cerebral aneurysms are associated with a deficiency of type III collagen would require one to postulate that cerebral aneurysms constitute two distinct entities, one associated with collagen deficiency and the other not, but both being indistinguishable in all other aspects such as epidemiology, risk factors, clinical presentation, radiological appearance, gross morphology, and outcome. This assumption, in the absence of critical epidemiological studies, remains unproven.

Dr. Pope suggests that our negative results are due to inherent difficulties in the techniques employed. This criticism is just as applicable to his results since we used the same techniques as he did. However, since one of our controls was a patient with Ehlers-Danlos syndrome type IV (E-D IV) and since this patient had, as expected, almost no detectable type III collagen, we do not feel that our results are due to methodological deficiencies.

With regard to E-D IV and cerebrovascular anomalies, Dr. Pope seems to confound carotid cavernous fistulas (CCFs) and cerebral aneurysms. These are not equivalent lesions. CCFs are relatively frequently reported in association with E-D IV. Dr. Pope’s own case report, cited as Fox et al in his letter, is of a CCF; and Case 1 of Krog et al, also cited by Dr. Pope, may have had bilateral, posttraumatic CCFs and not cerebral aneurysms. To our knowledge, cerebral aneurysms have not been shown to be statistically overrepresented in E-D IV. The case of Mirza et al, also cited by Dr. Pope, had multiple peripheral extracranial aneurysms (abdominal, brachial, renal) and a fusiform dilatation of the precavernous segment of the internal carotid artery, not a cerebral, “berry” aneurysm as were the aneurysms discussed in our report. Further, the patient reported by Mirza et al did not have E-D IV. Dr. Pope and collaborators’ identification of a genetic mutation in E-D IV is, of course, not germane to the discussion.

The suggestion has been made that “measuring collagen type III levels in cultured fibroblasts from asymptomatic relatives could be a promising alternative to angiography for detecting individuals at risk.” We are glad that Dr. Pope does not share this view, but it was this kind of overenthusiasm for the collagen deficiency hypothesis that prompted our short communication and our reaffirmation of the necessity of four-vessel cerebral angiography to demonstrate cerebral aneurysms in individuals at risk.

In summary, we feel that the collagen deficiency hypothesis of cerebral aneurysms has failed a critical test and is seriously undermined. The possibility remains that there may be a subtle defect in the collagen type III gene, or in any of the multiple steps in the processing of the genetic information toward the formation of the mature and functional collagen molecule, which may predispose to the formation of cerebral aneurysms. This hypothesis is presently being tested with our material.

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References

Occipital Infarction With Hemianopsia From Carotid Occlusive Disease

To the Editor:
We read the article by Pessin et al with considerable interest. It paralleled our recent experience in a patient with a similar presentation.

A 63-year-old right-handed male awoke from a nap with a throbbing bifrontal, biocipital headache and an awareness that he could not see objects in his left visual field. The patient had no prior history of neurological or vascular complaints. His past medical history was positive for stable insulin-dependent diabetes mellitus and hypothyroidism. He smoked half a pack of cigarettes per day for years. His general physical examination and his neurological examination were entirely normal except for a left homonymous hemianopsia. Routine laboratory studies were normal except for a glucose of 246, cholesterol of 205, and triglycerides of 377. Cardiac workup, including echocardiogram, was normal. Computerized tomogram of the brain (CT scan) showed a right occipital lucency (Figure 1). Duplex scan demonstrated a 90% right internal carotid artery (ICA) stenosis.

FIGURE 1. Contrast-enhanced CT scan showing right occipital infarction in distribution of posterior cerebral artery. Left side of body appears on right side of figure.
Cerebral angiography showed a tight right ICA stenosis. The right posterior cerebral carotid artery (PCA) filled from the right ICA injection (Figure 2). Right vertebral injection showed filling of the left PCA, but no filling of the right PCA. The patient was entered into a symptomatic endarterectomy trial and assigned to surgery plus aspirin treatment and has been asymptomatic in the 12 months following surgery.

Like the case of Pessin et al, our patient demonstrates an occipital infarction resulting from ipsilateral carotid disease. Unlike their case, there was no other clinical or CT evidence of ICA disease in our patient. Because there was no communication between the vertebrobasilar system and the right PCA, and the only major vessel feeding the right PCA was the right ICA in our patient, there is little doubt that the occipital infarction was associated with ICA disease.

This case provides further evidence that symptomatic infarction in the distribution of the PCA may be associated with extracranial ICA disease and may be the only manifestation of ICA disease. This association may not be as rare as once thought. In the absence of known cardiac or vertebrobasilar disease, one should consider a noninvasively documented ICA stenosis as a possible cause of PCA distribution infarction.

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