A Hybrid Approach for the Treatment of a Pure Arterial Malformation Located at an Accessory Middle Cerebral Artery. Cerebral Revascularization Followed by Endovascular Occlusion Using nBCA: Case Report

Pablo Albiña-Palmarola, MD [*‡§], **Peter Kurucz, MD**[‖], **Ali Khanafer, MD**[*], **Kamran Hajiyev, MD**[*], **Hansjörg Bäzner, MD**[¶], **Hans Henkes, MD**[*‡]

*Neuroradiologische Klinik, Klinikum Stuttgart, Stuttgart, Germany; [‡]Medical Faculty, University Duisburg-Essen, Essen, Germany; [§]Department of Anatomy, School of Medicine, Pontificia Universidad Católica de Chile, Santiago, Chile; [‖]Neurochirurgische Klinik, Klinikum Stuttgart, Stuttgart, Germany; [¶]Neurologische Klinik, Klinikum Stuttgart, Stuttgart, Germany

Correspondence: Pablo Albiña-Palmarola, MD, Neuroradiologische Klinik, Klinikum Stuttgart, Kriegsbergstraße 60, 70174 Stuttgart, Germany. Email: pablo.a.med@gmail.com
Hans Henkes, MD, Neuroradiologische Klinik, Klinikum Stuttgart, Kriegsbergstraße 60, 70174 Stuttgart, Germany. Email: hhhenkes@aol.com

Received, November 19, 2023; **Accepted,** February 16, 2024; **Published Online,** April 25, 2024.

Neurosurgery Practice 2024;5(2):e00089.

BACKGROUND AND IMPORTANCE: Pure arterial malformations (PAMs) are rare vascular lesions with unknown natural history. Described in 2013 and initially considered benign, recent evidence of ruptured cases and progressive angioarchitectural changes may support the need for interventional treatment for selected patients. We present the first case of an accessory middle cerebral artery PAM successfully treated using a hybrid approach, including microsurgical revascularization and endovascular occlusion using Glubran2/Lipiodol.

CLINICAL PRESENTATION: A 58-year-old woman with a history of spontaneous basal ganglia hemorrhage 2 years before presented to our clinic with a dilated, multilobulated, and overlapping lesion located at a dominant left accessory middle cerebral artery, compatible with the diagnostic criteria of a PAM. The lesion was near the previous bleeding site, and since perforating and cortical vessels arose from its distal lobule, a safeguarding superficial temporal artery-MCA bypass was considered necessary before the successful endovascular embolization of the malformation using nBCA (Glubran2/Lipiodol).

CONCLUSION: PAMs can be associated with intracranial bleeding; thus, active treatment is warranted for selected patients. A hybrid microsurgical-endovascular approach is feasible and can be considered the optimal treatment for specific complex anatomies.

KEY WORDS: Accessory middle cerebral artery, Endovascular devascularization, Glubran2/Lipiodol, Hybrid approach, Pure arterial malformation, Surgical revascularization

In 2013, McLaughlin et al[1] first described the term and criteria distinguishing a pure arterial malformation (PAM). The presence of dilated, overlapping, and tortuous vessels forming a mass of arterial loops with a coil-like appearance without any venous component defines this rare diagnosis. Less than 50 cases have been described in the literature, with the largest study consisting of 25 patients;[2] consequently, their etiology remains unknown.[3] Usually found in the anterior circulation,[4] PAMs were thought to have a benign natural history since most cases were discovered incidentally, leading some authors to recommend conservative management.[5] Nevertheless, the recent publication of patients presenting with hemorrhage or clinical deterioration has questioned this concept, especially when aneurysmal dilations or radiological progression are present.[4,6-11] Accordingly, only a handful of cases have reported treatment directed to the malformation itself.[4,6,9,12-15]

ABBREVIATIONS: AMCA, accessory middle cerebral artery; **PA,** posterior-anterior; **PAM,** pure arterial malformation.

Supplemental digital content is available for this article at neurosurgerypractice-online.com.

We report the case of a hemorrhagic PAM located on a dominant accessory middle cerebral artery (AMCA) successfully treated using a hybrid approach consisting of preventive superficial temporal artery-MCA bypass and endovascular embolization.

CLINICAL PRESENTATION

History and Examination

A 58-year-old woman presented to our clinic after suffering a spontaneous left basal ganglia hemorrhage secondary to an unidentified vascular lesion 2 years before (Figure 1A and 1B). Subarachnoid and intraventricular hemorrhage were also noted near the lesion (**Supplemental Digital Content 1**, http://links.lww.com/NS9/A12), and although the lesion was not treated, a ventriculoperitoneal shunt was installed. Regardless, she was asymptomatic and had no neurological deficit at the moment of consultation. Computed tomography angiography (CTA), MRI, and digital subtraction angiography revealed a proximal hypoplastic left MCA and a dominant AMCA arising from an elongated A1 segment (Figures 1 and 2). Shortly after originating near the posthemorrhage basal ganglia scar, the AMCA exhibited a dilated, multilobulated, overlapping morphology, causing contrast stagnation. The typical radiological signs of arterial dissection, namely signs of an intimal flap, intramural hematoma, or eccentric arterial wall thickening, were not identified,[16] and the absence of early venous drainage ruled out the diagnosis of an arteriovenous malformation. Together, these features suggested the diagnosis of a PAM[1,2] (Figure 2A-2C). After considering the previous bleeding, we determined that treatment was necessary. Consequently, informed consent was obtained from the patient.

Treatment

Superselective catheterization of the AMCA showed lenticulostriate arteries and frontal cortical vessels arising from a distal malformation lobule, forcing treatment suspension (Figure 2E). After a thorough review, a safeguarding and uneventful end-to-side superficial temporal artery-MCA (M4 branch of the superior trunk) bypass surgery was performed before embolization. Endovascular treatment followed. Under general anesthesia, the

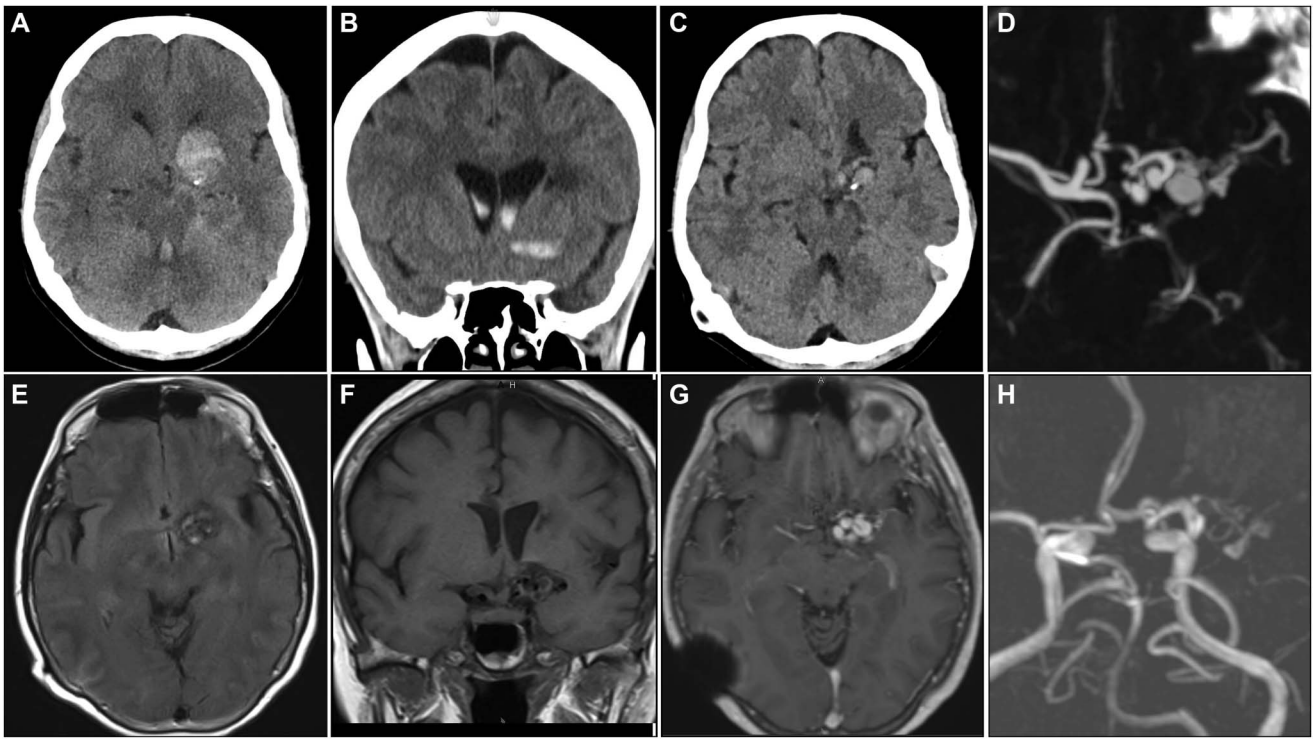

FIGURE 1. *Preoperative imaging. Acute episode: axial* **A** *and coronal* **B** *noncontrast CT showed a left hand basal ganglia hemorrhage with intraventricular hemorrhage and hydrocephalus. Two years later, the patient presented to our clinic for follow-up.* **C**, *A slightly hyperdense and irregular lesion with eccentric calcification at the left Sylvian cistern remains in the axial noncontrast CT and* **D**, *CTA revealed a complex coil-like multilobulated arterial lesion around the left M1 segment.* **E**, *Axial fluid-attenuated inversion recovery showed that the lesion was near the left basal ganglia scar and* **F**, *no signs of intramural hematoma or dissecting flaps were seen on the rest of the study (coronal T1-weighted imaging MRI);* **G**, *axial contrast-enhanced multiplanar reconstruction. A loss of signal inside and distal to the lesion in contrast to CTA was observed on TOF MRA, suggesting ectatic blood flow* **H**. *CTA, computed tomography angiography; MRA, magnetic resonance angiography; TOF, time-of-flight.*

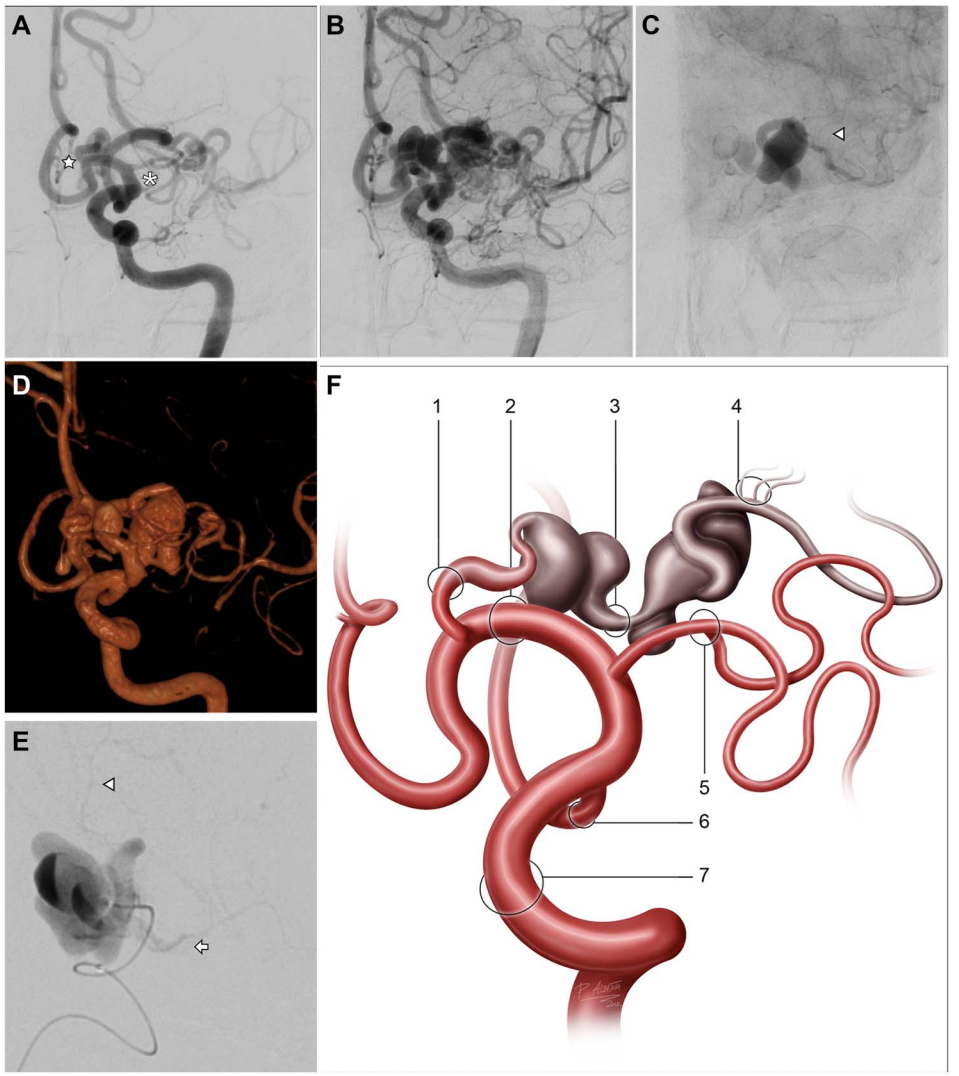

FIGURE 2. *Preoperative digital subtraction angiography. PA angiogram from early arterial to capillary phase after a left ICA injection* **A** *to* **C** *and 3-dimensional reconstruction* **D**, *identified 2 MCA-like vessels: a proximal hypoplastic one arising from the distal ICA (asterisk) and a second from an elongated A1 segment (star). Shortly after its origin, the dominant vessel exhibited a dilated, multilobulated, and overlapping morphology, with intraluminal contrast stagnation* **C** *and without signs of early venous drainage, compatible with a PAM. Some lenticulostriate arteries were observed distal to the lesion (arrowhead), prompting us to consider this vessel an AMCA. Superselective catheterization of the AMCA in lateral projection* **E**, *demonstrated that perforators (arrowhead) and cortical arteries (arrow) arose distal to the malformation, which forced the embolization to be suspended. Artist representation of the lesion's anatomy in a PA projection* **F**, *1: AMCA; 2: A1 segment; 3: PAM; 4: lenticulostriate arteries; 5: "true" MCA; 6: posterior communicating artery and posterior cerebral artery (fetal type); 7: intracranial ICA. This figure is original to this submission so no credit or license is needed. AMCA, accessory middle cerebral artery; ICA, internal carotid artery; MCA, middle cerebral artery; PA, posterior-anterior; PAM, pure arterial malformation.*

patient was given 500 mg aspirin IV and systemically heparinized, and an 8F Radifocus Introducer (Terumo) was inserted through the right femoral artery. An iatrogenic arterial dissection of the left cervical ICA was identified, prompting the administration of 180 mg ticagrelor through the nasogastric tube. Afterward, it was treated using a coaxial system consisting of a 6F Envoy (Cordis), a Phenom 21 microcatheter (Medtronic), a 0.014-inch Synchro2 200cm microguidewire (Stryker Neurovascular), and a 6/30 Solitaire AB stent (Medtronic). The bypass was patent and showed postcentral retrograde MCA territory irrigation (Figure

3A-3C, **Supplemental Digital Content 2** [http://links.lww.com/NS9/A13]). The AMCA was catheterized in a second session under local anesthesia. Selective intra-arterial injection of 8 mg propofol was tolerated without a focal neurological deficit. Successful embolization just proximal to the malformation was performed through a Marathon microcatheter (Medtronic) and an X10 microguidewire (Asahi), using 0.2 cc of Glubran2/Lipiodol (GEM/Guerbet) injected at a ratio of 1:1 (Figure 3D-3I).

Post-treatment and Follow-up

The procedure was well tolerated. Postoperative MRI demonstrated normal brain perfusion and no ischemic lesions, and the patient did not experience any clinical complications (Figure 4). The patient was discharged on the third day with a modified Ranking Score of 0. She remained on 90 mg ticagrelor twice a day for 6 months and aspirin 100 mg by mouth daily for life. The responsible ethics committee issued a waiver for approval for all case

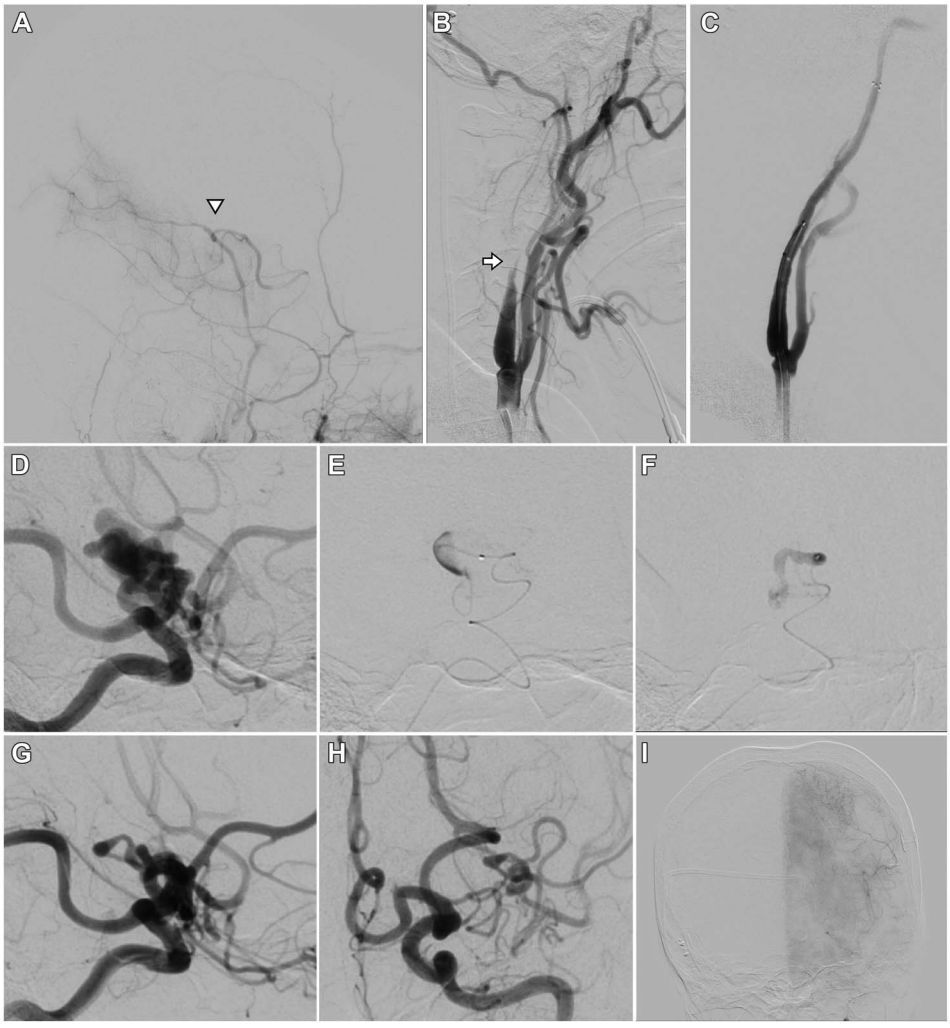

FIGURE 3. *Endovascular treatment. Lateral left external carotid artery angiogram* **A**, *demonstrated patency of the superficial temporal artery-MCA bypass and postcentral retrograde MCA territory irrigation.* **B**, *An iatrogenic left cervical ICA dissection was discovered and* **C**, *promptly resolved with a 6/30 Solitaire AB stent. Superselective catheterization of the pure arterial malformation was achieved using a Marathon microcatheter and an X10 microguidewire.* **D** *to* **F**, *After the patient tolerated an intra-arterial injection of 8 mg propofol without neurological deficit, the lesion was embolized using 0.2 cc of Glubran2/Lipiodol at a ratio of 1:1. Postembolization lateral* **G**, *and posterior-anterior projection* **H** *and* **I**, *after left ICA injection revealed the complete obliteration of the lesion. ICA, internal carotid artery; MCA, middle cerebral artery.*

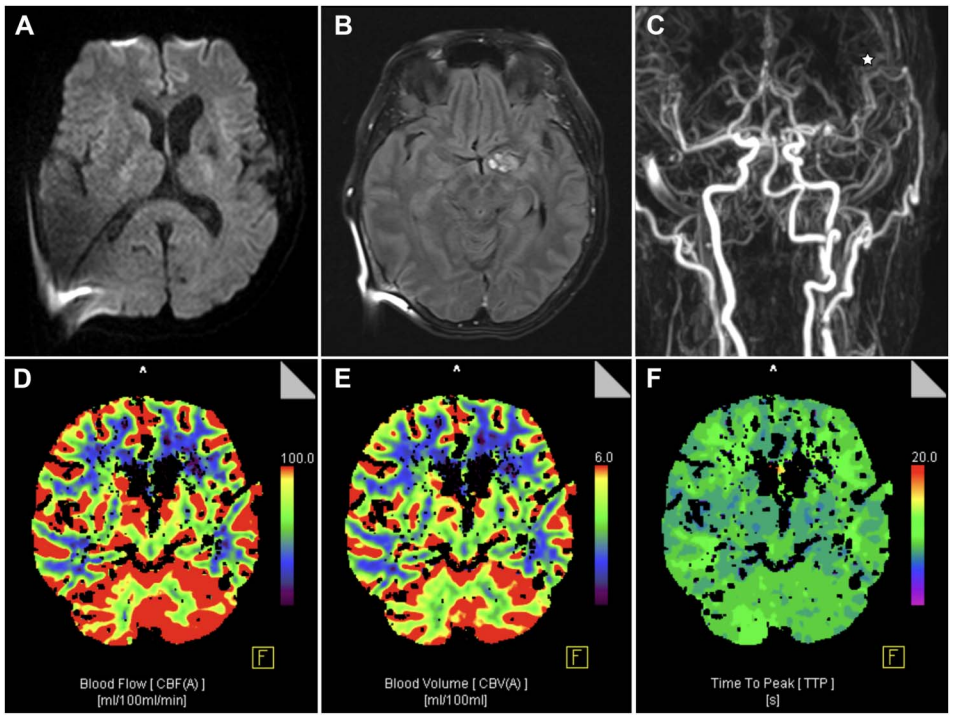

FIGURE 4. *Post-treatment imaging. One day after treatment, axial diffusion-weighted imaging* **A**, *and fluid-attenuated inversion recovery* **B**, *showed no signs of ischemic stroke. Contrast-enhanced MRA* **C**, *identified that the vascular territory of the MCA distal to the embolized lesion was successfully maintained by internal carotid artery and anterior cerebral artery leptomeningeal collaterals and through the external carotid artery-MCA bypass (star). Cerebral blood flow* **D**, *volume* **E**, *and time-to-peak parameters* **F**, *in arterial spin labeling MRI perfusion did not identify any signs of ischemia. MCA, middle cerebral artery; MRA, magnetic resonance angiography.*

reports, and the patient signed a statement allowing the publication of her case in an anonymous form.

DISCUSSION

Only one retrospective case series of 25 patients with long-term follow-up regarding PAMs exists,[2] emphasizing the rarity of this entity. After a mean follow-up of 44 months, no clinical deterioration or angioarchitectural progression were observed despite 2 patients requiring endovascular coiling for associated aneurysms and one being diagnosed with a PAM-related ischemic stroke. Interestingly, the authors concluded that these lesions had a benign natural history and could be safely treated with conservative management. However, since the introduction of PAM diagnostic criteria,[1] the publication of a handful of cases presenting with hemorrhage[4,6-11,15] and one who exhibited the emergence of a large partially thrombosed aneurysm[13] have questioned the allegedly benign nature of these lesions.

Feliciano et al presented an example of noninterventional management in a setting similar to our case, an MCA PAM characterized by M1 occlusion and a lenticulostriate artery aneurysm close to a right basal ganglia hematoma. After being unable to confirm any vascular reserve abnormality in the MCA's distal territory, revascularization was regarded unsuitable. Aneurysm occlusion was also considered significantly risky despite being the most probable cause of bleeding. Unfortunately, the patient's follow-up only lasted 1 year.[7] By contrast, regarding the possibility of angiographical progression, Yue et al[13] proposed that the presence of aneurysmal dilatations in PAMs could be an indication for treatment or, at the very least, close surveillance. The difficulty identifying these lesions from an acutely ruptured dissecting aneurysm, in which urgent treatment is the norm, has also been proposed as an argument in favor of active treatment.[10] Furthermore, it is unclear whether a prior bleeding episode increases the risk of subsequent hemorrhage, as has been found with intracranial aneurysms[17] and arteriovenous malformations.[18]

In less than 1% of cases, 2 MCA-like vessels are found. The terms AMCA and duplicated MCA are commonly used when the secondary vessel arises from the anterior cerebral artery and ICA, respectively. Regardless, this distinction lacks embryological grounds since both entities reflect the persistence of 2 separate arteries responsible for cortical supply.[19] According to Lasjaunias, the accessory vessel is defined by the presence of perforators arising from it, irrespective of its diameter. The true MCA is, therefore, purely cortical and usually proximal.[20] Although this concept has been challenged,[21] we applied these criteria in our case description. On rare occasions, aneurysms have been described in the presence of an AMCA; however, it is unknown if this configuration increases the risk of rupture or developing aneurysms.[19,21] On the other hand, PAMs have not been reported in association with an AMCA, making this the first case of its kind.

The previous hemorrhage and its anatomic relation with the malformation, the multiple dilated lobules, and the uncertain influence of AMCA location convinced us of the necessity to occlude and thus prevent a potentially devastating future bleeding episode. Yet, since part of the MCA territory relied on the affected AMCA, microsurgical revascularization was considered necessary. The relatively straightforward endovascular access to the proximal portion of the PAM through a dilated AMCA was regarded safer than microsurgical dissection and trapping, and the embolic properties of Glubran2/Lipiodol, as well as the ratio used (1:1) allowed for a quicker polymerization culminating in the successful occlusion of the malformation, without distal compromise.

Few examples of endovascular treatment have been published, including pseudoaneurysm coiling,[22] stent-assisted aneurysm coiling,[23] onyx embolization,[6] and delayed flow diversion.[15] Similarly, bypass revascularization has also been proposed[4,12]; however, this constitutes the first report of a hybrid approach directed at the malformation itself and the first PAM located on an AMCA.

Limitations

The mechanisms involved in PAM evolution are not fully understood, and more cases with longer follow-up periods are needed to evaluate the role of active treatment.

CONCLUSION

A hybrid approach is feasible, offering several advantages in treating anterior circulation PAMs.

Funding

This study did not receive any funding or financial support.

Disclosures

Hans Henkes is the coinventor of the Solitaire stent and the pRESET stent retriever and co-founder of phenox GmbH, femtos GmbH, and CONTARA GmbH, which are medical device companies developing and/or selling products for the EVT of neurovascular disorders. The other authors have no personal, financial, or institutional interest in any of the drugs, materials, or devices described in this article.

REFERENCES

1. McLaughlin N, Raychev R, Duckwiler G, Martin NA. Pure arterial malformation of the posterior cerebral artery: importance of its recognition. *J Neurosurg.* 2013;119(3):655-660.
2. Oushy S, Brinjikji W, Cloft HJ, Vine R, Lanzino G. Long-term clinical and mid-term radiographic follow-up of pure arterial malformations. *Acta Neurochir (Wien).* 2021;163(4):1181-1189.
3. Thatikunta M, Raman NV, Zieles KN, Ducis K, Jea A. An incidental pure arterial malformation in a child: case report and review of the literature. *Childs Nerv Syst.* 2020;36(11):2877-2881.
4. Lu X, Fang X, Huang Y, et al. Cerebral revascularization for the management of symptomatic pure arterial malformations. *Front Neurol.* 2021;12:755312.
5. Brinjikji W, Cloft HJ, Flemming KD, Comelli S, Lanzino G. Pure arterial malformations. *J Neurosurg.* 2018;129(1):91-99.
6. Chua MMJ, Gupta S, Essayed W, et al. Endovascular treatment of a ruptured posterior fossa pure arterial malformation: illustrative case. *J Neurosurg Case Lessons.* 2021;1(2):CASE2073.
7. Feliciano CE, Pamias-Portalatin E, Mendoza-Torres J, Effio E, Moran Y, Rodriguez-Mercado R. Color-coded digital subtraction angiography in the management of a rare case of middle cerebral artery pure arterial malformation. A technical and case report. *Interv Neuroradiol.* 2014;20(6):715-721.
8. Li Y, Sayyahmelli S, Baskaya MK. Spontaneous subarachnoid hemorrhage from a pure pial arterial malformation in the lateral cerebellomedullary junction: clinical images with a surgical video. *World Neurosurg.* 2020;135:214-216.
9. Munich SA, Brunet MC, Starke RM, Morcos JJ. Clipping of basilar perforator pure arterial malformation aneurysm: 2-dimensional operative video. *Oper Neurosurg.* 2019;17(2):E67.
10. Rosalind Lai PM, Patel NJ. Letter to the Editor. Pure arterial malformation: a rare vascular entity. *J Neurosurg.* 2018;130(1):335-336.
11. Wójtowicz K, Przepiórka Ł, Kunert P, Marchel A. Subarachnoid and intraventricular hemorrhage in a patient with a pure arterial malformation and two associated aneurysms in the posterior inferior cerebellar artery: a case report and literature review. *Cerebrovasc Dis Extra.* 2022;12(3):117-122.
12. Hanakita J, Miyake H, Nagayasu S, Nishi S, Suzuki T. Surgically treated cerebral arterial ectasia with so-called moyamoya vessels. *Neurosurgery.* 1986;19(2):271-273.
13. Yue H, Ling W, Hanmin C, et al. Progressive pure arterial malformations of the anterior cerebral artery. *World Neurosurg.* 2019;131:e52-e64.
14. Iwaki K, Arimura K, Nishimura A, Iihara K. Decompression surgery for pure arterial malformations in a 15 year old with acute, progressive visual impairment: illustrative case. *J Neurosurg Case Lessons.* 2021;1(4):CASE2037.
15. Marlow C, Cuoco JA, Ravina K, Sloboda CA, Entwistle JJ. Endovascular treatment of a ruptured pure arterial malformation and associated dysplastic middle cerebral artery dissecting aneurysm: illustrative case. *J Neurosurg Case Lessons.* 2023;5(21):CASE23150.
16. Iwama T, Andoh T, Sakai N, Iwata T, Hirata T, Yamada H. Dissecting and fusiform aneurysms of vertebro-basilar systems. MR imaging. *Neuroradiology.* 1990;32(4):272-279.
17. Greving JP, Wermer MJ, Brown RD, Jr, et al. Development of the PHASES score for prediction of risk of rupture of intracranial aneurysms: a pooled analysis of six prospective cohort studies. *Lancet Neurol.* 2014;13(1):59-66.
18. Stapf C, Mast H, Sciacca RR, et al. Predictors of hemorrhage in patients with untreated brain arteriovenous malformation. *Neurology.* 2006;66(9):1350-1355.
19. Shapiro M, Raz E, Nossek E, Chancellor B, Ishida K, Nelson PK. Neuroanatomy of the middle cerebral artery: implications for thrombectomy. *J Neurointerv Surg.* 2020;12(8):768-773.
20. Lasjaunias PBA, Brugge K. *Clinical Vascular Anatomy and Variations*, 2nd ed. Surgical Neuroangiography. Springer; 2011:XXII, 772.
21. Komiyama M, Nakajima H, Nishikawa M, Yasui T. Middle cerebral artery variations: duplicated and accessory arteries. *AJNR Am J Neuroradiol.* 1998;19(1):45-49.

22. Lanzino G, Burrows AM, Flemming KD, Cloft HJ. Pure arterial malformations of the posterior cerebral artery. *J Neurosurg.* 2014;120(2):575.
23. Liu TY, Xu N, Wan Z, et al. Diagnosis and treatment of pure arterial malformation: three case reports and literature review. *Medicine (Baltimore).* 2020;99(21):e20229.

Acknowledgments

Author contributions: Conception and design: PAP, HH. Acquisition of data: PK, HH. Analysis and interpretation of data: AK, KH, HB. Drafting the article: PAP, PK, AK. Critically revising the article: HB, HH. Approved the final version of the manuscript on behalf of all authors: PAP, HH. Study supervision: HH.

Supplemental digital content *is available for this article at neurosurgerypractice-online.com.*

Supplemental Digital Content 1. Figure 1. Bleeding pattern and relationship with malformation. At clinical presentation, subarachnoid hemorrhage located on the proximal Sylvian fissure (a) and a left basal ganglia hematoma (b) with intraventricular extension (c) were identified on NCCT. CTA showed an initially unidentified vascular lesion near the hemorrhage (d-g). A black asterisk shows the close proximity of the lesion with the hematoma. The patient's clinical history was unremarkable, without hypertension or any significant cardiovascular-associated risks. The hypothesis of a ruptured PAM as the bleeding source was supported by the hemorrhage pattern, localization, and morphological features of the malformation itself, which were identified later with DSA and MRI. CTA: computed tomography; MRI: magnetic resonance imaging; NCCT: noncontrast computed tomography; PAM: pure arterial malformation.

Supplemental Digital Content 2. Figure 2. Malformation and bypass angiographic evolution. DSA before treatment (a, b, c), one day after bypass surgery (d, e, f), and 4 months after PAM embolization (g, h, i). Left ICA injection: lateral (a, d, g) and posterior-anterior projections (b, e, h). Left ECA injection, lateral projection (c, f, i). The STA-MCA bypass was patent after surgery and remained so after embolization, allowing postcentral retrograde MCA territory irrigation, which became more apparent after successful embolization of the PAM was achieved. DSA: digital subtraction angiography; ECA: external carotid artery; ICA: internal carotid artery; PAM: pure arterial malformation; STA-MCA: superficial temporal artery—middle cerebral artery.