A Case of Vertebro-vertebral Arteriovenous Fistula Clinically Diagnosed as Segmental Arterial Mediolysis Complicated by Celiac Artery Aneurysm Suspected in a State of Impending Rupture

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Objective: Vertebro-vertebral arteriovenous fistula (VVAVF) is a rare disorder that often forms a high blood flow shunt. It is mostly due to trauma, but it is occasionally caused by systemic diseases. Segmental arterial mediolysis (SAM) is characterized by non-inflammatory/non-atherosclerotic segmental lysis of the arterial media and is often diagnosed due to intraperitoneal hemorrhage from a ruptured abdominal visceral artery.

Case Presentation: A 45-year-old male with right VVAVF presented with pulsating tinnitus and underwent coil embolization of the right vertebral artery around the fistula. Cerebral angiography disclosed multiple aneurysms in the head and neck region. Abdominal contrast-enhanced CT showed a dissecting aneurysm extending from the celiac artery to the splenic artery. Since impending rupture was suspected, the aneurysm was embolized later at the radiology department.

Conclusion: A case of VVAVF complicated by SAM is reported. If multiple arterial abnormalities are observed, systemic examination and appropriate intervention are important.

Keywords ▶ vertebra-vertebral arteriovenous fistula, segmental arterial mediolysis, endovascular treatment

Introduction

Vertebro-vertebral arteriovenous fistula (VVAVF) is a rare disorder that forms abnormal shunts between the vertebral artery and neighboring veins and often shows high blood flow shunts. Its symptoms are often pulsatile tinnitus and neck pain, but it occasionally induces myelopathy and radiculopathy secondary to venous dilation around the cervical spine.

Segmental arterial mediolysis (SAM) is a disease concept proposed by Slavin et al. in 1976 and is a degenerative disease that causes non-inflammatory/non-atherosclerotic segmental lysis of the arterial media. It is considered to be the primary cause of splanchnic artery aneurysms. Usually, it is often diagnosed due to abdominal pain or shock after rupture.

There have been sporadic reports on SAM complicating vertebral artery or splanchnic artery aneurysms, and the condition has often been detected due to rupture of some aneurysm. If abnormalities such as aneurysm and dissection of head and neck vessels are noted, it is recommended to perform general vascular assessment with systemic diseases such as SAM and fibromuscular dysplasia (FMD) in mind.

This report presents a patient who had an onset with VVAVF, was suspected to have SAM from the clinical history and angiographic findings, was suggested by contrast-enhanced CT to have a splanchnic artery aneurysm in a state of imminent rupture, and underwent endovascular treatment resulting in radical cure.
**Case Presentation**

The patient was a 45-year-old male complaining of pulsating tinnitus as a primary symptom. He had been diagnosed with hypertension half a year before and had since been treated with oral olmesartan, indapamide, and nifedipine. There was no history of trauma. He consumed about 360 mL of sake daily but had no history of smoking. His familial history included subarachnoid hemorrhage in his mother and grandmother on the father’s side.

The patient noted pulsating tinnitus with a sudden onset, and as this symptom persisted for about half a year, he consulted a local physician. Since head MRI suggested a right dural arteriovenous fistula, the patient was referred to our hospital. On examination at the outpatient clinic, he had alert and wakeful consciousness, showed no neurological deficits, and complained of pulsating tinnitus alone. No clotting abnormality was noted on blood chemistry tests.

On MRA, the delineation of the V3 and more distal segments of the right vertebral artery was poor, but the surrounding venous plexus, inferior petrosal sinus, and cavernous sinus were visualized (Fig. 1) and a diagnosis of VVAVF was made. Cerebral angiography was carried out, and right vertebral arteriography revealed an arteriovenous fistula in the V3 segment of the right vertebral artery (Figs. 2A and 2B). Left vertebral arteriography also showed an arteriovenous fistula in the V3 segment of the right vertebral artery via the vertebrobasilar junction. An aneurysm about 8 mm in diameter was also noted at a site distal to the fistula (Fig. 2C). By cone beam CT, the fistula was judged to be located in the V3 segment of the right vertebral artery (Fig. 2D).

The right vertebral artery showed marked tortuosity from the cervical region and multiple aneurysms (Fig. 2A). The cervical segment of the left internal carotid artery also presented findings suggestive of dissecting aneurysm (Fig. 2E).

Presently, pulsating tinnitus is the only symptom, but a vertebral artery aneurysm was large with a diameter of 12 mm and had ruptured before, multiple aneurysms were present in the right vertebral artery, and the shunt flow of the VVAVF was high, refluxed in the ipsilateral inferior petrosal sinus, and reached the cavernous sinus. Since these findings suggested the possibility of hemorrhage in the cervical region and induction of neurological symptoms by dilation of veins around the cervical spine, we selected embolization of the right vertebral artery including the aneurysms.

**Endovascular treatment**

Since the right vertebral artery was scheduled to be embolized with coils, oral administration of aspirin at 100 mg and clopidogrel at 75 mg was started from 5 days before the treatment to prevent periprocedural thromboembolism similarly to the treatment of cerebral aneurysms. Under local anesthesia, 8 Fr and 6 Fr sheaths were inserted into the right and left femoral arteries, respectively, and 8 Fr OPTIMO (Tokai Medical Products, Aichi, Japan) and 6 Fr Fubuki 90 cm Straight (Asahi Intecc, Aichi, Japan) were navigated to the right and left vertebral arteries, respectively. From the right vertebral artery, PX SLIM (Penumbra Inc., Alameda, CA, USA) was guided to the center of the aneurysm, and Headway 17 (Terumo, Tokyo, Japan) was wound nearly around the aneurysm and inserted into the proximal part of the aneurysm, using ASAHI Chikai black 14 (Asahi Intecc). The aneurysm and fistula were occluded by inserting eight Penumbra coils (Penumbra Inc.) and two Hydrocoil 14 (Terumo, Tokyo, Japan) through the PX SLIM. Since there was concern over recanalization, additional four Penumbra coils were inserted through the PX SLIM, and two DELTAMAX (Johnson and Johnson Inc., New Brunswick, NJ, USA) and 2 Hydrocoil 10 were inserted through the Headway 17, and embolization was successful.
Fig. 2  Preoperative digital subtraction angiography. (A) Right vertebral artery angiogram showing dilatation and tortuosity of the right vertebral artery and vessel wall irregularity at the V2 segment. (B) Right vertebral artery angiogram demonstrating A-V fistulae at the V3 segment (arrow). (C) Left vertebral angiogram showing retrograde flow drained into fistulae from the left vertebral artery through the right vertebral artery (arrow). Two flow-related dissecting aneurysms are identified near the fistulae (arrow head). (D) Cone beam CT image clearly showing the direct shunting point of the fistulae (arrow). (E) Left common carotid artery angiogram showing another aneurysm (double arrow) at the cervical segment of the left internal carotid artery.

Fig. 3  (A) Bilateral vertebral artery angiogram clearly showing vertebral aneurysms (black arrow head) and A-V fistulae drained into internal jugular vein and inferior petrous sinus (black arrow). (B) Intraoperative bilateral VA angiogram showing the tip of the Headway 17 micro-catheter (white arrow head) and the PX SLIM microcatheter (white arrow) navigated into the vertebral artery aneurysms. (C) Postoperative fluorography demonstrating complete occlusion of the vertebral artery aneurysms and the V3 segment of the right vertebral artery with hydrogel coils.
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completed after inserting a total of 18 coils (383 cm) (Fig. 3C). Complete occlusion of the right vertebral artery around the fistula was observed by final angiography (Fig. 4A), and occlusion of the fistula was also confirmed by left vertebral arteriography (Fig. 4B).

Postoperative course
Vascular murmur disappeared after the procedure, and no de novo neurological abnormalities were noted. CTA performed 2 days after the procedure for systemic vascular evaluation showed vascular dilation and septal formation from the celiac artery to the splenic artery, suggesting dissecting aneurysm (Fig. 5A).

The pancreas around the spleen showed low-density areas surrounding the vessel, and pseudoaneurysm or impending rupture was suspected. In addition, two saccular aneurysms were detected in the left renal artery (Fig. 5B).

Although the condition was asymptomatic, coil embolization was carried out for the prevention of rupture at the radiology department 14 days after the endovascular treatment at our department.

Antiplatelet medication was reduced to one drug 1 month after the procedure and terminated 1 year after the procedure. MRA performed 6 months after the procedure confirmed occlusion of the right vertebral arteriovenous fistula (Fig. 6A). No ischemic complications were observed during a follow-up period of 1 year and 5 months.

After 1 year, abdominal CT confirmed no change in the state of occlusion of the embolized area (Fig. 6B) or the renal artery aneurysms. Presently, the patient is followed up annually by imaging examinations.

Discussion
VVAVF is a rare disease, and surgical resection of a node and fistula of the proximal vertebral artery in 1917 is its first report. It is considered to be often caused by trauma, but there have been sporadic reports of VVAVF complicating FMD or neurofibromatosis type 1. Recently, endovascular treatment has been performed more frequently as a less invasive treatment. The devices used for endovascular treatment include detachable balloons, coils, ONYX for embolization and covered stents for fistula closure.

The natural history of VVAVF is unclear. Its symptoms also vary with the shunt flow volume and drainage route. Recently, complete occlusion has begun to be achieved by endovascular treatment, and complete occlusion leads to symptomatic resolution with rare recurrences. In the patient presented here, VVAVF was complicated by SAM, and arteriovenous fistula is surmised to have formed due to
rupture of a dissecting aneurysm. Concerning the treatment for arteriovenous fistulas, the possibility of radical cure is considered high by embolization from the dilated outflow vein to the vertebral artery, which is the feeding artery, through the fistula. However, as navigation of the catheter to the venous side via the fistula was difficult, we completely occluded the fistula with the vertebral artery aneurysm and the vertebral artery around it, resulting in the disappearance of VVAVF. However, there has been a report of the occurrence of new fistulas at other sites of the same vessel after closure of fistulas alone,\textsuperscript{21} and occlusion of the shunt area including the parent artery is considered more appropriate if the condition is complicated by a generalized disorder suggestive of vulnerability of the blood vessels themselves.

SAM was reported in 1976 by Slavin et al.\textsuperscript{5} Although it frequently affects the abdominal visceral arteries, it may occur in all organs including the retroperitoneum, head and neck region, and coronary artery. Kim et al.\textsuperscript{13} performed a systematic review of SAM and reported a mortality rate of 22\% once bleeding occurs. However, Michael et al. followed
up two patients by imaging examinations without surgery and reported complete remission in one patient and partial remission in one patient.\(^{14}\) It is necessary to perform conservative treatment with frequent examinations using imaging modalities and to make therapeutic interventions as necessary.

According to imaging findings, vascular wall irregularities such as dilation and stenosis are considered to occur frequently in SAM, and multiple findings resembling those of dissection are observed in typical cases. Inada et al.\(^{15}\) reported that SAM presented as dissection or pseudoaneurysm in 78% and had multiple aneurysms in 33% of the patients.

To definitively diagnose SAM, other systemic disorders or angiitis must be excluded. As there are no specific blood test or body surface findings such as Ehlers–Danlos syndrome and neurofibromatosis, the diagnosis is difficult, but it is possible if the characteristics of the disorder are understood.

In the differential diagnosis, SAM may be confused with FMD, but FMD is reported to be frequently observed in young to middle-aged women and to primarily affect the renal and internal carotid arteries, with extrarenal lesions being rare. In addition, FMD is accompanied by arterial stenosis and dilation and often presents with ischemic symptoms, but differs from SAM in that its lesions are mainly stenotic.

In our patient, angiitis was considered unlikely from the blood test results and clinical history, and vascular findings were more consistent with SAM than with FMD. Furthermore, systemic evaluation using imaging modalities is considered necessary for the future.

### Conclusion

In this patient, we suspected a systemic disorder from the findings on angiography performed for the treatment of VVAVF, found a celiac artery aneurysm considered from systemic contrast-enhanced CT scans to be in a condition of impending rupture, and treated both aneurysms endovascularly. Since the patient showed multiple vertebral artery and celiac artery aneurysms associated with VVAVF, SAM was clinically diagnosed based on the sites and images of the lesions.

When multiple aneurysms and shunt disorders are detected, it is important to carefully examine the condition with a systemic disorder in mind.

### Disclosure Statement

The first author and all of the coauthors have no conflicts of interest.

### References