Carotid-cavernous fistula poses no problem for diagnosis because of the impressive pulsating exophthalmos. In spite of a common belief that the lesion is usually easily and successfully treated by ligation of one or another artery leading to the fistula, the rate of cure is actually less than 50%.\(^1\) Reasons for such poor results are not clear, since the complications of treatment are not frequently reported, but cerebral ischemia and hemorrhage appear to predominate. The former is easily understood, but the pathogenesis of cerebral hemorrhage is not clear.\(^2\) Postmortem analysis of the following case showed an interesting series of lesions which could have accounted for the terminal massive intracerebral hemorrhage.

**Case Report**

The patient was a 32-year-old man who had suffered a head injury in a mine accident in 1948 at the age of 20. Examination at that time revealed ecchymoses around both orbits, old clotted blood in the right external auditory canal and in the right nostril, bilateral sixth nerve palsies, slight left facial weakness, impaired hearing on the right, and Babinski's signs that were suggestive bilaterally. The spinal fluid pressure was 170 mm, the fluid was xanthochromic and contained 150 mg\% protein. X-rays showed a fracture of the skull, possibly extending into the right orbit. While the patient was still in the hospital, he developed proptosis of the right eye associated with a bruit. Shortly thereafter the right internal carotid artery was ligated, following which a bruit developed over the left side, but this gradually subsided.

In June, 1956, the patient, now 28, was examined again when he complained of a pounding sensation in his right ear synchronous with the pulse. Proptosis of his right eye had remained unchanged in the last 5 years, and he still had double vision at times, impaired hearing on the right, and anosmia. Examination in November 1956 revealed pronounced proptosis of the right eye with obvious enlargement of the orbital veins in both the upper and lower lids and with an extremely large and tortuous vein carrying visibly pulsating blood near the medial canthus. The pulsations disappeared following manual occlusion of the right carotid artery above the ligation, but the whizzing bruit over the right globe, and the right frontal and temporal region could not be abolished. There was marked limitation of motion of the right eye, which was able to move only about 30° from the midpoint in any plane.

In February, 1957, a right carotid arteriogram showed complete occlusion of the right internal carotid artery; the contrast medium in the right external carotid artery passed through the orbital-ophthalmic system to enter the fistula in the right cavernous sinus. The right external carotid artery was ligated, but a retrograde injection of medium down the common carotid filled the right vertebral artery and demonstrated a massively enlarged posterior communicating artery feeding into the fistula in the right cavernous sinus.

**Operation.** In October, 1960, a right retrograde brachial cerebral arteriogram confirmed filling of the cavernous sinus from the markedly enlarged right posterior communicating artery (Fig. 1). On October 27, under general anesthesia with controlled hypotension, a right frontal craniotomy was performed with ligation of the intracranial portion of the right internal carotid and ophthalmic arteries. The patient did not regain consciousness. The pupils were dilated and fixed, there were bilateral Babinski's signs, and decerebrate posturing with progressive deterioration. The patient died the following morning at the age of 32, 12 years after the initial accident.

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Autopsy. The major pathological findings were confined to the central nervous system. The cavernous sinus and petrous bones were removed. Both petrous bones appeared to be dark red because of numerous tortuous blood vessels inside. The dura over the clivus was densely fibrous and there was a horizontal groove running across the clivus about 1 inch below the dorsum sellae. The groove was about 2 mm deep, lined by overgrown bony prominences with rough surfaces. This lesion was considered to be the site of the old basal skull fracture. The right ophthalmic artery was 4-5 mm in diameter engorged and occluded by a silver clip. The right internal carotid artery was occluded by silver clips at the level immediately distal to the carotid-ophthalmic junction. The proximal internal carotid artery in the neck was completely thrombosed and fibrous up to the siphon immediately proximal to the carotid-cavernous fistulous opening. The right cavernous sinus was bulging, fluctuant and blue, and the dura was tightly adherent to the underlying cranial nerves. The sinus was honeycombed with numerous small blood-filled chambers separated by white fibrous septa. All these small chambers communicated with a large chamber situated medial to the carotid artery and the cranial nerves. The carotid artery opened into this large chamber through a fistula approximately 1 cm in diameter on the medial wall of the internal carotid artery about 2 cm below the anterior clinoid process. The cavernous sinus communicated widely with irregular channels around the sella leading towards the tortuous vessels in the petrous bones.

The brain was markedly edematous, with marked transtentorial herniation on the right side and bilateral tonsillar herniation. There was a moderate amount of blood on the orbital surface of the right frontal lobe. The major blood vessels at the base of the brain showed no atherosclerotic changes. Both posterior communicating arteries were larger than the right internal carotid artery. The right anterior choroidal artery was actually larger than the normal sized right middle cerebral artery. About 2 cm from its origin, however, the right middle cerebral artery tapered down to a fine, firm, solid white strand for about 4-5 cm (Fig. 2); distal to this point it gradually resumed its normal size. Numerous perforating branches derived from the most proximal 2 cm.

There was a fresh hematoma of 200-300 cc in the right cerebral hemisphere, destroying the internal capsule, putamen, globus pallidus, and displacing the insula laterally. The cavity created by the hematoma was 5-6 cm in diameter coronally and 12-13 cm long anteroposteriorly. It had ruptured into the right lateral ventricle and both lateral ventricles were filled with blood clots and displaced to the left side (Fig. 2). The tegument of the midbrain was soft, with small punctate hemorrhages. The right cerebral peduncle was
compressed and displaced downward by the swollen and herniated right hippocampal gyrus. The hemorrhagic softening of the tegmentum extended into the rostral pons. The aqueduct was obliterated by compression and swelling. The fourth ventricle was filled with blood.

The most interesting features were found on microscopic study of serial sections of the right middle cerebral artery. The midportion was markedly atrophic and completely occluded by folds of wavy internal elastica and loose connective tissue in the center. The adventitia was rather thick, but the media was almost absent (Fig. 3). Large folds of internal elastica were similarly present in the proximal portion of the artery, but the internal elastica was interrupted at two sites and the defects were filled by thick connective tissue. The lumen of the dissecting aneurysm gradually appeared in the more proximal sections (Figs. 4 and 5). The lumen was located between the internal elastica and the adventitia and was generally surrounded by a thick layer of smooth muscle and filled with blood. Still more proximally the dissecting aneurysm communicated with a partially patent lumen inside the internal elastica of the middle cerebral artery. Most of the perforating branches feeding the basal ganglia received their blood

FIG. 3. Verhoff's elastica stain of midportion of right middle cerebral artery shows the complex folding of the collapsed elastica.

FIG. 4. Verhoff's elastica stain of proximal portion of right middle cerebral artery. Arrows indicate lumen of dissecting aneurysm surrounded by media.

FIG. 5. One of the dissecting aneurysms shown in Fig. 4.
supply through the dissecting aneurysm of the middle cerebral artery. The distal artery was patent.

**Discussion**

The cause of the right carotid-cavernous fistula was almost certainly the mechanical injury of the carotid artery associated with the basal skull fracture 12 years before the patient's death, since there was no suggestion of an aneurysm at the site of the fistula. The dissecting aneurysm of the right middle cerebral artery also must have occurred at the time of the injury and became thrombosed secondary to the dissection of blood between the internal elastic and the adventitia. As a result, folded sheets of the internal elastica filled the original lumen of the midportion of the middle cerebral artery. Most of the media atrophied, probably as the result of disuse, since no blood was flowing through the original lumen to subject the media to stretch. Some of the media, however, became reorganized into several channels that carried what little blood flow there was through the dissecting aneurysm into the lateral basal perforating arteries to the striatum. The internal elastic lamina, being relatively inert, persisted for the 12 years, irregularly pleated and folded in the site of the original lumen.

The inertness and stability of the elastica has not been widely emphasized, but we have seen a similar arterial change in a case of long-healed tuberculous meningitis with focal encephalomalacia, and Dr. Martin Netsky has shown us a case of an infantile middle cerebral artery thrombosis with an identical picture. Dr. Netsky measured the length of the internal elastica with a planimeter, compared it with a normal middle cerebral artery, and concluded that the internal elastica had not proliferated but just collapsed.

Wolman has recently reviewed the literature concerning dissecting aneurysms of intracranial arteries. He collected 14 reported cases and added three of his own. Of these 17 cases, 10 involved the middle cerebral artery, at least seven on the right side as in the present case. In three cases mechanical injury was considered the cause of the dissection. We found no evidence of new formation of elastica in the fibrous connective tissue surrounding the dissecting channels, but we too found proliferation of new smooth muscle fibers around the false channels. In our case the false channels were multiple and some of them communicated with the original lumen through defects in the elastica. In three other cases congenital defects of the arterial wall were considered by Wolman to be the causes of the dissection. Defects such as marked hypoplasia of the media were found, as in our case, but since the hypoplasia of the media occurred in only the thrombosed portion, we believe that atrophy rather than hypoplasia is the underlying mechanism, that is, it is the result of disuse and not the cause of the dissection.

In our case, the lateral perforating branches had unimpaired circulation, and they were fed by both true and false channels of the proximal middle cerebral artery. The distal middle cerebral artery was also patent and was fed by leptomeningeal anastomoses from both anterior and posterior cerebral arteries. The occlusion of the middle 2-3 cm of the middle cerebral artery apparently caused very little infarction of the brain at the time of the original accident. Any asymptomatic infarct that might have been present could not be studied adequately at postmortem because of the massive hemorrhage in the same vicinity. At the time of the injury, it is significant that only left facial weakness was present and subsequently no signs of right cerebral hemispheric damage were recorded.

From the clinical story, radiological studies, and autopsy findings, we have reconstructed the pathophysiological processes in this case. The patient had a basal skull fracture 12 years before his death. As a result of this injury he developed a right carotid-cavernous fistula and a dissecting aneurysm of the right middle cerebral artery. The middle cerebral artery was occluded, but blood flow persisted in the basal perforating branches through the dissecting aneurysm itself and in the cortical branches through collateral circulation from leptomeningeal anastomoses. Because of the pulsating right exophthalmos a few days after injury, the carotid-cavernous fistula was readily recognized and the right internal carotid artery was ligated. The pulsating exophthalmos subsided only temporarily. Since the patient did not return for further treatment for a period of 9 years, it seems likely that the relapse was gradual and progressive, probably due to gradually enlarging collaterals. Indeed, one arteriogram showed
that the fistula was fed by the ophthalmic-external carotid route, but ligation of the external carotid artery produced no significant change because of other collaterals demonstrated by subsequent arteriograms involving a huge posterior communicating artery fed by the vertebral-basilar arteries.5

Three years later the ophthalmic artery and the intracranial portion of the internal carotid artery were clipped intracranially. At the moment the internal carotid artery was suddenly occluded, the blood being supplied by the vertebral-basilar system through the greatly enlarged right posterior communicating artery was all turned into the right anterior and middle cerebral and anterior choroidal arteries. However, since the middle portion of the middle cerebral artery was already completely occluded by the old dissecting aneurysm, this sudden increase of blood flow must have presented too great a strain upon the small perforating branches derived from the dissecting aneurysm of the right middle cerebral artery and feeding into the basal ganglia and internal capsule. The result was the rupture of one or more of these branches with massive fatal hemorrhage. The occlusion of the middle cerebral artery was not identified on cerebral arteriography, but the leptomeningeal anastomotic supply to the distal artery was clearly shown and should have led to the conclusion that a proximal occlusion of the middle cerebral artery was present.

Summary

We have reported the details of a case of traumatic carotid-cavernous fistula. Autopsy studies 12 years after the injury demonstrated the following points: an asymptomatic dissecting aneurysm of the ipsilateral middle cerebral artery had developed but with preservation of the internal elastic, atrophy of the media, and reorganization of the dissecting channels.

Extensive collateralization of the vascular channels in the base of the skull had taken place. Marked dilatation of the ophthalmic and posterior communicating arteries had contributed to the persistence of the ophthalmic fistula in spite of multiple ligations of the internal and external carotid arteries in the neck. When, in the final treatment of the carotid-cavernous fistula a large part of the massive blood flow through the fistula was suddenly diverted into the relatively small perforating branches supplying the striatum, a fatal intracerebral hemorrhage resulted.

References

4. Netsky, M. Personal communication.