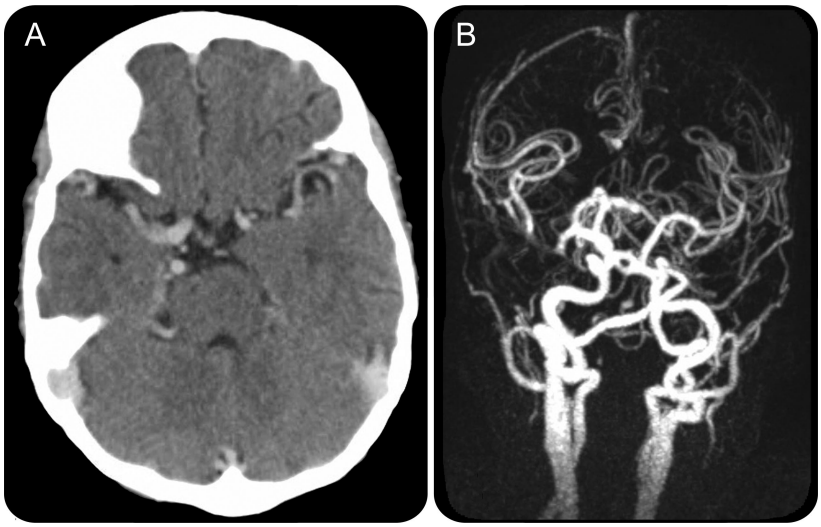
Cerebrovascular findings in an adult with cyanotic congenital heart disease

Figure Noncontrast head CT and magnetic resonance angiogram



(A) Noncontrast head CT showed markedly opacified intracranial vasculature secondary to hyperhemoglobinemia. (B) Magnetic resonance angiography demonstrated prominent hypervascularity with dilatation and ectasia of the cervical and cranial vasculature.

A 26-year-old man developed recurrent episodes of hemiparesis and dysarthria. He had cyanotic congenital heart disease (CCHD) with single-ventricle physiology. Chronic hypoxemia (baseline arterial oxygen saturation 70%) had caused secondary erythropoiesis; at this presentation, the hematocrit was 80% and the hemoglobin concentration exceeded 25 g/dL. Neuroimaging demonstrated marked hemoconcentration (figure, A) and hypervascularity (figure, B). Similar morphologic changes are observed in coronary arteries of adults with CCHD, with associated medial wall abnormalities including loss of smooth muscle, increased collagen, and duplication of internal elastic laminae.[1] This patient's symptoms were likely due to blood hyperviscosity, as he improved with hemodilution.

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