Case Report

Thrombosis of a Developmental Venous Anomaly in Inflammatory Bowel Disease: Case Report and Radiologic Follow-up

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Developmental venous anomalies (DVAs) are benign embryologic vascular variants, and before the advent of computed tomography and magnetic resonance imaging were supposed to be rare conditions. Usually, DVAs are asymptomatic and accidentally discovered during routine brain imaging studies, but sometimes they can be the cause of disabling neurologic symptoms. We describe the clinical and neuroradiologic follow-up of a 62-year-old man with a history of inflammatory bowel disease (IBD) presenting with new onset epilepsy and intracranial hemorrhage caused by thrombosis of a DVA who fully recovered after treatment with oral anticoagulant therapy. Patients with IBD have an increased risk of thrombosis because of inflammatory activity and the hypercoagulable state. Here we describe the first case of DVA thrombosis in a patient with IBD, and we show clinical and neuroradiologic follow-up after anticoagulant therapy. **Key Words:** Epilepsy—developmental venous anomaly—inflammatory bowel disease—intracranial hemorrhage—magnetic resonance imaging—stroke.

Developmental venous anomalies (DVAs) are presumably benign embryologic vascular variants caused by variation of the normal venous angioarchitecture, but their origin is still debated. DVAs are characterized by a cluster of venous radicles that converge into a collecting vein, resulting in the typical caput medusa appearance of the DVA.[1]

Some authors have proposed that DVAs result from a "hemodynamic need," leading to the recruitment of "transhemispheric anastomotic pathways," and therefore should be viewed as anatomic variations.[1,2] Other authors consider that thrombosis of normal parenchymal veins leads to the formation of DVAs,[1,3] and finally DVAs were suspected to be the result of disturbed fetal angiogenesis and regression.[4] DVAs are usually incidentally discovered on neuroimaging of the brain and occasionally associated with other vascular malformations, such as cavernous malformations (CMs), causing venous infarcts and hemorrhage.[5] Before the advent of computed tomographic (CT) and magnetic resonance imaging (MRI) scans, DVAs were considered rare, but now they are often observed accidentally.

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DVAs are benign vascular malformations, but seldom symptomatic DVAs have been described[6] due to thrombosis or occlusion.[7] The term cerebral DVA, coined by Lasjaunias et al,[2] is now widely used as a synonym for venous angioma, cerebral venous malformation, or cerebral venous medullary malformation. DVAs are encountered both in the pediatric and adult populations, with a slight predominance in males.[1,8] In the published literature, venous infarcts have been misdiagnosed as a tumor or demyelinating disease, sometimes biopsied,[9] and also surgical removed, causing catastrophic venous hemorrhage.[7] Moreover, therapy for DVA thrombosis is still debated.

Here we describe the case of a patient with thrombosis of DVA and show images from the onset of the symptoms until the resolution after starting anticoagulant treatment.

Case Report

A 62-year-old man presented with new onset focal involuntary clonic movements in his left leg. Clonic movements started on the left leg and spread to the left side of the body. Movements lasted a few minutes and appeared 3 times. At the onset of symptoms, neurologic examination revealed an alert patient with Jacksonian march on the left side of the body with clonic movements starting in the left leg and spreading to the left arm. He had a medical history of myocardial infarction 20 years earlier and a diagnosis of inflammatory bowel disease (IBD) done few months before the onset of neurological symptoms. He took acetylsalicylic acid 100 mg/day and mesalazine. A CT scan performed at the onset of symptoms was normal (Fig 1A). Seizures stopped after treatment with intravenous diazepam (10 mg) and antiepileptic therapy was started with carbamazepine (400 mg/day). MRI with contrast-enhanced magnetic resonance angiography (Fig 1B-1I) revealed a right DVA with a number of dilated medullary veins on the right hemisphere without the evidence of a collector draining vein. DWI was normal and gradient echo images revealed tiny cortical siderosis. An associated cavernoma was ruled out. An extensive coagulation and autoimmunity evaluation was performed and hereditary prothrombotic conditions, such as factor V Leiden mutation, prothrombin gene mutation 20210GA, antithrombin III, protein C, and protein S deficiency were ruled out.

The patient's condition worsened 4 days later. He had several seizures and developed right hemiplegia. A CT scan of the brain (Fig 2A) revealed a hemorrhage and edema in the right hemisphere in the same site of the venous anomaly. A new MRI scan of the brain revealed hemorrhage (Fig 2C and 2D) and vasogenic edema (Fig 2B). Digital subtraction angiography (DSA) revealed multiple tiny and serpiginous veins (Fig 2F and 2G; *arrows*) draining into right lateral ventricle veins, without a main collector vein. Low–molecular weight heparin (LMWH) and then oral anticoagulant therapy with

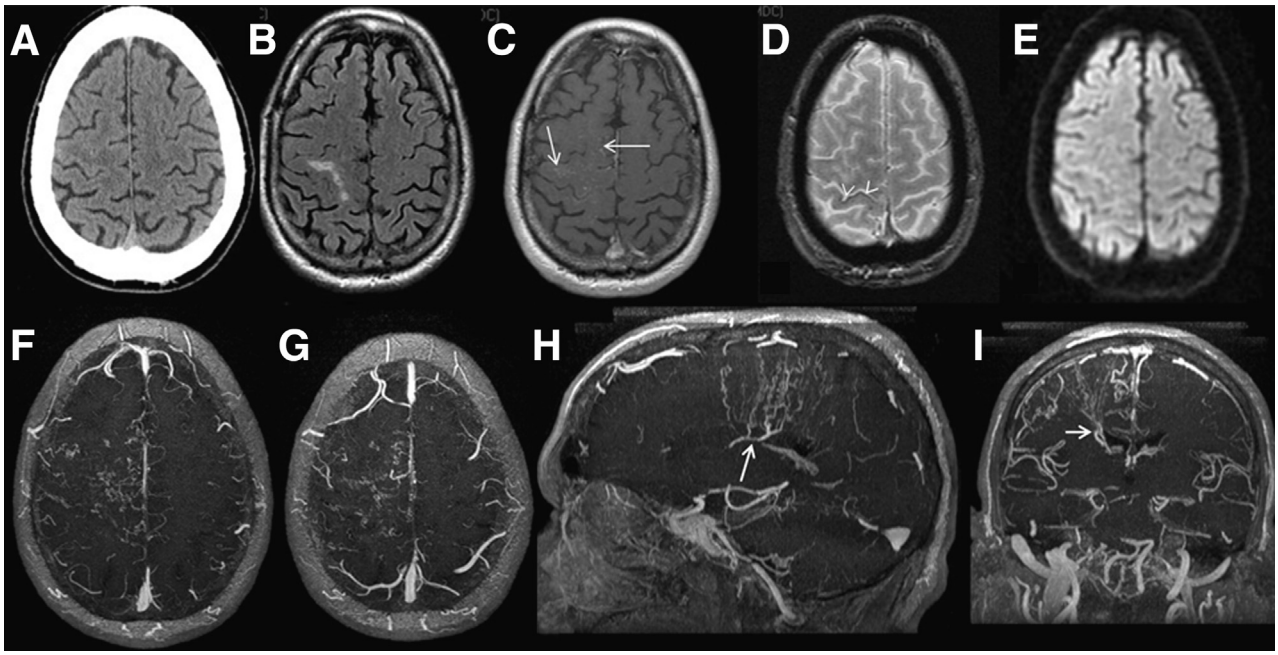

Figure 1. *An axial brain computed tomographic scan was normal (A), whereas magnetic resonance imaging (B-E) and magnetic resonance angiography with gadolinium (F-I) revealed a right frontoparietal venous anomaly. Fluid-attenuated inversion recovery imaging (B) revealed a signal intensity abnormality surrounding the developmental venous anomaly (DVA), and T_1-weighted imaging after gadolinium (C) revealed faint enhancement within the DVA drainage territory (arrows). Gradient echo imaging (D) revealed thin and subtle siderosis (arrowheads) and diffusion-weighted imaging (E) did not reveal areas of restricted diffusion. Brain magnetic resonance angiography axial (F-H), sagittal (H), and coronal (I) maximum intensity projection reformatted images revealed serpiginous medullary veins in the right frontoparietal lobes, draining into the deep circulation (right transmedullary veins; arrow in H and I) without evidence of a single collector vein. Few cortical veins of the right hemisphere drained into the superior sagittal sinus (H-I).*