We describe three cases of extracranial vertebral artery dissection that are unusual both in their modes of presentation and their associations with other pathologic conditions. The first patient had Marfan's syndrome and migraine; his dissection was asymptomatic and was diagnosed by chance at the time of repeat angiography following a previous internal carotid artery dissection. The second patient had systemic lupus erythematosus and presented with a subarachnoid hemorrhage attributed to an intracranial vertebral artery dissection by the demonstration of an extracranial dissection. The third patient had a minor basilar artery stroke in which dissection had occurred beside a congenital hemivertebra deformity. (Stroke 1990;21:618-625)

Cervical artery dissection accounts for approximately 4% of the cases of ischemic stroke in young adults, with the internal carotid artery most commonly affected. Vertebral artery dissections are less frequent; they usually affect the extracranial portion of the artery, and persons with such dissections present with ischemic strokes in the vertebrobasilar territory, preceded by cervical pain. The cause of vertebral artery dissection is unknown, although neck trauma (often apparently trivial) is frequently implicated. Of the so-called "spontaneous" cases of vertebral artery dissection reported, there is often an associated underlying disorder such as fibromuscular dysplasia, elastic tissue disease, arterial hypertension, or migraine. We describe three cases of vertebral artery dissection that are unusual both in their modes of presentation and in their associations with other pathologic conditions.

Case Reports

Case 1

A 31-year-old right-handed male smoker was admitted September 15, 1986. He gave a history of migraine without aura since the age of 18 years. He had once had difficulty in "finding his words" for 15 minutes in 1976; there had been no headache, and he had not sought medical care. The patient awoke at 2 AM the morning before admission with a severe right-sided headache typical of his usual migraine. Later that morning he felt nauseated and vomited. At 7 PM, he suddenly developed aphasia and right-sided hemiparesis; there was no cervical pain.

On admission at midnight there was partial recovery of function, with mild mixed aphasia and right-sided hemiparesis. An intermittent midsystolic cardiac murmur was audible. The patient was tall and thin, and there was mild laxity of his articular ligaments.

An early head computed tomogram (CT scan) was normal, but a second one a week later was suggestive of an infarct in the left middle cerebral artery territory.

Contrast echocardiography showed a patent foramen ovale and prolapse of the mitral and tricuspid valves without regurgitation. There was proximal aortic dilatation, but aortic valve function was normal. Blood count and erythrocyte sedimentation rate (ESR) and the results of hemostatic studies and urinary amino acid analyses were normal.

Four-vessel cerebral angiography on October 8, 1986, showed localized aneurysmal dilatation of the extracranial left internal carotid artery (Figure 1, left), with tapered narrowing of the more proximal portion of the vessel. The left vertebral artery appeared slightly irregular, but without an aneurysm (Figure 1, right). The remaining extracranial vessels were normal. The patient was treated with heparin for 3 weeks, followed by oral anticoagulant therapy.

Figure 1. (Facing page.) Case 1. Four-vessel cerebral angiograms on October 8, 1986. Left: Pseudoaneurysm of distal left internal carotid artery (arrow) with narrowing proximally. Right: Left vertebral artery appears slightly irregular in outline.
The patient recovered gradually so that by November 10 his neurologic examination was normal. Angiography was repeated on January 30, 1987. The left internal carotid artery aneurysm persisted, but the narrowing had disappeared. A localized pseudoaneurysmal dilatation of the second segment of the left vertebral artery at the C4 level was now evident, with narrowing of the lumen immediately distal (Figure 2, left). There had been no cervical pain, no evidence of basilar artery ischemia, no neck injury, and no unusual neck movement. A third angiogram on June 26, 1987, showed almost-complete resolution of the pseudoaneurysm, with persistent localized stenosis of the left vertebral artery (Figure 2, right).

The patient has remained well although he has reported three typical attacks of migraine with dysphasic auras.

Case 2

This 44-year-old woman presented with a subarachnoid hemorrhage (SAH) in June of 1987. In 1984, she had developed erythematous cutaneous lesions, mouth ulcers, arthralgia, alopecia, and hemolytic anemia. Her ESR was raised, and systemic lupus erythematosus (SLE) was confirmed by positive antideoxyribonucleic acid (DNA) antibody and homogeneously staining antinuclear antibody (ANA) titers of 160 and 10,000, respectively. Prednisolone (20 mg...
daily) and hydroxychloroquine (200 mg twice daily) were given with excellent response.

On June 3, 1987, the patient complained of posterior cervical pain of sudden onset that persisted for 12 days, when it became more severe and radiated to her arms. She then had a diffuse moderate headache.

On June 18, she had a seizure, became confused, and complained of a severe generalized throbbing headache. There was neck stiffness but no focal signs. CT scan showed blood in the third ventricle, and the cerebrospinal fluid was grossly hemorrhagic.

Four-vessel cerebral angiography on July 6 showed completely normal internal carotid and left vertebral arteries. By contrast, narrowing of the intracranial segment of the right vertebral artery was seen, with an aneurysm in its extracranial segment at the C1 level (Figure 3, left).

Blood count and serum complement levels and the results of hemostatic studies were normal. Her ESR was 27 mm/hr. Coombs' test and an assay for the lupus anticoagulant were negative, and her anti-DNA antibody and ANA titers were unchanged.

The patient recovered completely, and control angiography was performed 6 weeks later. The narrowing of the distal part of the right vertebral artery had virtually resolved, and the extracranial aneurys-
normal image was no longer visible (Figure 3, right), confirming the diagnosis of intracranial and extracranial vertebral artery dissection.

Case 3

This 37-year-old woman felt the abrupt onset of right-sided cervical and left shoulder pain while reversing her car in September of 1986. The pain resolved in 2 months.

On September 25, 1987, she noted the sudden onset of right cervical pain and 5 days later numbness of her right thumb and index finger. After 2 more days, her left cheek felt numb. On examination there was loss of light touch and pinprick sensation over the second division of the left trigeminal nerve and right C6 dermatome. There was scoliosis of the cervical spine, but the examination was otherwise normal.

A cervical radiograph showed a hemivertebra between C6 and T1, which produced a marked scoliosis with convexity to the right (Figure 4). A cervical myelogram showed no nerve root or spinal cord compression. Magnetic resonance imaging...
(MRI) of the brain and spinal cord was normal. Four-vessel angiography by the femoral route on December 21, 1987, showed that the right vertebral artery was normal above the scoliosis but abnormal below, with a small localized dilatation near its point of entry into the vertebral canal (Figure 5, left). Her pain and sensory signs resolved completely over 3 months.

A second angiogram on May 26, 1988, showed a decreased diameter of the dilated segment (Figure 5, right).

Discussion

In these three cases, vertebral artery dissection was diagnosed angiographically by the presence of an aneurysmal dilatation with (cases 1 and 2) or without (case 3) adjacent stenosis that improved or resolved on repeat studies. Dissection occurred in each case at a different level, midcervical in the first case, extracranial and intracranial in the second, and proximal in the third.

The clinical presentations were different from the classical association of abrupt craniocervical pain followed by signs of basilar ischemia. The first patient reported no symptoms, and his dissection was discovered by chance on repeat angiography 3 months after a left internal carotid artery dissection. "Silent" extracranial internal carotid or vertebral artery dissections have been reported in patients with multiple simultaneous dissections, but we are not aware of a report of silent extracranial vertebral artery dissection occurring during the first few months after carotid artery dissection. Had it not been for the initial dissection, angiography would not have been performed and the vertebral artery dissection would have remained undiagnosed. This suggests that extracranial vertebral artery dissection may be an underrecognized condition.

The second patient presented with a typical SAH and was found to have extracranial and intracranial vertebral artery dissection. SAH is a major manifestation of intracranial vertebral artery dissection, but the diagnosis of the underlying dissection is difficult and is most often made at surgery or at postmortem examination. Case 2 is remarkable in that the likely cause of her SAH was able to be determined by the demonstration of coexisting extracranial vertebral artery dissection suggested by the position of the aneurysmal image at the C1 level and by its normalization on subsequent angiograms. It is unlikely that the diffuse narrowing of the intracranial segment of the vertebral artery was due to arterial spasm given its localization and the absence of demonstrable intracranial aneurysm. It is more likely that the narrowing was akin to the "string" sign described for carotid artery dissection and that dissection of the vertebral and possibly basilar arteries was indeed the cause of the SAH. This case illustrates the importance of obtaining adequate angiographic views of the extracranial as well as the intracranial arteries in the investigation of SAH.

The third patient's symptoms (cervical pain, sensory deficit in the right C6 dermatome, and left cheek numbness) are difficult to analyze because the cervical pain and C6 signs were most likely due to the scoliosis itself. This does not, however, explain the cheek numbness, which despite a normal MRI was probably due to a small brainstem or thalamic infarct.

Our three cases are also unusual in their associations with other pathologic conditions. Dissection occurring at the site of a hemivertebra deformity (case 3) has not been previously described. The irregularity of the vessel lumen proximal to the apex of the scoliosis suggests some underlying intimal abnormality, but this is a matter for speculation.

There is no known association between cervical artery dissection and SLE or steroid therapy (case 2). It seems unlikely that SLE arteriopathy was a predisposing factor here given the absence of angiographic signs of arteritis and of laboratory indicators of SLE activity.

The first patient had two conditions known to be associated with cervical artery dissection, Marfan's syndrome and migraine. This patient fulfills one major criterion (dilated aortic root) and three minor criteria (asthenic build, joint laxity, and floppy mitral valve without incompetence) for the diagnosis of Marfan's syndrome. He also fulfills the International Headache Society's diagnostic criteria for migraine. The association of migraine with cervical artery dissection has been reported in individual cases and was shown to be statistically significant in a recent case-control study. In this patient, cerebral infarction could have been attributed to migrainous infarction or to embolism from the heart had cerebral angiography not been performed showing the dissecting aneurysm. This further illustrates the need for cerebral angiography to be performed early as part of the investigation of stroke in the young.

References


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