Arteriovenous malformation in the posterior fossa supplied by the external carotid circulation

Case report

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The authors report a case in which the blood supply of a posterior fossa arteriovenous malformation was derived entirely from the external carotid artery.

**Key Words**  • arteriovenous malformation  • external carotid artery  • posterior fossa

INTRACRANIAL arteriovenous malformations (AVM) derive their complicated blood supply from parts of the internal carotid system when the lesion is above the tentorium, and usually from the vertebrobasilar tree alone when it is infratentorial. Arteriovenous malformations fed by only one vessel from the intracranial carotid are rare. Dandy4 reported two such cases in his early series of eight intracranial malformations; one involved the middle cerebral artery and the other the vertebral. Although he described these cases before the days of angiography, his surgical observations were precise, and the results of surgical management by occlusion of a single vessel were so dramatic that there seems to be little doubt that these AVM's were fed by a single vessel.

We are reporting the even rarer occurrence of an AVM in the posterior fossa, fed from a single branch of the external carotid artery.

**Case Report**

A 46-year-old man was first admitted to the University of Iowa Hospitals on May 26, 1972, after having been awakened from sleep by an intense generalized headache and tonic extensor rigidity of the legs prior to loss of consciousness. Two months earlier he had noted "transient tingling" in the right leg and left thigh. Two days before admission he had been pitched "head first" from a bulldozer. The only neurological findings were lethargy and disorientation, neck rigidity, and bilateral Babinski's sign. Bilateral common carotid and right vertebral angiography demonstrated dilated ventricles and a posterior fossa AVM that filled only during left common carotid injection. Although he became brighter, he remained intermittently confused and required help in walking. Left vertebral angiography was planned to learn more about the AVM, but the patient insisted upon being discharged on June 19 before this could be done. Examination 1 month later disclosed tremor of the left leg and hyperactive lower tendon reflexes with Babinski's sign, bilateral hypesthesia below T-9,
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hypalgesia below T-10 on the right and L-4 on the left, and disturbed vibration sense below T-2 bilaterally. By October, 1972, the leg weakness had become worse and was associated with "shock-like" sensations and lancinating pain in the legs.

In January, 1973, he noted progressively severe neck stiffness and pain in both arms and scapular areas, all of which were relieved by wearing a supportive cervical collar. Bladder emptying became impaired, and he developed constipation. Cervical spine films revealed severe spondylosis. Since the patient had been without symptoms until the subarachnoid hemorrhage had occurred on May 26, 1972, the malformation appeared to be the major contributor to the myelopathy. Left vertebral angiography was normal. A selective left external carotid study demonstrated that the sole blood supply to the AVM was the posterior meningeal branch of the ascending pharyngeal artery (Fig. 1). A cervical myelogram confused the diagnosis by giving the impression of an "intramedullary mass" (Fig. 2).

Operation. On March 2, 1973, the left external carotid artery was ligated. A suboccipital craniectomy with C1-6 laminectomy then exposed the areas involved. The arachnoid of the cisterna magna was thick and opaque. There was a tangle of stout blood vessels densely adherent to each other and to the arachnoid in the region of the left cerebellar tonsil, the biventer, and between the cerebellar hemispheres over the inferior vermis. Two 10 × 5 mm bulbous, vascular, knuckle-like structures lay buried within the vermis. The medulla was yellow and bulbous and was constricted at the medullocervical junction (Fig. 3 left). The spinal cord was broad and translucently blue, displaced posteriorly, and rotated to the right side. The posterior roots from C-3 downward, especially on the right side, were splayed and thin. The cord appeared in places to be cystic, but no fluid was obtained from needling these sites.

Serial temporary occlusion of five large arterial trunks had no effect upon the turgidity of the malformation. As the AVM was traced beneath the left cerebellar hemisphere, about 2 cm behind the jugular foramen, a thick gray vessel 4 mm in diameter was found running into the vascular

Fig. 1. Anteroposterior (left) and lateral (right) views of selective left external carotid angiogram (February, 1973) demonstrating the malformation. The arterial supply is via the ascending pharyngeal (arrows) while the occipital artery is seen taking its normal lateral course.
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Fig. 2. Posteroanterior (left) and lateral (right) views of the cervical and cisternal Pantopaque myelograms show an "intramedullary mass effect" above C-7, and anteriorly the coils of a large twisting vessel that extends downward to T-11 (left). The malformation indents the Pantopaque column in the cisterna magna (right).

tangle. At its most lateral point, the vessel lay in relation to the inferior and lateral aspect of the foramen magnum, where it penetrated the dura to become extradural. When this vessel was occluded, the surging pulse of the AVM disappeared, and the turgidly erect vascular mass drooped in a collapsed state. The mass was resected (Fig. 3 right). The dentate ligaments were cut from C-1 through C-6, the cord then assumed a normal configuration. Exploration of the anterior spinal canal disclosed bony ridges at each intravertebral disc space; these had flattened the cord when the latter had been tethered by the dentate ligaments.

Complete cerebral angiography performed 9 months later showed that the left external carotid system filled from muscular anastomotic branches of the left vertebral artery (Fig. 4). The AVM was never demonstrated. The patient showed progressive improvement in his sensory and motor deficits.

Discussion

Arterial contributions from the external carotid system to an intracranial AVM have been only infrequently noted. In previously reported large series of AVMs, only a few references to lesions with an associated external carotid contribution were made. It is evident that some cases were common to a
FIG. 3. Left: Magnified operative view. A part of the malformation is seen in the left cervicomedullary junction, while an equally large portion is covered by arachnoid between the cerebellar tonsils. Note the tortuous dorsal venous channel. The spinal accessory nerve can be seen on the left as it approaches the jugular foramen. Right: Overall operative view after the vessels have been clipped and a major part of the AVM removed. The spinal cord is flattened ventrodorsally.

number of these series. This makes it difficult to know exactly how many malformations had external carotid components.

Tönns and Lange-Cossack\textsuperscript{13} mentioned seven patients in their group of 72 whose lesions were supplied by the external carotid but gave no specific information as to whether this was the sole supply. Similarly, Tönns, \textit{et al.},\textsuperscript{14} in describing the EEG activity related to their 130 supratentorial AVM’s, stated: “In the cases of arteriovenous aneurysms with influx from merely the external carotid artery, the electroencephalographic activity was normal.” They gave no further details.

Bergstrand, \textit{et al.},\textsuperscript{1} referred to two instances involving the dura and leptomeninges in which the AVM was presumably supplied from the external carotid system. They stated that in one there was “some connection to the left posterior cerebral artery.” The severe neurological disturbances that followed excision of these lesions led the authors to conclude that the cerebral circulation was in large part dependent on the external carotid system; in only one case was the blood supply of the AVM derived purely from the external carotid artery. The first of these cases is also included in the classic monograph of Olivecrona and Ladenheim,\textsuperscript{6} in the section entitled “External Carotid Lesions of the Brain,” they allude to five examples in which a lesion situated on or within the brain sub-
stance was fed "predominantly or exclusively" by the external carotid artery. The AVM in their Cases 4 and 5 had contributions from the posterior cerebral artery; the temporal lobe AVM described as a "classic example of a middle meningeal lesion" was also supplied by the posterior cerebral artery.

In the Cooperative Study of Subarachnoid Hemorrhage and Intracranial Aneurysms, among 421 supratentorial and 32 infratentorial AVM's, Perret found no record of such a lesion drawing its blood supply only from the external carotid artery. Although Ramamurthi and Balasubramanian reported two cases of AVM with a purely external carotid contribution, the lesions apparently involved neither the meninges nor brain substance, and seem to have been arteriovenous communications or fistulas.

Thus, supratentorial AVM's are only occasionally fed from the external carotid system and even more rarely from this circulation alone. Rarer still are AVM's in the posterior fossa receiving blood supply from the external carotid. Verbiest in reviewing the literature between 1914 and 1961, found 108 cases of posterior fossa AVM's, adding six of his own. Precise details of their blood supply were given in only occasional cases.

Ciminello and Sachs reported an intradural posterior fossa AVM supplied exclusively by the contralateral external carotid circulation, and found no previous similar report. The malformation in their case drained into veins over the right cerebellum and thence into the vein of Galen; it was fed only by "an enlarged posterior meningeal artery which arose from the occipital artery and entered the posterior fossa by way of the jugular foramen or condylar canal." Clipping of this single vessel, the posterior meningeal artery, obliterated the AVM.

The case that we report is quite similar to that of Ciminello and Sachs. We believe that the posterior meningeal branch of the ascending pharyngeal fed the malformation after its passage through the condyloid foramen.

Fig. 4. Postoperative arterial phases of the left vertebral angiogram. The left external carotid is filled via muscular anastomotic branches from the vertebral artery. No AVM is seen. Arrows indicate the position of the Heifetz clip on the feeder from the ascending pharyngeal artery.
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References


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