Microarteriovenous malformations

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This issue of the Journal of Neurosurgery includes an important report by Alén et al. from Professor Lo-bato’s service at the prestigious Universidad Complutense de Madrid. This group has had a long-standing interest in vascular malformations and has made important contributions to the literature, particularly on the less common types of vascular lesions. In this study, the authors presented an excellent retrospective analysis of their experience with microarteriovenous malformations (micro-AVMs), originally defined by Yaşargil as brain AVMs with a nidus smaller than 1 cm in diameter. Although small, these lesions are angiographically visible; however, at times, all that can be seen is a dilated early draining vein. The authors identified 28 patients with such lesions over a 10-year period. They emphasized that patients with these small AVMs almost always present with intracranial hemorrhage and that the hematoma is usually large and frequently results in significant focal neurological deficits. Among these 28 patients, the AVM was detected in all but 6 on the first digital subtraction angiography (DSA) study, usually performed within 24 hours of the hemorrhage. In 6 patients the initial DSA was normal, but, interestingly, MRI and dynamic MR angiography (MRA) with contrast revealed the AVM in 4 of these patients. This led to repeat DSA with superselective catheterization, which then demonstrated the AVM. In the other 2 patients, the AVM was demonstrated on late repeat angiography. Sixteen patients who had a superficial AVM were treated with excision. Deeper AVMs were treated with radiosurgery in 6 cases and endovascular embolization in 2. The remaining 4 patients did not receive any treatment because of their poor clinical condition. All patients treated with surgery and the 2 treated with embolization had occluded AVMs, as demonstrated by postoperative DSA. The AVM was occluded in 4 of the 6 patients treated with radiosurgery. Treatment did not induce a new neurological deficit in any patient, although several patients continue with significant neurological deficits resulting from the initial hemorrhage.

First, we want to address Yaşargil’s classification of small AVMs. In addition to his definition of micro-AVMs (angiographically identifiable AVMs with a nidus smaller than 1 cm, as demonstrated by surgery or pathological examination), he uses the term “occult AVM” to denote lesions not seen angiographically, not seen by the surgeon at exploration, and not demonstrated pathologically but presumed to have been responsible for the hemorrhage. He then uses the term “cryptic AVM” for lesions that are invisible on angiography and at surgery but can be recognized histologically if the pathologist carefully studies the hematoma. This classification leaves out a substantial number of lesions, to which we have previously alluded, that are angiographically invisible but can be seen at surgery either on the surface of the hematoma or deeper along the walls of the hematoma cavity when a thorough exploration is performed after evacuation of the clot, which, as we have suggested, is best done between 2 and 4 weeks after the hemorrhage when the hematoma is semi-liquified. We prefer to use the term “angiographically occult AVM” to include all AVMs that cannot be seen angiographically but can be detected at surgery, on pathological examination, or both. We hesitate to assume the presence of an occult AVM as the source of hemorrhage in cases in which the DSA is negative and no lesion is found at surgery or on pathological examination. Some patients may indeed have a very small AVM as the cause of their hematoma, but others could have an unrecognized tumor that bled, a hematological disorder, or some other etiology for their spontaneous hematoma, and it would seem imprudent to attribute all of these intracerebral hematomas of unknown etiology to a theoretical occult AVM.

We would also like to comment on whether these micro-AVMs, as defined by Yaşargil, are in any way different from the more common larger AVMs. Clearly, they are more difficult to diagnose, and as proved in this series, the gold standard for demonstrating these lesions, which is DSA, initially can be negative. One important finding of the authors is that enhanced MRI and dynamic MRA may actually reveal the malformation in some cases of initially negative DSA studies. This is important because it is generally held that DSA is the most sensitive test to diagnose small AVMs, and, in fact, the senior author of this editorial has long believed that MRA is unnecessary in these cases because the hematoma would mask the small AVM. Apparently this is not the case, and the contrast-enhanced MRA can indeed demonstrate these small lesions even
when the initial DSA may not. The more important question debated in the literature is whether these small AVMs bleed more frequently and/or result in larger hemorrhages than do larger AVMs. In a series of 168 patients with cerebral AVMs monitored after presentation without a prior hemorrhage, Brown and colleagues found, according to a multivariate statistical analysis, that AVM size was not predictive of future hemorrhage. Similarly, Stefani et al. have found no association between AVMs of a small size and hemorrhagic presentations in their analysis utilizing a multivariate model. On the contrary, some studies have suggested that small AVMs have a higher risk of hemorrhage. Spetzler et al. reported a significantly higher incidence of hemorrhagic presentation with small (< 3 cm) AVMs as compared with that for large (> 6 cm) AVMs (82% vs 21%). They also found significantly higher feeding artery pressures in small AVMs and suggested that differences in arterial feeding pressures may be responsible for the observed inverse relationship between the size of AVMs and the frequency and severity of hemorrhage. More recently, however, large natural history studies utilizing modern statistical methodology have shown that a large, not a small, size is an independent factor for hemorrhage.

To reflect the fact that their small size precludes presentation with other symptomatology, such as seizures, or detection by noninvasive brain imaging. It should be noted that in cerebral AVMs monitored after presentation without a prior hemorrhage, more than do larger AVMs. In a series of 168 patients with cerebral AVMs monitored after presentation without a prior hemorrhage, Brown and colleagues found, according to a multivariate statistical analysis, that AVM size was not predictive of future hemorrhage. Similarly, Stefani et al. have found no association between AVMs of a small size and hemorrhagic presentations in their analysis utilizing a multivariate model. On the contrary, some studies have suggested that small AVMs have a higher risk of hemorrhage. Spetzler et al. reported a significantly higher incidence of hemorrhagic presentation with small (< 3 cm) AVMs as compared with that for large (> 6 cm) AVMs (82% vs 21%). They also found significantly higher feeding artery pressures in small AVMs and suggested that differences in arterial feeding pressures may be responsible for the observed inverse relationship between the size of AVMs and the frequency and severity of hemorrhage. More recently, however, large natural history studies utilizing modern statistical methodology have shown that a large, not a small, size is an independent factor for hemorrhage.


due to the fact that the one single draining vein had been followed an arterialized vein toward the nidus. While operating on one of these micro-AVMs deep in the basal ganglia, the senior author of this editorial did exactly that and followed an arterialized vein without further diagnostic studies, at a period of 2–4 weeks after the hemorrhage when the hematoma is relatively liquefied and can be carefully removed and its walls can be inspected microsurgically. Importantly, we recommend these elective explorations only in cases in which the patient is relatively young and healthy and the hematoma is relatively superficial and accessible and not located in an eloquent area of the brain. In other words, we recommend elective evacuation of the clot and careful exploration to look for an etiology only when they can be performed with a very small risk of injuring the patient.

We end our editorial with a warning based on a very unhappy experience. In cases in which a micro-AVM is not visible on the surface, yet the surgeon can find an early draining vein, Alén et al. recommend what others have recommended under these circumstances: to follow the arterialized vein as it goes deeper in the brain toward the nidus. While operating on one of these micro-AVMs deep in the basal ganglia, the senior author of this editorial did exactly that and followed an arterialized vein toward the lesion. Unfortunately, in doing so, the arterialized vein was injured and bled profusely. The bleeding could be stopped only with rather indiscriminate coagulation in the area of hemorrhage, which resulted in occlusion of the draining vein. An immediate intraoperative arteriogram did not reveal any residual AVM, which was obviously related to the fact that the single draining vein had been occluded by coagulation. Several hours after surgery, the patient suffered a massive hemorrhage, which required emergent surgical evacuation. At surgery the hemorrhage was found to originate from a small residual nidus. The patient has remained incapacitated with aphasia and hemiplegia to this day. This warning is not to suggest abandoning the policy of following an arterialized vein...
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to a deeper lesion when there is no other practical way of finding it, but rather to recommend that it should be done with extreme care not to injure the vein. Moreover, with accurate neuronavigation nowadays, it may be safer to go directly to the lesion rather than risking hemorrhage from a single arterIALIZED vein.

We heartily congratulate Alén and colleagues for an excellent review of this important entity and for sharing with us through the years their careful observations about unusual vascular lesions.

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Disclosure

The authors report no conflict of interest.

References


Response

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We are very grateful to Drs. Elhhammady and Heros for their insightful comments about our paper and their affectionate words about our service in Madrid. We agree with them in using the term “angiographically occult AVMs” to include all AVMs that cannot be seen angiographically but can be detected at surgery or on pathological examination. This term comprises the cases of “occult AVMs” and “cryptic AVMs” in Yaşargil’s classification. It is important to emphasize that nowadays we should hesitate to assume the presence of an occult AVM when it is not seen on DSA and not found at surgery or on pathological examination. We think that most of these cases could have disease of another etiology that we should look for.

We think that these micro-AVMs are really different from the more common larger AVMs, because of the difficulty in making their diagnosis and because most of them are diagnosed after bleeding. It is difficult to ascertain if micro-AVMs are different from other AVMs because of the higher bleeding rate among micro-AVMs. Their small size and angiographic features (low-flow and low-volume AVMs) may explain the lack of compressive and/or ischemic symptoms due to steal phenomena. Moreover, the lack of a significant seizure history with micro-AVMs suggests that small intermittent microhemorrhages leading to epileptogenic deposits of hemosiderin are infrequent in these lesions.

With respect to diagnosis with MRA, although 3D time-of-flight provides higher spatial resolution than contrast-enhanced MRA, the latter better depicts the nidus of a small AVM when parenchymal bleeding is present, as contrast-enhanced MRA provides better background suppression without contamination from methemoglobin. Nevertheless, in our series, none of the 4 patients were diagnosed using MRA in the acute stage. These 4 patients had a first negative angiogram, and the MRA was performed several weeks after the bleeding, prompting the performance of a superselective angiogram. Even if MRA did not reveal the micronidus, we share the opinion that in a young patient with no other cause for a lobar hematoma, a second angiogram (with superselective catheterization) should be performed around 2–3 months after the initial hemorrhage.

Finally, we appreciate Dr. Heros’ warning about his unhappy experience. If the only draining vein has been sacrificed and there is doubt about resection of the nidus, an intraoperative angiogram could probably help. Anyway, these days, the need to follow a vein to find a micronidus is infrequent in our practice, as neuronavigation is routinely used to find the nidus of the micro-AVM at surgery. We usually meet with our neuroradiologists to draw exactly where the micronidus is located on the MRA, and sometimes we have to “fuse” the MR image with the selective angiogram to make the plan as accurate as possible.

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