Development of multiple cerebral arteriovenous malformations documented in an adult by serial angiography

Case report

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A 50-year-old woman with a parietal intracerebral hematoma was initially treated by hematoma evacuation. Initial pre-operative and follow-up angiograms obtained 6 months later demonstrated no pial arteriovenous malformations (AVMs). She suffered a subarachnoid hemorrhage 8 years later. Results of follow-up cerebral angiography revealed the development of previously undetected multiple cerebral AVMs. This appears to be the first reported case of the development of multiple cerebral AVMs in an adult, demonstrated on serial angiography.

KEY WORDS • arteriovenous malformation • cerebrovascular disease • angiography

The patient was readmitted to our hospital on December 24, 2001, again because of the sudden onset of severe headache and vomiting. Neurological deficits were not evident. Computerized tomography demonstrated slight perimesencephalic subarachnoid hemorrhage (Fig. 3) and right ICA angiograms revealed two pial AVMs (Fig. 4). One situated in the frontal lobe received its arterial supply from the right anterior cerebral artery, with venous drainage through the SSS. The other lesion, located in the right parietal lobe, was supplied by the branches of the middle cerebral artery and drained into the SSS. Furthermore, left vertebral artery angiograms displayed a third pial AVM in the left occipital lobe (Fig. 5), which was fed by the branches of the left posterior cerebral artery and drained into the parietooccipital vein. Magnetic resonance imaging demonstrated that the parietal lobe AVM was located a short distance from the previous hematoma cavity (Fig. 6). There was no conclusive proof that a previous hemorrhage caused the angiographically demonstrated AVMs. We plan to treat the AVMs by performing stereotactic radiosurgery.

Discussion

Newton and Cronqvist classified intracranial AVMs into three types on the basis of arterial supply: pure pial, mixed pial and dural, or pure dural types. Pial AVMs are generally thought to be developmental malformations, whereas dural AVMs may be either congenital or acquired. We report a case in which the development of multiple pial AVMs was demonstrated on serial cerebral angiography.

Abbreviations used in this paper: AVM = arteriovenous malformation; ICA = internal carotid artery; SSS = superior sagittal sinus.
The multiple vascular lesions discovered in our patient had the angiographic characteristics of true pial AVMs: first, because of their location within the cerebral parenchyma and second, because of their supply by parenchymal and not dura-based arteries as would be expected in a dural AVM. Third, venous drainage occurred through parenchymal veins. Because the pial AVM is generally considered to be a congenital anomaly, we must discuss why the initial and second angiograms obtained in this adult patient failed to demonstrate pial vascular malformations. There are various possible causes of nonvisualization of pial AVMs in cases of acute hemorrhage. Among the factors mentioned in the literature are compression of the malformation by a hematoma, posthemorrhagic edema, and vascular spasm of feeding arteries. All these may lead to thrombosis of the lesion. Nonetheless, the multiple AVMs presented here do not fit into this category because follow-up angiography was performed approximately 6 months after hemorrhage. A malformation may not be visualized on angiography for technical reasons, but the second angiogram (Fig. 2) was of high quality, and because the appropriate vessels were apparent, this explanation seems unlikely. Therefore, at least the frontal and parietal AVMs in the present case may have formed de novo, during the interval between the second and third angiograms. There is no conclusive proof that the occipital AVM was also a de novo lesion because vertebral angiography was not undertaken until the third admission.

To our knowledge, there have been only three relevant reports in the literature in which the presence of a pure pial AVM was ruled out on initial angiography but later discovered to have developed. Friedman, et al.,3 for example, documented the development of a cerebral AVM in an adult assessed by serial angiography. In their case, the cerebellar AVM developed 4 years after staged stereotactic radiosurgery for a dural arteriovenous fistula. These authors suggested that the most likely mechanism for the de novo development was local venous hypertension caused by the adjacent dural arteriovenous fistula. In addition to the possible role of local angiogenic factors, chronic venous hypertension may have dilated the venous end of parenchymal capillaries, ultimately causing ectatic dilation of the entire capillary network and precipitating direct arteriovenous connections. Schmit, et al.,12 demonstrated an acquired AVM forming in the site of a previous infarct in a patient with moyamoya disease. They speculated that a hyperangiogenic environment in combination with local angiogenic stimulation of cerebral infarction were unique causal factors. More recently, Bulsara, et al.,1 reported on an angiographically demonstrated de novo cerebral AVM in an adult patient who did not have a previous neurovascular abnormality. These reports raise the possibility that at least some pial AVMs arise later in life, perhaps as a consequence of
focal hyperangiogenesis driven by any number of potential mechanisms.

The present case is the first reported in which multiple and pure pial AVMs appear to have been newly formed, although Nussbaum, et al.,8 documented serial instead of simultaneous development of multiple AVMs. Multiple AVMs are rare, except in the Rendu-Osler-Weber and Wyburn–Mason syndromes, with an incidence in major series ranging from only 0.3 to 4%.2,4,10,13,14,16 Iizuka, et al.,4 divided multiple AVMs into two groups: congenital or acquired. According to their classification, the multiple AVMs presented here should be classified as acquired lesions. They speculated that acquired multiple AVMs can be due to angiogenesis (“sprouting” or “non-sprouting”) around a true AVM because of previous hemorrhage or ischemia, or to pial shunts associated with dural AVMs. The patient in the present case showed no symptom or sign of systemic disease, such as the Rendu-Osler-Weber and Wyburn–Mason syndromes. Neither venous hypertension nor cerebral infarction, speculated as factors underlying de novo AVMs, was demonstrated. The precise mechanisms responsible for the development of the multiple AVMs in the present case thus could not be defined, although there is a possibility that they could have been due to sprouting, caused by angiogenesis around a previous hemorrhage (Fig. 6).

Kader, et al.,5 recently described pediatric cases in which pial AVMs recurred after total resection of the original lesion had been verified. The present case of de novo AVM

Fig. 4. Selective right ICA angiograms of the lateral (left, early arterial phase), anteroposterior (center, early arterial phase), and oblique anteroposterior (right, late arterial phase) views, demonstrating two pial AVMs. The one located in the right frontal lobe (arrowheads) is fed by the right anterior cerebral artery and drains into the SSS. The other one situated in the right parietal lobe (arrows) is supplied by the branches from the middle cerebral artery and drains into the SSS.

Fig. 5. Left vertebral artery angiogram, anteroposterior view, displaying an occipital lobe AVM (arrow) supplied by branches from the posterior cerebral artery and draining into the parietooccipital vein.

Fig. 6. Magnetic resonance image demonstrating a parietal lobe AVM (arrow) located a short distance from the previous hematoma cavity (arrowhead).
and Kader and colleagues’ report are significant in that they call into question the theory that a pial AVM is an exclusively congenital phenomenon.

References


Manuscript received May 14, 2002. Accepted in final form September 16, 2002.
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