, no further attempt was made to treat this lesion and craniotomy was closed in the usual manner.

The patient returned to her preoperative condition on the day following surgery, but continued to be in a stuporous and hemiplegic state for several weeks. In the beginning of March, she became apallic and CT again showed marked ventricular dilatation. A ventriculoperitoneal shunt was substituted for the malfunctioning ventricular drainage with slight recovery of her conditions. In the middle of April, she developed uremia, and died of sudden cardiac arrest during hemodialysis for uremia on April 20. A proposal for necropsy was refused.

Discussion

We could not find a similar case in the literature. This case offers some problems to discuss.

The first problem relates to the etiology of the defect of the terminal portion of the ICA. There are two possibilities as to the nature of this lesion; one is congenital agenesis, and the other is the disappearance of previously occluded arteries. In our case, the terminal portion of the ICA, as well as the A1 and M1 segments, was completely absent without any history of cerebral ischemia. The right cerebral hemisphere was supplied by an artery arising from the right ACA, which was thought to be the accessory middle cerebral artery, though it was inappropriate to designate it as “accessory” because there was no primary middle cerebral artery. The right hemisphere was also supplied by several abnormal fine arteries arising from the right ICA and by the cortical branches of the ACA and PCA.

Embryonic failure of the primordium of an organ to appear is termed agenesis, and such may have been the case with the ICA here. Agenesis13,15 or hypoplasia11 of the ICA has rarely been reported, and it is entirely asymptomatic by itself. In our case, congenital agenesis is suggested as the nature of the defect based upon the complete absence of the arteries, the smooth continuity between the ICA and PCoA, the lack of ischemic symptoms, and the presence of the abnormal arteries arising from the right ICA and right ACA.

On the other hand, in all of the few reported cases of agenesis or hypoplasia of the ICA, the lesions are consistently noticed in the cervical as well as the intracranial segment of the ICA, and it is a common feature that the MCA was always normal with its blood supply assured either by the basilar artery or by the ACA through the circle of Willis.11,13,15 The present case, to the best of our knowledge, is the only one in which the defect was localized only at the terminal portion of the ICA.

There is another possibility, therefore, as to the nature of the defect. If the ICA was occluded at its bifurcation, would the terminal portion of the ICA as well as the A1 and M1 segments completely disappear? If the defect resulted from an acquired vascular occlusion, there should be some history suggesting cerebral ischemia, and even if the occlusion developed asymptotically after birth, some vestiges of the occluded vessels should be found in their original position. In our case, the abnormal fine arteries recognized around the aneurysm-like mass may be anastomotic channels developing after ICA occlusion, and the lack of ischemic symptoms may be explained by the presence of the accessory middle cerebral artery and leptomeningeal collaterals from the right ACA and right PCA.

However, complete disappearance of the arteries seems unlikely because the complete disappearance of occluded vessels has not so far been documented.7,10 In postmortem examination of Moyamoya disease in the adult, which is known as an insidiously developing and long standing occlusion of the terminal portion of the ICAs, some vascular structures always remain in situ.10 It is also reported that in cases of old carotid artery occlusion, some vestigial vessels are recognized in their original position.7 Fisher reported three cases of early-life carotid-artery occlusion.7 One of his cases had developed ICA occlusion at birth and succumbed to an intraventricular hemorrhage at the age of 56 years, and at autopsy the ICA, as well as the A1 and M1 segments, was shown to be extremely small and threadlike. Though occluded vessels may completely disappear after a very long period, these occluded vessels had persisted as threadlike cords in their original position 56 years after their occlusion.

If the defect discovered in our case had resulted from the disappearance of the occluded vessels which had normally developed in embryogenesis, the occlusion must have occurred very early in life, probably in intrauterine life, i.e., congenitally. We cannot conclude whether this defect was ICA agenesis or the result of the ICA occlusion. However, if it was agenesis, this report is the first one in which the defect was localized at only the distal portion of the ICA, and if it is due to vascular occlusion, this case offers an important example demonstrating that the occluded vessels may
completely disappear after a very long period.

The second problem for discussion relates to the nature of the tangled artery. Severe elongation and/or widening of the cerebral arteries is sometimes found in elderly patients with neurological symptoms, and it has been variously termed. Since first described comprehensively by Dandy,1 many cases have been reported. The basilar artery is most commonly involved, followed in frequency by the ICA, MCA, and ACA. The majority of patients with this disease are more than 40 years and usually hypertensive.3,12 The pathogenesis of such lesions appears to depend upon the association of hypertension, arteriosclerosis, and loss of arterial elements.

Reportedly, arteriosclerosis most often involves large cerebral arteries, e.g. the ICA, MCA, and basilar arteries. Smaller arteries such as the anterior and posterior communicating arteries are said to be spared by this degenerative change.1 In our case, arteriosclerosis seems to be an important etiological factor which contributed to the tangled artery. This case, however, is different both in the shape and location of the lesion from the one described above. Although all large basal arteries were elongated in our case, the tangled configuration of the arteries was localized at the right PCoA and PCA, which are usually insusceptible to arteriosclerotic changes.

What was the cause of this topographical difference? In our case, the PCA primarily originated from the ICA and there was a defect of the terminal portion of the ICA; therefore the PCoA and PCA might have been exposed to much hemodynamic stress which is thought to facilitate arteriosclerosis. If arteriosclerosis advances with age and is due to long lasting hypertension, these large PCoA and PCA might also undergo marked elongation. As the ICA and basilar artery are large in size and run in rather a wide cisternal space, they would not assume a tangled configuration when they had become markedly elongated. On the other hand, the PCoA and PCA are smaller arteries that run within a narrow space, and furthermore both of their ends are fixed to the ICA and the brain stem. Therefore, these arteries are obliged to assume a tangled form when arteriosclerotic elongation has taken place. As we interpreted the tangled artery to be a true aneurysm, to perform surgery may be a dangerous operation, as discussed before, and if these fine arteries played a role as collateral channels in ICA occlusion, the hemorrhage might have occurred from these fine arteries.

The third problem is whether this tangled artery can be differentiated from a true aneurysm preoperatively. Although repeated multiplane angiography including a stereoscopic view was taken, we could not exclude the possibility of aneurysm. Even in retrospective analysis of the angiograms, it was impossible to understand the anatomy of the lesion correctly. Magnification angiography might be of use for differentiation of the lesion. In any event, it is most important to know that such a tangled artery may be formed.

The final problem for discussion relates to the origin of the hematoma and treatment of the lesion. Intraventricular hemorrhage is known to result from many causes such as hypertension, intracranial aneurysms, arteriovenous malformations, intraventricular tumors, blood dyscrasias, and so on. Among these, three possibilities must be considered in our case. Intracranial aneurysms are most frequently associated with hypertensive intracerebral hemorrhage which is usually detected by CT with no difficulty. Intracranial aneurysms sometimes rupture into the ventricles with or without subarachnoid hemorrhage. Intraventricular hemorrhage may also result from rupture of abnormally dilated anastomotic channels developed in Moyamoya disease14 or ICA occlusion.7 In our case, hypertensive hematoma was ruled out peroperatively based on the CT findings showing no definite intracerebral hematoma, though hypertension must have played some role in the bleeding. Rupture of the abnormal fine arteries arising from the ICA was also excluded because they were not as prominent as those of Moyamoya disease.

Finally, we interpreted the tangled artery to be a true aneurysm, and therefore we supposed that this hematoma must have resulted from rupture of the aneurysm. However, operative findings disclosed that there was only the tangled artery instead of an aneurysm, and we could not show definitive evidence that this tangled artery had bled, though there was an aneurysm-like outpouch on the tangled artery. Since rupture of arteriosclerotic lesions is said to be exceedingly rare, other possibilities, i.e. intraventricular rupture of hypertensive intracerebral hematoma or rupture of abnormal fine arteries, must be considered. Especially if the ICA defect was caused by ICA occlusion, as discussed before, and if these fine arteries played a role as collateral channels in ICA occlusion,7 the hemorrhage might have occurred from these fine arteries.

As we interpreted the tangled artery to be a true aneurysm, it was reasonable for this patient to receive intracranial surgery. However, if we could preoperatively rule out aneurysm, to perform surgery may be a question. Some suitable method of treatment must be developed for this arteriosclerotic lesion as cerebral arterial ectasia also has a tendency to bleed.6,8

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