

## Headache in Cervical Artery Dissections

*Case History Submitted by Randolph W. Evans, MD*

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**Key words:** headache, cervical arterial dissection

**Abbreviations:** MRA magnetic resonance angiogram, ICAD internal carotid artery dissections, VAD vertebral artery dissections

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### CLINICAL HISTORY

A 36-year-old woman was eating dinner when she developed acute stabbing and throbbing pain going from the left anterior neck up to the left side of her face. There was no associated nausea, light sensitivity, visual symptoms, nor sensorimotor complaints. The pain was severe for about 5 hours. She went to an emergency room and received a narcotic injection, with benefit. The next morning the pain was still present but slight and then resolved that day. There was no history of head or neck trauma. For several months before admission she had been intermittently painting her house but had not done any painting of the house or ceilings for a number of weeks. Four days before the onset of pain she had her hair washed in a salon with her neck in an hyperextended position in the basin but had no associated symptoms. There was a history of intermittent tension-type headaches but no migraine and also of cervical and lumbar myofascial pain occurring intermittently. She had no history of hypertension, diabetes mellitus, ischemic heart disease, or cerebrovascular disease.

When I examined her the next day the general physical examination was normal except for left carotid bulb tenderness and left posterior cervical paraspinal tenderness, with full range of motion of

the neck. Neurological examination was entirely normal. A magnetic resonance image of the brain with magnetic resonance angiogram (MRA) sequence of the brain and neck revealed dissection of the upper cervical left internal carotid artery with marked stenosis of the arterial lumen and normal intracranial, vertebral, and basilar arteries. A cerebral arteriogram confirmed dissection of a long segment of the left internal carotid artery above the bulb and also demonstrated dissection of both vertebral arteries in their distal portions, with poor antegrade flow in the distal basilar artery. She was placed in a cervical collar and treated with heparin and warfarin. An ultrasound of the thoracic aorta and computed tomography of the chest were normal. An antinuclear antibody titer was elevated at 1/160 with a speckled pattern. Anti-DNA, Anti-SSA, SSB, and ANCA IgG, IgM levels were all negative. C3 and C4 levels were normal. An RPR was nonreactive. A urine homocysteine level was normal. At discharge her neurologic examination remained normal.

After discharge she was continued on warfarin, with an INR level ratio between 2 and 3. A repeat MRA study 9 months later showed only minimal residual irregularity in the cervical portion of the left carotid and both vertebrals. She was advised to stop warfarin, but she chose to continue to take a low dose of 2.5 mg daily for 15 additional months because of her concern about the possibility of recurrent thrombosis.

Four years later she experienced a few episodes of lightheadedness and a mild generalized "pressure"

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sensation in her head, lasting about 10 minutes. There was no head or neck trauma during this time. Magnetic resonance image of the brain was normal, and an MRA showed no change from the prior study. A cerebral arteriogram revealed a small short segment of dissection of the right internal carotid artery just superior to the carotid bulb and evidence of healing of the prior left carotid and bilateral vertebral dissections. She again was placed on warfarin. An antinuclear antibody titer was negative.

Three repeat MRA studies over the next 3 years showed evidence of healing of the prior multiple dissections. A skin biopsy was negative, demonstrating normal amounts of type I and type III procollagen and collagen. Warfarin was discontinued over 2 years ago, and she remains on daily aspirin.

**Questions.**—How often is headache the only manifestation of extracranial carotid or vertebral dissection? What are the features of the headaches associated with dissections? How common are multiple spontaneous dissections as evident in this case?

#### EXPERT COMMENTARY

With the remarkable advances made in noninvasive neurodiagnostic studies, a considerably larger number of patients with cervical artery dissections now are diagnosed. We now know that internal carotid artery dissections (ICAD) and vertebral artery dissections (VAD) are not rare and collectively carry an annual incidence rate of approximately 5 per 100,000 population.<sup>1</sup> Many important clinical points in connection with these dissections are now recognized, some of which are applicable to the case reported here.

First, in contrast to the initial reports based on autopsy observations, these dissections often convey a good prognosis; clinical recovery very good or excellent in approximately 85% of the patients, and resolution of or improvement in angiographic abnormalities occurs in 75% of the involved arteries. Although intracranial dissections (which especially in adults are much less common than extracranial dissections) carry a risk of subarachnoid hemorrhage, it would be most unusual for spontaneous extracranial dissections to rupture or bleed.<sup>2,3</sup>

Second, several distant clinical syndromes result from dissections. In ICADs these include hemicrania

and ipsilateral oculosympathetic palsy, hemicrania, and delayed manifestations of focal cerebral ischemia, hemicrania and ipsilateral lower cranial nerve palsies, or any combination of these with or without bruits or neck pain.<sup>1-3</sup> In a young patient, and especially if preceded or accompanied by headache, occurrence of lateral medullary syndrome either pure or with additional brainstem symptoms and signs, is highly suggestive of a VAD.<sup>4</sup> Even so, clinical variability is substantial. With the widespread availability of noninvasive neurodiagnostic studies, more and more cases of monosymptomatic, oligosymptomatic, or minimally symptomatic cervical artery dissections are identified that otherwise would have gone undiagnosed. Although actual figures are not yet available, today it is not rare to encounter patients with cervical artery dissection who present with headache only, bruit only, Horner's only, and so on. Furthermore, it is not unusual to identify asymptomatic cases, diagnosed either in the course of evaluation of dissection of a different vessel or evaluation of a different disease.

Third, multivessel dissections are common. In our series at least 20% of ICADs were bilateral, and for VADs the figure is even higher. In more than 8% of the patients with ICAD, we noted either a concomitant VAD or angiographic residue of a previous VAD. The associated VAD may be unilateral or bilateral. Among 200 patients with cervical artery dissections, five patients had concomitant bilateral ICAD and bilateral VAD.<sup>5</sup> Concomitant renal artery or other visceral artery dissections also can occur but only rarely. These plus several other observations raise the probability of an underlying disorder of the arterial wall in some dissection patients, likely involving an abnormality of elastin or fibrillin.<sup>1,6</sup>

Fourth, it is very uncommon to see recurrence of dissection in a previously dissected and healed cervical artery, but it is not so uncommon for recurrent dissection to occur in the other cervical arteries of the same patient. The risk of such recurrence is maximal within the first month (2%) after initial dissection and becomes much less thereafter (approximately 1% per year). The risk of recurrence is somewhat less for older patients and somewhat more for younger ones. In our patients we found the overall rate of recurrence at 10 years to be about 12%; for those

younger than 45 years the 10-year recurrence rate was 17%, whereas for those older than 45 years it was only 6%.<sup>5</sup>

Finally, despite the various clinical syndromes associated with cervical artery dissections, pain (headache, neck pain, or face pain) is the most common single manifestation and is noted in approximately 80% of the patients. Although the involved segment of the artery is in the neck, the resultant pain is much more commonly perceived in the head than in the neck. The headache is often steady and less frequently throbbing. It is typically unilateral, ipsilateral to the dissection, and often focal. Only sometimes is the headache bilateral, more so in VADs than in ICADs. In VADs the headache is typically occipital, whereas in ICADs the most common site is frontal, orbital, and periorbital, followed in order of frequency by pain in ear and mastoid area, temporal region, angle of mandible, cheek, and occipital region. Neck pain is more common in VADs than in ICADs (45% vs. 25%). In ICADs it is typically unilateral and involving the anterolateral aspect of the neck, whereas for VADs it is a bilateral or unilateral pain that involves the posterior neck.<sup>7</sup>

The case reported here highlights several of the important features of cervical artery dissections: headache may be the only manifestation of ICAD;

cervical artery dissection (in this case the accompanying VADs) can be asymptomatic or minimally symptomatic; occurrence of multivessel dissection is not unusual; and, although not especially common, recurrence of dissection may occur.

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