Editorial

Developmental venous anomalies with arterial supply

Stacey Quintero Wolfe, M.D., and Roberto C. Heros, M.D.

Department of Neurosurgery, University of Miami, Florida

This group of investigators from the prestigious Seoul National University Hospital present us with the largest series to date of developmental venous anomalies (DVAs) that, in contrast to typical DVAs, are characterized by early arterial filling during catheter angiography; in other words, they are “arterialized.” The authors collected a remarkable 15 cases during 14 years at their institution and provide us with excellent illustrations for 2 of these cases and a table describing the others. As detailed by the authors, these lesions appeared to be typical DVAs (venous angiomas) on magnetic resonance (MR) imaging, but on catheter angiography they were rapidly visualized during the arterial phase, which implies that they have a significant arterial supply; however, as opposed to typical arteriovenous malformations (AVMs), they do not have a discrete arterial nidus or the typical large feeding vessels seen in true AVMs (although in 1 case the authors describe an enlarged arterial branch that could be embolized).

In addition to the description of the imaging characteristics of these lesions, Im et al. provide us with a description of surgical findings in the 3 cases that were treated surgically. Essentially they found enlarged arterialized (red) veins intermingled with normal brain parenchyma without a typical arterial nidus. On histological studies in these 3 cases the authors found dilated veins with thickened hyalinized walls and vessels with elastic lamina, characteristic of true arteries interspaced with otherwise normal brain parenchyma. Clinically, these lesions clearly behaved very differently from DVAs; 8 patients presented with hemorrhage and 1 with seizures. Of the former, 1 patient had 2 separate intracerebral hemorrhages and another bled 3 times. The authors treated 3 of these patients with resection, and 2 of them developed a significant venous infarction, which was fatal in 1 of the cases. The third patient was treated with a lobectomy and did well. Six patients underwent Gamma Knife surgery (GKS), but although the authors are enthusiastic about this treatment, they have little to say about its effectiveness because only 1 patient had follow-up angiography, and in this case the lesion was decreased in size but not obliterated. It appears that their enthusiasm about this form of treatment is based on the fact that none of the patients who underwent radiosurgery had a hemorrhage during the follow-up period, but most of these patients were treated recently and therefore this information is not very valuable.

This excellent paper begs additional discussion of the following points. Are these lesions separate entities or are they simply hybrids on a continuous spectrum between capillary telangiectasias and true AVMs? How should we name these lesions? Can these lesions be identified and separated from ordinary DVAs by MR imaging? How should we treat them? Finally, and most importantly, should our generally relaxed therapeutic attitude (no therapy necessary) with respect to asymptomatic DVAs change? We will address these questions but will admit up front that we do not have definitive answers for any of them.

Clearly, the most interesting question that this article raises is whether these lesions are separate types of vascular malformations or simply part of a continuum of these lesions. The authors discuss very nicely some of the evidence in the literature concerning the existence of mixed vascular malformations. In addition to the literature discussed by the authors, the senior author of this editorial and his colleagues reported an interesting case of a patient with a typical DVA who upon initial presentation was found to have a true AVM with an angiographically visible nidus that drained into the DVA. The patient was followed conservatively and 10 years later, angiography showed a new, different arterial nidus draining into the same DVA; however, the previously demonstrated arterial nidus had thrombosed (disappeared) in the interval. This case appears to be different from the cases reported by Im et al., in that arterial nidi were definitely demonstrated, whereas the currently reported cases had no such nidus.

The frequent coexistence of cavernous angiomas and DVAs is very well known, and in fact it has been suggested that when there is a hemorrhage associated with a DVA, a cavernous angioma is almost certain to coexist and to have been the source of the hemorrhage. Clearly, this article raises the different possibility that the cause of the hemorrhage can be arterialization of the DVA rather than a coexisting cavernous angioma, which none of these patients appeared to have had. Perhaps the most thoughtful and detailed discussion of this problem was that given by Dr. Charles Wilson during one of his talks as Honored Guest of the Congress of Neurological Surgeons. Although Dr. Wilson was not specifically addressing lesions similar to the ones presented by these authors, he did refer to the frequent coexistence in his own practice of not on-
ly cavernous angiomas in relation to DVAs but also what he called “cryptic AVMs,” which essentially were small AVMs, confirmed pathologically, that could not be visualized angiographically. He postulated that the basic pathogenesis of these lesions was venous hypertension within the territory drained by the DVA, and, although the presence of venous hypertension in relation to DVAs had not been demonstrated by the time he wrote the article (and, to our knowledge, has not been proven to date), Dr. Wilson believed that the following mechanisms could hypothetically be responsible for venous hypertension: 1) outflow restriction at the site where the trunk of the DVA enters a central vein or a venous sinus; 2) undampened transmission of acute increases in intracranial venous pressure through the large venous radicles; and 3) “inconspicuous thrombosis of an angiomatous radicle.”

In general agreement with Wilson, our suspicion is that indeed these lesions are basically DVAs that have become secondarily arterialized for reasons that remain speculative, but that perhaps have venous hypertension as a common denominator. A more recent article reviewing 15 cases of coexisting DVAs and cavernous angiomas emphasizes the importance of the venous anomaly in the pathogenesis of these mixed lesions. The authors removed the cavernoma, leaving the DVA intact in 9 of these cases, and 3 of these patients suffered a recurrence. At surgery, AVMs in 2 cases and a telangiectasia in the third were found within the radicles of the DVA. These authors suggested that consideration be given to coagulating the DVA to prevent recurrence, and did so successfully in 6 primary cases and in the 3 recurrences. However, most experienced cerebrovascular surgeons, including the senior author of this editorial, would strongly warn against such a policy, and would recommend always attempting to leave the DVA intact, as we will discuss later.

Another thoughtful commentator with a career-long interest in vascular lesions of the brain, Dr. Sean Mullan, has gone as far as to suggest that, in common with DVAs, the fundamental pathological component of AVMs is actually the abnormal venous drainage that occurs because of a failure of normal development of the cortical venous mantle. Arterial fistulization of these abnormal veins results eventually in mature cerebral AVMs. We certainly would not go this far, because in our experience, most ordinary AVMs generally drain into what would otherwise be normal cerebral veins rather than into the very anomalous veins that characterize DVAs.

A related question is that of the nomenclature of these lesions. The authors suggest the name “venous-predominant parenchymal AVMs” to denote the fact that clinically they behave aggressively, perhaps as aggressively as otherwise ordinary AVMs. Other investigators, as discussed by the authors, have preferred to call these lesions by a variety of other names, such as “arterialized developmental venous anomalies,” “venous angiomas with arterial blood supply,” “mixed angiomas,” “venous angiomas with arteriovenous shunts,” and so on. Although to us none of these names appears perfect, we do not have a better suggestion. The authors’ proposal of calling them “venous-predominant AVMs” appropriately denotes the aggressive clinical behavior of these lesions, but inappropriately suggests that they should be treated as AVMs, which clearly is not the case, as we will discuss. On the other hand, although names that imply that this is just a variety of DVA or a peculiar form of DVA with arterIALIZATION more appropriately describe the imaging characteristics of the lesion, these names also, inappropriately, connote the benign behavior that we have come to associate with DVAs. Perhaps the best compromise is simply to call them “mixed vascular malformations,” and then, because these cases are relatively uncommon, describe exactly what we see on a case-by-case basis.

A very important issue is whether these lesions can be identified noninvasively. Clearly, as the authors recommend, these lesions are rare enough that we should not subject to catheter angiography every patient with an otherwise typical asymptomatic DVA found on computed tomography scanning or on MR imaging. Are there any clues on the MR image that would lead us to investigate more aggressively some of these patients? The authors’ answer is no. Apparently, all of their patients had typical DVAs on MR imaging and in none of them could an arterial nidus typical of an AVM be identified. Is it possible that, given the faster filling of these lesions, they could be identified by the routine use of more sophisticated MR sequences, such as perfusion MR imaging? To our knowledge, there is no answer to this question in the literature and therefore, at the present time, we have to admit that we do not know how to separate these more aggressive lesions from the otherwise typical DVAs noninvasively. What this article certainly tells us is that when a patient presents with hemorrhage and perhaps with seizures, more aggressive investigation with catheter angiography is to be recommended.

How about treatment? The authors treated successfully with lobectomy a patient who presented with hemorrhage of a lesion confined to the frontal lobe. A second patient who developed hemorrhagic infarction after partial resection was treated successfully by temporal lobectomy and complete excision in a second operation. It certainly appears reasonable to consider complete excision in those lesions that are relatively small and located in the frontal pole or the temporal pole, where the DVA does not appear to drain a significant portion of the brain beyond what can be resected by lobectomy. We suspect that this will be the case in a very small minority of these patients. The third patient who was surgically treated by the authors died of a hemorrhagic infarction, illustrating the danger of occluding the main draining channel of the DVA. This danger, of course, is well known and does not require further emphasis.

Because it appears that surgery would be indicated only in a small minority of these cases, are there other available treatments? Although the authors used embolization in 1 case, apparently this was the exception in their series (the lesion had a relatively enlarged arterial feeding vessel); the rest of the patients, as well as the great majority of those reported in the literature, have no arterial nidus or large arterial feeding vessels amenable to embolization. Is radiosurgery effective for these lesions? As stated earlier, the follow-up duration in this series is inadequate to answer this question. In 1993, Lindquist and colleagues had already suggested that GKS can be effective in the treatment of venous angiomas. They reported 13 cases of venous angiomas presenting prior to 1991, and in 2 of these cases there was an associated AVM. In those 2 cases they restricted the radiosurgical field to the AVM, which was obliterated on follow-up angiography. Clearly, we do not have solid evidence of the effectiveness of GKS for these lesions, and
we will have to await long-term follow-up in a larger number of cases. Having treated 6 patients in this manner, the authors will be in a position in the future to give us some information in this respect. For now, we would generally recommend being very conservative with the great majority of these lesions, even when they present with hemorrhage. We would recommend restricting surgery to drainage of life-threatening hematomas, taking particular care to preserve the main draining veins of the DVA. If a clear arterial nidus can be identified, of course, that should be removed when appropriate, again being very careful not to occlude the main draining vein of the DVA.

Finally, should we change our current attitude toward DVAs, given the accumulating evidence that some of these lesions are arterialized and behave aggressively? Our answer to this question is “not at the present time.” In other words, we would still be very relaxed and optimistic in our advice to asymptomatic patients who present with a typical DVA diagnosed on computed tomography scans, MR images, or both. The senior author of this editorial sees an average of 2–3 patients a month who have typical DVAs. These patients have been referred because of a “vascular malformation” discovered on a noninvasive study ordered for nonspecific reasons such as dizziness, headaches, or even “nerves.” These patients are generally told that these lesions are perfectly benign developmental anomalies that do not result in any untoward events and that they do not require further follow-up. Clearly this recommendation is reassuring to the vast majority of these patients, who otherwise might experience a significantly decreased quality of life due to anxiety if they were told that they had a lesion that could be dangerous and should be followed periodically with either noninvasive studies or, worse, with catheter angiography. Would the fact that indeed some of these patients may have the rare aggressive type of lesion described by these authors justify the anxiety that might be provoked in the large number of asymptomatic patients with a benign DVA if they were to be told of such a possibility? We believe that for the time being, these mixed lesions appear to be rare enough compared to the ordinary benign DVAs, that until noninvasive ways to identify the more aggressive lesions are available or until further evidence accumulates to indicate that these mixed lesions are more common than we think, we should continue to tell these patients to go about their lives without worrying about their developmental anomaly and without any need for follow-up imaging in the future.

We are indebted to these authors for providing the best characterization to date of these interesting mixed vascular malformations.

References

RESPONSE: On behalf of all participants in this study, we appreciate the detailed and constructive editorial comments by Drs. Wolfe and Heros.

We certainly share their conservative policy in the treatment of this rare subtype of vascular malformation when it is found incidentally. The point of our investigation was to separate this entity from typical DVAs, in which the angiographic appearance is identical in the venous phase and the natural course is benign. However, this rare subtype of vascular malformation, as described in our study, presumably follows a clinical course typical of AVMs, based on our observations. In the present study, clinically silent lesions showed low flow and slow circulation on angiography, whereas in the patients with prominent arterial components and very early venous drainage, the disease tended to follow an aggressive clinical course, and the fact that 3 patients experienced rebleeding episodes strongly supports this suggestion. The relatively aggressive clinical behaviors of these vascular malformations support the suggestion that this type of vascular malformation should not be considered a simple variant of DVA, but rather a particular type of AVM. For this reason, we designated these lesions “venous-predominant parenchymal AVM,” with the purpose of stressing their relatively aggressive clinical behaviors.

An important and practical issue is how to differentiate this rare type of lesion from a typical DVA noninvasively. Based on our observations, shortened circulation time in this lesion seems to be the most important difference from typical DVAs, and it would be possible to differentiate these if any noninvasive imaging method can show this difference in circulation time. We expect that time-resolved contrast-enhanced MR angiography can possibly differentiate these lesions by temporal separation of early-appearing veins in this rare type of vascular malformation. (DOI: 10.3171/JNS/2008/108/6/1139)

SO-HYANG IM, M.D.
Dongguk University Hospital
Gyeonggido, Korea

MOON HEE HAN, M.D.
Seoul National University Hospital
Seoul, Korea