

Development of a Posterior Fossa Cavernous Malformation Associated With Bilateral Venous Anomalies: Case Report

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ABSTRACT

Venous angiomas (VAs) and cavernous malformations (CMs) are common cerebrovascular malformations. Frequently, these lesions are found in close proximity. The interrelationship between VAs and CMs has not yet been adequately defined. The authors report a case of a 48-year-old man with progressive dysarthria, dysmetria, and ataxia. Eight years previously, magnetic resonance imaging (MRI) revealed a solitary CM and bilateral posterior fossa VAs. Later imaging after neurological progression revealed the presence of 2 rather than 1 CM adjacent to the VAs. The sequential imaging suggests a causal relationship between VAs and some CMs. Furthermore, the detailed MRI permitted radiosurgical treatment of these CMs. The occurrence of de novo CMs adjacent to VAs on future imaging studies in other patients may help confirm the etiology of at least a subset of CMs.

Key Words: Cavernous malformation, venous angiomas, radiosurgery.

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The association between cavernous malformations (CMs) and venous angiomas (VAs) is not uncommon.¹ The etiology of these vascular malformations and their interrelationship, however, remain the subject of much debate.²⁻⁵ Magnetic resonance imaging (MRI) has become an important tool for identifying these lesions, shedding light on the behavior and possible etiology of their frequent association and making radiosurgical targeting more precise.

Case Presentation

The patient was a 48-year-old right-handed man with progressive gait and speech difficulties. The symptoms dated back to 1993. At that time, an MRI study including T1, T2, and gradient echo sequences was obtained using an MRI with a 1.5-T field strength and subsequently reviewed at that time by both a neuroradiologist and a neurosurgeon. This MRI revealed a solitary pontomesencephalic CM in close proximity to bilateral VAs. He was originally told that this was a "congenital lesion" and would not require any further attention. Of note, the patient had no significant past medical or family history, and he had no prior surgeries.

Over the next 7 years, the patient had progression of his dysarthria, right arm dysmetria, and gait ataxia. MRI now revealed 2 CMs: 1 located in the right superior aspect of the pons, at the pontomesencephalic junction (Fig 1), and the other in the right dentate region of the cerebellum (Fig 2). MRI also revealed evidence of bilateral venous anomalies in the cerebellar hemispheres (Figs 3, 4).

After discussing various options with the patient, including further observation with repeat imaging, surgical resection, and radiosurgery, the decision was made to treat both of the CMs with gamma knife radiosurgery.^{6,7} We did not recommend additional observation because of the progressive nature of the patient's neurological symptoms as well as the chance for a hemorrhage that could result in neurological devastation. Surgical resection of both CMs would have been fraught with difficulties, including brainstem injury, cranial nerve palsies, worsening cerebellar dysfunction, and venous infarct as a result of damage to the adjacent VAs. During stereotactic planning, the anomalous venous drainage was excluded from the radiosurgical field. At 6 months after radiosurgery, the patient has improved articulation and right arm coordination.

Discussion

Venous anomalies are known to be the most common intracranial vascular malformation in both autopsy and clinical series.¹ VAs are believed to result from abnormal venous embryogenesis and are sometimes referred to as developmental anomalies.² CMs were also believed to be congenital in nature.⁵ In recent years, an association between CMs and VAs has often been reported.^{2,8,9} There appears to be an increased tendency for hemorrhage in CMs associated with venous anomalies as opposed to CMs alone.^{8,9} Some have postulated that higher pressure due to restricted venous disease may cause hemorrhage from a VA with subsequent formation of CMs.^{2,3,8,9} One case of CM development was postulated to have occurred as a result of

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Fig 1. Axial magnetic resonance imaging (TR = 21, TE = 7 with gadolinium) revealing a cavernous malformation within the superior aspect of the pons at the pontomesencephalic junction and bilateral venous anomalies.

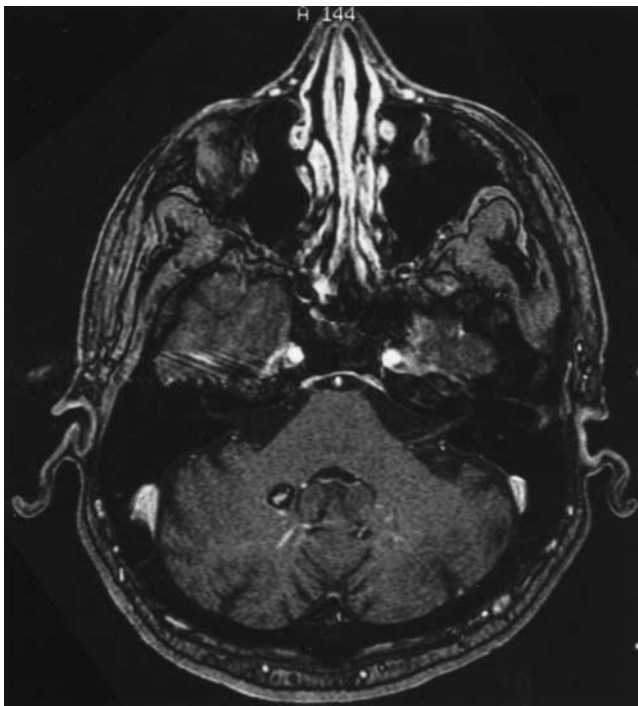


Fig 2. Axial magnetic resonance imaging (TR = 21, TE = 7 with gadolinium) depicting the deep cerebellar cavernous malformation not seen on imaging 7 years previously.

increased venous pressure induced by radiation therapy.⁸ Little et al¹⁰ demonstrated communication of CMs with the venous circulation and thereby highlighted the close relationship be-

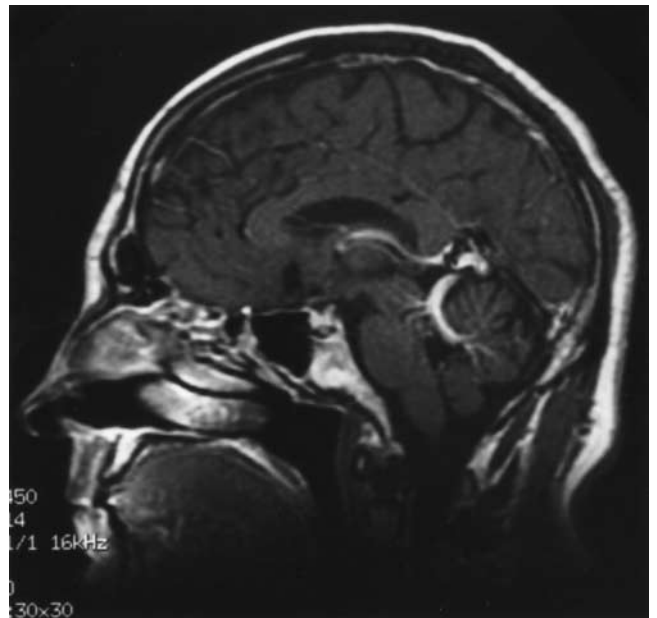


Fig 3. Sagittal magnetic resonance imaging (TR = 450, TE = 14 with gadolinium) illustrating the midbrain cavernous malformation in close proximity to the venous anomalies.

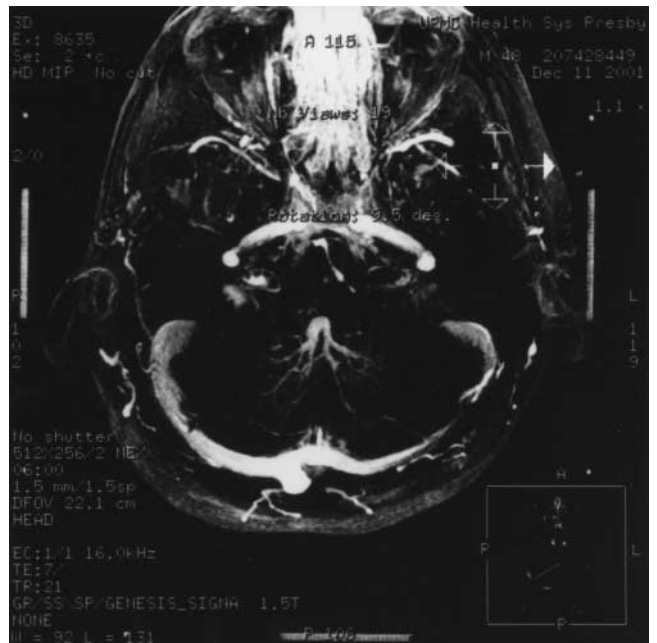


Fig 4. Axial magnetic resonance venogram reconstruction (TR = 21, TE = 7) depicting both the extensive bilateral cerebellar venous anomalies and the adjacent cavernous malformation.

tween these vascular malformations. Comey et al¹¹ noted parenchymal enhancement associated with combined cavernous and venous malformations and postulated that venous hypertension and blood brain barrier disruption are associated with selected venous malformations.

Others have theorized that VAs may facilitate formation of CMs via angiogenic growth factor release and red blood cell

diapedesis.⁴ Pluripotential cells may also be present in zones of active angiogenesis and demonstrate neoproliferation into the walls of cavernomas.^{2,12,13} The development of a second CM on follow-up imaging within the drainage region of bilateral VAs supports the notion that some CMs are acquired vascular lesions. This is the first case report to illustrate progressive CM occurrence adjacent to bilateral posterior fossa VAs. The intimate relationship depicted on MRI between the bilateral venous anomalies and CMs certainly suggests that the presence of one type of malformation could have led to the formation of the other. Although these images do not provide definitive proof of a causal relationship between VA-induced venous hypertension and the de novo formation of CMs, we believe that they provide an imaging correlate to the likely microscopic and biochemical factors linking these two types of cerebrovascular malformations. The fact that at least some CMs may be acquired lesions highlights the need for further investigation into the development and behavior of these vascular malformations.

With modern neuroimaging, many VAs and CMs are incidentally discovered. However, patients can frequently present with headaches, seizures, disequilibrium, subarachnoid hemorrhage, and parenchymal hemorrhage. The annual risk of hemorrhage for these lesions is less than 1%.^{5,6} As such, surgical intervention is generally reserved for patients with progressive neurological deficit or recurrent hemorrhage. Surgical resection of CMs can be curative. For those CMs not amenable to a conventional surgical approach, stereotactic radiosurgery has been shown to reduce the risk of hemorrhage.^{6,7,14,15} In addition to protecting against hemorrhage, radiosurgery also appears to shrink the lesion and reduce the incidence of seizures associated with some supratentorial CMs.^{7,14} When treated with radiosurgery, the venous anomalies are left outside of the radiosurgical field.

The presence of 2 CMs in close proximity to bilateral posterior fossa VAs hints at the complex interrelationship between these cerebrovascular malformations. Future imaging correlates may help further elucidate the etiology of these lesions. Conventional angiography is typically unable to adequately depict these vascular malformations. The ability of MRI and the relatively recent adaptations of magnetic resonance venography to delineate these lesions have further accentuated these imaging modalities. Moreover, detailed MRI facilitates the effective treatment of these lesions with radiosurgery.

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