De novo presentation of an arteriovenous malformation

Case report and review of the literature

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The authors report the case of a patient with a de novo arteriovenous malformation (AVM), indicating that the origin of these lesions may not always be congenital.

A 3-year-old girl who was struck by a car suffered a mild head injury and experienced posttraumatic epilepsy. The initial magnetic resonance (MR) image obtained in this child revealed only a small contusion in the left frontal lobe. Intractable epilepsy subsequently developed. A second MR image obtained almost 4 years after the injury demonstrated an AVM in the right posterior temporal lobe that was verified using angiography. The lesion was classified as a Spetzler–Martin Grade III AVM. The patient underwent embolization of the feeding vessels followed by gamma knife surgery. Fourteen months after treatment she was asymptomatic. Follow-up MR images demonstrate no evidence of an AVM and no changes in the white matter.

This case presents a de novo AVM that developed within approximately 4 years. The findings indicate that AVMs may not always be congenital and reinforce the concept that the natural history of AVMs is dynamic. Lesions may appear de novo, grow, and thrombose spontaneously.

KEY WORDS • arteriovenous malformation • gamma knife surgery • natural history

ARTERIOVENOUS malformations are shunts between an artery and the venous system that lie within a nidus without an intervening capillary bed. These lesions are thought to be congenital, but recent reports have challenged this assumption.17 We present a patient with a de novo AVM that was found incidentally during an evaluation for intractable seizures. The findings of a previous MR imaging study had been normal.

Case Report

History. This 3-year-old girl was struck by a car while crossing the street. She momentarily lost consciousness and was transferred to a local hospital where she was observed for 48 hours. On her initial evaluation, her Glasgow Coma Scale score was 15. Computerized tomography scans of the brain revealed a punctate contusion on the left frontal lobe. The next day the contusion was no longer evident on follow-up computerized tomography scans and she was discharged from the hospital.

The patient was asymptomatic for 2 weeks but then absence seizures developed and became generalized. She was examined by a pediatric neurologist and a regimen of antiepileptic medications was initiated. Electroencephalography demonstrated discharges from the left frontal region and slow wave activity. Magnetic resonance images of the brain revealed a hemorrhagic lesion in the left frontal region, which was the same location as the previous contusion (Fig. 1).

Examination. Control of the patient’s seizures became progressively more complicated. Different medications failed to provide adequate seizure control. As part of a comprehensive evaluation for intractable epilepsy, a new MR image was obtained 44 months after the first study (Fig. 2). A right posterior temporal lobe nidus, which corresponded to an AVM on subsequent MR angiography (Fig. 3 left) and conventional angiography (Fig. 3 center), was found. The lesion was classified as a Spetzler–Martin Grade III AVM.26

Operation. Treatment options, including surgery, embolization, and stereotactic radiosurgery, were discussed with the family. When the patient was 8 years of age, she underwent GKS (18 Gy delivered to the 50% isodose line) after undergoing embolization of the feeding vessels without complications (Fig. 3 right).

Postoperative Course. Fourteen months after emboliza-
tion and GKS, MR imaging (Fig. 4 left) and MR angiogra-
phy (Fig. 4 right) of the brain demonstrated no evidence of an AVM and no white matter changes on axial T2-weighted MR images. Approximately 6 months after treatment, her seizure activity began to decrease progressively. Ultimately, she became seizure free while taking Keppra (UBC Pharma, Inc., Smyrna, GA). She now attends a regular school.

Abbreviations used in this paper: AVM = arteriovenous malformation; GKS = gamma knife surgery; MR = magnetic resonance.
De novo arteriovenous malformation

**Fig. 1.** Axial T$_2$-weighted MR images. *Left:* Initial image revealing no evidence of vascular abnormalities. *Right:* Proton-density image demonstrating a hemorrhagic contusion in the left frontal pole.

**Fig. 2.** *Upper:* Follow-up axial T$_2$-weighted MR image revealing flow voids suggestive of an AVM in the posterior temporoparietal region 44 months after the initial MR image had been obtained. *Lower:* Sagittal T$_1$-weighted MR image demonstrating the nidus in the posterior aspect of the temporal lobe.

**Discussion**

The epidemiology of AVMs is dynamic. Some lesions have disappeared after spontaneous thrombosis or after a partial resection that might have changed the hemodynamic structure of the lesion, causing it to thrombose. Their dynamic nature is also supported by the findings of hemorrhage after lesions had been thought to have been eliminated by resection. Furthermore, clear evidence from long-term follow-up studies indicates that AVMs may enlarge or recur, especially in children. Some evidence supports the notion that there is a linear correlation between size and growth of small AVMs.

Technical factors may play a significant role in the evaluation of AVMs. Hematomas can compress the vessels, rendering the lesions occult during angiography. The AVM may later become evident after the collection has resolved. A spasm in these small vessels could also render them invisible on angiography. Hemorrhage causing partial thrombosis of the vessels eventually may become more evident when the clot is dissolved. Hyperemia after the resection of an AVM can affect the transit time of the dye and make postoperative angiograms difficult to evaluate.

De novo formation of AVMs is even more rare. To our knowledge, only three previous cases have been reported. The most recent case was criticized by some reviewers because retrospective analysis of some of the patient’s “negative” angiograms showed “faint” and “loose” vessels that 6 years later constituted a definitive AVM. The case reported by Ms. and coworkers and the present case demonstrate one end of the spectrum. It seems likely that an abnormality in the cerebral vessels may have been present before birth.

The growth of AVMs is established, and certain angiogenic mechanisms may recruit and incorporate new vessels within the lesion. Microhemorrhages associated with subsequent gliosis of adjacent brain tissue have been proposed as the cause. Basic fibroblast growth factor has been significantly elevated in the cerebrospinal fluid of patients with AVMs related to hemodynamic stress. The expression of fibroblastic growth factor is key for the development of a full-blown AVM, which might be present but undetected by angiography or MR imaging because these methods may be too insensitive to detect an AVM during the early stages of its development. Abnormalities of cerebral vessels are present in patients with other vascular conditions such as moyamoya disease. The delayed development of a full-blown AVM in a patient who simultaneously had a developmental venous anomaly leads us to infer that there was a congenital abnormality in the cerebral vessels.

That symptomatic AVMs are more prevalent in adults than children and that AVMs are rarely found incidentally during childhood indicate that these lesions grow to some degree. It is commonly believed that AVMs arise at approximately the 28th day of intrauterine life, but no in utero evidence of AVMs has been reported. In contrast, a congenital component is present in vein of Galen fistulas from birth or earlier and is commonly documented by prenatal ultrasonography. Dural arteriovenous fistulas are definitely acquired, especially after venous flow restriction and trauma. “Angiographically occult vascular malformation” is the term used to describe an existing vascular abnormality that cannot be detected by conventional angiography. The delayed presence of an AVM after a nondiagnostic angio-
graphic study associated with a suggestive medical history fits this category.\textsuperscript{18,29}

Radiation has been reported to contribute to the genesis of central nervous system tumors and vascular lesions such as cavernous malformations,\textsuperscript{3,7,14} but it has not been reported to occur with AVMs. Environmental factors must play a role in the growth of AVMs. Trauma is clearly involved with the development of dural arteriovenous fistulas, but whether they are involved with the development of AVMs is unknown. Our patient sustained a mild closed-head injury and an AVM subsequently developed. Whether the trauma played a role or was an unrelated phenomenon is unknown.

As suggested by Spetzler,\textsuperscript{25} AVMs should be considered in a dynamic context. A remodeling feature may explain their growth or spontaneous resolution.\textsuperscript{8,31} Findings in this case and others challenge the assumption that the origin of AVMs is always congenital.\textsuperscript{6}

Based on a longitudinal assessment,\textsuperscript{5} the risk of bleeding in patients with AVMs and no history of hemorrhage is almost 2% per year. In a retrospective analysis of AVMs with a nonhemorrhagic presentation, the risk of bleeding was 4% per year.\textsuperscript{20} The risk of bleeding (2–4%/year) is based on the time at which an AVM is discovered and does not involve its cause, whether congenital or acquired. Recognizing the dynamic features of AVMs, especially their growth, is important, especially in children in whom the lesions have a clear propensity to grow.

After treatment (surgery, radiosurgery, embolization, or multimodality treatments), a close follow-up study of AVMs is crucial, especially when they occur in children. Once the nidus has disappeared on angiography after surgery, it is necessary to schedule a late study (we routinely perform 3-year and 10-year follow-up angiography studies) to ensure that the AVM has been eliminated.\textsuperscript{10,11} Recently, MR imaging has been useful to define the presence of residual AVMs. In a comparison of MR imaging and angiography, Pollock, et al.,\textsuperscript{21} found that the specificity of MR imaging was 100% for identifying a residual nidus on delayed studies.

Conclusions

Evidence now indicates that AVMs are not always congenital lesions. Some congenital vascular abnormalities may be present, but the compact nidus develops as a result of unknown mechanisms.

The natural history of AVMs is dynamic and lesions may be observed at various points across a wide spectrum, with de novo formation on one side of the continuum, spontaneous thrombosis on the other side, and growth and recurrence after resection in the middle. Diagnosis of these lesions depends on one point in time and on the sensitivity of the instruments used for the diagnosis. Although angiography is still the diagnostic gold standard, it is limited in terms of the clinical course of AVMs. Patients who have undergone “complete resection” of an AVM should be observed over time. Follow-up angiography should be performed, especially in children, to confirm that a cure has been obtained and to prevent delayed hemorrhages. Magnetic resonance imaging has become very useful for identifying abnormal vessels that might become symptomatic if allowed to grow with no treatment.

References

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