Imaging Sciences Interesting Cases

CASE 13

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CLINICAL PRESENTATION: A 14-year-old male with one week history of right upper extremity numbness and new onset seizures.

IMAGING FINDINGS:

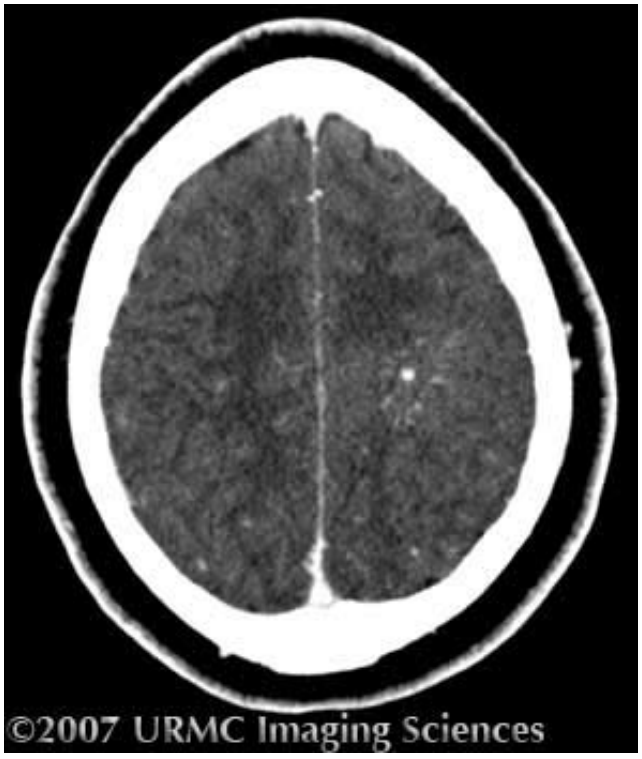

Figure 1: Contrast-enhanced CT demonstrates large central draining vein surrounded by small medullary venules in a spoke-wheel pattern in the left frontal lobe.

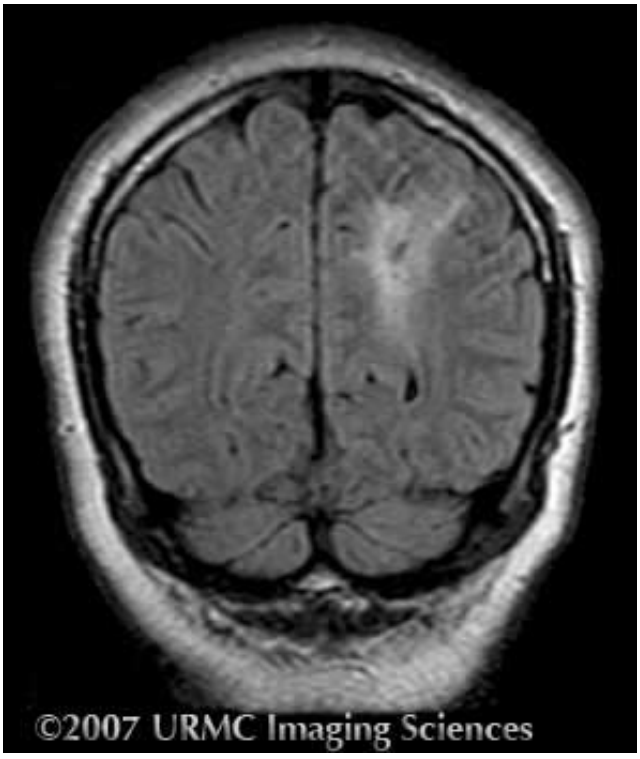

Figure 2: Coronal FLAIR image reveals signal void within central vein with surrounding edema.

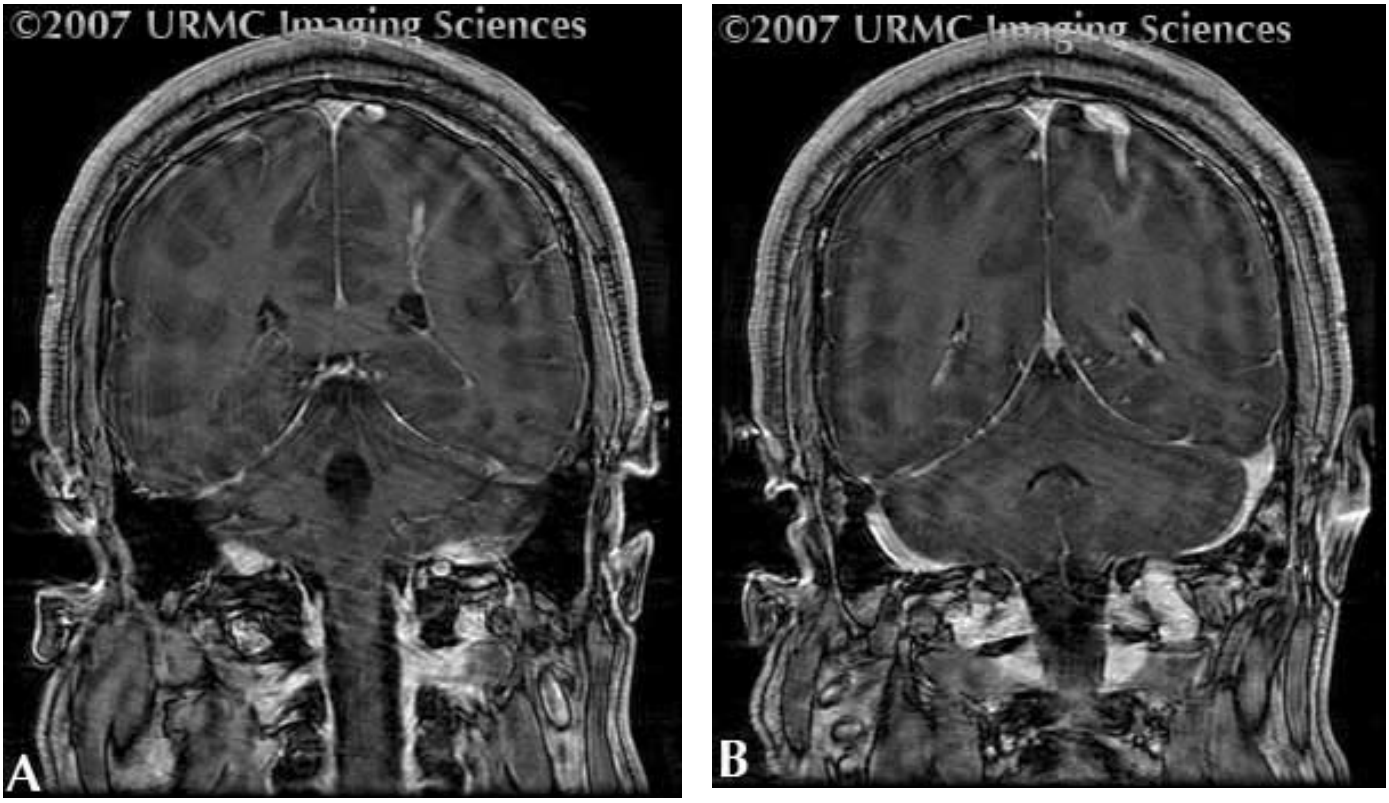

Figure 3. Two post-contrast coronal SPGR images demonstrate central draining vein emptying into superficial cortical vein, which ultimately drains into the superior sagittal sinus (not shown).

DIAGNOSIS: Developmental Venous Anomaly

DISCUSSION: Venous angiomas, also known as developmental venous anomalies (DVA) or venous malformations, are congenital anatomical variants of venous drainage within a region of the brain. They are the most common vascular anomaly within the brain and can be found in up to 2% of the population. Because this vascular anomaly does not entail abnormally formed vessels, the term developmental venous anomaly has been advocated.

DVA consists of a network of small medullary venules which converge into a central draining vein and empty into a superficial or deep cortical vein, depending on the location of the anomaly. This anomaly does not involve arteries. DVA forms as an alternate drainage pathway to the dural sinuses when normal venous drainage is absent. They are often found adjacent to the lateral ventricles, but may occur anywhere within the central neuraxis including the spine. The frontal-parietal lobes are the most common locations, followed by the cerebellum. Normal brain tissue can be found in between veins. Fifteen to thirty percent of DVA are associated with another cerebrovascular anomaly such as an arteriovenous malformation and capillary telangiectasia. An association also exists between DVA and head and neck vascular anomalies as well as hemangiomas.

DVA are typically clinically silent and diagnosed incidentally. The causal relationship between uncomplicated DVA and clinical symptoms is controversial. Hemorrhage is rare, but may occur in the presence of increased flow within the medullary veins with subsequent rupture. Spontaneous thrombosis has also been reported and may result in edema, ischemia or venous infarct.

DVA may be detected by contrast-enhanced CT, contrast-enhanced MR or angiography. The typical appearance consists of multiple small veins with the appearance of a spoke-wheel converging into a large central draining vein, which ultimately empties into a superficial cortical vein, subependymal ventricular vein or dural sinus. On non-contrast MR, a flow void within a large central vein may be seen. On long TR sequences, the slow flow within the central vein may result in high signal intensity secondary to gradient

moment nulling. If the vessel is oblique, the high signal intensity within the vessel is misregistered, and an adjacent low signal intensity is also noted. GRE sequences should also be performed to detect other vascular malformations that may be present. DVA will appear hypointense on GRE sequences secondary to deoxyhemoglobin within venous blood. Angiography demonstrates DVA during normal venous phase and persists through the late venous phase.

DVA are not treated, but may be followed up with MR if hemorrhage or thrombosis with associated edema or ischemia is detected. Careful distinction from arteriovenous malformations should be made as embolization or surgical removal of a DVA may result in catastrophic morbidity.

REFERENCES:

1. Lee C, Pennington MA, Kenney III CM. MR evaluation of developmental venous anomalies: Medullary venous anatomy of venous angiomas. AJNR Am J Neuroradiol. 1996 Jan;17(1):61-70. [PubMed]
2. Atlas SW, Do HM. Intracranial vascular malformations and aneurysms. In: Atlas SW. Magentic Resonance Imaging of the Brain and Spine. Lippincott Williams and Wilkins, Philadelphia, 2002. pg 875-879.