Atypical presentation of venous infarct

Developmental venous anomaly thrombosis propagating from clot in dural venous sinus

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Practical Implications

Consider venous ischemia in patients presenting with acute stroke-like symptoms, especially those with hypercoagulable states.

A 42-year-old woman with systemic lupus erythematosus (SLE) presented with right hemisensory loss and aphasia beginning 3 hours prior to arrival to the hospital. Two months prior to presentation, anticoagulation was started for a left transverse sinus to internal jugular thrombosis, which was diagnosed after development of a severe headache and vomiting (figure 1, A and C). There was an incidental finding of a patent left temporal developmental venous anomaly (DVA). The DVA drained the supramarginal and superior temporal gyri extending from the Sylvian fissure to the vein of Labbé (figure 1B).

At the current presentation, international normalized ratio was subtherapeutic and the patient was thrombocytopenic. CT and CT venogram images showed thrombosis of known left temporal DVA with associated surrounding venous congestion but without acute hemorrhage (figures 1, D–F, and 2). There was also thrombosis of the vein of Labbé (figure 2D) strongly suggestive of thrombus propagation from the transverse sinus to the DVA. MRI and magnetic resonance venogram confirmed venous congestion in the supramarginal and superior temporal gyri on susceptibility-weighted imaging (SWAN) (figure 2B) and demonstrated peripheral venous infarction on diffusion-weighted imaging (DWI) (figure 2C). During hospitalization, the patient's aphasia was intermittent and determined to be caused by cortical seizures. Her aphasia resolved with levetiracetam but right-sided sensory deficits persisted. Dilute Russell viper venom time ratio was 1.19, which is considered a negative test for lupus anticoagulant. Antiphospholipid antibodies were not elevated. The patient was discharged on therapeutic dose of warfarin.

DISCUSSION

Developmental venous anomalies are the most common cerebrovascular malformations.¹ They are characterized on cross-sectional imaging as an abnormal collection of dilated medullary veins with a prominent draining vein. They are commonly associated with cavernous

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Figure 1 Dural venous thrombosis propagating into developmental venous anomaly

(A–C) Presentation 2 months prior to current presentation. (A) Noncontrast CT demonstrates thrombosis of the left transverse sinus (white arrow). (B) CT venogram (CTV) demonstrates patent left temporal developmental venous anomaly (DVA) (green arrows). (C) CTV demonstrates nonocclusive thrombus in the left sigmoid sinus extending into the internal jugular vein (orange arrow). (D–F) Current presentation. (D) Noncontrast CT demonstrates thrombosis of the left temporal DVA (white arrow). (E) CTV now demonstrates partial occlusion of the distal DVA (yellow arrow). (F) Maximal intensity projection magnetic resonance venogram demonstrates thrombosis of the left transverse and sigmoid sinuses and of the internal jugular vein (blue arrows).

malformations.¹ The majority of DVAs are incidental findings, and symptomatic thrombosis of a DVA draining vein is a rare occurrence. Literature review identified 22 cases of DVA thrombosis.^{1,2} Of these, 19 were supratentorial and 3 were infratentorial. Due to the limited number of reported cases, predictive risk factors and treatment for DVA thrombosis have not been established. DVA thrombosis is presumed to have a clinical profile similar to that of dural sinus and cerebral venous thrombosis. Multiple factors predispose to venous thrombosis, including oral contraceptive use, thrombophilia, pregnancy, dehydration, protein C deficiency, and malignancy.^{1,3,4} Our patient's history of SLE and incompletely treated dural sinus thrombosis placed her at high risk for clot propagation. The most common symptoms of thrombosed DVA are headache, nausea/vomiting, seizures, and anatomic-specific focal neurologic deficit depending on the drainage pattern of the DVA.¹ Our patient's DVA was located in the superior temporal and supramarginal gyri, likely involving language pathways and serving as a focus for cortical seizures causing intermittent aphasia.

The pathophysiology of cerebral venous occlusion helps explain both clinical consequences and imaging findings of DVA thrombosis. On noncontrast CT, thrombus is identifiable as a tubular or crescentic hyperdensity. On MRI, thrombus demonstrates susceptibility on T2* and SWAN (figures 1D and 2B).^{1,4} Following contrast administration, a filling defect or lack of expected opacification of the affected vein should be present (figure 1E). Unlike arterial occlusion, venous occlusion results in development of venous hypertension due to insufficient drainage. This leads to cytotoxic, vasogenic, or more commonly mixed edema with or without associated venous infarct or intracranial hemorrhage.^{1,4} Contrast-enhanced CT may show parenchymal enhancement in the affected region (figure 2A). SWAN may identify venous congestion by demonstrating increase in capillary/venular deoxygenated blood (figure 2B). Over half (57%) of patients with venous thrombosis have parenchymal changes, most

Figure 2 CT venogram and MRI of thrombosed developmental venous anomaly

(A) CT venogram (CTV) demonstrates thrombosis of left temporal developmental venous anomaly (DVA) and enhancement of the left superior temporal and supramarginal gyri consistent with venostasis (white arrows). (B) Susceptibility-weighted MRI is a mirror image of the CTV (A), demonstrating susceptibility associated with thrombosed distal DVA (green arrow) and multiple foci of susceptibility in the brain parenchyma suggestive of venous stasis. (C) Diffusion-weighted MRI demonstrates restricted diffusion consistent with venous infarct (white arrows). (D) Contrast-enhanced T1-weighted MRI demonstrates partially thrombosed left vein of Labbé (orange arrows), a conduit for propagation of thrombus from the left transverse sinus into the DVA.

commonly vasogenic and cytotoxic edema, which can be distinguished on DWI.⁴ One-third of patients have parenchymal hemorrhage, which is more common in venous infarcts due to lack of smooth muscle in vein walls.^{1,2}

Current recommendations for treatment of DVA thrombosis are based on those for dural sinus thrombosis. In the absence of hemorrhage, systemic anticoagulation should be strongly considered to promote recanalization and to decrease propagation of the thrombus.^{1,3} In patients with severe intracranial hemorrhage or edema, decompressive craniotomy is sometimes performed.^{5,6}

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