Intracranial arterialized venous angioma: case report with new insights from functional brain MRI

Nestor D. Tomycz, Nirav A. Vora, Emanuel Kanal, Michael B. Horowitz, Tudor G. Jovin

ABSTRACT
We present the case of a 58-year-old man who suffered a left thalamic intracerebral hemorrhage. Brain magnetic resonance imaging (MRI) revealed an incidental venous angioma in the left frontal lobe. Further elucidated by cerebral angiography and functional MRI, this venous angioma exhibited arteriovenous shunting. The arterialized venous angioma represents an uncommon, “mixed” intracranial vascular lesion whose natural history remains unknown.

Key words: functional MRI • central nervous system venous angioma • vascular malformations

Most congenital intracranial vascular malformations fall into one of four groups: arteriovenous malformations (AVMs), capillary telangiectasias, cavernous malformations, and venous angiomas or developmental venous anomalies. Yet, some intracranial vascular lesions fall outside of this classification and have been dubbed “mixed” vascular malformations since they possess features of more than one type of classic vascular malformation; the hybrid venous angioma-cavernous malformation, venous angioma-AVM, cavernous malformation-AVM, and cavernous malformation-capillary telangiectasia have all been described (1).

The typical venous angioma is composed of a large parent vein that receives an array of radially-oriented tributary veins in a spoke-wheel configuration dubbed “the caput medusa”. Prospective studies on venous angiomas have demonstrated a very low rate of both symptomatic hemorrhage (0.34% per year) and neurologic symptoms; bleeding, when it rarely occurs, has been hypothetically blamed on putative neighboring cavernous malformations (2). Blood flow through venous angiomas is low and they are thought to drain normal brain. In contrast, the AVM is a true malformation formed from a nidus of tangled vessels in which high flow and pressure result from a direct communication between the arterial and venous systems.

The mixed vascular lesion which combines features of an AVM and a venous angioma has been called the venous angioma with arteriovenous shunt or arterialized venous angioma (1, 3). This vascular chimera has the architecture of a venous angioma and lacks an AVM-like nidus, but displays arteriovenous shunting during arterial phase angiography (3, 4). The rare coexistence of an AVM draining into a venous angioma has been described and this coupling may be the pathogenic successor of the arterialized venous angioma (5, 6).

Case report
A 58-year-old man developed acute right hemiparesis and right face numbness. Blood pressure upon presentation was 147/80 mmHg. His past medical history was significant for former intravenous heroin abuse, hepatitis A, and chronic hepatitis C. Noncontrast head computed tomography (CT) revealed a small acute hemorrhage (volume 5–10 mL) in the left thalamus with extension into the internal capsule. Laboratory studies were significant for a low platelet count of 49,000/mm³ and liver function tests were mildly elevated. Magnetic resonance imaging (MRI) of the brain demonstrated a vascular lesion in the left frontal lobe which satisfied all the imaging features characteristic of a venous angioma (Fig. 1). Formal cerebral angiography found an enlarged callosomarginal artery whose branches formed this atypical vascular structure in the left parasagittal frontal lobe. The lesion displayed the...
radial venous morphology of a venous angioma but arteriovenous shunting was appreciated via conspicuous early draining veins on digital subtraction angiographic imaging (Fig. 2). Early venous drainage was superficial through dilated cortical veins that emptied into the superior sagittal sinus. Utilizing a right-finger tapping regimen during functional MRI (fMRI), the left medial frontal lobe region harboring this arterialized venous angioma, in addition to the ipsilateral right cerebellum and left motor cortex, displayed blood oxygen level dependent (BOLD) signal activation (Fig. 3). The final diagnosis was intracerebral hemorrhage secondary to hypertension and thrombocytopenia with an incidental arterialized venous angioma. His right hemiparesis improved and he was transferred to rehabilitation on hospital day 4.

Discussion

AVMs may be symptomatic secondary to mass effect but the vascular steal phenomena, venous hypertension, associated aneurysms, and hemorrhage risk are at least partially attributable to their intrinsic arteriovenous shunting (7). Thus, one might predict that arteriovenous shunting may render a venous angioma less benign. Moreover, based on experience from the natural history of intracranial dural arteriovenous fistulas, one might suspect that the enlarged cortical venous drainage associated with this arterialized venous angioma

Figure 1. Axial T1-weighted MRI of the brain with gadolinium contrast reveals a left frontal parasagittal vascular malformation that resembles a venous angioma.

Figure 2. a, b. Cerebral angiography with contrast injection into the left internal carotid artery reveals through early (a) and late (b) arterial phases an enlarged left callosomarginal artery feeding a parasagittal vascular malformation with venous angioma morphology. Arteriovenous shunting is manifested by early venous drainage into dilated cortical vein tributaries of the superior sagittal sinus.

Figure 3. Functional MRI utilizing a right-finger tapping regimen reveals blood oxygen level dependent (BOLD) signal activation involving the brain region containing the arterialized venous angioma.
may lead to higher rates of hemorrhage in comparison to classic venous angiomas (8). Limited and conflicting data currently exist as to whether the risk of hemorrhage for a venous angioma is increased in the presence of arteriovenous shunting (3, 4, 9).

At a basic level, the presence of fMRI activation signal in the region of this arterialized venous angioma reminds us that intracranial venous anomalies play a role in normal physiological venous drainage and do not render the surrounding brain non-functioning. Moreover, that this lesion was found amongst zones of cortical activation on fMRI suggests that, unlike AVMS, arterialized venous angiomas do not seem to exclude normal brain. Similar to high flow vascular malformations such as AVMS, which have been associated with cortical reorganization of eloquent cortex as well as displacement of functional cortical regions independent of mass effect, arterialized venous angiomas, by virtue of possessing arteriovenous shunting, may influence brain function via hemodynamic perturbation (10, 11). Even if arteriovenous shunting is inefficient and potentially pathogenic, one must recognize that an arterialized venous angioma may exist interspersed within eloquent cortical regions as demonstrated by functional imaging in this patient.

The current mainstay treatment for a venous angioma is reassurance. Most physicians accept that they carry a very low risk of hemorrhage, seizure, and neurologic deficit (12). Few patients with venous angiomas go on to have formal cerebral angiography, so the prevalence of associated arteriovenous shunting is unknown. More common angiographic evaluation and advances in non-invasive dynamic neuroimaging may help physicians to characterize subtypes of venous angioma which, although indistinguishable by MRI, carry higher risks and warrant either closer follow-up or perhaps treatment. Although surgery for venous angiomas carries significant morbidity, less invasive endovascular technology may allow one to safely obliterate potentially dangerous arteriovenous shunting within arterialized venous angiomas while preserving normal venous drainage. The task at hand is to identify which, if any, venous angiomas merit treatment secondary to potentially morbid characteristics such as arteriovenous shunting or the presence of an accompanying vascular lesion with higher hemorrhagic risk.

References