The value of preoperative angiography in the surgical management of cervical hourglass neurofibroma

Case report

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The author describes the case of an unusually large extradural cervical hourglass neurofibroma in which preoperative angiography had revealed lack of filling of the ipsilateral vertebral artery, as well as the complete vascularization of the neurofibroma. Complete preoperative angiographic evaluation in all cases of cervical dumbbell neurofibromas is felt to be essential in planning the operative procedure.

KEY WORDS · cervical dumbbell neurofibroma · vertebral artery · arteriography

The first successful surgical attempt at removing an intraspinal tumor was reported by Gowers and Horsley in 1888. Since then, the advent of the x-ray film and contrast myelography has added very precise tools for the diagnosis of these tumors. Love and Dodge were the first to focus attention on the close contiguity of cervical dumbbell neurofibromas with the vertebral artery and to stress that this could pose a serious problem in surgical management. In spite of their concern, however, no suggestions were made to investigate these anatomical relationships by way of angiography. Since then, only two reports have appeared in the literature in which preoperative vertebral arteriography was felt to be essential in recognizing the size of the tumor and its exact position. Kriss and Schneider reported three cases of cervical neurofibromas in which myelography had been “unrevealing,” but in which arteriography demonstrated medial and anterior displacement of the vertebral artery by the hourglass tumor. Only in their second case did they demonstrate slight narrowing of the lumen of the vertebral artery. In none of their radiographs was there any actual visualization of the vascular supply of the tumors.

The second report can be found in Verbiest’s monograph, La Chirurgie Antérieure Et Latérale Du Rachis Cervical. In his cases, Verbiest had taken advantage of myelography as well as vertebral angiography, both of which he felt to be, “essential diagnostic procedures.” Findings similar to those described by Kriss and Schneider were reported.

In the following case, arteriography not only demonstrated the lack of filling of the ipsilateral vertebral artery but also gave the opportunity to describe, for the first time, the vascular supply of an unusually large extradural, hourglass cervical neurofibroma.

Case Report

A 67-year-old woman was admitted in
January, 1968, because of progressive difficulty with the use of the left hand, accompanied by pain in the thumb, and later by pain in the left arm and in the upper scapular and nuchal areas. For a few months, the patient had also noted tingling in the tips of the fingers of the right hand, as well as some weakness in her legs. The patient denied any sphincter involvement.

**Examination.** A large, nonpulsating mass was found in the left supraclavicular lower lateral cervical region, separable from and posterior to the carotid artery, which was quite evidently displaced anteriorly. The mass measured approximately 5 × 6 cm.

Hyperextension of the head elicited a radicular type of pain radiating into the left upper scapular region, as well as along the outer arm, down to the elbow. There was moderate weakness of abduction and external rotation of the left arm, as well as marked weakness of the left triceps and dorsiflexors of the left hand and wrist. The grip of the left hand and interossei function were markedly compromised. There was advanced atrophy of the small muscles of the left hand. The biceps reflexes were both hypoactive, whereas the triceps reflexes were very hyperactive. The Hoffman sign was negative bilaterally but both plantar responses were equivocally extensor. There was loss of vibratory sensation in the arms and legs, most marked in the distal portion of the upper extremities. There was a peculiar hyperesthesia of the entire left arm. The patient could not identify objects with the left hand. Her gait was unstable but was not felt to be truly ataxic.

X-ray films of the cervical spine revealed an extensive, destructive process of the lateral masses of C-4, 5, and 6 on the left side. There was excavation of the lateral and posterior borders of the vertebral bodies from C-4 to C-6. The individual intervertebral foramina from C-3 to C-7 were no longer visible. Indeed, only a single large foramen was present (Fig. 1). Cervical myelogram disclosed a complete block of the Pantopaque column at the level of the body of C-6. The upper contour was very irregular, suggesting an extradural mass. The cervical cord was clearly seen to be displaced to the right. The spinal fluid protein content was 115 mg%.

Because of the certainty of having to deal with the vertebral artery, the tumor-artery relationship was clarified by arteriography. A left retrograde brachial arteriogram revealed lack of filling of the left vertebral artery. The blood supply to the tumor was clearly demonstrated (Fig. 2). The nonvisualization of the vertebral artery was felt to be due to the progressive, extraluminal pressure exerted by the very large tumor mass which had totally encased and eventually occluded the vertebral artery. The rare possibility of aplasia of the artery could not be ruled out.

**Operation.** The operative removal of the tumor was carried out in two stages. As advised by Love and Dodge, the laminectomy was performed first. The tumor was entirely extradural. No vascular structure resembling the vertebral artery or its venae comitantes could be recognized. Because of the
Preoperative angiography for cervical neurofibroma

Fig. 2. Upper Left: Left retrograde brachial arteriogram showing excellent filling of the axillary and subclavian arteries. The left vertebral artery is not visualized. The ascending cervical artery (arrow 1) originates from the subclavian artery opposite to the origin of the internal mammary artery. The thyrocervical trunk is well shown. Its main three branches (inferior thyroid artery, arrow 2; superficial cervical artery, arrow 3; transverse scapular artery, arrow 4) contribute, with the ascending cervical artery, to the vascularization of the tumor. The extraspinal, soft-tissue mass is more apparent. Note the deviation of the trachea to the right. Upper Right: Later view showing the anastomotic channels between these arteries, within the tumor mass (arrows). Lower Left: Late arteriolar-early capillary phase showing "corkscrew" appearance of tumor vessels (thin arrow). A "tumor blush" is also evident. Early venous filling is apparent (wide arrow). No filling of radicular or medullary vessels. Lower Right: Venous phase showing fairly sizable venous lake (arrow). The tumor mass is now clearly outlined by a substantial "blush." Peculiar beady-looking vessels are seen throughout.

J. Neurosurg. / Volume 36 / January, 1972
preoperative information obtained by arteriography, the surgeon was free from concern about the possibility of interfering with the blood supply of the cord or brain stem. The patient tolerated the procedure well and, in fact, much of the arm and shoulder pain was relieved within 24 hours.

The second stage procedure was carried out about 3 weeks later, and complete tumor removal was accomplished. Again, in spite of the grossly distorted anatomy, all the neurovascular structures within the extraspinal compartments of the neck were easily identified. Nothing resembling the vertebral artery could be seen.

Discussion

This unusual case emphasizes several points.

If more than one intervertebral foramen is involved by the expansile growth of a tumor, then the neoplasm is probably growing in the extradural space. Our case is unusual in that four foramina had been eroded as the neoplasm grew to huge proportions, both intraspinally and in the lateral cervical compartments.

The patient had exhibited symptoms and signs of cord compression, radicular involvement, and brachial plexus dysfunction. Nevertheless, in spite of the size attained within the intraspinal compartment, cord symptoms and findings had been rather mild, owing to the very slow growth of the tumor.

Arteriography has not yet come to play a major role in the diagnosis and preoperative evaluation of intraspinal tumors. However, in cases where a cervical hourglass neurofibroma is suspected, arteriography should be a routine preoperative study and should include visualization of all major branches arising from the aortic arch. One should carefully investigate the possibility of an anomalous origin of the vertebral arteries, and the presence of carotid-basilar anastomoses. Particular attention should be paid to branches that feed the tumor mass, for it is conceivable that they may also contribute to the vascularization of the cervical spinal cord or the structures of the posterior fossa; subtraction technique may be valuable in ascertaining this point.

In this case the vertebral artery had probably undergone thrombosis without causing any recognizable difficulty in the vascularization of the cord (via the radicular arteries) or the brain stem.

The angiographic pattern (Fig. 2) of this spinal neurofibroma was characterized by fairly large anastomotic channels among the different arterial feeders, a corkscrew appearance of tumor vessels seen in the late arteriolar and early capillary phases, tumor blush, first noted in the late arteriolar phase, but persisting through the capillary and late venous phases, early venous filling occurring in the late arteriolar and early capillary phase, irregular venous channels in the capillary—early venous phase, and large venous lakes in the venous phase.

Excellent postoperative bone regeneration and stability of the cervical spine can be expected in spite of extensive bone destruction caused by the tumor and additional bone removal during the surgical procedure.

The angiographic demonstration of the vascular supply to this tumor seems to favor the extraspinal origin of the dumbbell neurofibromas postulated by some authors.

References


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Received for publication January 11, 1971.
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