A Case of Spontaneous High-flow Vertebral Arteriovenous Fistula Treated by Trans-arterial Target Embolization Using Balloon Remodeling Technique

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Objective: Spontaneous vertebral arteriovenous fistula (VAVF) is a rare vascular disorder. Herein, we report a case of high-flow VAVF with ipsilateral persistent primitive trigeminal artery (PPTA) treated by trans-arterial target embolization (TE) using balloon remodeling technique (BRT).

Case Presentation: A 58-year-old woman with a medical history of non-tuberculous mycobacterial lung disease had complained of right-sided pulsatile tinnitus for 3 years; however, there were no symptoms due to brain or spinal cord dysfunction. DSA revealed high-flow VAVF in the right V3 segment at the C2 vertebral body level and ipsilateral PPTA. The multi-planar reconstruction (MPR) images of cone-beam CT (CB-CT) obtained from 3D rotational angiography (3D-RA) in the arterial phase were useful to evaluate the morphology of VAVF. The fistula was single and positioned in the V3 segment at the C2 vertebral body level. The venous side of the fistula had expanded into a spheroid-like shape. A planned trans-arterial TE was performed 39 months after diagnosis due to recurring lung hemorrhage. Using BRT, TE was successful by packing back from the spheroid-like space with coils. The spheroid-like space was located slightly caudal to the fistula, and occlusion balloon inflated just distal to the fistula was helpful to navigate the microcatheter. After 12 months, DSA showed no recurrence of VAVF.

Conclusion: The MPR images of CB-CT obtained from 3D-RA were useful for evaluation of the morphological structure of VAVF. The BRT was useful for successful TE.

Keywords ▶ vertebral arteriovenous fistula, three-dimensional-rotational angiography, target embolization, balloon remodeling technique, pulsatile tinnitus

Introduction

Vertebral arteriovenous fistula (VAVF) is a rare vascular disorder defined as abnormal direct communications of the extracranial vertebral arterial trunk or its branches into the neighboring vertebral vein or vertebral venous plexus or internal jugular vein. There may be more than one fistula, and the structure on the venous side is often complicated. We experienced a case of spontaneous high-flow VAVF accompanied with persistent primitive trigeminal artery (PPTA) and reported its anatomical evaluation and treatment.

Case Presentation

A 58-year-old woman with a medical history of non-tuberculous mycobacterial lung disease had complained of right-sided pulsatile tinnitus for 3 years. During an active state, the patient’s tinnitus was minimal, but increased when she pointed her head down. This prompted the administration of a brain checkup system employing head MRI and MRA, called the brain-dock, which is prevalent in Japan. The brain MRI showed no abnormalities and the intracranial MRA revealed the right PPTA.
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and intracranial MRA showed an abnormal structure around the right vertebral artery (VA) at the C1-3 cervical vertebral body level, wherein the right VA appeared discontinuous (Fig. 1A and 1B). The patient was referred to our hospital. The patient suffered from tinnitus. There were no symptoms due to brain or spinal cord dysfunction related to steal, venous hypertension, or mechanical compression. She did not have any mixed connective tissue diseases, such as neurofibromatosis. Right vertebral angiography (VAG) revealed high-flow VAVF between the right VA trunk and vertebral vein (Fig. 1C). The left VAG retrogradely imaged the right VA up to VAVF (Fig. 1D). Right internal carotid angiography revealed the basilar artery (BA) via right PPTA and also retrogradely imaged the right VA up to VAVF (Fig. 1E).

The 3D rotational angiography (3D-RA) in the arterial phase was performed using Innova IGS 630 angiography system (GE Healthcare, Chalfont St Giles, UK) equipped with a 30 × 30 cm flat-panel detector and a rotation speed of 40 degree/sec. A total of 16 mL of the undiluted contrast agent was injected at a rate of 2.5 mL/sec and recorded at 30 fps (Fig. 2A and 2B). Evaluation of VAVF was enabled using the multi-planar reconstruction (MPR) images obtained from cone-beam CT (CB-CT) of right vertebral 3D-RA; (Fig. 2C and 2D). The fistula was single and positioned in the V3 segment at the C2 vertebral body level, between the transverse process of C2 and the transverse process of C1. The maximum diameter of the fistula was 2.3 mm. The venous side of the fistula had expanded into a spheroid-like shape measuring 5.5 × 5.5 × 7.8 mm. Most of the shunt flow through the fistula flowed retrogradely into the internal and external vertebral venous plexuses. In the vertebral venous plexuses, most of the shunt flow flowed downward, but some flowed upward into the suboccipital cavernous sinus via marginal sinus and veins around the occipital condyle and then drained into the internal jugular vein. Some shunt flow through the fistula directly and retrogradely flowed into the suboccipital cavernous sinus.

We decided to perform trans-arterial target embolization (TE) with packing the venous spheroid-like space beyond the

![Fig. 1](A and B) Cervical and intracranial MRA shows discontinuity of the right VA and the right PPTA. (C) Right VAG reveals high-flow VAVF. (D) Left VAG retrogradely images right VA and VAVF. (E) Right ICAG shows right PPTA, and retrogradely images right VAVF. ICAG: internal carotid angiography; PPTA: persistent primitive trigeminal artery; VA: vertebral artery; VAG: vertebral angiography; VAVF: vertebral arteriovenous fistula
fistula. If the coil mass would be unstable in the venous spheroid-like space, short segment internal trapping (SSIT) would be performed because BA was perfused via PPTA and left VA. However, the procedure was postponed because of patient’s lung hemorrhage, which recurred a year later. Change in treatment for her mycobacterial disease halted her lung hemorrhage for more than a year, after which she underwent endovascular treatment, 39 months after diagnosis.

The patient was treated with 100 mg of aspirin once daily for 1 week and 200 mg of cilostazol twice daily for 3 days before trans-arterial TE. Under intravenous anesthesia with dexmedetomidine hydrochloride, a 7 Fr long sheath was placed in the right femoral artery. The bilateral VAG revealed no apparent change in the blood flow hemodynamics. The right vertebral 3D-RA was performed under conditions previously adopted. The MPR images obtained from CB-CT of right vertebral 3D-RA revealed that the maximum diameter of the fistula had increased slightly to 2.8 mm (Fig. 3). The venous spheroid-like space had also enlarged and measured 7.7 × 7.2 × 9.2 mm. Under full heparinization, we placed a 7 Fr FUBUKI Catheter (Asahi Intecc, Aichi, Japan) into the right VA and the following three catheters were advanced coaxially. An Excelsior SL-10 microcatheter (Stryker, Kalamazoo, MI, USA) was navigated in the right VA, distal to the fistula using CHIKAI microguidewire (0.014 inch; Asahi Intecc), which would be used if SSIT was to be performed. A Scepter C occlusion balloon catheter (4 × 10 mm; Microvention, Tustin, CA, USA) was navigated in the right VA just distal to the fistula using Traxcess microguidewire (0.014 inch; Microvention). A Headway 17 microcatheter (steam shaped to 30 degrees; Microvention) was placed in the venous spheroid-like space through the fistula using CHIKAI microguidewire. The venous spheroid-like space was located slightly caudal to the fistula; however, with the Scepter C balloon inflation just distal to the fistula, the microcatheter somehow managed to follow the microguidewire. Using the balloon remodeling technique (BRT), a stable coil frame was made in the spheroid-like space with an 8 mm × 27 cm coil (HydroFrame 18;
According to a recent review article, 280 cases of VA VFs have been reported. Among these cases, approximately half are known to be caused by trauma, including penetrating injury, indirect blunt injury such as chiropractic-induced, falls, and iatrogenic injury. Iatrogenic factors include incorrect puncture of the jugular vein or carotid artery, and faulty insertion of a pedicle screw. Conversely, the remaining half of all VA VFs are spontaneous and idiopathic, and one-third of the spontaneous VA VFs is associated with diseases such as neurofibromatosis, Ehlers–Danlos syndrome, and fibromuscular dysplasia that may result in dysplasia of the arterial smooth muscle cells. The present case had no obvious history of trauma and no apparent physical findings related to mixed connective tissue diseases, but she had ipsilateral PPTA. The exact cause of persistence of the primitive vessel into adulthood is not completely clear; however, it is well known that PPTA can be associated with several other
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Fig. 4 DSA at target embolization. (A) Fluoroscopy shows a stable coil frame made by the first coil in the spheroid-like space. (B) After spheroid-like space is filled with six coils, right VAG shows a narrow space between the right VA and the spheroid-like space (surrounded by black arrowheads). (C) Fluoroscopy shows small coil mass in the narrow space. (D) Right VAG reveals occlusion of the shunt. VAG: vertebral angiography; VA: vertebral artery

vascular anomalies and disorders, including aneurysms, arteriovenous malformations, and carotid-cavernous fistula. In addition, PPTA has been reported in the context of neurofibromatosis. Therefore, this is the first case of VAVF with PPTA; there may be some developmental or genetic relationship between them.

Regarding the position of the fistula, one half of traumatic VAVFs were located at the V2 segment (from the transverse foramen of C6 to the transverse foramen of C2), and almost half of spontaneous VAVFs, including our patient, were located at the V3 segment (from the transverse foramen of C2 to dura). For the successful treatment of VAVF, it is necessary to evaluate the number, position, and size of the fistula and venous structure around the fistula. There could be more than one fistula, or it may be too large for TE. One of the methods to pre-evaluate whether TE is possible employs a non-detachable balloon catheter, which navigates from VA to the venous side through the fistula. TE is possible if VAVF is occluded when the non-detachable balloon is inflated. A newly established method to evaluate the fistula and to predict whether TE will succeed is 3D-RA. The introduction of the DSA machine equipped with a flat-panel detector has easily enabled 3D-RA. In the present case, the MPR images obtained from contrast enhanced CB-CT for 3D-RA created in any direction enabled evaluation of VAVF that revealed the point of fistula and showed that the coil mass was stably placed in the venous spheroid-like space near the fistula, as expected. In addition, high-resolution CB-CT, akin to 3D-RA, can be used to evaluate VAVF. According to the report, the radiation dose of high-resolution CB-CT is about 3.5 times than 3D-RA, but uses 3–5 times less contrast agent than 3D-RA.

The ideal goal of treatment is the occlusion of the fistula and preservation of VA patency. Surgical treatments, such as direct suture of fistula orifice, and ligation of the fistula orifice with or without carotid-vertebral bypass are invasive procedures that have largely fallen out of favor. Progress in endovascular treatment due to advances in medical devices such as detachable balloons, coils, and stents has made it
easier to perform more surgeries. Early in endovascular treatment, latex balloons were often used and later shifted to coils.\textsuperscript{2,5,9,10} Balloons and coils have been used to perform TE or SSIT; TE preserves VA patency and therefore should be the first option. When fistulas cannot be identified, or are multiple or if too large for TE, SSIT would be selected. Among others, liquid embolic material may be used in combination with the coils,\textsuperscript{2,9,11} and the use of stents has increased in recent years.\textsuperscript{12–14} Covered stents are considered useful because they can preserve the antegrade blood flow of the VA and might be able to close even large fistulas. However, poor adhesion between VA and stent caused the failure of the fistula closure.\textsuperscript{12} Recently, a case report on the use of flow diverting stents for VAVF was published.\textsuperscript{14} However, four flow diverting stents were used in the study, and the patient’s symptoms persisted until ceasing the administration of antithrombotic agents. Moreover, covered and flow diverting stents are off-label use in Japan.

When performing TE, the BRT would be essential to navigate and stabilize the microcatheter and protect coil migration. In the present case, the microcatheter needed to be navigated caudally from the fistula, and inflation of the occlusion balloon just distal to the fistula made it easy to guide the microcatheter caudally. Most VAVF have a high-flow shunt, and it may be relatively easy for a microcatheter to pass through the fistula, but the venous structure of VAVF is complicated, and it is sometimes difficult to advance the microcatheter toward the venous side. Furthermore, if SSIT is the only option, double catheter technique or tandem balloon technique may be useful to control the flow of VA and stabilize the coils.\textsuperscript{15}

### Conclusion

A case of spontaneous high-flow VAVF with ipsilateral PPTA is reported. To evaluate the venous structure of VAVF, the MPR images of CB-CT obtained from 3D-RA were very helpful. VAVF was successfully treated by trans-arterial TE with BRT.

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### Disclosure Statement

None of the authors have any conflicts of interest.

### References