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Thrombosis of a Developmental Venous Anomaly with Hemorrhagic Venous Infarction

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A 17-year-old female presented with a 4-day history of severe left frontal headache, nausea, and vomiting that was worst in the morning. Neurologic examination revealed an alert, conversant teenager with a right facial droop and no other focal neurological signs. Medical history was significant only for occasional headaches and a 1-year history of oral contraceptive use.

Head CT showed a small amount of hemorrhage, edema, and a probable thrombosed venous structure in the left frontal lobe (Figure 1, panel A). MRI of the brain showed flow voids in the left frontal region consistent with a developmental venous anomaly (DVA), with surrounding edema (Figure 1, panel B), scattered T2 hypointensity consistent with areas of hemorrhage, and weakly positive diffusion-weighted imaging consistent with a small hemorrhagic venous infarction. There was also significant edema and mass effect. MRA was normal. MRV showed a partially thrombosed deep vein draining the left frontal lobe. Thrombosis of the DVA and absence of other vascular malformations was confirmed by digital subtraction angiography (DSA). The partially occlusive clot within the venous collector of the DVA is well delineated on the sagittal view of the post-contrast MRI and on the lateral projections of the angiogram. (Figure 2, panels A and B).

Comment

DVAs are fairly common and generally benign congenital vascular abnormalities, present in approximately 2.6% of the general population.¹ Thrombosis of a DVA is extremely rare, and most symptomatic complications arise from DVAs that are arterialized or associated with other vascular malformations, like cavernomas or arteriovenous malformations.¹ This patient had no associated vascular malformations, but she did have a 1-year history of oral contraceptive use and was also heterozygous for the Factor V Leiden R506Q mutation. It is likely that the combination of these thrombotic risk factors along with sluggish circulation in the DVA drainage system allowed the thrombus to form.

Oral contraceptives were discontinued. Anticoagulation was considered, but not given due to ICH and patient preference. Long-term aspirin therapy was recommended to prevent further

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thrombosis. The patient recovered well and at the 3-month follow-up visit had only flattening of her right nasolabial fold.

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Figure 1. Axial Head CT and MRI of the Brain showing Developmental Venous Anomaly Thrombosis

A, Non-contrast head CT showing a thrombosed DVA (arrow) with hemorrhagic infarction in the left frontal lobe. B, Axial T2-weighted MRI showing the typical appearance of a DVA in the left frontal region (arrow), with surrounding edema consistent with venous infarction.

Figure 2. Sagittal Contrast-Enhanced MRI of the Brain and Digital Subtraction Angiography confirming DVA Thrombosis

A, Sagittal MRI showing an intraluminal defect consistent with partially occlusive thrombosis of the main DVA collector (arrows). B, Digital subtraction angiography confirming subocclusive thrombosis of the DVA collector (arrows), delayed drainage with contrast stagnation in the DVA territory (caput medusae).