Pseudoaneurysm of the extracranial vertebral artery

Case report

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The unusual association of a giant extracranial vertebral artery pseudoaneurysm, intracranial aneurysms, and extracranial carotid occlusion in a woman with neurofibromatosis is presented. Pain as a result of expansion of the mass in the soft tissue of the neck led to her seeking evaluation. Herniation of the mass intraspinally between the occiput and C-1 resulted in myelopathy. Following balloon occlusion of the vertebral artery, the mass and associated symptoms resolved without the need for direct resection. The salient features of these unusually associated problems are discussed.

KEY WORDS: neurofibromatosis • pseudoaneurysm • vertebral artery • aneurysm

Due to its relatively protected location, the extracranial vertebral artery in its second and third portions is infrequently injured by penetrating trauma, although it is vulnerable to blunt trauma or rotational injury. Uncommon nontraumatic lesions of the distal extracranial vertebral artery include dissection, arteriovenous fistulae, and aneurysm formation. A case of neurofibromatosis associated with intracranial aneurysms and extracranial carotid artery occlusion, in which the most striking feature was a progressively enlarging distal extracranial vertebral artery pseudoaneurysm, is the subject of this report. The size and location of this aneurysm led to the choice of endovascular occlusion of the vertebral artery proximal to the aneurysm neck, which resulted in an excellent recovery. The interrelationship between neurofibromatosis, arterial lesions, and trauma is discussed in this case report.

Case Report

This 52-year-old Caucasian woman was admitted for evaluation of a painful enlarging mass on the left side of her neck. Two weeks previously she had undergone an elective abdominal hysterectomy and on the 1st postoperative day had noted neck pain which became progressively more severe. She was subjected to two chiropractic manipulations in an effort to obtain relief. After the second manipulation the chiropractor noted a mass on the left side of her neck. In spite of oral analgesics, the neck pain became intolerable and the mass continued to enlarge.

The patient's medical history was significant for von Recklinghausen's disease, manifested by multiple peripheral neurofibromas. She had no symptoms of cerebral ischemia and was not hypertensive. There was no history of blunt or penetrating trauma to the head or neck.

Examination. There were numerous café-au-lait spots over the patient's trunk and extremities as well as multiple cutaneous neurofibromas varying in size from 2 mm to 1.5 cm. A 4 × 5-cm mass, with an associated bruit, was present in the left posterior aspect of the neck extending from the skull base down to approximately C-4. A bruit was also heard over the right carotid artery. Neurological examination revealed decreased hearing in the left ear and weakness of the left trapezius and sternocleidomastoid muscles. Her gait was mildly broad-based, but she was able to tandem-walk. No specific abnormalities of cerebellar function were evident. The muscle stretch reflexes were normal and plantar stimulation resulted in a flexor response.

Over the next 10 days the patient's neurological condition slowly worsened and the mass increased in size to 7 × 9 cm. She developed weakness in the left deltoid muscle, hyperactive reflexes in the left lower extremity, left ankle clonus, and left-sided dysmetria. A contrast-enhanced computerized tomography (CT)
scan revealed moderate lateral ventricular dilatation and a large mass in the soft tissues of the left side of the neck (Fig. 1 left). The central portion was filled with contrast medium, but the outer layers were of variable density suggesting partial thrombosis. At the skull base, the mass herniated through the foramen magnum and compressed the lower portion of the medulla (Fig. 1 right).

Transfemoral cerebral angiography revealed numerous abnormalities. The right internal carotid artery was completely occluded at its origin. A 9-mm saccular aneurysm arose from the cavernous portion of the left posterior cerebral artery, as well as a 3-mm saccular aneurysm from a branch of the left posterior cerebral artery. At the C-2 level, the left vertebral artery filled a large aneurysmal sac (Fig. 2). No filling was seen distal to the aneurysm. Injection of the right vertebral artery revealed it to be large and mildly tortuous. It filled the basilar artery, its branches, and the right posterior inferior cerebellar artery (PICA). The left PICA was not visualized on either vertebral artery injection. A second angiogram was obtained the following day in an attempt to demonstrate filling of the left PICA. With temporary balloon occlusion of the left vertebral artery, contrast material injected into the right vertebral artery did not reflux into the left PICA.

**Embolization.** Two days later, the patient underwent placement of a detachable balloon in the left vertebral artery just proximal to the neck of the aneurysm. It was technically impossible to place a second balloon distal to the neck of the aneurysm. After inflation of the balloon, somatosensory evoked potentials and brain-stem auditory evoked responses were monitored for 20 minutes prior to detachment of the balloon. These studies showed no alteration following balloon inflation or release. Although the pulsations in the mass ceased, there was no immediate change in the neurological findings.

**Postembolization Examination.** Six weeks after balloon occlusion, the neck mass had diminished in size. The patient's neck pain had completely resolved as had her muscle weakness. The reflexes were normal and the ataxia and ankle clonus had resolved. A follow-up CT scan showed no further enhancement of the aneurysm and a decrease in the displacement of the medulla (Fig. 3). She had returned to work and was functioning at her premorbid capacity.

**Discussion**

The extracranial vertebral artery is arbitrarily divided into three divisions. Due to the unique bone protection inherent to the second portion and the deep anatomical location of the first, reports of vertebral artery injury to these two segments are rare. Trauma to the neck causing pseudoaneurysm formation in the first and second portion is most often penetrating in nature (stabbing, gunshot, or shrapnel). The frequency of survival in these cases is further reduced by the first portion's proximity to the common carotid artery, where over 50% of penetrating injuries cause fatal hemorrhage. Blunt nonpenetrating trauma to the cervical spine has been shown to cause similar arterial damage.

In contrast, the distal second and third portions are more often injured by nonpenetrating trauma. In the atlanto-occipital joint complex, the vertebral artery is fixed at two sites: proximally as it exits from the transverse foramens of C-1 and distally as it traverses the posterior atlanto-occipital membrane. Apart from these sites of fixation, the vertebral artery can be compressed or stretched by cervical rotation. In cadaver studies, the luminal diameter of the vertebral artery could be reduced 90% by movements within the normal range. Pure rotation was the most effective in reducing flow, but this was also achieved using rotation combined with extension or flexion. Similar results were achieved in
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asymptomatic volunteers where vertebral artery occlusion was induced in eight of 43 subjects by varying the head position.12

Chiropractic manipulation resulting in vertebral artery injury and stroke is well documented.5,16,19,26 In 1980, Krueger and Okazaki19 reviewed this problem, finding 28 previously reported cases; they added 10 of their own. From examination of the type of cervical manipulation used, it appears that pure rotation followed in frequency by combinations of rotation along with flexion, extension, tilting, or stretching leads to vertebral artery damage. Although some authors have reported transient neurological deficits,25 the majority of patients develop symptoms either at the time of manipulation (followed by a progressive neurological decline) or after a brief asymptomatic interval. Our patient's persistent neck pain after each treatment could be related either to the enlarging mass lesion or to arterial spasm induced at the time of initial injury. In other cases with persistent headache, arterial spasm and occlusion have been visualized on angiography.19,26

Neurofibromatosis and Ehlers-Danlos syndrome are both diseases with a generalized abnormality of connective tissue composition, and have been associated with single cases of nontraumatic pseudoaneurysms.2,25 Stenosis or occlusion of the internal carotid artery and middle cerebral artery have been reported as a complication of neurofibromatosis in children and adolescents.29 Fibromuscular dysplasia (FMD) is a similar disease in that there is disorganized fibrosis of the arterial intima, media, or adventitia. Cases of FMD with nontraumatic vertebral artery involvement have been reported more frequently.22,27 On the intracranial portion of the cerebral angiogram, our patient showed multiple saccular aneurysms and occlusion of one extracranial carotid artery, pointing to the diffuse nature of her vasculopathy. This diffuse involve-

ment has been noted in both neurofibromatosis and FMD.6,23,27 While it is suspected that irregularities of the intima and/or media allow these aneurysms to develop, the exact pathogenesis is unknown. In the case described, we were unable to determine the exact cause of the pseudoaneurysm. Nonspecific neck pain could have led to the cervical manipulation with subsequent vertebral artery damage. However, a spontaneous pseudoaneurysm could also have developed, leading to the neck pain and the chiropractic manipulation following which the neck mass was noted.

Symptoms secondary to involvement of the vertebral artery by a pseudoaneurysm are variable and result from either mass effect or arterial insufficiency. Headache and neck pain are frequent. A partial or complete Horner's syndrome may develop. Fullness of the oropharynx, dysphagia, or a brachial plexus neuropathy may occur secondary to compression of adjacent tissue. In a similar manner, a radiculopathy or phrenic nerve paresis may develop. As in this case, a large pseudoaneurysm with intraspinal or intracranial extension can lead to cervical myelopathy or brain-stem or lower cranial nerve dysfunction. Symptoms of lateral or medial medullary ischemia or infarction may occur secondary to emboli or thrombosis of the vertebral artery or PICA. The distinguishing feature of a vertebral artery pseudoaneurysm is a palpable mass which is nontender, nonfluctuant, possibly pulsatile, and accompanied by a

FIG. 2. Angiogram, lateral view, showing the left vertebral artery (large arrow) and demonstrating the neck (arrowhead) and angiographic border (small arrows) of the pseudoaneurysm.

FIG. 3. Axial computerized tomography scan 2 months after vertebral artery occlusion showing a decrease in the size of the pseudoaneurysm (arrows) and resumption of the normal shape and position of the caudal medulla (arrowhead).
thick or bruit. The most reliable clinical sign of vertebral artery injury is the persistence of a bruit or thrill when the common carotid artery is transiently occluded.18

Arterial kinking or the formation of an aneurysm or pseudoaneurysm along the second portion of the vertebral artery may cause sufficient widening of the cerebral intervertebral foramen or foramen intertransversarium to be seen on radiographs of the cerebral spine. However, more frequent causes of such an anomaly are neurofibroma, meningioma, fibroma, lipoma, or traumatic meningocele. Cervical myelography, CT, and magnetic resonance imaging may be useful in cases of myelopathy to delineate the borders and extent of extradural lesions. Angiography is required in all cases to: 1) verify the vascular anomaly; 2) delineate the neck of the pseudoaneurysm; and 3) define the involvement of the remainder of the vertebrobasilar system. The usefulness of arteriography was illustrated in a study of Meier, et al. Three years after instituting routine four-vessel cervical angiography in all cases of severe trauma to the neck, they noted a rise in the diagnosis of vertebral artery damage to 19.4% of all cases of cervical vascular injury from the 3% recorded for the 16 years prior to changing the policy.

Historically, the surgical treatment of vertebral artery injuries resulted in high rates of morbidity and mortality. As early as 1836, Sanson commented that “wounds of this vessel are beyond the resources of art.” In a review of 42 extracranial vertebral artery injuries in 1893, Matas reported a combined mortality rate of 80%. A review of acute vertebral artery injury by Fogelman and Stewart reported a 50% mortality rate. Vertebral artery ligation alone carries an 8% chance of central nervous system ischemia. Direct surgical approaches to the vertebral artery are made difficult by the bone canal formed by the foramina of the transverse process, an extensive periarterial venous plexus, numerous sources of collateral blood flow, and the risk of vertebrobasilar ischemia and infarction.

Some authors have reported successful treatment of carotid-cavernous and vertebro-vertebral fistulas, with conservation of normal carotid or vertebral blood flow, by the placement of detachable intravascular balloons. Ideally, the aneurysm should be trapped by placing a balloon above and below it. This accomplishes two things: 1) it protects against migration of a balloon if it should deflate; and 2) it prevents retrograde filling from the other vertebral artery. In this case we could not place a distal balloon for technical reasons and settled for occlusion of the neck of the pseudoaneurysm where it arose from the vertebral artery after adequate collateral blood flow was verified. We did not consider that aneurysm resection was indicated. Gradual resorption of the clot within the pseudoaneurysm occurred over a period of weeks with resolution of the myelopathy. While leakage and eventual collapse of balloons placed for arterial occlusion is the rule, studies have shown them to retain 80% to 90% of their original volume for a week or longer. Arterial occlusion for this length of time should allow for formation of a well-organized thrombus with vessel occlusion. Temporary occlusion prior to balloon detachment is useful in detecting potential ischemic deficits. During temporary balloon occlusion of the left vertebral artery, this patient’s clinical status and electrophysiological studies did not suggest vertebrobasilar ischemia.

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