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Bilateral Carotid and Vertebral Rete Mirabile Presenting with Subarachnoid Hemorrhage Caused by the Rupture of Spinal Artery Aneurysm

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Rete mirabile (or carotid rete) is a normal structure that plays physiological roles in the lower mammals. However, the rete does not exist in the normal carotid circulation of humans. Carotid rete mirabile (CRM) is a rare condition compensating for congenital dysplastic internal carotid artery. Arterial plexus at the cavernous region, which supplies intradural internal carotid artery instead of the aplastic cavernous portion of internal carotid artery, looks like the "rete mirabile" seen in the lower mammals, and is a characteristic angiographical finding of CRM. In addition to the CRM, existence of segmental occlusion and tortuous collaterals of vertebral artery, so-called carotid and vertebral rete mirabile (CVRM), is a very rare condition. We report a 70-year-old female patient with bilateral CVRM presenting with subarachnoid hemorrhage (SAH) caused by the rupture of a cervical spinal artery aneurysm. Our patient is the oldest, compared with the previously reported four patients with CVRM. Moreover, this is the first report of ruptured spinal artery aneurysm as a cause of SAH associated with CRM/CVRM. To avoid rebleeding in the patient, we successfully treated the patients by performing coil embolization of the remaining spinal aneurysms. In patients with CVRM, aneurysm formation of the cervical spinal artery may be a reasonable consequence because of the hemodynamic stress on the spinal artery as a collateral pathway. Detailed evaluation of the cervical spinal arteries should be performed to detect or to rule out ruptured aneurysm in patients with SAH associated with CVRM.

Keywords: internal carotid artery; rete mirabile; subarachnoid hemorrhage; vascular anomaly; vertebral artery
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Introduction

Rete mirabile (or carotid rete) is a normal structure that plays physiological roles in the lower mammals. However, the rete does not exist in the normal carotid circulation of humans. Carotid rete mirabile (CRM) is a rare condition compensating for congenital dysplastic internal carotid artery (ICA). Arterial plexus at the cavernous region, which supplies the intradural ICA instead of the aplastic cavernous portion of ICA, looks like the "rete mirabile" seen in the lower mammals, and is a characteristic angiographical finding of CRM. Only 29 cases of CRM have been reported in the literature (Chng et al. 2004; Mahadevan et al. 2004; Mikami et al. 2005; Kim et al. 2006; Li et al. 2006; Şahin et al. 2010). CRM is sometimes associated with the vertebral so-called rete mirabile. We report a very rare geriatric case of bilateral carotid and vertebral rete mirabile (CVRM) presenting with subarachnoid hemorrhage (SAH) caused by the rupture of spinal artery aneurysm.

Clinical Report

A 70-year-old female patient presenting with SAH was admitted to our hospital. SAH was considered Fisher group 3, as judged by thick subarachnoid clots on computed tomography scans. Initial computed tomography scans showed SAH dominantly distributing in the posterior cranial fossa. She had no significant past medical history or familial history. Cerebral three-dimensional computed tomography angiography showed no cerebral artery aneu-

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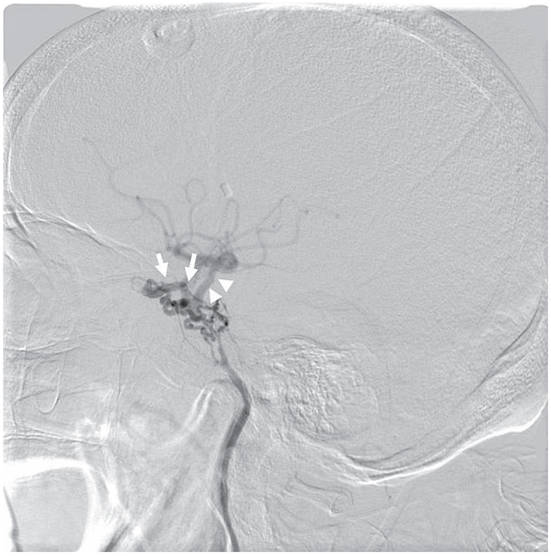

Fig. 1. The right internal carotid arteriogram.
Lateral projection of the right internal carotid arteriogram shows tapering at the petrous portion of internal carotid artery (ICA), absence of the cavernous portion of ICA, and tortuous arterial network (typical "rete mirabile") filling into the dilated right ophthalmic artery (arrows) supplying intracranial ICA of normal appearance (arrowheads).

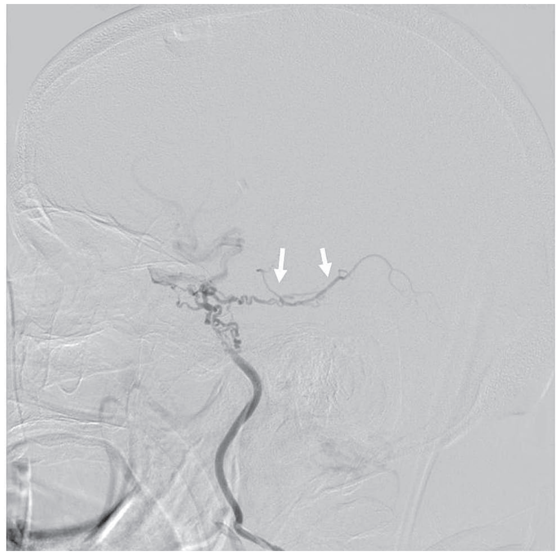

Fig. 2. The left internal carotid arteriogram.
Lateral projection of the left internal carotid arteriogram revealed findings similar to the contralateral ICA. One of the tortuous vessels around the left cavernous sinus also anastomosed into the left superior cerebellar artery (arrows).

a

b

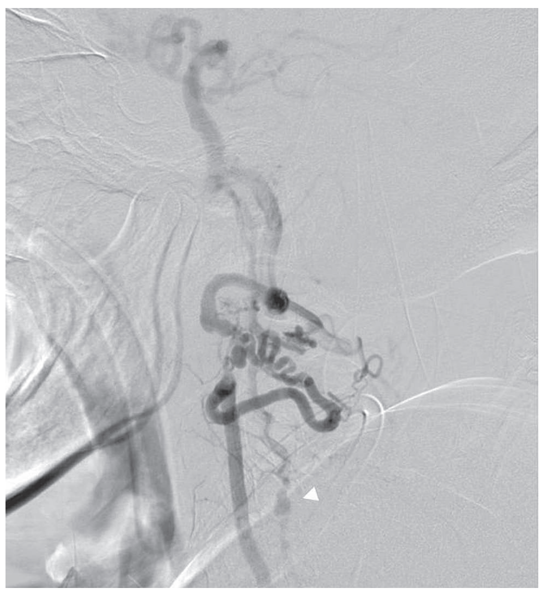

Fig. 3. The left vertebral arteriograms.
Anteroposterior (a) and lateral (b) projection of the left vertebral arteriograms indicate dominant left vertebral artery (VA) occluded segmentally at the level of C2-3 vertebral bodies. Left vertebral arteriograms show not only tortuous arterial network filling into the distal VA, but also the dilated lower radiculomedullary artery (arrows) connecting to the basilar artery via prominent anterior spinal artery (small arrows). There is a saccular aneurysm (arrowhead) on the left lower radiculomedullary artery.

rysm. Cerebral angiography was performed on day 1. The right internal carotid arteriography (ICAG) showed tapering at the petrous portion of ICA, absence of the cavernous portion of ICA, and tortuous arterial network (typical "rete mirabile") filling into the dilated right ophthalmic artery supplying the intracranial ICA of normal appearance (Fig. 1). The left ICAG had a similar appearance (Fig. 2). Left vertebral arteriography (VAG) showed the dominant left vertebral artery (VA) was occluded segmentally at the level of C2-3 vertebral body (Fig. 3). Left VAG showed not only a b

Fig. 4. The right vertebral arteriograms.
Anteroposterior (a) and lateral (b) projection of the right vertebral arteriograms indicate termination of the right VA into the right posterior inferior cerebellar artery. There is a prominent posterior meningeal artery, arising from the distal end of right VA, which fills into the contralateral intradural VA (arrows). Dilated right lower radiculomedullary artery, similarly to the left, feeds the basilar artery via anterior spinal artery. There is a saccular aneurysm (arrowhead) also on the lower right radiculomedullary artery.

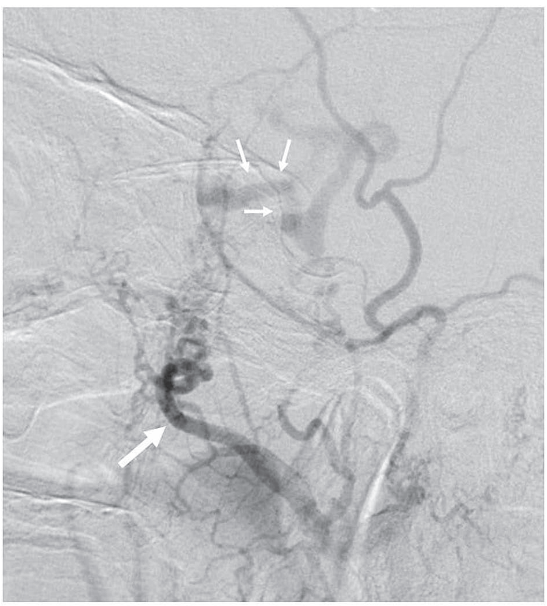

Fig. 5. The right external carotid arteriogram.
Lateral projection of the right external carotid arteriogram shows arterial plexus between the distal internal maxillary artery (large arrow) and the ipsilateral ophthalmic artery (arrows).

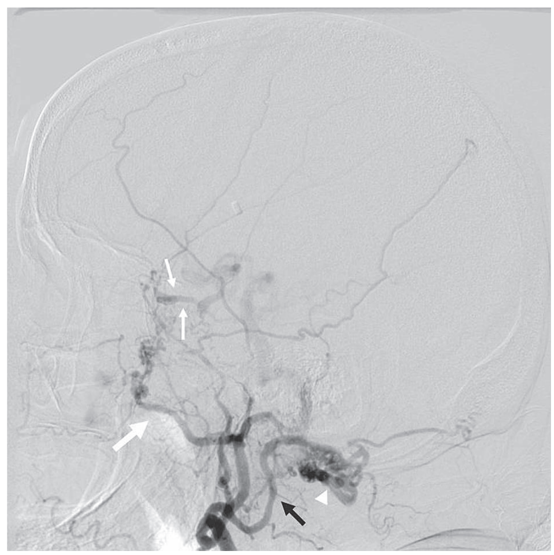

Fig. 6 The left external carotid arteriogram.
Lateral projection of the left external carotid arteriogram (ECAG) reveals findings similar to the contralateral ECAG. There is an arterial network between the distal internal maxillary artery (large arrow) and the ipsilateral ophthalmic artery (arrows). Left ECAG also shows a prominent occipital artery (black arrow) anastomosing into the left intradural VA via tortuous arterial network at the level of left VA occlusion (arrowhead).

tortuous arterial network filling into the distal VA, but also the dilated lower radiculomedullary artery connecting to the basilar artery via anterior spinal artery (Fig. 3). There was a saccular aneurysm on the left lower radiculomedullary artery (RMA) (Fig. 3). The right VAG indicated termination of the right VA into the right posterior inferior cerebellar artery, and a prominent posterior meningeal artery, arising from the distal end of right VA, filled into the contralateral intradural VA. Dilated right lower RMA, similarly to the left, fed the basilar artery via anterior spinal artery. There was a saccular aneurysm also on the lower right RMA (Fig. 4). The right (Fig. 5) and left (Fig. 6) external carotid arteriography (ECAG) showed arterial plexus between the distal internal maxillary artery to the

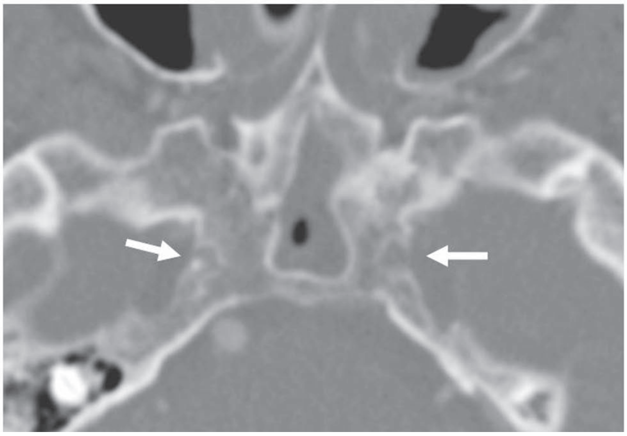

Fig. 7. Computed tomography.
A source image of the three-dimensional computed tomography angiography indicates hypoplastic carotid canal bilaterally (arrows).

ipsilateral ophthalmic artery. Left ECAG also revealed a prominent occipital artery anastomosing into the left intradural VA via tortuous arterial network at the level of the left VA occlusion (Fig. 6). In conclusion, there were not only segmental occlusions of the bilateral ICA, but also segmental occlusion of the bilateral VA just below the dural penetration. Carotid canal was hypoplastic bilaterally (Fig. 7). CRM was bilaterally observed as a connection from the petrous ICA to ipsilateral intradural ICA. Vertebral rete was also observed at the occluded segment of the left VA. Therefore, we diagnosed this condition as bilateral carotid and vertebral rete mirabile (CVRM). In this patient, one of lower cervical spinal aneurysms on the bilateral RMAs seemed to be the cause of SAH. It was compatible with the distribution of SAH, which was thick in the posterior cranial fossa and cervical spinal canal in this patient.

To avoid rebleeding, we planned coil embolization of the spinal aneurysm. We decided to embolize the left RMA aneurysm because it was larger than the contralateral RMA aneurysm. On day 7, we tried intrasaccular coil embolization using a micro-catheter (Excelsior™ SL-10, Stryker Japan, Osaka, Japan) and detachable coils. However, we could only perform a proximal RMA occlusion with four detachable coils (a 2.5 mm × 6 cm coil, and three 2 mm × 6 cm coils of GDC-10 UltraSoft™, Stryker Japan, Osaka, Japan) because of the proximal arterial tortuosity. Left RMA aneurysm had disappeared on the follow-up magnetic resonance angiography, on which the contralateral RMA aneurysm was enlarged. Therefore, we performed right RMA proximal occlusion by a microcatheter (Excelsior™ SL-10, Stryker Japan, Osaka, Japan) and three detachable coils (a 2.5 mm × 6 cm coil and a 4 mm × 4 cm coil of GDC-10 UltraSoft™, Stryker Japan, Osaka, Japan, and a 1.5 mm × 1 cm coil of ED COIL-10 Extra Soft, Kaneka, Osaka, Japan) on day 19. Spinal aneurysms had disappeared on the follow-up magnetic resonance angiography. Fortunately, despite our concern about impaired intracranial circulation pressure, no infarct was observed in the posterior circulation after performing embolization twice. She was transferred to another hospital for further rehabilitation with modified Rankin scale of 4.

Discussion

Rete mirabile is a normal structure in the lower mammals such as sheep, cow, and pig. In these species, the rete has physiological roles, such as heat exchanger or pressure absorber for the intracranial arterial blood flow (Edelman et al. 1972). Of course, the rete does not exist in the normal human development process (Jones and Wetzel 1970). Rete may not be a retrograde developmental shift in humans. The cause of rete mirabile as a collateral pathway from extradural to intradural arteries has not been established yet. We can only say that the rete may be a rare configuration of the collaterals anastomosing the extradural and intradural arteries.

The angiographic frequency of CRM was reported as 0.01% (Occleshaw and Garland 1969). To the best of our knowledge, 29 cases of CRM have been reported since 1966 (Chng et al. 2004; Mahadevan et al. 2004; Mikami et al. 2005; Kim et al. 2006; Li et al. 2006; Şahin et al. 2010). Most of the CRM/CVRM cases were Asian patients (Mahadevan et al. 2004). Among these 29 cases, only four cases were associated with vertebral rete mirabile (Itoyama et al. 1993; Hyogo et al. 1996; Mahadevan et al. 2004; Li et al. 2006). The present patient is the fifth case of CVRM. The age of the 29 patients with CRM/CVRM ranged from 2 to 68 (mean 40.4 years). Therefore, our patient is the oldest patient with CRM/CVRM amongst reported cases.

In the previous reports, twelve cases of CRM/CVRM were associated with SAH (Mahadevan et al. 2004; Mikami et al. 2005). However, ruptured aneurysm was detected in only 3 of the 12 patients. The present patient is the first report of ruptured cervical radiculomedullary arterial aneurysm as a cause of SAH associated with CRM/CVRM. In patients with CRM/CVRM, aneurysm formation on the collateral vessels may be a reasonable consequence because of the hemodynamic stress. Spinal arteries can work as a collateral pathway in cases of CVRM. Detailed evaluation of the cervical spinal arteries should be an important part of the evaluation to detect or rule out ruptured aneurysm in case of SAH associated with CVRM.

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Conflict of Interest

All authors declare no conflict of interest.

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