Ischemic Complication of a Cerebral Developmental Venous Anomaly: Case Report and Review of the Literature

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Abstract: We report a case of a nonhemorrhagic infarct associated with a thrombosed developmental venous anomaly (DVA), with secondary gliosis and Wallerian degeneration. The initial MRI scan showed an acute ischemic infarct in the region of the DVA, seen as a region of restricted diffusion on diffusion-weighted imaging (DWI), with later development of encephalomalacia and Wallerian degeneration on follow-up MRI. No blood products were seen. We believe that thrombosis of the collector vein of a DVA with associated infarction is a rare but possible complication that should be considered within the proper clinical setting and can be easily and confidently diagnosed by means of DWI. Index Terms: Developmental venous anomaly—Infarct—MRI—Diffusion-weighted imaging.

Developmental venous anomalies (DVAs; venous angiomas) are presumably benign embryologic vascular variants that are usually incidentally discovered on enhanced CT or MRI of the brain. Although hemorrhagic complications have been described in association with DVAs, isolated ischemic events remain much less frequent manifestations of this entity. In this report, we present a case of initially misdiagnosed venous infarct secondary to a cerebral DVA. Misdiagnosis may occur as a result of the heterogeneous imaging characteristics of venous infarcts in general (1). In 1996, Bakac and Wandl (1) reported nine cases of venous infarcts misdiagnosed as arterial infarcts or primary intracerebral hemorrhage. In the literature, venous infarcts have even been misdiagnosed as tumors or demyelinating diseases, and many times, they have been unnecessarily biopsied (2,3). The availability of diffusion-weighted imaging (DWI) makes it much easier to diagnose venous infarcts such as in our case.

CASE REPORT

A 26-year-old female patient presented to an outside hospital with complaints of acute numbness and weakness involving the upper and lower extremities on the right side. Her symptoms progressed over 2 days to the point that she could no longer use her right arm and she could not walk.

She had been in excellent health before these symptoms. She gave birth to her second child via an uncomplicated vaginal delivery 40 days before admission to the hospital. Two weeks postpartum, the patient went back on birth control pills and resumed smoking. She noted nonspecific headaches for several weeks before admission. Her past medical history was notable for a mother who had a deep venous thrombosis of the leg at the age of 35 years during a period of immobility caused by back pain.

Initial MRI of the brain was performed and revealed the presence of an area of bright signal intensity on DWI in the left frontoparietal region (Fig. 1). On the postcontrast images, this area of restricted diffusion corresponded to the location of a DVA, with a collector vein draining to the surface of the left lateral ventricle (Fig. 2). MR angiography of the intracranial and cervical vessels was normal. Routine laboratory tests, including hematology and coagulation studies, were normal.

On presentation to our hospital 2.5 months after the onset of her symptoms, the patient's symptoms had improved, with only a mild gait abnormality and minimal sensory deficit. Her physical examination revealed decreased pinprick sensation on the right side compared with the left side, with preservation of the temperature, vibration, and light touch sensations. Motor examination revealed a slight right pronator drift with slowed fine finger movements and clumsiness of movements of the right leg.

Further testing included a normal echocardiogram. The following additional laboratory studies were either negative or within the range of normal: antinuclear antibody, anticardiolipin antibodies, antithrombin III, factor V Leiden mutation, protein C, protein S, activated protein C resistance, and homocysteine.

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Repeat MRI performed at our institution 4.5 months after the episode of weakness revealed an area of complex signal intensity with cystic and solid components in the left frontoparietal region at the same location as the previously seen DWI abnormality (Fig. 3). No mass effect or volume loss was noted. Changes of Wallerian degeneration were seen on the left side as high signal intensity on T2-weighted and fluid-attenuated inversion-recovery-weighted images along the course of the corticospinal tracts (Fig. 4). On postcontrast images, the previously described branching vascular structure compatible with a DVA was again noted in the periphery of the gliotic area. The major venous sinuses were patent as documented by MR venography. There was no evidence of blood products or other abnormal signal intensity lesions in the rest of the brain.

**DISCUSSION**

Developmental venous anomalies, formally known as venous angiomias, are the most frequent cerebral vascular anomalies, with a reported incidence between 0.48% (4,5) and 2.56% (6). The term developmental venous anomaly was suggested by Lasjaunias et al. (7), after postulating that venous angiomias are actually embryologic variants of venous drainage rather than real vascular anomalies.

The main suggested etiology for the formation of DVAs entails an embryologic accident that results in either arrested formation or thrombosis of the developing venous drainage of a specific region. This is followed by a secondary compensatory mechanism in which embryologic medullary venules persist and cluster in a large draining vein. The DVA collector then penetrates the cortex to drain either into cortical veins or sinuses or into subependymal veins and then into the deep venous system (8). On angiography, the region drained by a DVA has no other "normal" venous drainage system (9). This was also proven in patients who underwent surgical
FIG. 3. Axial T2-weighted image of the brain obtained 4.5 months after the onset of symptoms showing an area of complex cystic and solid components in the left frontoparietal region at the same location as the previously seen area of restricted diffusion, compatible with gliosis secondary to previous infarction.

ligation of DVAs and consequently developed massive venous infarcts in the corresponding territory (10).

Histologically, DVAs are characterized by a composition of thickened hyalinized veins with interspersed normal neural parenchyma (4).

On CT scan, they are rarely detectable without contrast administration. After contrast administration, they appear as linear areas of enhancement radiating either toward the ependymal surface of the ventricles or toward the venous sinuses. On MRI, they may show a characteristic flow void appearance on T1- and T2-weighted images, with normal nongliotic surrounding parenchyma. When small, they might not be detected on MRI. Administration of gadolinium further demonstrates the medullary veins and draining collector vein.

On cerebral angiography, DVAs are usually incidentally identified as deep medullary veins during the early or middle venous phase, accompanied by a single large draining vein.

There has been a slight controversy in the literature concerning the nature of these lesions. In a recent review by Topper et al. (4), DVAs are thought to be benign entities that are unlikely to become symptomatic. The authors evaluated a large series of patients and concluded that symptoms such as seizures, vertigo, syncope, tinnitus, and headache are not related to the venous anomaly itself. Intracerebral hemorrhage in their series only occurred in association with cavernous angiomas, which are known to be prone to bleeding (4). An association between venous angiomas and cavernous angiomas has been reported in the literature (11). Nevertheless, it remains unclear whether the hemorrhagic event is really caused by the DVA or is always associated with an occult cavernous angioma that may or may not be detected as a result of factors such as its size or the presence of large hematomas.

Some authors have suggested that hemorrhagic events associated with DVAs can be the result of thrombosis of the draining vein (12). This leads to increased pressure within the venous system and secondary hemorrhage (12). Hemorrhagic transformation is known to be a common complication of venous infarcts. Keiper et al. (13) detected subcortical hemorrhage in 9 of 24 (38%) subjects with cerebral venous thrombosis. Similarly, most of the previously reported complicated DVAs were those that presented with hemorrhagic complications (12,14).

Conversely, nonhemorrhagic ischemia associated with

FIG. 4. Areas of hyperintensity seen on axial fluid-attenuated inversion-recovery-weighted images of the brain 4.5 months after the onset of symptoms, compatible with changes of Wallerian degeneration (open black arrows).
DVA has been rarely reported in the literature. To our knowledge, there are no more than seven reported cases of nonhemorrhagic infarcts associated with DVAs (15–21). The suggested etiology of DVA-associated infarcts is thrombosis of the draining vein. This has been documented by means of angiography by Konan et al. (15) as an endoluminal clot inside the DVA collector in a patient who presented with a cerebellar infarct. The authors suggested that the frequently seen area of stenosis at the dural opening of a DVA, which was documented by Truwit (22) in 1992, might have a role in initiating the thrombotic event.

We believe that the same predisposing factors for dural sinus thrombosis in the central nervous system apply for the thrombotic event associated with the DVA in our patient. It has been demonstrated that the puerperium and oral contraceptive use predispose for dural sinus thrombosis (23,24). A recent study conducted by De Bruijn et al. (25) showed that women using oral contraceptives have a 13-fold risk of dural sinus thrombosis compared with age-matched control subjects, with the risk becoming 30-fold when those patients have an additional hereditary hypercoagulable state such as protein C or S deficiency, antithrombin deficiency, or factor V Leiden mutation (25). Our patient had a negative hypercoagulability screen but still had the risk factors of recent delivery, oral contraceptive use, and smoking, which we assume to have contributed to the thrombotic event involving the DVA.

CONCLUSION

Developmental venous anomalies are benign vascular anomalies that are usually incidentally discovered on imaging studies performed for unrelated problems. Although infrequent, hemorrhagic transformation and, much less frequently, ischemic complications have been reported. Resection or radiosurgery remains contraindicated, because these DVAs drain normal brain parenchyma.

REFERENCES