Endovascular occlusion of a ruptured transitional aneurysm associated with a developmental venous anomaly

Case report

ANDREW F. DUCRUET, M.D., CHRISTOPHER P. KELLNER, M.D., E. SANDER CONNOLLY JR., M.D., AND PHILIP M. MEYERS, M.D.

Department of Neurological Surgery, Columbia University Medical Center, New York, New York

In 1966, McCormick classified cerebrovascular malformations into 4 categories: AVM, venous angioma, CM, and capillary telangiectasia. Although mixed vascular malformations have been described, McCormick's classification is still widely accepted. Veinous angiomas, now referred to as DVAs or cerebral venous malformations, are the most common cerebrovascular lesion in both autopsy and imaging studies. Despite a high incidence, these lesions are rarely a cause of intracerebral hemorrhage. Due to the vanishingly small risk of hemorrhage, most reports support a conservative approach to DVAs, and few authors suggest any form of treatment.

In this case, we describe the unusual presentation of intracerebral hemorrhage associated with a mixed vascular lesion: a DVA with associated arteriovenous shunting more typical of a pial AVM and an aneurysm transitioning from PerA to draining vein. Mixed vascular lesions form a subset of DVAs that may show increased risk of hemorrhage and may therefore necessitate treatment. This is the first report documenting the successful use of endovascular embolic occlusion of the transitional aneurysm in a lesion of this variety, and it supports the argument for aggressive management of the subset of DVAs that present with hemorrhage. We provide a description of the case along with a discussion of the origins, frequency, and concerns associated with this lesion and its management.

Case Report

History and Examination. This 33-year-old man without significant medical history developed acute onset of gait ataxia, vertigo, occipital headache, and mild left leg weakness. Brain CT scans obtained without ad-
dition of contrast material revealed an acute right frontal intraparenchymal hemorrhage above the corpus callosum (Fig. 1a). Catheter angiography demonstrated a right frontal DVA with associated arteriovenous shunting, and a 3-mm aneurysm arising from the transition between the right PerA and draining vein (Fig. 1b–d). The vascular malformation drained predominantly to the right internal cerebral vein. Over several days, the aneurysm increased in size from 3 mm at initial diagnosis to 5 mm at the time of its endovascular occlusion (Fig. 2a and b).

**Operation and Postoperative Course.** Microcatheter arteriography was performed in the right PerA and a perforator branch arising from this artery, from which the aneurysm formed (Fig. 2c and d). Embolization and occlusion of the transitional aneurysm was then performed using Cordis N-butyl cyanoacrylate (Tru-Fill, JNJ Cordis) mixed with glacial acetic acid and iodized oil (Lipiodol). A control angiography study showed complete occlusion of the aneurysm, without other arterial or venous branch vessel occlusion (Fig. 3). The patient was discharged on hospital Day 12 without residual neurological deficits, and he continues to be in good health through 10 months of ongoing follow-up.

**Discussion**

Due to the high incidence but rare clinical presentation of DVAs, they are thought to be relatively benign lesions. A small subset of these lesions, however, is associated with other cerebrovascular abnormalities, such as AVMs, CMs, venous anomalies, and AVFs. It has been suggested that these mixed cerebrovascular lesions carry an increased risk of hemorrhage, and therefore merit increased monitoring and potentially even treatment. It is unknown whether increased morbidity results from an inherent developmental abnormality in these vessels or if abnormal blood flow and high pressures result in aneurysm formation.

Because of the histological similarities between DVAs and the dilated, single-layered veins of the fetal cerebral vasculature, it has been suggested that DVAs are vestigial remnants of the fetal circulation.
proposed a coherent theory linking the development of DVAs and pial AVMs, suggesting that both form when the fetus is 40–80 mm in length, during the period of time when pial-to-dural venous connections regress. If excessive numbers of veins draining the cortical venous plexus to the dural venous plexus occlude or if the occlusion spreads into the cortical venous plexus itself, conditions favoring formation of a DVA are created. A deep network of veins that drain to the cortical venous plexus through a single adjoining vein is then likely to develop. The arrangement of an abnormal deep venous network (referred to as a “star”), an adjoining draining vein, and fewer pial veins contains all the elements of a DVA.

Some authors have theorized that this initial malformation then sets the stage for other cerebrovascular abnormalities. If the “star” cluster of deep veins forms a thrombosis, the cluster may form an AVF as the clot is absorbed, as well as recruit an artery to its nidus.4,5,28 Furthermore, chronic venous hypertension of 2–3 months duration without associated venous or sinus thrombosis has been shown experimentally to induce dural or subcuneous AVF.5,20 We suspect that the AVF in this case developed in response to the venous hypertension observed secondary to stenosis of the venous outflow of the DVA (Fig. 1d) and suggested by collateral venous drainage to the superior sagittal sinus, representing a clinical manifestation of the basic mechanisms proposed by the aforementioned experimental studies.

Hemorrhage in the presence of a DVA may result from one of a number of reasons. Case reports have described hemorrhage following thrombotic occlusion of the associated draining veins.2 Other authors have found evidence of CMs in the presence of DVAs, and have suggested that occult CMs may play a large role in DVA hemorrhage.15 Other studies, however, have examined ruptured DVAs histologically, and found no evidence of associated CMs.23 Rarely, venous aneurysms have been reported as a potential source of bleeding.22

**Conclusions**

With significant associated venous hypertension,
arteriovenous shunting, and a transitional aneurysm, the DVA in this case exhibited multiple factors increasing the risk of hemorrhage. After observing a dramatic increase in the size of the aneurysm from 3 to 5 mm over a 1-week period, we suspected that this probably represented a dynamic lesion secondary to abnormally high flow or pressure through the DVA. Due to the symptomatic presentation and the dynamic nature of the lesion, we believed that treatment was merited, and endovascular embolization of the aneurysm was successfully performed. Once the perforating vessel harboring the aneurysm was successfully catheterized, glue was used to embolize the vessel, thus occluding the aneurysm. This avoided the ischemic complications involved with either embolization of the large PerA or compromise of the venous outflow of the DVA. This technique was also used because we believed that catheterizing the aneurysm itself would lead to rupture of its parent artery. In summary, this case provides a rare example of a dynamic venous lesion and the first documented case of its effective treatment with endovascular obliteration of an associated ruptured transitional aneurysm.

Disclaimer

The authors do not report any conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

References


Address correspondence to: Andrew F. Ducruet, M.D., Department of Neurological Surgery, Columbia University, 630 West 168th Street, Room 5-454, New York, New York 10032. email: afd12@columbia.edu.