Developmental venous anomaly with symptomatic thrombosis of the draining vein

Report of 2 cases

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Developmental venous anomalies (DVAs) are generally considered a benign and asymptomatic finding on CT and MR imaging. The authors report 2 cases of spontaneous thrombosis of the draining vein of a DVA depicted on CT and MR imaging. One patient presented with a nonhemorrhagic transient ischemia, which was successfully treated with anticoagulant therapy. The second patient presented with ischemia complicated by hemorrhagic conversion. (DOI: 10.3171/JNS.2008.109.12.1119)

Key Words • developmental venous anomaly • neuroimaging • thrombosis

Developmental venous anomalies (DVAs) are the most frequently encountered cerebral vascular malformation, and their incidence is as high as 2.5%.4 While generally considered incidental findings and unlikely to be the source of symptoms, a small number of such cases will demonstrate complications such as venous infarction, nonhemorrhagic ischemia, and intracranial hemorrhage due to thrombosis of the common draining vein. The authors report imaging findings in 2 patients with DVAs that became symptomatic after thrombosis of the draining vein. These cases demonstrate that DVAs can sometimes cause hemorrhagic and nonhemorrhagic ischemic complications, and this possibility must be considered in the appropriate clinical setting.

Case Reports

Case 1

This 38-year-old man, without a significant medical history, was found unresponsive and drooling. Initial physical examination demonstrated right facial droop, decreased sensation to light touch and pressure over the anterior aspect of his tongue, and mild dysarthria. Initial noncontrast head CT scanning (Fig. 1A) demonstrated a left frontal lobe linear high-attenuation vascular structure extending from the frontal horn of the left lateral ventricle to the cortex. Magnetic resonance imaging revealed a linear hypointense vascular structure on T2 and FLAIR sequences with slightly hyperintense T1 signal corresponding to the high-attenuation CT abnormality. This structure exhibited susceptibility effects on diffusion-weighted sequences and gradient echo images with signal loss (Fig. 1B). Contrast-enhanced T1-weighted imaging (Fig. 1C) and contrast-enhanced MRV (Fig. 1D) did not show contrast enhancement through the draining vein. However, multiple small enhancing vessels were demonstrated adjacent to the abnormality in the characteristic “caput medusae” configuration of a DVA. There was no evidence of an associated cavernoma. The patient’s symptoms resolved within 24 hours of anticoagulant therapy. Follow-up MR imaging (Fig. 1E) after 8 weeks of anticoagulant therapy demonstrated a patent prominent transmedullary draining vein with normal contrast enhancement. The patient continued to be symptom free up to 6 months following initial presentation.

Case 2

This 52-year-old man, with a history of an arteriovenous malformation that was diagnosed in 1982, presented to an outside institution with a 3-day history of eye pain and a 1-day history of ataxia, nausea, and vomiting. The initial noncontrast CT scan demonstrated punctate hemorrhages (Fig. 2A) and, on the reformatted images, a high-attenuation linear structure perpendicular to the cortex in the right parietal lobe (Fig. 2B). Magnetic resonance imaging revealed

Abbreviations used in this paper: DVA = developmental venous anomaly; MRV = MR venography.
T1 hyperintensity within this linear structure in the right parietal lobe (Fig. 2C). There was abnormal T2 and FLAIR signal intensity in the adjacent cortex and white matter of the right parietal and occipital lobes consistent with edema. No restricted diffusion was visualized in the corresponding region to suggest acute infarction. Contrast-enhanced and 2D time-of-flight MRV demonstrated no flow within this structure. Multiple enhancing vessels adjacent to the abnormality in the characteristic “caput medusae” configuration of a DVA were visualized with signal dropout on gradient echo imaging (Fig. 2D), characteristic for the susceptibility effects of thrombus. Three hours following the initial MR imaging the patient suddenly lost consciousness. An emergency noncontrast head CT demonstrated high-attenuation blood within the right parietooccipital lobe (Fig. 2E) consistent with acute intraparenchymal hemorrhage. The patient was immediately taken to the operating room for a craniectomy and clot removal. He survived but, at the time of discharge to an extended care facility, he was not responding to verbal commands.

Discussion

Developmental venous anomalies, also termed venous angiomas, are the most commonly encountered intracranial vascular lesion noted on routine brain imaging. Fortunately, as most DVAs are asymptomatic, treatment is not considered necessary and surgery is not an option because these provide venous drainage of normal brain. Developmental venous anomalies are characterized by a network of dilated medullary veins, surrounding and draining into a large central vein, which runs perpendicular to the cortex or ventricular ependyma. Unlike other vascular lesions such as cavernous angiomas, the intervening brain parenchyma within a DVA is normal, suggesting that a DVA represents a developmental anomaly rather than a pathological lesion. Considering this fact, Lasjaunias et al. proposed that the name be changed from “venous angioma” to “developmental venous anomalies.” These authors believed that DVAs represented nonpathological variations of venous drainage.

The exact pathogenesis of a DVA is unclear, although many believe these to be congenital lesions. Developmental venous anomalies can be found in children in whom normal draining veins are absent in the region of the DVA. The DVA, as well as its draining vein in such cases, is the primary venous drainage system. Currently, a DVA is thought to represent a primary dysplasia of capillaries and small transcerebral veins or a compensatory mechanism caused by an intrauterine thrombosis of normal venous pathways. The draining vein can empty into
the superficial venous system such as the cortical veins, the dural venous sinuses, the deep subependymal veins, or some combination. Because the DVA is a compensatory mechanism, the parenchyma being drained has no other normal venous drainage system. This is supported by the occurrence of venous infarction whenever the draining vein is thrombosed or surgically ligated. In addition, DVAs have been implicated in the genesis of cavernous malformations, and it is possible that thrombosis of a DVA may represent the factor eventually leading to development of the cavernous malformations.

The preceding cases demonstrate what has been proposed as the inciting event leading to DVA-associated ischemia and hemorrhage: thrombosis of the draining vein. Hemorrhage is a well-known complication of venous thrombosis and is thought to occur from increased venous pressure in the parenchyma drained by the thrombosed vein. Previous authors have demonstrated hemorrhagic or nonhemorrhagic infarcts resulting from thrombosis of the draining vein of a DVA. Angiographic documentation demonstrating an endoluminal clot has been provided by Konan et al., and MR and CT imaging demonstration of thrombosis of the draining vein leading to intracerebral hemorrhage has been provided by Merten et al.

We have demonstrated the progressive imaging findings associated with thrombosis of the transmedullary draining vein preceding both nonhemorrhagic ischemia and intracranial hemorrhage. In the former case, the patient was treated with anticoagulation that led to resolution of the presenting symptoms and recanalization of the thrombosed vein on follow-up imaging. Treatment of DVAs is not recommended since this will often result in a venous infarction. This has led to DVAs being considered incidental and benign entities, but reports of hemorrhagic and nonhemorrhagic complications in the literature are growing.

The imaging findings in these cases are characteristic. The CT studies revealed a linear structure of high attenuation that could be confused with subarachnoid hemorrhage. Thin-section CT reconstructions may allow this distinction. Magnetic resonance imaging performed acutely may show low signal in the thrombosed vein due to the susceptibility effects of acute clot. It is for this reason that susceptibility weighted images or gradient echo images are essential since they could demonstrate both the intravascular clot and any acute parenchymal hemorrhage. In addition, a DVA should not have any significant abnormal signal in surrounding brain on T2-weighted images, but abnormalities on FLAIR imaging would be expected in a case of symptomatic thrombosis.

We believe that increased awareness of the possibility of thrombosis of a DVA is necessary so that the parenchymal abnormality, which does not conform to a usual vascular territory, is not mistaken for a tumor and to establish an early diagnosis so that therapy may be initiated to prevent progression to a completed venous infarction or hemorrhage.

**Disclaimer**

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References


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