

Spontaneous Intracranial Carotid Artery Dissection

Michael S. Pessin, MD, Lester S. Adelman, MD, and Nancy R. Barbas, MD

A previously healthy 46-year-old woman had the abrupt onset of aphasia and right hemiplegia from a large left-hemisphere infarct. At postmortem examination the cause of the infarct was dissection and occlusion of the intracranial carotid artery. No preexisting abnormality was found to account for the dissection. This uncommon cause of stroke and its pathogenesis are discussed. (*Stroke* 1989;20:1100–1103)

Spontaneous (nontraumatic) intracranial internal carotid artery (ICA) dissection is an uncommon cause of cerebral infarction. The diagnosis may be suggested by angiography when an irregular, thin column of dye (the string sign^{1–3}) is seen as part of the occlusive process, but often only an intracranial occlusion, which may be indistinguishable from embolism,^{4–9} is noted. Pathologic documentation is usually required to establish the diagnosis of dissection and to clarify its pathogenesis.

Several conditions have been implicated in the pathogenesis of spontaneous intracranial dissection. These include congenital defects in the media or internal elastic lamina, fibromuscular dysplasia, cystic medial necrosis, trivial trauma, and migraine.^{4,6–12} In a few patients, no preexisting abnormalities have been discovered, adding further to the mystery of this condition.^{2,5,13} The present patient falls in the latter category, in which no cause for the dissection was identified.

Case Report

A 46-year-old woman arose and prepared for work at 6:00 AM and was found on the floor shortly thereafter by her husband. She was vomiting but awake and mute, not moving her right side. She was taken to a local hospital and then to New England Medical Center. On examination, her blood pressure was 130/80 mm Hg and her pulse was 58 beats/min. She was lethargic and followed no verbal instructions. A left gaze preference was noted, but her eyes moved fully to the right with oculocephalic maneuvers. She had right lower face weakness, and her right arm and leg moved poorly to painful

stimulation compared with her left arm and leg. Bilateral extensor plantar responses were present. There were no carotid or ocular bruits.

There was no history of transient ischemic attacks, migraine, or neck or head trauma. She did not have hypertension, diabetes, or heart disease, and she was not taking oral contraceptives. She smoked cigarettes.

Results of computed tomography (CT scan) and lumbar puncture were normal. Eight hours later she was comatose, with bilateral decorticate posturing and pupils poorly reactive to light. CT scan (Figure 1) at that time showed a large infarct in the territories of the anterior cerebral artery (ACA), middle cerebral artery (MCA), and posterior cerebral artery (PCA). Despite medical measures to control her increased intracranial pressure, she died on the second hospital day.

Gross examination of the brain revealed massive swelling and softening of the left cerebral hemisphere involving the territories of the ACA, MCA, and PCA. There was a medial temporal lobe herniation on the left. The left posterior communicating artery was large; the P1 segment of the PCA was small. The anterior communicating artery (AComA) was of average size. The left ICA, as it entered the cavernous sinus, had a hematoma in its wall, without significant narrowing of the lumen. The distal intracavernous ICA was grossly normal. Red-brown, granular material obstructed the lumen of the left ICA in its most proximal intracranial portion. It was not possible to tell grossly if this material was in the lumen of the vessel or in its wall. This process extended along the ICA, past the orifice of the ACA, and involved the proximal MCA in the sylvian fissure (Figure 2). The carotid arteries in the neck were widely patent and showed only mild atherosclerosis. Horizontal sections of the cerebral hemispheres showed a recent infarct involving almost all of the left cerebral hemisphere, includ-

From the Departments of Neurology (M.S.P., L.S.A., N.R.B.) and Pathology (L.S.A.), Tufts University School of Medicine and the New England Medical Center, Boston, Massachusetts.

Address for reprints: Michael S. Pessin, MD, New England Medical Center, Box 314, 750 Washington Street, Boston, MA 02111.

Received March 3, 1989; accepted April 3, 1989.

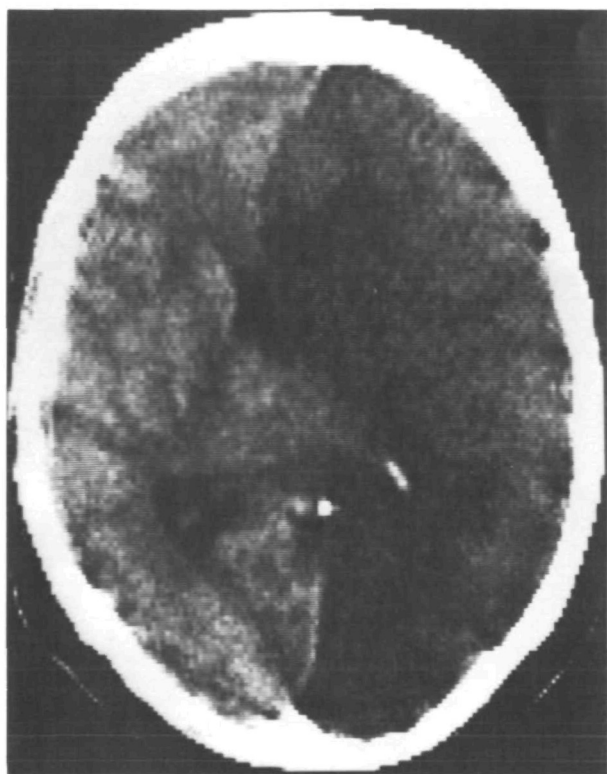


FIGURE 1. Noncontrast computed tomogram in 46-year-old woman showing low-attenuation area involving anterior, middle, and posterior cerebral artery territories with marked midline shift and compression of left lateral ventricle.

ing the basal ganglia. Transverse sections of the brainstem showed elongation and distortion of the midbrain and rostral pons, but no secondary brainstem hemorrhages.

Microscopic examination of the intracavernous portion of the left ICA showed a recent dissection with an intimal tear and an intramural hematoma located in the center of the media (Figure 3, left). The vessel showed no preexisting abnormality. A section of the grossly spared distal intracavernous artery showed no dissection and no other abnormality of the vessel wall. The intracranial ICA was severely narrowed by an intramural dissection between the internal elastica and the media (Figure 3, right). There was recent thrombus in the lumen, some attached to the wall and some lying free in the lumen. Similar changes were seen in the proximal portion of the MCA. The dissection did not involve the ACA, but a microscopic bit of recent thrombus was seen in the lumen of this vessel. The distal left MCA, right MCA, and the right ICA were all histologically normal.

Discussion

Postmortem examination established dissection as the mechanism for this patient's intracranial ICA occlusion and massive hemispheric infarct. Dissection was present in two separate areas of the ICA,

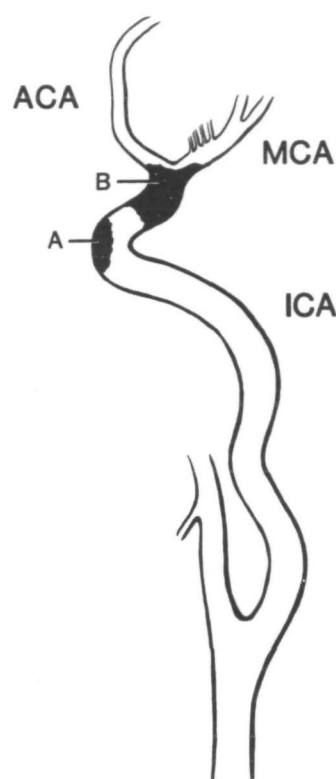


FIGURE 2. Diagram of location of lesions found in left carotid artery. Sites of dissection in siphon (A) and in intracranial vessels (B) are indicated. Area uninvolved by dissection is seen between A and B. ICA, internal carotid artery; MCA, middle cerebral artery; ACA, anterior cerebral artery.

with a "skip" area between them free of any abnormality (Figure 2). These separate areas of ICA dissection probably occurred nearly simultaneously since both hematomas showed recent blood clot histologically. The reason for the normal skip area is unclear, but its length of >1 centimeter suggests that there was more than one dissection. This case reemphasizes the difficulty in deciding on the importance of intimal tears in the pathogenesis of dissections since an intimal tear was found in one area of dissection but not in the other. The intramural hematomas were in slightly different locations in the two areas of dissection. The hematoma in the intracavernous dissection, associated with the intimal tear, was located in the center of the media; the hematoma in the intracranial dissection was found between the elastica and media. Dissections have been reported in both locations, but those in the media are more often found in the vertebrobasilar system and are associated with hemorrhage, whereas subendothelial hematomas are more characteristic of carotid dissection with resulting luminal stenosis and occlusion.^{9,11,12,14}

The inclusion of the ACA territory in the hemispheric infarct is unexpected in a patient with a patent AComA and ACA. It may have resulted from local embolism of the ICA thrombus to the

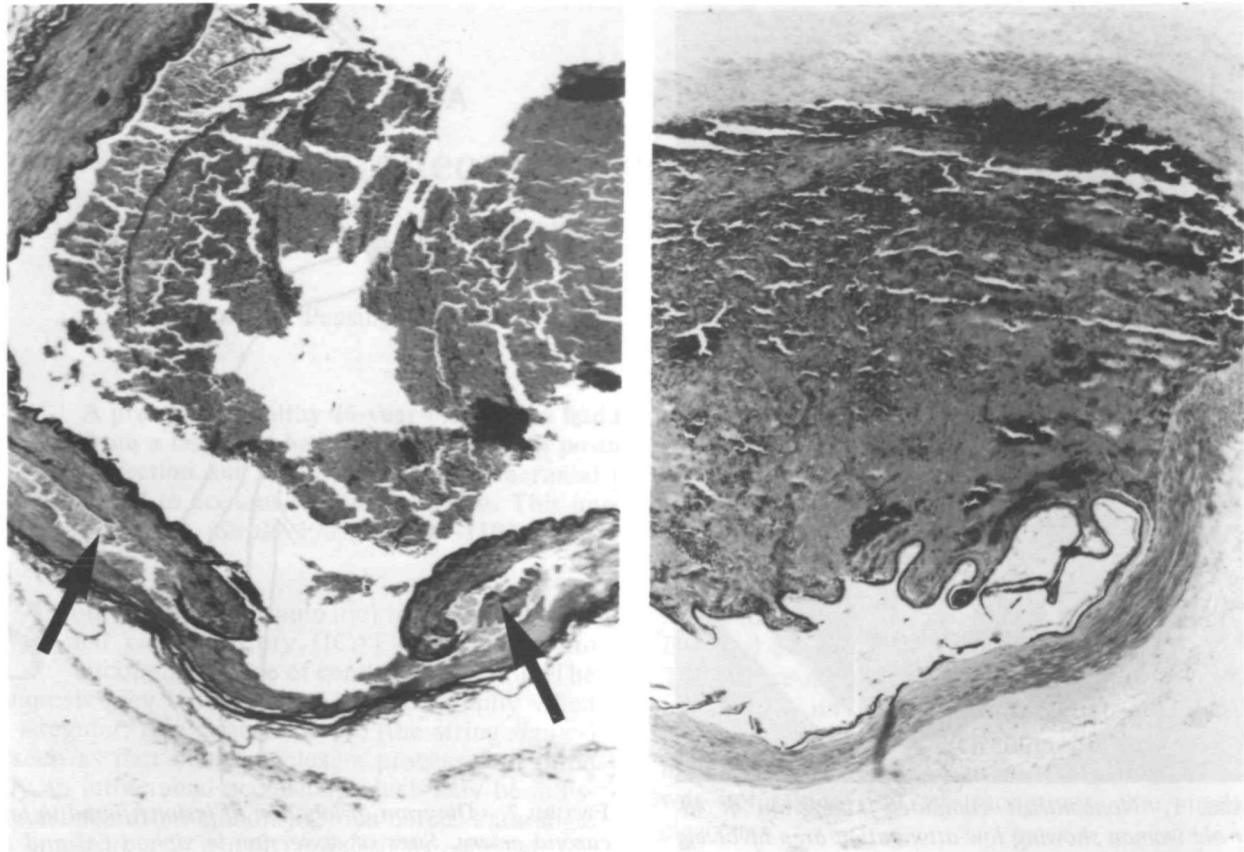


FIGURE 3. Photomicrographs of two areas of dissection. Left is from siphon (A in Figure 2). There is defect in intima elastica and part of media at lower part of photomicrograph. Dissection is indicated by arrows. Elastica stain, $\times 34$. Right is from intracranial carotid artery (B in Figure 2). Large dissection between elastica and media has reduced lumen to slit. Elastica stain, $\times 34$.

ACA with subsequent fragmentation. This hypothesis is supported by the microscopic particle of thrombus found in the ACA. Either local embolism or hemodynamic insufficiency affecting the left PCA may have resulted in the inclusion of the PCA territory in the huge hemispheric infarct since the left PCA arose primarily from the ICA.

Our patient had no preexisting arterial condition to explain the occurrence of her dissection, nor was trauma, infection, or migraine a factor. Histologic examinations of intracranial dissection have frequently identified defects, presumably congenital, in the internal elastic lamina.^{4,6-12} Gaps in, fragmentation of, or disruption of the internal elastic lamina have been the usual findings implicated in the pathogenesis of dissection. In some cases there has been an association with other conditions such as fibromuscular dysplasia, moyamoya disease, and migraine.^{9,11} Other patients have not shown preexisting arterial abnormalities.^{2,5,13} Whether trivial trauma, such as persistent coughing, sneezing, or vomiting, can precipitate dissection even with preexisting defects remains unknown.

Our patient is unusual because of the two distinct areas of dissection and the absence of any preexisting underlying defect. Further pathologic studies of

such cases may help clarify the pathogenesis and may help identify patients at risk for stroke.

References

- Ojemann RG, Fisher CM, Rich CJ: Spontaneous dissecting aneurysm of the internal carotid artery. *Stroke* 1972; 3:434-440
- Hochberg FH, Bean C, Fisher CM, Roberson GH: Stroke in a 15 year old girl secondary to terminal carotid dissection. *Neurology* 1975;25:725-729
- Fisher CM, Ojemann RG, Roberson GH: Spontaneous dissection of cervico-cerebral arteries. *Can J Neurol Sci* 1978; 5:9-19
- Wolman L: Cerebral dissecting aneurysms. *Brain* 1959; 82:276-291
- Spudis EV, Scharyj M, Alexander E, Martin JF: Dissecting aneurysms in the neck and head. *Neurology* 1962;12:867-875
- Nedwich A, Haft H, Tellem M, Kauffman L: Dissecting aneurysm of cerebral arteries. Review of the literature and report of a case. *Arch Neurol* 1963;9:477-484
- Chang V, Newcastle NB, Harwood-Nash DCF, Norman MG: Bilateral dissecting aneurysms of the intracranial internal carotid arteries in a 8 year old boy. *Neurology* 1975; 125:573-579
- Manz HJ, Vester J, Lavenstein B: Dissecting aneurysm of the cerebral arteries in childhood and adolescence. Case report and literature review of 20 cases. *Virchows Arch [A]* 1979;384:325-335
- Hart RG, Easton JD: Dissections of cervical and cerebral arteries. *Neurol Clin* 1983;1:155-182

10. Adelman LS, Doe FD, Sarnat HB: Bilateral dissecting aneurysms of the internal carotid arteries. *Acta Neuropathol (Berl)* 1979;29:93–97
11. Alexander CB, Burger PC, Goree JA: Dissecting aneurysms of the basilar artery in 2 patients. *Stroke* 1979;10:294–299
12. Deck JHN: Pathology of spontaneous dissection of intracranial arteries. *Can J Neurol Sci* 1987;14:88–91
13. Grosman H, Fornasier VL, Bonder D, Livingston KE, Platis ME: Dissecting aneurysm of the cerebral arteries. Case report. *J Neurosurg* 1980;53:693–697
14. Caplan LR, Baquis GD, Pessin MS, D'Alton J, Adelman LS, Dewitt LD, Ho K, Izukawa MD, Kwan ES: Dissection of the intracranial vertebral artery. *Neurology* 1988;38:868–877

KEY WORDS • carotid artery diseases • dissection