Short Communications

Spontaneous Dissection of the Extracranial Vertebral Arteries

L. R. Caplan, M.D.,* C. K. Zarins, M.D.,† and M. Hemmati, M.D.‡

SUMMARY Though the syndrome of carotid artery dissection is well known, "spontaneous" vertebral artery dissection is rarely recognized. We now report clinical and radiologic findings in five patients with presumed vertebral dissection, one pathologically confirmed.

Mean age was 35.2 years (range 27-41). Two were men; three women. None had hypertension, vascular disease, or trauma. Headache and neck or occipital pain was prominent in all, often preceding other symptoms. Four of five patients had unilateral partial altered medullary syndromes, in one accompanied by medial medullary signs. One patient had a cerebellar infarct. Angiography in four patients showed severe irregular stenosis of the distal extracranial vertebral artery (three bifurcally). A fifth patient with irregular stenosis above the vertebral origin had verified extensive dissection in the resected segment. No patient developed late ischemia. Repeat angiography in three showed healing.

We conclude that spontaneous vertebral artery dissection, though rare, has recognizable clinical and radiologic features.

"SPONTANEOUS" DISSECTION of the internal carotid artery causes prominent neck and facial pain, headache, ipsilateral Horner's syndrome, episodes of ipsilateral cerebral hemisphere and retinal ischemia, and sometimes "migrainous" visual accompaniments. The syndrome was analyzed in depth by Fisher et al.1,2 and subsequently has been widely reported.3-5 Migraine6,7 and fibromuscular dysplasia8 have been associated with arterial dissection. Intramural clot separates the media often compromising the lumen; clot can dissect into the lumen through the torn intima. Because of intimal and endothelial disruption platelet nidi can form in the lumen and reduced flow potentiates the development of intraluminal clot. Distal ischemia is caused by embolization of luminal clot or reduced distal flow. Dissections and intimal tears are usually traumatic and are termed "spontaneous" when no definite trauma is recalled by the patient. Though "spontaneous" dissection of the vertebral artery has been reported,1,9-14 the clinical and radiologic features are less well known than the findings in carotid dissection. We now report clinical and radiologic findings in 4 patients with presumed vertebral artery dissection and radiographic and pathologic findings in 1 patient with surgically proven vertebral dissection. The clinical and radiologic signs of spontaneous vertebral artery dissection closely resemble those described in vertebral artery trauma and chiropractic manipulation.15-23

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Case Reports

Patient 1*

D.J., a 31 year old woman awakened at 3 am on 4/17/82 with pain in the left occiput and neck and a left retroorbital headache. She had no important prior illnesses, but had a history of frequent prior headaches. She recalled no unusual recent neck motion or trauma. Two hours after awakening, she suddenly noted tingling at the corner of her left mouth and tongue, dizziness, vomiting, and a sensation that her body was being pulled to the left. Later her right hand felt cold. On examination, blood pressure and general examination were normal. Neurological abnormalities included: coarse horizontal nystagmus to the left, left facial hypalgesia, diminished left corneal response, and left limb dysmetria. She veered to the left when she walked. CT was normal. Bilateral vertebral angiography showed moderately severe narrowing and irregular stenosis of the third segment of the right vertebral artery and severe stenosis of a short segment of the third portion of the left vertebral artery (fig. la, b). Warfarin anticoagulation was begun. Neurologic examination returned to normal and she had no symptoms. A left vertebral angiogram 5 months after onset was normal with no residual stenosis (fig. lc). Warfarin was stopped. Two years after the original episode, the patient had no neurologic signs or symptoms.

Patient 2‡

J.R., a 39 year old man had noted for 10 years that alcohol use was followed by left ptosis and decreased sweating on the left face. In the past he had been in several car accidents and experienced other head traumas. Three months before admission he noted gait...
Patient 3

C.V., a 27 year old woman had a ski accident in 1975 but did not injure her head or neck. In February,
1978 she had a single spell of spinning dizziness. During October, 1979 she had unaccustomed generalized headaches. She was using oral contraceptives. In November, 1979, while driving a car, she suddenly developed dizziness, blurred vision, and slurred speech. A severe headache accompanied these symptoms. She staggered and could not walk. The symptoms became maximal within the first hour. Examination revealed: normal blood pressure, decreased pin sensation on the left face and right limbs, pendular vertical nystagmus, horizontal nystagmus especially to the right, clumsiness of the left hand, weakness of the left tongue, right hemiparesis, and bilateral extensor plantar responses. Angiography revealed marked irregularity of the third segment of the left vertebral artery as it exited from the transverse foramen of the axis and the fourth segment of this vessel as it entered the spinal canal (fig. 3). The basilar artery and its distal branches were visualized. However, there was persistent retrograde filling of the basilar artery from a left internal carotid artery injection. The right vertebral artery was hypoplastic and poorly visualized. She was not given anticoagulants. Examination in 1984 revealed persistent horizontal nystagmus, atrophy of the left tongue, and right hemiparesis. She had had no subsequent attacks.

Patient 4§

J.L., a 41 year old man noted one day in September, 1983 pain in the left neck and occiput. Several days later he discovered that neck turning would make him feel dizzy and lose his balance. On the day of admission, he noted spinning dizziness and could not walk. Examination revealed rotatory nystagmus on left lateral gaze and lethargy. CT, SER and BAER were nor-

§Referred by Dr. L. Barry of Denver, Colorado.
mal. At angiography the left vertebral artery did not fill distal to PICA. The right vertebral artery had a smooth tapered narrowing in the third segment of the vessel both at its exit from the transverse foramen of the axis and when it curved over the posterior arch of the atlas (fig. 4a, b). He was given corticosteroids. When he left the hospital, he felt much improved and walked well. Because of persistent headache, difficulty concentrating and discouragement 2 months later he had repeat angiography which showed a normal right vertebral artery (fig. 4c). The left vertebral artery was identified fluoroscopically in the neck but films were not obtained. By February of 1984, neurological examination was normal except for slight left nystagmus, slightly diminished pin perception in the left face, and slight overshoot of the left arm on rapidly dropping the arm from an elevated position. In May, 1984 he was still psychologically disturbed, but had no new neurological symptoms or signs.

Patient 5
R.L., a 38 year old woman had a past history of seizures, asthma, nasal polyps and extreme sensitivity to aspirin. On 3/18/84 3 minutes after inadvertently taking an aspirin containing pill she became dyspneic and arrived at a nearby hospital cyanotic and in circulatory collapse. Neurologic screening examinations were normal. She was resuscitated including intubation. Approximately 36 hours later, she complained of severe generalized headache and dizziness especially if she moved her head. CT four days later showed a left cerebellar infarct with deviation of the fourth ventricle to the right. She was given steroids. Neurological examination confirmed left limb dysmetria and slight gait ataxia. Left subclavian injection using arterial digital subtraction angiography showed an irregular narrowing in the first segment of the left vertebral artery just before entering the transverse foramen of C6. There was a pseudoaneurysm in the middle of this narrowed segment (fig. 5). On 4/16/84, a common carotid to vertebral artery bypass was created using a saphenous vein graft. The proximal diseased segment was removed. The specimen was sectioned serially and revealed a large subintimal hematoma compressing the lumen and friable irregular intima beyond the clot (fig. 6). Intraoperative angiography revealed normal distal flow and graft patency. The postoperative course was uneventful and she has had no further symptoms during the five months since surgery.

Discussion
Our case material includes five patients, three women and two men, all were young (27 to 41 years, average 35.2 years) and none had prior or current evidence of hypertension or cardiovascular disease. Two had a history of prior headaches compatible with common migraine. One patient was using oral contraception at the time of the stroke. Pain was a prominent and early finding in all patients, and usually was located in the occiput and posterior neck. Pain preceded other symptoms in four of five patients, and the fifth had

Figure 3. a, AP view and b, lateral view of left vertebral artery (11/79), showing severe irregularity of the third and fourth segments.
frequent unaccustomed generalized headache in the months before her stroke. Retroorbital and temporal headache accompanied the stroke in two patients. All 5 patients had generalized headache with their strokes. In four of five patients, neurological abnormalities included elements of the lateral medullary syndrome as follows: paresthesia and decreased pin perception on the ipsilateral face (4), nystagmus (3) (a fourth patient had diplopia and oscillopsia at onset but a month later had no nystagmus), "crossed" pin loss in the contralateral limbs (4), ipsilateral "cerebellar type" limb dysmetria (3), gait ataxia (4), and ipsilateral Horner's syndrome (1). Only one of these four patients had additional findings not usually associated with lateral medullary infarction; these signs were ipsilateral tongue paralysis and crossed hemiplegia. Combined ipsilateral, medial and lateral medullary ischemia has been reported in vertebral artery occlusion.23,24 This was the only patient in whom the vertebral artery dissection extended intracranially and was also the only patient with a significant fixed residual neurologic deficit. The patient without lateral medullary findings had a cerebellar infarct confirmed by CT. Lateral med
Figure 5. AP view of the left subclavian artery (digital subtraction angiography). The irregular first segment of the left vertebral artery (single arrow) has a pseudoaneurysm (arrow head). Post aneurysmal stenosis (double arrows) is seen.

ullary syndrome and cerebellar infarction are the two commonest findings in groups of patients with occlusion of the third or fourth portion of the vertebral artery.26 The neurologic deficit developed abruptly in three patients, and seemed to evolve gradually during days to weeks in the other two patients. Only two patients had known preexisting trauma, in one presumably not directly affecting the head or neck. In one patient the onset followed resuscitation which could have led to an unusual neck position. Treatment included anticoagulants in two patients and corticosteroids in two patients. Follow up information revealed that no patient developed new neurologic symptoms or signs after their original stroke (five to 55 months, average 21 months).

Angiography revealed extensive lesions in the distal extracranial vertebral arteries in four patients. In three the lesion was clearly bilateral, and in the fourth the contralateral vertebral artery was poorly opacified. Irregular stenosis usually began in the third segment of the artery as it exited from the transverse foramen of the axis and commonly extended to the horizontal portion of this segment as it traveled over the posterior arch of the atlas. In one patient the stenosis extended to the fourth portion of the vertebral artery as it entered the spinal canal just before entering the foramen magnum. In the fifth patient, the stenotic portion was in the first segment for the left vertebral artery with what appeared to be a small pseudoaneurysm in the middle of the affected segment. When the angiographic lesions were bilateral, the symptoms always were in the territory of the more severely affected vertebral artery. Repeat angiography showed definite improvement in three patients, and interoperative angiography was normal in a fourth. Angiography has not been repeated in the remaining patient. No patient had associated fibromuscular dysplasia and carotid angiography was available on all patients.

Pathological material was available only in the fifth patient, whose abnormal vertebral artery segment was removed at the time of a common carotid to vertebral artery bypass. The intima was friable and the endothelium partially denuded. There was a large subintimal hematoma with more luminal encroachment than could be predicted from angiography.

"Spontaneous" dissection of the extracranial vertebral artery has been rarely reported. Table 1 describes data from the present series of patients and table 2 includes data from prior reports. In three reports, the vertebral artery dissections were accompanied by bilateral internal carotid artery dissection;8, 10, 11 the clinical findings in these patients were referable to hemisphere ischemia and no definite posterior circulation

Figure 6. Transverse section of left vertebral artery. Luminal stenosis due to subintimal dissection of blood. The space between the intima and media is filled with organizing thrombus (approx. 3 weeks old).
### Table 1. Spontaneous Dissection of Extracranial Vertebral Artery — Presently Reported Cases

<table>
<thead>
<tr>
<th>Authors</th>
<th>Age</th>
<th>Sex</th>
<th>Predisposing factors</th>
<th>Pain locus</th>
<th>Clinical syndrome</th>
<th>Vessel lesion locus</th>
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<td>1</td>
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<td>L occiput neck</td>
<td>L lat medullary</td>
<td>R VA-C2</td>
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<td></td>
<td>2</td>
<td>M</td>
<td>Past trauma</td>
<td>Headache R jaw</td>
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<td>R VA-C2</td>
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<td></td>
<td>3</td>
<td>F</td>
<td>Past ski accident</td>
<td>Headache</td>
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<td>L VA C2-C1</td>
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<td></td>
<td>4</td>
<td>M</td>
<td>—</td>
<td>Headache L neck</td>
<td>L lat medullary</td>
<td>L VA C2-C1</td>
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<td>5</td>
<td>F</td>
<td>Recent intubation</td>
<td>Headache</td>
<td>L cerebellar</td>
<td>L VA T1-C6</td>
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</table>

VA = vertebral artery; ICA = internal carotid artery; PICA = posterior inferior cerebellar artery.

Signs were noted. The patient of Fisher et al’s was mentioned en passant and was not described in detail. Prior reports included nine patients, six women and three men. Ages ranged from 26–56 years (average 37.5 years). In two patients symptoms began during sports activities (skiing and playing softball). In another patient posterior circulation ischemia began postoperatively, possibly related to neck position during surgery, intubation, or anesthesia. This circumstance is similar to our patient 5 whose symptoms began after resuscitation with intubation. Headache or face or neck pain was a prominent feature in six patients and was not mentioned in the other three patients. In the six patients with recognized posterior circulation symptoms, three had features of the lateral medullary syndrome combined in one with medial medullary ischemia. In five patients most of the signs resolved satisfactorily; only two were left with important residual deficits and two patients died. In only three patients was there confirmation of the dissection, two at ne-

### Table 2. Spontaneous Dissection of Extracranial Vertebral Artery — Previously Reported Cases

<table>
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<tr>
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<th>Clinical syndrome</th>
<th>Vessel lesion locus</th>
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<td>Alpert et al^-10-</td>
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<td>Oral contraceptives</td>
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<td>R ICA origin</td>
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<td>Mokri et al^-11-</td>
<td>7</td>
<td>F</td>
<td>Migraine fibromus-</td>
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<td>Ringel et al^-8-</td>
<td>8</td>
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<td>Bradac et al^-12-</td>
<td>9</td>
<td>F</td>
<td>Migraine</td>
<td>—</td>
<td>R lat “brainstem”</td>
<td>L VA C2-C1</td>
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<td>Goldstein^-13-</td>
<td>10</td>
<td>F</td>
<td>—</td>
<td>L headache</td>
<td>R lat &amp; medial medul.</td>
<td>R VA near orgn.</td>
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<td>Bladin^-14-</td>
<td>11</td>
<td>F</td>
<td>—</td>
<td>Occiput neck</td>
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<td>L post crbrl. artery</td>
<td>R VA near orgn. to C2</td>
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<td></td>
<td>12</td>
<td>M</td>
<td>Migraine</td>
<td>R occiput neck</td>
<td>“Bulbar” paralysis</td>
<td>R VA C4-C2</td>
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<td>Bostrom et al^-9-</td>
<td>13</td>
<td>F</td>
<td>Post-op</td>
<td>—</td>
<td>Vomiting</td>
<td>R VA C4-C2</td>
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<td>Sudden post-op death</td>
<td>L VA</td>
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<tr>
<td>Fisher et al^-1-</td>
<td>14</td>
<td>M</td>
<td>—</td>
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<td>R lat medullary</td>
<td>R VA C3-C1</td>
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VA = vertebral artery; ICA = internal carotid artery; PICA = posterior inferior cerebellar artery.
cropsy and one at surgery. In two series, the authors were uncertain of the nature of the lesion and called the process "idiopathic regressing arteriography" and "reversible angiopathy."10 In ten vertebral arteries the site of the lesion was specified; six affected the distal extracranial artery from C3-C4, two lesions began near the origin and two began at C6. Migraine (3) and fibromuscular dysplasia (1) were occasional accompanying conditions.

It is now well known that vertebral artery dissection can follow trauma, neck manipulation, or sudden head or neck turning.27 At least 15 patient reports are available in which the vascular lesion affected the extracranial vertebral artery and was well delineated angiographically or at post-mortem.15-22 Among these 15 patients, 10 vertebral dissections followed chiropractic manipulation and one followed neck manipulation by a dentist; other incitants included automobile accidents (2), a direct blow to the neck by a log (1), and turning abruptly while driving a car (1). In 14 patients, the vascular lesion was located in the distal extracranial third segment of the vertebral artery as it occurred behind the atlas. The horizontal limbs of this distal vascular loop were often irregularly stenosed, and sometimes contained pseudoaneurysmal dilatations. In only one patient who had direct trauma to the neck by a moving log did the lesion involve the proximal vertebral artery just distal to its origin. The first and third portions of the vertebral artery are movable whereas the second and fourth segments are relatively fixed by bony attachments; this anatomical fact probably explains the predilection for tearing in the third and first portions of the artery. In contrast, atherosclerosis invariably affects the origin of the vertebral artery or the intracranial fourth segment.26 Only temporal arteritis and oral contraceptive use have been associated with lesions affecting the distal extracranial vertebral artery before it pierces the dura. Occipital and nuchal pain and generalized headache were often noted in patients with traumatic dissections. The clinical syndrome usually included features of lateral medullary or cerebellar infarction. In an additional report not analyzed above because angiography was not available, young people usually during or after vigorous physical exertion had acquired lateral medullary infarcts with a benign late course;28 these lesions were most likely also related to dissection caused by vigorous neck turning. Although we have called our cases "spontaneous" dissections, they may well have had unrecognized trauma or sudden neck motion forgotten or deemed insignificant by the patient and so not reported to the physicians.

In the absence of pathological material from each patient, we can not be absolutely certain that four of our patients had dissections. Features that favor dissection include: (1) the prominence of pain; (2) absence of known atherosclerotic or other vascular disease; (3) location of the vascular lesions; (4) close resemblance angiographically to cases of "traumatic dissection," and (5) improvement or normalization of the lesions in repeat angiography.

Dissection of the vertebral artery probably occurs more commonly than is presently recognized. The important features of the syndrome are (1) frequent history of sudden neck movement, trauma, chiropractic manipulation, or intubation; (2) prominent headaches, occipital or nuchal pain; (3) clinical findings compatible with lateral medullary or cerebellar infarction; (4) at angiography, irregular stenosis of horizontal loops of the distal extracranial vertebral arteries, usually bilaterally, as they loop behind the atlas. The syndrome should be especially considered in young patients with
no known predisposing vascular risk factors for atherosclerosis or cerebral embolism.

Acknowledgments

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References

Spontaneous dissection of the extracranial vertebral arteries.
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