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...
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. However, they can be distinguished by DSA with the absence or presence of ischemic or hemorrhagic lesions.
an arteriovenous shunt or nusid. 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 before and after 2013, when PAMs were first defined by McLaughlin et al. 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. The other is a case of a PAM in the left PCA, and the operation was coiling for a focal aneurysm pouch. 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, whereas the other had ruptured aneurysms of the basilar apex clipped. 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, 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.

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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.
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.

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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.
| 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-Widzgowska 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, and 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 suprACLINIC 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 suprACLINIC 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 suprACLINIC ICA, PCoA, P1, and rt suprACLINIC ICA | PHACE syndrome, fever, and hypotonia | Tightly coiled and markedly dilated | None | None |

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| Case No. | Authors & Year | Age (yrs), Sex | Location | Symptoms | Appearance | Surgical Treatment for PAM | Clinical Follow-up |
|----------|----------------|----------------|----------|----------|------------|---------------------------|-------------------|
| 19       | Shankar et al., 2009\(^2\) | 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\(^2\) | 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\(^4\) | 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\(^2\) | 35, M | Lt PICA | Vertigo | Tightly coiled distal PICA | None | 6 mos, no change |
| 25       | Sorenson et al., 2016\(^2\) | 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 | 35, F | Lt PCoA and PCA | Headache | Arterial tortuosity with aneurysm dilatations and calcified | None | 84 mos, no change |
| 37       | Brinjiki et al., 2018 | 20, F | Rt ICA, PCA, and Lt PCA | Trauma | Tortuous and tightly coiled | None | 1 month, no change |
| 38       | Yue et al., 2019 | 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 | 37, F | BA apex (ruptured aneurysm) | Partial CNIII palsy | 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.