Aneurysm of the Left Middle Cerebral Artery Caused by Myxoid Degeneration of the Vessel Wall

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Background Myxoid degeneration of arterial walls may result in dissection and dissecting aneurysms in extracranial and intracranial portions of cerebral arteries. Rarely, saccular aneurysms may also develop on that basis, but thus far these have only been reported in the cervical portions of the carotid arteries. We describe a case of a nondissecting aneurysm of the left middle cerebral artery caused by myxoid degeneration of the media.

Case Description A 39-year-old man had acute onset of frontal headache and neck stiffness. Computed tomographic scan and lumbar puncture established the presence of subarachnoid hemorrhage. Angiography demonstrated a left middle cerebral artery aneurysm. This was clipped and the wall biopsied. Microscopically the entire thickness of the vascular wall showed marked myxoid alterations. No dissection was present. The patient had an uneventful recovery.

Conclusions This observation confirms that myxoid degeneration of arterial walls may cause aneurysms with eventual rupture even in the absence of dissection. Rare cases of this type of aneurysm have been reported to occur in the cervical portions of the carotid arteries, but this is the first reported observation of such changes involving an intracerebral artery. (Stroke. 1994;25:2283-2286.)

Key Words • aneurysm • middle cerebral artery • myxoid degeneration • subarachnoid hemorrhage

Intracranial aneurysms have a spectrum of causes. The most common variant is the so-called congenital saccular aneurysm, which is believed to develop on the basis of focal developmental absence of the muscularis media of the artery, the internal elastic membrane, or both; with some secondary, possibly hemodynamic, factors also playing a role in the development of an aneurysm at the area of structural defect. Acquired weakness of arterial walls may be the result of atherosclerosis, particularly in older subjects, and may be the result of the different forms of vasculitides at any age. "Mycotic" aneurysms are the result of a (usually bacterially) infected embolus lodging in an artery; the infectious process may subsequently erode the arterial wall, starting at the intimal layer and extending into the media. Tumor emboli may have a similar erosive effect, particularly emboli from atrial myxomas, which may be responsible for multiple intracranial aneurysms. Uninhibited elastase activity in patients with α1-antitrypsin deficiency may have similar consequences. Constitutional structural weakness of collagen and elastic fibers may also affect arterial walls, eg, in the Ehlers-Danlos syndrome, and fistulas as well as aneurysms may follow. Such structural weakening of the arterial media may also lead to dissection, and this process may involve the cervical carotid arteries but may also occur intracranially, resulting in subarachnoid hemorrhage.

Myxoid degeneration may also contribute to the weakening of arterial walls. This process brings about a replacement of the normal connective tissue, smooth muscle, and elastic fibers by primitive myxoid connective tissue similar to that found in Wharton's jelly of the umbilical cord. In the heart itself this process may involve the cardiac valves, making them abnormally soft and pliable ("floppy valves"). In its advanced form these myxoid changes may lead to dissolution of the media, as in cystic necrosis, a process particularly characteristic of full-blown Marfan's syndrome but occasionally also encountered in the aorta, its major tributaries, or in the pulmonary arteries without the patient having the characteristic habitus of Marfan's syndrome ("formes frustes").

Beyond the largest blood vessels, myxoid degeneration and medionecrosis of other types involving arterial walls are seldom seen. When present, they may result in dissection, with or without secondary rupture. Barnes and Jacoby reported mucoid changes with cystic medionecrosis in the common carotid artery with resulting aneurysm of the nondissecting type. A similar case with involvement of the extracranial (cervical) portion of the internal carotid artery was reported by Martinez et al.

To our knowledge, no case of intracranial myxoid degeneration of arterial walls with nondissecting aneurysm formation has been reported thus far. We present a case of advanced myxoid degeneration of a segment of the left middle cerebral artery with resultant aneurysm formation and subarachnoid hemorrhage in a 39-year-old man.
Case Report

History and Physical Findings

A 39-year-old white man had acute onset of frontal headache and neck stiffness while watching a sporting event on television. He continued to experience headaches 5 days after the initial onset of symptoms. Computed tomographic (CT) scan of the head revealed subarachnoid hemorrhage in the left sylvian fissure and over the convexity. Lumbar puncture showed 24,000 red blood cells and xanthochromia. He was transferred to the University of Kansas Medical Center.

The patient's medical, social, and family histories were unremarkable. He had no history of arterial hypertension and was normotensive at the time of admission. He had no features of a marfanoid habitus (eg, unusual height, arachnodactyly, ocular problems suggesting luxation of the lens) or features of Ehlers-Danlos syndrome (hyperextendability of joints, abnormal stretchability of the skin). There was no history of other connective tissue diseases in the family. Neither the patient nor his family had history of vascular malformations, renal disease of any kind, or cerebrovascular diseases, including documented aneurysms.

Radiological Findings

The CT examination was repeated and images of the head obtained without the intravenous administration of iodinated contrast material. The results were compared with the previous CT scans of the outside hospital. These images again demonstrated the presence of acute blood in the subarachnoid space of the left sylvian fissure. The lateral third and fourth ventricles were normal in size, shape, and position. Apart from the subarachnoid blood, no other abnormality was noted.

Selective left internal carotid angiography demonstrated an oval 8x5-mm middle cerebral artery aneurysm. An unusual feature of this aneurysm was that it arose distal to the trifurcation of the middle cerebral artery. Delayed filling of the aneurysm was another unusual angiographic feature of this aneurysm (Fig 1). On the initial arterial phase images, the aneurysm did not opacify but only became obvious in the late arterial phase. No definite arterial spasm was noted. Selective right carotid angiography and right and left vertebral angiography were normal.

Operation

A left pterional craniotomy was performed, and the dura was opened. The arachnoid was incised and the left sylvian fissure split and explored. An aneurysm of the middle cerebral artery was identified quite distal

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Pathological Findings

The material submitted as "aneurysm" consisted of two small pieces of tissue, the larger measuring 0.6×0.3×0.2 cm and the smaller 0.2×0.2×0.1 cm in greatest dimensions. They were very soft and pliable and had a pale tan color. They were embedded in their entirety. Microscopic sections of both pieces showed longitudinally cut segments of an artery. The intimal lining of the lumen was irregularly folded. No embolic material, myxomatous or otherwise, was seen within the lumen, but the wall itself had undergone marked alterations. Much of the internal elastic membrane was absent, and only small discontinuous fragments could be found on sections stained with the Verhoeff–van Gieson technique for elastic fibers (Fig 2). Masson's trichrome stain showed total absence of smooth muscle elements of the media. Instead, a thick layer of primitive-appearing connective tissue replaced the full thickness of the arterial wall (intima, media, and adventitia), with the most pronounced changes affecting the media. The intima and the inner portion of the media were composed of myxoid tissue with spidery multipolar cells that had replaced the normal fibroblasts and smooth muscle cells, but some collagen fibers were still seen between cells (Fig 3). The central and outer portions of the media had a very loose texture, with rather widely spaced multipolar cells arranged in a fashion similar to that seen in tissue cultures, and there were no collagen fibers between them, only a very thin mucoid-myxoid ground substance (Fig 4). These areas were very similar to the primitive myxoid tissue (Wharton's jelly) of the umbilical cord in the fetus and newborn. Colloidal iron stain for acid mucopolysaccharides gave a very strong positive reaction for the full thickness of the vascular wall; the intensity of the staining paralleled that of the mucus-producing goblet cells in the small intestinal mucosa used as positive controls for the colloidal iron stain (Fig 5). There were no inflammatory changes, and

Results of Microbiological Studies

Smears as well as aerobic and anaerobic cultures from the clot were negative for the presence of bacteria.
atherosclerosis was also absent. No dissection of the vascular wall was observed.

The microscopic changes were those of severe myxoid degeneration of the arterial wall, and it appeared that the tensile weakening of the wall by this tissue alteration was the likely cause for the formation of the aneurysm in this segment of the left middle cerebral artery.

Discussion

Saccular as well as fusiform aneurysms of cerebral arteries have as their common background weakening of the tensile strength of a given segment of the artery. The reasons, as indicated, include congenital developmental defects and acquired injuries to the vessel walls in the form of atherosclerosis, various types of vasculitides, and erosion by infectious emboli or by embolizing tumor particles, particularly those originating from atrial myxomas.

There may also be an underlying constitutional disorder present in these individuals. For example, uninhibited elastase activity in patients with α1-antitrypsin deficiency may contribute to weakening of the arterial walls, and the same holds for structural weaknesses in collagen and/or elastic fibers in Ehlers-Danlos syndrome and in Marfan’s syndrome. It is in the latter disease that myxoid alterations of the wall may set the stage for further structural damages, most commonly in the form of "spontaneous" (usually meaning nontraumatic) dissections. With or without the established diagnosis of Marfan’s syndrome in the affected patients, such changes may involve the cerebral circulation, most often in the extracranial areas but occasionally also within the cranium. While dissection is the most common complication of myxoid degeneration (and its most severe form, cystic medionecrosis), in two instances "regular" (nondissecting) aneurysms have also been observed in cervical portions of carotid arteries in adults. This should not be surprising because the above-mentioned loss of tensile strength of the vascular walls, the common denominator of all saccular and fusiform aneurysms, is one of the principal complications of myxoid alterations in connective tissues of the body. This is well known in cases of floppy valve syndrome (also seen within and without the full Marfan’s complex), in which the clinical significance of myxoid changes in the cardiac valves and the chordae tendineae lies in the loss of tensile strength of these structures, with resulting mechanical dysfunction of the valvular apparatus. In the case of arteries, in addition to the more common intimal tears and dissection, this structural weakness may lead to nondissecting aneurysms as well, as was found in our patient. The myxoid changes may also bring about an overall increase in wall thickness, a feature that was quite prominent in our case as judged from the surgical specimen but also observable radiologically. The slow passage of blood through the portion of the artery just proximal to the aneurysm very likely was due to the reduction of the lumen by circumferential thickening of the arterial wall involved by myxoid changes.

In summary, although two extracranial cases had been reported earlier, our case appears to be the first reported instance of intracranial aneurysm formation resulting from myxoid alterations of the arterial wall.

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