Clinical/Scientific Notes

Sashank Prasad, MD
Robert W. Hurst, MD
Scott E. Kasner, MD

POSTPARTUM THROMBOSIS OF A DEVELOPMENTAL VENOUS ANOMALY

Developmental venous anomaly (DVA) is a common congenital vascular abnormality that typically has a benign natural history. The lesion likely occurs after atresia or thrombosis of normal venous structures, leading to compensatory retention of embryologic medullary venules.[1,2] These venules form an umbrella- shaped arrangement (caput medusae) and cluster into a large central vein that drains into the deep or superficial venous system.[1,2] Spontaneous thrombosis of the central vein of a DVA can rarely occur, and lead to symptomatic nonhemorrhagic venous infarction.[2-7]

Most reported cases of DVA thrombosis demonstrate no underlying predisposition.[2-6] One report of nonhemorrhagic venous infarction attributes thrombosis to hypercoagulability induced by puerperium, oral contraceptive use, and smoking.[7] We report a case of DVA thrombosis occurring in a postpartum patient on hormonal contraception.

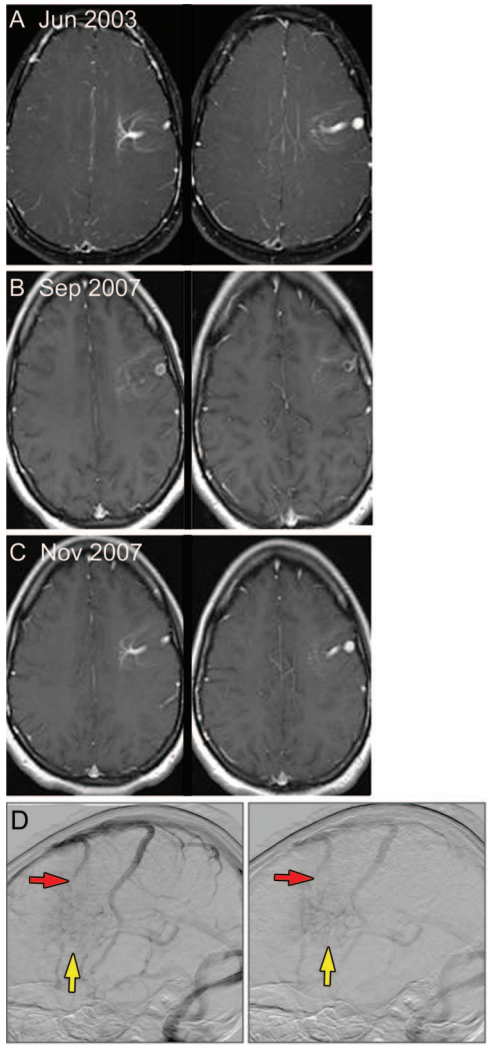
Case report. A 16-year-old G1P1 woman presented after three brief simple motor seizures involving the right arm and face occurring over 3 days. She denied persistent symptoms. After the first seizure, she had presented to another hospital and was treated with phenytoin, but did not undergo any neuroimaging studies.

Four years previously, she had three episodes of right face, arm, and leg shaking, lasting 2 minutes, without altered consciousness. A routine EEG revealed occipital slowing, but no epileptiform discharges. Brain MRI revealed multiple small vessels draining into an enlarged vein in the left frontal lobe, representing a venous angioma (figure, A). The patient was lost to follow-up, and no antiepileptic agents were started.

Three weeks prior, in the 40th week of pregnancy, she was presumptively diagnosed with mild preeclampsia on the basis of elevated blood pressure (146/96 mm Hg) but without proteinuria or edema. She had a 20-minute episode of right face and arm shaking with secondary generalization, which was presumed to be an eclamptic seizure. She was treated with IV magnesium and diazepam. A head CT was reportedly negative. She underwent emergent Cesarean section. She recovered fully, and no additional studies were performed.

The patient had no other medical illnesses, and there was no relevant family history. Immediately after her delivery, she had received the contraceptive

Figure MRI and angiography findings



Gadolinium-enhanced T1-weighted MRI revealed left frontal developmental venous anomaly (A, June 2003; B, September 2007, thrombosis within draining vein, with absent flow-void; C, November 2007, resolution of thrombosis). Conventional angiogram (D, left internal carotid artery injection, oblique view) revealed thrombosis of the draining vein (red arrow) of a DVA (yellow arrow). There is delayed emptying of the medullary veins into the late venous phase. See video on the *Neurology*® Web site at www.neurology.org.

Supplemental data at www.neurology.org

depot medroxyprogesterone acetate (DMPA). She was on no other medications. She denied smoking.

The entire examination was normal, including mental status, cranial nerves, strength, sensation, coordination, reflexes, and gait.

Noncontrast CT scan of the head revealed a region of hyperdensity within the left frontal lobe. Brain MRI revealed a partially thrombosed venous angioma with surrounding areas of swelling and edema (figure, B). Diffusion-weighted imaging revealed no evidence of restricted diffusion. Gradient echo sequences showed susceptibility artifact within the venous angioma and many of its periventricular branches. Postgadolinium images demonstrated dilated medullary veins and hyperintensity within an enlarged superficial draining vein. Digital subtraction angiography was performed, and injection of the left common carotid artery demonstrated a tangle of small vessels in the posterior left frontal lobe filling during the late venous phase, compatible with a venous angioma (figure, D; video). Emptying of the medullary veins was delayed, suggesting thrombosis of the major draining vein. No aneurysm or arteriovenous malformation was noted and the dural venous sinuses were patent.

Blood cell counts and coagulation profiles were normal. Protein C and S levels, antithrombin III level, and homocysteine level were normal. Activated protein C resistance was not present. Antinuclear antibody, anticardiolipin antibody, anti β2-glycoprotein antibody, factor V Leiden mutation, and prothrombin gene mutation were negative.

The patient was treated with warfarin (goal INR 2.0–3.0) and levetiracetam. At 3-month follow-up, she reported no further seizures. Repeat MRI and MRA demonstrated resolution of thrombosis within the DVA (figure, C). Warfarin was discontinued. At 6-month follow-up, levetiracetam was discontinued.

Discussion. Although DVAs are considered to be relatively benign, there is growing recognition that they may cause venous infarction or hemorrhage. Our patient demonstrates that thrombosis of a DVA may occur in the hypercoagulable state induced by puerperium and hormonal contraception, and possibly preeclampsia, which are risk factors for venous sinus thrombosis. Management of this condition should include evaluation for inherited thrombophilia and short-term anticoagulation. Given the relatively benign natural history of DVAs, the risks of hemorrhage during the course of short-term anticoagulation are expected to be minimal, while the benefits of anticoagulation for cerebral venous sinus thrombosis may be substantial. Surgical resection of a DVA is not recommended since the vessel may be the only venous drainage for the surrounding brain parenchyma.[1]

From the Departments of Neurology (S.P., S.E.K.) and Radiology (R.W.H.), Hospital of the University of Pennsylvania, Philadelphia.

Disclosure: The authors report no disclosures.

Received March 28, 2008. Accepted in final form July 3, 2008.

Address correspondence and reprint requests to Dr. Sashank Prasad, Department of Neurology, Hospital of the University of Pennsylvania, 3W Gates Bldg., 3600 Spruce Street, Philadelphia, PA 19104; sashank.prasad@uphs.upenn.edu

1. Agazzi S, Regli L, Uske A, Maeder P, de Tribolet N. Developmental venous anomaly with an arteriovenous shunt and a thrombotic complication: case report. J Neurosurg 2001;94:533–537.
2. Peltier J, Toussaint P, Desenclos C, Le Gars D, Deramond H. Cerebral venous angioma of the pons complicated by nonhemorrhagic infarction: case report. J Neurosurg 2004;101:690–693.
3. Lai PH, Chen PC, Pan HB, Yang CF. Venous infarction from a venous angioma occurring after thrombosis of a drainage vein. AJR Am J Roentgenol 1999;172:1698–1699.
4. Konan AV, Raymond J, Bourgouin P, Lesage J, Milot G, Roy D. Cerebellar infarct caused by spontaneous thrombosis of a developmental venous anomaly of the posterior fossa. AJNR Am J Neuroradiol 1999;20:256–258.
5. Masson C, Godefroy O, Leclerc X, Colombani JM, Leys D. Cerebral venous infarction following thrombosis of the draining vein of a venous angioma (developmental abnormality). Cerebrovasc Dis 2000;10:235–238.
6. Vieira Santos A, Saraiva P. Spontaneous isolated nonhaemorrhagic thrombosis in a child with development venous anomaly: case report and review of the literature. Childs Nerv Syst 2006;22:1631–1633.
7. Hammoud D, Beauchamp N, Wityk R, Yousem D. Ischemic complication of a cerebral developmental venous anomaly: case report and review of the literature. J Comput Assist Tomogr 2002;26:633–636.

M. Bitoun, PhD
J.A. Bevilacqua, MD, PhD
B. Eymard, MD, PhD
B. Prudhon, MSc
M. Fardeau, MD
P. Guicheney, PhD
N.B. Romero, MD, PhD

A NEW CENTRONUCLEAR MYOPATHY PHENOTYPE DUE TO A NOVEL DYNAMIN 2 MUTATION

Autosomal dominant centronuclear myopathy (CNM) is a rare congenital myopathy mostly characterized by delayed motor milestones, slowly progressive muscle weakness, and bilateral ptosis.[1] Mutations in the *DNM2* gene encoding dynamin 2 (DNM2), a large GTPase involved in membrane trafficking, have been identified in CNM.[2,3] Mutations in the middle domain of the protein are mostly associated with the slowly progressive mild late-onset CNM,[2] while mutations in the C-terminal part of the Pleckstrin homology (PH) domain cause a more severe neonatal phenotype.[3] In addition, mutations in the N-terminal part of PH domain have been reported in intermediate and axonal Charcot-Marie-Tooth disease (CMT).[4-6] Here, we report a novel