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Decompression surgery for pure arterial malformations in a 15 year old with acute, progressive visual impairment: illustrative case

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BACKGROUND The authors document the first case of pure arterial malformations (PAMs) of the posterior communicating artery (PCoA), which were successfully treated with microsurgical clipping of the main body of the PAMs. PAMs are defined as dilated, overlapping, and tortuous arteries with a coil-like appearance and/or a mass of arterial loops without any associated venous component. Although PAMs usually have a benign history and are often incidental findings, this case presented with acute progression of visual field impairment.

OBSERVATIONS Because the patient's right optic tract was affected by the loop of PAMs of the PCoA, the authors performed microsurgical clipping of the main body of the PAMs using endoscopy, which ceased the progression of symptoms without any complications.

LESSONS There have been several reports of PAMs receiving surgical treatment for accompanying lesions. However, in this case, the lesion to the main body of PAMs was the cause of visual field impairment and was successfully treated with microsurgical clipping.

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KEYWORDS pure arterial malformations; progressive symptoms; posterior communicating artery; decompression surgery

In 2013, McLaughlin et al. defined pure arterial malformations (PAMs) as dilated, overlapping, and tortuous arteries forming a mass of arterial loops with a coil-like appearance in the absence of any arteriovenous connection.[1] The majority of PAMs have a benign clinical course and are treated conservatively.[2] Several cases of PAMs have been treated surgically,[3–6] but most surgically treated cases were those involving aneurysms accompanied by PAMs.[4–6] Here, we describe for the first time a case of a PAM with acute progressive symptoms that warranted surgical treatment targeting the main body of the PAM instead of any accompanying lesions.

Illustrative Case

History and Examination

A 15-year-old boy with narrowing of the visual field, which had been progressing for a year, was referred to our hospital. He and his family had no history of cerebrovascular disease, and he was not receiving any medication. A cranial computed tomography (CT) scan showed abnormal vessels in the left interpeduncular cistern, which was suspected to reflect an arteriovenous malformation (AVM). He was referred to our hospital for evaluation of the lesions.

The patient's neurological examination and visual field test done via Goldmann perimetry revealed left homonymous hemianopsia (Fig. 1C). A cranial CT angiogram showed a dilated, tortuous, and coil-like vessel at the right P1 and P2 segments of the posterior cerebral artery (PCA) and posterior communicating artery (PCoA) (Fig. 1A and B). A digital subtraction angiography (DSA) revealed a dilated and coil-like right PCoA and tortuous PCA without arteriovenous shunting, which was diagnosed as a PAM (Fig. 2). Constructive interference in steady-state (CISS) magnetic resonance imaging (MRI) and three-dimensional (3D) fusion images, with MRI and DSA, showed that the abnormal vessels were compressed in the right optic tract (Fig. 3A and B). These findings led us to consider his abnormal angioarchitecture to be the cause of his visual disturbance.

Treatment

At first, the patient and his parents opted for a conservative approach, without surgical treatment. However, the patient subsequently

ABBREVIATIONS 3D = three-dimensional; AVM = arteriovenous malformation; CT = computed tomography; CISS = constructive interference in steady-state; DSA = digital subtraction angiography; ICA = internal carotid artery; MCA = middle cerebral artery; MRI = magnetic resonance imaging; PAM = pure arterial malformation; PCA = posterior cerebral artery; PCoA = posterior communicating artery.

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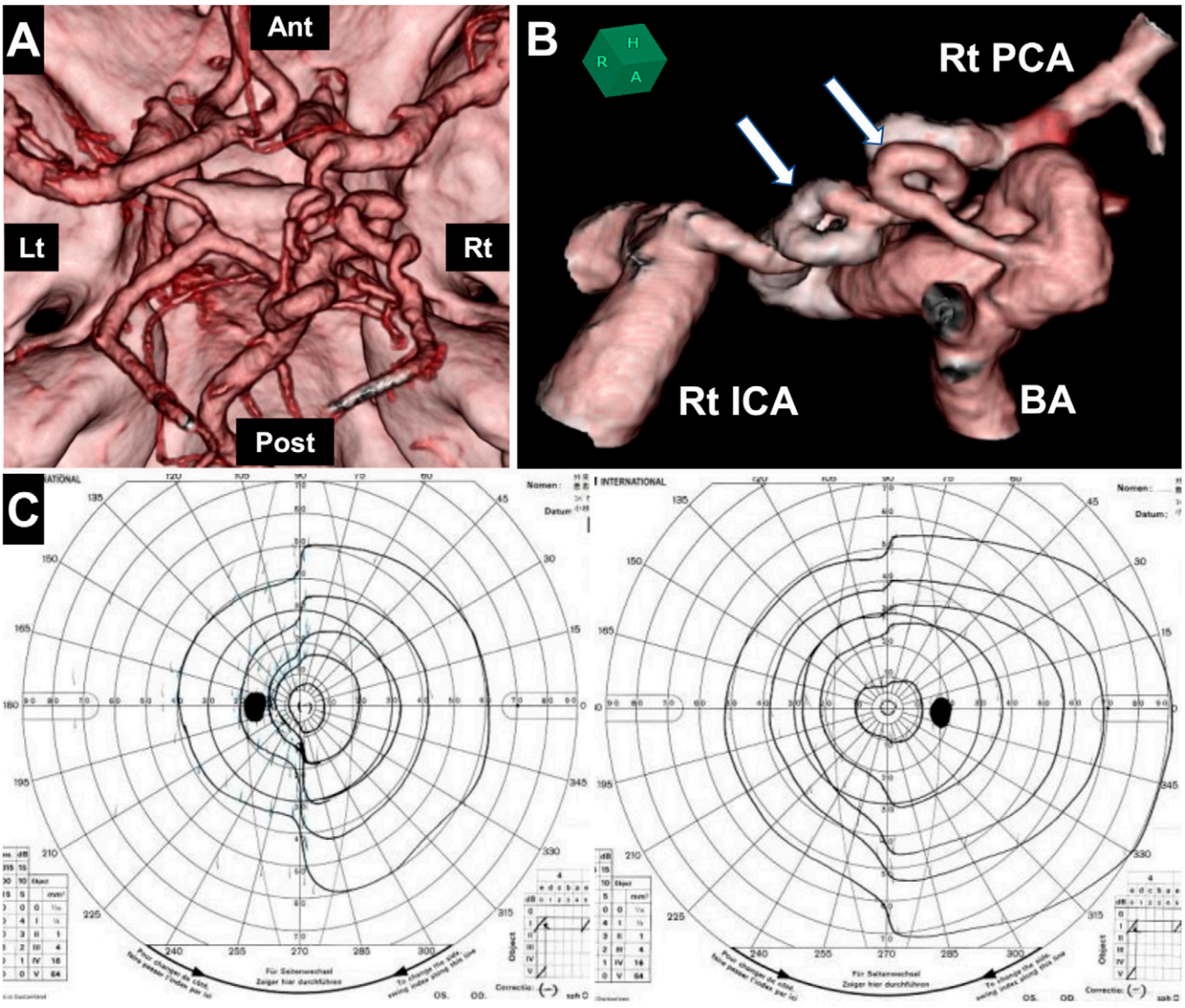

FIG. 1. 3D CT angiograms and visual field test by Goldmann perimetry at first visit. **A:** Whole arteries, as seen from above. **B:** The lesions seen on the right side. The right PCoA has double loops (*arrows*). **C:** The left eye (*left*) and the right eye (*right*). Visual field test at the first visit showed a left homonymous hemianopsia. Ant = anterior; BA = basilar artery; Post = posterior.

opted for surgical management 3 months later because of worsening symptoms (Fig. 3C).

For the procedure, the patient was placed in a supine position, and a right fronto-parieto-temporal craniotomy was performed. The right internal carotid artery (ICA), PCoA, and right optic tract were exposed using a transsylvian approach (Fig. 4A). We confirmed that the loop of the PCoA affected the right optic tract by endoscopy (Fig. 4B). The PAM was challenging to mobilize, but we managed to clip the PCoA to decompress the PAM (Fig. 4C and D). Because the motor evoked potentials monitoring remained unchanged with clipping of the PCoA distal to the perforating arteries, we performed complete clipping of the PCoA. The right optic tract was successfully decompressed without any signs of neurological worsening (Fig. 4E and F). The patient's postoperative course was uneventful, with the cessation of symptom progression. The patient underwent MRI follow-ups the next day and at three months, a year, and two years after surgery and DSA three months after surgery. No hemorrhagic or ischemic lesions appeared, and loops of the right PCoA were not seen. Though complete decompression of the optic tract was made, the patient has not fully regained intact visual fields yet. We think it is long-term compression. However, his field of vision gradually improved (Fig. 4G), and he felt no inconvenience in daily life by his latest follow-up.

Discussion

Observations

We document the case of a patient with acute progressive symptoms caused by a PAM of the PCoA requiring decompressive surgery. PAMs can be mistaken for an AVM, arteriovenous fistula, intracranial dolichoectasia, or intracranial arterial dissection.[2] However, they can be distinguished by DSA with the absence or presence of

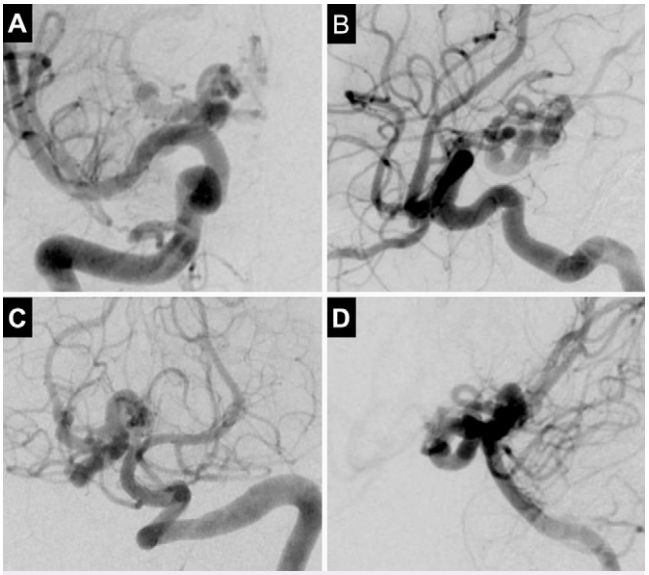

FIG. 2. Catheter angiograms. **A:** Anteroposterior view of right ICA angiogram. **B:** Right lateral view of right ICA angiogram. **C:** Anteroposterior view of the left vertebral artery angiogram. **D:** Right lateral view of the left vertebral artery angiogram. The patient's right PCoA and PCA had a tortuous, dilated, and coil-like appearance without arteriovenous shunting.

an arteriovenous shunt or nidus. Our case was also distinct from dolichoectasia because the present lesions were redundant to the point of looking like a mass of arterial loops with a coil-like appearance. Moreover, patients with dolichoectasia tend to be older than those with PAMs; they also have a history of cardiovascular risk factors. Because several examinations showed that the walls of abnormal vessels in our case remained parallel within the tortuous segment, we were able to distinguish the present case from those of dissections, which was confirmed intraoperatively.

Table 1 describes the case list of definitive and probable PAMs[1–26] before and after 2013, when PAMs were first defined by McLaughlin et al.[1] Previous reports have shown that PAMs are usually asymptomatic, have a benign course, and are treated using observation. In our review of previous reports, there were only three cases (7.7%) wherein the patient was symptomatic. Only four cases (10.3%) were treated surgically. Still, two of them did not present with any progressive symptoms preoperatively, such that it remained unclear whether invasive treatment was necessary. One is a case of a PAM in the right ICA, middle cerebral artery (MCA), and left PCA, but the surgery was an external carotid artery–ICA bypass for moyamoya disease.[3] The other is a case of a PAM in the left PCA, and the operation was coiling for a focal aneurysm pouch.[4] But the only symptom was headache, and it was not certain it was caused by lesions. The remaining two cases both had symptoms that occurred by aneurysms accompanied with PAMs, and they underwent surgical treatment. One had an unruptured giant aneurysm of the anterior cerebral artery that got trapped by the parent artery, causing obstructive hydrocephalus,[5] whereas the other had ruptured aneurysms of the basilar apex clipped.[6] Our case was different in that it presented with acute progressive symptoms without an aneurysm portion of the PAM, and his symptoms were due to the main body of lesions affecting the right optic tract, requiring decompressive surgery.

Although PAMs usually have a benign natural history, aneurysms associated with the lesions have the potential to grow and rupture,[5,6] such that patients need to be followed up periodically. Moreover, as in our case, a PAM located close to cranial nerves can cause progressive neurological symptoms despite the absence of an aneurysm, necessitating timely surgery.

Lessons

To the best of our knowledge, we report the first case of a PAM in which the main body caused progressive symptoms, requiring decompressive surgery with the assistance of an endoscope. PAMs typically have an uneventful clinical course. However, if there are advanced neurological symptoms associated with lesions, surgical intervention should be considered.

We also present clear intraoperative endoscopic pictures of a PAM affecting the patient's right optic tract. This is the first picture of a PAM captured by an endoscope.

Acknowledgments

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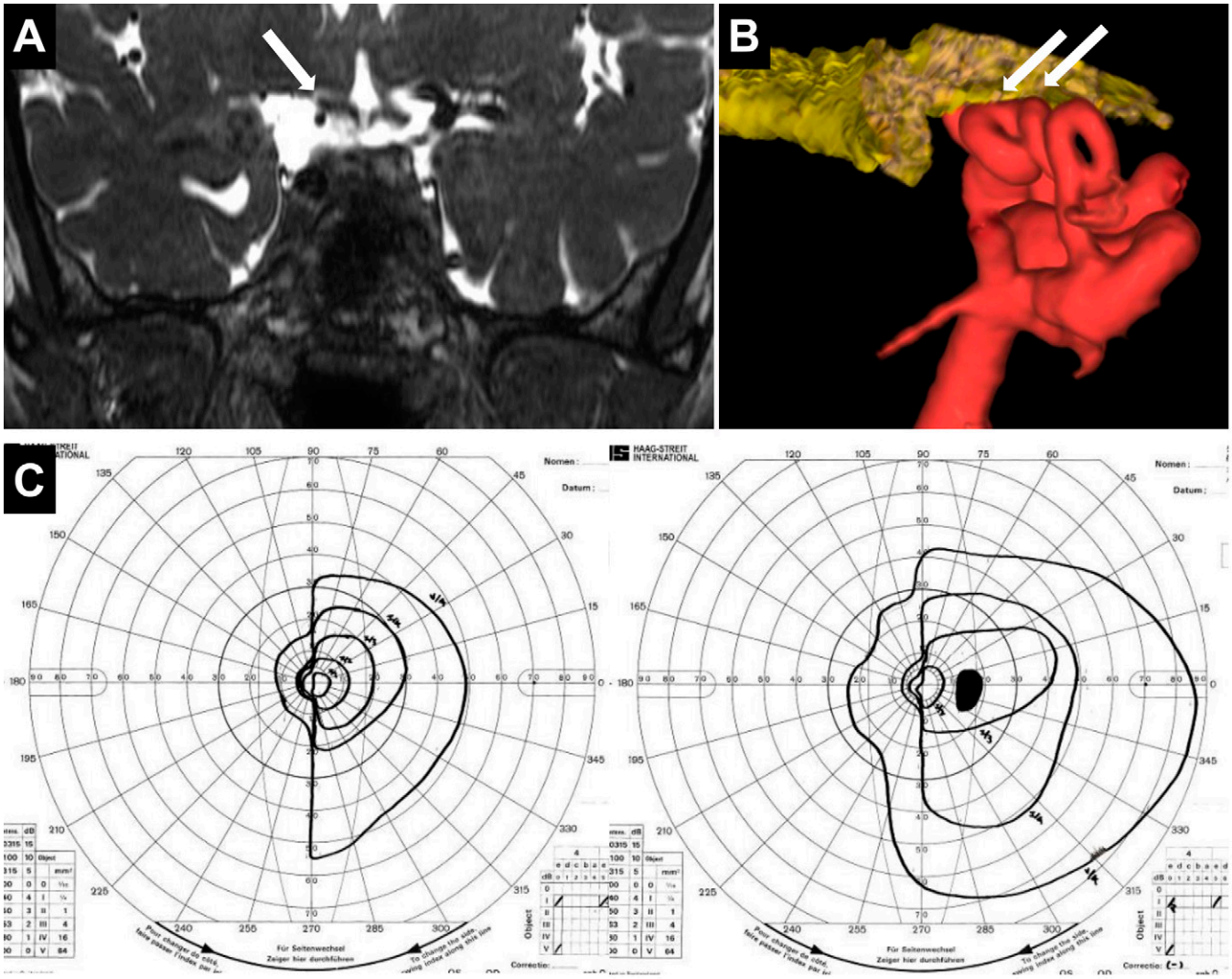

FIG. 3. A: Coronal view of CISS MRI. **B:** 3D image created using CISS images. The yellow fiber is the optic nerve. The loops of the right PCoA were touching the undercompartment of the right optic tract (*arrows*, **A** and **B**). **C:** Visual field test three months after the first visit. The symptoms progressed.

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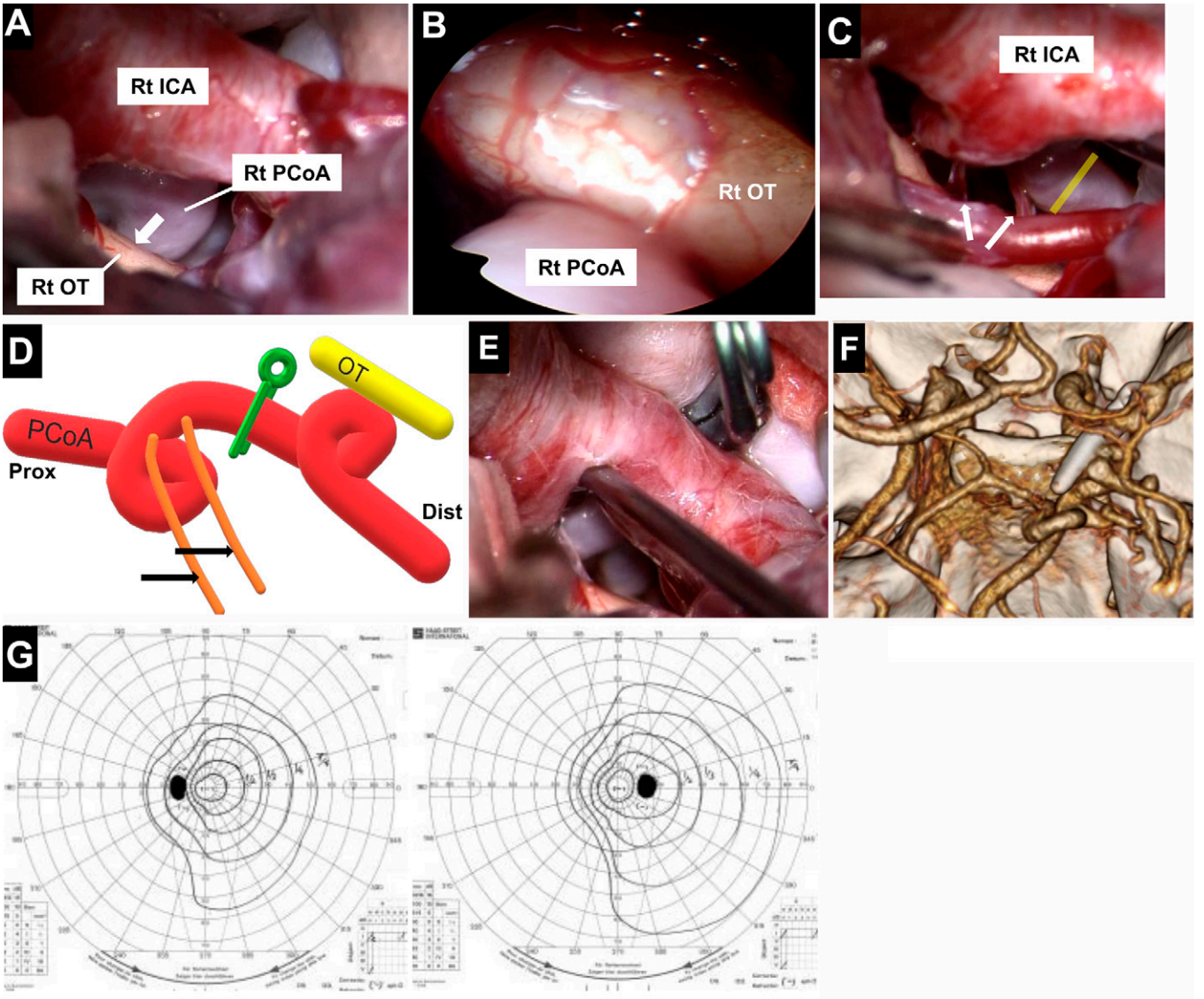

FIG. 4. A: Microscopic view before clipping. The endoscope was placed at the position of the *white arrow*. **B:** Endoscopic view demonstrating the right PCoA compressing the right OT. **C:** Microscopic view before clipping. *White arrows* show perforating arteries from the PCoA. The *yellow line* shows the clipping line. **D:** Clipping schema. We clipped the right PCoA distal to the perforating arteries (*black arrows*). **E:** Microscopic view after clipping. Right PCoA separated from the right OT. **F:** 3D CT angiogram after surgery. The loops of the right PCoA were not seen. **G:** Visual field test at 24 months after surgery. The visual field impairment, especially of the left eye, was gradually improving. Dist = distal; Prox = proximal; OT = optic tract.

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: Arimura, Iwaki, Iihara. Acquisition of data: Iwaki. Analysis and interpretation of data: Arimura, Iwaki. Drafting the article: Iwaki, Nishimura. Critically revising the article: Iwaki. Reviewed submitted version of manuscript: Arimura, Iwaki. Approved the final version of the manuscript on behalf of all authors: Arimura. Administrative/technical/material support: Iwaki. Study supervision: Iihara.

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TABLE 1. Literature review of pure arterial malformations

Case No.	Authors & Year	Age (yrs), Sex	Location	Symptoms	Appearance	Surgical Treatment for PAM	Clinical Follow-up
1	Sacks et al., 1969[7]	2, M	Bilat A2 segment of ACA	Viral encephalitis	Tightly coiled, moderately dilated	None	None
2	Wolpert et al., 1972[8]	21, M	Bilat pericallosal artery	Seizure	Moderately coiled, tortuous, and calcified	None	None
3	Thompson et al., 1976[9]	39, M	Distal lt ACA	Seizure	Enlarged, moderately tortuous ACA and pericallosal artery	None	3 yrs, no change
4	Kryst-Widźgowska et al., 1980[10]	72, F	Distal bilat ACA	Aphasia, rt-sided hemiplegia (infarction)	Dolichoectasia, moderately tortuous	None	None
5	Tsukamoto et al., 1985[11]	37, F	Bilat pericallosal artery	Mania	Moderately coiled, dilated, and calcified	None	None
6	Yamada et al., 1985[12]	17, F	Lt supraclinoid ICA, MCA, and ACA	Nausea, vomiting	Tightly coiled, moderately ectatic cluster of vessels, and calcified	None	None
7	Yamada et al., 1985[12]	40, F	Rt supraclinoid ICA, MCA, and ACA	Rt-sided hemiparesis	Tightly coiled, moderately ectatic cluster of vessels, calcified	None	None
8	Hanakita et al., 1986[3]	43, F	Rt distal ICA, proximal MCA, and lt PCA	Dysarthria	Tightly coiled, dilated vessels, and stenotic lesion of MCA	EC-IC bypass and wrapped ectatic MCA with muscle	None
9	Araki et al., 1987[13]	25, F	Rt MCA, ACA, and PCA	Rt hemimegalencephaly	Tightly coiled MCA and generalized ectasia of distal vasculature in rt hemisphere	None	None
10	Doran et al., 1995[14]	14, F	Bilat pre- and supracallosal segments of ACAs	Seizure	Tightly coiled, moderately dilated, calcified, thickening of medial frontal lobes, and delayed washout	None	None
11	Abe et al., 1997[15]	32, M	Sylvian branches of lt MCA	Cortical dysplasia, seizure	Plexiform arterial network with tortuous vessels	None	None
12	Kanemoto et al., 1998[16]	41, F	Lt MCA	Seizure (ipsilateral cavernoma)	Tightly coiled, moderately dilated, and elongated	None	None
13	Vanslambrouk et al., 2000[17]	5, M	Lt ICA, PCoA, PCA, MCA, and lt SCA	Minimal rt hemiparesis (brainstem compression)	Tightly coiled and moderately dilated	None	None
14	Metry et al., 2001[18]	1, F	Lt MCA and supraclinoid ICA	PHACE syndrome	Tightly coiled and markedly dilated	None	None
15	Uchino et al., 2003[19]	35, F	Rt SCA	Severe headache	Tightly coiled and moderately dilated	None	2 yrs, no change
16	Beringer et al., 2004[20]	49, M	Bilat pericallosal	Intermittent frontal headache	Tightly coiled, mildly dilated, calcified, and associated stenosis	None	Several mos, no change
17	Baccin et al., 2007[21]	4, F	Lt supraclinoid ICA and PCoA	PHACE syndrome and rt-sided hemiparesis (infarction)	Tightly coiled and markedly dilated	None	16 mos, no change
18	Baccin et al., 2007[21]	1, F	Lt supraclinoid ICA, PCoA, P1, and rt supraclinoid ICA	PHACE syndrome, fever, and hypotonia	Tightly coiled and markedly dilated	None	None

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TABLE 1. Literature review of pure arterial malformations

Case No.	Authors & Year	Age (yrs), Sex	Location	Symptoms	Appearance	Surgical Treatment for PAM	Clinical Follow-up
19	Shankar et al., 2009[22]	26, F	Distal rt PCA	Incidental	Tightly coiled, not dilated, and associated with cortical dysplasia	None	None
20	McLaughlin et al., 2013[1]	24, F	Lt PCoA and P2 with saccular aneurysm	Frequent headaches and dizziness	Tightly coiled, moderately dilated, and focal saccular aneurysm	None	30 yrs, no change
21	McLaughlin et al., 2013[1]	8, F	Lt supraclinoid ICA and proximal M1	Sinus infection	Tightly coiled, dilated vessel, and focal aneurysm	None	None
22	Lanterna et al., 2014[23]	1, M	Lt PCoA and PCA	Infarct from moyamoya disease	Tightly coiled, dilated vessel, and associated with ipsilateral moyamoya disease	None	None
23	Feliciano et al., 2014[24]	42, M	Rt MCA	Headache with basal ganglia hemorrhage	Markedly ectatic distal M1 with superimposed cluster of aneurysms	None	1 yr, no change
24	Sako et al., 2016[25]	35, M	Lt PICA	Vertigo	Tightly coiled distal PICA	None	6 mos, no change
25	Sorenson et al., 2018[26]	17, F	Proximal lt PICA	Migraine	Coil-like configuration in its proximal portion and ectatic	None	None
26	Brinjiki et al., 2018[2]	10, F	Lt supraclinoid ICA, PCoA, and PCA	Severe lt-sided headaches	Multilobulated pseudoaneurysm of supraclinoid ICA, partially calcified, coil-like tortuosity, dilatation of PCoA, PCA and stenosis of lt M1	Coil embolization of a focal aneurysm pouch	72 mos, no change
27	Brinjiki et al., 2018[2]	19, F	Lt MCA	Incidental	Coil-like tortuosity of distal lt MCA lenticulostriate vessel with superimposed multilobulated aneurysm, mild preceding stenosis	None	36 mos, no change
28	Brinjiki et al., 2018[2]	27, F	BA	Headache, lt hemibody numbness, and facial droop	Tortuous BA, tightly wound, mildly dilated, and no focal aneurysmal dilatation	None	5 mos, no change
29	Brinjiki et al., 2018[2]	25, F	Lt supraclinoid ICA and MCA	Headache	Tortuous, coil-like appearance of supraclinoid ICA, M1 with 3 focal aneurysms, and calcified	None	60 mos, no change
30	Brinjiki et al., 2018[2]	25, F	Lt ACA	Headache after minor trauma	Tortuous A2 and mildly dilated	None	30 mos, no change
31	Brinjiki et al., 2018[2]	34, F	Lt ACA	Light trauma	Tortuous A2, mildly dilated, and calcified	None	12 mos, no change
32	Brinjiki et al., 2018[2]	38, F	Lt PICA	Transient hand numbness	Tortuous and tightly coiled	None	2 mos, no change
33	Brinjiki et al., 2018[2]	11, M	Lt PCoA	Incidental	Tortuous, coil-like appearance of supraclinoid ICA and PCoA	None	1 mo, no change
34	Brinjiki et al., 2018[2]	17, M	Rt SCA	Headache	Tortuous SCA, tightly wound, moderately dilated, and no focal aneurysmal outpouching	None	26 mos, no change
35	Brinjiki et al., 2018[2]	47, F	Rt ACA	Prior thunderclap headache	Tortuous artery, mildly dilated, partially calcified, and delayed venous drainage	None	27 mos, no change

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TABLE 1. Literature review of pure arterial malformations

Case No.	Authors & Year	Age (yrs), Sex	Location	Symptoms	Appearance	Surgical Treatment for PAM	Clinical Follow-up
36	Brinjiki et al., 2018[2]	35, F	Lt PCoA and PCA	Headache	Arterial tortuosity with aneurysm dilatations and calcified	None	84 mos, no change
37	Brinjiki et al., 2018[2]	20, F	Rt ICA, PCA, and lt PCA	Trauma	Tortuous and tightly coiled	None	1 month, no change
38	Yue et al., 2019[5]	45, M	Proximal rt ACA	Headache, vomiting	Dilated, distorted, and tortuous artery vessels with some aneurysmal structures inside	Surgical trapping	3 mos, no change
39	Munich et al., 2019[6]	37, F	BA apex	Partial CNIII palsy (ruptured aneurysm)	Dilated, tortuous BA	Clipping of aneurysm	None
40	Our case	15, M	Rt PCoA and PCA	Lt homonymous hemianopsia	Dilated, tortuous, and coil-like rt P1 and P2 segment of PCA and PCoA	Clipping of loop of PCoA, decompression of optic tract	2 yrs, symptoms improved

ACA = anterior cerebral artery; BA = basilar artery; CNIII = third cranial nerve; EC-IC = extracranial-intracranial; PHACE = posterior fossa malformations, hemangiomas, arterial anomalies, coarctation of the aorta and other cardiac defects, and eye abnormalities; PICA = posterior inferior cerebellar artery; SCA = superior cerebellar artery.