Progressive Pure Arterial Malformations of the Anterior Cerebral Artery

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BACKGROUND: **Pure arterial malformations (PAMs) are rare intracranial vascular anomalies. As recently reported, PAMs have a benign natural history and can be treated conservatively. However, their etiology, natural history, and treatment have not yet been fully elucidated.**

METHODS: **We present a rare case of the progression of a PAM involving the anterior cerebral artery. Magnetic resonance imaging showed a mass located in the supra-sellar region associated with obstructive hydrocephalus. Digital subtraction angiography showed evolution of the arterial anomaly with progression compared with the studies 3 years earlier. Surgical trapping of the parent artery with electrophysiological monitoring was performed. Combined with previous data, the clinical features and therapeutic strategies for this unusual entity are discussed.**

RESULTS: **The patient recovered uneventfully after surgery. Postoperative digital subtraction angiography confirmed complete obliteration of the lesion. Axial computed tomography also showed shrinkage of the aneurysm, with improvement of the hydrocephalus. Our review of the reported data showed only 4 patients with a definite or probable PAM who had undergone surgery. To the best of our knowledge, the present case represents the first report of changes in the vascular architecture of a PAM during the follow-up period.**

CONCLUSION: **Whether the aneurysmal component of PAMs merits invasive treatment has remained controversial. The findings from our case raises the possibility of evolution for some patients with PAMs and stresses the importance of scheduled follow-up noninvasive imaging studies to rule out progression of these nosological entities, especially PAMs with an "aneurysm-like" component.**

INTRODUCTION

Pure arterial malformations (PAMs) are rare intracranial vascular anomalies defined as dilated, coiled, and tortuous arterial loops and/or a mass of arterial loops without an associated early venous shunt.[1] However, their etiology, natural history, and treatment have not been fully elucidated. Recent reports have shown that PAMs result from a congenital anomaly and have a benign natural history and, as such, will generally be managed conservatively.[2] We report the case of a patient with a typical PAM of the anterior cerebral artery (ACA) who had experienced progression and evolution of the PAM to symptomatic aneurysmal dilatation. To the best of our knowledge, the present study represents the first report of changes in the vascular architecture of a PAM during follow-up observation. Our case raises the possibility of evolution for some of these cases and stresses the importance of scheduled follow-up imaging at various intervals for these patients after the original diagnosis.

CASE REPORT

Clinical History

In 2015, a 42-year-old man was evaluated for frequent headaches. No significant previous medical history was found. Head

Key words
- Arterial malformation
- Developmental arterial anomaly
- Dilative arteriopathy
- Intracranial arterial dissection
- Intracranial arterial dolichoectasia
- Vascular disorder

Abbreviations and Acronyms
ACA: Anterior cerebral artery
AComA: Anterior communicating artery
CT: Computed tomography
DSA: Digital subtraction angiography
ICA: Internal carotid artery
MRI: Magnetic resonance imaging
PAM: Pure arterial malformation
PComA: Posterior communicating artery

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computed tomography (CT) showed a high-density lesion in the right internal carotid cistern and no signs of mass effect or bleeding (Figure 1). CT angiography and digital subtraction angiography (DSA) revealed that the anomaly was formed by coiled loops of the right A1 segment of the ACA. The morphological characteristics of the vascular malformation included dilated, distorted, and tortuous artery vessels and the absence of an early venous shunt, consistent with the diagnosis of a PAM (Figure 2). His symptoms were not considered related to the arterial anomaly, and he was treated medically for the headaches. His condition remained stable, and no obvious symptoms had developed until 3 years later when he presented with severe headaches and vomiting. On questioning, he reported experiencing headaches, dizziness, and gait instability for the previous month. Bilateral papilledema was present on funduscopic examination. A brain CT scan showed a large mass in the suprasellar region with bilateral ventriculomegaly suggestive of obstructive hydrocephalus. Magnetic resonance imaging (MRI) confirmed a suprasellar mass with a mixed signal on T1-weighted sequences and a flow-void signal on T2-weighted sequences, indicating an intraluminal thrombus (Figure 3). Catheter angiography showed that the arterial anomaly involving the right A1 segment of the ACA had evolved, and aneurysmal structures were clearly visible inside the malformation. Compared with the findings obtained 3 years earlier, 2 obvious structural changes were observed. First, a giant aneurysm with thrombosis had appeared at the end of the lesion, which was supplied by a small branch of the ACA and was not present in the first image. Second, the right distal branches of the ACA were not supplied by the ipsilateral internal carotid artery (ICA). The left ICA supplied the contralateral distal branches of the ACA through the anterior communicating artery (AComA), inconsistent with the previous findings (Figure 4). Although no manifestations of acute or chronic ischemia or recent hemorrhage were found, all the symptoms were closely related to obstructive hydrocephalus.

Management, Postoperative Course, and Follow-Up Observations

We planned to occlude the aneurysm and relieve the mass effect. Therefore, through a pterional approach combined with a frontobasal interhemispheric approach, we exposed the aneurysm but did not try to excise it because of the risk of injuring critical adjacent structures. Instead, we elected to clip and trap the parent artery. The patient recovered with no new neurological deficits. The 3-month follow-up imaging studies showed shrinkage of the mass and resolution of the hydrocephalus (Figure 5). Follow-up DSA and CT angiography revealed no residual PAM and filling of the right A2 segment from the left ICA injection through the AComA (Figure 6).

Previously Reported Cases and Review of the Reported Data

Using a search of the reported data conducted via PubMed and the keywords "pure artery malformation," we identified 17 similar cases confirmed as PAMs in 7 reports in PubMed that were consistent with the clear definition of PAMs first proposed by McLaughlin et al.[3] in 2013. The clinical data of the reported cases are presented in Supplementary Table 1.[1-5] Of the 17 patients, 4 were men and 13 were women. The ratio of male to female patients was ~1:3. Headache was the most common symptom (10 of 17; 58.8%). For all the cases, except for 1, the PAM was confirmed by DSA (94.1%). For the single case, PAM had been confirmed by magnetic resonance angiography. The most commonly involved arteries were the posterior communicating artery (PComA)/posterior cerebral artery and supraclinoid ICA/M1 segment of the middle cerebral artery (4 of 17 cases each; 23.5%), followed by the ACA and posterior inferior cerebellar artery (3 of 17 cases each; 17.6%). All reported cases of definite or probable PAMs involving the ACA are listed in Supplementary Table 2.[1,6-14] No previously reported cases involved the A1 segment of the ACA. In most cases, the distal segment of the ACA, such as the A2 and pericallosal artery, was affected.

Of the 17 confirmed and previous suspected cases of PAMs, most patients underwent conservative treatment and medical management of their presenting symptoms. Only 4 patients received invasive therapy (Table 1).[4,15-17] The first case was described by Hanakita et al.[15] in 1986. Their patient underwent extracranial–intracranial bypass of the affected artery, and the ectatic middle cerebral artery was wrapped with muscle. The investigators reported that the arterial ectatic change had decreased in size after surgery.[15] The second patient had experienced short-lasting, primarily left-sided, headaches and had a complex vascular anomaly involving the PComA. He underwent coil embolization of the larger, pseudoaneurysm component. The remaining 2 patients had undergone resection of the associated cavernous angioma because of seizures and extracranial–intracranial bypass and synangiosis for associated Moyamoya disease. All the patients with of confirmed and probable PAMs showed results on the imaging analysis or clinical follow-up examinations at different times indicating a good prognosis.

DISCUSSION

We have described the case of a patient with a vascular anomaly of the right ACA consistent with a PAM that showed evolution to a symptomatic aneurysmal formation with a mass effect during a 3-year period. "Pure arterial malformations," defined as dilated, overlapping, and tortuous arteries forming a mass of arterial loops with a coil-like appearance in the absence of arteriovenous shunting, were first characterized by McLaughlin et al.[3] in 2013, although similar cases had been previously reported and characterized differently. Until recently, the definite cause and pathogenesis of PAMs have remained unclear.[1] Many potential etiologies have been proposed for these lesions. These etiologies have included a congenital defect resulting in arterial dysplasia, an insult from a viral infection or somatic mutation, a chronic healed dissection, and genetic, inflammatory, immunological, and degenerative factors.[17] Patients with arteriovenous malformations or arteriovenous fistulas can be easily distinguished by the absence of a nidus and an early venous shunt.[18] Other entities, such as intracranial arterial dolichoectasia,[19,20] intracranial arterial dissection,[21,22] fusiform aneurysm,[23,24] dilative arteriopathy, and developmental arterial anomaly,[25,26] should also be

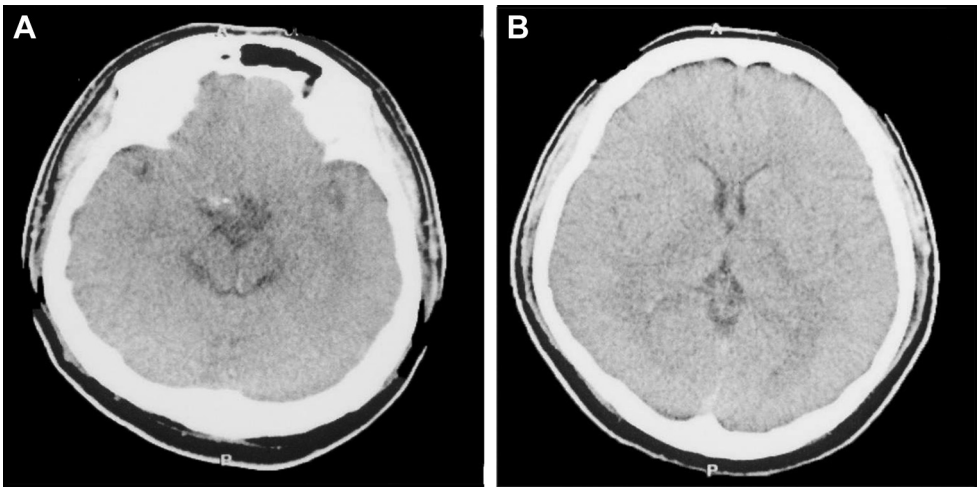

Figure 1. (**A,B**) Axial brain computed tomography scans showing slight calcification in the right internal carotid cistern but no other positive findings.

differentiated from PAMs. The characteristics of PAMs and the differential diagnosis from other intracranial vasculopathies are listed in **Supplementary Table 3**.

The ACA has seemed to be the more commonly affected site; however, no clear preference has been found, and the anomaly can affect any of the intracranial vessels.[1] PAMs at different locations

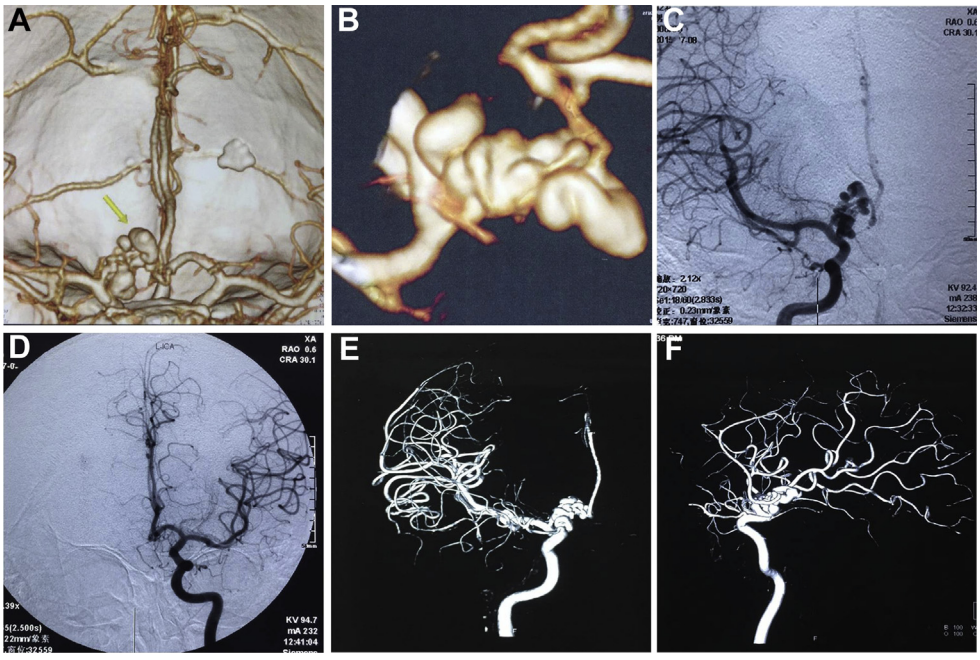

Figure 2. (**A,B**) Computed tomography angiograms showing a cluster of abnormal vessels in the right anterior cerebral artery (ACA) with a coil-like configuration. (**C**) Angiogram after anteroposterior right internal carotid artery (ICA) injection demonstrating the dilated proximal aspect of A1, representing a coiled mass of dilated elongated and tortuous vessels with no early venous drainage, suggestive of a pure arterial malformation (PAM). (**D**) Angiogram after anteroposterior left ICA injection showing the normal left ACA. (**E,F**) Three-dimensional reconstruction of the rotational angiographic view of the right ICA showing a dilated, distorted, and tortuous A1. The distal branch of the right ACA can also be seen.

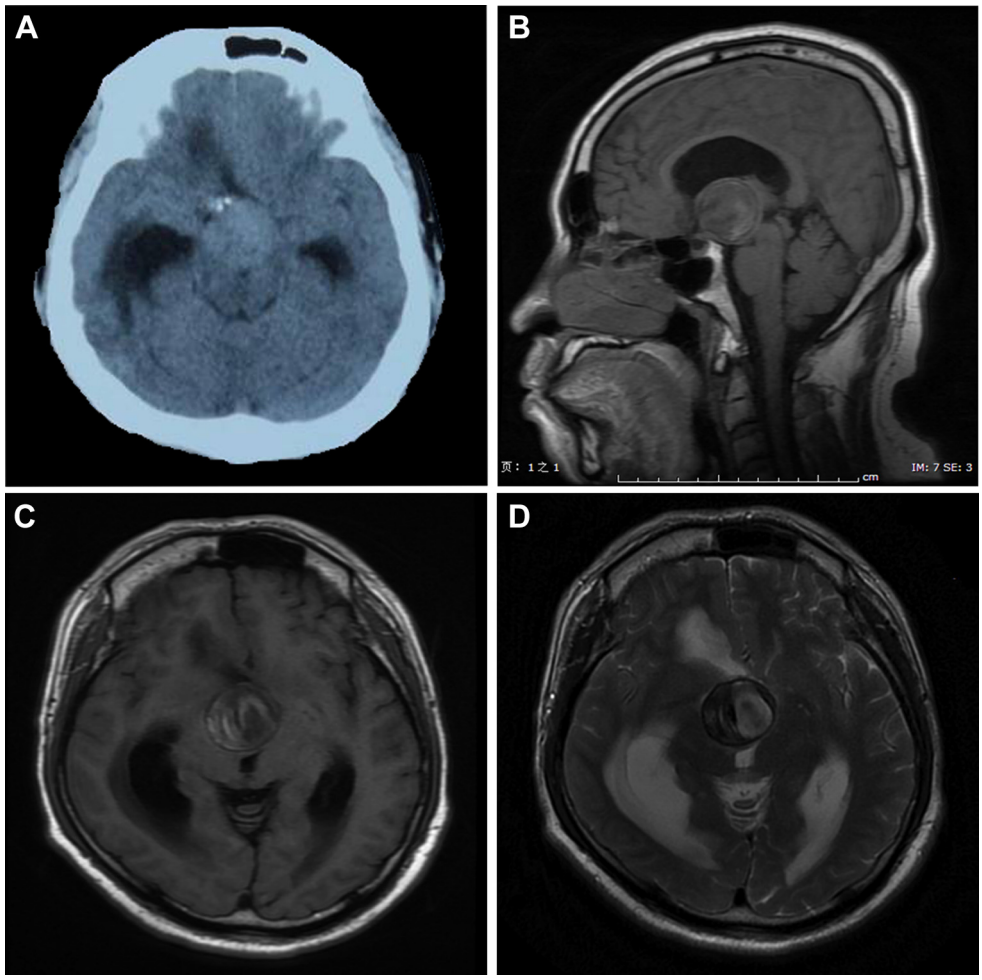

Figure 3. (**A**) Axial brain computed tomography scan showing a large mass located in the suprasellar region, accompanied by dilation of the bilateral ventricle. (**B,C**) Sagittal and axial T1-weighted magnetic resonance images showing a large suprasellar mass with a heterogeneous signal. The supratentorial ventricle was dilated by obstruction of the cerebrospinal fluid pathway. (**D**) Axial T2-weighted magnetic resonance image showing a large suprasellar mass with a flow-void signal, indicating a giant aneurysm with thrombosis.

have had varied angioarchitectural characteristics. PAMs involving distal branches of the ACA will be ectatic and moderately tortuous with a loose coil configuration. However, similar to lesions involving the M1 and PComA/posterior cerebral artery, a PAM involving the proximal A1 will exhibit dilatation, elongation, and tortuosity. Therefore, we believe that PAMs involving the proximal arteries will be quite different from lesions involving distal branches in the same artery. The hemodynamic effects and the course of the artery might be the main factors leading to these differences.

Brinjikji et al.[1] reported a series of 12 patients and found no changes during the clinical follow-up period in any of the patients. In addition, for 6 patients, the findings from the imaging studies confirmed no interval changes during the follow-up period. Furthermore, in a few reported studies, the longest period that the patients had maintained their vascular structure was 30 years.[3] However, in the present case, 2 obvious structural changes were detected on the follow-up imaging examinations within only 3 years. The first was a large aneurysm containing a thrombus that had appeared at the end of the lesion. The second was that the right A2 and its distal branches were not supplied by the right ICA. The left ICA supplied the contralateral A2 and its distal branches through the AComA, which was not observed on the previous DSA examinations. To the best of our knowledge, the present study is the first report of a case in which the angioarchitecture of a PAM changed during the follow-up period.

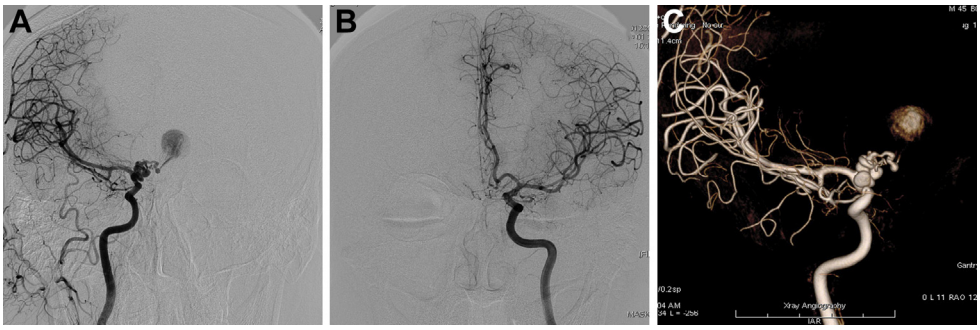

Figure 4. (**A**) Repeated angiogram after anteroposterior right internal carotid artery (ICA) injection 3 years after his initial presentation demonstrating the previous pure arterial malformation of the right A1. A giant aneurysm with thrombosis had appeared at the end of the lesion, supplied by a tiny branch of the A1. The distal branch of the anterior cerebral artery (ACA) was no longer observed compared with the first image. (**B**) Repeated angiogram after anteroposterior left ICA injection showing the normal left ACA. The distribution of the right A2 was supplied by the left A1 through the opening of the anterior communicating artery. (**C**) Three-dimensional reconstruction of rotational angiographic view of the right ICA showing a tightly coiled, distorted, and tortuous A1 segment with some aneurysmal structures inside the pure arterial malformation. A giant aneurysm with thrombosis had appeared at the end of the lesion, and the distal branch of the right ACA was no longer observed.

The patient had no history of hypertension, smoking, alcohol consumption, drug abuse, or diabetes mellitus, all of which are risk factors for aneurysm formation.[27] The reason for these changes remains unclear. However, we believe that the main contributor to these changes was the hemodynamic factor. PAMs consist of a number of dilated, overlapping, and tortuous arteries. In addition, the caliber of the perforator of the A1 segment is very small. Imaging studies of the blood flow have shown that it slows when it passes through this coil-like collection of arteries and gradually swirls, leading to dilation, thrombosis, and occlusion of the artery. Cerebral autoregulation is the physiological mechanism involved in the maintenance of cerebral blood flow during afferent vessel occlusion and plays a role in preventing tissue infarction.[28] Therefore, the patient had no symptoms when the distal part of the right A1 became chronically occluded.

Most researchers believe that PAMs have a benign natural history and can be safely treated with conservative management. Our review of the reported data showed that only 4 patients with a definite or probable PAM had undergone surgery (**Table 1**).[4,15-17] Whether invasive treatment is necessary for this lesion has remained a controversial topic. In 1 case, the reason was not reported; in 2 other cases, the patients were only treated for a coexisting disease (i.e., cavernous angioma in 1 and Moyamoya disease in the other). In 1 case, a larger, pseudoaneurysmal component of the PAM, which was suspected as the responsible lesion, was embolized with a coil. For our unique case, we initially considered that the symptom of headache was not directly related to the PAM. Therefore, conservative treatment and follow-up observation were implemented. However, the patient had presented to our hospital with severe headache, dizziness, and gait instability after 3 years. Cranial CT and MRI showed obstructive hydrocephalus due to suppression of the cerebrospinal fluid pathway by the suprasellar mass. Further examination using DSA and MRI confirmed that the suprasellar mass was a giant aneurysm containing many thrombi. Although no bleeding occurred, all symptoms were closely related to those of obstructive hydrocephalus. Therefore, we treated our patient to remove the lesion and relieve the hydrocephalus. From our analysis, we suspect that the associated aneurysmal component of the PAM was most likely the lesion responsible for the symptoms, such as hemorrhage and the mass effect. This is similar to unruptured intracranial aneurysms coexisting with arteriovenous malformations, which, theoretically, are responsible for the increased risk of bleeding and size progression. However, in the case series reported by Brinjikji et al.,[1] 4 patients had an associated aneurysmal component, and clinical and imaging changes did not occur during the follow-up period in 4 and 3 cases, respectively. Therefore, whether the aneurysmal component of PAMs merits invasive treatment has remained controversial. However, at the least, close surveillance should be advised for patients with PAMs with an "aneurysm-like" component. Finally, the real factors affecting the treatment strategy require further exploration and discussion.

In our patient, clipping and remodeling of the A1 artery were not feasible, and arterial grafting and revascularization after excision of the lesion were also very difficult. The right A2 and its distal branches were supplied by the left ICA through opening of the AComA. No obvious perforator vessels of A1 were found in any phase on repeated DSA studies, providing a reliable anatomical basis for surgical trapping of the lesion. The pterional approach, combined with the frontobasal interhemispheric approach, was chosen to surgically trap the affected A1 and clear the thrombi. For

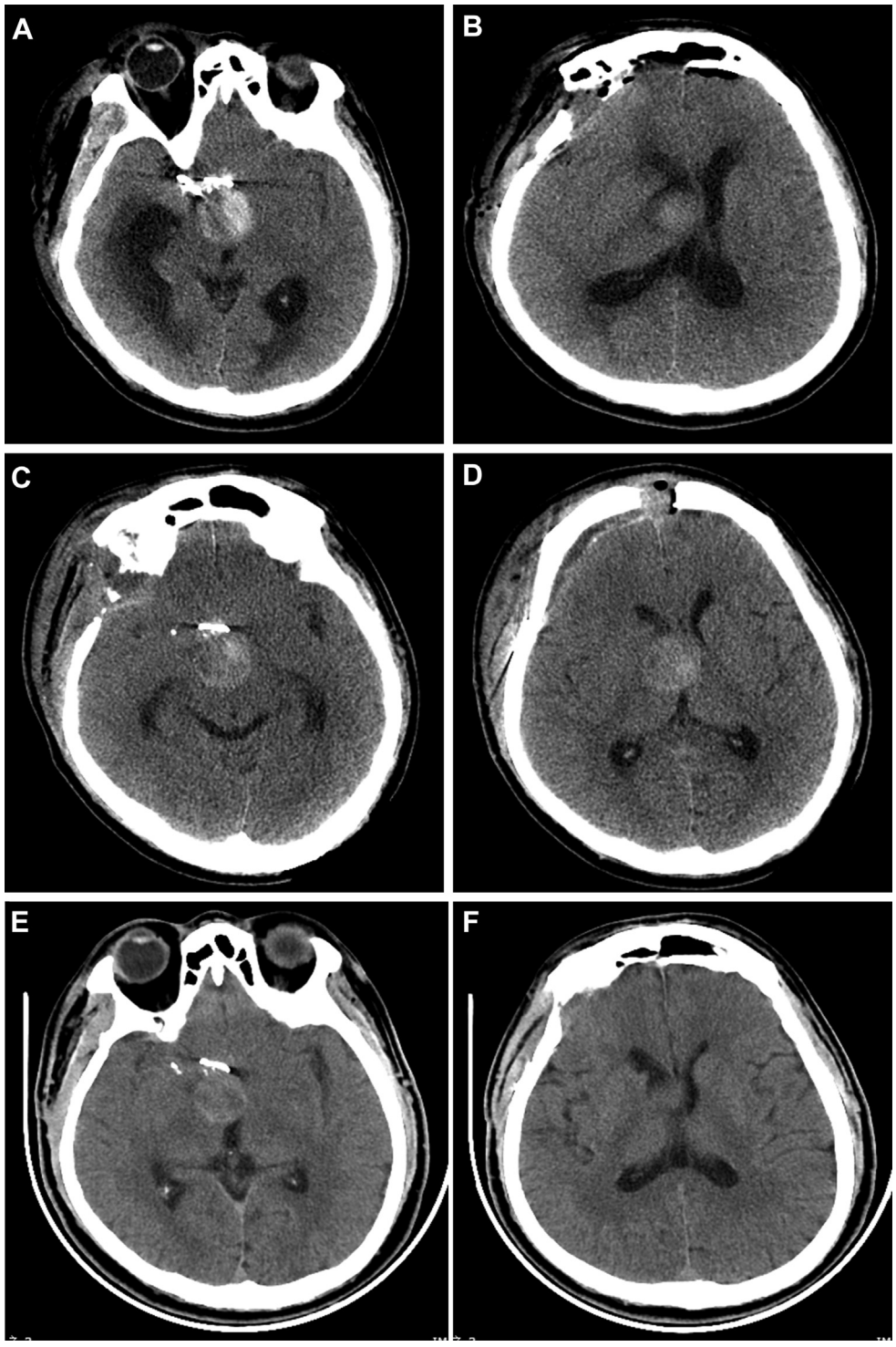

Figure 5. (**A,B**) Computed tomography (CT) scan showing a high-density lesion inside the suprasellar mass. The hydrocephalus had not improved, and no surgical-related infarction was observed on the first day after surgery. (**C,D**) Compared with the previous CT scan, the CT scan taken 14 days after surgery showed that the mass had not obviously decreased. However, the hydrocephalus had improved slightly. (**E,F**) CT scans at the 3-month follow-up examination showing that the mass had shrunk and the hydrocephalus had completely resolved.

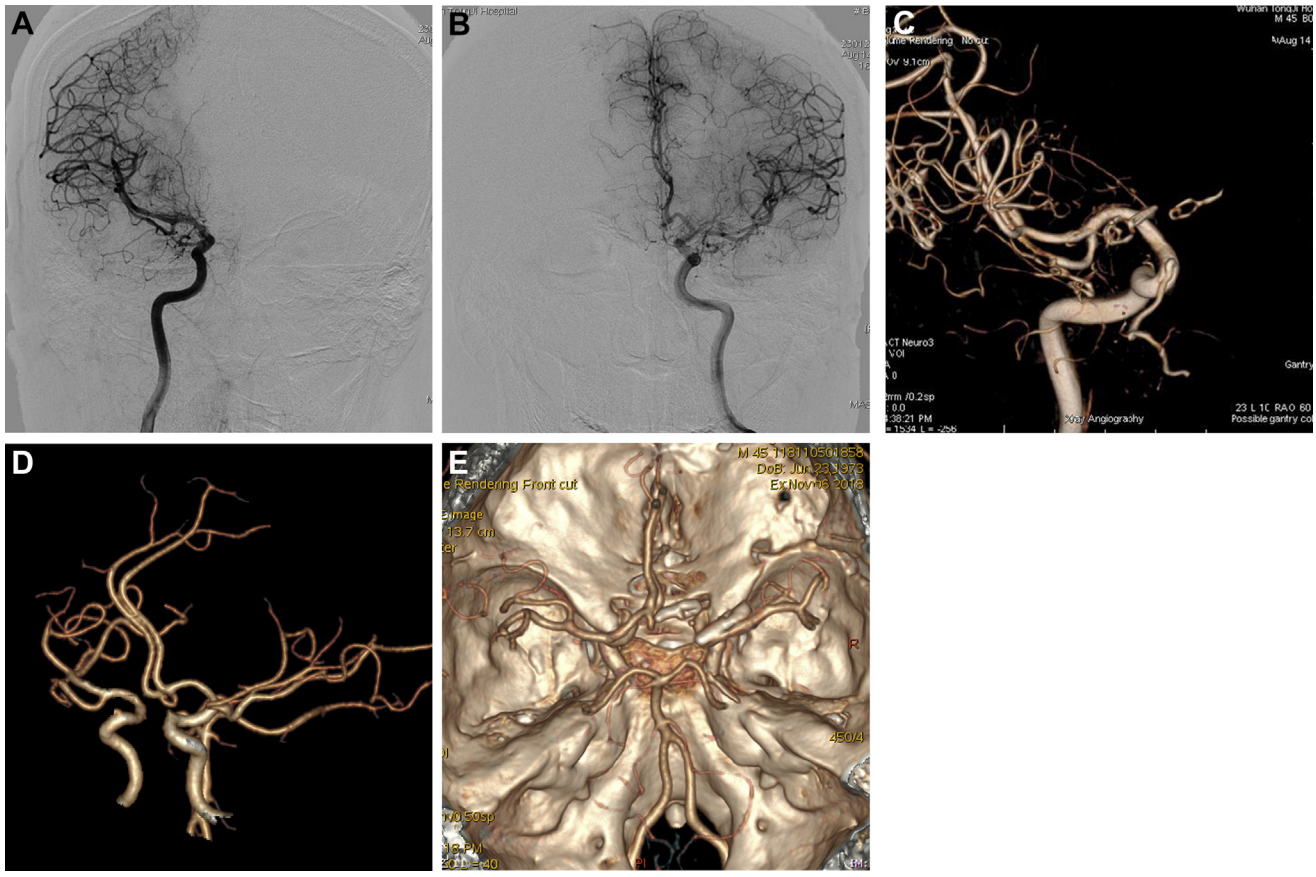

Figure 6. Anteroposterior view of follow-up digital subtraction angiograms of (**A**) right and (**B**) left internal carotid artery demonstrating that the previous pure arterial malformation (PAM) was no longer observed and that the distal branches of the right anterior cerebral artery were supplied by the left A1 through the opening of the anterior communicating artery. (**C**) Three-dimensional reconstructed computed tomography scan showing that the PAM had disappeared, and 2 clips were observed to be attached at both ends of the right A1. (**D,E**) Computed tomography angiogram at the 3-month follow-up examination showing that the PAM had disappeared and the diameter of the right A2 was the same as that of the left.

electrophysiological monitoring, we first attached 2 temporary clips (model nos. 240, 210 [Aesculap, Tuttlingen, Germany]) on both sides of the PAM. No significant decrease was found in the somatosensory evoked potentials or motor evoked potentials, and the procedure time was >30 minutes. Moreover, the abnormal vessels were no longer observed on intraoperative indocyanine green videoangiography. Therefore, we changed the clips to 2 permanent clips (model nos. 740, 712 [Aesculap]), trapping the affected vessel. Considering the high risk of injury to important structures owing to the high volume and deep location of the aneurysm, we decided not to excise the aneurysm or clear the thrombi. We expected that the volume of the aneurysm would decrease and that the hydrocephalus would improve after trapping. The patient had no neurological dysfunction after surgery, and the hydrocephalus had significantly improved at the 3-month CT follow-up visit. Follow-up DSA revealed no remaining PAM and showed that the distribution area of the distal branches of the right ACA was supplied by the left A1 through the AComA. This intervention proved to be the simplest method to solve this complex problem and was effective. Occasionally, ACA reconstruction with a bypass, such as superficial temporal artery—ACA anastomosis and A3 side-to-side in situ anastomosis to preserve the peripheral flow have been used; however, such procedures can be challenging.[29]

CONCLUSION

PAMs usually have a benign natural history, and these patients will usually receive conservative treatment and medical management of the presenting symptoms. However, because of our results, we strongly recommend that close follow-up observation and reevaluation of this rare entity should occur to determine the possibility of PAM progression, especially for PAMs with an "aneurysm-like" component. We expect more reports of similar cases will improve the understanding of the natural history and clinical significance of

Table 1. Patients with Pure Arterial Malformations Who Had Received Invasive Treatment

Pt. No.	Investigator	Age (years)/Sex	Location	Symptoms	Appearance	Reasons for Treatment	Treatment Modality	Outcome
1	Hanakita et al.,[15] 1986	43/F	Right distal ICA, proximal M1, left PCA	Dysarthria	Tightly coiled, dilated vessels; stenotic lesion of MCA	ND	EC-IC bypass, wrapped ecstatic MCA with muscle	Good
2	Kanemoto et al.,[16] 1998	41/F	Left M1 segment of MCA	Seizure	Distended and elongated MCA with complex coiling	Seizure	Resection CA; not for PAM	Good
3	Lanzino et al.,[17] 2014	10/F	Left PComA	Short-lasting, primarily left-sided headache	Complex vascular anomaly involving PComA	Suspected existence of complex dissecting PA	Coil embolization of larger PA component	Good
4	Lanterna et al.,[4] 2014	1/M	Left PComA and PCA	Hemispheric stroke from moyamoya	Tortuous and redundant PComA extending to P2 segment (left side); moyamoya (right side)	Moyamoya	EC-IC bypass and synangiosis for moyamoya (right side)	Good
Present case	He et al.	45/M	Right proximal ACA	Headache, vomiting	Dilated, distorted, and tortuous artery vessels with some aneurysmal structures inside	Hydrocephalus from giant aneurysm containing thrombus	Surgical trapping	Good

Pt. No., patient number; F, female; ICA, internal carotid artery; PCA, posterior cerebral artery; MCA, middle cerebral artery; ND, not described; EC-IC, extracranial—intracranial; CA, cavernous angioma; PAM, pure arterial malformation; PComA, posterior communicating artery; PA, pseudoaneurysm; M, male; ACA, anterior cerebral artery.

PAMs. However, further studies on the etiologies, susceptibility factors, genetic characteristics, and treatment strategies for PAMs are needed.

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REFERENCES

1. Brinjikji W, Cloft HJ, Flemming KD, Comelli S, Lanzino G. Pure arterial malformations. *J Neurosurg*. 2018;129:91-99.
2. Sorenson TJ, Brinjikji W, Flemming KD, Lanzino G. Pure arterial malformation of the posterior inferior cerebellar artery with interspersed adipose tissue: case report. *J Neurosurg Pediatr*. 2018;22:261-264.
3. McLaughlin N, Raychev R, Duckwiler G, Martin NA. Pure arterial malformation of the posterior cerebral artery: importance of its recognition. *J Neurosurg*. 2013;119:655-660.
4. Lanterna LA, Brembilla C, Gritti P. Letter to the editor: pure arterial malformation of the posterior cerebral artery. *J Neurosurg*. 2014;121:1007-1008.
5. Sako T, Uchino A, Saito N. Pure arterial malformation of the posterior inferior cerebellar artery diagnosed by MR angiography. *Neuroradiol J*. 2016;29:283-285.
6. Beringer W, Alenghat J. Pericallosal artery ectasia with associated stenosis. *AJNR Am J Neuroradiol*. 2004;25:1197-1198.
7. Tsukamoto Y, Nakata H, Soejima T, et al. Bilateral pericallosal arterial ectasia. *Neuroradiology*. 1985;27:271-274.
8. Doran SE, Deveikis JP, Chandler WF. Dolichoectasia of the anterior cerebral arteries in an adolescent. *AJNR Am J Neuroradiol*. 1995;16:1548-1550.
9. Wolpert SM, Carter BL, Ferris EJ. Lipomas of the corpus callosum. An angiographic analysis. *Am J Roentgenol Radium Ther Nucl Med*. 1972;115:92-99.
10. Kryst-Widźgowska T, Koztowski P, Binkiewicz M, et al. The angiographic and scintigraphic picture of dolichoectasia of the anterior cerebral artery. *Eur J Nucl Med*. 1980;5:387-389.
11. Sacks JG, Lindenburg R. Dolicho-ectatic intracranial arteries: symptomatology and pathogenesis of arterial elongation and distention. *Johns Hopkins Med J*. 1969;125:95-106.
12. Thompson JR, Weinstein PR, Simmons CR. Cerebral arterial dolichoectasia with seizure. Case report. *J Neurosurg*. 1976;44:509-512.
13. Yamada K, Hayakawa T, Ushio Y, Mitomo M. Cerebral arterial dolichoectasia associated with moyamoya vessels. *Surg Neurol*. 1985;23:19-24.
14. Araki Y, Takagi Y, Mineharu Y, et al. Rapid contralateral progression of focal cerebral arteriopathy distinguished from RNF213-related moyamoya disease and fibromuscular dysplasia. *Childs Nerv Syst*. 2017;33:1405-1409.
15. Hanakita J, Miyake H, Nagayasu S, Nishi S, Suzuki T. Surgically treated cerebral arterial ectasia with so-called moyamoya vessels. *Neurosurgery*. 1986;19:271-273.
16. Kanemoto Y, Hisanaga M, Bessho H. Association of a dolichoectatic middle cerebral artery and an intracranial cavernous hemangioma—case report. *Neurol Med Chir*. 1998;38:40-42.
17. Lanzino G, Burrows A, Flemming KD, Cloft HJ. Pure arterial malformations of the posterior cerebral artery. *J Neurosurg*. 2014;120:575-577.

18. Bikmaz K, Erdem E, Krisht A. Arteriovenous fistula originating from proximal part of the anterior cerebral artery. *Clin Neurol Neurosurg*. 2007;109:589-591.

19. Gutierrez J, Sacco RL, Wright CB. Dolichoectasia-an evolving arterial disease. *Nat Rev Neurol*. 2011;7:41-50.

20. Del Brutto VJ, Ortiz JG, Biller J. Intracranial arterial dolichoectasia. *Front Neurol*. 2017;8:344.

21. Sikkema T, Uyttenboogaart M, Eshghi O, et al. Intracranial artery dissection. *Eur J Neurol*. 2014;21:820-826.

22. Sikkema T, Uyttenboogaart M, van Dijk JM, et al. Clinical features and prognosis of intracranial artery dissection. *Neurosurgery*. 2015;76:663-670 [discussion: 670-671].

23. Xu K, Yu T, Guo Y, Yu J. Study and therapeutic progress on intracranial serpentine aneurysms. *Int J Med Sci*. 2016;13:432-439.

24. Kandemirli SG, Cekirge S, Oran I, et al. Intracranial serpentine aneurysms: spontaneous changes of angiographic filling pattern. *AJNR Am J Neuroradiol*. 2018;39:1662-1668.

25. Abe T, Singer R, Marks MP, et al. Arterial vascular abnormality accompanying cerebral cortical dysplasia. *AJNR Am J Neuroradiol*. 1997;18:144-146.

26. Shankar JJ, Banerjee ST, Hogan M, terBrugge K, Lasjaunias P, dos Santos MP. A rare case of cerebral cortical dysplasia with arterial vascular dysplasia. *Can J Neurol Sci*. 2009;36:757-760.

27. Jabbarli R, Dinger TF, Darkwah Oppong M, et al. Risk factors for and clinical consequences of multiple intracranial aneurysms: a systematic review and meta-analysis. *Stroke*. 2018;49:848-855.

28. Kennedy McConnell F, Payne S. The dual role of cerebral autoregulation and collateral flow in the circle of Willis after major vessel occlusion. *IEEE Trans Biomed Eng*. 2017;64:1793-1802.

29. Mitsuhara T, Ikawa F, Hidaka T, Kurokawa Y, Yonezawa U. Ruptured dissecting aneurysms of the A1 segment of the anterior cerebral artery: three case reports and a review of the literature. *Neurosurg Rev*. 2018;41:409-414.

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SUPPLEMENTARY DATA

Supplementary Table 1. Description of Reported Cases Confirmed as Pure Arterial Malformation

Pt. No.	maxInvestigator	Age (years/ Sex	Location	Symptoms	Diagnostic Method	Appearance	Treatment Modality	Clinical Follow-Up
1	McLaughlin et al.,[3] 2013	24/F	Left PComA, PCA	Frequent headache, dizziness	DSA	Tightly coiled, moderately dilated, focal saccular aneurysm	Conservative	30 Years, no change
2	Lanterna et al.,[4] 2014	1/M	Left PComA, PCA	Hemispheric stroke from moyamoya	DSA	Tortuous, redundant PComA extending to P2 segment (left side); moyamoya (right side)	EC-IC bypass and synangiosis for moyamoya (right side)	None
3	McLaughlin et al.,[3] 2013	8/F	Left supraclinoid ICA and M1	Isolated headaches	DSA	Coiled mass of dilated, elongated, tortuous vessels; focal aneurysm	Conservative	9 Months, no change
4	Sako et al.,[5] 2016	35/M	Left PICA	Vertigo	MRA	Extremely coil-like tortuosity of vessel with no venous component	Conservative	None
5	Sorenson et al.,[2] 2018	17/F	Left proximal PICA	Migraine	DSA	Coil-like configuration in its proximal portion and ecstatic	Conservative	None
6	Brinjiki et al.,[1] 2018	10/F*	Left PComA	Left-sided headache	DSA	Complex vascular anomaly involving PComA	Coil embolization of the larger, pseudoaneurysm component	3 Years, good
7	Brinjiki et al.,[1] 2018	19/F	Left MCA	Incidental	DSA	Coil-like tortuosity of distal left M1; lenticulostriate vessel superimposed multilobulated aneurysm; mild preceding stenosis	Conservative	3 Years, no change
8	Brinjiki et al.,[1] 2018	27/F	BA	Headache, numbness	DSA	Tortuous BA, tightly wound, mildly dilated, no focal aneurysmal dilatation	Conservative	5 Months, no change
9	Brinjiki et al.,[1] 2018	25/F	Left supraclinoid ICA, M1	Headache	DSA	Moderately dilated with 3 loops and 3 superimposed small aneurysms, calcified	Conservative	5 Years, no change
10	Brinjiki et al.,[1] 2018	25/F	Left A2 segment of ACA	Headache	DSA	Moderately dilated and 2 loose coil-like loops	Conservative	30 Months, no change
11	Brinjiki et al.,[1] 2018	34/F	Left A2 segment of ACA	Incidental	DSA	Corkscrew-like appearance, dilated distally	Conservative	1 Year, no change
12	Brinjiki et al.,[1] 2018	38/F	Left PICA	Transient hand numbness	DSA	Tortuous, tightly coiled	Conservative	2 Months, no change
13	Brinjiki et al.,[1] 2018	11/M	Left PComA	Incidental	DSA	Tortuous, 3 tightly coiled loops along its course	Conservative	1 Month, no change
14	Brinjiki et al.,[1] 2018	17/M	Right SCA	Headache	DSA	Tortuous and tightly coiled	Conservative	26 Months, no change

Pt. No., patient number; F, female; PComA, posterior communicating artery; PCA, posterior cerebral artery; DSA, digital subtraction angiography; M, male; EC-IC, extracranial—intracranial; ICA, internal carotid artery; PICA, posterior inferior cerebellar artery; SCA, superior cerebellar artery; ACA, anterior cerebral artery; MRA, magnetic resonance angiography; BA, basilar artery.
*This case was also reported by Lanzino et al.[17] in 2014.

Continues

Supplementary Table 1. Continued

Pt. No.	maxInvestigator	Age (years/ Sex	Location	Symptoms	Diagnostic Method	Appearance	Treatment Modality	Clinical Follow-Up
15	Brinjiki et al.,[1] 2018	47/F	Right distal ACA	Headache	DSA	Tortuous, mildly dilated, coil-like configuration, delayed venous drainage	Conservative	27 Months, no change
16	Brinjiki et al.,[1] 2018	35/F	Left supraclinoid ICA, M1, PComA	Headache	DSA	Tortuous, markedly ecstatic, coil-like configuration extending to proximal M1 and PComA	Conservative	7 Years, no change
17	Brinjiki et al.,[1] 2018	20/F	Right supraclinoid ICA; right P2, left P2 segment of PCA	Incidental	DSA	Moderately tortuous and loosely coiled (right ICA); tightly coiled, moderately tortuous right (right and left PCA)	Conservative	1 Month, no change

Pt. No., patient number; F, female; PComA, posterior communicating artery; PCA, posterior cerebral artery; DSA, digital subtraction angiography; M, male; EC-IC, extracranial—intracranial; ICA, internal carotid artery; PICA, posterior inferior cerebellar artery; SCA, superior cerebellar artery; ACA, anterior cerebral artery; MRA, magnetic resonance angiography; BA, basilar artery.
*This case was also reported by Lanzino et al.[17] in 2014.

Supplementary Table 2. Description of Reported Cases of Definite or Probable Pure Arterial Malformations Involving Anterior Cerebral Artery

Pt. No	Investigator	Sex/Age (years)	Location	Symptom	Appearance	Treatment	Follow-Up
1	Brinjikji et al.,[1] 2018	25/F	Left A2	Headache	Moderately dilated with 2 loose coil-like loops	Conservative treatment and observation	30 Months, no change
2	Brinjikji et al.,[1] 2018	34/F	Left A2	Headache	Moderately dilated, corkscrew like appearance, calcified	Conservative treatment and observation	12 Months, no change
3	Brinjikji et al.,[1] 2018	47/F	Right distal ACA	Headache	Tortuous, mildly dilated, coil-like configuration, delayed venous drainage, calcified	Conservative treatment and observation	27 Months, no change
4	Beringer et al.,[6] 2004	49/M	Bilateral pericallosal artery	Intermittent frontal headache	Tightly coiled, mildly dilated calcified, associated stenosis	Conservative treatment and observation	Several months, no change
5	Tsukamoto et al.,[7] 1985	37/F	Bilateral pericallosal artery	Mania	Moderately coiled, dilated, calcified	Conservative treatment and observation	None
6	Doran et al.,[8] 1995	14/F	Bilateral pre- and supracallosal segments of ACAs	Seizure	Tightly coiled, moderately dilated, calcified, thickening of medial frontal lobes, delayed washout	Conservative treatment and observation	None
7	Wolpert et al.,[9] 1972	21/M	Bilateral pericallosal artery	Seizure	Moderately coiled, tortuous, calcified	Conservative treatment and observation	None
8	Kryst-Widźgowska et al.,[10] 1980	72/F	Bilateral distal ACAs	Aphasia, right-sided hemiplegia	Dolichoectasia, moderately tortuous	Conservative treatment and observation	None
9	Sacks et al.,[11] 1969	2/M	Bilateral A2	Viral encephalitis	Tightly coiled, moderately dilated A2s	Conservative treatment and observation	None
10	Thompson et al.,[12] 1976	39/M	Left distal ACA	Seizure	Enlarged, moderately tortuous ACA, pericallosal artery	Conservative treatment and observation	3 Years, no change
11	Yamada et al.,[13] 1985	17/F	Left supraclinoid ICA, M1, ACA	Nausea, vomiting	Tightly coiled, moderately ecstatic cluster of vessels, calcified	Conservative treatment and observation	None
12	Yamada et al.,[13] 1985	40/F	Right supraclinoid ICA, M1, ACA	Right-sided hemiparesis	Tightly coiled, moderately ecstatic cluster of vessels, calcified	Conservative treatment and observation	None
13	Araki et al.,[14] 1987	25/F	Right MCA, ACA, PCA	Right hemimegalencephaly	Tightly coiled MCA, generalized ectasia of distal vasculature in right hemisphere	Conservative treatment and observation	None

Pt. No., patient number; F, female; ACA, anterior cerebral artery; M, male; ICA, internal carotid artery; MCA, middle cerebral artery; PCA, posterior cerebral artery.

Supplementary Table 3. Characteristics and Differential Diagnosis of Pure Arterial Malformations Versus other Intracranial Vasculopathy

Variable	PAMs	AVM or AVF	Serpentine Aneurysm	ICAD	IAD	Developmental Arterial Anomaly	Venous Anomalies or Malformation
Etiology	Unclear (congenital defect. insult from viral infection, somatic mutation)	Congenital nature	Unclear, transformation from saccular (fusiform, dolichoectatic) aneurysm; Coanda effect	More often acquired subtype (aging, male sex, arterial hypertension)	Hypertension, oral contraceptives, recent infection, head trauma	Dysfunction of embryogenesis	Dysfunction of embryogenesis
Susceptible population	Younger females	Younger ages	More often young males	Older men	VA dissection (males); ICA dissection (females)	No preference	No preference
Risk factors	No definitive factors	No definitive factors	No definitive factors	Hypertension, smoking history, genetic, immunological, infectious, inflammatory factors	Sports habits, strenuous physical activity	No definitive factors	No definitive factors
Common symptoms	Headache or incidental found	Hemorrhage, seizure	Mass effect and (hemorrhagic or ischemic) stroke	Ischemic or hemorrhagic stroke, compression of surrounding structures	SAH and ischemic symptoms; mass effect	Asymptomatic; seizure	Asymptomatic; hemorrhage (coexistent with CAs)
Angiographic characteristics	Dilated, overlapping, tortuous arteries forming mass of arterial loops with coil-like appearance	Abnormal connections between arteries and veins, with or without nidus	Partially thrombosed aneurysms with patent, tortuous vascular lumen coursing through aneurysm	Elongated, tortuous, dilated arteries at base of brain; vessel still recognizable	Double-lumen sign; pearl-and-string sign; morphological changes of involved segment	Network-like cluster of small dilated and ecstatic arteries	Wedge collections of dilated medullary veins, caput medusae shape
Artery involved	Yes	Yes; termed feeding artery	Yes	Yes	Yes	Yes	Normal
Vein involved	Not affected	Yes; termed draining vein	Not affected	Not affected	Not affected	Not affected	Yes
Preferred vessels	More proximal dilated arteries	No preference	Seventy (69.3%) involved anterior circulation; more often MCA (47.5%)	Vertebrobasilar system and ICAs	ICA and VA	Often involves distal branches of intracranial vessels	No preference
Risk of bleeding	Low	Relatively high	Relatively low	Low	Moderate	Low	Extremely low
Treatment	Observation	Observation, surgery (resection), endovascular therapy, radiotherapy, or combination	Observation; surgery (bypass); endovascular therapy (PAO)	Observation; endovascular therapy (stent)	Observation; medication (anticoagulant or antiplatelet); endovascular therapy (stent); surgery	Observation	Observation

PAM, pure arterial malformation; AVM, arteriovenous malformation; AVF, arteriovenous fistula; ICAD, intracranial arterial dolichoectasia; IAD, intracranial arterial dissection; VA, vertebral artery; ICA, intracranial carotid artery; SAH, subarachnoid hemorrhage; CAs, cavernous malformations; MCA, middle cerebral artery; PAO, parent artery occlusion.