Cervical Epidural Arteriovenous Malformation Occurring with a Spinal Neurofibroma

Case Report

Harry B. Hoffman, M.D.,* and Merwyn Bagan, M.D.
Department of Radiology and Division of Neurological Surgery, The Johns Hopkins University School of Medicine and The Johns Hopkins Hospital, Baltimore, Maryland

Vascular lesions of the spinal cord are a well-recognized entity, having been comprehensively reviewed by Elsberg,3 Wyburn-Mason,4 and more recently by Odom.5 Epidural vascular lesions are commonly hematomas or hemangiomas. Arteriovenous fistulas involving the vertebral vessels in the neck have occasionally been reported.1,2,5 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 subtraction.

Case Report

History. The patient, a 17-year-old righthanded 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, and 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.
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