Pure arterial malformation of the posterior cerebral artery: importance of its recognition

Case report

NANCY MCLAU GLIN, M.D., PH.D.,1 RADO SLAV RAYCHEV, M.D.,2 GARY DUCKWILER, M.D.,2 AND NEIL A. MARTIN, M.D.1

Departments of 1Neurosurgery and 2Radiology, Neurointerventional Radiology, David Geffen School of Medicine at UCLA, Los Angeles, California

The finding of dilated, elongated, and tortuous vessels on brain imaging should prompt clinicians to determine what vascular anomaly is present. Importantly, not all suspicious serpentine flow voids are manifestations of arteriovenous malformations or arteriovenous fistulas. Other types of intracranial vasculopathies should also be considered. The authors report a rare case of dilated, tortuous, and redundant left posterior communicating artery and left P2 segment of the posterior cerebral artery identified in a young healthy adult that remained stable over a 30-year period. Dynamic and 3D images were critical for determining the type of vascular anomaly and for guiding appropriate management. The authors propose that this case represents a pure arterial malformation and discuss its distinguishing features.

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KEY WORDS • arterial malformation • developmental anomaly • dilatative arteriopathy • dolichoectasia • cerebral artery ectasia • posterior cerebral artery • cerebral aneurysm • vascular disorders

Patients are referred to neurosurgeons specialized in cerebrovascular pathologies following the identification of dilated vessels on brain CT scans, CT angiograms, MR images, or MR angiograms. Not all suspicious serpentine flow voids are manifestations of arteriovenous malformations or arteriovenous fistulas. Other entities that should be considered are purely arterial anomalies or malformations (dolichoectasia, fusiform aneurysm, dissections) or purely venous anomalies or malformations (venous varix, developmental venous anomaly, or venous angiomas). However, differentiating between arteries and veins on these static 2D imaging modalities is difficult and often impossible. Dynamic imaging modalities such as a catheter angiogram and 3D images such as those obtained with 3D angiography may be essential to determine the type of vascular anomaly or malformation present and, ultimately, to guide management.

We report a rare case of dilated, elongated, and redundant left PCoA and left P2 segment of the PCA that remained stable over 3 decades, based on detailed radiographic documentation (reports and images), in an otherwise healthy individual. The catheter angiogram and the 3D images were critical in characterizing this vascular malformation. We propose that this case represents a pure arterial malformation, emphasizing the importance of clinical history and appropriate imaging with long-term follow-up for safe counseling and management.

Case Report

History. A 54-year-old woman in good health was
referred to our clinic for evaluation of a possible arteriovenous malformation. At age 24, she had undergone brain imaging prompted by the development of more frequent headaches. Head CT scanning and catheter angiography showed the presence of abnormal vessels in the suprasellar and preponticular cisterns, interpreted as an arteriovenous malformation. No treatment had been recommended. At age 37, she was brought to the emergency department after suffering an acute episode of dizziness that spontaneously resolved. Brain MRI at the time revealed a small cystic pituitary lesion and again showed the abnormal vessels in the same region. She was also discovered to be pregnant at this visit, and regular imaging follow-up was recommended for a supposed Rathke cleft cyst. Recently, while gardening, the patient sprayed an insecticide product in her face and eyes. She later developed diplopia, unsteadiness, and myalgia. Symptoms improved within the following 7 hours and were completely resolved within 24 hours. She did not note any headaches throughout this episode. Brain MRI and MRA showed again the infracentimetric pituitary cystic lesion, as well as the abnormal vessels in the left suprasellar and preponticular cisterns (Fig. 1). The catheter angiogram showed a dilated, tortuous, and redundant left PCoA and left P₂ segment of the PCA. The redundant vascular loops formed a compact purely arterial mass. A 6-second frame acquisition allowed following the tortuous and redundant loops of the involved PCoA and P₂ arteries (Fig. 3). A small wide-necked aneurysm at the origin of the left PCoA measuring 2.6 mm (dome) and 3.4 mm (neck) was visualized. Reviewing her most recent MRA images acquired 3 months before the angiograms, although this aneurysm was already present, it was very difficult to separate from all vessel loop superpositions. It had not changed in size and morphology. The vascular anomaly was consistent with a dilated, elongated, and redundant left PCoA and P₂ segment, which, based on the detailed radiographic description of the prior angiogram, had not changed in appearance for 3 decades. The absence of a nidus and early venous drainage were noted.

**Management.** Given the circumstances in which the recent symptoms occurred, it was not believed that these symptoms were related to the arterial anomaly. No treatment was recommended for this vascular anomaly. The patient was, however, counseled to undergo a follow-up MRA in 6 months to reevaluate the small PCoA aneurysm. If this examination documents stability of the left PCoA aneurysm, catheter angiography will be recommended in 2 years to evaluate for an interval aneurysm change (size, morphology, or both) or for a new aneurysmal formation that could potentially be obscured by the vessel loop superposition.

**Discussion**

**Distinction From Arteriovenous Malformations or Fistulas**

Accurate characterization of the architecture of abnormal vessels is essential prior to discussing any avenues of management. Appropriate characterization of the vasculopathy is important because treatment and prognosis vary depending on the type of the vascular anomaly. Arteriovenous malformations represent abnormal connections between arteries and veins that would irrigate and drain normal brain, respectively. This transition can occur through an intervening network, a nidus, or be a direct process, termed fistulous arteriovenous malformation. If the abnormal vessels are extraparenchymal, the abnormal communication may form directly between pial arteries and pial veins, termed a pial arteriovenous fistula.

![Fig. 1. Serial sagittal T1-weighted MR images obtained in 2005 (A), 2008 (B), and 2011 (C).](image-url)
or directly between dural arteries and dural veins, termed a dural arteriovenous fistula. Recognizing these two vascular abnormalities and detailing their angioarchitecture is important, as associated findings may increase the risk of hemorrhage or of treatment. The absence of a nidus and a venous component makes this reported case distinct from cases of arteriovenous malformations or fistulas.

**Distinction From Dilatative Arteriopathy**

However, abnormal vessels visualized with a brain CT, CTA, MRI, or MRA do not always imply abnormal arterial to venous connections (Fig. 5). Indeed, dilated vessels seen with these imaging modalities can be purely arterial or purely venous. The most common entity presenting as elongated, tortuous, and dilated arteries at the base of the brain is an intracranial dilatative arteriopathy (dolichoectasia). This acquired subtype is more common, affecting adults over 40 years of age, and is most often diagnosed in the 6th and 7th decades of life. It most commonly involves the vertebrobasilar artery and ICA. Recent studies have proposed that the acquired subtype of dilatative arteriopathy may be part of a systemic ectatic vasculopathy as larger diameters in thoracic aortas and coronary arteries have been noted in these patients. Congenital intracranial dilatative arteriopathy is less frequent, affects children and young adults, and frequently involves branches of the ICA. Genetic, infectious, inflammatory, immunological, and degenerative factors may all cause or contribute to the formation and/or progression of dilatative arteriopathy.

The present case was distinct from dilatative arteriopathy (dolichoectasia), which presents as a dilated and elongated vessel along the course of a recognizable artery (Fig. 5B). In pure arterial malformations, in addition to being overlapping, dilated, and tortuous, the involved vessels are redundant to the point of looking like a mass of arterial loops with a coil-like appearance. In addition, although children, adolescents, and young adults diagnosed with dilatative arteriopathy generally have genetic, infectious, inflammatory, immunological, or degenerative predisposing factors that may cause or contribute to the formation and/or progression of dilatative arteriopathy, these were all absent in our patient.

**Distinction From Developmental Arterial Anomaly (or Arterial Vascular Dysplasia)**

Similar to developmental venous anomalies, there have been few reports of cases of arterial vascular dysplasia or of developmental arterial anomaly. Reported developmental arterial anomalies most often involve distal branches of intracranial vessels and present as a network-like cluster of small dilated and ectatic arteries.
They appear to some extent as collections of small dilated veins clustered in one area around a larger vein characteristic of developmental venous anomalies. Dysembryogenesis may occur not only in intracranial veins but also in intracranial arteries, which may be linked to structural defects in the arterial wall and migrational abnormalities. Magnetic resonance images should be reviewed attentively to search for a possible associated cortical dysplasia, which has been described for developmental vascular anomalies. The configuration of pure arterial malformation differs from developmental arterial anomalies, with the former involving more proximal dilated vessels featuring redundant arterial loops forming a compact mass of overlapping vessels instead of a network-like cluster of small distal vessels. Unlike in other reported cases of developmental arterial anomaly, no cortical anomaly was found in this patient.

**Distinction From Intracranial Arterial Dissections**

The lesion in the present case was distinct from an intracranial dissection because the walls of the dilated vessel remained parallel along the entire course of the tortuous segment. The dilation and tortuosity not only involved the left PCoA but also extended beyond the junction of the PCoA with the PCA to involve the P2 segment. Both of these features are not typically seen in intracranial dissec-
Over the past 25 years, the senior author (N.A.M.) has encountered 2 other cases of developmental arterial anomalies compatible with purely arterial malformations. One patient was referred to our institution following a subarachnoid hemorrhage. Investigation revealed a distal ICA aneurysm with what was thought to be an arteriovenous malformation. Careful review of the angiogram showed a tangle of dilated arteries with no draining veins. Although the aneurysm and the tangle of arteries were in close proximity, it was not felt that both were related. The patient was brought to surgery for clipping of the ruptured suprachainoid ICA aneurysm. During the procedure, a mass of redundant arteries originating from the middle cerebral artery/lenticulostriates with macroscopically normal appearing vessel walls (no evidence of atherosclerosis) was observed. The second case was a patient referred for evaluation of a possible arteriovenous malformation involving the M1 segment. The dilated and tortuous middle cerebral artery showed no venous component. This patient was not brought to the operating room and was managed conservatively.

In summary, recognizing the presence of abnormally dilated vessels on images from brain CT, CTA, MRI, or MRA should initiate a differential diagnosis, including rarer entities such as purely arterial and purely venous anomalies. As illustrated in the present case, dynamic and 3D imaging may be required to determine the nature of the vascular anomaly and to guide appropriate management. The diagnosis of a purely arterial malformation should be raised in the presence of dilated, overlapping, and tortuous arteries forming a mass of arterial loops with a coil-like appearance in the absence of any venous component.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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