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Endovascular treatment of a ruptured posterior fossa pure arterial malformation: illustrative case

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BACKGROUND Pure arterial malformations (PAMs) are rare vascular anomalies that are commonly mistaken for other vascular malformations. Because of their purported benign natural history, PAMs are often conservatively managed. The authors report the case of a ruptured PAM leading to subarachnoid hemorrhage (SAH) with intraventricular extension that was treated endovascularly.

OBSERVATIONS A 38-year-old man presented with a 1-day history of headaches and nausea. A computed tomography scan demonstrated diffuse SAH with intraventricular extension, and angiography revealed a right posterior inferior cerebellar artery–associated PAM. The PAM was treated with endovascular Onyx embolization.

LESSONS To the authors' knowledge, only 2 other cases of SAH associated with PAM have been reported. In those 2 cases, surgical clipping was pursued for definitive treatment. Here, the authors report the first case of a ruptured PAM treated using an endovascular approach, showing its feasibility as a treatment option particularly in patients in whom open surgery is too high a risk.

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KEYWORDS pure arterial malformation; subarachnoid hemorrhage; endovascular; posterior fossa

Pure arterial malformations (PAMs) are poorly understood intracranial vascular entities that are commonly mistaken for arteriovenous malformations (AVMs), aneurysms, or arteriovenous fistulas (AVFs).[1–6] They are distinguishable by their characteristic dilated and tortuous coil-like appearance without associated early venous drainage on digital subtraction angiography (DSA).[5–7] A small percentage of them may also be associated with an aneurysm.[8] Although their etiology and natural history are not well established, these entities have been considered benign and managed conservatively.[5,6,9] However, recent publications show that PAMs can be progressive and may not be as benign as originally thought, with the potential to rupture.[6,7,10–12]

We describe the case of a patient who presented with symptomatic diffuse subarachnoid hemorrhage (SAH) with intraventricular extension secondary to a PAM originating from the right posterior inferior cerebellar artery (PICA), which was successfully treated with Onyx embolization (eV3 Covidien). This case underscores the potential of PAMs to rupture and demonstrates that endovascular intervention is an effective option for treatment.

Illustrative Case

A 38-year-old African American nonsmoking man with recently diagnosed hypertension (not receiving medications) presented with a 1-day history of progressive headaches and nausea in the absence of any recent trauma or falls. He had no known family history of aneurysms or other vascular abnormalities. On arrival to the emergency department, his initial blood pressure was 173/116 mm Hg. His neurological examination was notable for mild confusion but was otherwise nonfocal with a Glasgow Coma Scale score of 15. Computed tomography demonstrated a spontaneous SAH with intraventricular extension, classified as a Hunt-Hess grade (HH)1 and modified Fisher grade 4 (Fig. 1). Subsequent computed tomography angiography (CTA) revealed an abnormal tangle of vessels in the right

ABBREVIATIONS AVF = arteriovenous fistula; AVM = arteriovenous malformation; CTA = computed tomography angiography; DSA = digital subtraction angiography; EVD = external ventricular drain; HH = Hunt-Hess; ICU = intensive care unit; MRI = magnetic resonance imaging; PAM = pure arterial malformation; PBD = post-bleed day; PICA = posterior inferior cerebellar artery; SAH = subarachnoid hemorrhage; SOFIA = soft torqueable catheter optimized for intracranial access.

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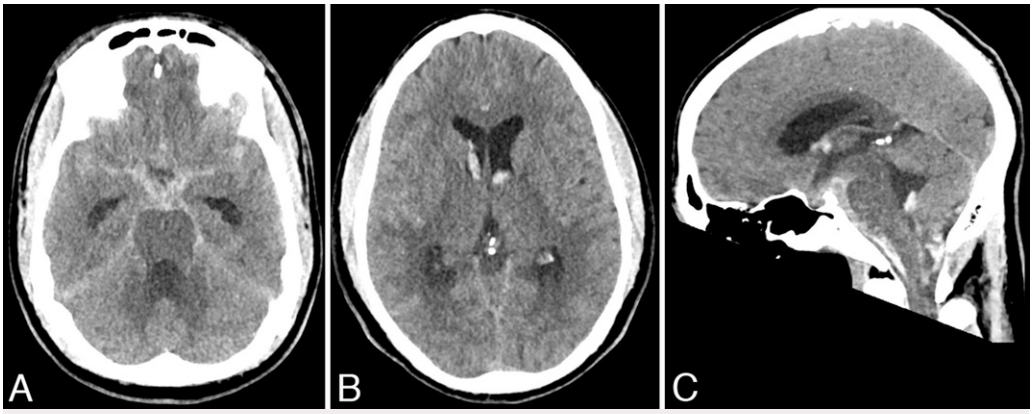

FIG. 1. CT images, axial (**A**, **B**) and sagittal (**C**) views, demonstrating diffuse SAH and intraventricular hemorrhage.

cervicomedullary junction, initially concerning for an AVM originating from the right PICA (Fig. 2).

On transfer from the emergency room to the intensive care unit (ICU), he became progressively somnolent (HH4) and hypertensive to 190/110 mm Hg, concerning for intracranial hypertension. A right frontal external ventricular drain (EVD) was placed, with an opening pressure >30 cm H_2O. Improvement in wakefulness was observed after EVD placement; the patient, however, remained disoriented and incapable of following commands. The patient subsequently underwent cerebral angiography, which revealed a dilated coil-like artery branching off the right PICA without associated early venous drainage, confirming the diagnosis of PAM (Fig. 3).

Endovascular Embolization Course

Intraarterial access was obtained by catheterization of the right common femoral artery with an 8-French sheath. A Neuron MAX (Penumbra, Inc.) catheter was positioned in the right subclavian artery, and the diagnostic catheter and glidewire were exchanged for a SOFIA (soft torqueable catheter optimized for intracranial access, MicroVention) intermediate catheter. The SOFIA catheter was positioned in the right vertebral artery, and an Echelon 10 (Covidien) microcatheter was used to specifically select the PAM branching off the right PICA. Quantities of 0.4 mL of dimethyl sulfoxide and 0.5 mL of ethylene vinyl alcohol copolymer (Onyx) were injected over 5 minutes, and successful embolization was confirmed by selective angiography of the right vertebral artery (Fig. 4).

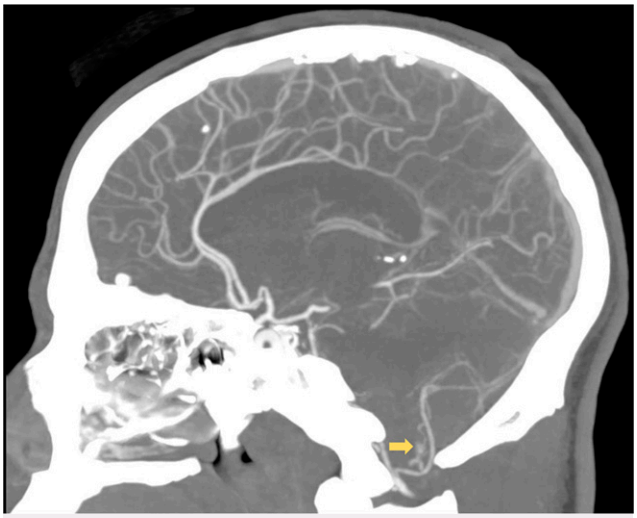

FIG. 2. CTA, sagittal view, demonstrating an abnormal tangle of vessels in the right cervicomedullary junction (*arrow*).

Postoperatively, the patient was transferred back to the ICU in stable condition. He was found to have bilateral lower extremity weakness, with abnormal T2 prolongation at the dorsal cervicomedullary junction and upper cervical cord visualized by magnetic resonance imaging (MRI), concerning for a subacute stroke (Fig. 5). This may have occurred because of Onyx propagation to a lateral medullary artery arising from the right PICA that was not observed on the CTA scan or the angiogram. The patient's postoperative course was complicated by paroxysmal sympathetic hyperactivity secondary to SAH-associated dysautonomia on postbleed day 18 (PBD 18), which gradually resolved. His ICU stay was also complicated by aspiration pneumonia and sepsis causing an acute respiratory distress syndrome and leading to a tracheostomy and percutaneous endogastric tube placement to facilitate his ICU management. No vasospasm was detected on multiple CTA scans and transcranial Doppler ultrasound studies throughout his ICU admission. He was transferred out of the ICU on PBD 32 and discharged to an inpatient rehabilitation facility on PBD 40. His neurological examination findings continued to improve throughout his admission and at the time of discharge revealed an alert and oriented man with no cranial nerve palsies, 4/5 strength in his right upper and bilateral lower extremities, and 3/5 strength in his left upper extremity. At the time of writing this article, he continues to be in rehabilitation, where he is receiving physical, occupational, and speech therapy. The patient currently is able to ambulate with assistance, with full strength in his right upper and bilateral lower extremities and 4/5 strength in his left upper extremity. He has not presented with additional episodes of bleeding. A follow-up cerebral angiogram obtained at 3 months from the initial insult demonstrated complete obliteration of the PAM without evidence of recurrence or additional intracranial abnormalities.

Discussion

PAMs were first characterized by McLaughlin et al. in 2013.[3] They provided a framework for differentiating PAMs from other entities. This included (1) lack of venous involvement, separating them from AVMs or

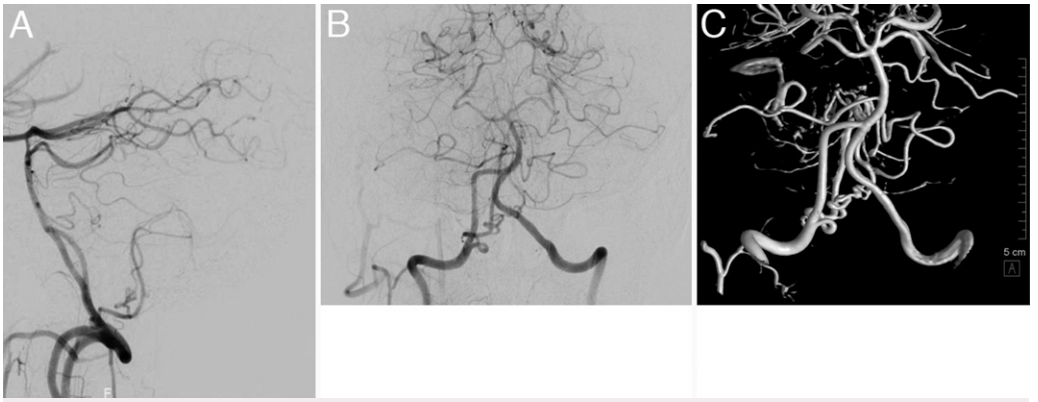

FIG. 3. Cerebral angiography with right vertebral injection, lateral (**A**) and posterior-anterior views (**B**, **C**), demonstrating a dilated "coil-like" artery branching off the right PICA with no associated early venous drainage.

AVFs; (2) location (i.e., proximal); and (3) absence of cortical dysplasia. These latter 2 criteria differentiate PAMs from arterial dysplasia, which occurs more distally and in the presence of cortical dysplasia. Differences can be more subtle between dolichoectasia and PAMs, where the key differentiating factor is the "coil-like" serpentine appearance of PAMs in contrast to the elongated appearance of dolichoectasias. PAMs can also be associated with local aneurysms.[3,6,8,11,13,14] Although there is no accepted preferential vascular territory, the anterior cerebral artery and posterior cerebral artery branches are the most commonly reported.[8] Altogether, selective cerebral angiography is the most important diagnostic tool in differentiating PAMs from other vascular anomalies (particularly AVMs), as it demonstrates a lack of arteriovenous shunting.[3–5] Despite this, PAMs can still be difficult to identify purely on the basis of imaging. Our institution has previously reported 2 cases presenting with symptomatic SAH and radiographic appearances of PAMs on DSA, with a dilated coil-like appearance and no evidence of early venous drainage; however, both cases were confirmed to be aneurysmal ruptures in surgery.[11,15] This highlights that

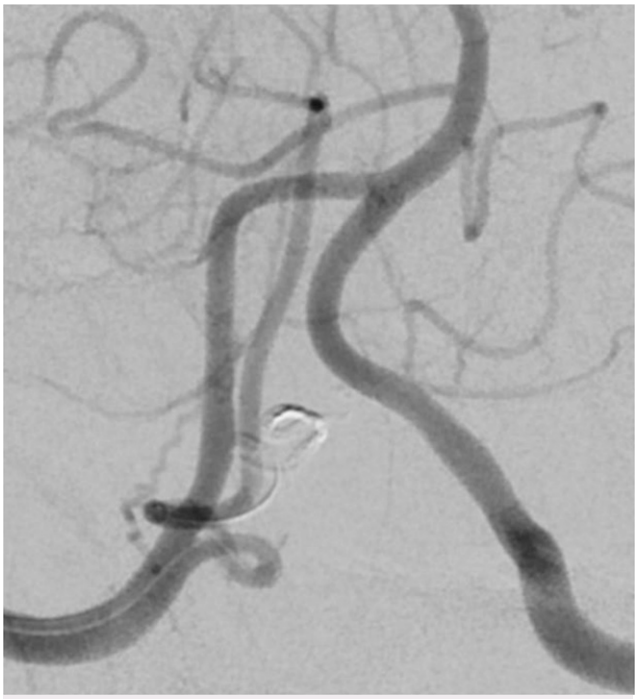

FIG. 4. Cerebral angiography after Onyx injection demonstrating successful embolization of the PAM.

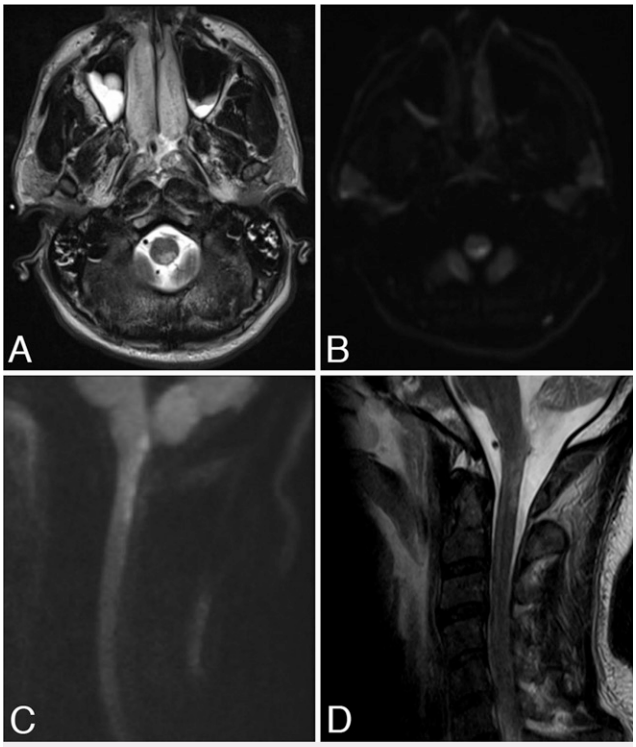

FIG. 5. MRI, axial (**A**, **B**) and sagittal (**C**, **D**) views, with abnormal T2 prolongation (**A**, **C**) and diffusion-weighted imaging restriction (**B**, **D**) at the dorsal cervicomedullary junction and upper cervical cord concerning for a subacute stroke.

even if imaging is convincing for a PAM, patients should still be closely followed.

PAM cases are most commonly diagnosed incidentally, largely in young women.[14] Of those presenting with symptoms, the majority present with headaches, with other presentations including seizures, neurological deficits, and rarely hemorrage.[2,3,7,10,13,16] A review of the literature revealed that a majority of patients presenting with non-hemorrhagic PAMs or similar arterial entities are conservatively managed.[1–3,8,16–22] Doran et al. presented the case of a 14-year-old girl who presented with complex partial seizures and was found to have dolichoectasias of bilateral anterior cerebral arteries.[21] Two occurrences of pure arterial dysplasia associated with surrounding cortical dysplasia have been reported following a motor vehicle accident and during a workup for headache, respectively, both without evidence of intracranial hemorrhage.[17,18] McLaughlin et al. highlighted the case of a 54-year-old woman who presented with acute diplopia and unsteadiness after accidentally getting sprayed in the face with insecticide.[3] She had originally presented 3 decades earlier with headaches and was found to have a lesion concerning for a suprasellar AVM, for which she was managed conservatively. MRI on presentation showed a stable suprasellar abnormality; however, DSA revealed the lesion to actually be ectatic left posterior communicating and left posterior cerebral arteries consistent with a PAM. Feliciano et al. presented the case of a 42-year-old man with sudden severe headache and left hemiparesis who was found to have a right basal ganglia hemorrhage.[13] CTA revealed a vascular lesion of the right middle cerebral artery. Subsequent DSA demonstrated a PAM consisting of ectatic collaterals from the right internal carotid and anterior cerebral arteries. In this case, there was a small medial lenticulostriate aneurysm in the vicinity of the hematoma. Uchino et al. presented the case of a 35-year-old man who had sudden-onset headache without intracranial hemorrhage and was found to have an extremely tortuous right superior cerebellar artery that at first was concerning for an AVM.[19] Brinjikji el al. described a series of 12 patients with PAMs who had no clinical changes on follow-up, with 6 of 12 confirming stable imaging on follow-up.[8] Importantly, in all of these cases, no intervention was offered, given the expected benign nature of these lesions. A recent publication by Liu et al. presented a case of a 53-year-old man with 2 weeks of left-sided paresthesias, with DSA demonstrating left-sided moyamoya disease and a dilated tortuosity of the left posterior communicating artery with an associated aneurysm.[14] There was no associated hemorrhage. Endovascular treatment with stent-assisted coil embolization was performed, and DSA at the 6-month follow-up showed resolution of the vascular dilation as well as the aneurysm.

Recent literature demonstrates that PAMs may demonstrate progression, challenging the notion that these typically take a benign course. Yue et al. recently presented a case of a 42-year-old man with progression of a right anterior cerebral artery PAM 3 years after the initial diagnosis, when he had presented with headaches.[6] Progression was identified by the development of a large aneurysm causing symptomatic obstructive hydrocephalus. There was also the development of vascular collaterals from the left internal carotid artery supplying the contralateral A2 through the anterior communicating artery, which were not present on initial imaging, and the right A2 was no longer supplied by the right internal carotid artery. There was no hemorrhage. This patient subsequently underwent open surgery for clipping of the parent vessel. Postoperative DSA showed shrinkage of the aneurysm, resolution of hydrocephalus, and no residual PAM.

The goal of both open and endovascular techniques is to safely reconstruct the parent vessel while obliterating the possible foci of rupture. Open surgical interventions can provide direct visualization of the lesion and a lasting ligation but may present an elevated risk that may not outweigh benefits in the elderly or in those with multiple decompensated comorbidities, high HH grade, or deep lesions. Surgery can be associated with prolonged surgical time, infection, bleeding, transfusions, and direct tissue damage, among other things. On the contrary, endovascular intervention in experienced hands may offer a more favorable option for high-risk patients. Embolization of the most distal aspect of the PAM may be safely achieved by decreasing the systemic blood pressure or balloon occlusion of the parent vessel to decrease reflux of embolic material. Embolization may be limited by severe tortuosity of the vessels and can be associated with stroke, vessel injury, or contrast-related adverse outcomes.

Observations

To our knowledge, there have only been 2 other reports of SAH-associated PAMs. These were both treated via open surgical ligation.[7,10] We understand that that although our example was complicated by stroke, the endovascular option, with its constant evolution, represents a feasible alternative for treatment with results comparable to those of surgery.

Lessons

PAMs remain a poorly understood vascular entity. It is hypothesized that these lesions may result from both congenital causes, including embryologic defects or insults during intracranial arterial development, and acquired causes, such as infections or trauma.[8,20] In our patient, there was no known preceding trauma or family history of vascular anomaly. His history was only significant for hypertension, which may have incited the rupture episode. That said, more work is needed to investigate whether PAMs are associated with impaired vessel integrity. Although 2 published cases have used open surgical intervention for the treatment of hemorrhagic PAMs, we show that endovascular treatment is also an alternative option. Endovascular treatment may be more favorable in high-risk patients who are unable to be medically optimized to undergo open surgery due to the nature of the setting and shorter surgical times.

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Disclosures

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

Author Contributions

Conception and design: Chua, Essayed, Ziayee, Aziz-Sultan. Acquisition of data: Chua, Gupta, Aziz-Sultan. Analysis and interpretation of data: Chua, Vicenty-Padilla. Drafting the article: Chua, Gupta, Essayed, Donnelly, Vicenty-Padilla. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Chua. Study supervision: Izzy, Aziz-Sultan.

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