

## Endovascular treatment of a ruptured pure arterial malformation and associated dysplastic middle cerebral artery dissecting aneurysm: illustrative case

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**BACKGROUND** Pure arterial malformations are characterized as unique cerebrovascular lesions with a dilated, coil-like appearance and tortuous arteries without early venous drainage. Historically, these lesions have been described as incidental findings with a benign natural history. However, pure arterial malformations can rarely demonstrate radiographic progression and develop associated focal aneurysms with an unclear risk of rupture. Whether radiographic progression of these lesions or the presence of an associated aneurysm warrants treatment remains controversial.

**OBSERVATIONS** A 58-year-old male presented with sudden-onset left hemiparesis. Computed tomography revealed a large, acute, right frontotemporoparietal intraparenchymal hemorrhage with underlying irregular curvilinear calcifications. Diagnostic cerebral angiography revealed a dysplastic right middle cerebral artery dissecting aneurysm along the M2 segment associated with a pure arterial malformation, which was treated with endovascular flow diversion in a delayed fashion.

**LESSONS** Pure arterial malformations with associated focal aneurysms may not exhibit a benign natural history as once thought. Intervention should be considered for ruptured pure arterial malformations to mitigate the risk of rerupture. Asymptomatic patients with a pure arterial malformation with an associated aneurysm should at least be followed closely with interval radiographic imaging to evaluate for malformation progression or changes in aneurysmal morphology.

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**KEYWORDS** aneurysm; cerebrovascular; endovascular; pure arterial malformation; vascular lesion; vascular malformation

Pure arterial malformations are rare intracranial vascular entities characterized by a dilated, coil-like appearance typically associated with calcifications.<sup>1-6</sup> Given their tortuous appearance on digital subtraction angiography, these cerebrovascular lesions are commonly mistaken for arteriovenous malformations or arteriovenous fistulas; however, they can be distinguished from such vascular anomalies on the basis of their lack of early venous drainage.<sup>1-6</sup> Although their etiopathogenesis remains ambiguous, pure arterial malformations are often considered incidental findings and thought to have a benign natural history.<sup>1,6-9</sup> Nevertheless, recent reports have illustrated that these lesions may exhibit a more aggressive course, because they have been associated with radiographic progression in angioarchitecture, aneurysm development, and rupture of the parent pure arterial malformation or associated focal aneurysms.<sup>9-17</sup> Here, we describe a case of a 58-year-old male with a large intraparenchymal hemorrhage suspected to be secondary to

rupture of a dysplastic right middle cerebral artery dissecting aneurysm associated with a pure arterial malformation. We review the current literature on pure arterial malformations to better characterize the natural history of these lesions and consider the indications for intervention.

### Illustrative Case

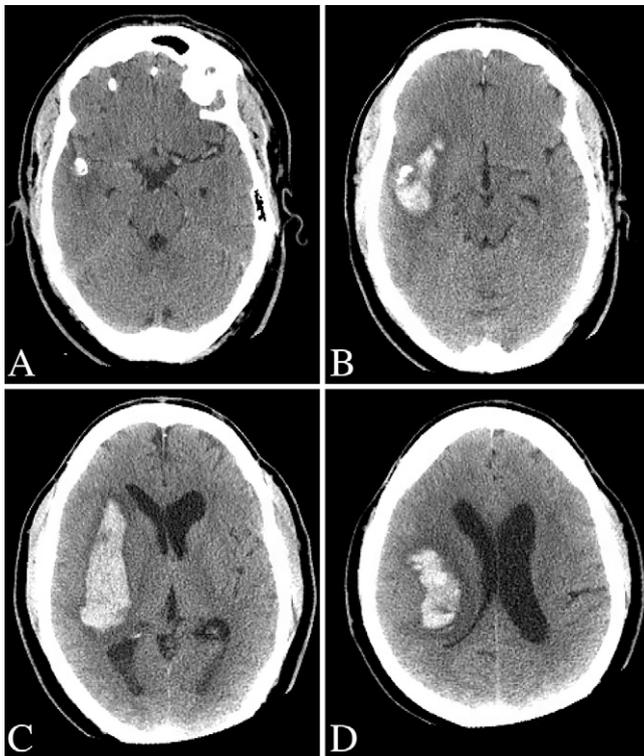
#### History and Examination

A 58-year-old male presented to an outside hospital emergency department with sudden-onset left hemiparesis noticed a few hours prior to presentation. His past medical history was notable only for hyperthyroidism. Computed tomography of the head showed a large, acute, right frontotemporoparietal intraparenchymal hemorrhage (Fig. 1). Notably, there was an irregular curvilinear calcification within the right temporal lobe hemorrhage suspicious for an underlying vascular

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**FIG. 1.** Initial head computed tomography scans (A–D) show a large, acute, right frontotemporoparietal intraparenchymal hemorrhage. Irregular curvilinear calcification within the right temporal lobe hemorrhage was suspicious for an underlying vascular malformation.

malformation, prompting further work-up. A diagnostic cerebral angiogram revealed an abnormal cluster of dilated vessels with a tight, coil-like appearance off the distal M2 and proximal M3 branches of the right middle cerebral artery. Additionally, the parent M2/M3 branches appeared dysplastic with focal aneurysmal outpouchings. There was no evidence of an early draining vein or compact nidus to suggest the presence of an arteriovenous malformation or arteriovenous fistula. There was no evidence of tumor blush. These findings were consistent with a dysplastic right middle cerebral artery dissecting aneurysm along the M2 segment associated with a pure arterial malformation (Fig. 2). The patient was placed in the intensive care unit with repeat imaging to ensure stability of the hemorrhage. He was dispositioned to inpatient rehabilitation. He then presented 3 months later to our outpatient cerebrovascular clinic to establish care after relocating. Given the recent symptomatic hemorrhage, treatment was recommended.

### Endovascular Treatment

Surgical options as well as endovascular options were considered. Elective endovascular flow diversion with a FRED Jr flow redirection endoluminal device (MicroVention) of the M2 segment was pursued to secure the dysplastic middle cerebral artery dissecting aneurysm. Intraarterial access was obtained via catheterization of the right radial artery using a micropuncture kit. A 7-French slender introducer sheath was placed into the right radial artery. A Rist radial access guide catheter (Medtronic) with a 5-French SIM2 diagnostic catheter (Cordis) was introduced into the system over a

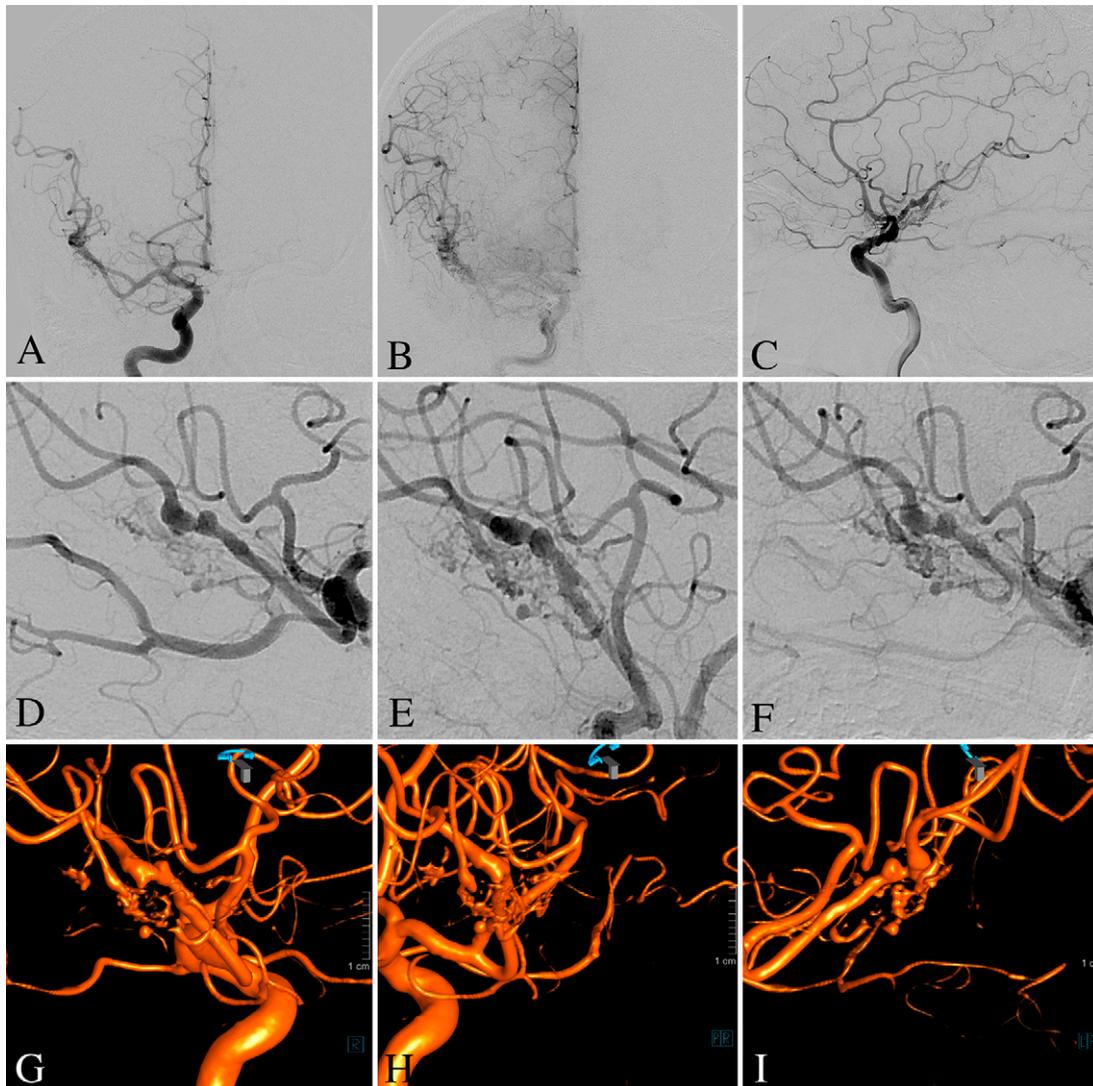
35 Glidewire (Terumo) and navigated to the right internal carotid artery. Heparin (50 U/kg) was administered, with a therapeutic activated clotting time being obtained (>250 seconds). The Rist guide catheter was placed into the petrous segment of the right internal carotid artery. A Headway 21 microcatheter (MicroVention) was advanced over an Aristotle 14 microwire (Scientia Vascular) into the distal right M2 segment of the middle cerebral artery. A FRED Jr measuring 2.5 mm × 25 mm was then deployed from the distal M2 segment to the proximal M2 segment across the entirety of the dissecting aneurysm and pure arterial malformation. Deployment was uneventful. Angiographic runs were obtained, showing good position of the FRED Jr without parent vessel compromise and interval reduction in filling of the pure arterial malformation (Fig. 3). Interestingly, a repeat diagnostic cerebral angiogram at 6 months showed occlusion of the M2 segment just proximal to the deployed flow diverter despite compliance with dual antiplatelet therapy (Fig. 4). The dysplastic middle cerebral artery dissecting aneurysm was no longer filling consequent to proximal vessel occlusion. The pure arterial malformation exhibited similar flow and appeared unchanged in size compared with the prior angiogram. Given that the dysplastic aneurysm was secured and the patient remained neurologically stable in the interval 6 months, conservative measures were pursued. At the 18-month clinical follow-up, he exhibited persistent left hemiparesis but without any new symptomatology since his initial hospitalization.

## Discussion

### Observations

Pure arterial malformations are characterized as unique cerebrovascular lesions with a dilated, coil-like appearance and tortuous arteries typically associated with calcifications.<sup>1–6</sup> They were first described in 2013, and McLaughlin et al.<sup>1</sup> provided a framework for differentiating pure arterial malformations from other cerebrovascular lesions, including 1) lack of venous involvement, 2) proximal location, and 3) absence of cortical dysplasia. The absence of a compact nidus and early venous drainage favors a pure arterial malformation as compared with the more commonly known arteriovenous malformation.<sup>18</sup> However, other cerebrovascular entities should be considered, including arterial dolichoectasia, arterial dissection, fusiform aneurysm, developmental arterial anomaly, and dilative arteriopathy.<sup>19–24</sup> Dolichoectasia and pure arterial malformations exhibit subtle differences. In contrast to the elongated appearance of dolichoectatic vessels, pure arterial malformations typically exhibit coil-like vessels described to be in a serpentine distribution.<sup>6,19,20</sup> Additionally, arterial dysplasia is differentiated from pure arterial malformations from the latter two criteria whereby arterial dysplasia occurs more distally and in proximity to cortical dysplasia.<sup>23,24</sup> Numerous etiologies have been proposed for these lesions, including an arterial insult from a viral infection; congenital defects resulting in dysplastic vessels; chronic healed dissection; somatic mutation; or other inflammatory, immunological, and genetic factors.<sup>3,6,9</sup>

Historically, pure arterial malformations have been described as incidental findings with a benign natural history.<sup>1,6–9</sup> Brinjikji et al.<sup>6</sup> described their experience with these lesions in the largest series to date, consisting of 12 patients. The most common clinical presentation leading to imaging was headache, as reported in 58% of the cohort.<sup>6</sup> Four lesions exhibited an associated focal aneurysm.<sup>6</sup> Only one patient underwent coil embolization of a focal aneurysmal pouch associated with the pure arterial malformation.<sup>6</sup> The remaining 11 patients were managed conservatively.<sup>6</sup> Importantly, none of



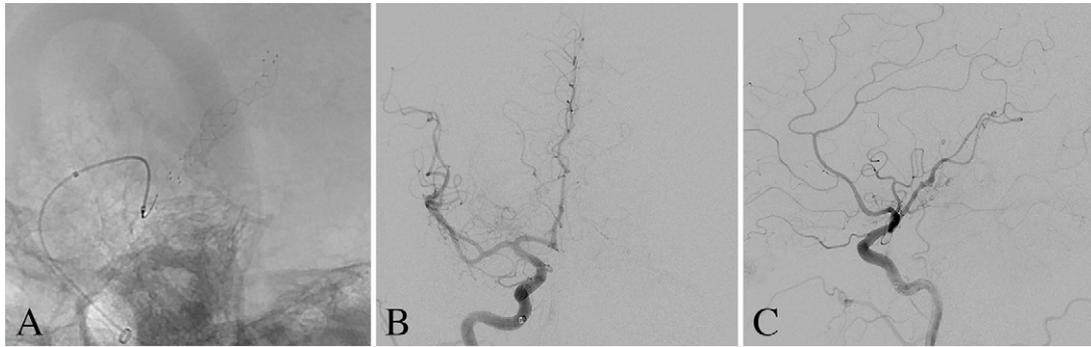
**FIG. 2.** Initial diagnostic cerebral angiograms show an abnormal cluster of dilated vessels with a tight, coil-like appearance off the distal M2 and proximal M3 branches of the right middle cerebral artery. Additionally, the parent M2/M3 branches appear dysplastic with focal aneurysmal outpouchings. Anterior-posterior views (A, B), lateral view (C), oblique views with increased magnification (D–F), and three-dimensional reconstructions (G–I).

the cases were associated with intracranial hemorrhage, infarct, or disability at the mean 29-month follow-up.<sup>6</sup> Moreover, none of the patients within the cohort demonstrated a permanent neurological deficit.<sup>6</sup> Given the relatively low morbidity and absence of complications, the authors concluded that pure arterial malformations are likely associated with a benign natural history.<sup>6</sup> However, other authors have described pure arterial malformations with a more aggressive course complicated by radiographic progression in angioarchitecture, aneurysmal development, or rupture of the parent malformation or associated focal aneurysms.<sup>9–17</sup>

Yue et al.<sup>9</sup> described a case of a 42-year-old male found to have an anterior cerebral artery–associated pure arterial malformation on work-up for frequent headaches. The lesion was deemed noncontributory to the patient’s headaches and was managed conservatively.<sup>9</sup> Three years later, he developed intractable headaches and was found to have bilateral papilledema.<sup>9</sup> Repeat imaging

revealed evolution of the known malformation with two significant structural changes: 1) the development of a giant aneurysm with thrombosis causing obstructive hydrocephalus, and 2) the right distal branches of the anterior cerebral artery were not supplied by the ipsilateral internal carotid artery.<sup>9</sup> The authors secured the aneurysm via clipping and trapping of the parent vessel.<sup>9</sup> Follow-up digital subtraction angiography revealed no residual pure arterial malformation and filling of the right A2 segment from the left internal carotid artery injection via the anterior communicating artery.<sup>9</sup>

Rupture of a pure arterial malformation or associated focal aneurysm is exceedingly uncommon.<sup>11–17</sup> Our English-language MEDLINE online literature review identified only seven cases of a ruptured pure arterial malformation or associated aneurysm (Table 1).<sup>11–17</sup> Of the reported cases, the source of rupture was attributable to a pure arterial malformation vessel in two cases and an associated focal aneurysm in five cases.<sup>11–17</sup> The malformation was localized to the



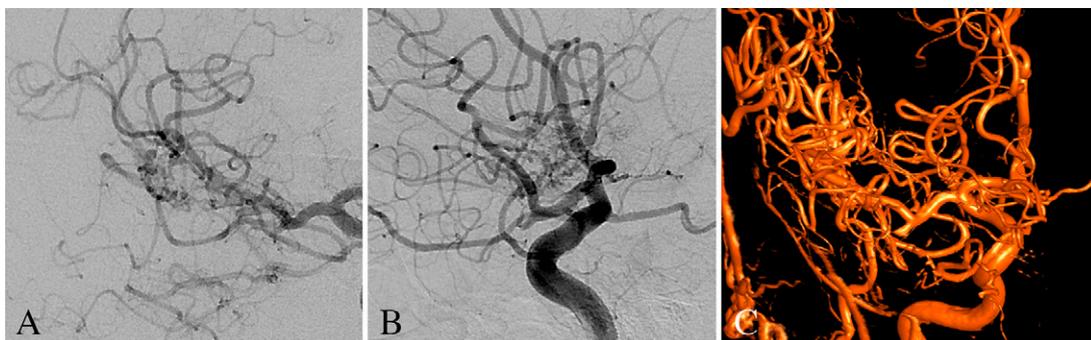
**FIG. 3.** Diagnostic cerebral angiograms obtained subsequent to deployment of the M2 branch flow diverter, showing interval reduction in filling of the pure arterial malformation. Unsubtracted image shows deployment of the flow diverter (A). Anterior-posterior view (B) and lateral view (C) are also shown.

posterior circulation in five cases and anterior circulation in one case.<sup>11,12,14–17</sup> Interestingly, one case reported the coexistence of two pure arterial malformations in the anterior and posterior circulation.<sup>13</sup> The majority of cases underwent intervention (six of seven cases).<sup>11–16</sup> Of the cases treated, surgical intervention (e.g., aneurysm clipping, clip trapping, bypass, parent artery occlusion) was pursued in five of six cases, whereas endovascular ethylene vinyl alcohol copolymer (Onyx) embolization was implemented in one case.<sup>11–16</sup>

First reported in 2014, experience with ruptured pure arterial malformations has remained limited to single-case reports.<sup>11–17</sup> Feliciano et al.<sup>17</sup> described a case of a 42-year-old male who presented with sudden-onset severe headache and left hemiparesis, with imaging demonstrating a right basal ganglia intraparenchymal hemorrhage. Digital subtraction angiography demonstrated a right middle cerebral artery malformation with convoluted and ectatic collateral vessels supplying the distal middle cerebral artery territory with a proximal occlusion as well as an associated medial lenticulostriate artery aneurysm—all of which was characterized as a pure arterial malformation with an associated focal aneurysm.<sup>17</sup> The patient was treated conservatively with resolution of his presenting symptomatology at the 1-year follow-up.<sup>17</sup> Munich et al.<sup>16</sup> described a case of a ruptured basilar perforator aneurysm associated with a

pure arterial malformation managed surgically. Li et al.<sup>14</sup> reported a case of a 77-year-old who presented with sudden-onset headache and was found to have subarachnoid and intraventricular hemorrhage attributable to multiple aneurysmal outpouchings of a distal anterior inferior cerebellar artery pure arterial malformation. The patient underwent craniotomy with trapping of each aneurysmal outpouching within the pure arterial malformation.<sup>14</sup> Moreover, Chua et al.<sup>12</sup> reported a case of a 38-year-old male found to have diffuse subarachnoid hemorrhage with intraventricular extension. Digital subtraction angiography revealed an abnormal tangle of vessels within the right cervicomedullary junction with a dilated coil-like vessel branching from the right posterior inferior cerebellar artery without the presence of early venous drainage consistent with a pure arterial malformation.<sup>12</sup> The authors secured the pure arterial malformation with endovascular Onyx embolization.<sup>12</sup> The details of previously reported cases of ruptured pure arterial malformations or associated focal aneurysms can be found in Table 1.<sup>11–17</sup>

Here, we describe a case of a large symptomatic intraparenchymal hemorrhage with an underlying dysplastic middle cerebral artery dissecting aneurysm and associated pure arterial malformation. Given the presence of hemorrhage, we opted to secure the dissecting aneurysm via endovascular flow diversion. We suspected that the associated aneurysmal component of the pure arterial malformation was



**FIG. 4.** Repeat diagnostic cerebral angiograms at 6 months showing occlusion of the M2 segment just proximal to the deployed flow diverter despite compliance with dual antiplatelet therapy. The dysplastic middle cerebral artery dissecting aneurysm is no longer filling consequent to proximal vessel occlusion. The pure arterial malformation exhibits similar flow and appears unchanged in size compared with the prior angiogram. Lateral view (A), anterior-posterior view (B), and three-dimensional reconstruction (C) are shown.

**TABLE 1. Previously reported cases of ruptured pure arterial malformations**

Authors & Year	Age (yrs)/ Sex	Source of Hemorrhage	Location	Appearance	Symptoms	Imaging	Management	FU	Outcome
Wojtowicz et al., 2022 <sup>11</sup>	65/F	Aneurysm associated w/ PAM	Cervicomedullary junction (PICA)	Tortuous, dilated artery w/ 2 saccular aneurysms	Sudden-onset HA	Diffuse, thick SAH w/ IVH	Surgical (aneurysm clipping)	1 yr	Intact
Chua et al., 2021 <sup>12</sup>	38/M	PAM vessel	Cervicomedullary junction (PICA)	Dilated, "coil-like" artery	HAs & nausea	HH1, FG4 SAH	Endovascular (Onyx embolization)	3 mos	Improving LUE weakness (medullary stroke)
Lu et al., 2021 <sup>13</sup>	53/F	PAM vessel	Supracarotid ICA & PCA	Dilated, tortuous, & redundant vessels, coiled loops	Sudden-onset HA & LOC	SAH in cisterns & Sylvian fissure	Surgical (proximal occlusion & STA-P2 bypass)	1 yr	Near-complete recovery
Li et al., 2020 <sup>14</sup>	77/M	Aneurysm associated w/ PAM	Cerebellomedullary junction (distal AICA)	Collection of tortuous arteries, "coil-like"	Sudden-onset HA, nausea, & confusion	HH3, FG3 SAH	Surgical (clip trapping)	6 wks	Return to baseline
Lai & Patel, 2018 <sup>15</sup>	45/F	Aneurysm associated w/ PAM	PCA	Dilated, tortuous vessel	NR	SAH	Surgical (clipping)	NR	NR
Munich et al., 2019 <sup>16</sup>	NR	Aneurysm associated w/ PAM	BA apex	NR	Partial CN III palsy	Hemorrhage	Surgical (parent artery occlusion)	NR	NR
Feliciano et al., 2014 <sup>17</sup>	42/M	Aneurysm associated w/ PAM	Basal ganglia (MCA)	Convoluted, ectatic vessels providing distal MCA supply	Sudden-onset HA, left-sided weakness, nausea, & vomiting	Basal ganglia IPH	Conservative	1 yr	Return to baseline

AICA = anterior inferior cerebellar artery; BA = basilar artery; CN = cranial nerve; FG = Fisher grade; FU = follow-up; HA = headache; HH = Hunt-Hess grade; ICA = internal carotid artery; IPH = intraparenchymal hemorrhage; IVH = intraventricular hemorrhage; LOC = loss of consciousness; LUE = left upper extremity; MCA = middle cerebral artery; NR = not reported; PAM = pure arterial malformation; PCA = posterior cerebral artery; PICA = posterior inferior cerebellar artery; SAH = subarachnoid hemorrhage; STA = superficial temporal artery.

the etiology of the intraparenchymal hemorrhage. This resembles focal aneurysms coexisting with arteriovenous malformations, which may be responsible for the higher risk of hemorrhage and progression in size. However, in the case series by Brinjikji et al.,<sup>6</sup> 4 of 12 patients exhibited associated focal aneurysms without a corresponding change in radiographic appearance, rupture, or clinical symptomatology at follow-up. Indeed, rupture of a parent pure arterial malformation or associated aneurysm should warrant intervention to mitigate the risk of rerupture. However, it remains controversial whether asymptomatic patients with these lesions should be treated.

## Lessons

Pure arterial malformations remain exceedingly rare cerebrovascular lesions with an unclear natural history. Malformations with associated focal aneurysms may not exhibit a benign natural history as once thought, because recent reports have demonstrated the potential for hemorrhage secondary to rupture of the parent pure arterial malformation vessel or associated focal aneurysm.<sup>11–17</sup> Symptomatic hemorrhage attributable to a ruptured pure arterial malformation or associated focal aneurysm warrants treatment to mitigate the risk of recurrent hemorrhage. However, the risk of rerupture remains unknown. In the absence of hemorrhage, patients with focal aneurysms associated with pure arterial malformations should be closely monitored with interval diagnostic cerebral angiography and contrast-enhanced magnetic resonance imaging with vessel-wall imaging to evaluate for radiographic progression. Further studies are necessary to better understand the natural history of these lesions and determine the threshold for intervention.

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## Disclosures

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

## Author Contributions

Conception and design: Cuoco, Marlow, Entwistle. Acquisition of data: Cuoco, Marlow, Ravina. Analysis and interpretation of data: Cuoco, Marlow, Ravina. Drafting the article: Cuoco, Marlow, Ravina. Critically revising the article: Cuoco, Marlow, Ravina, Sloboda. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Cuoco. Study supervision: Entwistle.

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