Cervical Epidural Arteriovenous Malformation Occurring with a Spinal Neurofibroma

Case Report

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Vascular lesions of the spinal cord are a well-recognized entity, having been comprehensively reviewed by Elsberg,† Wyburn-Mason,‡ and more recently by Odom.¶ Epidural vascular lesions are commonly hematomas or hemangiomas. Arteriovenous fistulas involving the vertebral vessels in the neck have occasionally been reported.1,2,3 Nearly all of these fistulas have resulted from penetrating neck injuries, and the venous component has not primarily involved the epidural space. To our knowledge, only a single case of congenital arteriovenous malformation of the vertebral vessels has been reported.5 The purpose of this report is to present an unusual case having this rare epidural vascular anomaly in addition to a spinal neurofibroma. We also wish to emphasize the method of angiographic study using selective arterial catheterization with subtrac-

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History. The patient, a 17-year-old right-handed Negro male, was admitted on December 17, 1965, with complaints of weakness of the right arm and pain in the neck and right shoulder. While lifting a heavy object in August, 1965, he turned his head from right to left and immediately felt pain in the neck. This later radiated to the right shoulder. The pain was intermittent, occurring 3 to 4 times per day and at night. Cervical motion and coughing increased the pain, which was worse nocturnally. In November, 1965, his neck was stiff and he had a limited range of cervical motion. Numbness began in the right index finger and then involved the remaining fingers of that hand. A progressive weakness of the right arm and hand developed. The pain pattern changed in that the intermittent shooting pains radiated from the right side of the neck to the ipsilateral shoulder and distally to the forearm. Analgesics relieved the pain. On admission he was unable to use the right arm in feeding and dressing himself. There was no history of blunt or penetrating neck trauma.

The patient’s 43-year-old mother was thought to have neurofibromatosis. His 16-year-old brother had multiple cafe-au-lait spots, pedunculated cutaneous neurofibromas, and hypertension.

Examination. Pertinent physical findings were: blood pressure 120/70 mm Hg, cafe-au-lait spots, a 6-cm flat, subcutaneous, non-tender mass on the medial aspect of the right leg. The right arm showed a flail defect due to paresis of the right spinal accessory nerve and atrophy of the pectoralis major, deltoid, biceps, and lower trapezius muscles. Tone was diminished in the right arm, which he was unable to elevate above his head; the weakness was greater proximally than distally. Perceptions of pinprick and temperature were diminished in the C-2 through T-2 dermatomes. The deep tendon reflexes were hypoactive in the right arm but equal in the legs. The plantar response was flexor bilaterally. The range of cervical motion was decreased.

Lumbar cerebrospinal-fluid pressure was 280 mm of water, the protein 640 mg%, the Pandy positive, and the cell count 6 red-blood cells per cubic millimeter.

Plain films of the cervical spine showed enlargement of the right neural foramen at C2-3 with erosion of the posterior aspects of the vertebral bodies (Fig. 1). A cervical myelogram showed nearly complete obstruction of the cranial flow of contrast medium at the level of C-5 (Fig. 2). A linear filling defect in the contrast column was noted immediately below the level of obstruction.

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First Operation. On December 24, 1965, a laminectomy was performed from C-2 through C-7 with partial removal of facets of C2-3 and C5-6. A purplish mass was noted in the right gutter. This was completely epidural. The dura was opened and the spinal cord was noted to be rotated 90° from right to left with considerable tension on the roots (Fig. 3). On palpation through the open dural sac, epidural tumor was felt anteriorly from C-2 through C-6. No intradural component of this lesion was found. Biopsy of the purplish mass at C5-6 resulted in a sudden hemorrhage. When the bleeding had been controlled, a network of arterial vessels was noted in the epidural space at C5-6. The epidural tumor at C2-3 was then biopsied; this too resulted in brisk hemorrhage. The patient required 3500 cc of whole blood during the operation. Biopsy specimens were interpreted as normal blood-vessel wall.

Postoperative Studies. The following definitive angiographic studies were performed postoperatively to determine the nature and extent of the vascular lesion:

1. A selective right-vertebral angiogram demonstrated filling of a large arteriovenous malformation located within the cervical vertebral canal (Fig. 4). The venous component extended virtually the entire length of the cervical region and drained inferiorly into the superior mediastinum.

2. A selective left-vertebral angiogram showed nearly the entire bolus of contrast medium refluxed down the distal right-vertebral artery with only minimal, transient filling of the basilar artery (Fig. 5). Radicular branches from both vertebral arteries were
3. Separate catheter injections of the contrast medium into both proximal subclavian arteries excluded the possibility of additional vessels feeding the malformation from the thyrocervical and costocervical subclavian branches.

4. Bilateral selective internal and external carotid cerebral angiograms showed major feeding of the malformation from the occipital branch of the right external carotid artery via the distal right vertebral artery through suboccipital communications (Figs. 7, 8). Both posterior cerebral arteries were well filled from the internal carotids via their respective posterior-communicating arteries. No feeding of the malformation was demonstrated from the left external carotid artery.

On January 24, 1966, the right vertebral and occipital arteries were to be ligated at the atlanto-occipital junction and superior nuchal line respectively. However, after intubation, cardiac arrest occurred. The patient responded to external massage, but the operative procedure was cancelled.

Second Operation. The patient underwent a resection of the arch of the atlas and clipping of the right vertebral artery on January 28, 1966. At this time, the right lateral masses of C-2 and C-3 vertebrae were removed, and pinkish-red extradural tissue was noted. This was separated from the dura by blunt dissection and a biopsy obtained.
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FIG. 5. Anteroposterior projection of selective left vertebral angiogram showing reflux down the distal right vertebral artery.

FIG. 6. Right posterior oblique projection of selective left vertebral angiogram demonstrating minor feeding vessels at C3-4.

(Fig. 3). Attempt at further removal of this lesion caused brisk hemorrhage. The biopsy diagnosis was neurofibroma.

Further ligation of the malformation was planned for February 7, 1966. During induction of anesthesia, the patient sustained a cardiac arrest and died.

Discussion

We believe that there are three interesting
features of this case, as described below:

1. The presence of a cervical epidural arteriovenous malformation. The malformation was presumed to be congenital in origin in view of no known history of neck trauma. Although one case of congenital arteriovenous malformation of the vertebral vessels has been previously reported, the exact location of the malformation was not demonstrated; therefore, it is not known whether its venous component was epidural.

2. The presence of another abnormality (spinal neurofibroma). This was the primary diagnosis in view of the clinical history, family history of neurofibromatosis, and radiological changes on the plain cervical spine films. The myelographic findings were initially interpreted as representing congested vascularity secondary to a spinal tumor at a higher level. We wish to stress the importance of performing preoperative angiography in cases demonstrating myelographic findings which could be produced by primary vascular lesions unrelated to other suspected disease processes.

3. The importance of using selective arterial catheterization techniques in demonstrating vascular lesions of this type. Successful surgical management depends upon accurate visualization of the precise origin and number of feeding vessels. This can best be achieved with selective catheter techniques. One must study all vessels of the vertebral-basilar and carotid systems to avoid missing unsuspected, minor feeders. This complete angiographic approach is also essential for determining all routes of cerebral blood flow prior to vertebral ligation.
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Corresponding frontal projection of selective right external carotid angiogram.

The patient’s lack of neck mobility prevented the use of standard frontal and lateral cervical projections. Therefore, serial angiography of the cervical area was performed in oblique projections to visualize the area of interest with a minimum of superimposed structures. A photographic subtraction technique was used in an effort to eliminate confusing skeletal structures and residual myelographic contrast medium in the cervical area.

Summary

We have presented a case of cervical epidural arteriovenous malformation and have described various neuroradiological aids we found essential to a complete visualization of the malformation and its origins. We have also commented on the presence of a verified cervical-spinal neurofibroma in the same individual.

References