A recurrent cerebral arteriovenous malformation in an adult

Case report

PATRICK J. CODD, B.S., ALIM P. MITHA, M.D., AND CHRISTOPHER S. OGILVY, M.D.

Department of Neurosurgery, Massachusetts General Hospital, Boston, Massachusetts

Arteriovenous malformations are generally considered to be cured following angiographically proven complete resection. However, rare instances of AVM recurrence despite negative findings on postoperative angiography have been reported in both children and adults. In this paper, the authors present the case of a 33-year-old woman with 2 AVM recurrences. This patient represents the oldest case of recurrent AVM, and the first adult double recurrence reported in the literature. The case is presented, the radiological and surgical features are considered, and the literature on recurrent AVMs is reviewed. (DOI: 10.3171/JNS/2008/109/9/0486)

KEY WORDS • arteriovenous malformation • postoperative angiography • recurrent arteriovenous malformation

Case Report

History. This 33-year-old, otherwise healthy woman originally presented to another hospital’s emergency department in March 1997 after being awoken from sleep by the sudden onset of severe left occipital headache associated with a right-sided homonymous hemianopia. Examination at the time of presentation revealed 0/4 deep tendon reflexes bilaterally in the patellar and biceps region, and no other motor or sensory abnormalities. Admission CT scans were notable for a left occipital lobe hematoma with a rim of surrounding edema (Fig. 1 left). Subsequent catheter angiography revealed a 7-mm Spetzler–Martin Grade III AVM (size < 3 cm = 1 point, eloquent occipital lobe cortex = 1 point, deep draining vein = 1 point) supplied by the calcarine branch of the left PCA, and notable for a single early filling, deep draining vein (Fig. 1 right). The patient was then referred to our institution.

First Operation and Postoperative Course. Seven weeks later, the patient underwent left occipital craniotomy for evacuation of the hematoma, and microsurgical resection of the AVM with intraoperative imaging guidance. The AVM was resected without complication and immediate postoperative angiography demonstrated no residual AVM (Fig. 2). Her immediate postoperative course was uneventful and, over the 3 months following resection, the patient had a nearly complete recovery of her visual field. Four months after resection, during tapering of her prophylactic Dilantin therapy, she developed episodes of visual hallucinations consisting of floating colors. Despite normal outpatient and ambulatory electroencephalogram findings, these episodes were diagnosed as simple partial occipital lobe seizures and were well controlled with oral antiepileptic drugs.
Recurrence. The patient was otherwise in good health until December 2004, 7 years after her initial resection. At that time, she presented to our emergency department with sudden onset of severe headache, flashing colors, dizziness, and a worsened right-sided homonymous hemianopia. The admission CT scan demonstrated a 3 × 3–cm hemorrhage in the left occipital region in which the prior AVM resection had been performed (Fig. 3 left). Catheter angiography showed a small left occipital Spetzler–Martin Grade II AVM (size < 3 cm = 1 point, eloquent occipital lobe cortex = 1 point, superficial draining vein = 0 points) at the anterior margin of the resection bed, which was again fed by the left calcarine artery and early cortical venous drainage into the SSS (Fig. 3 right).

Second Operation. Three weeks later, the patient underwent a second microsurgical resection of the occipital AVM recurrence with stereotactic guidance. Noted at the time of surgery were several large feeding vessels and a recurrent AVM nidus. Through careful microdissection the lesion was elevated away from the prior resection cavity, and then removed in its entirety. Postoperative angiography again showed no evidence of residual AVM (Fig. 4). The patient had an uneventful postoperative course and was discharged on the 4th postoperative day in excellent condition, and without new neurological deficit.

Second Postoperative Course and Additional Treatment. Because of the AVM recurrence, follow-up angiography...
raphy was recommended at 1 year after the second resection. This was notable for an asymptomatic collection of small abnormally dilated and tortuous vessels, without shunting or early filling of the draining veins, in the anterior margin of the previous AVM resection cavity (Fig. 5). Ten months later, a second follow-up angiogram confirmed the presence of a recurrent Spetzler–Martin Grade III AVM (size < 3 cm = 1 point, eloquent occipital lobe cortex = 1 point, deep draining vein = 1 point) again fed by the calcarine branch of the left PCA with shunting to an occipital vein draining to the vein of Galen (Fig. 6A and B). This represented a second recurrence despite negative findings on postoperative angiography following the second resection. Given the location and size of the recurrent nidus, radiation was believed to be the best treatment option. Subsequently, the patient completed therapy with 18 cGy of stereotactic proton radiosurgery in February 2007. She tolerated the procedure well and continues to be followed by our service. Follow-up imaging after radiosurgery is not yet available.

Discussion

Angiographically proven resection of cerebral AVMs is generally thought to eliminate the risk of future hemorrhage, therefore not requiring routine follow-up angiography. However, recurrence of these lesions, defined as local reappearance following treatment and angiographically confirmed absence of residual nidus or early filling of the draining veins, has been reported.\textsuperscript{10,17} To date, 24

Fig. 3. \textit{Left}: Axial CT scan notable for a 3 × 3–cm hyperdense lesion in the left occipital lobe consistent with acute hemorrhage. \textit{Right}: Left VA catheter angiography study demonstrating a recurrent left occipital AVM at the anterior margin of the previous resection bed (arrow), fed by the left calcarine artery, with early venous drainage to the SSS (arrowheads).

Fig. 4. Anteroposterior (left) and lateral (right) left VA catheter angiography studies obtained immediately postoperatively following the second resection. No residual AVM is noted.
cases of recurrent AVM following gross-total microsurgical resection have been reported in the English language literature. Of these, 20 cases are in the pediatric population, and the remaining ones are in adults.

Yaşargil first described a pediatric case of recurrent AVM in a 17-year-old adolescent presenting with intracerebral hemorrhage. This child had negative findings on postoperative angiography, with AVM recurrence in the right frontal opercular area 7 years after the initial resection. Kader et al. reported on 5 patients ranging from 6 to 11 years old who had AVM recurrences. All patients in this series had negative results on postoperative angiograms, with recurrences 1 to 9 years after initial resection. In a study of 132 patients with cerebral AVMs, Kondziolka et al. described 2 children with recurrent temporal lobe AVMs despite negative results on postoperative angiograms. Similarly, Klimo et al. report 5 cases of recurrent AVMs in children with immediate postoperative angiograms showing complete resection. Three of these recurrences were identified during routine follow-up imaging, 1 patient presented with new seizures, and another with new headache. Together, these studies demonstrated a recurrence latency following initial treatment ranging from 6 months to 9 years. In all but 1 case with a second recurrence, a long-term cure was achieved following a second resection.

Although AVM recurrence and rehemorrhage have
been reported in pediatric patients, it remains extremely rare in the adult population with AVMs. Previously, only 4 cases of adult recurrent AVM following gross-total microsurgical resection and negative results on postoperative angiograms had been reported in the English language literature. All 4 previously reported recurrences were in men between the ages of 19 to 28 years, with recurrence latency between 4 and 10 years following initial resection. Even rarer is a double recurrence of an angiographically proven completely resected AVM. In fact, only a single case of double recurrence has previously been described, which occurred in a 9-year-old boy with 6- and 8-month latency periods between resections. Our case, therefore, represents the oldest patient with recurrent AVM to date, and the first report of a double recurrence in an adult.

In all previously reported cases in adults, as well as in the case described in this paper, postoperative angiography within 10 days of operation was clearly negative for residual AVM. Angiography is the current gold standard for confirming complete resection of an AVM. Current practice is to obtain images as soon as possible following resection to assess the risk of possible postoperative hemorrhage from a residual nidus. If a residual nidus is found, immediate reexploration should be performed due to the risk of hemorrhage in the early postoperative period. It is standard practice at our institution to perform immediate postoperative angiography while the patient is still anesthetized, so that reoperation can be expedited should it be necessary. Investigators in numerous large case series examining long-term results in the treatment of cerebral AVMs have reported no risk of hemorrhage following resections in which postoperative angiography confirmed the absence of a residual nidus or draining vein. In our patient, postoperative angiography confirmed the absence of residual AVM after each resection. Despite this, long-term recurrence of the AVM resulted in rehemorrhage after the first resection, and was diagnosed on follow-up angiography after the second resection.

Some theories of AVM recurrence suggest that the "recurrent" AVM actually represents a residual nidus that went unseen on postoperative angiography. Factors such as vessel spasm, brain edema with compressive mass effect, or temporary thrombosis of residual AVM vasculature in the immediate postoperative period could prevent this residual nidus from being detected on angiography studies obtained immediately postoperatively. Subsequent relief of spasm, resolution of edema, or recanalization of feeding vessels may then expand and reveal the residual AVM, resulting in further symptoms or hemorrhage. In several of the recurrent AVMs reported in the literature, however, follow-up angiography yielded negative findings 3–6 months after the initial postoperative imaging, falling outside the temporal window suggested by this theory.

Others have proposed theories for true AVM recurrence. Pellettieri et al., for instance, described the "hidden compartment" model of AVM recurrence. This theory suggests that angiographically unfiled areas of an AVM could exist, located either contiguous with or adjacent to a nidus, which can subsequently blossom under the new local hemodynamic state following resection. The authors suggest that this may explain AVM regrowth, and would account for difference in feeding vessels between initial and recurrent lesions. Sano et al. proposed a similar mechanism, suggesting that AVM growth may be due to a "reserve nidus," or an abnormal vascular group adjacent to the primary nidus that subsequently grows to become a recurrent lesion capable of rehemorrhage.

In the case presented here, we cannot exclude with absolute certainty the possibility of residual nidus, because the absence of such was not documented sufficiently late in the postoperative period. This is also an issue with the previously reported cases of recurrent AVM in adults. Late postoperative angiograms, taken after the resolution of pathophysiological processes that could explain an undetected residual nidus, would help to resolve this issue but are not standard practice after complete resection. We believe, however, that our case represents a true AVM recurrence because immediate postoperative angiograms after both resections demonstrated no obvious spasm or thrombosis that could have potentially concealed a residual nidus. In addition, we believe that differing patterns of venous drainage between recurrences (deep drainage at presentation, superficial drainage to the SSS with the first recurrence, and deep drainage with the second recurrence) also suggest true recurrences versus the evolution of a residual nidus.

Kader et al., hypothesized that AVM growth is not limited to passive abnormal capillary formation during embryogenesis, but is an active angiogenic process that continues through childhood, eventually reaching a "mature" state where growth no longer occurs. These authors also suggest that a resection performed during this active growth stage in patients in whom proangiogenic tissue remains locally following surgery could increase the risk for recurrence. They also propose that proangiogenic factors such as VEGF might play a role in AVM recurrence. Sonstein et al. examined VEGF expression in 4 cases of recurrent pediatric AVMs, and found that all 4 specimens had a high degree of expression, compared with 1 in 7 patients with nonrecurrent pediatric AVM and 2 of 8 nonrecurrent adult specimens. They also noted that this immunoreactivity specifically labeled the abnormal vessels themselves. Thus, while the underlying cause of AVM recurrence in both the pediatric and adult population is presently unclear, there is evidence supporting a role for vessel-associated VEGF and active lesion regrowth.

Conclusions

Recurrence of AVMs following microsurgical excision and negative findings on postoperative angiography is rare in the adult population. However, it should be considered in the setting of recurrent neurological symptoms or new intracranial hemorrhage even after angiographically proven complete resection. Routine follow-up imaging to monitor for recurrence in adults is generally not necessary; however, in rare patients with recurrent AVM such as in the case presented here, long-term clinical and angiographic follow-up may be required.
Adult recurrent arteriovenous malformation

Disclaimer

The authors do not report any conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

References


Address correspondence to: Christopher S. Ogilvy, M.D., Neurovascular Surgery, Department of Neurosurgery, Massachusetts General Hospital, 55 Fruit Street, VLB 710, Boston, Massachusetts 02114-2698. email: cogilvy@partners.org.