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Developmental Venous Anomaly Associated with Hemi-Parkinson's Syndrome

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Disclosures can be found in Additional Information at the end of the article

Abstract

Objective and Importance: Developmental venous anomalies (DVAs) are common congenital anomalies of intracranial venous drainage. In general, it is felt that DVAs replace the normal venous drainage system and, unless associated with another lesion such as a cavernous malformation, should not produce symptoms. The present case raises important questions regarding the potential for DVAs to become symptomatic and the role of venous hypertension in this process.

Clinical Presentation: We present the case of a 23-year-old woman who presented with rapidly progressive hemi-parkinsonism consisting of unilateral tremor, micrographia, gait difficulty, and cogwheel rigidity. MRI demonstrated a large DVA draining the region of the contralateral basal ganglia with associated high signal intensity on T2-weighted MR imaging and gadolinium enhancement. Arteriography demonstrated only the DVA with the trunk of the angioma draining forward toward the inferior petrosal sinus instead of into the deep venous system. A focal stricture of the main venous trunk was noted. Retrograde venography confirmed a tight stricture at the entrance of the venous trunk into the cavernous sinus.

Conclusions: Although DVAs are generally considered benign variants of normal venous drainage, it is possible that in select cases, venous outflow restriction may result in symptomatic venous hypertension. This may be particularly problematic in the setting of a large DVA when multiple radicular veins drain into a single large venous channel.

Categories: Neurosurgery
Keywords: parkinson's disease, cerebral vascular malformation, developmental venous anomaly, venous angioma, venous hypertension

Introduction

Developmental venous anomalies (DVAs) are congenital anomalies of intracranial venous drainage that replace the normal draining veins of the surrounding brain [1, 4]. They have been found in infants and children, and it has been postulated that they may result from intrauterine occlusion or maldevelopment of normal medullary veins [4]. We describe the case of a patient with a DVA that formed the venous drainage of the basal ganglia. The patient presented with progressive hemi-parkinsonism,possibly related to venous hypertension. To our knowledge, this is one of the first reports of such a sequence of events. This case suggests the potential importance of venous outflow restriction in the development of symptoms in a DVA.

Case Presentation

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This 23-year-old right-handed woman presented with three month history of progressively worsening right-sided tremor, loss of right hand dexterity and walking difficulty. Neurological examination revealed the unilateral tremor, micrographia, cogwheel rigidity, and gait difficulty. Magnetic resonance imaging (MRI) scan demonstrated the large DVA with contrast enhancement and abnormal signal change within the associated basal ganglia (Figure *1*). T2-weighted imaging revealed signal change compatible with edema in the basal ganglia. Diffusion-weighted imaging showed no true ischemia but did reveal abnormal signal hyperintensity presumably related to the significant edema.

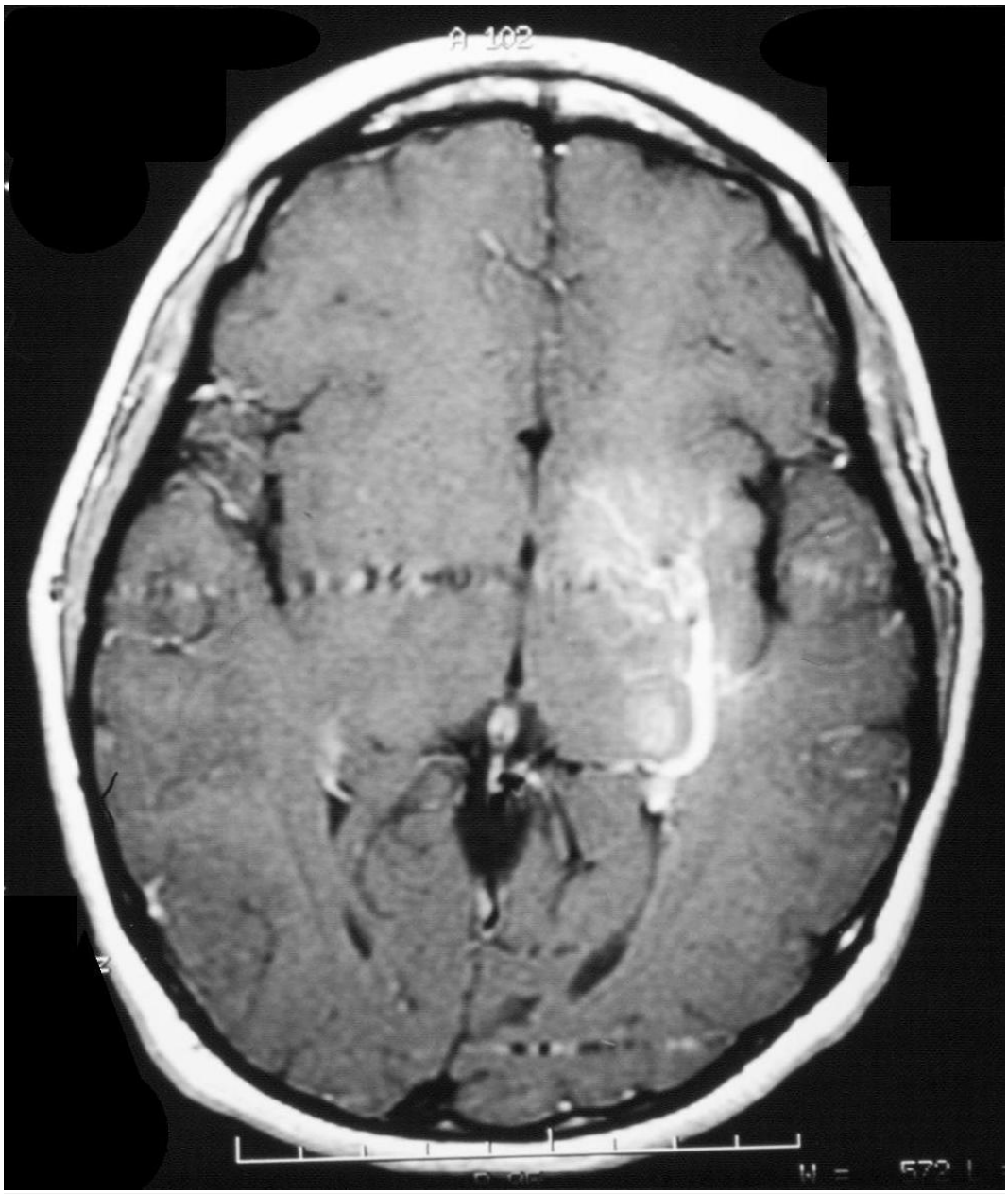

FIGURE 1: Axial, contrast-enhanced, T1-weighted MR image shows the large DVA with associated contrast enhancement and abnormal signal change within the surrounding parenchymal tissue.

Cerebral angiography confirmed the large DVA and also showed a stenosis involving the large venous trunk representing the primary outflow of the DVA (Figure *2*).

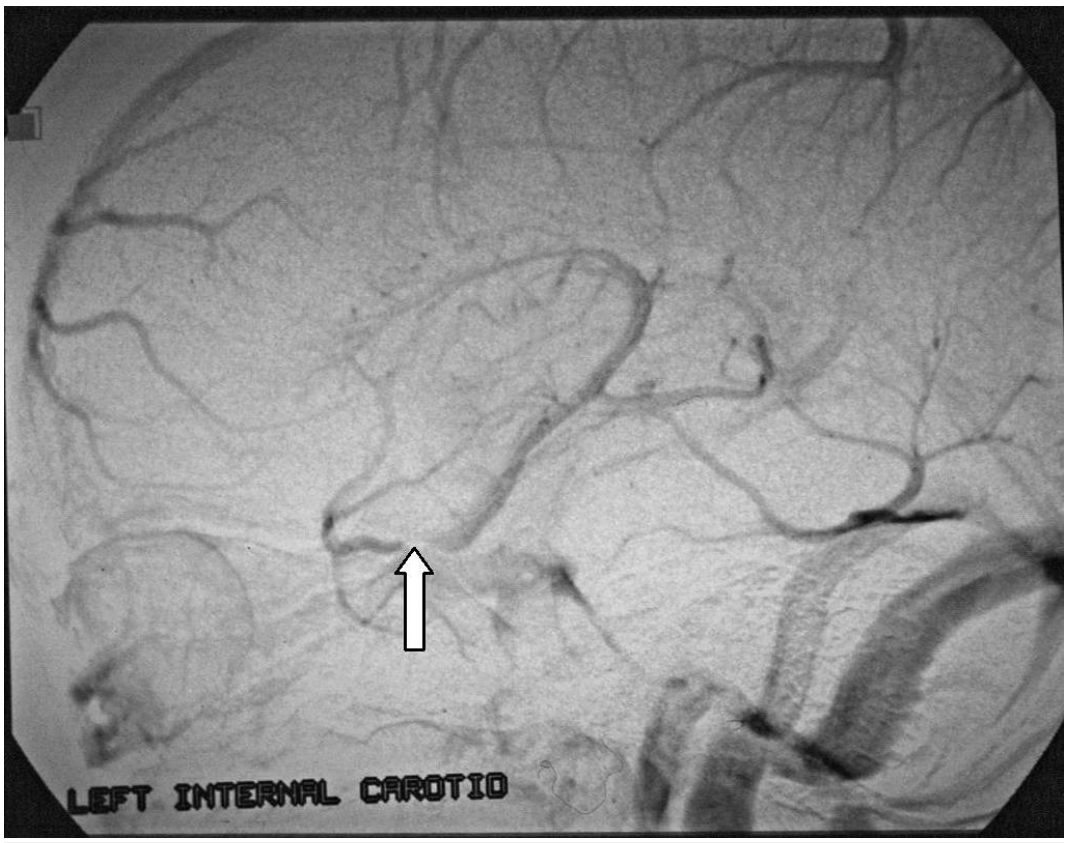

FIGURE 2: Lateral venous phase angiographic injection of the left internal carotid artery demonstrates the large caput type appearance of a DVA.

Lateral venous phase angiographic injection of the left internal carotid artery demonstrates the large caput type appearance of a DVA with stenosis (arrow) involving the primary venous outflow of the DVA

The possibility of balloon dilatation of the venous stricture was considered, but a catheter could not be advanced across the narrowing. The patient was managed with dopamine and responded well with a significant improvement in her symptoms. In addition, she was maintained on aspirin at 325 mg per day. At two year follow-up, she remained stable neurologically and then relocated from the area when she was lost to follow-up.

Discussion

The DVA represents a congenital malformation composed entirely of veins with normal intervening neural parenchyma in which a network of small medullary veins converges into a large central venous channel that drains into the superficial or deep venous system [1, 4]. This configuration results in the classic “caput medusa” appearance on angiography. Prior to the development of high resolution MR imaging, it had been felt that DVA’s could result in bleeding, and some surgeons aggressively removed DVA’s in the setting of hemorrhage, particularly in the posterior fossa. Over time, it became clear that these DVA’s were almost always associated with a cavernous malformation that represented the true source of bleeding in these cases [1]. In general, surgery is reserved for those cases in which an associated cavernous malformation can be identified as the source of bleeding. Because the DVA represents a normal and often critical venous drainage pathway for blood flow exiting the brain, direct surgical treatment aimed at excising or partially removing a DVA is contraindicated.

The notion that a DVA can produce symptoms by itself is controversial, but isolated reports have described either obstructive hydrocephalus, venous infarction, hemorrhage, or other symptoms

arising from a DVA without another associated vascular abnormality [3, 5-15]. Based on our case and these other isolated reports, it would appear that stenosis or occlusion of one of the venous radicles or the main trunk of the lesion could result in venous hypertension or ischemia. In fact, the abnormal drainage pattern might predispose to such a situation if the main draining trunk were compromised given the potential lack of normal collateral venous drainage channels.

The documented venous outflow limitation associated with abnormal MR signal surrounding the DVA in our case further supports the likely relationship between the DVA and the development of basal ganglia symptoms in our patient. It is possible that over time, individuals with similar venous outflow limitation will develop new venous collateral pathways which could result in symptomatic improvement.

Conclusions

Although DVAs are traditionally felt to be benign entities, the clinical picture and imaging studies in this case suggest that DVAs may become symptomatic. We postulate an impaired venous outflow resulting in “venous ischemia” as the pathophysiology.

Additional Information

Disclosures

Human subjects: Consent was obtained by all participants in this study.

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