Editorial

Fusiform middle cerebral artery aneurysms

ROBERTO C. HEROS, M.D.

Department of Neurosurgery, University of Miami, Florida

In this editorial comment I discuss two related but very different papers appearing in this issue of the Journal. In the first paper, Day, et al., put forth the important hypothesis that essentially all fusiform middle cerebral artery (MCA) aneurysms that are not related to atherosclerosis develop as a result of arterial dissection. The strength of this paper lies in the thoroughness and eloquence with which the authors develop the rationale for their hypothesis, which is based on extensive experience, including 40 cases from their institutions and an exhaustive review of the literature, which yielded an additional 62 cases; its weakness is the lack of pathological confirmation. The second paper, by Horie, et al., also involves giant fusiform MCA aneurysms, but these authors propose that there are three different mechanisms of rupture that have as a common denominator weakness of the internal elastic lamina (IEL) with intimal thickening. The strength of this paper is that it presents pathological findings in two of the cases; unfortunately, the case in which the authors postulate that dissection was the final mechanism of rupture was not studied pathologically. The obvious weakness of this paper is that it is based on only three cases.

The idea that some fusiform MCA aneurysms are essentially dissecting lesions is not new, and a few cases in which dissection is certainly suggested as a mechanism have been presented in the literature and are discussed by the authors. What is novel is the concept that essentially all of these lesions that are not caused by atherosclerosis are dissecting aneurysms. Dr. Day and his colleagues have been discussing this hypothesis for years in formal presentations; however, in this article, they put forth their concept in a very elegant fashion and support it with very convincing, although inconclusive evidence, which includes at least one case that was documented to progress in the expected manner of a dissection (Fig. 6A and B). They also present two examples of what clearly appear to be dissecting aneurysms that presented with hemorrhage (Fig. 3A–C). Unfortunately, however, there is no documentation of the evolution of these aneurysms, and the question of whether they would have eventually progressed to become large or giant fusiform MCA aneurysms, as the authors speculate that they could, remains unanswered.

In addition to supporting evidence from magnetic resonance imaging and angiographic studies, these authors provide beautiful artistic sketches of their proposed mechanism of dissection that leads initially to rupture or to stenosis and/or occlusion of the lumen, with possible ischemic symptoms. With time, these lesions can grow into larger, fusiform aneurysms or into classic giant “serpentine” aneurysms (Fig. 8). The weakness of the paper, the lack of pathological confirmation, is perfectly understandable in view of the fact that, because most of these lesions involve the main trunk of the MCA (M1 segment), the aneurysm was left in situ at surgery. This was properly done because of the almost inevitable presence of important perforating vessels coursing from the segment in which the aneurysm was located.

The value of the paper by Horie, et al., is that in two of their cases they were able to resect the aneurysm completely and to study its pathological features carefully. Excision of the aneurysm was possible in these two cases because it was not located in the main trunk of the MCA, but rather in a division in which perforating vessels are often not present or are of less significance. In these two cases, the striking finding was of a fragmented or completely absent IEL and a thickened intima. In the first case the authors concluded that the rupture had been a “classic” one from a weakened arterial wall, and in the other case they concluded that the rupture was caused by a hemorrhage within an atheroma; they found no evidence of dissection in either of these cases. The third case, in which unfortunately there was no pathological confirmation because the surgeon left the aneurysm in place, was attributed to dissection given the appearance of the lesion at surgery. These authors have provided three beautiful composite pictures illustrating the diagnostic studies, the treatment used, and the pathological findings in the two cases in which the lesions were resected.

In terms of aneurysm origins, after a careful reading of these two papers and leaning on my own personal experience, I have become convinced that indeed many fusiform lesions of the MCA are due to arterial dissection. Nevertheless, I have not been able to convince myself that all or perhaps even most fusiform MCA aneurysms that are not due to atherosclerosis are due to dissection. It is clear that Horie
and his colleagues could find no evidence of dissection in their pathological study of the aneurysm that was not due to atherosclerosis. Although Day, et al., illustrate some cases of small, fusiform aneurysms of the MCA that clearly have the typical appearance of dissecting lesions, the three cases they present of a large, a giant, and a serpentine fusiform aneurysm of the MCA have none of the features that we have come to recognize as typical of a dissection. Additionally, it is difficult to understand why, if most of these lesions are dissecting aneurysms, they behave so differently clinically from dissecting aneurysms of the supraclinoid portion of the internal carotid artery (ICA) and of the intracranial segment of the vertebral artery (VA), which so frequently present with subarachnoid hemorrhage, have a marked tendency to rebleed early, and generally do not grow to giant size.

Day, et al., provide two interesting and reasonable explanations for these differences. They think that, because both the ICA and the VA are tethered to the dura as they enter the intracranial compartment, the dissection tends to rupture because of this tethering, as opposed to MCA dissections in which, because of the lack of tethering, “the pulsatile and expansive forces in a growing dissection may be dissipated by elongation and the development of ectasia, with gradual enlargement of the dissecting segment resulting in the formation of a giant or serpentine aneurysm.” Furthermore, they review evidence that in some of the MCA dissections that have been studied pathologically, the intramural hemorrhage was observed to occur between the elastica and the media (subintimal dissection) as opposed to studies of VA dissections in which the splitting appeared to take place between the media and the adventitia (subadventitial dissection), which could explain the potential for a more frequent presentation with hemorrhage. It is also interesting that the authors performed pathological studies in two of their patients in whom a fusiform MCA aneurysm occurred during the 1st year of life. In neither of these two cases were they able to find a dissection, and rather found “an aneurysm wall composed of a thin layer of fibrous tissue without an elastica interna,” lending support to the hypothesis of Horie, et al., that in most of these cases, a defect in the IEL is the basic underlying process.

In reference to treatment, these two papers illustrate clearly the advantage of distal revascularization and proximal occlusion for definitive treatment of these aneurysms. It is important to note that the aneurysms in the cases presented by Horie, et al., were located in divisions of the MCA rather than in the main trunk. Therefore, they presumed, correctly, that a relatively low-flow bypass from the superficial temporal artery would be sufficient to maintain distal circulation. Nevertheless, I fully agree with Day and his colleagues that a high-flow bypass, either with saphenous vein or with an interposed arterial graft, is preferable (I would say necessary) whenever the main trunk of the MCA is to be occluded. It is also important to realize that aneurysms in the main trunk of the MCA cannot be “trapped” or excised, as were the two aneurysms reported by Horie, et al., which were located more distally in a division of the MCA. The obvious reason for this is that the important lenticulostriate perforating vessels come mostly from the M trunk, and excision of that segment, with its perforating vessels, would in most instances lead to a profound neurological deficit.

It has been remarkable to me to see in several of my own cases that were treated by M1 occlusion proximal to the aneurysm and a distal bypass, how the aneurysm becomes almost totally thrombosed, and yet a very small channel supplying the perforating branches remains open due to retrograde flow from the distal bypass. I also agree with Day, et al., that once these aneurysms become symptomatic, they should be treated aggressively because of their poor natural history, as those authors have discussed. I would go even further and say that once the lesions reach a very large or giant size, if the patient is relatively young and healthy, they should be treated prophylactically to avoid having to treat the aneurysms at a time when they have become symptomatic because of major cerebral edema. In these circumstances the results of the treatment are likely to be poor, as happened in one of the cases discussed by Day, et al., and also in a case that we reported on previously.1

In brief, these two papers provide convincing evidence that at least some fusiform MCA aneurysms are due to dissection, and the one by Day, et al., strongly suggests that dissection may be the source of many, if not most, of these aneurysms. Both papers reinforce the notion that these lesions ought to be treated aggressively when they become symptomatic or reach a large size, and that the preferred treatment is proximal arterial occlusion with distal revascularization. Both of these excellent papers are thought-provoking and both are beautifully illustrated.

Reference


RESPONSE: The comments by Dr. Heros are much appreciated, and underscore the difficulties with pathological confirmation of the various hypotheses about the genesis of these most interesting lesions. In my opinion, the presence of weakness of the IEL with intimal thickening (the common underlying substrate proposed by Horie, et al.) is not excluded by our suggestions. When dissections occur in large extracranial vessels, the separation of layers is generally easily identified, both pathologically and on neuroimaging. Somewhere along their course, both the ICA and VA go through a transition into an intracranial vessel, with shedding of most of their adventitia. This “transitional segment” is more elongated and apparent in the carotid artery as it traverses the cavernous sinus, supported by the surrounding venous wall and dura mater. The VA, however, goes through a much more abrupt transition. Both of these regions may be sites where transitional defects may occur, predisposing the vessel to dissection. Inside the brain and a fully mature intracranial vessel, the intralaminar nature of a dissection may be less obvious, at least in most cases. Speculating further, the possibility that some type of congenital vessel defect, as described by Horie, et al., could be the underlying substrate that predisposes the vessel to degenerate, dissect, generally weaken, and evolve into the variations described in our article, is perfectly reasonable. One day, perhaps, the “perfect” case will come along and we will be able to document the pathological features and natural evolution of fusiform MCA aneurysms.