AJNR

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AJNR Am J Neuroradiol 1998, 19 (7) 1274-1275
http://www.ajnr.org/content/19/7/1274.citation

Commentary

Venous Hypertensive Encephalopathy

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Dural arteriovenous fistulae (DAVFs) in adults are generally regarded to be acquired lesions that may follow trauma, surgery, or thrombosis of a dural sinus (1, 2). Normal dural sinuses contain small arteriovenous anastomoses which do not appear to create any problem (3). In a setting of venous hypertension possibly in association with local angiogenic factors or congenital abnormalities of the dura, however, there can be enlargement of these preexisting arteriovenous anastomoses. The arteriovenous shunting then exacerbates the venous hypertension. Venous hypertension can then be even further compounded by vessel wall hypertrophy or thrombosis of the draining veins in response to the abnormal high flow in these vessels, which can cause stenosis or occlusion of these draining veins. This whole process can result in a wide spectrum of clinical presentations, and the fistulae can vary from asymptomatic lesions to those which are severely disabling or even life-threatening.

In this issue of *The American Journal of Neuroradiology,* Hurst et al (page 1267) describe a series of patients with dementia related to DAVFs. Unlike patients with parenchymal arteriovenous malformations who may present with dementia, patients with dural fistulae have only minimal, if any, arterial contribution from pial arteries to the arteriovenous shunt. Therefore there is no likelihood that arterial steal is the source of these symptoms. Venous hypertension is the culprit, and this is well demonstrated by angiography and even, as in this series, by pathologic findings. All patients had angiographic evidence of venous occlusive disease with redistribution of flow. Extensive congestion of cerebral cortical and deep veins could be seen, with marked slowing of the arteriovenous transit time. Cross sectional imaging showed parenchymal edema remote from the fistula site. Autopsy results found in one patient provided pathologic evidence of chronic venous hypertension. With all these pathologic changes, it is no wonder these patients exhibited signs of impaired cognitive function. By angiographic criteria alone, these DAVFs have characteristics of aggressive lesions. Awad et al (4) have shown that leptomeningeal venous drainage, variceal or aneurysmal venous dilatation, or involvement of the deep venous system are features strongly correlated with progressive neurologic deficits and even intracranial hemorrhage.

Other authors have described focal central nervous system deficits associated with DAVFs (4–6). These types of symptoms are much less common than bruit, headache, and visual changes, but, like intracranial hemorrhage, appear to be associated with drainage of the fistula to cortical or deep cerebral veins. In vein of Galen malformations, another condition causing venous hypertension, Quisling and Mickle (7) showed the degree of brain parenchymal damage is roughly proportional to measured venous pressures. Lasjaunias et al (6) reported several cases of cognitive deficits from DAVFs, again with signs of impaired venous drainage of the brain, though dementia as the presenting symptom of a DAVF is unusual.

There are a number of endovascular and open surgical methods available for treating DAVFs (4–6, 8–10). One can occlude the arterial feeders by filling the fistula with liquid adhesives, sclerosing agents, particles or coils. Large particles or coils are less likely to pass through dangerous anastomoses or into vasa nervora supplying cranial nerves than are liquid embolic agents. These larger emboli, however, produce occlusion well proximal to the site of the fistula, and allow a high rate of recurrence since collateral vessels can continue to fill the lesion. Liquid adhesives provide more distal penetration of the fistula and a more permanent occlusion, but require more skill and training if complications are to be avoided. Transvenous embolization obliterates the fistula from the venous side, and usually involves occluding the vein or sinus that drains the fistula. Coils are usually used for this purpose. Care must be taken not to disturb the venous drainage of the brain by this procedure. Some DAVFs cannot be treated with this technique because the sinus involved with the fistula also participates in drainage of normal structures. The venous drainage pathways in each patient must be thoroughly understood before contemplating therapy.

Early surgical methods involved simple ligation of the major arterial branches supplying the fistula, but resulted in a high recurrence rate because of the extensive collateral pathways leading to the dura. More modern methods have added sectioning the dura around the involved sinus, cauterizing or ligating the feeding arteries in the process. Arterialized veins that redistribute the flow to parenchymal veins can also be interrupted. This approach is very effective in achieving long-term control of symptoms, although residual or recurrent fistulae can be seen on occasion. A more radical and definitive approach is to resect the involved sinus or vein and the adjacent dura completely (4). One may also employ techniques that combine endovascular and open surgical procedures such as preoperative embolization of arterial feeders and intraoperative embolization of surgically exposed arteries and veins.

Treatment of DAVFs may be merely palliative; larger arterial inputs are occluded in hopes of reducing the degree of venous hypertension and reducing

the symptoms. This appears to have been Hurst et al's therapeutic approach. This treatment may also have a curative objective. Occlusion or removal of the draining vein can be effective if all arterial feeders converge on a single venous structure as long as it is not a required draining pathway for normal brain. Alternatively, one must, using either endovascular or surgical methods, interrupt *all* arteriovenous shunts to achieve a cure. Long-term cures with transarterial embolization are less common than with transvenous occlusion of the draining vein or with surgical treatment of the fistula. When transarterial embolization has produced complete obliteration of the DAVF, it often occurs by a mechanism of slowing flow through an already diseased draining sinus. Thrombosis of the sinus follows and results in the occlusion of all of the arteriovenous shunts. This process is similar to the spontaneous thrombosis of fistulae that occasionally occurs, especially with DAVFs involving the cavernous sinuses. On the other hand, thrombosis of a portion of the venous outlets of a DAVF can be the cause of acute worsening of symptoms, if there is exacerbation of venous hypertension with disturbance of drainage of normal structures, and especially if there are persistent arteriovenous shunts. Any transarterial embolization with agents that are known to have a significant chance of recanalization, such as polyvinyl alcohol particles, could not be expected to reliably produce long-term cures. Consequently, these agents are usually used for embolization immediately prior to definitive surgical treatment of the DAVF.

There is no question that the patients in this series by Hurst et al improved after embolization. One could argue that a more aggressive approach should be taken for these lesions, and that this clinical improvement after partial treatment provides a stronger argument for more definitive treatment of these troublesome lesions. Additionally, the aggressive character of these types of fistulae is well-known so why allow such lesions to recur and cause further mischief? All of these lesions could have been more permanently occluded with either a transvenous embolization, transarterial embolization with more permanent agents, or combined embolization plus surgery. Each patient with a DAVF must be evaluated individually, and a therapeutic plan devised that takes into account the patient's clinical status, the arterial and venous architecture of the lesion, and the various endovascular and surgical therapeutic options that are available. Aggressive lesions often require aggressive therapy.

Traditionally, DAVFs have been considered "cured" if all the arteriovenous connections have been interrupted either surgically or with permanent embolic agents. It is ironic that many "cures" are accomplished by occlusion of the draining sinus, even though occlusion of the sinus may have been the inciting factor in the process of creating these fistulae. We have certainly seen complete "cures" from treatment, with development of a new arteriovenous fistula remote from the original lesion years later (11). These cases of recurrent or multiple lesions suggest that, once a patient has developed a DAVF, it is as though restless dura is just waiting to create a new lesion. All of the DAVF treatments, whether endovascular or directly surgical, in many ways are quite primitive and do not address the pathophysiologic basis of these lesions. It may eventually come to be that recanalization or bypass of occluded sinuses, in addition to pharmacologic inhibition of local angiogenic factors, may provide the most durable treatment of these difficult lesions. Although our understanding of these lesions and the treatments we have to offer for them are primitive, strategies such as ones presented in this important paper by Hurst et al can point the way to a greater understanding of this problem.

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