Recurrence of a cerebral arteriovenous malformation after surgical excision

Case report

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Total surgical removal of an arteriovenous malformation (AVM) of the brain should eliminate any future risk of hemorrhage. Although postoperative angiography is commonly used to confirm complete excision of an AVM, regrowth of the lesion with subsequent hemorrhage after angiographically documented obliteration has been observed in children.8,9,15,16 However, as the following case report demonstrates, AVMs in adults may also be at risk for regrowth despite an angiogram confirming complete removal.

Key Words • arteriovenous malformation • recurrence • prognosis • cerebral hemorrhage

Case Report

While at work on March 10, 1984, this previously healthy 19-year-old man developed a left frontotemporal headache with subsequent emesis and loss of consciousness. He was noted to have a right hemiparesis and was taken to a local hospital where his symptoms and signs began to resolve.

Examination. A computerized tomography (CT) scan demonstrated a left frontal intracerebral hematoma. The patient was transferred to a second hospital where he was treated with intravenous dexamethasone. His neurological status gradually improved and he was discharged on March 26. At the time of his admission to our institution on April 29, he was neurologically normal except for a slight right central facial paresis. Cerebral angiography

demonstrated an AVM low in the left frontotemporal area, which was supplied by the left internal carotid, middle cerebral, and anterior cerebral arteries; two large draining veins were also identified (Fig. 1).

First Operation. On May 1, 1984, a left frontotemporal craniotomy without preoperative embolization was performed for removal of the AVM. The surgeon believed at the time that a complete excision was achieved. Histopathological analysis of the surgical specimen verified that the lesion was an AVM.

Postoperative Course. Postoperatively, the patient retained some subtle right facial weakness; he was otherwise neurologically normal and had a routine recovery without complication. He was not given prophylactic anticonvulsant medication. Seven days after his operation, he underwent left internal and external carotid angiography that was interpreted at that time (and again on recent review) as showing no evidence of residual AVM (Fig. 2). He returned to work and resumed his normal activities without restrictions. He remained free of symptoms for more than 2 years.

In September 1986, the patient had three generalized tonic–clonic seizures, at least one of which was preceded by aphasia and rhythmic jerking of the right upper extremity. He was started on a course of phenytoin, but despite a therapeutic serum level, he had one further seizure. An electroencephalogram obtained on November 3, 1986,
demonstrated a moderate focal slow-wave abnormality with occasional spikes and sharp waves in the left frontotemporal area. His neurological examination revealed no abnormalities and he was started on a course of carbamazepine in addition to the phenytoin. However, he experienced another generalized major motor seizure that was preceded by aphasia and right-sided jerking and was followed by aphasia and a right hemiparesis that resolved over approximately 10 minutes. Subsequently he was maintained on phenytoin, experiencing only occasional auras but no other seizure activity.

In July 1993, when he was 28 years old, the patient returned to our institution after experiencing five generalized tonic–clonic seizures during a 24-hour period. Although he described a mild generalized headache on the previous day, he denied any other neurological symptoms aside from the seizures. His neurological examination was normal and his serum phenytoin level was 12.5 μg/ml. A contrast-enhanced CT scan indicated probable recurrence of the AVM (Fig. 3). Left internal and external carotid angiograms demonstrated a rapidly filling 3.0 × 2.0 × 2.5-cm AVM in the region of the previously resected AVM, which was fed by branches of the left internal carotid, middle cerebral, and anterior cerebral arteries (Fig. 4). This lesion was similar in configuration to the original lesion but was larger.

Second Operation. On July 16, 1993, a secondary left frontotemporal craniotomy was performed and the recurrent AVM was resected as a single specimen. Again, the surgeon believed that a complete excision of the lesion was achieved, and again, histopathological examination of the specimen verified the diagnosis of an AVM.

Postoperative Course. A left common carotid angiogram obtained 2 days postoperatively again failed to demonstrate a residual AVM (Fig. 5). The patient’s postoperative course was uneventful. He was started on a course of phenytoin and discharged on the 6th postoperative day with no neurological deficit. Postoperative magnetic resonance (MR) studies performed 4 months and 17 months after the second resection showed only the expected postoperative changes with no evidence of a residual or recurrent AVM. When last seen on December 12, 1994, the patient had had no seizure-like events for 8 months and he remained neurologically normal.

Discussion

Angiography has been relied upon to confirm complete excision of an AVM. Several large series in the literature reporting long-term surgical results in the treatment of cerebral AVMs have shown that those patients with negative postoperative angiograms (that is, ones that demonstrate no residual nidus or early draining veins) do not experience rehemorrhage.1–3,5–7,11,14 This is in accord with the general consensus that complete surgical removal of an AVM should eliminate the risk for an associated hemorrhage. If a late hemorrhage does occur postoperatively, it is often attributed to an incomplete resection with a residual AVM nidus5,2,11 or, perhaps, an intracerebral hemorrhage into a softened area of the brain.4 A few patients with recurrent hemorrhage have been reported in whom the surgeon thought that he had achieved a complete
resection, but this was not documented angiographically.\textsuperscript{1,13}

The phenomenon of AVM regrowth and rehemorrhage after negative postoperative angiography has been reported in children.\textsuperscript{8,9,15,16} Yaşargil\textsuperscript{13,16} published the largest series of such AVM regrowths. He described six pediatric patients ranging in age from 4 to 17 years; however, only one patient had a documented negative angiogram obtained 10 days postoperatively. In this patient, the AVM was located in the right frontal opercular area and recurred in the same location; this recurrence was discovered 7 years postoperatively, following a subarachnoid hemorrhage. In addition, Kader,\textit{et al.}\textsuperscript{8} described a series of five pediatric patients with recurrent AVMs who presented initially with a hemorrhage at 6 to 11 years of age. All five patients had negative postoperative angiograms; however, recurrence of the AVM was noted 1 to 9 years later. Finally, Kondziolka,\textit{et al.}\textsuperscript{9} found among 132 children with AVMs of the brain, two who developed rehemorrhage due to regrowth of a temporal lobe AVM despite a negative postoperative angiographic study.

Regrowth of childhood AVMs may be related to the inherent nature of the development of an original abnormally vasculature. Padget\textsuperscript{15} has hypothesized that a cerebral AVM arises from an aberrant connection (a fistula) between a primitive artery and a relatively large vein overlying the developing cerebral cortex. It has been theorized further that the reduced resistance to blood flow in such arteriovenous fistulas may cause the abnormal arteries to dilate relative to normal arteries.\textsuperscript{10} These hypotheses about the early evolution of AVMs are consistent with the diffuse patterns of arteriovenous shunting observed in children as opposed to the more distinct fistulous pattern seen in adults.\textsuperscript{10} If more “mature” AVMs develop along a continuum as the brain develops, then children are more likely to have immature vasculature as part of their AVM. Immature vessels left in the surgical bed may not be angiographically visible but still may retain the ability to regrow and form a new malformation in the same location. Our patient is the oldest patient reported in the literature with an AVM that recurred after a complete surgical resection (without preoperative embolization) had been documented angiographically. The theory described above that attributes regrowth of AVMs to the presence of immature vasculature is less tenable in this reported case, because the patient’s cerebral vasculature should have reached a more “mature” state of growth by his age.

There are also other plausible explanations for the regrowth of our patient’s AVM. For example, a small nidus of the AVM could have been left at the end of the first operation; this residual portion of the AVM might not have been visualized on the postoperative angiogram because of local compression of the residual vessels by cerebral swelling or because of spasm of one or more persistent feeding arteries. Recanalization of thrombosed vessels, which can occur after radiosurgery or embolization, should not have played a role in the regrowth of our patient’s AVM because, presumably, the abnormal vessels were severed at the time of surgery. Another explanation, although unsubstantiated, could be the existence of an angiogenic growth factor that promotes the initial development and potential regrowth of these anomalous vessels. Further studies would need to be performed to elucidate this concept as a causative factor in the development and recurrence of AVMs.

Regrowth of AVMs and recurrent hemorrhages from residual AVMs raise an important question regarding the optimum timing for obtaining a postoperative angiogram. Most surgeons agree that angiography should be performed to exclude any residual AVM that may require further treatment;\textsuperscript{1,14} however, angiography that is performed too soon after surgery may not provide accurate data. The first negative postoperative angiogram in this patient was obtained on the 7th postoperative day, which is in accord with today’s current standards. Usually by the 7th to 10th postoperative day, edema and vessel spasm are less of a concern and should not interfere with the angiographic visualization of a residual AVM. If the angiogram raises the question of a residual nidus or demonstrates suspicious vascular tufts, then obtaining a delayed angiogram some months later should be seriously considered. However, considering the rarity of AVM regrowth in adults, this need not be performed routinely in all adults with negative postoperative angiograms. This approach may not be appropriate for children because their risk for AVM recurrence is higher.\textsuperscript{8,9,15,16} In this group of patients, repeat angiography 6 to 12 months after an initial negative postoperative study has been suggested to exclude a recurrence.\textsuperscript{8}

Regardless of the cause of recurrent AVMs, the outcome of this patient, which occurred despite the initial “complete” resection, would imply that adult patients may still be at risk for regrowth of an AVM with the potential for rehemorrhage despite postoperative angiographic.

\textit{J. Neurosurg. / Volume 84 / May, 1996}
demonstration of AVM obliteration. Although the authors do not propose that additional follow-up angiography be performed routinely after the initial negative postoperative angiogram in adults, we do believe that such patients should be followed clinically. If symptoms and signs consistent with AVM regrowth were to appear, CT or MR imaging would be indicated at that time, as well as angiography.

References


Manuscript received August 16, 1995. Accepted in final form November 17, 1995. Address reprint requests to: Eric M. Gabriel, M.D., Division of Neurosurgery, Box 3807, Duke University Medical Center, Durham, North Carolina 27710.