Pure arterial malformations

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OBJECTIVE Over the last half century, there have been isolated case reports of purely arterial malformations. In this study, the authors report a consecutive series of patients with pure arterial malformations, emphasizing the clinical and radiological features of these lesions.

METHODS Pure arterial malformations were defined as dilated, overlapping, and tortuous arteries with a coil-like appearance and/or a mass of arterial loops without any associated venous component. Demographic characteristics of the patients, cardiovascular risk factors, presentation, radiological characteristics, and follow-up data were collected. Primary outcomes were new neurological symptoms including disability, stroke, and hemorrhage.

RESULTS Twelve patients meeting the criteria were identified. Ten patients were female (83.3%) and 2 were male (16.6%). Their mean age at diagnosis was 26.2 ± 11.6 years. The most common imaging indication was headache (7 patients [58.3%]). In 3 cases the pure arterial malformation involved the anterior cerebral arteries (25.0%); in 4 cases the posterior communicating artery/posterior cerebral artery (33.3%); in 2 cases the middle cerebral artery (16.6%); and in 1 case each, the superior cerebellar artery, basilar artery/anterior inferior cerebellar artery, and posterior inferior cerebellar artery. The mean maximum diameter of the malformations was 7.2 ± 5.0 mm (range 3–16 mm). Four lesions had focal aneurysms associated with the pure arterial malformation, and 5 were partially calcified. In no cases was there associated intracranial hemorrhage or infarction. One patient underwent treatment for the pure arterial malformation. All 12 patients had follow-up (mean 29 months, median 19 months), and there were no cases of disability, stroke, or hemorrhage.

CONCLUSIONS Pure arterial malformations are rare lesions that are often detected incidentally and probably have a benign natural history. These lesions can affect any of the intracranial arteries and are likely best managed conservatively.

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KEY WORDS vascular malformation; aneurysm; vascular disorders

OVER the last half century, there have been isolated case reports of purely arterial malformations defined as dilated, overlapping, and tortuous arteries with a coil-like appearance and/or a mass of arterial loops without any associated venous component.^{1–3,5–7,9–11,15–21,23–31} Now defined as “pure arterial malformations,” these lesions are commonly mistaken for aneurysms or arteriovenous malformations on noninvasive imaging.^{16,17,19,20} Unlike aneurysms and arteriovenous malformations, they are thought to have a benign natural history and are generally managed conservatively. Herein we report on a consecutive series of 12 patients with pure arterial malformations, with an emphasis on the clinical and imaging manifestations of these lesions.

Methods

Patient Selection and Disease Definition

Following IRB approval, we searched the senior author's intracranial aneurysm and vascular malformation database for cases of pure arterial malformation, dissecting aneurysm, and dissection and contacted a second author to identify any additional cases from his institution. We also searched our institution's imaging database for reports indicating vessel dilation or dolichoectasia. In defining pure arterial malformations, we used the definition put forth by McLaughlin et al.: dilated, overlapping, and tortuous arteries forming a mass of arterial loops with a coil-like appearance in the absence of any arteriovenous

ABBREVIATIONS ACA = anterior cerebral artery; AICA = anterior inferior cerebellar artery; BA = basilar artery; ICA = internal carotid artery; MCA = middle cerebral artery; PCA = posterior cerebral artery; PCoA = posterior communicating artery; PICA = posterior inferior cerebellar artery; SCA = superior cerebellar artery.

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TABLE 1. Description of cases in our single-institution series of patients with pure arterial malformations

Case No.	Age (yrs), Sex	Reason for Imaging	Lesion Location	Imaging Appearance	Max Diameter (mm)	Associated Symptoms	Imaging FU	Clinical FU
1	10, F	Severe lt-sided HAs	Lt supraclinoid ICA, PCoA, & PCA	Multilobulated pseudoaneurysm of supraclinoid ICA, partially calcified, coil-like tortuosity & dilatation of PCoA & PCA, stenosis of left M ₁	13	Yes	72 mos, no change	72 mos, no change
2	19, F	Incidental	Lt MCA	Coil-like tortuosity of distal lt M ₁ & lenticulostriate vessel w/ superimposed multilobulated aneurysm, mild preceding stenosis	16	No	NA	36 mos, no change
3	27, F	HA, lt hemibody numbness, & facial droop	BA	Tortuous BA, tightly wound, mildly dilated, no focal aneurysmal dilatation	5	Possibly	NA	5 mos, no change
4	25, F	HA	Lt supraclinoid ICA & M ₁	Tortuous & coil-like appearance of supraclinoid ICA & M ₁ w/ 3 focal aneurysms, calcified	8	No	60 mos, no change	60 mos, no change
5	25, F	HA after minor trauma	Lt ACA	Tortuous A ₂ , mildly dilated	4	No	1 mo, no change	30 mos, no change
6	34, F	Light trauma	Lt ACA	Tortuous A ₂ , mildly dilated, calcified	4	No	3 mos, no change	12 mos, no change
7	38, F	Transient hand numbness	Lt PICA	Tortuous, tightly coiled	3	No	NA	2 mos, no change
8	11, M	Incidental	Lt PCoA	Tortuous & coil-like appearance of supraclinoid ICA & PCoA	5	No	NA	1 mo, no change
9	17, M	HA	Rt SCA	Tortuous SCA, tightly wound, moderately dilated, no focal aneurysmal outpouching	4	No	NA	26 mos, no change
10	47, F	Prior thunder-clap HA	Rt ACA	Tortuous artery, mildly dilated, partially calcified, delayed venous drainage	4	No	1 mo, no change	27 mos, no change
11	35, F	HA	Lt PCoA/PCA	Arterial tortuosity w/ focal aneurysm dilatations, calcified	15	No	84 mos, no change	84 mos, no change
12	20, F	Trauma	Rt ICA, P ₂ & lt P ₂	Tortuous, tightly coiled	3	No	NA	1 mo, no change

FU = follow-up; HA = headache; NA = not available.

connection.²⁰ Only cases that met this definition as agreed upon by the study's authors were included.

Imaging Analysis

For each patient, we performed a comprehensive evaluation of all associated brain imaging studies. CT and MR images were evaluated for the presence of acute or chronic infarction; white matter disease; vascular calcifications; T1 signal abnormality of the vessel wall indicating presence of dissection; malformations of cortical development; and abnormal enhancement. Cross-sectional and conventional angiographic images were evaluated for evidence of aneurysms or stenosis associated with the pure arterial malformation, maximum diameter, location, and interval growth if follow-up imaging was available.

Clinical Chart Review

All patients received a follow-up telephone call from a research nurse to determine if the patients had suffered any disability, stroke, or intracranial hemorrhage since they last presented to our institutions. Modified Rankin

Scale scores were collected for all patients. Follow-up was performed 1 month prior to submission of this article. Clinical charts were reviewed to determine the indication for the imaging examination in which the pure arterial malformation was discovered, any history of trauma, any association between the vascular abnormality and symptoms, and any new neurological symptoms including infarction, transient ischemic attack, or hemorrhage associated with the lesion at follow-up. Baseline characteristics including demographic variables and history of cardiovascular risk factors such as smoking, hypertension, hyperlipidemia, coronary artery disease, and diabetes mellitus were collected. We also collected data on any personal or family history of connective tissue disease and aneurysm.

Statistical Analysis

This study is a descriptive study. No statistical comparisons were made. Continuous variables are reported as the mean $\pm$ standard deviation and range, and categorical variables are reported as number (%). Data analysis was performed using the SAS-based statistical software package JMP version 12.0 (www.jmp.com).

Results

Demographic and Baseline Characteristics

Twelve patients were included, 10 females (83.3%) and 2 males (16.6%). Their mean age at diagnosis was 26.2 ± 11.6 years (range 10–47 years). One patient had a history of hyperlipidemia, 1 had a history of hypertension, and 1 had a history of nicotine dependence. No patients had a history of connective tissue disease or a family history of intracranial aneurysm. One patient had a congenital bicuspid aortic valve with mild dilation of the thoracic aorta. Eleven of the 12 patients had undergone conventional cerebral angiography.

Patient Presentation

The most common symptom leading to imaging was headache (7 patients [58.3%]). Of the 7 patients with this symptom, 1 had a history of thunderclap headache and 1 had headache associated with left hemibody numbness and facial droop. Of the 5 patients who did not present with headache, 2 had their lesions discovered during evaluation for pituitary abnormality, 2 had their lesions discovered incidentally during evaluation for minor head trauma, and 1 had her lesion discovered incidentally during evaluation for transient hand numbness. Four patients (33.3%) reported a history of minor trauma. No patients had a history of major trauma resulting in skull or facial fractures or intracranial hemorrhage. No patients had any history of ischemic or hemorrhagic stroke.

Imaging Findings

In 3 cases (25.0%) the pure arterial malformation involved the anterior cerebral arteries (ACAs); in 4 cases the posterior communicating artery (PCoA)/posterior cerebral artery (PCA) (33.3%); in 2 cases (16.6%) the middle cerebral artery (MCA); and in 1 case each, the superior cerebellar artery (SCA), basilar artery (BA)/anterior inferior cerebellar artery (AICA), and posterior inferior cerebellar artery (PICA). The mean maximum diameter was 7.2 ± 5.0 mm (range 3–16 mm). Four lesions had focal aneurysms associated with the pure arterial malformation, and 5 lesions were partially calcified.

Eleven patients had undergone MRI, and in no case was intracranial hemorrhage or infarction present. Also absent was intramural hematoma suggesting dissection. In 1 patient (Case 7) we noted cortical thickening and parenchymal calcifications in the pure arterial malformation territory. Six patients had serial imaging of their lesions (median 31.5 months, range 1–84 months), and in no cases were there changes in the size or angioarchitecture of the lesion.

Clinical Outcomes and Management

None of the patients with pure arterial malformations had a permanent neurological deficit. In 1 case the pure arterial malformation was thought to be associated with the patient's symptoms (Case 1). This patient has been reported on in a prior publication and had presented with left-sided headaches that were thought to be related to the pure arterial malformation, which was located on the left

FIG. 1. **A:** Case 1. Pure arterial malformation of the PCoA and PCA in a 10-year-old girl with corkscrew-like appearance of the PCoA (arrows). **B:** Case 2. Pure arterial malformation of the left MCA in a 19-year-old woman. Image shows coil-like tortuosity of the distal left M₁ segment and a lenticulostriate vessel with superimposed multilobulated aneurysm and mild preceding stenosis (arrow). **C:** Case 3. Pure arterial malformation of the BA in a 27-year-old woman with a tightly wound coil of the proximal BA and right A₁ segment. **D:** Case 4. Pure arterial malformation of the supraclinoid segment of the left ICA and left M₁ segment, which is moderately dilated with 3 loops and 3 superimposed small aneurysms, in a 25-year-old woman. Figure is available in color online only.

supraclinoid segment of the internal carotid artery (ICA), PCoA, and PCA.¹⁷ This was the only patient who underwent treatment of the pure arterial malformation (coil embolization of a focal aneurysm pouch). Eleven patients were managed conservatively with observation and recommendations for follow-up MR angiography. All 12 patients have had clinical follow-up indicating an mRS score ≤ 2 (median duration 19 months, mean 28.9 months, range 1–84 months), and no patients have reported new symptoms related to the lesion. A summary of the cases is provided in Table 1. Representative images obtained in Cases 1–8 are provided in Figs. 1 and 2. Images and descriptions of Cases 9–12 are provided in Figs. 3–6.

Discussion

In this consecutive case series of 12 patients with intracranial pure arterial malformations, we found that these lesions are generally asymptomatic, probably have a benign natural history, and exhibit a heavily female predominance. In only 1 of the cases was the pure arterial malformation potentially associated with the patient's symptoms, and in 1 additional case the lesion was possibly associated with the patient's symptoms. Only 1 patient went on to receive treatment of an aneurysm associated with the pure arterial malformation, while in the remainder of the

FIG. 2. **A:** Case 5. Pure arterial malformation of the left A₂ segment, which is moderately dilated and has 2 loose coil-like loops, in a 25-year-old woman. **B:** Case 6. Pure arterial malformation of the A₂ segment in a 34-year-old woman. The A₂ segment has a corkscrew-like appearance and is dilated more distally. **C:** Case 7. Pure arterial malformation of the left PICA in a 38-year-old woman. The vessel has 2–3 tightly coiled loops just distal to its origin. **D:** Case 8. Pure arterial malformation of the left PCoA with 3 tightly coiled loops (arrows) along its course, in an 11-year-old boy. Figure is available in color online only.

cases, the lesions were managed conservatively. We found that these lesions can affect any of the intracranial vessels, although the most common locations were the ACAs and PCoA-PCA complex. Among patients who underwent imaging or clinical follow-up, there were no instances of new symptoms related to the lesion, no cases of growth or change in the appearance of the lesion, and no cases of rupture. These findings suggest that pure arterial malformations are probably benign lesions.

Prior Reported Cases

A summary of previously reported cases of definite or probable pure arterial malformations is provided in Table 2. While our study is likely the first consecutive case series of patients with pure arterial malformations, there have been at least 20 reported cases reported in the literature over the last half century that meet the definition put forth by McLaughlin et al.^{1–3,5–7,9–11,15–21,23–31} Similar to our series, the previously described lesions impacted all segments of the intracranial arterial tree, with lesions affecting the distal ACAs and supraclinoid segment of the ICA/PCoA/PCA accounting for over half of reported cases. In general, patients with these lesions are younger (median age 25 years), and the vast majority of patients are female. Similar to our series, the pure arterial malformations were incidental findings in over 85% of the previously reported cases, and in only 1 case did the patient undergo treatment of a pure arterial malformation. Follow-up of these lesions is limited to fewer than 10 cases, but in all cases there were no new symptoms attributed to these lesions and no changes in the size or angioarchitecture of the lesion.^{3,5,7,19,20,24,26,28}

One interesting finding from our review of the literature is the similarity in the angioarchitecture shared by these lesions depending on their location. In general, le-

FIG. 3. Case 9. This 17-year-old boy underwent MRI for headaches that were refractory to medical therapy. **A:** MR angiogram demonstrating an incidentally noted abnormal cluster of vessels in the region of the right SCA (arrow). There was no evidence of infarct or hemorrhage on MRI (not shown). **B:** Left vertebral artery angiogram revealing a mildly dilated right SCA that was tortuous and tightly coiled without an associated aneurysm or evidence of arteriovenous shunting (arrow). The patient was managed conservatively and, in 18 months of clinical follow-up, has remained asymptomatic.

FIG. 4. Case 10. This 47-year-old woman had a history of thunderclap headache. **A:** Axial CT scan showing parenchymal calcifications of the right frontal lobe. **B:** In addition, there was some faint wispy enhancement of the underlying brain parenchyma on MRI. This was thought to represent a malformation of cortical development with an associated venous drainage anomaly. A dilated and tortuous distal ACA branch was also noted in the region of this abnormality on the T1-weighted image and the contrast-enhanced image. **C:** Conventional angiogram demonstrating a mildly dilated distal ACA branch that has a coil-like configuration and no associated arterial aneurysm or arteriovenous shunting. **D:** There was delayed venous egress in the region of the abnormal enhancement on MRI as well, which was thought to represent a complex developmental venous anomaly. The patient was managed conservatively as her particular vascular abnormality was not thought to correspond to her symptoms.

sions involving the distal ACAs are characterized as ectatic, moderately tortuous, and a looser coil configuration.^{5,6,15,23,26,27,30} Furthermore, calcifications of distal ACA lesions are relatively common in previously reported cases. These features are similar to those in our cases of lesions involving the ACAs. Additionally, lesions of the MCA or PCoA-PCA are often composed of a tightly wound coil of vessels with superimposed clusters of aneurysms and calcifications.^{3,9,16,17,19–21,29,31} Previously reported lesions of the SCA and PICA also bear a remarkably similar resemblance to our cases in that the vessels have the appearance of a tightly wound coil without substantial ectasia or any associated aneurysm.^{24,28}

Of the cases reported in the literature, there were 3 cases in which the characteristics of a pure arterial malformation were surgically confirmed. The first case was of a 2-year-old boy with a viral encephalitis in which Sacks and colleagues provided an image of a tortuous and coiled

FIG. 5. Case 11. This 35-year-old woman presented following light trauma, and a complex calcified vascular lesion was incidentally found in the region of the left supraclinoid segment of the ICA on an axial non-contrast CT scan (**A**). Follow-up MR angiogram (**B**) confirmed a 5-mm left PCoA aneurysm as well as a 2-mm aneurysm of the left supraclinoid segment of the ICA. Cerebral angiograms (**C and D**) demonstrated a markedly ectatic and tortuous supraclinoid segment of the ICA with a coil-like configuration extending to the proximal M₁ segment and PCoA. The patient was managed conservatively, and in 7 years of imaging and clinical follow-up she has had no imaging change in the appearance of the lesion and has had no new neurological symptoms.

A₂ segment.²³ In the case of a 41-year-old woman with a left frontal cavernoma and a pure arterial malformation of the left MCA, Kanemoto et al. provided an intraoperative photograph demonstrating a distended and elongated MCA with complex coiling.¹¹ In their case of a 43-year-old female, Hanakita et al. reported performing a bypass of the ectatic vessel with wrapping of the vessel with muscle. During surgical exploration, they found that the ectatic ICA was constructed of double loops, some of which were very thin in portions.⁹

Distinguishing Features

Distinguishing pure arterial malformations from other vascular abnormalities that could have a similar appearance is important so as to avoid unnecessary invasive procedures. Catheter angiography is essential in the characterization of these lesions. It provides the best spatial resolution, which allows one to determine the presence of the tightly coiled loops, and provides excellent temporal resolution, which allows for evaluation of arteriovenous shunting.^{19,20} While it is easy to mistake a complex pure arterial malformation for an arteriovenous malformation nidus, catheter angiography makes distinguishing be-

FIG. 6. Case 12. This 20-year-old woman underwent noncontrast CT evaluation following trauma. Cerebral angiograms demonstrated a tightly coiled and moderately tortuous right and left P₂ segment (**A and B**) and a moderately tortuous and loosely coiled supraclinoid segment of the right ICA (**C**).

tween a pure arterial malformation and an arteriovenous malformation or arteriovenous fistula easy because there is absolutely no evidence of arteriovenous shunting in a pure arterial malformation.^{19,20}

Pure arterial malformations can be mistaken for intracranial dolichoectasia as well.^{19,20} However, there are a number of important distinguishing features between the two. Dolichoectasia predominantly affects the vertebro-basilar system and ICAs. Branch arteries such as the distal ACAs, PCAs, PICA, SCAs, and PCoA are rarely involved. The angioarchitecture of a pure arterial malformation also differs substantially from a dolichoectatic vessel. While the vessel in dolichoectasia is elongated, dilated, and tortuous, the vessel is still recognizable. However, in a pure arterial malformation the tortuosity is often so severe that the vessels are overlapped and have the appearance of a mass or cluster or arterial loops. Furthermore, the demographic characteristics of patients with dolichoectasia and pure arterial malformations differ vastly, as an intracranial dolichoectasia tends to affect older men with a history of hypertension and smoking, whereas, in our series and in cases reported in the literature, pure arterial malformations predominantly affect younger females without a history of cardiovascular risk factors.^{8,22} While dolichoectasia can occur in younger individuals, these patients often have a known connective tissue disease, genetic abnormality, or a history of viral infection that resulted in the dolichoectasia. In the MCA territory, pure arterial malformations must be differentiated from the so-called serpentine aneurysms, which represent a completely separate pathological entity.³²

Potential Etiologies

The pathogenesis and etiology of pure arterial malformations are unclear. A number of authors have speculated as to the origin of these bizarre lesions. Potential etiologies that have been discussed in the literature include: 1) a congenital defect or an insult resulting in an arterial dysplasia; 2) an insult such as a viral infection or somatic mutation occurring later in life and affecting a particularly vulnerable arterial segment (i.e., segmental arterial vulnerability); or 3) a chronic healed dissection. An underlying connec-

tive tissue disease is another possibility, but it is important to note that none of our 12 cases had any history of underlying connective tissue disease, and in a recent review of patients with connective tissue diseases at our institution, no patients had any such abnormalities.^{12,13} Undoubtedly, many of these lesions, particularly the calcified ones, were likely present for a period of time prior to detection. Ultimately, the most likely explanation for these lesions is that they are the phenotypic manifestation of a wide range of etiologies or are the result of a combination of the aforementioned factors.

Undoubtedly there are cases in the literature in which the arterial dysplasia seen in a pure arterial malformation is congenital or embryological in nature. In 5 of the previously reported cases and in 1 of the cases in our series (Case 8) there were cortical or white matter dysplasias in the area supplied by the pure arterial malformation, including focal cortical dysplasias, hemimegalencephaly, or agenesis of the corpus callosum.^{1-3,21,30} Since arterial development is thought to be stimulated by the development of the overlying brain parenchyma, it seems reasonable that the arterial abnormalities accompanied the formation of the cortical abnormalities.⁴

There are certainly cases in which an underlying genetic or infectious insult would result in the formation of these pure arterial malformations. It is possible that some of the lesions we have observed that involve the supraclinoid segment of the ICAs or the MCAs are due to a remote viral infection, as viruses such as varicella zoster have a tropism for these vessels.¹⁸ There is at least 1 previously reported case in the literature in which a patient with varicella vasculopathy had serial imaging that demonstrated progressive development of a tightly coiled arterial malformation of the supraclinoid segment of the ICA and M₁ segment with an associated proximal stenosis.¹⁸ One autopsy case involving a pure arterial malformation of the bilateral A₂ segments has been described in a 2-year-old child who died following a viral encephalitis.²³ Ultimately, this probably represents the minority of cases because varicella vasculopathy is often associated with severe neurological symptoms including acute ischemic stroke and hemorrhage.

TABLE 2. Review of the literature

Authors & Year	Age (yrs), Sex	Location	Appearance	Symptoms	Infarct, Hemorrhage, or Compression	Surgical or Endovascular Treatment	FU
Beringer & Alenghat, 2004	49, M	Bilat pericallosal	Tightly coiled, mildly dilated calcified, associated stenosis	Intermittent frontal HA	None	None	Several mos, no change
Tsukamoto et al., 1985	37, F	Bilat pericallosal	Moderately coiled, dilated, calcified	Mania	None	None	None
Doran et al., 1995	14, F	Bilat pre- & supracallosal segments of ACAs	Tightly coiled, moderately dilated, calcified, thickening of medial frontal lobes, delayed washout	Szs	None	None	None
Wolpert et al., 1972	21, M	Bilat pericallosal	Moderately coiled, tortuous, calcified	Szs, agenesis of corpus callosum	None	None	None
Kryst-Widzowska et al., 1980	72, F	Bilat distal ACAs	Dolichoectasia, moderately tortuous	Aphasia, rt-sided hemiplegia	Unrelated infarct	None	None
Sacks & Lindenburg, 1969	2, M	Bilat A ₂ s	Tightly coiled, moderately dilated A ₂ s	Viral encephalitis	None	None	None
Thompson et al., 1976	39, M	Distal lt ACA	Enlarged & moderately tortuous ACA & pericallosal artery	Szs	None	None	3 yrs, no change
McLaughlin et al., 2013	24, F	Lt PCoA & P ₂ w/ saccular aneurysm	Tightly coiled, moderately dilated, focal saccular aneurysm	Frequent HAS & dizziness	None	None	30 yrs, no change
Lanterna et al., 2014	1, M	Lt PCoA & PCA	Tightly coiled, dilated vessel, associated w/ ipsilateral moyamoya	Infarct from moyamoya	None	None	None
Vanslambrouk et al., 2000	5, M	Lt ICA, PCoA, PCA, MCA, & lt SCA	Tightly coiled, moderately dilated	Minimal rt hemiparesis	Brainstem compression	None	None
Baccin et al., 2007	4, F	Lt supraclinoid ICA & PCoA	Tightly coiled, markedly dilated	PHACE syndrome, rt-sided hemiparesis	Infarct due to PAM	None	16 mos, no change
	1, F	Lt supraclinoid ICA, PCoA, P ₁ ; rt supraclinoid ICA	Tightly coiled, markedly dilated	PHACE syndrome, fever, & hypotonia	None	None	None
McLaughlin et al., 2013	8, F	Lt supraclinoid ICA & proximal M ₁	Tightly coiled, dilated vessel, focal aneurysm	Sinus infection	None	None	None
Metry et al., 2001	1, F	Lt MCA & supraclinoid ICA	Tightly coiled, elongated, moderately dilated	PHACE syndrome	None	NA	None
Yamada et al., 1985	17, F	Lt supraclinoid ICA, M ₁ , & ACA	Tightly coiled, moderately ectatic cluster of vessels, calcified	Nausea & vomiting	None	None	None
	40, F	Rt supraclinoid ICA, M ₁ , & ACA	Tightly coiled, moderately ectatic cluster of vessels, calcified	Rt-sided hemiparesis	None	None	None
Feliciano et al., 2014	42, M	Rt MCA	Markedly ectatic distal M ₁ w/ superimposed cluster of aneurysm	HA w/ basal ganglia hemorrhage	Hemorrhage due to PAM	None	1 yr, no change
Kanemoto et al., 1998	41, F	Lt MCA	Tightly coiled, moderately dilated & elongated	Szs, ipsilateral cavernoma	None	None	None
Araki et al., 1987	25, F	Rt MCA, ACA, & PCA	Tightly coiled MCA, generalized ectasia of distal vasculature in rt hemisphere	Rt hemimegalencephaly	None	None	None

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TABLE 2. Review of the literature

Authors & Year	Age (yrs), Sex	Location	Appearance	Symptoms	Infarct, Hemorrhage, or Compression	Surgical or Endovascular Treatment	FU
Hanakita et al., 1986	43, F	Rt distal ICA & proximal M ₁ ; Lt PCA	Tightly coiled, dilated vessels, stenotic lesion of MCA	Dysarthria	None	EC-IC bypass, wrapped ectatic MCA w/ muscle	None
Uchino et al., 2003	35, F	Rt SCA	Tightly coiled, moderately dilated	Severe HA	None	None	2 yrs, no change
Shankar et al., 2009	26, F	Distal rt PCA	Tightly coiled, not dilated, associated w/ cortical dysplasia	Incidental	None	None	None
Sako et al., 2016	35, M	Lt PICA	Tightly coiled distal PICA	Vertigo	None	None	6 mos, no change
Abe et al., 1997	32, M	Sylvian branches, Lt MCA	Plexiform arterial network w/ tortuous vessels	Cortical dysplasia, Szs	None	None	None

EC-IC = extracranial-intracranial; PAM = pure arterial malformation; PHACE = Posterior fossa malformations, Hemangiomas, Arterial anomalies, Coarctation of the aorta and other cardiac defects, and Eye abnormalities; Szs = seizures.

Another reported theory for the etiology of these lesions is a chronic healed dissection.^{16,17,19,20} Arterial dissections can heal in a variety of ways with fusiform dilatations, stenoses, and bizarrely shaped complex aneurysms.¹⁴ In none of the cases in our series did we find evidence of a recent dissection on MRI (i.e., abnormal T1 signal in the vessel wall), even in patients who were recently symptomatic. Interestingly, in none of the cases included in this study was there any evidence of prior infarct or hemorrhage on MRI or CT, and infarct and hemorrhage were each present in only 1 case each among cases of possible pure arterial malformation in our literature review. There are some reports in the literature of chronic dissections that have taken on a bizarre, tortuous appearance with clusters of aneurysms, but these lesions are often associated with prior subarachnoid hemorrhage.¹⁴

Management

As described in our series and in numerous previously reported cases, the best management strategy for many of these patients is conservative management with serial follow-up imaging and management of risk factors such as smoking and hypertension. There have only been 2 reported cases in the literature in which surgical or endovascular treatment was used to treat these lesions, one from our own institution that has been previously described and the other described by Hanakita et al., who performed an extracranial-intracranial bypass of the affected vessel.^{9,17} In each of these cases, it is not clear whether invasive treatment was absolutely necessary, and in our own case, in retrospect, the ipsilateral transient headaches were most likely unrelated to the malformation in question. Even in the minority of patients who have presented with symptoms related to the pure arterial malformation, conservative management has proven to be effective. In 2 previously reported cases, pure arterial malformations were associated with symp-

toms including stroke and basal ganglia hemorrhage due to rupture of a tiny lenticulostriate aneurysm associated with a pure arterial malformation, and in both cases recurrent ischemic or hemorrhagic events were absent at 16 and 12 months of follow-up.^{3,7} The most common recommendation at our institution for patients with pure arterial malformations is serial imaging follow-up with head MR angiography to ensure that there is no change in the characteristics of the lesion.

Limitations of the Study

Our study has limitations. First, there are incomplete imaging data. For this reason, it is difficult to accurately characterize the imaging natural history of these lesions. Nonetheless, our clinical follow-up is up to date, and no patients experienced any new symptoms or disability related to the aneurysm. Our small sample size is another limitation. We do not have any data on the histopathological characteristics of the lesions, nor do we have information on the surgical/anatomical characteristics of these lesions. Explanations regarding the etiology of these lesions are purely putative. Since these lesions have only recently been defined, the definition of is still in flux. Nonetheless, the cases that we presented meet the criteria put forth by McLaughlin et al.^{19,20} Lastly, with so few reported cases and the short follow-up period, it is difficult to completely characterize the natural history of these lesions.

Conclusions

Pure arterial malformations are exceedingly rare lesions that are often detected incidentally and probably have a benign natural history. They can affect any of the intracranial arteries and are characterized as dilated, overlapping, and tortuous arteries with a coil-like appearance and/or mass of arterial loops without any associated ve-

nous component. These lesions are most likely more common than reported. Further case series on pure arterial malformations are needed to better educate the neurovascular community on the clinical and radiological characteristics of these lesions so they are not mistaken for dangerous lesions such as brain arteriovenous malformations and dissecting intracranial aneurysms.

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Disclosures

Dr. Lanzino is a consultant for Medtronic.

Author Contributions

Conception and design: all authors. Acquisition of data: Brinjikji. Analysis and interpretation of data: Brinjikji. Drafting the article: Brinjikji, Lanzino. 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: Brinjikji. Statistical analysis: Brinjikji. Study supervision: Lanzino.

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