Coexistence of anterior choroidal artery and posterior cerebral artery retia mirabilia presenting with subarachnoid hemorrhage: illustrative case

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BACKGROUND A rete mirabile is a rare vascular anomaly, with posterior cerebral artery (PCA) involvement being especially rare. Its pathogenesis has been speculated as a remnant of “distal annexation” between the primitive anterior choroidal artery (AchA) and the PCA at this site, but the exact mechanisms remain unclear.

OBSERVATIONS A 29-year-old man presented with subarachnoid hemorrhage. Arteriovenous malformation in the medial temporal lobe was initially suspected, but an arteriovenous shunt was not detected. First, conservative treatment was administered; however, rebleeding occurred 1 month later. Carotid angiography revealed a network-like cluster of blood vessels at the choroidal point of the AchA, suggesting a rete mirabile; these vessel clusters led to the persistent temporo-occipital branch of the AchA. Furthermore, an aneurysm was detected at the junction between the rete mirabile and the persistent temporo-occipital branch of the AchA. Additionally, vertebral angiography demonstrated a rete mirabile at the P2 segment. These findings suggested the coexistence of AchA and PCA retia mirabilia. Consequently, the aneurysm was clipped using a subtemporal approach to prevent re-rupture, and the postoperative course was uneventful.

LESSONS This first report of coexisting AchA and PCA retia mirabilia supports the remnant of distal annexation between the primitive AchA and the PCA as the reason for rete formation at this site.

https://thejns.org/doi/abs/10.3171/CASE23580

KEYWORDS rete mirabile; arteriovenous malformation; cerebral aneurysm; posterior cerebral artery; anterior choroidal artery; pure arterial malformation

A rete mirabile (“wonderful net” in Latin) is a complex arterial network primarily found in lower mammals, such as pigs, cows, goats, cats, and sheep. The purpose of this distinctive vascular architecture is believed to be related to thermoregulation, pressure absorption, or blood flow regulation. However, rete mirabile is a rare vascular anomaly that does not exist in humans during normal development. The most accepted mechanism suggests that a rete mirabile results from embryonal agenesis, regression, or annexation of normal vessels, which help to compensate for the blood flow. However, the exact mechanisms have not yet been established. A rete mirabile commonly involves the internal carotid and vertebral arteries, which have been described as carotid rete mirabile and vertebral rete mirabile, respectively. A rete mirabile of the posterior cerebral artery (PCA) is an extremely rare lesion that has been previously reported in only two cases. Moreover, the PCA and anterior choroidal artery (AchA) are closely related developmentally. The proximal segment of PCA (P1) is derived from the posterior division of the internal carotid artery (ICA). In contrast, the distal segment of the PCA (P2–4) is annexed from the primitive AchA in the anterior division of the ICA during the embryonic stage. This process is called “distal annexation.” The pathogenesis of PCA rete formation has been hypothesized to be the incidence of a remnant of distal annexation between the primitive AchA and the PCA during embryogenesis. To our knowledge, this is the first report of coexisting AchA and PCA retia mirabilia, supporting the remnant of distal annexation between the primitive AchA and the PCA as the reason for rete formation at this site.
the remnant of distal annexation between the primitive AchA and the PCA as the underlying pathogenesis of rete formation at this site.

**Illustrative Case**

A 29-year-old man with severe headache was transferred to our hospital for subarachnoid hemorrhage. He had no neurological deficit and had clear consciousness (World Federation of Neurosurgical Societies grade I). Brain computed tomography (CT) showed a diffuse subarachnoid hemorrhage of Fisher grade 3 (Fig. 1A); CT angiography showed an abnormal cluster of blood vessels at the choroidal point of the AchA and the P2 segment of the PCA (Fig. 1B). An arteriovenous malformation (AVM) of the medial temporal lobe was initially suspected. Although there were clusters of blood vessels like a nidus, we could not detect an apparent arteriovenous shunt from the initial cerebral angiography. Subsequently, conservative treatment was performed, and a follow-up angiography was scheduled to understand the correct vessel architecture. However, 1 month later, the patient’s headache returned with a sudden onset after the initial hemorrhage. Brain CT showed a fresh hematoma at the right ambient cistern, suggesting rebleeding (Fig. 1C and D).

Consequently, follow-up cerebral angiography was performed, which demonstrated no arteriovenous shunt indicating an AVM. Carotid angiography revealed a network-like cluster of blood vessels at the choroidal point of the AchA, suggesting a rete mirabile; this network-like cluster of blood vessels led to the persistent temporo-occipital branch (Fig. 2). Furthermore, a small aneurysm was found at the junction between the rete mirabile and persistent temporo-occipital branch on the AchA (Fig. 2). The tiny posterior communicating artery was bifurcated at the proximal portion of the AchA (Fig. 2B). In addition, vertebral angiography revealed the network-like cluster of blood vessels, suggesting the occurrence of a rete mirabile at the P2 segment (Fig. 3). The peripheral branch of the original PCA was almost replaced by the AchA, which suggests hyperplastic AchA type 4 according to the Takahashi classification. These results suggested the coexistence of AchA and PCA retia mirabilia with flow-related aneurysms.

The subarachnoid hemorrhage was suspected to be caused by a re-rupture of the rete mirabile–associated aneurysm. Therefore, a temporoparietal craniotomy via the subtemporal approach was performed in a hybrid operating room. The rete mirabile–associated aneurysm was clipped to prevent re-rupture (Fig. 4A and B). There were no complications associated with the surgical procedure. Moreover, intraprocedural and postoperative angiography revealed the disappearance of the aneurysm (Fig. 4C and D). The patient had no complications postoperatively and was discharged on postoperative day 17 with a modified Rankin scale score of 0. No recurrence was observed during the 6-month follow-up.

**Patient Informed Consent**

The necessary patient informed consent was obtained in this study.

**Discussion**

**Observations**

To our knowledge, this is the first report of coexisting AchA and PCA retia mirabilia, supporting the remnant of distal annexation
between the primitive AchA and the PCA as the reason for rete formation at this site.

The term “rete mirabile” is used to describe multiple arteries or arterioles that freely interconnect and reconstitute the absent or hypoplastic segments of the parent artery. A rete mirabile usually exists in lower mammals, such as pigs, cows, goats, cats, and sheep. It does not exist in humans during normal development and is a rare vascular anomaly. Angiographically, the prevalence of rete mirabile is approximately 0.01% in humans, and most cases have been reported in Asia. A rete mirabile involving the ICA, which is the most common location, has been classically described as a carotid rete mirabile. With regard to the posterior circulation, retia mirabilia involving the vertebral artery, PCA, and posterior inferior cerebellar artery have been previously reported. In the two cases of PCA rete mirabile reported earlier, one was asymptomatic, whereas the patient in the other case died from subarachnoid hemorrhage due to rete mirabile-associated aneurysm. The primitive AchA has cortical branches to the temporal, parietal, and occipital lobes in the early embryonic stage that transform into the P2–4 segments of the PCA distal to the posterior communicating artery. This process is described as distal annexation. Hyperplastic AchA, which has cortical branches, is a rare anomaly due to incomplete distal annexation between the primitive AchA and the PCA. Zheng et al. reported on a PCA rete mirabile; the persistence of the embryonic anastomosis between the primitive AchA and the PCA was speculated to be the cause. In our case, retia mirabilia coexisted in the AchA and PCA, supporting a remnant of distal annexation between the primitive AchA and PCA.

The most common presentation of the rete mirabile is subarachnoid hemorrhage (34.2%), followed by incidental findings (20%). Subarachnoid hemorrhage is caused by a rete mirabile–associated aneurysm, a fragile rete mirabile vessel, and hemodynamic stress. Treatment of a ruptured aneurysm associated with a rete mirabile is challenging. Open surgery for aneurysm clipping has a risk of bleeding from fragile vessels of the rete mirabile. Furthermore, endovascular access is often restricted because of the complicated vascular structure of the rete mirabile, as in our case. The rebleeding rate of rete mirabile–associated aneurysms remains unknown. In our case, rebleeding occurred 1 month after the first bleeding, and we performed a surgical intervention for the aneurysm.

Notably, a rete mirabile can be misdiagnosed as an AVM. The vessel structure of a rete mirabile resembles the nidus of an AVM. In addition, the rete mirabile of pigs has been used as an animal model of AVM in many studies. The rete mirabile is an arterial network that perfuses normal brain tissue and has no arteriovenous shunt. Moreover, resection or occlusion of a rete mirabile can cause ischemia in that perfusion area. Therefore, cerebral angiography studies must be read carefully to avoid misdiagnosing the rete mirabile and AVM. The use of super-selective angiography may help in distinguishing the presence of an arteriovenous shunt. Pure arterial malformation was defined as dilated, overlapping, and tortuous arteries forming a mass of arterial loops with a coil-like configuration in the absence of any venous component. Pure arterial malformation involves all segments of the intracranial artery, and it also needs to be differentiated from an AVM and a rete mirabile. The natural history of a pure arterial malformation is presumably benign, and conservative management may be recommended. Notably, the rete mirabile has a complex arterial network composed of smaller vessels than those involved in pure arterial malformation. In our case, cerebral angiography showed network-like clusters composed of small vessels. Furthermore, unlike in a pure arterial malformation,
vessel dilation and overlapping arteries were not apparent. Thus, our findings are inconsistent with those characterizing pure arterial malformation. However, further studies are warranted to clearly distinguish rete mirabile from pure arterial malformation.

A rete mirabile can be associated with a cerebral aneurysm, spinal aneurysm, carotid-cavernous fistula, pial arteriovenous fistula, AVM, vein of Galen aneurysmal malformation, pseudoxanthoma elasticum, PHACE (posterior fossa anomalies, hemangiomas, arterial anomalies, cardiac anomalies, and eye anomalies) syndrome, vertebral artery stenosis, and Dieulafoy’s ulcer. Previous reports have shown that a rete mirabile was associated with cerebral aneurysms in 20% of cases. Hemodynamic stress from complex vessel architecture of the rete mirabile may induce flow-related aneurysm formation. Therefore, follow-up involving brain imaging should be recommended if a rete mirabile is diagnosed. Recently, four-dimensional flow magnetic resonance imaging has been reported to help visualize and evaluate flow-related aneurysms of rete mirabile networks.

Lessons
This is the first report of coexisting AchA and PCA retia mirabilia, supporting the remnant of distal annexation between the primitive AchA and PCA as the reason for rete formation at this site.

References

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: Yamaguchi, Kanematsu, Teshima. Acquisition of data: Yamaguchi, Shimada, Korai, Sogabe, Teshima, Suzuki. Analysis and interpretation of data: Yamaguchi, Miyake, Sogabe. Drafting of the article: Yamaguchi. Critically revising the article: Yamaguchi, Takagi. Reviewed submitted version of the manuscript: Yamaguchi, Ishihara, Haboshi, Suzuki, Takagi. Approved the final version of the manuscript on behalf of all authors: Yamaguchi. Study supervision: Takagi.

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