Development of a cerebral arteriovenous malformation documented in an adult by serial angiography

Case report

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This 61-year-old man presented in 1989 with the acute onset of vertigo and vomiting. On examination, the patient had mild dysmetria of the right arm and nystagmus on lateral gaze bilaterally. No evidence of intracranial hemorrhage was noted on CT scans. Examination of the cerebrospinal fluid revealed no subarachnoid hemorrhage or xanthochromia. Magnetic resonance imaging revealed flow voids in the region of the right tentorium associated with abnormal signal in the right cerebellar hemisphere. An angiogram demonstrated a right-sided tentorial dural arteriovenous fistula (DAVF) (Fig. 1). This fistula was supplied primarily by branches of the right occipital, ascending pharyngeal, and internal maxillary arteries, and by multiple small transsaccular branches of the internal carotid arteries bilaterally. Distal branches of the right superior and anterior inferior cerebellar arteries also contributed to the DAVF. Venous drainage was into the vein of Galen, the right transverse sinus, and inferiorly into the sigmoid sinus. Transverse and sigmoid sinuses were patent bilaterally. No cortical venous drainage was present.

The patient’s symptoms resolved over the next 2 weeks. Because of his clinical improvement and the unclear association between the patient’s symptoms and the DAVF, he was treated conservatively. The patient remained asymptomatic for 5 years. Follow-up MR images and angiograms obtained at that time revealed a venous varix that had developed in this interval in the posterolateral aspect of the DAVF. Because this was believed to increase his risk of intracranial hemorrhage,2 the patient was referred for neurosurgical evaluation. After discussing microsurgical resection or staged radiosurgery with transarterial embolization, the patient opted for the latter treatment strategy.19,28 Stereotactic radiosurgery was performed using a combination of stereotactic MR imaging and cerebral angiography to visualize the fistula. Five isocenters of radiation were used to cover a prescription isodose volume of 2.8 cm³ (Fig. 1). The radiation dose to the margin was 20 Gy, and the maximum radiation dose was 40 Gy. Transarterial embolization of the right external carotid artery supplying the malformation was then performed with no complications.

The patient remained asymptomatic, and a follow-up angiogram was obtained 2 years after radiosurgery (Fig. 2). The right-sided tentorial DAVF was slightly smaller. However, a parenchymal AVM was seen in the superior cerebellar vermis that had not been seen on the earlier imaging studies. The AVM received its arterial supply from the left SCA. Venous drainage was via a vein to the junction of the vein of Galen and the straight sinus. In light of the patient’s age and asymptomatic state, no additional intervention was recommended.

Two years later (4 years after radiosurgery), the patient had another severe episode of vertigo and nausea that re-
solved over several days. There was no evidence of subarachnoid hemorrhage on CT scans. However, a lumbar puncture was not performed at that time. An angiographic study of the cerebral area revealed complete obliteration of the right-sided tentorial DAVF (Fig. 3). The AVM in the superior cerebellar vermis had increased in size. Its arterial supply at this point was from the SCAs bilaterally. The patient chose repeated radiosurgery of the cerebellar AVM after being informed of the potential risks and benefits of observation, surgical resection, or radiosurgery. He remains asymptomatic 3 months after radiosurgery.

**Discussion**

Cerebral AVMs are believed to be congenital lesions. Although the exact mechanism of AVM development remains to be elucidated, ontological investigation of the cerebral vasculature has supported this hypothesis. In the 4th week of development, the primordial vascular plexus within the embryonic brain differentiates into arterial, venous, and capillary components. Artiovenous malformations result from abnormal persistence of direct arterial-to-venous connections without an intervening capillary bed. Dural-based AVFs are pathologically distinct from AVMs. In DAVFs, the abnormal arteriovenous connections lie completely within the leaves of the dura. Typically, these are acquired lesions, occurring as a result of trauma, surgical procedure, or venous sinus thrombosis. The lesion discovered in our patient has the angiographic characteristics of a true parenchymal AVM. First, the lesion is located within the cerebellar parenchyma. Second, the AVM is supplied by superior vermian and other smaller parenchymal arteries, not dura-based arteries as would be expected for a DAVF.

Formation of an AVM in an adult has not been described previously. Two possibilities exist for the discovery of the AVM in our patient: it may have formed de novo in the interval between the two angiograms, or it may have been present initially but was small enough to elude detection on neuroimaging. If the lesion did indeed form de novo, an alternative mechanism for AVM development must exist in addition to the accepted embryological mechanism. In children, AVM formation and enlargement is likely an ongoing process within the developing brain. This hypothesis is based largely on reports that surgically excised AVMs in children in whom resolution of the lesion is seen on postoperative angiograms may later demonstrate recurrence and enlargement. However, postoperative recurrence has also been reported in adults after presumed complete excision. It can also be postulated that radiation exposure from radiosurgery of the dAVF might play a role in this case. Larson, et al., have described cerebral cavernous malformation development after radiation treatment of children. In their report, six young patients aged 9 to 19 years were discovered to have cavernous malformations within irradiated brain at a median of 68 months after radiation therapy. We believe that radiation is unlikely to have been a causative agent in the case presented here because of the short latency between radiosurgery and discovery of the AVM (24 months), and the extremely low radiation dose to that region (< 2 Gy).

The most likely mechanism for the de novo development of the AVM in this case is local venous hypertension secondary to the adjacent DAVF. In addition to the possible role of local angiogenic factors, chronic venous hypertension may have dilated the venous end of parenchymal capillaries, ultimately causing ectatic dilation of the
entire capillary network and precipitating direct arteriovenous connections. This is analogous to the formation of mucosal AVMs in the gastrointestinal tract, which are thought to occur by a similar mechanism. It has been suggested previously that both cerebral AVMs and DAVFs form as a result of congenital or acquired abnormalities of venous drainage. In our patient, it is possible that abnormalities of the local venous drainage may underlie the development of both lesions.

The second possibility for the delayed discovery of the AVM is that the lesion was in fact present at the time of the initial neuroimaging but was undetected. With modern neuroimaging procedures, the lesion would have had to be smaller than 1 cm to have been below the resolution of MR imaging (Fig. 4). However, because the MR images and the angiograms were of high quality, and the appropriate vessels were studied for each angiogram, this explanation seems unlikely. Nonetheless, if indeed the AVM had been present from the outset but was small, what then caused its dramatic enlargement and detection? Hemodynamic factors may underlie the enlargement of existing AVMs. The cause of the enlargement of the AVM in this case could have been the decrease in size and blood flow of the nearby DAVF after the initial stereotactic radiosurgery and embolization. As the blood flow across the DAVF decreased, the arteriovenous pressure gradient across the AVM may have consequently increased, thereby causing increased blood flow through the AVM and its enlargement.

Although AVMs may fail to be visualized in either MR imaging or conventional angiographic studies, no AVM has been reported to be occult to both imaging modalities. Arteriovenous malformations that have hemorrhaged may not be visualized on angiograms as a result of compression from the hematoma and/or thrombus within the lesion. These AVMs may attain spontaneous obliteration or may reexpand and be visualized on subsequent angiograms. Some AVMs that have not bled but that result in seizures, mass effect, or headache can on rare occasions be undetected in angiographic studies. In the case of Hashim, et al., reviewed 50 reported cases of such AVMs, in which the lesions were not detected on angiograms but were demonstrated on CT or pathological examination. In no case did an occult AVM later appear in follow-up angiographic studies.

Conclusions

In this report we document the development of a cerebral AVM in an adult by serial angiography. Although the vast majority of AVMs in the cerebrum are congenital in origin, some patients may develop parenchymal lesions as a result of local factors such as venous hypertension. It is likely that this is a rare occurrence, and should not significantly affect our thinking regarding the natural history of AVMs.
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References
30. Sabin F: Preliminary note on the differentiation of angioblasts and the method by which they produce blood-vessels, blood-plasma and red blood-cells as seen in the living chick. Anat Rec 13:199–204, 1918

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