Reversible Angiopathy

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SUMMARY A 26 year old woman presented with a subacute onset of headache, hypertension, and neurologic impairment. Angiography revealed severe extracranial carotid and vertebral artery disease. Long segmental stenoses, lumen irregularities, and aneurysmal dilatations were noted. She improved with medical management and was followed for one year. Because recurrent neurologic symptoms developed, angiography was repeated and disclosed normal vasculature. “Spontaneous dissections” with recovery are the most probable explanation for this phenomenon.

CEREBROVASCULAR disease in young adults often poses difficult diagnostic and management problems. We encountered a young woman who presented with neurologic impairment associated with marked abnormality of the extracranial portions of the carotid and vertebral arteries. The patient completely recovered from her initial deficits. However, one year later, because of recurrent episodic neurologic symptoms, the patient underwent repeat angiography and all the previous abnormalities had disappeared.

Case Report

A 26 year old woman was transferred to St. Luke’s Episcopal Hospital on 9/7/78 because of severe extracranial vascular disease. For about 6 weeks she had minor recurrent generalized headaches. Two weeks before admission she awoke in the morning with a severe right-sided headache associated with tenderness at the angle of the right mandible. The headache increased in severity and after 1 or 2 days was associated with nausea and vomiting. She periodically saw flashing lights and zig-zag lines in front of both eyes with no lateralization. She had two episodes of brief loss of consciousness without seizure phenomena when getting out of bed at night. There was a low grade fever of 100°F. Past medical and family history were unremarkable. The patient was drowsy, tangential in her replies, and complained of poor memory. General physical examination revealed a blood pressure of 150/110, pulse 86 and regular, and temperature of 98.6°F. A right carotid bruit was heard intermittently. Neurologic examination disclosed dysomia and right-left confusion. She could not spell 3 letter words backwards. On face-hand test there was bilateral extinction of hands. She had a left facial paresis affecting frontalis and a slight right arm drift with pronation. Bilateral grasp reflexes were elicited.

Chest x-ray and CT scan were normal. White blood cell count was 12,000 with a normal differential. Hemoglobin was 13.6 grams. Calcium, phosphorus, blood urea nitrogen, fasting blood sugar, uric acid, cholesterol, bilirubin, alkaline phosphatase, creatinine, lactic dehydrogenase, serum glutamic-oxaloacetic transaminase were normal. Serology was negative. Cryoglobulins were absent. Sedimentation rate was 18. Antinuclear antibody was negative and protein electrophoresis was normal. Electrocardiogram was normal. An electroencephalogram showed theta activity in temporal leads bilaterally. Review of angiography from the referring hospital was carried out and no further angiography was done. The intracranial circulation was entirely normal. There was severe, extensive, irregular narrowing of both internal carotid arteries. The narrowing began just past the origin of the internal carotid artery and involved the vessel to its entrance into the carotid canal. Aneurysmal dilatations were seen just above the bifurcation on the right side. The common and external carotid arteries were normal. The posterior circulation filled with the right carotid injection because of reflux into the vertebral artery. The posterior circulation could be seen with the left carotid injection because of collateral flow between the external carotid and muscular branches of the vertebral artery, a phenomenon compatible with internal carotid artery stenosis or occu-

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sion (figs. 1 and 2). The left vertebral was diffusely and irregularly narrowed throughout its length (fig. 3). There was mild dilatation, then narrowing, of the proximal portion of the right vertebral artery (fig. 4). The abdominal aorta was normal as were the extrarenal portions of the renal arteries. In the intrarenal circulation there were multiple beaded, narrowed, and obstructed small arteries.

Over a period of one week there was steady improvement and her mental status returned to normal. A mild left facial paresis persisted. Steroid therapy was tapered and discontinued during this time. Blood pressure returned to a normal range of 130/75. She was discharged on propranolol 20 mg t.i.d., dipyridamole 50 mg b.i.d., aspirin X grains b.i.d., and cyproheptadine 4 mg t.i.d.

The patient was seen approximately every one to two months after discharge. In October 1978 her blood pressure was 100/70 and propranolol was discontinued. Headaches were infrequent and cyproheptadine was stopped. In May 1979, she had spells of depersonalization associated with headache lasting a few hours. Neurologic examination was normal except for left facial synkinesias. An electroencephalogram was normal. Occasional frontal headache, “fullness” in the neck, and impaired thinking were recurrent symptoms. Regional cerebral blood flow studies showed impaired perfusion in the left frontal region on 8/14/79. Superficial temporal-middle cerebral artery bypass was considered and the patient was admitted to Franklin Hospital in San Francisco. Angiography was repeated and disclosed normal vasculature (fig. 5).
Discussion

Spontaneously reversing extracranial vascular disease has a limited differential diagnosis. This includes vasculitis, idiopathic regressing arteriopathy and spontaneous dissection. Other entities to consider are vasospasm and fibromuscular dysplasia.

Spasm would not be expected to produce such long segmental narrowing as was noted in the high cervical region of both internal carotid arteries. The left vertebral artery was selectively catheterized, but the catheter in the common carotid artery was always considerably below the carotid bifurcations. There were beaded areas and aneurysmal dilatations which are not seen in vasospasm particularly in the right internal carotid artery. Lastly, and most important, the patient had fever, hypertension, and neurologic symptoms prior to performance of the angiograms.

Fibromuscular dysplasia was a second consideration. This diagnosis would not explain some of the patient’s systemic complaints. The pathology of fibromuscular dysplasia is generally above the carotid bifurcation at the level of C2 higher than the radiographic abnormalities observed in this patient.1,2

Some features of the angiogram, particularly the beaded appearance suggesting mural aneurysms, would be consistent with this diagnosis,2,8 but it is not known to be reversible.3,6

The clinical presentation raised the question of vasculitis. The patient experienced a low grade fever, headache, and neck tenderness for several days prior to admission to the hospital. Hypertension was present and renal angiograms showed significant abnormalities. Laboratory data did not support this diagnosis as the sedimentation rate, antinuclear antibody, and serum protein electrophoretic patterns were all normal. The lesions seen in the angiograms have not been seen in patients with proven vasculitis.

Among rare vasculitides, Takayasu’s arteritis fits part of this patient’s clinical picture. It is most common in young women often beginning with fever and associated with hypertension. This patient, however, had a lesion in the upper portion of the internal carotid artery and lacked the typical involvement of the aortic arch and common carotid arteries.

“Spontaneous dissection” of carotid and vertebral arteries is an uncommon disorder.11-16 Most patients are men in the age range of 35 to 60. Hypertension is
**Figure 3.** Right subclavian injection demonstrates initial dilatation of the right vertebral artery followed by mild narrowing. The subclavian artery is normal.

**Figure 4.** There is severe, diffuse irregular narrowing of the entire length of the left vertebral artery. The subclavian artery is normal.
common. Presenting manifestations include unilateral headache, Horner's syndrome, and focal neurologic symptoms associated with single vessel involvement, but multiple lesions have been described. The angiographic abnormality is most often located 1.5 cm to 3 cm above the bifurcation and may extend up to the base of the skull. Long irregular filling defects with a narrow column of contrast are seen on angiography. This finding has been referred to as the "string sign." There is one report of several cases with some spontaneous improvement on repeat angiography two to sixteen months later. The angiographic features of our patient resemble those described in this disorder. The prodromal symptoms for several weeks prior to the onset of neurologic signs are not a feature of the described cases.

Idiopathic regressing arteriopathy has been described by Mokri et al. Of the three cases reported, all were in the age range of 35 to 42. There was sudden onset with headache and two patients had focal neurologic findings including a hemiparesis. One patient had a partial Horner's syndrome and another was hypertensive. Angiographic abnormalities included multiple vessel involvement with severe stenosis and occlusion. Saccular aneurysm formation, long segmental stenosis of the cervical portion of the internal carotid artery, and arterial dissections were noted. The disease process began at the level of the bifurcation and extended as far as the carotid canal. The angiographic findings of our patient are similar to this description. It is very probable that these cases were variants of "spontaneous dissections."

Spontaneously reversing extracranial vascular disease is not commonly identified. Whether this is an under-recognized phenomenon remains to be seen. The benign course of the majority of these patients indicates that medical management is ordinarily preferable to surgical intervention.

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