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Case reports in endovascular and interventional neurology, volume III - 2023

Edited by
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Case reports in endovascular and interventional neurology, volume III - 2023

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Multiple telescoping flow diverter technique in endovascular treatment of a vertebrobasilar dissecting aneurysm: case report

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A 64-year-old man presented with headache and dizziness. A vertebrobasilar dissecting aneurysm was identified via computed tomography angiography and high resolution magnetic resonance imaging. Perioperatively, standard oral dual antiplatelet drugs were given. Two flow diverters were telespcoped for endovascular treatment of the aneurysm. Postoperatively, there were no signs of cerebral infarction and no new symptoms. At the 6-month follow-up, digital subtraction angiography showed that the aneurysm was almost completely occluded, with no other complications. This case serves as a reference for using the multiple telescoping flow diverter technique to treat vertebrobasilar dissecting aneurysm.

KEYWORDS

flow diverter, telescoping technique, vertebrobasilar, dissecting, aneurysm

Introduction

Surgical treatment of complex intracranial aneurysm is problematic, and endovascular treatment has high rates of complications and recurrence. Using a flow diverter (FD) makes treating this kind of aneurysm simpler and safer (1). The advantages of a flow diverter include low porosity and high metal coverage rates. Some studies, however, report that overlapping flow diverters increase the risks of the endovascular treatment and do not benefit the aneurysm cure rate (2).

It remains inevitable that in cases with large or giant aneurysms with extremely wide necks, or longer spindle-shaped aneurysms, two or more flow diverters must be inserted, through a telescoping technique. Studies that have reported the efficacy and safety of telescoping flow diverters in complex intracranial aneurysms treatment are limited (3, 4).

Here we report the case of a vertebrobasilar dissecting aneurysm treated with a multiple telescoping flow diverter technique, which may serve as a reference for the procedure.

Case presentation

A 64-year-old man visited a local hospital due to headache and dizziness. The head computed tomography angiogram (CTA) showed an aneurysm, and he was referred to our hospital in February 2022.

The patient had a family history of subarachnoid hemorrhage. Digital subtraction angiography revealed a fusiform aneurysm at the left vertebral artery V4 segment to the proximal segment of the basilar artery, with a maximum diameter and length of about

10 and 60 mm, respectively; the right vertebral artery was slender, and the right vertebral artery V4 segment was occluded (Figure 1). High resolution magnetic resonance imaging confirmed that the lesion was vertebrobasilar dissecting aneurysm (Figure 2). Considering the characteristics of the aneurysm, we decided to use a flow diverter for the treatment. Because of the large area of the lesion, a single flow diverter could not completely cover the aneurysm, so two flow diverters were implanted via a telescoping technique.

The patient was administered oral antiplatelet drugs (aspirin 100 mg, and clopidogrel 75 mg, 1×/d) 5 days before the procedure. After 5 days, thromboelastography was performed. The arachidonic acid and adenosine diphosphate inhibition rates were 82 and 48%, respectively. Under general anesthesia, a 6F 90-cm long sheath (Cook, USA) was placed in the left subclavian artery, a 5F 115-cm Navien catheter (Medtronic, USA) was placed in the V3 segment of the left vertebral artery, and a microcatheter (Shanghai MicroPort, China) was inserted into the parent artery.

The first 5.5 × 45 mm Tubridge flow diverter (Shanghai MicroPort, China) was delivered to the head of the aneurysm via the microcatheter. The distal end of the first flow diverter was opened by pulling out the microcatheter, and the location of the distal end of the first flow diverter was determined. The first flow diverter was then slowly released in a push-based manner. After the first flow diverter was totally released, the microcatheter was conveyed to the distal end of the first flow diverter under the guidance of the flow diverter push guidewire. The second Tubridge flow diverter (6.0 × 45 mm) was delivered and opened inside the first, and then released slowly by combining a push and tension-reduction technique. The two flow diverters overlapped by ∼25 mm (Figures 3A, B).

The angiography performed after the release of the flow diverter showed that the contrast agent was obviously retained in the aneurysm and the parent artery was patent [O'Kelly-Marotta (OKM)] grade B (5) (Figure 3C). Dyna computed tomography showed that the patency of the two flow diverters was satisfactory, in good agreement with the parent artery, and the entire lesion was covered (Figure 3D).

There were no signs of cerebral infarction after the operation, and the patient recovered well. He had no new symptoms after discharge and continued to receive dual antiplatelet aggregation therapy for 6 months. Six months after the operation, the follow-up digital subtraction angiography showed that the aneurysm was almost completely occluded, with OKM grade C (Figure 4). The antiplatelet strategy was then changed to aspirin (100 mg, 1×/d) with clinical follow-up.

Discussion

Numerous clinical studies have reported the safety and efficacy of flow diversion in treating large intracranial and dissecting aneurysms (1, 6, 7). In recent years, flow diversion to treat vertebrobasilar aneurysms has also been applied, although off-label (8, 9). Few studies have evaluated flow diverter telescoping techniques for treating vertebrobasilar aneurysms. Yet, these techniques offer two advantages. First, multiple telescoping flow diverters to treat aneurysm of the neck effect significantly benefit blood flow changes, and promote thrombopoiesis in the aneurysm. Secondly, for large or dissected aneurysms, a single flow diverter is insufficient to cover the diseased vessels; for long-term efficacy, telescoping is required to completely cover the proximal and distal ends of the aneurysm (10).

The flow diverters now commonly used in China include the pipeline embolization device (PED) and Tubridge. The latter is longer; the maximum length is 45 mm. However, the Tubridge is weaker radially and more highly porous compared with the PED, larger aperture in design can reduce the incidence of perforator occlusion events. This is why the Tubridge flow diverter was chosen for the present case.

Dissecting aneurysms in the vertebrobasilar artery have a complex neurovascular anatomy (11). Placement of flow diverters inevitably results in coverage of basilar artery branches and perforators, the occlusion of perforator may be the most common cause of ischemia events (12). Having a perforators of the basilar artery occluded after FD implantation may occur by 2 mechanisms: the profile of the FD mechanically blocks the orifice of the perforators; or tiny thrombi form on the surface of the FD, which are then carried downstream by flowing blood causing perforators embolism. The smallest branching arteries of the basilar trunk are the perforators, which tend to have a diameter in the range of 80 to 940 μm, with a mean value of 400 μm (13).The Tubridge FD is a self-expanding device, it is composed of 46–62 nickel–titanium alloy microfilaments of 35 μm, and the pore size varies between 1,100 and 2,500 μm, depending on the final FD morphology and selection of the proper size adapted to the vessel diameter. The large-size Tubridge (>3.5 mm), which was mostly used in the posterior circulation, has less decreases the shortening rate after its full opening and offers lower pore attenuation (14). Tubridge FD has larger aperture in design, thus decreases the risk of mechanically blocks the perforators. In addition, in the present case, the overlapping area of the two FDs was selected at the proximal segment of the basilar artery with relatively few perforators to increased security.

Rigorous adequate standardized antiplatelet therapy (with reference to the thromboelastogram) can effectively reduce perforators thrombi embolic events. In the present case, the patient had no ischemic events occurred perioperatively. Rigorous testing of platelet function and subsequent regimen adjustments were crucial to minimize the risk of ischemic complications. Despite this, implantation of multiple FDs should be prudent in vertebrobasilar dissecting aneurysm, which helped to reduce thromboembolic complications.

In our patient, the flow diverter was deployed from the aneurysmal distal to the proximal, which is preferable to proximal-to-distal. Firstly, after the release of the first flow diverter, the re-superselection of the microcatheter can be conducted more easily in the channel of the first flow diverter. Secondly, when telescoping from the distal to the proximal aneurism (with reference to the heart), the first flow diverter and the pushing guidewire can be

Abbreviations: FD, flow diverter; CTA, Computed tomography angiography; HR-MRI, High resolution magnetic resonance imaging; DSA, Digital subtraction angiography; OKM, O'kelly-Marotta.

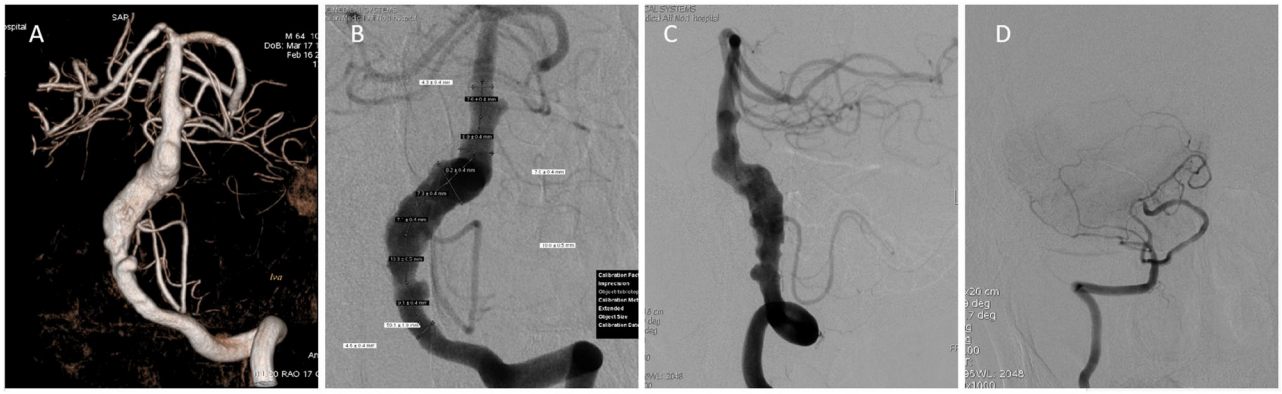

FIGURE 1
Preoperative angiography. **(A)** Three-dimensional angiography reveals a vertebrobasilar aneurysm. **(B)** Anteroposterior angiography measurement data shows the maximum outer diameter and the length of the aneurysm. **(C)** Lateral angiography reveals a vertebrobasilar aneurysm. **(D)** Anteroposterior angiography shows a slender right vertebral artery and occulded V4 segment.

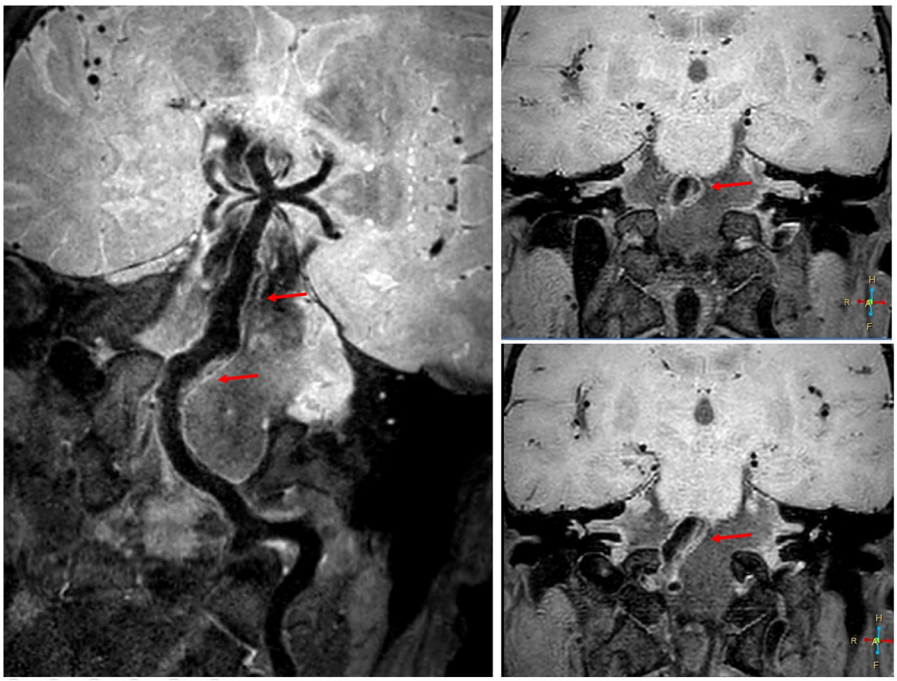

FIGURE 2
High resolution magnetic resonance imaging before the operation. High resolution magnetic resonance imaging reveals a vertebrobasilar dissecting aneurysm (red arrow).

used to support the microcatheter. Compared with telescoping from proximal to distal, the guidewire support is longer, which can support the microcatheter more stably for re-superselection. In addition, it is necessary to ensure that the first flow diverter is firmly anchored in the distal parent artery through distal-to-proximal telescoping.

In this patient, the first flow diverter was anchored about 10 mm in the distal parent artery. A sufficient anchoring length at the distal end is especially important to ensure the stability of the distal flow diverter (15). Studies have confirmed that healing of the aneurysm is promoted when the flow diverter is attached to the wall of the tube after it is implanted. However, attachment is also associated with thrombosis and long-term stenosis and occlusion in the flow diverter itself (16). The telescoping technique may lead to poor adhesion of the flow diverter wall, requiring further treatment.

In the present case, a J-tip guidewire dilation in the flow diverter was performed after the Tubrige implantation to promote adherence of the flow diverter. The present case shows that vertebrobasilar dissecting aneurysm can be treated using telescoping flow diverters. However, we await the long-term results.

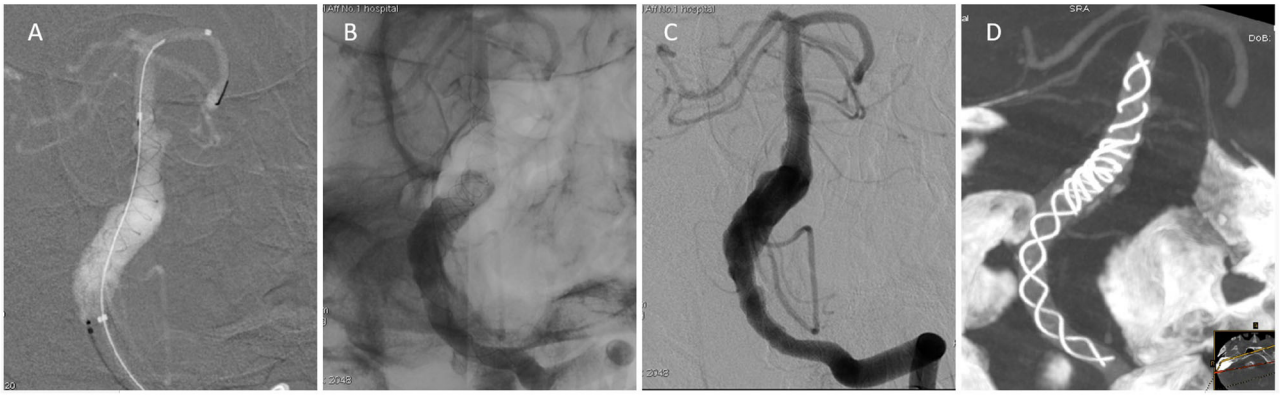

FIGURE 3
Angiography during the operation. **(A)** The first flow diverter was released in the distal end of the aneurysm. **(B)** The second flow diverter was telescoped from the distal to the proximal. **(C)** Angiography after release of the two flow diverters showed retention of contrast agent in the aneurysm. The parent artery was patent. **(D)** Postoperative Dyna-CT showed that the two flow diverters were satisfactorily open.

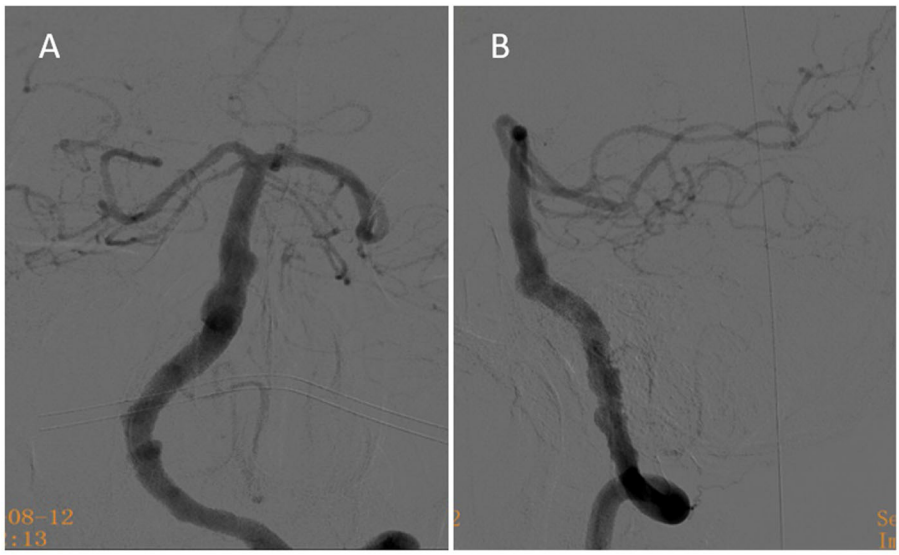

FIGURE 4
Six-month follow-up angiography. **(A)** At the 6-month follow-up, anteroposterior angiography showed that the aneurysm was nearly completely occluded. **(B)** At the 6-month follow-up, lateral angiography reveals the aneurysm was nearly completely occluded.

Conclusions

This report is evidence that vertebrobasilar dissecting aneurysm can be successfully treated with a telescoping flow diverter technique with two flow diverters, and the treatment is feasible for this type of aneurysm. A prospective randomized study is needed to strengthen the evidence and determine the best conditions for applying flow diverter technology.

Data availability statement

The raw data supporting the conclusions of this article will be made available by the authors, without undue reservation.

Ethics statement

The studies involving human participants were reviewed and approved by First Affiliated Hospital of Dalian Medical University. The patients/participants provided their written informed consent to participate in this study. Written informed consent was obtained from the participant/patient(s) for the publication of this case report.

Author contributions

M-YW and FW performed most of the investigation, data analysis, and wrote the manuscript. Y-SL, X-BA, and TP contributed to interpretation of the data

and analyses. All authors have read and approved the manuscript.

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The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Hemifacial spasm caused by unruptured fusiform vertebral aneurysm treated with endovascular coil embolization: a case report

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Hemifacial spasm due to fusiform aneurysm of the vertebral artery is extremely rare. The lateral spread response (LSR) is routinely used to monitor hemifacial spasms during microvascular decompression to predict the degree of postoperative remission of hemifacial spasm. We report a case of hemifacial spasm caused by an unruptured fusiform vertebral aneurysm treated with intravascular intervention and monitoring of LSR. A 59-year-old man was admitted to the hospital with a left facial spasm that gradually worsened for 1 year. Preoperative cerebrovascular angiography indicated fusiform aneurysms in the intracranial segment of the left vertebral artery close to the left facial nerve. The patient underwent parent artery occlusion and aneurysm embolization, and LSR was monitored intraoperatively. After intraoperative aneurysm embolization, LSR disappeared immediately. The postoperative review of cerebrovascular angiography indicated that the parent artery and aneurysm were embolized successfully, and the patient's left facial spasm was relieved after surgery. Hemifacial spasm caused by the vertebral artery fusiform aneurysm can be safely and effectively treated by parent artery occlusion and aneurysm embolization. Meanwhile, intraoperative LSR monitoring can be used to predict postoperative efficacy.

KEYWORDS

hemifacial spasm, occlusion, vertebral artery, fusiform aneurysm, lateral spread response

1. Introduction

Hemifacial spasm (HFS) is typically caused by the curvature and elongation of the facial nerve into the brain stem root exit zone (REZ) and branches of the vertebrobasal nervous system or the vertebral artery (VA). Microvascular decompression (MVD) is the most effective hemifacial spasm treatment (1, 2). The LSR is widely used to guide MVD and predict postoperative efficacy as an objective electrophysiological monitoring indicator of HFS (3). The occurrence of HFS caused by vertebral fusiform artery aneurysms is extremely rare. A case of HFS caused by an ipsilateral fusiform vertebral artery aneurysm is described. In the meantime, intraoperative electrophysiological monitoring was used to record changes in LSR before and after intervention to predict postoperative efficacy.

2. Case report

A 59-year-old man presented with paroxysmal involuntary spasm in the left orbital area that gradually worsened 1 year ago before admission to our hospital. After admission, three-dimensional CT reconstruction examination of the intracranial artery showed that the intracranial segment of the left vertebral artery was locally enlarged and nodular, with a size of ~1.5 cm × 1.3 cm. Magnetic resonance imaging revealed an aneurysm of the intracranial segment of the left vertebral artery, which was appressed to the left facial nerve (Figure 1). Cerebral angiography revealed an aneurysmal enlargement in the V3 segment of the left vertebral artery, with no obvious aneurysm neck and a maximum transverse diameter of 1.2 cm (Figure 2A). No other possible causative lesions for the hemifacial spasm were identified.

Because the aneurysm is fusiform and the parent artery is tortuous, it is difficult to clip the aneurysm directly during craniotomy. Concurrently, there was a certain distance between PICA and aneurysm, so PAO was selected. We intended to use parent artery occlusion (PAO) and coil embolization during surgery after evaluating our patients. The patient was given the following medications as preoperative and perioperative measures: oral aspirin 100 mg/day for 3 days before surgery with clopidogrel 75 mg/day, intraoperative tirofiban acid injection for arterial thrombosis prevention and papaverine hydrochloride injection heparin 80IU/Kg were given intravenously before the operation to prevent cerebral vasospasm, and then, half heparin was added every hour until 1,000 IU was maintained. The patient was then operated under general anesthesia. A unilateral femoral artery was punctured by the Seldinger method, a 6F guide sheath was placed, and a 5F angiography catheter was placed. The internal carotid artery and vertebral artery were overselected for standard angiography, and then, 3D-DSA was performed on the left vertebral artery. The 5F guide catheter (Johnson & Johnson, USA) was superselected for the V2 segment for further angiography, and PAO was performed after the indications were clear. An Excelsior SL-10 microcatheter (Stryker, USA) was superselected through the ipsilateral vertebral artery under the guidance of a microguide wire into the aneurysm, and seven coils (MicroVention, USA) were used to fill the aneurysm completely (Figures 2B, C). 2 hours after the surgery, the sheath was removed, and the femoral artery puncture point was pressed for 15 min, the elastic bandage "8" word compression bandage could be used when there was no active bleeding, total bed rest, and the lower limb was immobilized for 12 h. The marginal mandibular branches of the facial nerve were stimulated with fixed frequency and intensity, the LSR was recorded in the mentalis and orbicularis oculi muscle, and the LSR was monitored throughout the operation. After intraoperative embolization of the aneurysm, electrophysiological monitoring showed that LSR disappeared (Figure 3). Postoperative DSA examination indicated complete embolization of the aneurysm and parent artery, and the patient's left facial spasm was significantly relieved. A month later, the patient's symptoms completely disappeared.

To prevent thrombosis and ischemic events after surgery, the patient was instructed to take aspirin 100 mg/d and clopidogrel 75 mg/d orally the following day. Clopidogrel was discontinued 6 weeks later, and asprin was prescribed for life for a year.

3. Discussion

HFS is a functional neurological disease characterized by involuntary spasms of the muscles innervated by the facial nerve and its branches, which is mainly caused by the direct compression of the REZ area by the branch of the vertebrobasilar system or the curvature and elongation of the vertebral artery (VA) itself (1). HFS can also be caused by rare aneurysms, arteriovenous malformations, and tumors in the cerebellopontine angle. HFS caused by fusiform vertebral aneurysm is extremely rare. Four cases have been reported to date (Table 1) (11, 12). Most vertebral saccular aneurysms with HFS can be clipped by craniotomy, and the REZ region can be explored at the same time to separate the aneurysm from the adhesion of the facial nerve root, which can cure both aneurysm and HFS (11, 13). However, because most vertebral

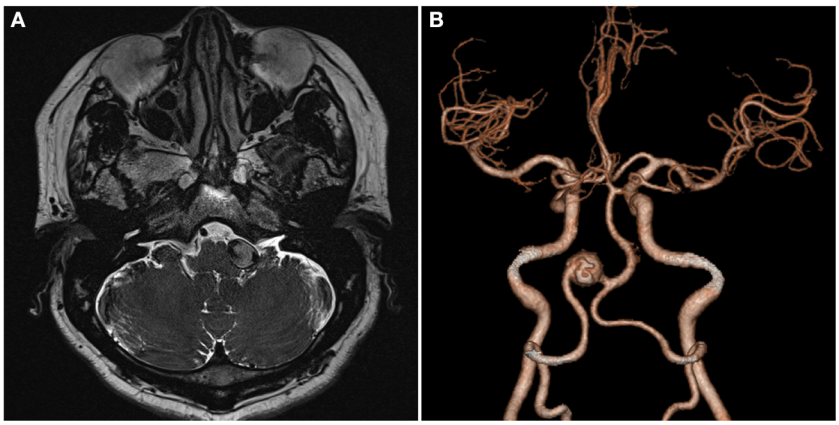

FIGURE 1
Images of the patient upon admission. **(A)** Axial T2-weighted MR image shows a fusiform aneurysm as a flow void signal left of the pons. **(B)** Three-dimensional CT angiography shows VA fusiform aneurysm. VA, vertebral artery.

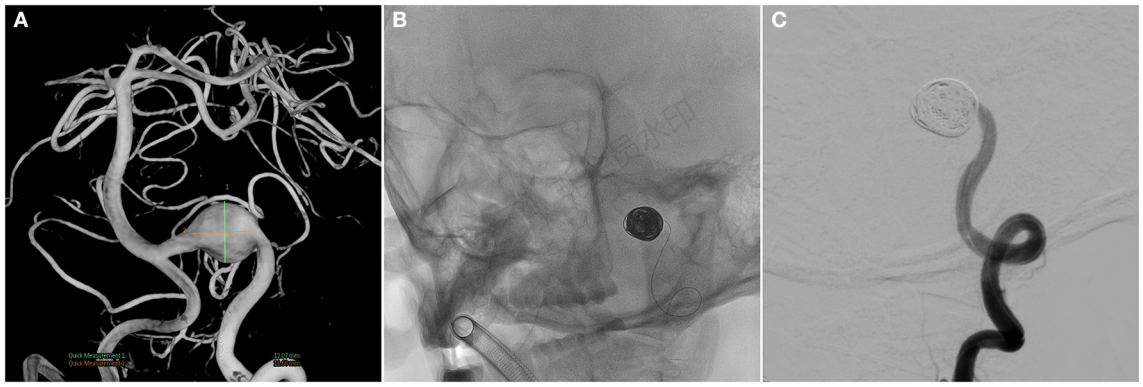

FIGURE 2
(A) Cerebral angiography before and after endovascular treatment and preoperative cerebral angiogram shows a fusiform aneurysm at the V3 segment. **(B, C)** Angiograms after endovascular treatment show the complete occlusion of left VA, including aneurysm. VA, vertebral artery.

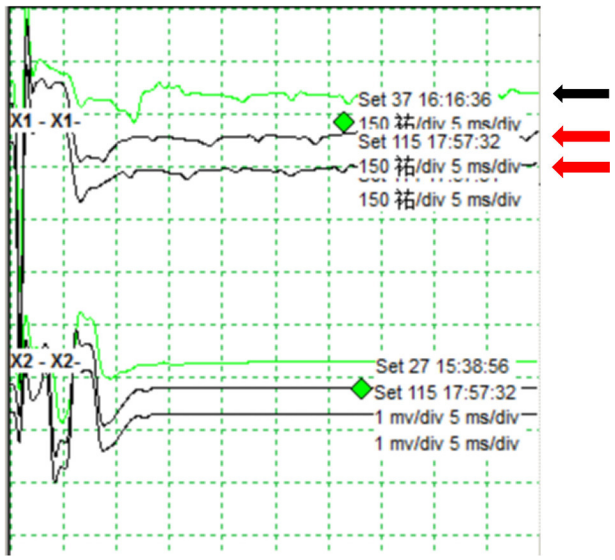

FIGURE 3
Monitoring of the lateral spread response during endovascular surgery. The LSR shows before embolization of the aneurysm (black arrow). The LSR disappears after intraoperative embolization of the aneurysm (red arrow).

aneurysms are fusiform and direct clipping is difficult, the majority of them were treated with isolated and interventional embolization (14). The patient in this case had a fusiform aneurysm at the V3 segment of the left vertebral artery, and imaging revealed that the aneurysm was fusiform with no clear aneurysmal neck and a large aneurysm volume. Endovascular treatment was chosen due to the simultaneous distortion of the vertebral artery and the proximity of the aneurysm to the brain stem and vulnerable cranial nerves.

The primary endovascular interventions for fusiform vertebral artery aneurysms are PAO and parent artery reconstruction (PAR). Before surgery, PAO may need to perform a balloon occlusion test (BOT) to confirm the compensation of the contralateral vertebral artery to the basilar artery or ipsilateral PICA, as well as good blood supply from anterior to posterior circulation. Most patients are candidates for PAO. PAO has the advantage of improving the degree of distal aneurysm embolization, reducing the number of coils, and alleviating the mass effect. It can be used to treat cranial nerve compression caused by long lesion segments, irregular shapes, or large aneurysms. However, proximal PICA occlusion is easily caused, resulting in a significant increase in the incidence of ischemic events, especially when the lesion involves PICA or the diseased vessel is the basilar artery's dominant supplying artery. PAR is appropriate for patients who are unable to tolerate BOT tests, have a small aneurysm, and are not expected to rupture prematurely (15, 16). According to the preoperative imaging examination, the aneurysm tumor was fusiform and larger than 1.5 cm in size, and the PICA measured during DSA was approximately 1 cm away from the distal end of the aneurysm. PAR may cause ischemic events in the PICA or other perforator vessels in the long run. In contrast, PAO is less likely to cause ischemic events. As a result, PAO is chosen to simultaneously embolize aneurysms. During the procedure, a single catheter was used to gradually release the coil. Open surgery was another option for this patient. Direct clipping is exceedingly challenging in this situation because there is not a definite neck of the aneurysm. Tsunoda et al. (17) reported a case of surgical removal using V3-radial artery graft-V4 bypass and occipital artery-posterior inferior cerebellar artery bypass for a giant thrombosed aneurysm. In this approach, the complete restoration of the distal vertebral artery blood flow is consistent with the physiological function of the human body. While the procedure is being performed, the connection between the cranial nerve and aneurysm can be investigated, as well as any potential involvement of further vessels. However, endovascular techniques are believed to be less intrusive than open surgical manipulation. The risk of surgery was particularly enhanced by the size of the aneurysm and its near proximity to peripheral nerves and blood vessels in this case. Meanwhile, another drawback of surgery is that it takes a long time and is extremely complicated and invasive.

When the zygomatic branch of the facial nerve is stimulated in patients with HSF, an abnormal electrical change in the other branches is detected, which is known as LSR. The mechanism by which it occurs is unknown (18). The compression of responsible

TABLE 1 Summary of reports of symptomatic hemifacial spasm caused by aneurysm of the vertebral artery.[α]

Author (year)	Age, sex	Side	Type	Period of HFS	Size of aneurysm (mm)	Treatment	LSR	Decomp	Outcome
Moriuchi et al. (4)	62, F	Left	Saccular	6 years		Neck clipping	-	+	Disappeared immediately
Tsuchiya et al. (5)	71, F	Right	Fusiform	11 years	-	Neck clipping	-	+	Disappeared immediately
Sato et al. (6)	53, M	Left	Saccular	2 years	15×8×4	Coil embolization	-	-	Disappeared 6 months later
Murakami et al. (7)	49, F	Right	Fusiform	9 months	-	Coil embolization	+	-	Disappeared 6 months later
Uchino et al. (8)	59, M	Left	Fusiform	18 months	-	MVD only	-	+	Disappeared immediately
Nakagawa et al. (9)	55, F	Left	Fusiform	2 years	13	Coil embolization	-	-	Disappeared 3 months later
Lee et al. (4)	69, M	Left	Fusiform	5 years	-	Extracranial VA ligation	-	+	Disappeared immediately
Iida et al. (10)	59, M	Left	Saccular	2 months	5.5	Coil embolization	-	-	Disappeared immediately
Our case	59, M	Left	Fusiform	1 year	15 × 13	Coil embolization	+	-	Disappeared 1 month later

[α]F, female; M, male; Decomp, decompression; HFS, hemifacial spasm; LSR, lateral spread response.

blood vessels, demyelination of facial axons, and formation of reverse afferent stimuli via pseudosynapses all contribute to the production of LSR. When the responsible vessel is separated from the nerve, the LSR should vanish immediately, allowing the effectiveness of MVD to be monitored. However, because HFS is rarely caused by the aneurysm itself, the mechanism of HFS remission following endovascular treatment of aneurysms remains unknown. Nakagawa et al. (9) reported a patient with HFS caused by a contralateral fusiform aneurysm, whose symptoms improved gradually 3 months after aneurysm interventional embolization. The gradual improvement in the aberrant excitatory circuit at the REZ may parallel the gradual decrease in facial nerve compression following intravascular treatment of the aneurysm, alleviating the hemifacial spasm. The reduction of the aneurysm mass effect was attributed to the remission of HFS symptoms. Similarly, the relief of ophthalmic paralysis after endovascular therapy for cavernous sinus aneurysm and the relief of oculomotor nerve paralysis after endovascular therapy for posterior communicating aneurysm have been reported (19, 20). Iida et al. (10) reported a case of a patient with HFS caused by the intravascular treatment of the saccular aneurysm of the vertebral artery. The authors believed that the relief of HFS symptoms was due to the relief of aneurysm pulsation rather than the relief of compression. In this case, the direct electrophysiological monitoring of the aneurysm revealed that LSR disappeared immediately after aneurysm embolization, and the patient's spasm of the right hemifacial muscle was immediately relieved after surgery. The aneurysm volume did not decrease significantly after aneurysm embolization, but we discovered that LSR disappeared immediately after endovascular therapy, so we believe that aneurysm pulsation is important in HFS. However, when monitoring MVD, it has been observed that LSR disappears in some patients after opening the dura mater, releasing cerebrospinal fluid, and pulling the cerebellum. Thirumala et al. (3) hypothesized that the surgical procedure preceding the separation of the responsible vessels could alter the local anatomical relationship between the responsible vessels and the facial nerve, acting as a temporary decompression. As a result, whether there will be slight morphological changes after aneurysm endovascular treatment, resulting in changes in the occupying effect, needs to be confirmed further. Liu et al. (21) proposed that only a "just right" compression can generate and trigger the symptom. "Just right compression" means an appropriate compression with a certain specific frequency, amplitude, and angle. In fact, when PAO is conducted in this case, changes in some of the parameters of the compression, such as angiodynamics, will cause symptoms to halt. Simultaneously, the abnormal impulse generation would cease.

4. Limitation

The main limitation of this study is the lack of enough cases. We cannot provide controlled clinical research to demonstrate the availability and efficacy of our strategy. Following a direct procedure, such as MVD, HFS typically disappears right away. However, it takes time for HFS to recover after endovascular therapy (10). Although the symptoms completely disappeared after a month, a more-than-6-Month follow-up is needed. If the spasm recurs, open surgery such as a bypass or MVD could be an excellent choice.

5. Conclusion

HFS caused by a fusiform aneurysm of the vertebral artery is extremely rare. In this case, endovascular treatment can not

only prevent aneurysm rupture but also successfully relieve HFS. Intraoperative monitoring of LSR can effectively predict postoperative HFS efficacy. The reason may be the disappearance of aneurysm wall pulsation after endovascular therapy, but the specific reason needs to be further verified. Endovascular therapy can be a safe and effective way to relieve HFS caused by compression of vertebral aneurysms.

Data availability statement

The original contributions presented in the study are included in the article/Supplementary material, further inquiries can be directed to the corresponding author.

Ethics statement

Written informed consent was obtained from the individual(s) for the publication of any potentially identifiable images or data included in this article.

Author contributions

PH put forward research ideas and took the responsibility of communicating with the patient's family and obtaining the authorization of this study. HJ was responsible for drafting articles and revising the article. ZL was responsible for literature searches and final proofreading. All authors contributed to the article and approved the submitted version.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Supplementary material

The Supplementary Material for this article can be found online at: https://www.frontiersin.org/articles/10.3389/fneur.2023.1203751/full#supplementary-material

SUPPLEMENTARY VIDEO 1
Patient had involuntary left facial spasm before Interventional operation.

SUPPLEMENTARY VIDEO 2
On the first day after Interventional operation, the patient's left facial spasm were significantly relieved.

SUPPLEMENTARY VIDEO 3
The patient's symptoms did not completely disappear.

SUPPLEMENTARY VIDEO 4
The patient's symptoms completely resolved after 1 month.

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Case report: Mechanical thrombectomy for acute basilar artery occlusion via persistent hypoglossal artery

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Persistent hypoglossal artery (PHA) is a rare carotid-vertebrobasilar anastomosis in adults. Here, we report a case of mechanical thrombectomy for acute basilar artery occlusion via the PHA. A 44-year-old man was admitted to our stroke unit with an unstable gait and aphasia for 2 h. The baseline National Institutes of Health Stroke Scale (NIHSS) score was 4, but the clinical symptoms continued to worsen. Computed tomography angiography showed the absence of the basilar artery and an abnormal anastomosis between the anterior and posterior circulation. Clinical symptoms continued to worsen, and endovascular treatment was scheduled. PHA was demonstrated and basilar artery occlusion was confirmed using digital subtraction angiography. Mechanical thrombectomy with a stent retriever and aspiration was performed via the PHA, and modified thrombolysis in cerebral infarction level 3 was achieved. The patient underwent intravenous antiplatelet therapy after the operation, and follow-up neuroimaging revealed multiple small infarcts in the cerebellum and medulla oblongata. The patient was discharged after 10 days for further rehabilitation, with an NIHSS score of 25. At 10 months follow-up, the NIHSS score decreased to 18. Recognition of this rare variation is particularly important for interventional strategy determination and rapid recanalization of basilar artery occlusion.

KEYWORDS

persistent hypoglossal artery, acute ischemic stroke, basilar artery occlusion, thrombectomy, case report

Introduction

Basilar artery occlusion (BAO) is the most serious type of acute ischemic stroke. Failure to achieve rapid recanalization can lead to high mortality and disability rates and poor clinical prognosis (1). The safety and efficacy of mechanical thrombectomy (MT) for the treatment of acute large-vessel occlusion in the anterior circulation have been confirmed in many studies (2). Multiple randomized controlled trials (3–6) have shown promising results for MT for acute BAO, with better clinical outcomes and reduced mortality.

Persistent hypoglossal artery (PHA) is a rare carotid-vertebrobasilar anastomosis in adults. Few studies have reported BAO accompanied by PHA. In this case, the patient with BAO received MT, and PHA was confirmed during the operation. Such variation is of great importance in the determination of an endovascular treatment (EVT) strategy for BAO, which includes the establishment of MT access with sufficient supporting strength and the evaluation of compensatory situations.

Case description

A 44-year-old man was admitted to our stroke unit with unstable gait and dysarthria for 2 h. The baseline National Institutes of Health Stroke Scale (NIHSS) score was 4 and the Modified Rankin Scale (mRS) score was 1. As the clinical symptoms progressively worsened, the NIHSS score increased to 33 and the mRS score to 4 within 95 min. The patient had a heavy smoking history and primary hypertension treated with nifedipine. In addition, the patient had a history of undefined intracranial hemorrhage; therefore, thrombolytic therapy was a contraindication.

Multimodal computed tomography (CT) was performed, and intracranial hemorrhage was excluded (Figure 1A). CT angiography (CTA) showed the absence of the basilar artery (BA) and an abnormal anastomosis between the internal carotid artery (ICA) and the intracranial segment of the vertebral artery (VA) located in the hypoglossal canal (Figures 1B, C). The size of the area with cerebral blood flow >30% and delay time <3 s were determined using CT perfusion. The mismatch volume was 104 mL (Figure 1D), determined using MIStar software (Apollo Medical Imaging Technology, Melbourne, Australia). Digital subtraction angiography was performed via the right radial access and showed complete BAO with a modified Thrombolysis in Cerebral Infarction (mTICI) level of 0. PHA originated from the right ICA and anastomosed to the right VA. Right subclavian artery angiography revealed hypoplasia of VA origin. Left middle cerebral artery occlusion with collateral vessel formation and a slender left VA were also observed (Figures 1E–H).

Treatment

Great difficulties were encountered in the establishment of MT access with sufficient supporting strength via transradial access; therefore, transfemoral access was selected. A triaxial system, including a five French aspiration catheter (Navien, Medtronic, Irvine, CA, USA), microcatheter (Prowler Select Plus, Cerenovus, Bridgewater, NJ, USA), and stent retriever (Solitaire AB, Medtronic), was chosen for MT. A 4 × 20 mm Solitaire AB stent was deployed from the P1 segment of the left posterior cerebral artery (PCA) via the PHA for thrombectomy.

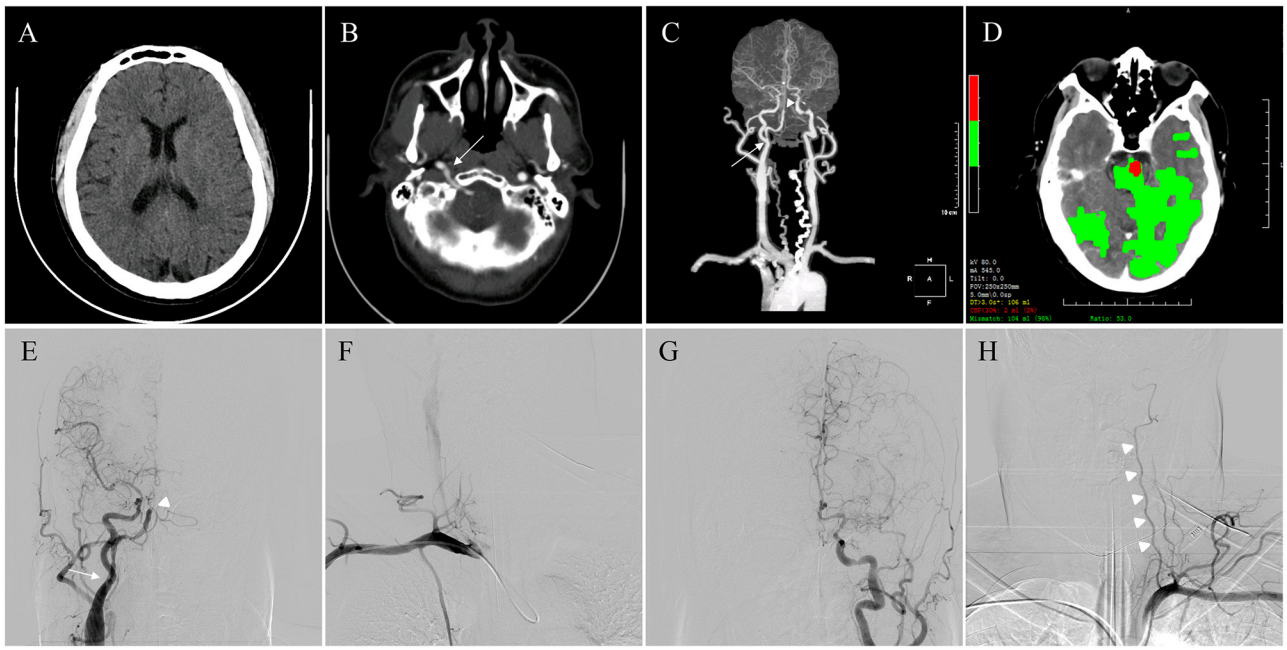

FIGURE 1
Preoperative multimodal neuroimaging evaluation. **(A)** Intracranial hemorrhage was excluded through computed tomography (CT). **(B)** Cross-sectional contrast CT revealed abnormal anastomosis (arrow) between the internal carotid artery (ICA) and the intracranial segment of the vertical artery (VA) located in the atlantooccipital space. **(C)** CT angiography (anterior-posterior view) showed basilar artery occlusion (arrowhead) and that right VA (arrow) originated from the ICA. **(D)** Volumes of infarct core (red area) and penumbra (green area) were evaluated automatically from CT perfusion to calculate the mismatch volume. **(E)** Right hemispheric angiography showed complete occlusion of the middle segment of the basilar artery (arrowhead) and that persistent hypoglossal artery (PHA) (arrow) originated from the right ICA. **(F)** Right subclavian artery angiography showed the absence of VA origin. **(G)** Left middle cerebral artery occlusion was observed with collateral vessel formation. **(H)** Slender left VA (arrowheads) demonstrated in left hemispheric angiography.

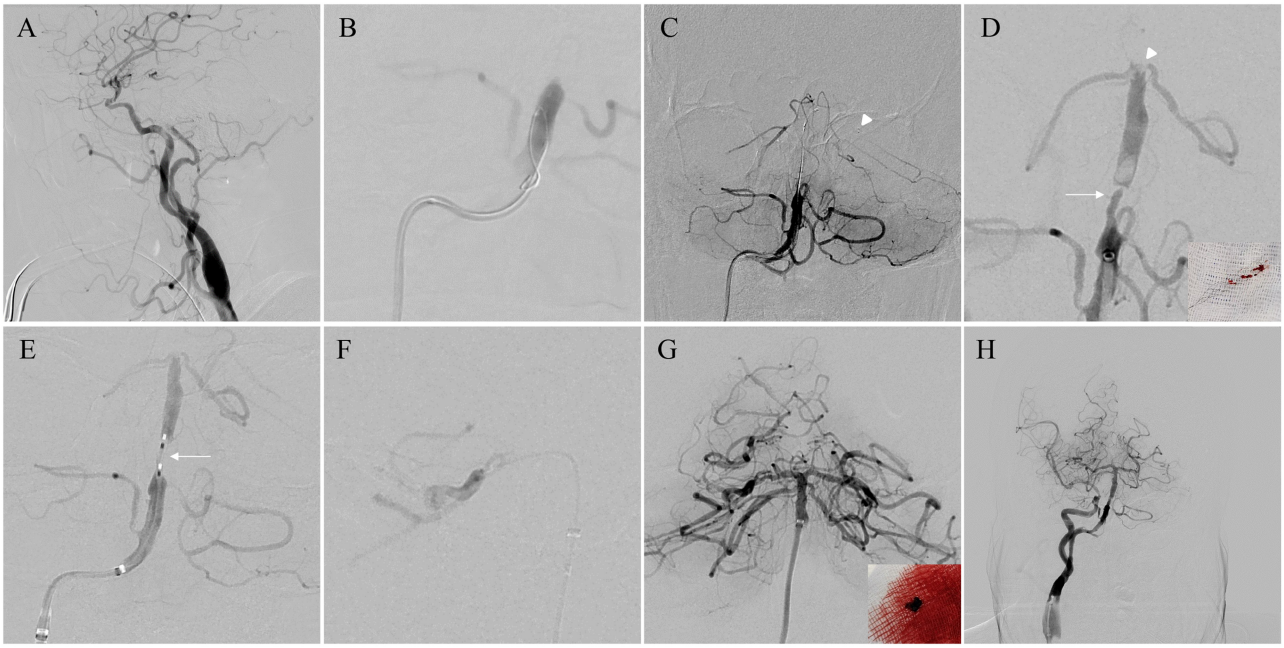

FIGURE 2
Surgical process. **(A)** Lateral view as a working projection for the establishment of access for thrombectomy. **(B)** Angiography via the microcatheter to confirm the real lumen of the distal basilar artery (BA). **(C)** The distal part of the stent retriever (arrowhead) was deployed from the left posterior cerebral artery. **(D)** After the first thrombectomy, part of the clot was removed with the stent (right corner), and cerebral angiography showed the migration of residual thrombosis to the tip of the BA (arrowhead) and severe stenosis in the BA trunk (arrow). **(E)** Angioplasty with a 3 × 9 mm balloon (arrow) in the stenotic portion of the BA. **(F)** The second thrombectomy was performed from the right persistent hypoglossal artery (PCA) with proximal aspiration. **(G)** Complete recanalization of the BA and the clot from the aspiration catheter (right corner). **(H)** Modified Thrombolysis in Cerebral Infarction (mTICI) level 3 forward flow was confirmed after 20 min.

Angiography showed thrombotic migration to the tip of the BA and severe stenosis in the BA trunk. After angioplasty with a 3 × 9 mm noncompliant balloon (Gateway, Stryker Neurovascular, Kalamazoo, Michigan), thrombectomy was performed again from the right PCA with proximal aspiration via a 5 French distal access catheter (Silver Snake, TonBridge Medical, Zhuhai, China). Complete recanalization of BA flow was achieved at an mTICI level of 3. Postoperative CT revealed no obvious intracranial hemorrhage, and a loading dose of tirofiban was administered (Figures 2A–H).

Outcome and follow-up

The patient received half loading-dose of intravenous antiplatelet therapy (Tirofiban) and the systolic blood pressure was strictly maintained between 130 and 140 mmHg after the operation. Postoperative neuroimaging follow-up revealed multiple small, newly developed infarcts in the cerebellum and medulla oblongata (Figures 3A–E). CTA performed 3 days after the operation showed good patency of the BA trunk (Figure 3F). The patient was discharged after 10 days for further rehabilitation, with an NIHSS score of 25 and a mRS score of 5. At 10 months follow-up, the NIHSS score decreased to 18 and the mRS score decreased to 4.

Discussion

Few studies have reported the occurrence of BAO accompanied by PHA (Table 1). During early development of the human embryo, various anastomotic channels exist between the carotid artery and vertebrobasilar system, which play critical roles in irrigating the posterior circulatory bed before it fully develops (7). PHA is usually involved in the 12 to 14 mm embryonic stage. Rarely, it fails to regress and persists into adult life, with an incidence of 0.03–0.09% (8). PHA is generally accompanied by other anomalies such as hypoplasia of the VA or PCA. Due to its unique anatomical and hemodynamic characteristics, PHA is a potential risk factor of posterior circulation ischemic events (9). However, cases of PHA combined with acute BAO have rarely been reported.

Earlier recanalization has been correlated with a better prognosis for acute BAO. Owing to its various clinical symptoms, advanced CT evaluations are essential to select suitable cases for patients with BAO (10). Based on the MIStar software, the ischemic penumbra and core infarction could be identified, even though the diagnostic value may be limited because of the influence of the blood flow velocity and the bony structure of the posterior fossa. In addition, CTA is helpful in distinguishing potential variations so that appropriate access catheters can be selected effectively to shorten recanalization time. Hence, the application of advanced CT evaluations is

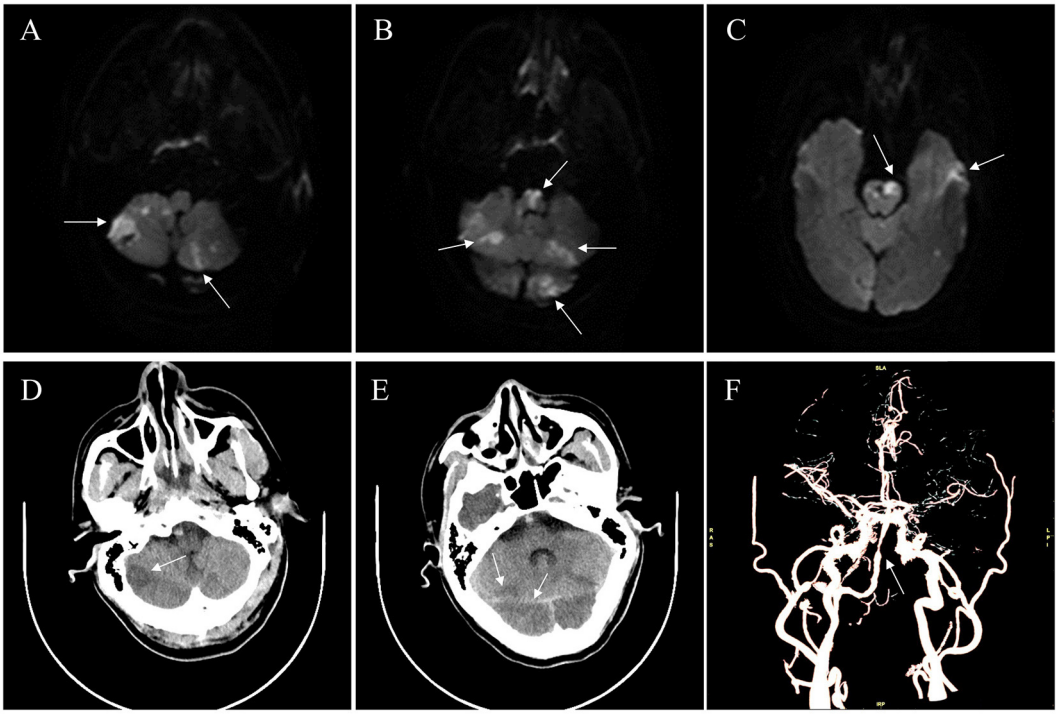

FIGURE 3
Neuroimaging follow-up. **(A–E)** Postoperative follow-up revealed multiple small newly developed infarcts (arrows) in the cerebellum and medulla oblongata on diffusion weighted imaging and computed tomography (CT). **(F)** Postoperative CT angiography confirmed the general patency of the BA trunk, with slight stenosis (arrow).

TABLE 1 Previous cases of BAO accompanied by PHA.

Previous literature	Age (years)/sex	Clinical presentation	Medical history	Baseline NIHSS	Imaging findings	TOAST type	EVT strategy	Outcome/ follow up
Kawano et al. (11)	76/Male	Consciousness lost	Severe dilated cardiomyopathy and cardiac dysfunction	N/A	PHA, BAO and occlusion of left ICA and VA	Cardioembolism	N/A	Died after 3 days
Voronovich et al. (12)	48/Male	Unresponsive with flaccid extremities	Atrial septal defect repairment	28	PHA and BAO	Cardioembolism	Aspiration	Aphasia at discharge
See et al. (8)	70/Female	Nausea, dizziness and dysphasia on awakening, followed by left hemiparesis and decreased consciousness.	Ischemic cardiomyopathy and cardiac defibrillator implantation	From 4 to 22	PHA, BAO and atherosclerotic changes at ICA	Stroke of undetermined cause	Aspiration	Mechanical ventilation and appropriate motor function improvement of limbs at discharge
Park et al. (13)	83/Female	Acute unresponsive mental deterioration onset	Hypertension	20	PHA and BAO	Stroke of other determined cause	Aspiration	Drowsy and mild dysarthria after 1 month
Present study	44/Male	Unstable gait and dysarthria	Smoking, hypertension and undefined intracranial hemorrhage	From 4 to 33	PHA and BAO	Large-artery atherosclerosis	Stent retriever, aspiration and angioplasty	NIHSS score was 25 at discharge and decrease to 18 at 10 months follow-up

BAO, basilar artery occlusion; PHA, persistent hypoglossal artery; NIHSS, National Institutes of Health Stroke Scale; TOAST, Trial of Org 101072 in Acute Stroke Treatment; EVT, endovascular treatment; ICA, internal carotid artery; VA, vertebral artery.

particularly important in surgical decision-making for BAO combined with PHA.

Previous studies (11–13) on BAO accompanied by PHA have mostly attributed it to cardioembolic stroke, and aspiration alone has a high rate of recanalization (Table 1). In this case, the mechanism of BAO was considered to be ischemic stroke based on intracranial atherosclerotic stenosis (ICAS), which is highly prevalent in East Asian populations. Although bridging intravenous treatment may confer benefits for ICAS, the patient had a history of intracranial hemorrhage; therefore, direct MT was selected. To the best of our knowledge, this is the first report of the treatment of BAO via PHA using a stent retriever with aspiration. Similar to the pathogenesis of ICAS, rescue angioplasty after MT is essential for maintaining forward blood flow. More importantly, although the BA was completely recanalized with an mTICI level of 3, embolic debris migration and limited collateral circulation may have resulted in a very poor prognosis. Despite the aforementioned methods, flow control in the proximal part of the ICA with a balloon-guiding catheter may also be more effective and should be seriously considered in subsequent clinical practice (14).

Conclusion

BAO accompanied by PHA is extremely rare, and relevant literature is lacking. Preoperative multimodal CT evaluations are helpful in identifying such vascular variations to determine an interventional strategy for earlier recanalization. Proximal flow control, aspiration with a balloon-guiding catheter, and the use of intra-arterial alteplase or tirofiban may improve the clinical outcomes of ischemic stroke based on ICAS.

Data availability statement

The raw data supporting the conclusions of this article will be made available by the authors, without undue reservation.

Ethics statement

The studies involving human participants were reviewed and approved by the Institutional Review Board (IRB) of Huashan Hospital, Fudan University, China (approval number KY2015-256). The patients/participants provided their written informed consent to participate in this study. Written informed consent was obtained from the individual(s) and/or minor(s)' legal guardian/next of kin for the publication of any potentially identifiable images or data included in this article.

Author contributions

XZ, JW, ZC, YL, YD, XC, CG, and YG contributed to the conception and design, acquisition and interpretation of the case data, drafting and revision of the article, particularly for important intellectual content, final approval of the published version, and agreement to be accountable for the accuracy or integrity of the article. All authors contributed to the manuscript and approved the submitted version.

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Conflict of interest

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Case report: Delayed quadriplegia from traumatic carotid cavernous fistula: a rare case with perimedullary venous drainage

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Background: Carotid cavernous fistula (CCF) refers to the abnormal arteriovenous communication between the carotid system at the skull base and the sphenoid cavernous sinus, which is caused by trauma in almost 75% of cases. The drainage of venous blood to the spinal cord represents a distinctive mechanism, which is commonly observed in dural arteriovenous fistula (DAVF), and typically manifests clinically as progressive myelopathy. However, it is a rare occurrence in clinical practice that traumatic carotid cavernous fistula (TCCF) causes delayed quadriplegia through perimedullary venous drainage.

Case presentation: We report the case of a 29-year-old male patient who was admitted to the hospital with a sudden onset of headache and quadriplegia. The patient had previously lost his right eye in a traffic accident 5 years ago. Cerebral angiography showed a high-flow direct CCF on the right side, accompanied by obvious drainage of cerebellar and perimedullary veins. We successfully performed coil embolization for the CCF, and the symptoms of the patient gradually improved after the operation. During follow-up at sixth-months, the patient regained the ability to walk independently.

Conclusion: We experienced a rare case of TCCF with quadriplegia. Utilizing coil embolization, we achieved successful improvement in the patient's condition. However, the mechanism and the best treatment of CCF drainage through the perimedullary vein are still unclear. We need to further explore the pathophysiological information of CCF venous drainage.

KEYWORDS

carotid cavernous fistula, perimedullary drainage, quadriplegia, coil embolization, case report

Introduction

Carotid cavernous fistula (CCF) refers to the abnormal arteriovenous communication between the carotid system at the skull base and the sphenoid cavernous sinus, usually manifested as pulsatile exophthalmos, conjunctival congestion, and intracranial murmur (1). CCF is a rare but not unique disease, with traumatic causes being the most common, accounting for approximately 75% of cases (2). Traumatic carotid cavernous fistula (TCCF) only occurs in 0.2% of the brain or in maxillofacial trauma (3) clinically, and the symptoms largely depend on the direction of venous drainage of the cavernous sinus (4, 5). While venous blood flowing to

the spinal cord is a rare drainage method commonly observed in dural arteriovenous fistula (DAVF), its clinical manifestations are predominantly progressive myelopathy. According to the classification scheme proposed by Cognard et al., this type of vascular disease is classified as Cognard V (6). However, there are few cases of tetraplegia caused by TCCF, resulting in limited information regarding its mechanism. This manuscript describes a case of TCCF presenting as quadriplegia through intraspinal drainage.

Case presentation

A 29-year-old male patient was admitted due to an acute onset of severe headache and quadriplegia. The patient's medical history can be traced back to a traffic accident five years ago, during which he sustained head and abdominal injuries. Although a cranial computed tomography (CT) scan performed at the time of the accident showed no abnormality, the accident eventually led to blindness in the patient's right eye. Over the past five years, the patient has remained asymptomatic with no clinical manifestations.

The patient was admitted to West China Hospital of Sichuan University (Chengdu, China) through the emergency department, presenting with an abrupt onset of severe headache and bilateral lower limb paralysis upon awakening during the night. A comprehensive neurological assessment of the patient elucidates right-sided blindness, graded bilateral upper limb muscle strength of 2, and graded bilateral lower limb muscle strength of 0. The patient demonstrated reduced upper limb tendon reflexes and a complete absence of knee and ankle reflexes in both lower limbs, concurrently presenting positive pathological indicators bilaterally. Upon admission, an urgent cranial CT scan showed no significant abnormalities, while head and neck computed tomography angiography (CTA) demonstrated an anomalous vascular mass in the right cavernous sinus area, contiguous with the internal carotid artery (ICA), accompanied by dilated and tortuous perimedullary vascular structures (Figure 1). The patient's spinal cord magnetic resonance imaging (MRI) examination revealed medullary and cervical cord edema, along with multiple tortuous vascular flow voids (Figure 2). These findings raised the suspicion of a carotid cavernous fistula (CCF). Subsequently, a cerebral angiography was expeditiously conducted, with the digital subtraction angiography (DSA) revealing a high-flow CCF on the right side draining into the intraspinal venous plexus and cerebellar vein (Figure 3A). The CCF's drainage into the spinal canal led to spinal cord edema and ischemia, providing an explanation for the patient's limb muscle weakness symptoms. Then contrast agent was injected into the external carotid artery, and no dural vessel branches supplying the CCF were found (consistent with Barrow Type A CCF). The patient was diagnosed with a traumatic carotid cavernous fistula (TCCF).

Based on the analysis of cerebral angiography, the treatment plan for this case was scheduled for right CCF coil embolization *via* femoral artery access. Prior to the procedure, a traditional femoral artery puncture was performed, and systemic heparinization was administered to prevent thrombus formation during the intervention. Subsequently, a neurovascular guidewire was used to guide the placement of the catheter into the petrous segment of the ICA, ensuring accurate positioning for subsequent steps. During the surgery, intraoperative DSA was employed to confirm the precise location of the fistula's opening. With the assistance of the micro guidewire, the microcatheter was carefully advanced to the site of the CCF's orifice. Utilizing precise placement, five coils were deployed to embolize and effectively occlude the abnormal arteriovenous communication (Figure 3B). The entire procedure was smoothly executed, and immediate post-interventional DSA demonstrated successful closure of the fistula, absence of draining veins, and preservation of the integrity of the ICA (Figure 3C).

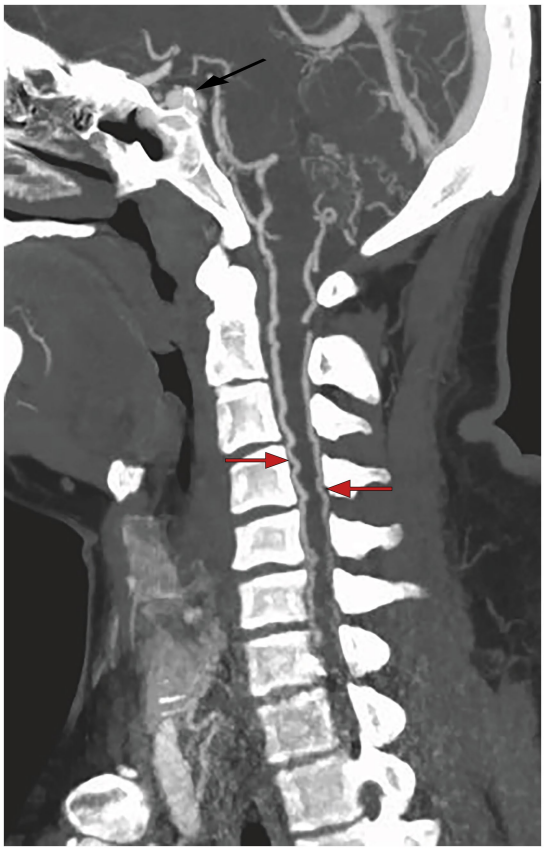

FIGURE 1
Preoperative CTA imaging showed that there was an abnormal vascular mass in the right cavernous sinus area, and prominent vascular structures could be seen around the medulla.

The patient's postoperative recovery displayed promising improvements. On the third day after the operation, the patient experienced a noticeable reduction in headache intensity, along with improvement in upper limb muscle strength, and slight toe movement. At the 1 month follow-up, the patient's limb strength continued to improve gradually, with the ability to walk assisted by crutches. Encouragingly, during the 6-month follow-up after the operation, the patient's upper limb muscle strength had essentially returned to normal, and both legs were capable of slow independent walking, significantly enhancing the patient's quality of life.

Discussion

Traumatic carotid cavernous fistula (TCCF) is a rare complication of craniofacial trauma. It was first reported in 1835, and is mostly seen

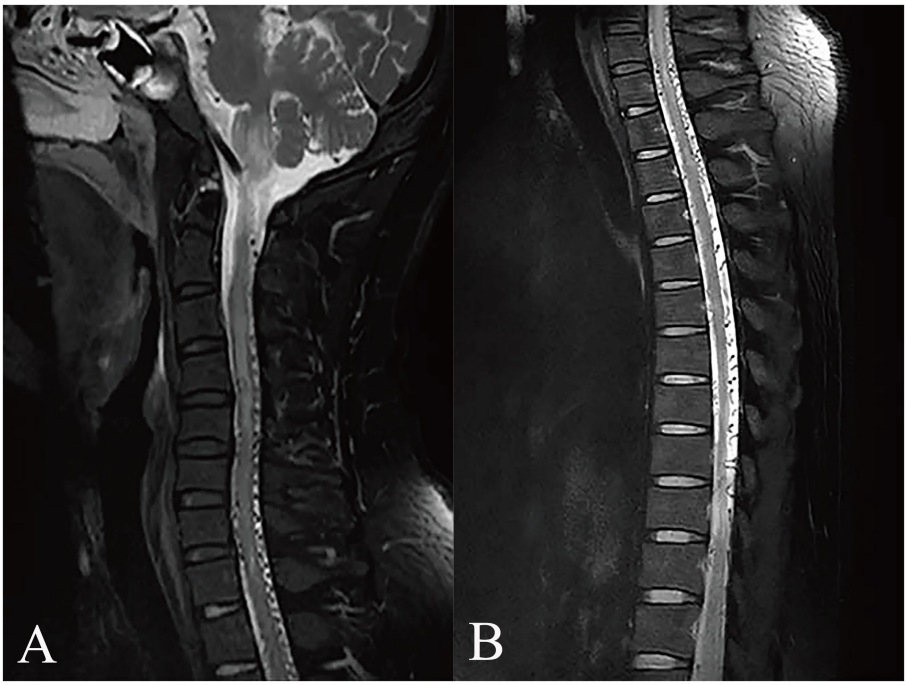

FIGURE 2
MRI examination of the affected region. The MRI scan reveals medullary and cervical cord edema with multiple tortuous vascular flow voids **(A)**. Additionally, tortuous vascular flow voids are also observed in the thoracic and lumbar cord regions **(B)**.

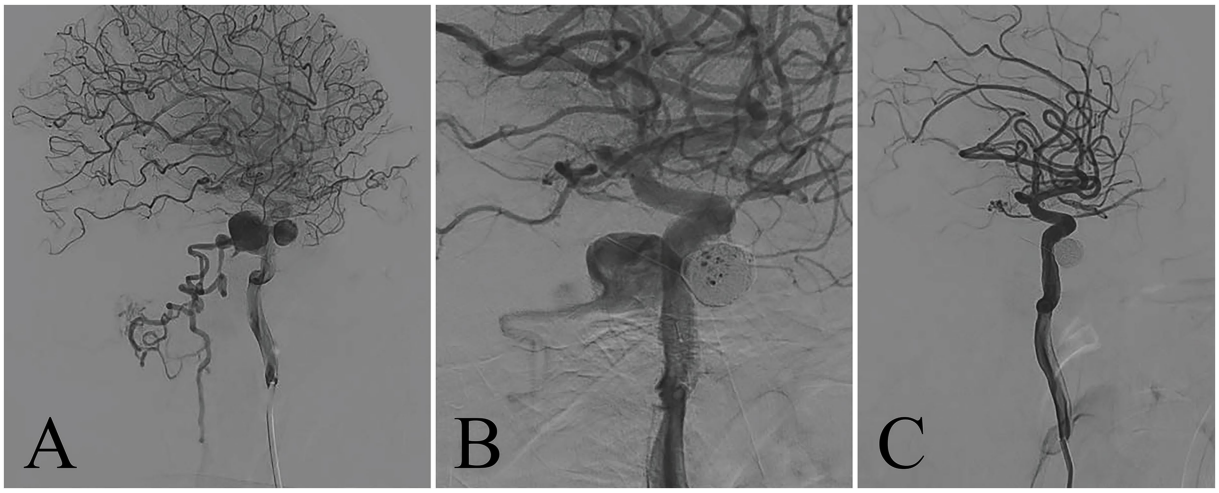

FIGURE 3
Angiography of right internal carotid artery. The preoperative DSA showed that the cavernous sinus was abnormally filled and blood flowed back through the perimedullary vein **(A)**. Intraoperative DSA for coil embolization **(B)**. After embolization of the cavernous sinus with arterial coil, the fistula was completely occluded **(C)**.

in middle-aged males. Incidents of forceful trauma can result in fractures of the skull base. The fracture fragment may directly perforate, displace, or tear the internal carotid cavernous sinus (CS) segment, leading to the rapid ingress of arterial blood into the cavernous sinus and the consequent formation of TCCF (7). In 1985, Barrow et al. (8) classified CCF into four types (type A to type D) based on the arterial architecture. TCCF represents a direct communication between the C4 segment of the internal carotid artery (ICA) and the CS, which usually belongs to high flow disease. When TCCF occurs, arterial blood tends to flow rapidly into the cavernous sinus, which increases the pressure in the sinus. Consequently, The hemodynamic alterations in the CS promote retrograde or clockwise blood flow through multiple veins (9). In 1972, Houser et al. (10) initially proposed that the clinical manifestations of CCF mainly depend on the direction of blood drainage within the CS. Diverse drainage patterns result in distinct clinical symptoms.

Typically, venous drainage patterns of TCCF can be categorized into the following four groups (11):

(1) Anterior Drainage: It involves forward flow toward the ophthalmic vein, medial canthus vein, or facial vein, eventually reaching the external jugular vein. Most CCF cases are drained to the ophthalmic vein (the most typical venous drainage pattern), resulting in the ophthalmic triad (consisting of exophthalmos, bulbar conjunctival edema, and orbital murmur) corresponding to CCF (12). (2) Posterior Drainage: The fistula may drain backward toward the superior petrosal sinus or inferior petrosal sinus, finally emptying into the internal jugular vein and draining to the pterygoid plexus through a guide vein (13). Imaging studies have demonstrated a significant increase in the flow velocity of the internal jugular vein in such cases. (3) Upward Drainage: It is drained upward to the cerebral cortex or deep vein through the lateral fissure vein. The clinical manifestations often include headaches, increased intracranial pressure, and even subarachnoid hemorrhage. (4) Contralateral Drainage: In certain instances, drainage occurs through the contralateral CS, leading to bilateral pulsating exophthalmos (14). However, it is extremely rare for TCCF to result in quadriplegia through intraspinal drainage, and only 3 cases have been reported so far (15–17). Given the unclear pathogenesis and prognosis of this particular case, we conducted a comprehensive review of the clinical characteristics of this case and the three cases previously reported in the literature.

Ricolfi. F et al. (15) described the first case of CCF with perimedullary drainage leading to myelopathy in 1999. In this case, the patient had tetrapareis, sphincter disturbance, and bulbar signs at admission. MRI revealed perimedullary vessels, and high signal intensity in the swollen medulla and cervical cord. Selective angiography demonstrated a right-sided CCF supplied by meningeal branches of the ICA and external carotid artery (ECA), draining into the superior ophthalmic vein anteriorly and posteriorly into the cervical spinal cord veins via the superior petrosal sinus and lateral mesencephalic veins. In this case, they chose to take an embolization treatment by mixing a mixture of histoacryl and lipiodol in the middle meningeal and sphenopalatine arteries and polyvinyl alcohol foam particles (PVA) particles in the ascending pharyngeal artery. However, no anticoagulant therapy was administered after embolization. This patient's neurological condition rapidly deteriorated and they passed away 5 days later. A study published in 2011 described another case of TCCF with perimedullary drainage (16). This patient had a clear history of trauma and presented to the hospital with tetraparesis. Angiography revealed a direct CCF with prominent pontine mesencephalic and perimedullary venous drainage. The physician performed coil embolization of the TCCF, which successfully improved the patient's symptoms to walk with the aid of external assistance. Two years ago, a study (17) reported the third case of TCCF with perimedullary venous drainage. In this case, the patient developed progressive gait disturbance, hyperreflexia, hypoesthesia, and pulsatile tinnitus one month after an anterior skull base fracture. Cervical MRI revealed spinal cord edema and serpentine signal flow voids, which were eventually confirmed to be TCCF with drainage into the perimedullary veins through DSA. The treatment approach chosen by the physician involved the combined use of coils and the Onyx liquid embolic system to occlude the fistula. Postoperatively, the patient experienced the complete resolution of symptoms.

In our presented case, the patient had significant spinal cord symptoms with progressive aggravation. Cerebral angiography confirmed a high-flow CCF on the right side with drainage into the spinal canal and the cerebellar veins. It is noteworthy that the patient's right eye was blind from trauma 5 years ago and the left eye did not show any abnormal symptoms and signs. We finally opted to perform TCCF embolization using coils. Postoperatively, at the six-month follow-up, the patient demonstrated remarkable progress and was able to walk independently.

Venous drainage to the spinal cord is a recognized drainage modality for DAVF, first proposed by Woimant et al. (18) in 1982. Since its initial description, several case reports and case series have provided detailed accounts of this vascular condition (19–21). According to the classification by Cognard et al. (6), this particular vasculopathy has been classified as type V based on clinical and angiographic correlation. Patients with Cognard type V DAVF often present with non-specific clinical symptoms. Due to elevated spinal venous pressure, 62% of patients exhibit progressive myelopathy, while 31% display bulbar dysfunction associated with dysautonomic signs (22). In addition, it is difficult to make a timely diagnosis of Cognard type V DAVF in clinical practice because the MRI findings of lesions in this category may resemble those seen in inflammatory demyelinating diseases, infarctions, and intramedullary tumors. Here, we report a case of TCCF presenting as tetraplegia through intramedullary drainage. It is of concern that the two reported cases in this context had eye-related symptoms prior to the onset of spinal cord symptoms. In the first case, treatment initially involved embolization because of moderate right-sided proptosis with conjunctival hyperemia, associated with a spontaneous CCF. The identification of CCF with perimedullary drainage occurred only 1 year later. The second patient was extremely similar to our case, also suffering from blindness in the right eye due to a history of trauma. This suggests that the occurrence of the rare perimedullary drainage pattern in CCF may be related to abnormal ophthalmic vein function, leading to a failure in proper drainage through this vein.

We hypothesize that the patient's traumatic accident causing blindness in the right eye 5 years ago may have resulted in impaired ocular venous circulation, gradually leading to spontaneous thrombosis. Following the discovery of the TCCF, the arterial blood flow directly bypasses the fistula ostium into the CS. The massive shunting of blood flow from the ICA leads to the reduction of blood supply to the affected intracranial artery and the opening of collateral circulation.

The venous system, on the other hand, has increased blood flow and elevated pressure, leading to the development of abnormal drainage channels. Venous blood within the CS begins to drain toward the cerebellar veins and intraspinal region due to obstruction caused by spontaneous thrombosis in the ophthalmic veins. Thus, abnormal perimedullary veins were clearly visualized on CTA imaging in this patient. Escalated blood flow and increased venous pressure in the perimedullary veins gradually led to venous dilation, causing spinal venous hypertension syndrome. This explains the patient's initial presentation of progressive quadriplegia. A study by Hassler et al. (23) demonstrated that the pressure on the intradural perimedullary vein was 60–87% higher than the mean systemic arterial blood pressure in spinal DAVF surgery. Because of the lack of valves, the intramedullary veins are directly affected by the increased pressure in the perimedullary venous plexus. Consequently, arteriovenous pressure gradients within the spinal cord decrease, leading to the development of extracellular edema and congestive myelopathy. This provided a theoretical basis for our conjecture.

Undoubtedly, this case has introduced a novel perspective for clinicians, emphasizing the importance of considering the possibility

of progressive myelopathy arising from intracranial vascular malformations. Currently, due to the rarity of this disease, a significant proportion of clinicians lack familiarity and precious therapeutic experience in managing it. However, it is crucial not to overlook the potentially life-threatening nature of TCCF through perimedullary venous drainage, underscoring the significance of early recognition, diagnosis, and treatment. We firmly advocate that CCF with venous drainage into the brainstem, cerebellum, and spinal cord should be included in the differential diagnosis of progressive myelopathy, contributing to early diagnosis and optimizing patient outcomes. In addition, in future instances resembling these cases, particular attention should be paid to the patient's ocular symptoms and the reflux of ocular veins, as such observations might offer valuable insights into the underlying mechanism of these cases.

Conclusion

We encountered an exceptional and rare case of TCCF with quadriplegia. The application of coil embolization demonstrated notable efficacy in improving the patient's condition. However, the precise mechanism and optimal treatment strategy for CCF drainage through the perimedullary vein remain enigmatic. Further exploration of the pathophysiological aspects related to CCF venous drainage is imperative to gain a comprehensive understanding and valuable insights.

Data availability statement

The original contributions presented in the study are included in the article/supplementary material, further inquiries can be directed to the corresponding author.

Ethics statement

Written informed consent was obtained from the individual(s) for the publication of any potentially identifiable images or data included in this article.

Author contributions

Y-HM wrote the manuscript and edited the images of the article. Y-HM, RS, and S-HL revised the existing literature together. Y-HM, SL, and TW collected patient data and provide corresponding explanations. C-WZ completed the surgery and designed this study. All authors have contributed to the article and approved the submitted version.

Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Case report: Usefulness of angioscopy in determining antiplatelet drug reduction after carotid artery stenting

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We report a case in which neointima was confirmed by angioscopy and antiplatelet drug administration was reduced 2months after carotid artery stenting (CAS). A patient in their 80s was scheduled to undergo resection for renal cancer; however, he also had right cervical internal carotid artery stenosis. Because this was a risk for general anesthesia, CAS was performed after first starting dual antiplatelet therapy. Urologically, early reduction of antiplatelet drugs was necessary for a nephrectomy. Although no obvious neointima could be identified on ultrasound 2months after CAS, thin neointima was observed using angioscopy. Based on the above results, we reduced the antiplatelet drug administration, and then the nephrectomy was performed. Ultimately, no cerebral infarction occurred in the perioperative or postoperative periods. Angioscopy allows for visual confirmation of thin neointima. If sufficient neointima can be confirmed, antiplatelet drug reduction can be performed more safely and reliably.

KEYWORDS

angioscopy, angiography, carotid artery stenting, dual antiplatelet therapy, neointima

Introduction

Before carotid artery stenting (CAS), dual antiplatelet treatment (DAPT) is necessary to prevent thrombosis (1–4). Long-term DAPT carries the risk of bleeding problems; thus, it is preferable to reduce the dose as soon as possible (3, 4). However, it is unclear how long it should be continued following CAS. If the struts of the stent are suitably coated with neointima, the risk of thrombosis is decreased, and antiplatelet medications may be stopped early. However, because early neointima is thin, it can be challenging to detect using carotid ultrasonography (CUS) or angiography. In this work, we describe a case in which CUS was unable to identify the neointima, but macroscopic inspection using angioscopy could detect the presence of a thin neointima, thereby allowing for a reduction in antiplatelet medication 2 months after CAS.

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Case report

A patient in their 80s was scheduled to undergo resection for renal cancer, but he was referred to our department because right cervical internal carotid artery stenosis was suddenly discovered when using CUS for screening before general anesthesia. Right common carotid artery angiography revealed 82% stenosis (North American Symptomatic Carotid Endarterectomy Trial method), even though it was asymptomatic (Figures 1A,B); therefore, after beginning DAPT (aspirin and clopidogrel), CAS utilizing CASPER Rx (Terumo, Tokyo, Japan) was conducted (Figures 1C,D). In terms of urology, an early nephrectomy was required along with a reduction in antiplatelet medication.

Two months after CAS, another workup with angiography and angioscopy was conducted because it was challenging to detect the neointima within the stent using CUS (Figure 2A). After systemic heparinization, right common carotid angiography showed a radiolucent gap between the stent and lumen of the artery, suggesting neointimal formation (Figure 2B). After that, thin neointimal development was observed throughout the stent, except for the external carotid artery orifice, where the lumen was visually inspected with an angioscope VISIBLE (Intertec Medicals, Osaka, Japan) under proximal blood flow obstruction with a balloon catheter (Figure 2C). The obstruction time was less than 1 min and there were no ischemic symptoms.

Based on the above findings, we determined that the risk of thrombosis was low, decreased the antiplatelet drug administration to one (only aspirin), and then conducted the nephrectomy at the department of urology. In the end, there was no perioperative or postoperative cerebral infarction.

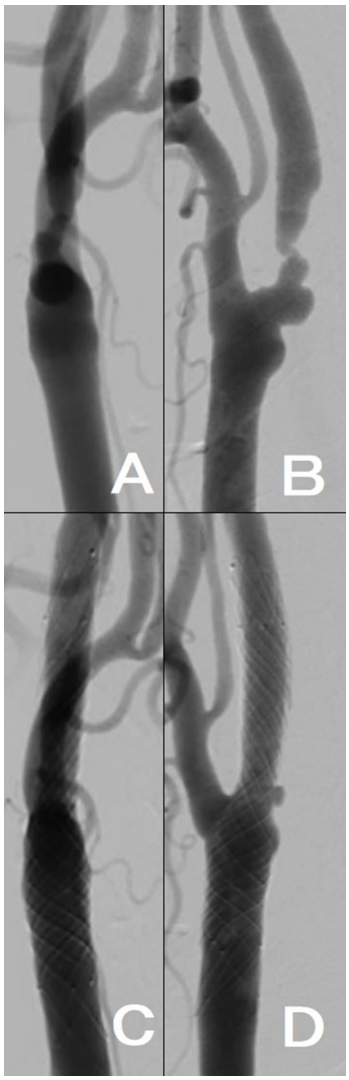

FIGURE 1
(A,B) Right internal carotid artery origin was 82% stenosed (North American Symptomatic Carotid Endarterectomy Trial method) according to the results of right common carotid artery angiography (ICA). **(C,D)** CASPER Rx was placed at the right ICA; the blood flow improved. [**(A,C)**, anteroposterior view; **(B,D)**, lateral view].

Discussion

Although there are accounts of angioscopy evaluations of plaque before and after (5–7), there are no reports of angiography evaluations of neointima sometime after CAS. In this report, CUS was conducted 2 months after CAS, but no neointima could be identified. However, investigations on coronary arteries have indicated that neointima are typically seen 1–1.5 months following the implantation of a bare-metal stent (8). Although it is challenging to decide whether or not to reduce the antiplatelet drug administration based on the findings of CUS alone, direct confirmation of the neointima by angioscopy may allow a safer

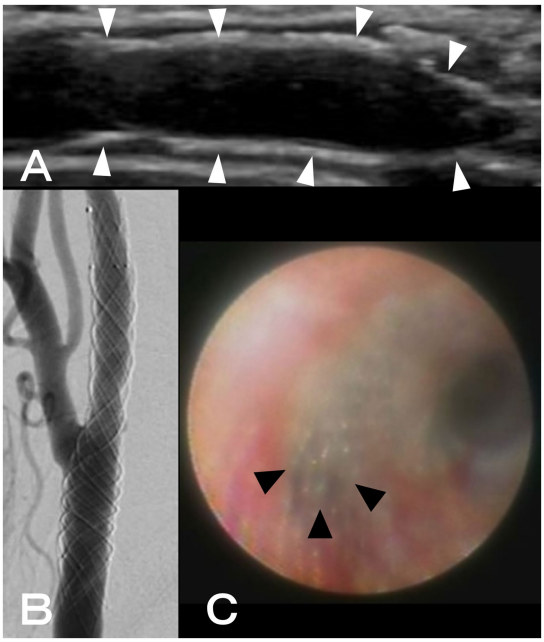

FIGURE 2
(A) Using ultrasonography, neointimal formation in CASPER Rx could not be detected (white arrowheads: CASPER Rx). **(B)** A radiolucent gap between the stent and artery lumen was discovered during right common carotid angiography. **(C)** Using angioscopy, thin neointimal development was seen throughout, except for the external carotid artery orifice (black arrowheads: the orifice of the external carotid artery).

and more reliable decision to reduce the dose. Other intravascular ultrasound devices exist, such as intravascular ultrasound and optical coherence tomography; however, similar to CUS, identification is problematic if the neointima is thin. In general, neointima may be formed more or less 2 months after CAS, and it is possible that antiplatelet medication can be lowered or stopped at this point. The timing of neointima development may vary if a stent with a different shape, such as a single layer, is employed over a dual layer stent, such as the one used in this study. On the one hand, a dual layer is less likely to form neointima because of the increased amount of metal; on the other hand, it is more likely to form neointima because of the increased scaffolding.

A drawback of an angioscope is that, depending on the blood vessel's diameter, it is rigid and has a narrow field of vision, making it challenging to thoroughly inspect the interior of a stent. It is a somewhat more intrusive test than conventional angiography because proximal blood flow blockage is required to provide a decent viewing field.

Conclusion

Thin neointima that is not visible under CUS can be visually confirmed using angioscopy. Antiplatelet drug lowering can be conducted more securely and consistently if enough neointima can be verified. Antiplatelet medication may typically be lowered or stopped at that point because in-stent neointima formation is complete 2 months after CAS.

Data availability statement

The original contributions presented in the study are included in the article/Supplementary material, further inquiries can be directed to the corresponding author.

Ethics statement

The studies involving human participants were reviewed and approved by Osaka Police Hospital. The patients/participants provided their written informed consent to participate in this study. Written informed consent was obtained from the participant/patient(s) for the publication of this case report.

Author contributions

KF, MS, SA, TM, YuS, HH, YMu, RM, RT, YaS, and YMo contributed to the work described in this paper, involved in the clinical management of the patient, and revised the manuscript. KF and MS conceived and designed the experiment, and drafted the manuscript. SA and YM supervised and coordinated the study and the manuscript. All authors contributed to the article and approved the submitted version.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Supplementary material

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Case report: Retrograde endovascular recanalization of vertebral artery occlusion with non-tapered stump via the deep cervical collateral

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Introduction: Vertebral artery (VA) occlusive disease is the major cause of posterior circulation ischemic stroke. Endovascular recanalization has been reported as a feasible treatment for patients with symptomatic VA occlusion refractory to optimal medical therapy. However, VA occlusion with non-tapered stump exhibits a low technique success rate when treated by antegrade endovascular therapy because of increased difficulty in passing the guidewire into the occluded segment. Herein, we presented a novel endovascular approach to recanalize chronically occluded VA with a non-tapered stump using a retrograde method *via* the deep cervical collateral, which has not been reported before.

Case presentation: The present case was a patient with VA ostial occlusion with non-tapered stump and distal severe stenosis of the left VA who had recurrent posterior circulation transit ischemic attacks under optimal medical therapy. CT angiography demonstrated proximal non-tapered occlusion and distal severe stenosis of the left VA, and that the right VA did not converge with the left VA into basilar artery. Endovascular treatment was recommended and performed on this patient. However, antegrade endovascular recanalization of the left VA origin occlusion failed because the micro guidewire was unable to traverse the occluded segment. Fortunately, robust collateral from the deep cervical artery to the V3 segment of the left VA developed, in which we advanced the micro guidewire to the V3 segment of the left VA and reversely passed the micro guidewire through the occluded segment. Then, the occlusion and stenosis of the left VA were successfully resolved with angioplasty and stenting. After the procedure, the patient reported no neurological symptoms under medical therapy during 3-month follow-up.

Conclusion: Antegrade endovascular recanalization of VA occlusion with a non-tapered stump is a challenge. The retrograde endovascular method *via* the cervical collateral may be an alternative for this type of VA occlusion, which requires further exploration.

KEYWORDS

vertebral artery occlusion, non-tapered stump, ischemic stroke, endovascular recanalization, retrograde, deep cervical collateral

Introduction

Vertebral artery (VA) occlusive disease is a major cause of posterior circulation ischemic events, which accounts for approximately 20–32% of the transient ischemic attacks (TIAs) or ischemic strokes in the posterior circulation (1, 2). Atherosclerosis is the common etiology for this disease with the VA ostium as the most common site prone to be involved (3, 4). Current management of VA occlusive disease includes anti-thrombotic therapy, risk factor modification, open surgery, and endovascular treatment (5). Although the optimal strategy for preventing stroke occurrence in symptomatic patients with VA occlusion is controversial and empirical, the perspective that open surgery and endovascular treatment are important complementary treatments for symptomatic VA stenosis or occlusion refractory to optimal medical therapy seems to be well recognized (5–7). Open surgery including bypass surgery, vertebral endarterectomy, and hybrid surgery has been considered as a therapeutic option for VA occlusion (8–10), which, however, is not commonly performed due to the complexity and serious complications of these procedures (6). Recently, endovascular revascularization has been reported as a feasible treatment for VA occlusion with a high success rate of 86% and a low rate of periprocedural complications of 12% (5, 6). However, VA occlusion with a non-tapered stump exhibits a relatively low technique success rate of 71.4% when treated by antegrade endovascular revascularization because of increased difficulty in finding the access for the micro guidewire to pass through the occluded segment (6). In this report, we presented a novel approach to endovascular recanalize chronically occluded VA with a non-tapered stump using a retrograde method via the deep cervical collateral, which, to the best of our knowledge, has not been reported before.

Case presentation

A 57-year-old man with a previous history of hypertension and stroke was referred to our hospital with transient episodes of dizziness, diplopia, and left-side numbness for 2 weeks. Each episode lasted for 5 to 30 min without unconsciousness and then completely relieved. After the onset of these symptoms, the patient was first admitted to a local hospital where he received optimal medical therapy including dual antiplatelet agents (aspirin 100 mg/day and clopidogrel 75 mg/day) and risk factor modifications. However, the patient still experienced another two episodes of TIAs under optimal medical treatment. His radiological examination in the local hospital suggested multifocal intracranial and extracranial atherosclerosis (data not shown). The patient was, therefore, transferred to our hospital for further management. After admission, the patient underwent a series of diagnostic evaluations. Neurological examination showed no permanent neurological disability with a National Institute of Health Stroke Scale score of 0. Laboratory tests including blood routine examination, hepatorenal function, lipid profile, homocysteine level, glycosylated hemoglobin level, and coagulation function were all normal. Multimodal computed tomography (CT) was also performed, and non-contrast CT demonstrated a hypodensity lesion in the left thalamus (Figure 1A). CT angiography (CTA) showed proximal non-tapered occlusion and distal severe stenosis of the left VA (Figure 1B), and that the right VA did not converge with the left VA into the basilar artery (Figure 1C). Based on these findings, the diagnosis of TIA due to VA occlusion was made. Since the patient had recurrent neurological symptoms despite optimal medical management, endovascular recanalization of the left VA was recommended.

The procedure was performed under local anesthesia. After the femoral artery puncture, an 8F artery sheath was inserted, and the patient was intra-arterially heparinized to achieve an activated clotting time of more than 250 s. An 8F guiding catheter (Cordis, Florida, USA) and a 5F diagnostic catheter (Cordis, Florida, USA) were delivered to the left subclavian artery proximal to the VA ostium under the guidance of a 0.035-in loach guidewire (Terumo, Tokyo, Japan). After retracting the loach guidewire, an initial digital subtraction angiography (DSA) was performed *via* the diagnostic catheter, which demonstrated non-tapered occlusion of the V1 segment and severe stenosis (approximately 80%) of the V4 segment of the left VA. In the beginning, multiple attempts were performed with the coaxial assembly of a PT 0.014-in micro guidewire (Boston Scientific, Boston, USA) and an Excelsior SL-10 microcatheter (Stryker, Michigan, USA) to facilitate navigation across the occluded segment but failed (Figure 2A). At the moment, robust deep cervical collateral to the distal V3 segment of the left VA and a tapered stump of the distal part of the occluded segment were noted (Figure 2B). Therefore, the exchange of a Synchro 0.014-in micro guidewire (Stryker, Michigan, USA) was performed to reach the distal V3 segment through the left deep cervical artery, which then reversely traversed the occluded segment to the left subclavian artery successfully (Figures 2C, D). A Neuro RX 2.75 ×15 mm balloon (Sinomed, Tianjin, China) was subsequently advanced to the VA ostium along the micro guidewire to dilate the occluded segment (Figure 2E). After balloon dilation, the occluded segment was successfully recanalized but remained in severe stenosis (Figure 2F). We retracted the balloon and advanced a 5F intermediate catheter (Tonbridge, Zhuhai, China) with a Transend 0.014-in micro guidewire inside (Stryker, Michigan, USA), which passed the recanalized segment and was placed in the left VA and posterior cerebral artery. DSA revealed severe stenosis of the V4 segment and antegrade filling of the basilar artery and both posterior cerebral arteries (Figure 2G). Under the guidance of an angiogram, a NOVA 4.0 ×15 mm balloon-expandable stent (Sinomed, Tianjin, China) was implemented in the stenotic segment (Figures 2H, I). To prevent restenosis of the V1-V2 segment, a RX 4.0 ×18 mm balloon-expandable stent (Abbott, California, USA) and a Bridge 4.0 ×18 mm rapamycin drug-eluting stent (Microport, Shanghai, China) were consecutively implanted in the recanalized segment (Figure 2J). Finally, the 8F guiding catheter, intermediate catheter, and micro guidewire were carefully retracted.

After the procedure, the patient underwent a repeated CTA examination, which indicated successful recanalization of the left VA (Figure 1D). Dual antiplatelet therapy and risk factor control strategies were continued in this patient. During the follow-up of 3 months, the patient reported no neurological symptoms. Figure 3 summarizes the timeline of the present case.

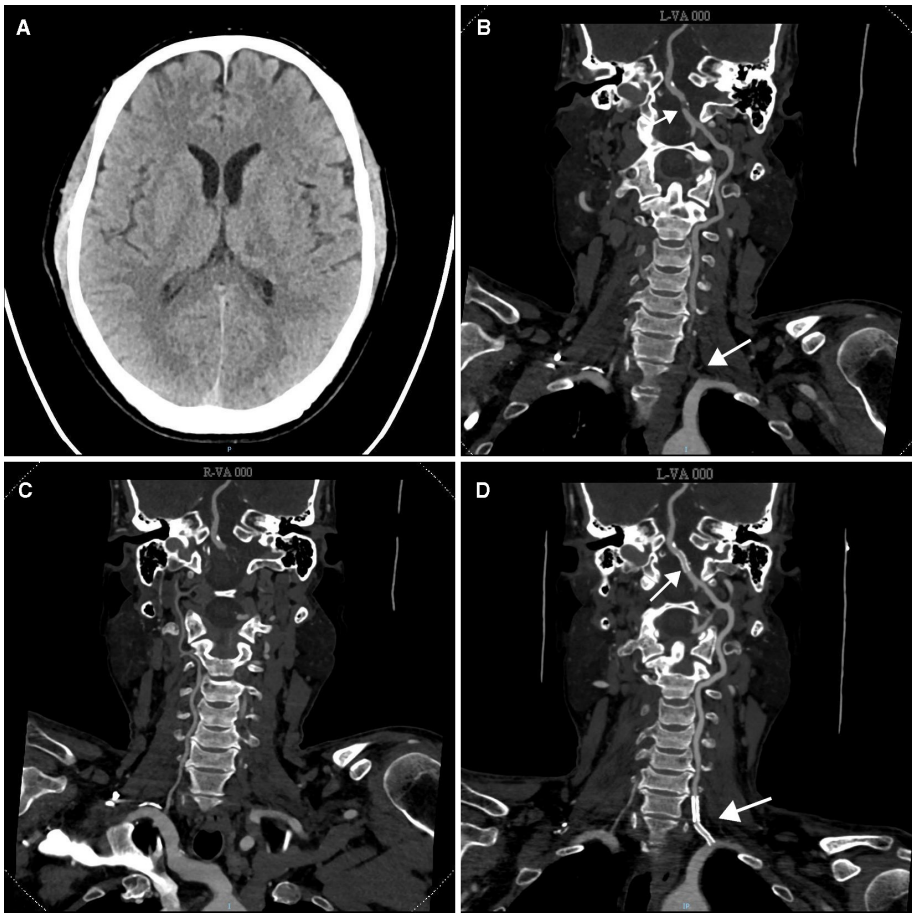

FIGURE 1
Pre- and post-treatment CT evaluation of the patient. **(A)** Pre-treatment non-contrast CT scan demonstrated a hypodensity lesion in the left thalamus. **(B)** Cervical and cerebral CT angiography (CTA) showed proximal occlusion with a non-tapered stump (white arrow) and distal severe stenosis of the left vertebral artery. **(C)** CTA showed that the right vertebral artery did not join in the basilar artery. **(D)** Post-treatment CTA indicated that the left vertebral artery was recanalized with stent placed inside (white arrow).

Discussion

VA occlusive disease is a common cause of TIAs or strokes in the posterior circulation (1, 2), and VA ostial occlusion accounts for approximately 9% of posterior circulation infarcts (11). The mechanisms of ischemic cerebrovascular events caused by VA occlusive disease include hemodynamic impairment and vertebral artery stump syndrome (12, 13). Based on the angiographic findings, we speculated that the mechanism of the recurrent neurological symptoms of ischemia in the present patient was hemodynamic impairment due to VA occlusive disease. Although the patient did not undergo perfusion examination, DSA demonstrated antegrade filling of the basilar artery, which suggested that the source of blood supply to the posterior fossa might mainly come from the abundant collateral from the deep cervical artery. However, severe stenosis of the V4 segment of the left VA limited blood flow to the posterior fossa, and the right VA did not converge with the left VA into the basilar artery, which resulted in hemodynamic impairment. Therefore, relieving the stenosis of intracranial VA was the key to improve the hemodynamics and alleviate symptoms in this patient.

The two main strategies to regain the blood supply to the posterior fossa from an occluded VA are open surgery and endovascular treatment, which have been considered as complementary alternatives for patients with recurrent ischemic events under optimal medical therapy (6). Open surgery is limited to be performed in clinical practice because of technical complexity and high rates of morbidity and mortality (6, 14). Recently, several reports have evaluated the efficiency and safety of endovascular recanalization for symptomatic VA occlusion (5, 6, 14, 15). The technique success rate of endovascular recanalization of VA occlusion ranges from 86.0 to 91.3%, and the rate of periprocedural complications was 4.3–12.0% (5, 6), suggesting the feasibility of this treatment. Since the patient had recurrent neurological symptoms under optimal medical treatment, endovascular treatment was recommended. However, we encountered challenges in navigating the occluded segment of the non-tapered VA occlusion with a micro guidewire, which was the first step to recanalize the artery. This was not surprising as recent studies demonstrated that the stump morphology is a strong predictor of successful recanalization for extracranial VA occlusion, where the technique success rate of an occlusion with a tapered stump is significantly higher than

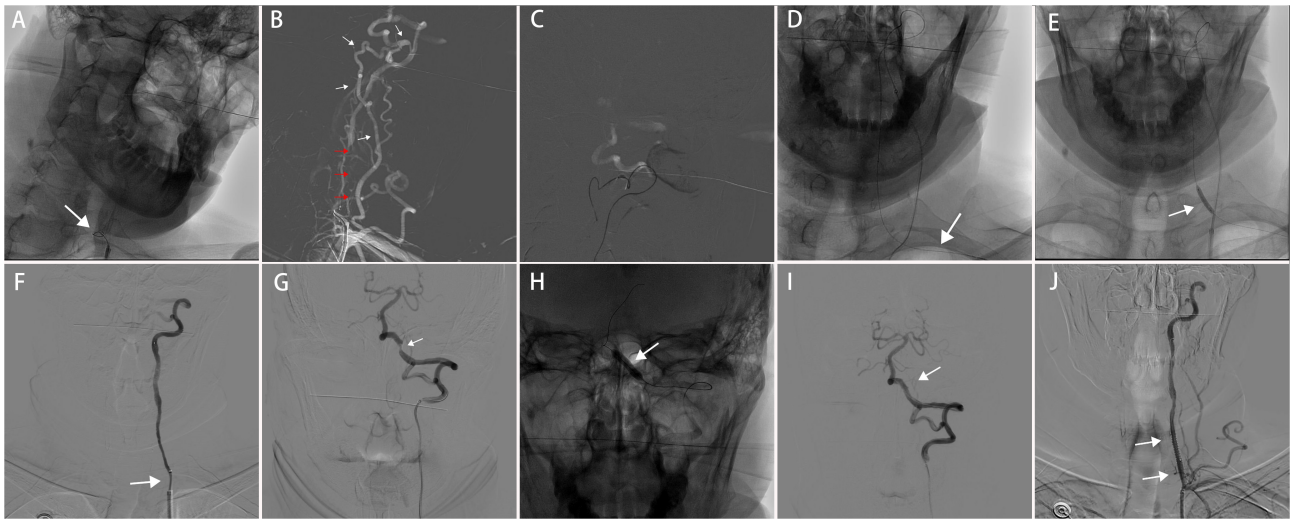

FIGURE 2
Endovascular procedure of the patient. **(A)** The micro guidewire was unable to across the occluded segment of the left vertebral artery. **(B)** Angiogram showed abundant collateral from the left deep cervical artery to the V3 segment of the left vertebral artery (white arrow), a tapered stump in the distal part of the occluded segment of the left vertebral artery (red arrow). **(C, D)** The micro guidewire reversely traversed the occluded segment of the left vertebral artery via the left deep cervical artery and was placed in the left subclavian artery [white arrow in **(D)**]. **(E)** Balloon dilation of the left vertebral artery occlusion (white arrow). **(F)** Occluded left vertebral artery was successfully reopened but remained with severe stenosis (white arrow). **(G)** Angiogram showed severe stenosis of the V4 segment of the left vertebral artery (white arrow). **(H)** A balloon-expandable stent was implemented in the stenotic segment of the left vertebral artery (white arrow). **(I)** After stenting, the stenosis of the V4 segment has been relieved (white arrow). **(J)** Stent placement of the V1 segment of the left vertebral artery to prevent restenosis (white arrow).

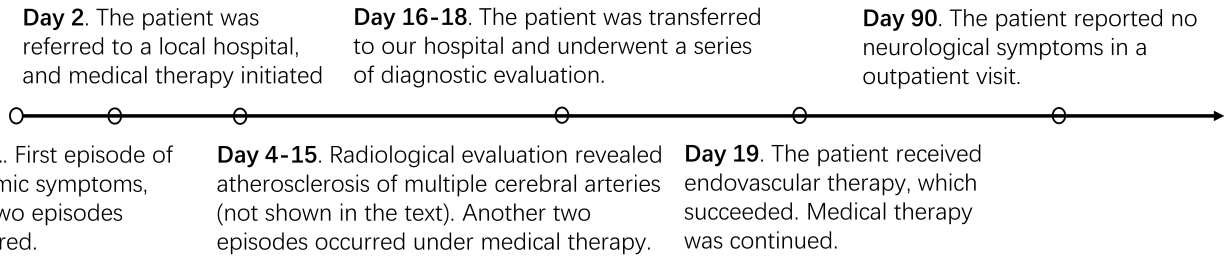

FIGURE 3
The timeline of the present case.

that of a non-tapered stump occlusion (6). A non-tapered stump might hamper the micro guidewire to find the access to cross the occluded segment. Hence, it has long been a challenge to antegrade endovascular recanalization of a non-tapered occlusion.

Fortunately, the development of robust collateral flow from the deep cervical artery arising from the subclavian artery provided a new path for the micro guidewire to reversely cross the occluded segment in this patient. During the natural course of VA occlusion, collateral circulation may develop from the cervical arteries to the V2–V3 segment of VA (16), which could serve as an alternative routine to endovascular recanalization of the non-tapered VA occlusion when antegrade navigation failed. In a previous report, Ji et al. performed endovascular intervention for a patient with vertebral artery stump syndrome using retrograde recanalization *via* the ascending cervical artery when attempts of antegrade recanalization failed (17). Antegrade endovascular recanalization of a non-tapered occlusion is challenging due to morphologic and hemodynamic factors described above, where blood flow from the brachiocephalic trunk to the distal right subclavian artery may divert from the original direction of the micro guidewire. While the distal part of the occluded segment may have a tapered stump, the blood flow is usually static. The retrograde endovascular method takes advantage of morphologic and hemodynamic factors to recanalize a non-tapered occlusion. Thus, our report, together with the report Ji et al., suggested that the cervical collateral to the VA might be a new solution for endovascular recanalization of non-tapered VA origin occlusion. This, however, depends on the extent of the collateral developed.

Instead of recanalizing the occluded segment, antegrade recanalization of the stenosis of the V4 segment via the deep cervical artery was also theoretically possible, which, however, was not considered due to the acute tortuosity at the junction between the two vessels and the difficulty in delivering balloon and stent to the diseased site via the deep cervical artery. In addition, if the

endovascular treatment had been a failure in this patient, open surgery would be considered.

Conclusion

Antegrade endovascular recanalization of VA occlusion with a non-tapered stump has long been a challenge due to morphologic and hemodynamic factors. The retrograde endovascular method *via* the cervical collateral may be an alternative for this type of VA occlusion when antegrade recanalization fails, which requires further exploration.

Patient perspective

The perspective of the patient was obtained in an outpatient visit during a follow-up of 3 months. The patient appreciated the doctors for solving his symptoms by endovascular recanalization of the occluded VA. He was amazed at the unique technique that the neurointerventionist performed. In addition, the patient agreed to publish his de-identified medical records.

Data availability statement

The raw data supporting the conclusions of this article will be made available by the authors, without undue reservation.

Ethics statement

The studies involving human participants were reviewed and approved by the Medical Ethics Committee of Zhongnan Hospital of Wuhan University (2023070K). The patients/participants provided their written informed consent to participate in this study. Written informed consent was obtained from the patient for the publication of any potentially identifiable images or data included in this article.

Author contributions

HQ: conceptualization and design, literature review, and manuscript preparation. ZK: data collection, literature review, and manuscript preparation. DS: patient management and manuscript review. BM: patient management, manuscript review, and funding acquisition. JZ: manuscript review and supervision. All authors contributed to the article and approved the submitted version.

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Conflict of interest

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Case report: Navigating the challenges: successful mechanical thrombectomy in a case of persistent primitive hypoglossal artery

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Persistent primitive hypoglossal artery (PPHA) is a highly uncommon abnormal connection between the internal carotid artery (ICA) and vertebral artery (VA), with reported incidences ranging from 0.027 to 0.26%. Attempting endovascular intervention in such cases presents a considerable challenge as it carries a higher risk of embolization and other procedure-related complications that may affect a wide area of the brain. We present a case study involving the utilization of mechanical thrombectomy (MT) to treat an ischemic stroke in the M1 segment of the middle cerebral artery (MCA) despite the presence of PPHA. Performing mechanical thrombectomy in an anomalous vascular connection is feasible; however, it necessitates heightened vigilance, thorough knowledge of the anatomy, and utmost caution.

KEYWORDS

persistent primitive hypoglossal artery, mechanical thrombectomy, stroke, MCA, PPHA

Background

During fetal circulation, an intricate network of anastomoses is established between the carotid and basilar arteries, involving the hypoglossal, trigeminal, optic, and proatlantal arteries. In the normal course of embryogenesis, carotid–vertebro-basilar anastomoses typically regress and cease to exist (1). However, in rare instances, their persistence can occur, representing a carotid–basilar anastomosis that endures beyond the usual developmental period. Persistent primitive hypoglossal artery (PPHA) is an example of these developmental connections between the carotid and vertebro-basilar arteries. The reported prevalence of this anomalous artery in angiographic studies ranges from 0.03 to 0.09% (2). This unique artery is considered a vascular anomaly, which originates from the internal carotid artery (ICA) between the C1 and C2 vertebral levels and then passes through the hypoglossal canal and joins the vertebro-basilar system, creating an anastomosis (3). PPHA can be a potential route for emboli originating from the heart or proximal internal carotid artery (ICA) to reach the posterior circulation, resulting in ischemic stroke (4). Moreover, in patients with PPHA, there is a rare occurrence of simultaneous embolic infarcts in both the anterior and posterior circulation. Additionally, a

PPHA and carotid artery dissection can be associated with recurrent cerebral infarction in both the anterior and posterior circulation (5, 6).

Case presentation

We are reporting a case of a 43-year-old lady healthcare worker with a modified Rankin score (mRS) of zero at baseline, presented to the National Institute of Cardiovascular Diseases (NICVD), Karachi, Pakistan, with right-sided hemiparesis, aphasia, and slightly altered level of consciousness of 3-h duration. Her physical examination revealed right-sided facial drooping and left-sided gaze preference with an overall NIHSS of 18. Her past medical history was positive for ischemic heart disease. Her baseline hemodynamics were within normal limits with a blood pressure (BP) of 131/63 mm Hg.

Investigations

With the provisional diagnosis of stroke, the hyper-acute stroke team was activated and the patient was shifted for computed tomography angiography (CTA) within 15 min of her presentation. Her non-enhanced CT scan is shown in Figures 1A1, 2.

Her subsequent CTA (Figures 1B1–3) revealed an occluded left-sided middle cerebral artery (MCA) at the M1 segment. However, an interesting and uncommon occurrence was the bifurcating left internal carotid artery (ICA) at the level of C2, after the common carotid artery (CCA) bifurcation on axial images (Figures 1B1–3) and 3D images as shown in Figure 2, respectively. The actual ICA was present medially, while laterally, it was PPHA, arising from the cervical part of the ICA and continuing as the left vertebral artery connecting the anterior and posterior circulation. PPHA is a fetal connection between anterior and posterior circulation, which failed to regress as expected in the normal developmental process. However, she fulfilled the mechanical thrombectomy (MT) criteria as per American Stroke Association Guidelines 2018 (7) and subsequently shifted to the catheterization laboratory for direct MT without bridging with thrombolytic therapy.

Her baseline digital subtraction angiography (DSA) further confirmed the anomalous connection of left ICA with the left vertebral artery (VA) as PPHA, which resulted in the filling of the entire posterior circulation through left ICA injection, along with an occluded M1 segment of left as shown in Figure 3.

Treatment

We performed her thrombectomy very cautiously to minimize the risk of embolization in the posterior circulation and avoid unnecessary injection. We performed a direct first pass aspiration technique (ADAPT) only once, which resulted in the successful recanalization of MCA; however, there was some embolization to an A3 segment, as shown in Figure 3.

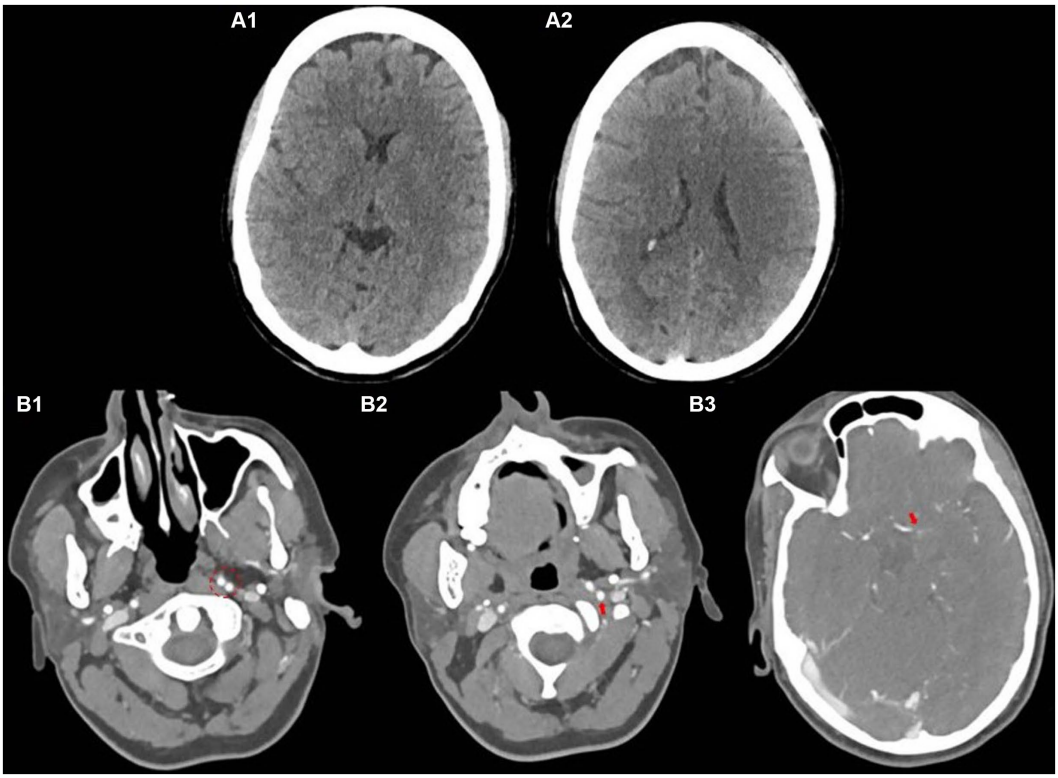

FIGURE 1
Baseline non-contrast CT scan **(A)**, CT angiography showing bifurcating ICA **(B1)**, persistent primitive hypoglossal artery **(B2)**, and occluded left MCA **(B3)**.

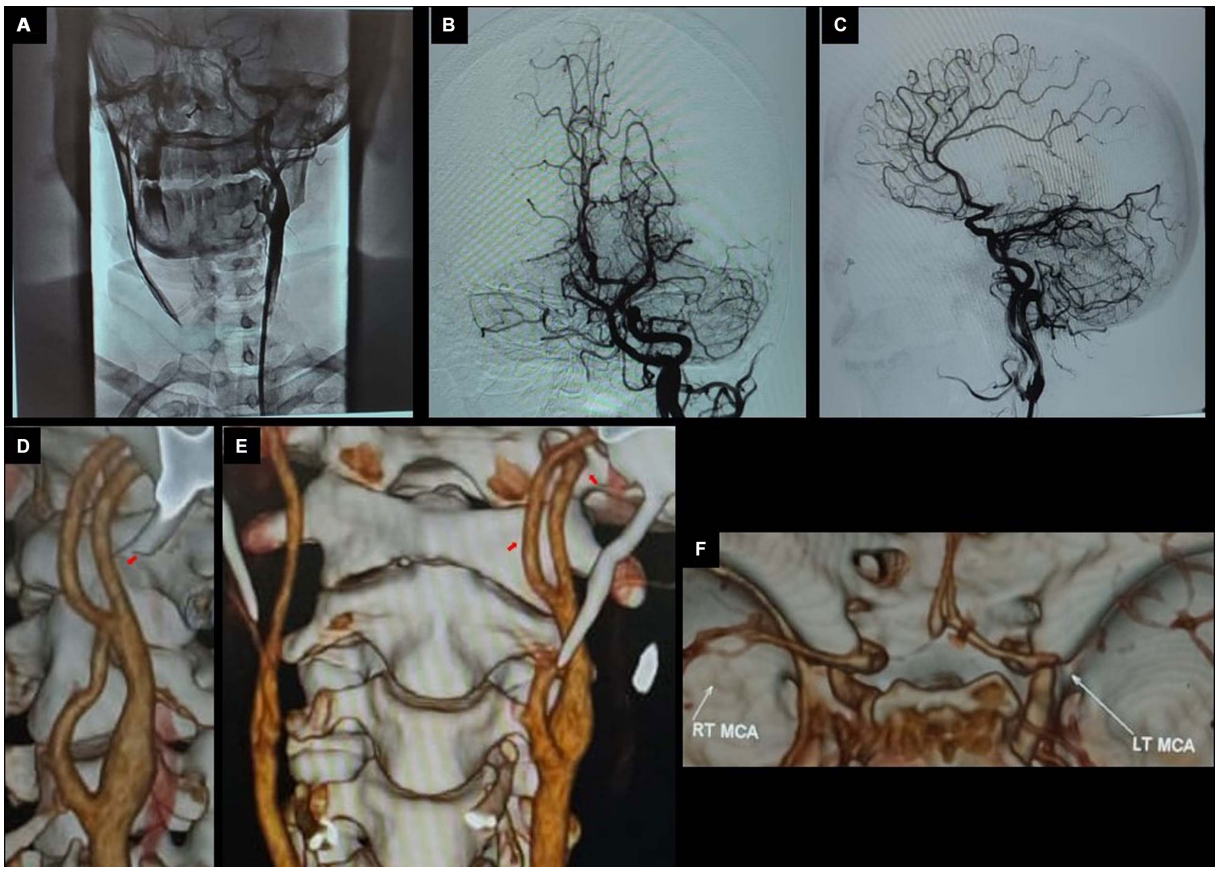

FIGURE 2
Pre-thrombectomy; LAO view at CCA bifurcation **(A)**, town's view **(B)**, lateral view **(C)**, and 3-D images showing bifurcating ICA **(D)**, persistent primitive hypoglossal artery **(E)**, and occluded left MCA **(F)**.

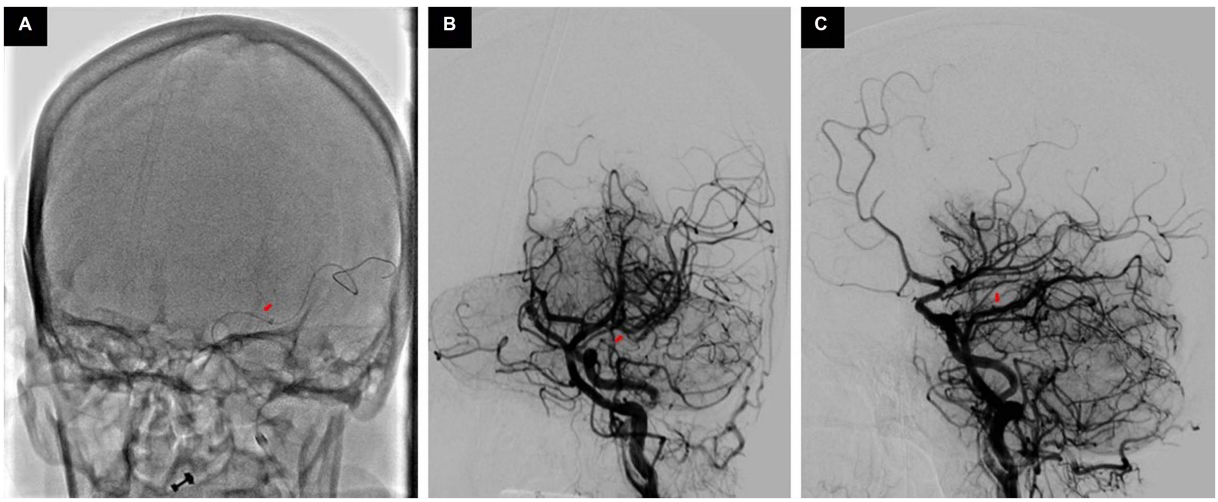

FIGURE 3
ADAPT is being performed **(A)**, Post-thrombectomy town's view **(B)** and lateral view **(C)** showing recanalized MCA.

Outcome and follow-up

Post-MT, her BP remained stable and below 140 mmHg, which is well within target as mentioned in our institutional protocol post-MT with TICI IIb flow. She was then shifted to our stroke unit, where she was monitored and managed in accordance with standard clinical practice guidelines. Her post-MT CT scan after 24 h showed no hemorrhage; however, she developed subtle hypodensity with an

ASPECT of 9 (Figure 4). She was kept in the hospital and treated with IV mannitol along with a single antiplatelet agent and other symptomatic treatment. She was discharged home after 4 days with an mRS of 4. She was subsequently followed in the outpatient department, and after 3 months of follow-up, her mRS improved to 3.

Consent

We have obtained written consent from the patient's attendant for reporting this case report.

Discussion

The PPHA belongs to a category of carotid–basilar anastomoses, which also encompasses the persistent trigeminal, otic, and proatlantal intersegmental arteries. These arteries are designated based on their respective cranial nerve associations. PPHA is a unique and infrequent vascular condition representing an embryonic connection between the ICA and the vertebro-basilar artery system. Specifically, the hypoglossal artery originates from the ICA, typically between the C1 and C2 vertebral levels, and courses through the hypoglossal canal to establish a connection with the vertebro-basilar arterial network. It manifests as a consequence of the incomplete regression of pre-segmental arteries responsible for nourishing the posterior circulation during the early stages of fetal brain development (8). Notably, the left PPHA is more prevalent, accounting for approximately 65% of cases (9).

PPHA ranks as the second most prevalent persistent carotid–vertebro-basilar anastomosis, with an estimated incidence ranging from 0.027 to 0.26%, as mentioned above (10). It exerts substantial hemodynamic alterations on both the carotid and vertebro-basilar vascular systems, potentially correlating with the presence of intracranial vascular anomalies. It typically represents an incidental discovery during angiography, often remaining entirely asymptomatic.

In its presence, the vertebral arteries usually exhibit hypoplasia, the ipsilateral ICA may be entirely absent, and under the circumstances involving occlusion of the contralateral ICA, the PPHA can assume the exclusive role of supplying the circle of Willis. The literature has documented a limited number of cases in which the PPHA serves as the sole source of cerebral blood supply (8). The co-occurrence of intracranial aneurysms alongside PPHA was observed at an incidence rate of approximately 26% (11). An especially noteworthy consequence of PPHA is the occurrence of simultaneous anterior and posterior circulation infarctions in conjunction with carotid atherosclerosis. While this occurrence is infrequent, a limited number of cases have been documented in the existing literature (6).

Only 10–20% of all ischemic strokes qualify for endovascular treatment based on involvement of large vessels and time of presentation since last known to be well (12, 13). There exists a limited timeframe within which this intervention can be successfully performed. As this time elapses, the effectiveness of reperfusion diminishes. Current American Stroke Association Guidelines (ASA) recommend the provision of endovascular treatment up to the window of 6 h, which can be extended up to 24 h since last known to be well with the help of perfusion studies (14).

However, there were no significant differences between the mechanical thrombectomy and control groups regarding rates of symptomatic intracranial hemorrhage or 90-day mortality.

Performing mechanical thrombectomy has always been a complex task due to the varying tortuosity (twisting and turning) of intracranial blood vessels. It requires the use of advanced tools such as guiding catheters, aspiration catheters, micro-catheters, or stent retrievers, often in a multi-step telescoping manner. Moreover, when dealing with anomalous anatomy, the procedure becomes more challenging than in a typical normal setting. However, certain steps, including more selective engagement, use of a balloon-guide catheter, use of concomitant thrombolytic therapy, and if patients presented within 4.5 h, may reduce the risk of embolization in the non-involved territory.

The existence of this abnormal connection poses a significant risk to the overall circulation during the procedure, increasing the likelihood of distal embolization or other mechanical complications.

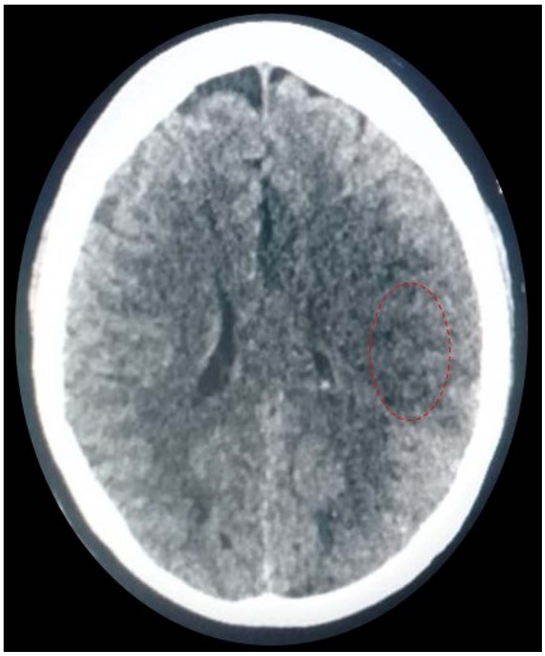

FIGURE 4
Post-thrombectomy; non-contrast CT scan showing subtle hypodensity in left MCA territory with an ASPECT of 9.

Learning points/take home messages

- Performing mechanical thrombectomy in the presence of an anomalous vascular connection is feasible; however, it necessitates heightened vigilance, thorough knowledge of the anatomy, and utmost caution.
- The procedure becomes particularly challenging due to the inherent risks of embolization and other potential complications that can concurrently impact both the anterior and posterior circulation.
- Careful considerations and meticulous execution are essential to minimize the potentially disastrous consequences associated with this unique anatomical condition.

Data availability statement

The original contributions presented in the study are included in the article/Supplementary material, further inquiries can be directed to the corresponding author.

Ethics statement

The studies involving human participants were reviewed and approved by the National Institute of Cardiovascular Diseases (NICVD). The patients/participants provided their written informed consent to participate in this study. Written informed consent was obtained from the individual's legal guardian/next of kin for the publication of any potentially identifiable images or data included in this article.

Author contributions

KK, IL, JS, and SF conceived the idea of the case report. KK, IL, JS, SF, SA, and BS were involved in the assessment diagnosis and management of the patients. KK, IL, JS, SF, SA, BS, AK, FZ, AA, and NQ were involved in data acquisition, manuscript writing, and critical review of the manuscript. All authors contributed to the article and approved the submitted version.

Conflict of interest

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Supplementary material

The Supplementary material for this article can be found online at: https://www.frontiersin.org/articles/10.3389/fneur.2023.1248506/full#supplementary-material

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Resection of ruptured aneurysm associated with bilateral anomalous posterior inferior cerebellar anastomotic arteries: case report and review of literature

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Introduction: Aneurysms on the posterior inferior cerebellar artery (PICA) may not be the major part of intracranial aneurysm. Especially, an aneurysm located on the bilateral posterior inferior cerebellar anastomotic artery has abnormal anatomical characteristics in the vessel wall and then causes stroke including subarachnoid hemorrhage. This case report explores the direct resection of a ruptured aneurysm associated with the bilateral anomalous anastomotic artery of PICA.

Methods: The case report discusses a 53-year-old woman who suffered from sudden severe headache and vomiting for more than 3 h admitted to our hospital. Emergency computed tomography (CT) revealed subarachnoid hemorrhage (SAH) in the third and fourth ventricles. Preoperative 3 Dimensions-digital subtraction angiography (3-D DSA) indicated a ruptured aneurysm located on the bilateral posterior inferior cerebellar anastomotic artery. Postoperative pathological findings indicated the characteristics of parent artery PICA and control aneurysm. The authors performed an overview of PICA aneurysms with anomalous variation in the Pubmed, Web of Science, and Medline databases. The search was until 1 August 2023. Related terms "posterior inferior cerebellar artery" And "aneurysm" AND "anatomical variants" were used to search the review. The reasons for anomalous variation anastomosis between bilateral PICAs were analyzed.

Results: The aneurysm was resected successfully. Post-operative 3-D DSA revealed the disappearance of the aneurysm. The vessel wall of anastomotic PICA showed neovascularized hyperplasia, abnormal arrangement of smooth muscle, CD31+ endothelial cells, and SMA+ smooth muscle cells. In contrast, when it came to aneurysm, the wall at the location of the fracture thinned, which could be used to explain that the local nodular protrusion was formed and CD31+ endothelial cells existed. No neurological deficits were found at her 1-year follow-up visit (mRS score of 0).

Conclusion: Direct resection of ruptured aneurysm associated with bilateral anomalous posterior inferior cerebellar anastomotic arteries was an effective treatment and careful consideration of the anatomical characteristics concerning the interesting aneurysm and the variant PICA was critical for sate treatment. Also, the literature on the lesion was reviewed.

KEYWORDS

subarachnoid hemorrhage, anatomical variants, aneurysm, posterior inferior cerebellar artery anastomosis, surgical treatment

Introduction

Posterior inferior cerebellar artery aneurysms are rare and account for less than 3% of incidence (1). Due to its variants in the process of fetal development (2), arterial network concerning PICA indicates various pathological characteristics and may be an essential factor associated with some posterior circulation vascular diseases. Additionally, the mentioned features affect the collateral circulation and hemodynamics of PICA, causing aneurysms which can be a culprit of stroke. As to the PICA aneurysm with variant vessel, coiling, direct clipping, and trapping, the aneurysm with PICA-PICA anastomosis had different outcomes (1, 3–5). Especially, patients with PICA aneurysms suffered postoperative posterior ischemic stroke and nerve paresis, which makes the options concerning the variant PICA aneurysm unfavorable and negative.

The authors report one case of a ruptured aneurysm located on the bilateral posterior inferior cerebellar anastomotic artery. The ruptured aneurysm was resected leaving no neurological deficits, and further pathological characteristics between the variant PICA and control aneurysm were shown. In short, direct resection of the ruptured aneurysm was performed on the patient, allowing for the anatomical characteristics and imaging findings. To the best of our knowledge, the present study is the first research concerning direct resection of the aneurysm associated with the bilateral Anomalous Posterior Inferior Cerebellar Artery Anastomotic Arteries. Related literature on the lesion was reviewed and studied.

Case description

A 53-year-old woman suffering from sudden severe headache and vomiting for more than 3 h was admitted to our hospital. Emergency CT revealed subarachnoid hemorrhage in the localization of the third and fourth ventricles (Figures 1A,B). Preoperative DSA and 3-D DSA indicated a ruptured aneurysm located on the bilateral posterior inferior cerebellar anastomotic artery (Figures 1C–E). The intraoperative image indicated an aneurysm located on the bilateral posterior inferior cerebellar anastomotic artery with some arterial branches on the back of the medulla (Figures 1F,G).

Concerning the anatomical characteristics and imaging findings, direct coiling was considered dangerous and may bring about ischemic events. Direct clipping of the aneurysm in the fourth ventricle may cause ischemic events and recurrence following clipping for anastomosis between bilateral PICAs of the anatomical characteristics. The woman was positioned in the right lateral park-bench position. Suboccipital craniotomy in the midline was performed. The aneurysm was exposed by a suboccipital approach in the midline. Bilateral posterior inferior cerebellar arteries were trapped prior to resection of the ruptured PICA aneurysm (Figures 1H,I). After trapping the anastomosis between bilateral PICAs, the aneurysm was resected successfully and intraoperative ICG showed the disappearance of the aneurysm (Figures 1J,K). Post-operative CT (Figure 1L), post-operative DSA, and 3-D DSA revealed the obliteration of the aneurysm (Figures 1M–P). Postoperative pathological findings indicated the characteristics of the variant aneurysm. The woman was discharged on the seventh postoperative day (mRS score of 0) and no neurological deficits were found at her 1-year follow-up visit.

Pathological characteristics

An in-depth study of the histopathological findings was conducted for the treatment of the disease which helped the authors understand the pathogenic mechanism.

Postoperative pathological findings indicated significant differences between the present variant posterior inferior cerebellar anastomotic artery (V-PICA) and another aneurysm (the clinical features of another patient just suffering an aneurysm that was not mentioned in the present study).

Variant posterior inferior cerebellar anastomotic artery

HE staining (Figure 2A) showed that the vessels were twisted, with uneven thickness of the vessel wall, obvious thickening of part of the vessel wall, and rupture of part of the vessel wall with bleeding. Mechanized neovascularization and hyperplasia and lumen sizes were observed at remote bleeding sites (original magnification ×40).

HE staining (Figure 2B) revealed that the smooth muscle of the malformed vessel wall was disordered, the lumen of the ruptured vessel was connected with the remote mechanized hemorrhage, and vascular endothelial cell proliferation was observed (original magnification ×100).

HE staining (Figure 2C) indicated that the partial vascular wall was disordered with remote hemorrhage accompanied by thrombosis, fibrin aggregation, and mechanical angiogenesis with lumen varying in size, and vascular endothelial cell proliferation could be seen in some thrombotic areas (original magnification ×100).

CD31 immunohistochemical staining (Figure 2D, original magnification ×100) revealed a positive expression of vascular endothelial cells, vascular lumen disorder, and some new vessels in the vascular wall along the original vessel wall, and the lumen size was different, which shows mechanical thrombosis.

Aneurysm

HE staining (Figure 3A, original magnification ×40) showed that the arterial vessel wall was broken, the vascular endothelium extended outside the vessel wall along the fracture site, and the bleeding at the

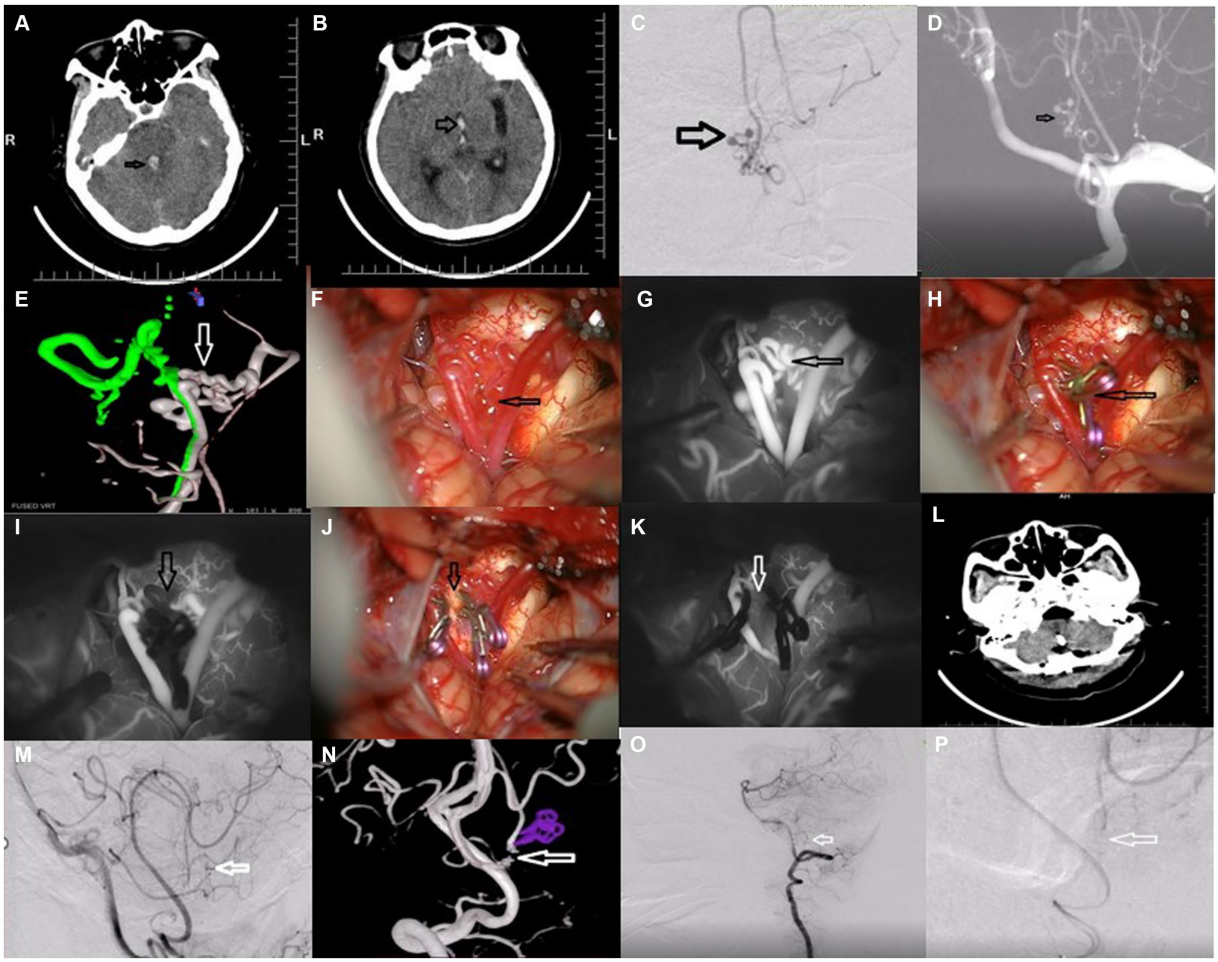

FIGURE 1
Resection of ruptured aneurysm associated with bilateral anomalous posterior inferior cerebellar anastomotic arteries.

fracture site was significantly thinned. It could be found that local nodular protrusion formed when the vessels were in the filling state.

CD31 immunohistochemical staining indicated positive staining for vascular endothelial cells (Figure 3B, original magnification ×100).

Literature search and analysis

The authors performed an overview of PICA aneurysms with anomalous variation in the Pubmed, Web of Science, and Medline databases. The search was until 1 August 2023.

Related terms "posterior inferior cerebellar artery" And" aneurysm" AND "anatomical variants" AND "English" were used to search the databases. The authors followed the PRISMA guideline previously reported and manually extracted relevant references. Furthermore, all searched articles were carefully checked and reviewed (Table 1). In eight reviewed articles, different patients suffered SAH, headache, intra-ventricular hemorrhage, obtundation, quadriplegia, giddiness, nausea, vertigo, and right hypoacusis. The ages of patients ranged from 19 to 78 years. Endovascular treatment, direct clipping of PICA-PICA anastomosis combined with trapping of the related PICA aneurysm, coil embolization of the aneurysm, and further parent vessel occlusion and posterior fossa decompression by suboccipital craniectomy for rebleeding, reimplantation bypass, and trapping of the aneurysm were taken into consideration. Various neurological statuses at discharge and follow-up are shown in Table 1.

Follow-up

The patient returned to normal life without postoperative complications. In addition, no neurological disturbances occurred at 1-year follow-up (mRS score of 0).

Discussion

Although dominantly originating from the vertebral artery, it is well known that PICA has various variations. The variations may be responsible for some intracranial vascular diseases (3–5), including aneurysms. There were various clinical features of the V-PICA aneurysm. In the eight reviewed articles (Table 1), the symptoms included SAH, headache, intra-ventricular hemorrhage, obtundation, quadriplegia, giddiness, nausea, vertigo, and right hypoacusis (2–9).

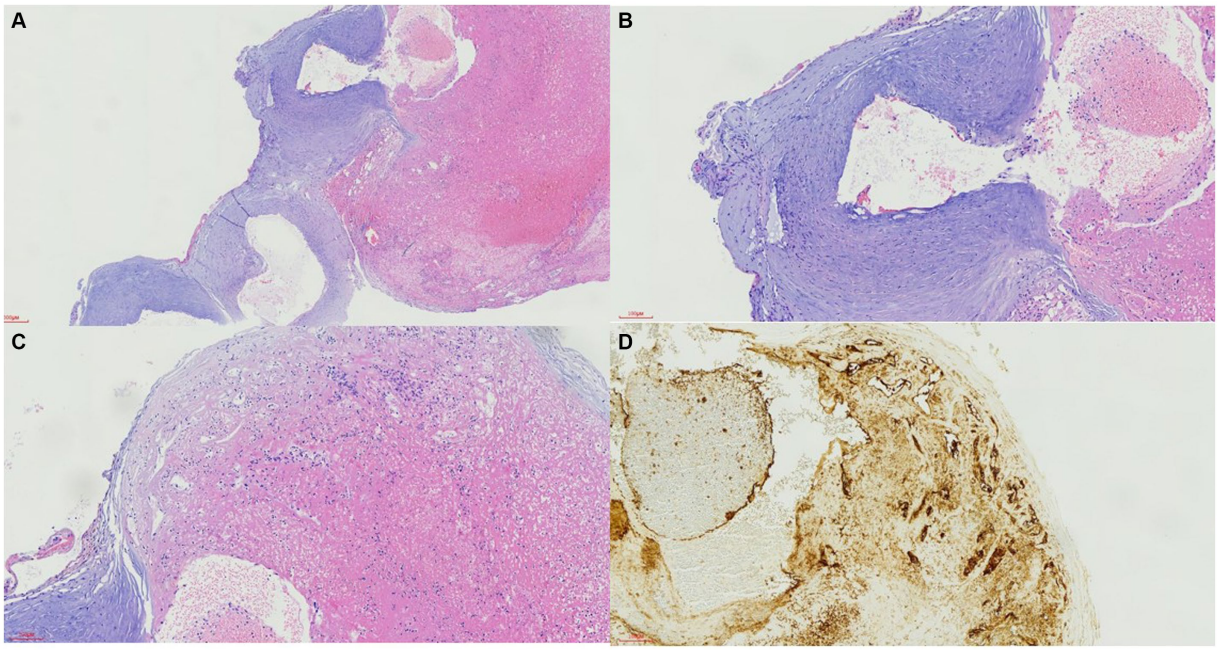

FIGURE 2
Pathological findings in the vessel wall of variant posterior inferior cerebellar anastomotic artery.

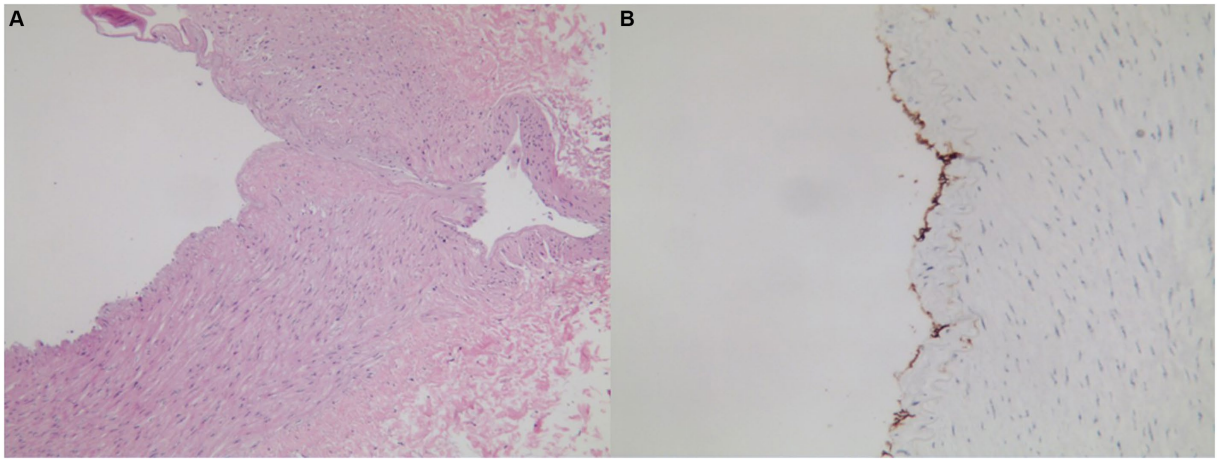

FIGURE 3
Pathological findings in the vessel wall of control aneurysm.

In our present case, the woman suffered a sudden severe headache and vomiting for more than 3 h before being admitted to our hospital, and preoperative CT indicated SAH.

Some embryonic arterial characteristics concerning V-PICA can be utilized to explain the aforementioned intracranial vascular diseases, especially PICA aneurysms. Usually, in the first 28 days, fetal development occurs in neural arteries and then in lateral vertebrobasilar anastomosis at 29 days (2). At that time, bilateral PICA had anastomosis and collateralization, and then the above anastomosis regresses. Additionally, the fetal anastomosis developed into the independent PICAs in most individuals (1). However, the other few individuals continued to have the features of fetal anastomosis and were unable to regress. Additionally, anastomosis and collateralization became anomalous and fragile (6), which was related to hemodynamic disturbance, then the hemodynamic stress owing to the mentioned anastomosis and collateralization led to vascular disease.

In the present case, the authors indicated that due to anomalous variation anastomosis between bilateral PICAs, the bilateral PICA suffered hemodynamic stress. The left PICA was dominant and the right PICA was non-dominant (Figure 1G). Due to the fragile anastomosis and collateralization between the two PICAs, the aneurysm occurred and was prone to rupture. Interestingly, the

TABLE 1 Overview of PICA aneurysm with anomalous variation.

References	Sex, age	Clinical sign	Diseased region	Treatment	Neurological status at discharge	Follow-up
Joseph et al. (6)	19, male	SAH	AICA and bulbar artery supply to PICA	Coil embolization of the An	Left Horner, s syndrome	4 months
Fujimura et al. (2)	50, male	Headache, obtundation, and quadriplegia	PICA-AICA anastomotic Artery	Coil embolization of An and occlusion of the PA	moderate disturbance of consciousness and quadriparesis	-
Baskaya et al. (7)	44, female	SAH and IVH	AICA-PICA anastomotic Artery	Clipping	Hydrocephalus and lower cranial nerve paresis	3 years
Chin et al. (3)	54, male	Cerebellar hematoma and IVH	Bihemispheric PICA	Coil embolization of An, and further PVO and posterior fossa decompression by suboccipital craniectomy for rebleeding	mRS 1	6 months
Dammers et al. (8)	52, female	SAH	Extracranial PICA	Clipping	No major neurological deficits	7 days
Germans et al. (9)	49，male	SAH	LSA-PICA	Clipping	Uneventful	6 months
Lang et al. (4)	78, male	SAH	Double-origin PICA	Reimplantation bypass and trapping of the aneurysm	Glasgow coma scale score of 15 and mRS 3	5 months
Jerry et al. (5)	39, male	Vertigo and right hypoacusis	AICA-PICA common trunk variant	Endovascular treatment	Intact neurological status	7 months
Present study	53, female	SAH and IVH	Bilateral PICA anastomosis	Resection of the An	mRS 0	1 year

PICA, Posterior inferior cerebellar artery; An, Aneurysm; PA, Parent artery; IVH, Intraventricular hemorrhage; mRS, modified Rankin score; PVO, Parent vessel occlusion.

ruptured PICA aneurysm was located at the anastomosis and collateralization, which was similar to the formation of the anterior communicating artery aneurysm with one side having a dominant A1 segment and the other side having a non-dominant A1 segment. At the same time, the pathological findings revealed the differences between the present V-PICA and the control aneurysm (Figures 2, 3). In all, the anomalous anastomosis and collateralization were the culprits of the formation and rupture of the present aneurysm at the bilateral PICA anastomosis.

The pathological or etiology characteristics of V-PICA are dramatically different from those of other intracranial vascular lesions. In the present study, we found some interesting characteristics associated with the vessel wall in V-PICA and aneurysm. The vessel wall of V-PICA showed neovascularized hyperplasia, abnormal arrangement of smooth muscle, CD31+ endothelial cells, and SMA+ smooth muscle cells. In contrast, when it came to aneurysm, the wall at the location of the fracture thinned, which could be used to explain that the local nodular protrusion was formed and CD31+ endothelial cells existed.

Why are the treatments for V-PICA aneurysm dramatically different? In fact, because of a lack of a full understanding of the clinical features, the optimal strategy associated with V-PICA aneurysms is in debate. However, when it comes to the effective intervention for V-PICA aneurysms, various aspects are feasible and available. Previous studies show that endovascular treatment should be recommended as an ideal option following careful consideration of vascular features preoperatively, including sacrificing the parent artery PICA and its collateralization (2, 3, 5, 6). Directly clipping the ruptured V-PICA aneurysm can be performed in some corresponding patients, who may have higher postoperative complications (7–9). Even reimplantation bypass and trapping of the aneurysm may be done to prevent postoperative posterior circulation stroke (4). Compared with a perforating artery, anatomical variations and surgical access may give rise to sooner difficulties in treating PICA aneurysms, except that a perforating artery has a rich anastomotic network (10). PICA anastomotic arteries or the double origin of the two sides of PICAs had anomalous characteristics concerning anatomy. In the present study, we performed complete resection of the ruptured PICA aneurysm without postoperative neurological dysfunctions. To the best of our knowledge, the present study is the first study concerning direct resection of the V-PICA aneurysm.

However, more attention should be paid to some items. Firstly, preoperative analysis of the anatomical variations in PICA should be performed carefully. Secondly, the experience is limited for the present case and further studies concerning the lesion should be done for proper clinical options. Thirdly, other interventions can also be taken into consideration for mitigating the risk.

Conclusion

Direct resection of ruptured aneurysm associated with bilateral anomalous posterior inferior cerebellar anastomotic arteries was an effective treatment, and careful consideration of the anatomical characteristics concerning the interesting aneurysm and the variant PICA was critical for sate treatment.

Data availability statement

The datasets presented in this article are not readily available because of ethical and privacy restrictions. Requests to access the datasets should be directed to the corresponding author.

Ethics statement

The studies involving humans were approved by the Ethics Committee of the First Affiliated Hospital of Soochow University.

The studies were conducted in accordance with the local legislation and institutional requirements. The participants provided their written informed consent to participate in this study. Written informed consent was obtained from the individual(s) for the publication of any potentially identifiable images or data included in this article.

Author contributions

QH: Resources, Writing – original draft, Writing – review & editing. ZW: Writing – original draft. TL: Data curation, Resource, Writing – original draft. YH: Resources, Supervision, Writing – original draft, Writing – review & editing.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Case report: Right vertebral and carotid artery anomalies with an aberrant right subclavian artery in two patients

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Abnormal origins of the vertebral artery with supra-aortic vessel variants are exceedingly uncommon. Herein, we present two cases of the vertebral artery originating from the right common carotid artery associated with the right subclavian artery arising separately as the initial branch of the aortic arch, followed by the right common carotid artery. We reviewed the embryology of the anomalous origins of the vertebral and subclavian arteries. These variants can significantly affect surgical planning and cause severe clinical symptoms.

KEYWORDS

right subclavian artery, anomalous origin, right vertebral artery, aberrance, aortic arch

Introduction

Abnormal origins of the right vertebral artery (RVA) are uncommon, occurring at a rate of 0.18% in the general population (1). Right subclavian artery (RSCA) variants are primarily observed in vascular ring anomalies of the aortic arch, with incidence rates ranging from 0.5% to 2% (2). Therefore, RVA anomalies with an aberrant RSCA are extremely rare. To the best of our knowledge, an abnormal origin of the RVA associated with the RSCA arising as the first branch of the aortic arch has not been described in the literature (1, 3). We present two cases of the RVA originating from the right common carotid artery (RCCA), associated with the RSCA arising separately as the initial branch of the aortic arch, followed by the RCCA.

Case reports

Case 1

A 65 years-old woman presented with a history of acute severe headaches the previous year after an emotional trauma. She has no significant family or past medical history. Brain CT revealed a diffuse subarachnoid hemorrhage and a right posterior communicating aneurysm (PComA) was seen on CT angiogram. This patient was successfully treated with microsurgical clipping. An initial cerebral angiogram was performed for follow-up. Digital subtraction angiography (DSA) imaging demonstrated that the RVA arose from the RCCA and that the RSCA was the first branch arising from the aortic arch, followed by the RCCA (see Figure 1).

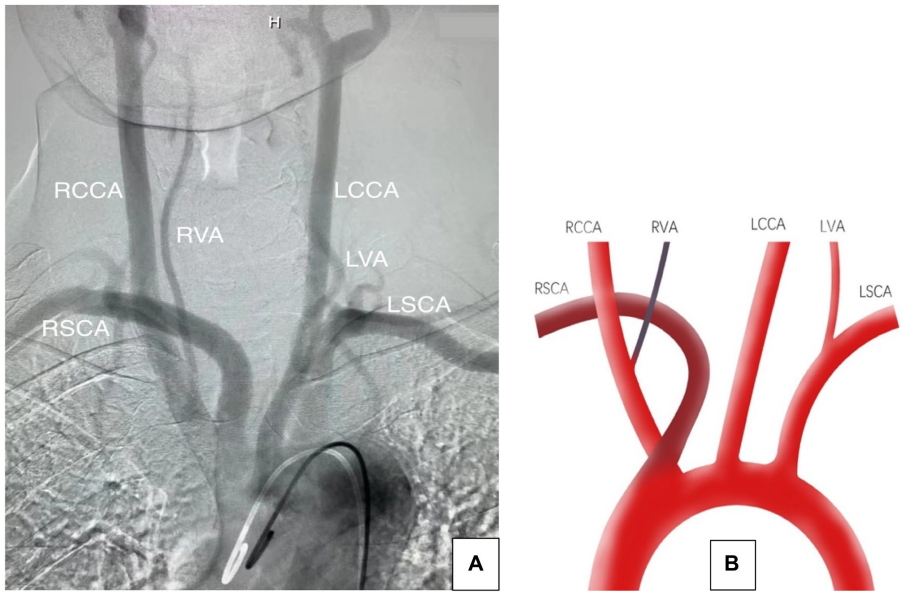

FIGURE 1
(A) Aortic arch angiogram showing the aberrant origin of the right subclavian artery and the right vertebral artery. **(B)** Illustration showing that the RVA originated from the RCCA, both the RCCA and the RSCA originated directly from the aortic arch, and the RSCA was the first branch, followed by the RCCA. LCCA, left common carotid artery; LSCA, left subclavian artery; RVA, right vertebral artery; RCCA, right common carotid artery; RSCA, right subclavian artery; LVA, left vertebral artery.

Case 2

A 60 years-old man with hypertension presented with a history of severe acute-onset headaches. Computed tomography angiography (CTA) of the head identified a left PComA aneurysm and a subarachnoid hemorrhage. In addition, arch CTA demonstrated that the RVA originated from the RCCA, part of the left subclavian artery (LSCA) was missing, the brachiocephalic trunk was absent, the RCCA and the RSCA had separate origins from the aortic arch, and the RSCA was the first branch to emerge, followed by the RCCA. DSA was performed and demonstrated that the LSCA had diverted blood from the RVA. A coiling procedure successfully treated the left PComA aneurysm and the patient's neurological condition was excellent at discharge. No intervention was performed for the LSCA, and the patient was managed with an antiplatelet regimen when necessary (see Figure 2).

Discussion

In a typical aortic arch anatomy, the vertebral arteries are conventionally identified as the primary branches of the subclavian arteries. The most common variation of the vertebral arteries is in the left vertebral artery, with an incidence rate of 2.4%–5.8% (3, 4). In contrast, variations in the origin of the RVA are less common, with an incidence rate of 0.18% (1). If the RVA arises from the RCCA, its association with RSCA abnormalities also increases (5–8). Although the embryology of abnormal RVA and RSCA is complex, understanding embryonic development and branching patterns can help explain these abnormalities.

During the early stages of embryonic development, the right ventral aorta forms the right common and brachiocephalic arteries. At the 7 mm stage, seven cervical intersegmental arteries (CIA) emerge on the aortic arch (9–11). At the 10–12 mm stage, the first to sixth CIAs anastomose longitudinally to form the VA (12). At the 14–17 mm stage, the horizontal segments of the first six CIAs degenerate and disappear, and the seventh CIA combined with the fourth aortic arch form the subclavian artery (5). Thus, the VA commonly arises from the subclavian artery, which is the largest and most proximal branch (Figure 3).

Nevertheless, incomplete involution of one of the first six CIAs leads to various anomalous origins of the VA (13). If the horizontal segments of the first or second CIA persist, an aberrant VA origin may develop in the external or internal carotid artery. If the third, fourth, or fifth CIA persist, the VA may originate from the RCCA. If the sixth CIA persists, an abnormal VA origin may arise from the aortic arch (Figure 4).

In addition, at the 40 mm stage of embryonic development, the remaining vessel of the right third aortic arch combines with the right half of the fourth ventral aorta to form the brachiocephalic trunk (BT). The possible cause of missing the BT is that the seventh CIA originates directly from the ascending aorta or that the right aortic arch partially degenerates, resulting in separate emergence of the RCCA and RSCA. The RSCA arises first, followed by the RCCA (Figure 4).

Although RVA and RSCA variations are only anatomical, and most are asymptomatic, it is essential to recognize these rare anomalies before surgery. When the RVA originates anomalously from the RCCA and its branches, injury to, or intraoperative blockage of, the corresponding carotid artery during surgery may

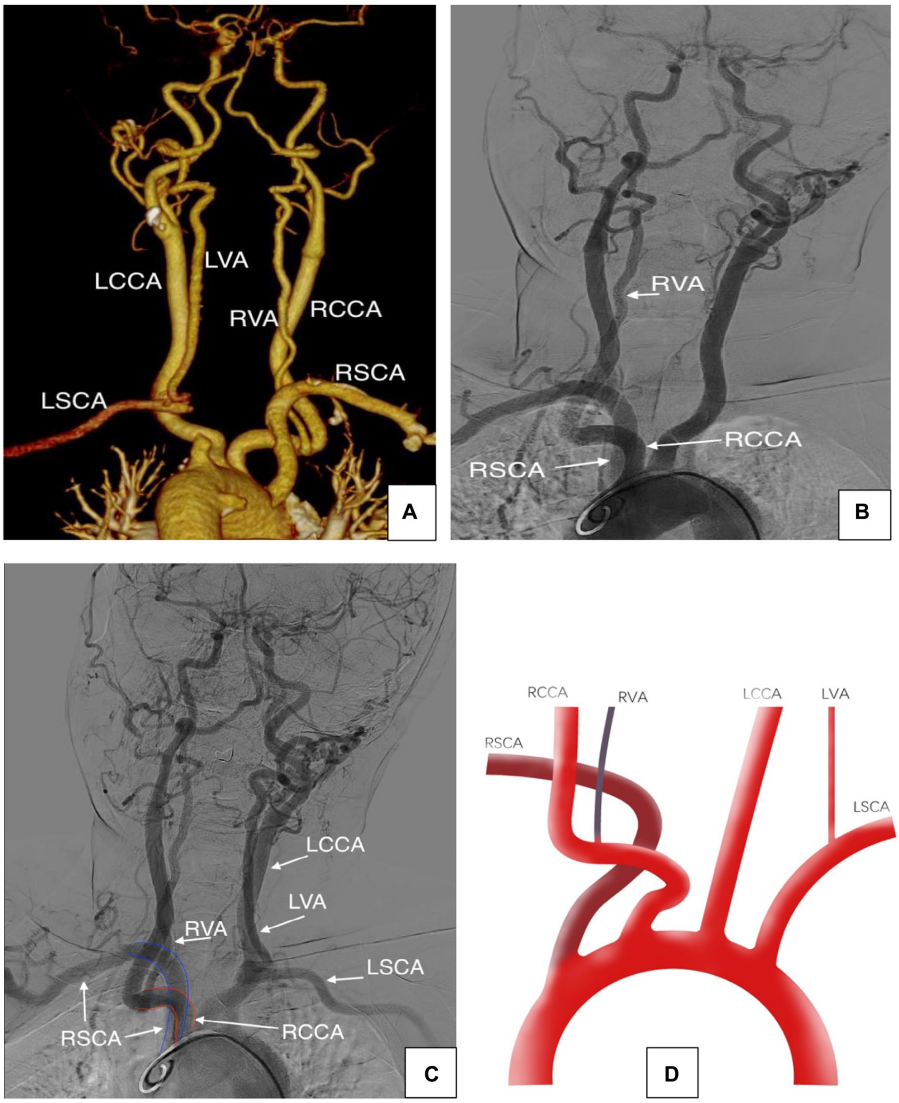

FIGURE 2
(A–C) Arch CTA and DSA showing the aberrant origin of the right subclavian artery and the right vertebral artery. **(D)** Schematic representation of the case showing that the RVA originated from RCCA and the RSCA was the first branch to arise from the aortic arch, followed by the RCCA.

cause fatal brain ischemia. When the RSCA originates anomalously from the aortic arch, surgical procedures and endovascular interventions involving the arch may pose significant challenges. Additionally, the angles of the opening of the variant RVA and RSCA are always tricky, and super selection for endovascular treatment is often difficult. Therefore, detailed information on these anatomical variations is of broad concern to clinicians.

Conclusion

The RVA from an RCCA associated with an RSCA arising as the first branch, followed by the RCCA, is extremely rare. The embryology of abnormal RVA and RSCA is complex. Physicians should identify these abnormalities before surgery.

Data availability statement

The original contributions presented in the study are included in the article/supplementary material, further inquiries can be directed to the corresponding author.

Ethics statement

Ethical review and approval was not required for the study on human participants in accordance with the local legislation and institutional requirements. Written informed consent from the patients/participants or patients/participants' legal guardian/next of kin was not required to participate in this study in accordance with the national legislation and the institutional requirements. Written informed consent was obtained from the individual(s) for the

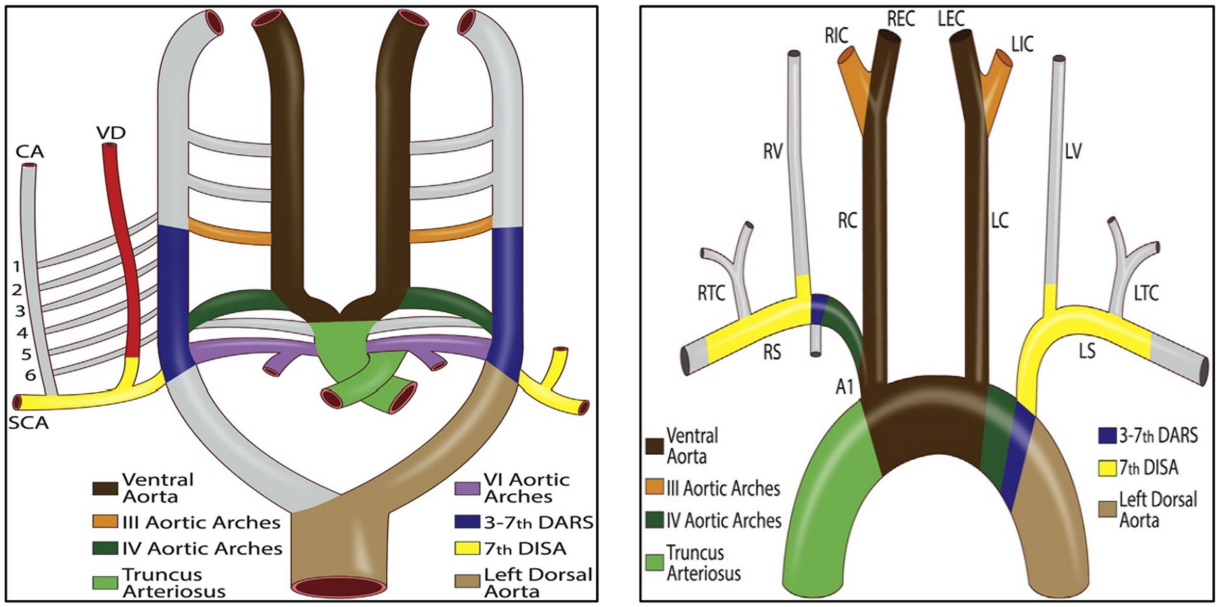

FIGURE 3
Schematic diagram of the primitive ventral and dorsal aorta (1). DARS, dorsal aortic root segments; VD, right vertebral artery; CA, ascending cervical artery; DISA, dorsal intersegmental arteries.

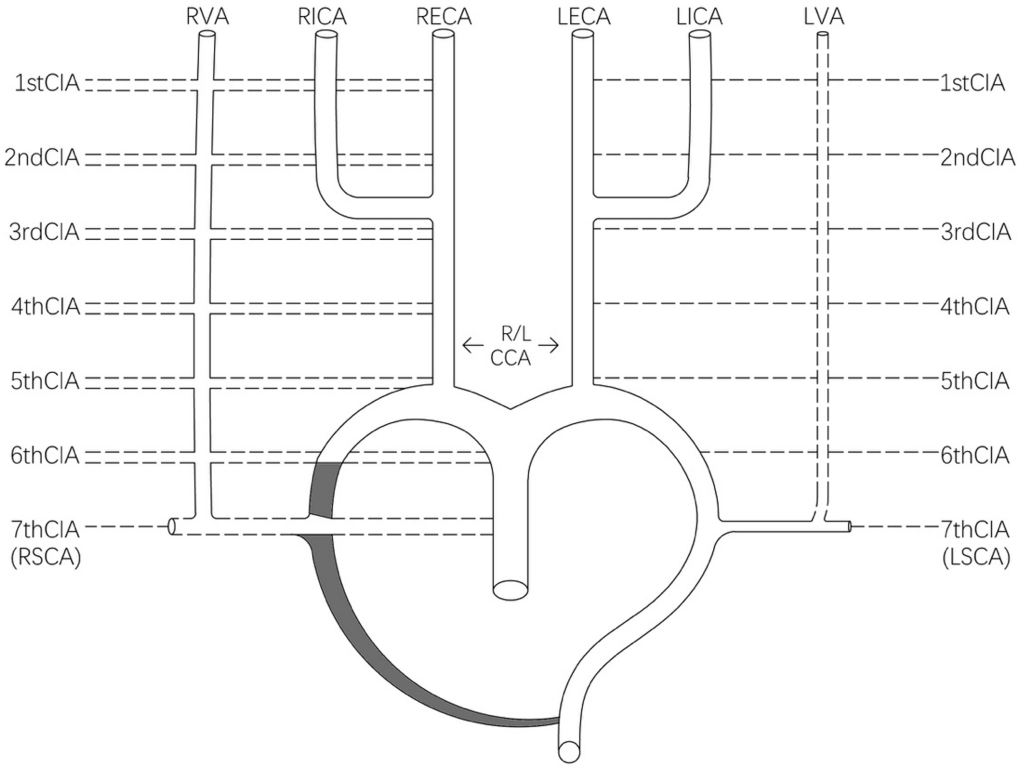

FIGURE 4
Schematic of the various anomalous origins of the VA. CIA, cervical intersegmental artery; R/LCCA, right/left common carotid artery; RICA, right internal carotid artery; LICA, left internal carotid artery; RECA, right external carotid artery; LECA, left external carotid artery.

publication of any potentially identifiable images or data included in this article.

Author contributions

JC: Writing – original draft, Writing – review & editing. LL: Formal analysis, Methodology, Writing – review & editing. XK: Data curation, Writing – review & editing. CW: Data curation, Formal analysis, Methodology, Supervision, Writing – review & editing.

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Complex torcular dural arteriovenous fistula leading to cortical venous reflux-induced severe varix and subsequent bilateral cerebral hemispheric hemorrhage: a case report

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Background and importance: Dural arteriovenous fistulas (dAVFs) with cortical venous reflux (CVR) are associated with a higher incidence of intracranial hemorrhage (ICH). We report a rare case of a complex torcular dAVF with severe cortical veins (CV) varix leading to extensive bilateral cerebral hemorrhages. This discovery suggests a potential new subtype of dAVF. The case underscores the necessity of a comprehensive understanding of hemodynamic changes in dAVFs and the importance of considering venous compensatory capacity in treatment. This case challenges existing classifications and treatment strategies for dAVFs, highlighting the need for further research and discussion within the neurosurgical community.

Clinical presentation: A 56-year-old male was admitted to the hospital presenting with dizziness, fatigue, and numbness. Brain CT scans revealed extensive bilateral cerebral hemorrhages. Digital subtraction angiography (DSA) identified a complex torcular dAVF. No cerebral sinus venous thrombosis was detected, but a venous variation in the left transverse sinus was observed. Preoperative DSA demonstrated the patient's well-developed venous compensatory ability. Subsequently, the patient underwent transarterial embolization. The patient made a good recovery. Follow-up DSA and MR angiography at 3 months and 1 year post-treatment showed no recurrence.

Conclusion: DAVFs are rare lesions, prone to ICH, particularly when CVR is involved. We report a rare case of CVR with severe varix leading to hemorrhagic lesions in both cerebral hemispheres. Our aim is to alert neurosurgical colleagues worldwide to this potential new subtype and to evaluate treatment options, in order to assist those who may encounter such cases in the future.

KEYWORDS

dural arteriovenous fistula, intracerebral hemorrhage, cortical venous reflux, superior sagittal sinus, venous compensation, venous variation

Background

Dural arteriovenous fistulas (dAVFs), abnormal shunts situated within the dura mater (1), constitute 10–15% of all intracranial vascular malformations (2). Predominantly affecting individuals aged between 50 and 60 (3), the clinical manifestations of dAVFs can vary, largely dependent on their location (4). When cortical venous reflux (CVR) is involved, the dAVF is considered high-grade, leading to an elevated risk of hemorrhage, increased surgical complexity, and a less favorable prognosis (3, 5–10).

Existing classifications primarily focus on the angiographic presentations of dAVFs, incorporating the adverse impacts of intracranial hemorrhage (ICH) and Non-Hemorrhagic Neurological Deficits (NHND) (3, 5, 6). However, these classifications may not fully capture the significance of CVR, potentially resulting in an underestimation of disease severity and influencing treatment decisions. Furthermore, the role of the intracranial venous system in dAVFs is often overlooked, despite its potential substantial influence on both etiology and prognosis (11). An effective preoperative evaluation of the venous system can significantly improve patient survival rates (12). In this report, we present a unique case of a dAVF leading to severe cortical venous varices, which resulted in widespread hemorrhagic lesions in both cerebral hemispheres due to CVR. We also discuss the valuable insights and implications derived from this case.

Clinical presentation

A 56-year-old male patient with a history of deep vein thrombosis (DVT) and chronic headaches, but no history of head trauma, surgery, or other conditions such as intracranial infection or hypertension, was admitted to the hospital due to dizziness, fatigue, and numbness. Neurological examination revealed clear consciousness and slightly weakened limb strength. Blood tests, including complete blood count and coagulation index, were completely normal (Table 1, Supplementary material). Ophthalmological examination revealed papilledema (Figure 1, Supplementary material). Brain CT scans showed multiple scattered hemorrhagic foci in both cerebral hemispheres. Digital subtraction angiography (DSA) of the cerebral vessels suggested an arteriovenous shunt around the torcular. The shunt was supplied by bilateral occipital arteries (OA), the left middle meningeal artery (MMA) occipital branch, and the vertebral artery (VA) meningeal branch, and drained into the superior sagittal sinus (SSS) and cortical veins (CV). No cerebral sinus venous thrombosis (CVST), but a venous variation in the left transverse sinus (TS) was found, impeding its normal venous drainage. The patient became somnolent the next day, with increased hemorrhage in the left frontal and parietal lobe. Upon further examination, it was observed that the cerebral hemorrhage had significantly intensified compared to the time of admission (Figure 1). An emergency plan was made to embolize the shunt arterially. Informed consent was obtained before treatment.

Under local anesthesia, a 6-Fr Envoy guiding catheter (Cordis) was placed in the internal carotid artery (ICA), and a Microcatheter (Hyperform 4 × 7 mm, America MTI/EV3) was navigated into the left MMA occipital branch and left OA branches. ONYX (Medtronic, Irvine, CA, United States.) glue was used for embolization of the dural arteriovenous fistula. After several attempts, the final angiography showed no branches supplying the fistula from the left internal and external carotid arteries, but a small amount of supply from the right OA remained, suggesting a slight residual shunt (Figure 2).

Postoperative CT scanning revealed a high-density area indicating a cast of ONYX glue in the SSS. In the following week, intracranial hematoma continued to progress but eventually absorbed. Follow-up DSA 3 months after surgery demonstrated no recurrence of the dAVF and the varix in the bilateral cerebral hemispheres was markedly reduced. A one-year postoperative MR review indicated no recurrence of dAVF, although thrombosis was observed in the SSS (Figure 3), the patient's condition and vision improved, with no significant neurological abnormalities.

Discussion

Hemorrhage due to dAVF accumulation in the SSS is not uncommon, as documented in previous studies (13, 14). The distinguishing feature of this case, however, is the severe varix of the draining CV, leading to widespread hemorrhagic lesions in the bilateral cerebral hemispheres, a phenomenon rarely reported. This indirectly suggests an extraordinarily high pressure within the SSS, leading us to postulate that the blood within the SSS essentially has arterial characteristics, with a pressure and impact force equivalent to arterial blood. We term this phenomenon 'venous arterialization', which further exacerbates the risk of hemorrhagic events (15). To date, no clinical studies have proposed this concept.

The preoperative cerebral hemorrhage patterns observed in our patient warrant particular attention. The initial CT scan revealed multiple hemorrhagic foci across both cerebral hemispheres, predominantly attributable to the severe varix of the draining cortical veins. This finding underscores the significant impact of venous varicosities on cerebral hemorrhage. Subsequently, the second CT scan demonstrated an increased hemorrhage in the left frontal and parietal lobes. This progression can be directly ascribed to the elevated pressure in the SSS, indicative of a distinct hemorrhagic mechanism compared to the initial presentation. These observations highlight the complex interplay between venous features and hemorrhagic patterns, particularly in the context of venous arterialization and its hemodynamic consequences (16).

The etiology of dAVFs is multifaceted, encompassing trauma, surgery, venous stenosis, or CVST (17). In this case, DSA revealed an anatomical variation in the torcular area, a variation that accounts for about 8% of transverse sinus anomalies (18), leading to the absence of the left transverse sinus in this patient. We postulate that this could be a pivotal factor in the formation of the dAVF in this case. Previous studies have provided evidence that congenital anomalies in the venous system's anatomy may be associated with the occurrence of intracranial vascular

Abbreviations: AVF, arteriovenous fistula; dAVF, dural AVF; DSA, digital subtraction angiography; DVT, deep vein thrombosis; MMA, middle meningeal artery; OA, occipital artery; VA, vertebral artery; SSS, superior sagittal sinus; TS, transverse sinus; CV, cortical veins; CVST, cerebral sinus venous thrombosis; ICH, intracranial hematoma; NHND, Non-Hemorrhagic Neurological Deficits; CVR, cortical venous reflux; CVD, cortical venous drainage.

TABLE 1 Classification of dAVFs.

A. Borden classification and Cognard classification							
Natural course	Borden classification			Cognard classification			
	Type	Venous drainage site	CVR	Type	Venous drainage site	Flow pattern in sinus	CVR
Benign	I	Dural sinus	No	I	Dural sinus	Antegrade	No
Benign				IIa	Dural sinus	Retrograde	No
Aggressive	II	Dural sinus	Yes	IIb	Dural sinus	Antegrade	Yes
Aggressive				IIa + b	Dural sinus	Retrograde	Yes
Aggressive	III	Cortical vein	Yes	III	Cortical vein		Yes without venous ectasia
Aggressive				IV	Cortical vein		Yes with venous ectasia
Aggressive				V	Cortical vein with spinal medullary drainage		Yes

B. Modification to angiographic classification systems for DAVFs proposed by G. J. Zipfel et al.							
Modified type	Borden-Shucart Type	Cognard Type	Venous Drainage	CVD	Annual Risk (%)	Treatment Recommendation	
					ICH	Death	
1	I	I, IIa	dural sinus	No	<1	0	Elective treatment for intractable symptoms
2 w/aCVD	II	IIb, IIa + b	dural sinus	Yes	1.4–1.5	0	Elective treatment to prevent ICH/NHND
2 w/sCVD	II	IIb, IIa + b	dural sinus	Yes	7.4–7.6	3.8	Immediate treatment to prevent ICH/NHND
3 w/aCVD	III	III, IV, V	CVD	Yes	1.4–1.5	0	Elective treatment to prevent ICH/NHND
3 w/aCVD	III	III, IV, V	CVD	Yes	7.4–7.6	3.8	Immediate treatment to prevent ICH/NHND

CVR, cortical venous reflux; dAVF, dural arteriovenous fistula. CVD, cortical venous drainage; aCVD, asymptomatic CVD; sCVD, symptomatic CVD.

variations (19–21). However, this perspective is not commonly reported in cases of dural arteriovenous fistulas.

In this case, we observed a significant increase in pressure in the SSS due to 'venous arterialization'. We hypothesize that this could be associated with the specific anatomical location of the lesion, the complex and diverse arterial supply to this area, the relatively singular venous drainage outlet, and the patient's own venous variation. These factors may collectively contribute to a significant alteration in hemodynamics (16).

The patient's bilateral inferior anastomotic veins, both cavernous sinuses, and the basal sinus were notably well-developed. MR imaging further revealed that the left CV connected to the superior and inferior petrosal sinuses via the cavernous sinus, ultimately draining into the sigmoid sinus, underscoring a pronounced venous compensatory capacity. This capacity largely determined the favorable postoperative prognosis. From embryonic development to adulthood, some veins degenerate during growth (18), but this does not imply a complete loss of function in adulthood. Under certain pathological conditions, some degenerated veins may be reactivated (22, 23). Through intraoperative DSA observation, we have reason to conclude that some collateral venous circulation in this patient has been reopened. This finding suggests that we should pay more attention to the importance of intracranial veins in the management of dAVFs.

In the current case of dAVF, the transarterial approach was unequivocally the superior choice, aligning with existing guidelines (24). Prior to embolization, an attempt was made to access the contralateral sigmoid-transverse sinus, which proved unsuccessful. The dAVF was primarily supplied by bilateral OA, the left MMA occipital branch, and the VA meningeal branch. In contrast, the venous drainage was predominantly through the SSS, which was under high pressure. The transarterial approach offered several advantages, including reduced risk of redirecting blood flow to alternative venous pathways, preservation of functional venous systems, and minimization of complications unique to the transvenous route, such as abducens nerve palsy (25). Before the Onyx era, transvenous approach was the mainstay of endovascular treatment for cure of dAVF. Modern transvenous techniques involve retrograde catheterization of the affected sinus and cortical veins, using microcoils, liquid embolic agents, or a combination for occlusion (26). However, the transvenous approach necessitates meticulous patient selection to achieve complete occlusion and avoid complications. Given the complex arterial supply and singular, high-pressure venous drainage in this case, the transarterial route was indeed the most judicious choice.

The shift from using n-BCA to Onyx as the embolic agent of choice in dAVF treatment is indicative of evolving clinical perspectives,

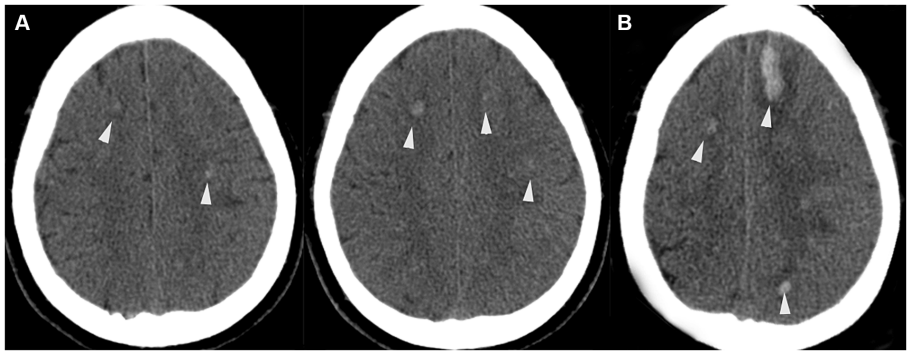

FIGURE 1
(A) Admission axial CT scan, indicating multiple scattered hemorrhagic foci in both cerebral hemispheres (white arrowheads). **(B)** Preoperative axial CT scan showing the progression of hemorrhage in the left frontal lobe (white arrowheads).

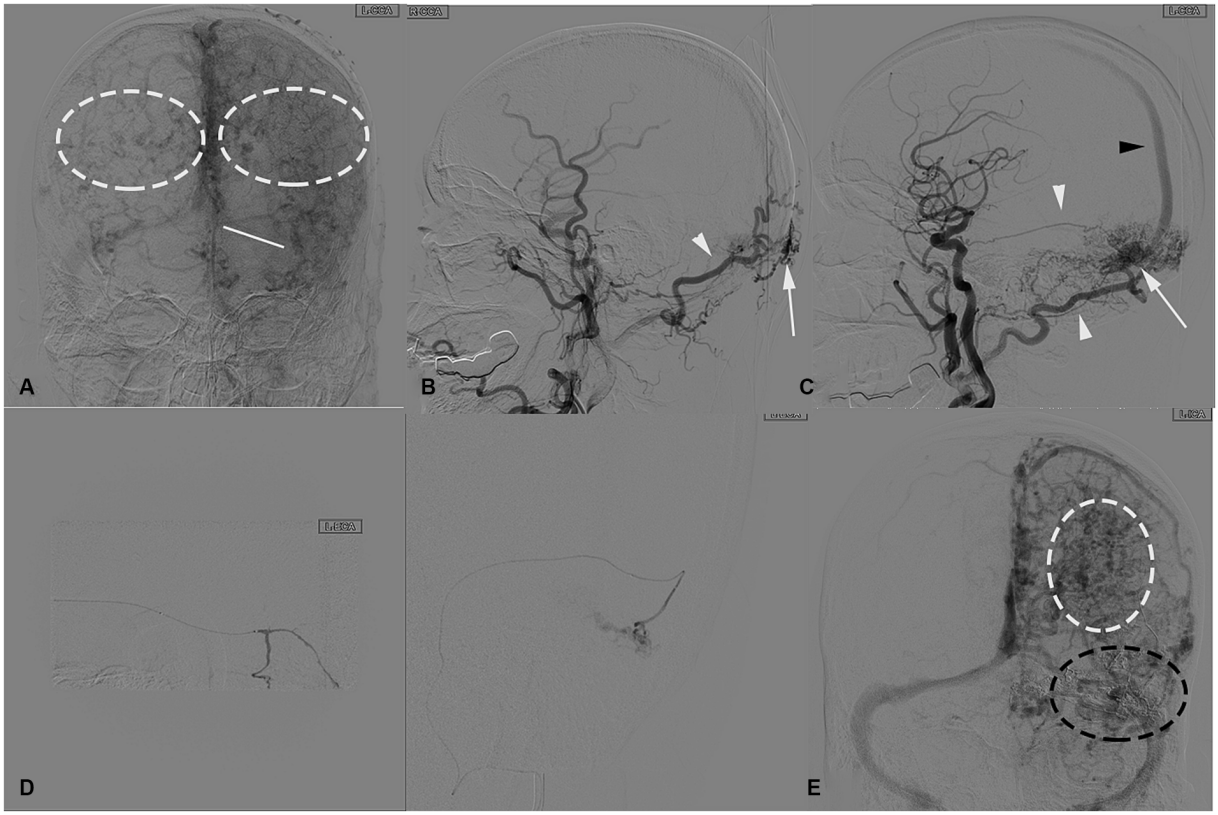

FIGURE 2
(A) Anteroposterior view of the left common carotid artery angiography venous phase before endovascular surgery, a congenital absence of the left transverse sinus is shown (white solid line) and severe varix of bilateral cortical veins (white dashed circles). In the lateral view of the left common carotid artery angiography, **(B,C)** demonstrating an arteriovenous shunt (white arrowheads). The arteriovenous shunt is fed by the left occipital artery (white arrow), left MMA occipital branch (white arrow), and drains into SSS (black arrow) around the torcular area. **(D)** Superselective angiography of the left occipital artery and MMA branch obtained before ONYX glue injection, demonstrating multiple arterial channels flowing into the fistula.
(E) Intraoperative angiography obtained after ONYX injection shows a cast of glue occluding the fistula, including all the left feeding arteries and part of the compensatory veins (black dashed circle), with increased cortical vein varix (white dashed circle).

driven by the quest for better control and reduced complications. While n-BCA has been a staple for high-flow dAVFs, its limitations, such as reduced controllability and a higher risk of complications, have made Onyx a more favorable choice (27). Onyx's longer injection time allows for a more controlled embolization, particularly beneficial when venous routes are compromised (28). This case employed Onyx 20, aligning with its established safety profile and efficacy. However, it's crucial to acknowledge that even with the optimal choice of Onyx and a transarterial approach, we observed some unintended glue migration into the SSS in postoperative imaging. This serves as a cautionary note on the inherent complexities and risks involved, emphasizing the need for meticulous technique and material selection.

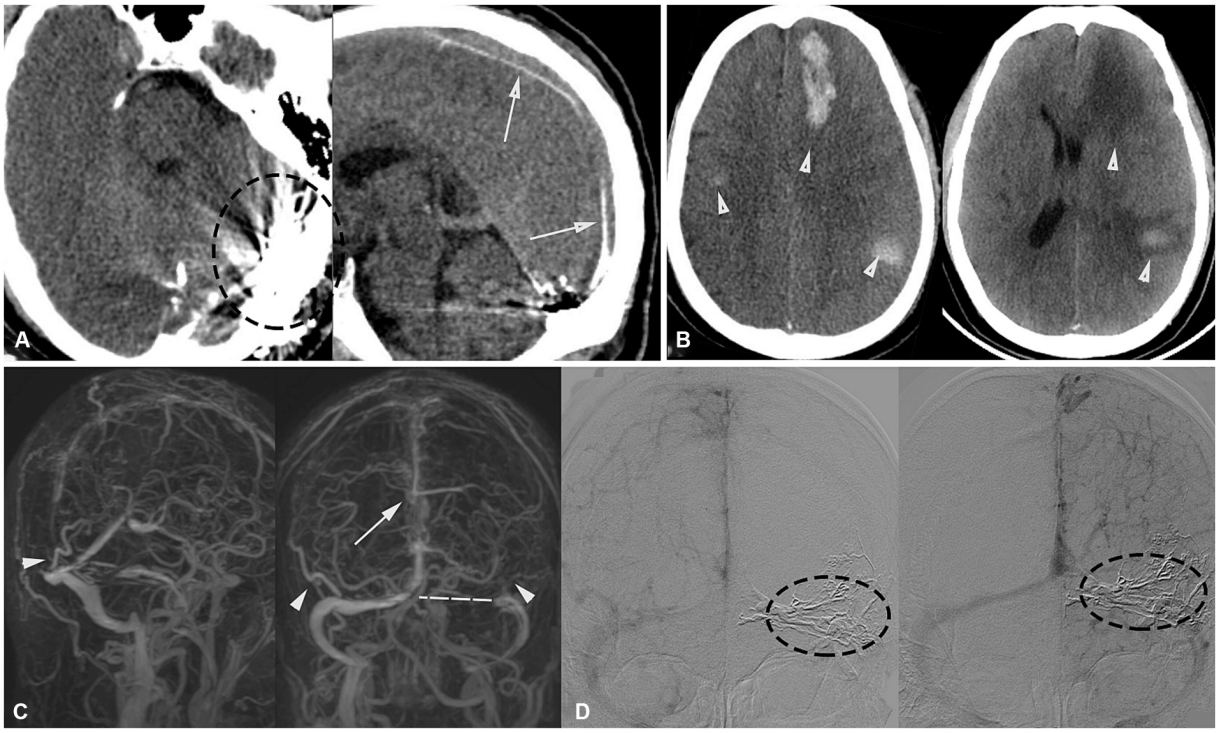

FIGURE 3
(A) Post-embolization axial and sagittal CT scans show a high-density area, demonstrating a cast of glue occluding the fistula (black dashed circle), including feeding arteries and the SSS (white arrow). **(B)** Postoperative follow-up CT revealed hemorrhage in the left frontal lobe, posterior temporal region, and right temporal lobe. Discharge review showed that the hematoma had been essentially absorbed (white arrowhead). **(C)** Postoperative follow-up MRV images in oblique posterior and posterior views revealed well-developed bilateral Labbé veins (white arrowheads), a variation in the left transverse sinus (white dashed line), and the presence of thrombosis in the SSS (white arrow). **(D)** DSA 3-month follow-up compared to post-surgery shows a significant reduction in cortical venous varix (left), with the fistula completely occluded and no recurrence.

The newer agents like Squid and PHIL, although promising, are yet to be widely adopted in clinical practice (29, 30).

According to the prevalent classifications (6, 7) (Table 1), this case falls under Cognard Classification IV or Borden classification III. It is crucial to recognize the historical significance of the DjinDjian classification (31) in laying the groundwork for our understanding of cranial AVFs, which has substantially influenced subsequent classifications, including those referenced in our study. Transarterial embolization remains the preferred treatment for this type (24). Although Zipfel et al. (5). have expanded the classification to account for ICH and NHND, our case presents unique challenges to these existing frameworks. The severe cortical venous varicosities in our patient, leading to bilateral widespread scattered hemorrhages, highlight the limitations of current classifications and suggest the need for their refinement. Future classifications could benefit from differentiating between unilateral and bilateral hemorrhages, as well as incorporating a grading system for venous ectasia severity. Additionally, considering venous anomalies in the classification could provide a more comprehensive understanding of these conditions and their implications for treatment. Furthermore, integrating a post-embolization therapeutic classification could offer valuable insights into the outcomes and efficacy of interventional treatments, enhancing our approach to managing these complex cases. Unlike merely indicating the presence of CVR or ICH, the occurrence of these kinds of hemorrhages implies higher intracranial pressure, which poses a challenge to our treatment choices, necessitating a careful evaluation of the patient's venous compensatory ability before determining the final approach.

Conclusion

This case study presents a rare instance of a complex torcular dural arteriovenous fistula leading to severe cortical venous reflux-induced varix and subsequent bilateral cerebral hemispheric hemorrhage. It offers novel insights into the etiology, hemodynamics, classification, and venous compensation of this condition. The case underscores the need for further research and a broader understanding of these aspects, emphasizing the importance of considering venous compensatory capacity in treatment planning. It challenges existing classifications and calls for more attention to this potential subtype in the neurosurgical community.

Patient perspective

"I first came to the hospital's neurosurgery outpatient clinic because I was feeling dizzy and tired. The doctors ran a bunch of tests, including a digital subtraction angiography (DSA), which showed something wasn't right with the blood vessels in my brain. They moved me quickly to the ICU, and I remember being aware of what was happening. But by the next day, things started to get really

uncomfortable, and everything around me seemed blurry and confusing.

At the time, I did not understand why I was feeling that way. It was only later that I learned from my family that the doctors had found my bleeding was getting worse, and that's why they rushed me into surgery. I do not recall much about the procedure itself, just waking up to anxious faces and the beeping of machines. After the surgery, I spent a week in the ICU, which was filled with pain and uncertainty, and I had to rely on my family and doctors to piece together what happened during those days.

As time went on, the doctors told me and my family that my bleeding was under control and the hematoma in my brain was slowly being absorbed. That period was a rollercoaster of emotions, from despair to hope. Follow-up checks 3 months later showed that my arteriovenous fistula had not come back, and the varices in my brain were much better. Even though a check-up a year later found a clot in my superior sagittal sinus, the doctors said it did not need any special treatment.

Now, I can pretty much take care of myself. I get the occasional slight headache, but there are no serious neurological issues, and my vision has improved. Looking back, that time was as much a challenge as it was a period of growth for me. I'm deeply grateful for the skill and care of the doctors and nurses. They did not just treat my condition; they gave me a chance at a new life".

Limitations

A limitation of this case is the missing ophthalmological imaging from an outpatient visit. Only written records are available. Nonetheless, follow-up imaging adequately addresses this inconvenience.

Data availability statement

The raw data supporting the conclusions of this article will be made available by the authors, without undue reservation.

Ethics statement

Written informed consent for the publication of any potentially identifiable images or data included in this article was obtained from the individual(s) and their next of kin.

Author contributions

ZL: Conceptualization, Data curation, Formal analysis, Investigation, Methodology, Software, Writing – original draft. PH: Conceptualization, Software, Visualization, Writing – original draft. QC: Conceptualization, Project administration, Supervision, Writing – original draft. SM: Data curation, Formal analysis, Writing – original draft. JL: Conceptualization, Supervision, Writing – original draft. YF: Methodology, Writing – original draft. WJ: Investigation, Writing – original draft. XT: Project administration, Supervision, Writing – review & editing. SW: Conceptualization, Funding acquisition, Project administration, Resources, Supervision, Writing – review & editing.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Supplementary material

The Supplementary material for this article can be found online at: https://www.frontiersin.org/articles/10.3389/fneur.2023.1303234/full#supplementary-material

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False positive angiographic aneurysm of the anterior segment of the M1 bifurcation of the middle cerebral artery: a case report

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Occlusion of an intracranial arterial branch, resulting in a false positive aneurysm on vascular imaging, is extremely rare, with only a few reports in the literature and mostly in the posterior circulation artery or the middle cerebral artery (MCA) bifurcation. We report a case of a 69 years-old woman with a subacute infarct lesion in the left frontal lobe, for whom both computed tomographic angiography (CTA) and digital subtraction angiography (DSA) of the cerebral vessels showed aneurysms in the anterior segment of the M1 bifurcation of the middle cerebral artery (MCA) and in the bifurcation of the MCA. The aneurysm in the MCA bifurcation was found during craniotomy, whereas the anterior segment of the M1 bifurcation had intact branch vessels with severe atherosclerosis and no aneurysm was present. The branch vessel of M1 was presumed to be atherosclerotic occlusion resulting in the distal vessels without contrast filling on CTA and DSA, and only the occluded stump at the beginning of the vessel was filled with contrast, showing an aneurysm-like morphology, which was very confusing. This case highlights to neurologists that the diagnosis of aneurysm by cerebrovascular CTA or DSA must be carefully differentiated to avoid misdiagnosis, especially if the unruptured aneurysm is in an uncommon location in combination with ischemic cerebrovascular disease.

KEYWORDS

middle cerebral artery (MCA), occluded stump, false-positive aneurysm, CTA, MRA, DSA

Introduction

Occlusion of an intracranial arterial branch, resulting in a false positive aneurysm on vascular imaging, is extremely rare, with only a few reports in the literature and mostly in the posterior circulation artery or the MCA bifurcation (1–3). A so-called false positive aneurysm is actually a stump of a proximal artery following a distal arterial occlusion. To expand the clinical diagnostic thinking of neurologists, we report a rare case of a false positive angiographic aneurysm of the anterior segment of the M1 bifurcation of the MCA combined with a true aneurysm in the MCA bifurcation.

False positive aneurysms at the M1 bifurcation of the middle cerebral artery (MCA) are rare. Neurologists should be cautious when diagnosing aneurysms using computed tomography angiography (CTA) or digital subtraction angiography (DSA). It is important to differentiate accurately to avoid misdiagnosis, especially for unruptured aneurysms in rare locations

associated with ischemic cerebrovascular disease. Magnetic resonance imaging (MRI) fusion techniques may be helpful in the differential diagnosis of false positive aneurysms.

Case report

A 69 years-old woman with a history of hyperlipidemia suddenly developed dysphasia and left-sided facial palsy during a walk 1 week previously and was hospitalized because her symptoms continued to worsen. Diffusion-weighted imaging (DWI) revealed a subacute infarct lesion in the left frontal lobe (Figure 1A). CTA of cerebral vessels revealed a cystic aneurysm in the anterior segment of the M1 bifurcation of the left MCA, approximately 3.6 mm × 4.2 mm and a cystic irregular aneurysm in the MCA bifurcation, approximately 4.7 × 4.8 mm (Figure 1C). Magnetic resonance angiography (MRA), DSA (Figures 1D,E), and CTA findings were consistent, but the MCA bifurcation showed a defect in the MRA (Figure 1B). Considering that the patient had two aneurysms in the MCA, especially the risk of rupture of the cystic irregular aneurysm in the MCA bifurcation, a craniotomy was performed to clip the aneurysms. The 3Dslicer and Freesurfer software (4) were used to simulate the surgical approach before the operation to clearly show the location and morphology of the abovementioned two aneurysms (Figure 1F).

During surgery, after separating the lateral fissure vein, the MCA bifurcation aneurysm was exposed and clipped (Figure 1G). We continued to investigate along the posterior part of the M1 bifurcation and found a branch of the M1 bifurcation with an intact vessel in the anterior part of the M1 bifurcation, which showed obvious atherosclerotic changes, and the beginning of the vessel here was misdiagnosed as an aneurysm before surgery, which was very confusing (Figures 1G,H). After repeated confirmation and careful judgment, we diagnosed that the aneurysm shown on angiography should be a vascular stump after stenosis or occlusion caused by atherosclerosis in the distal vessel, and not an aneurysm, so no further operation was performed.

After surgery, she was discharged from the hospital after 2 weeks of medication and presented with residual mild symptoms of speech disfluency. At the 6 months follow-up, the patient exhibited significant recovery, demonstrating restored speech function, and was maintained on long-term oral anticoagulants and lipid-lowering agents.

Discussion

A meta-analysis that included 68 studies of 94,912 patients from 21 countries found that the overall prevalence of unruptured intracranial aneurysms was approximately 3.2% in people without other comorbidities (5). Another systematic evaluation including 12,781 patients with a definite diagnosis of TIA/light stroke yielded a prevalence of 5.1% for unruptured intracranial aneurysms (6). From this, we hypothesized that the probability of intracranial aneurysm is higher in patients with combined ischemic cerebrovascular disease than in patients without ischemic cerebrovascular disease; therefore, in this case, cerebrovascular imaging was performed, and two aneurysms were found.

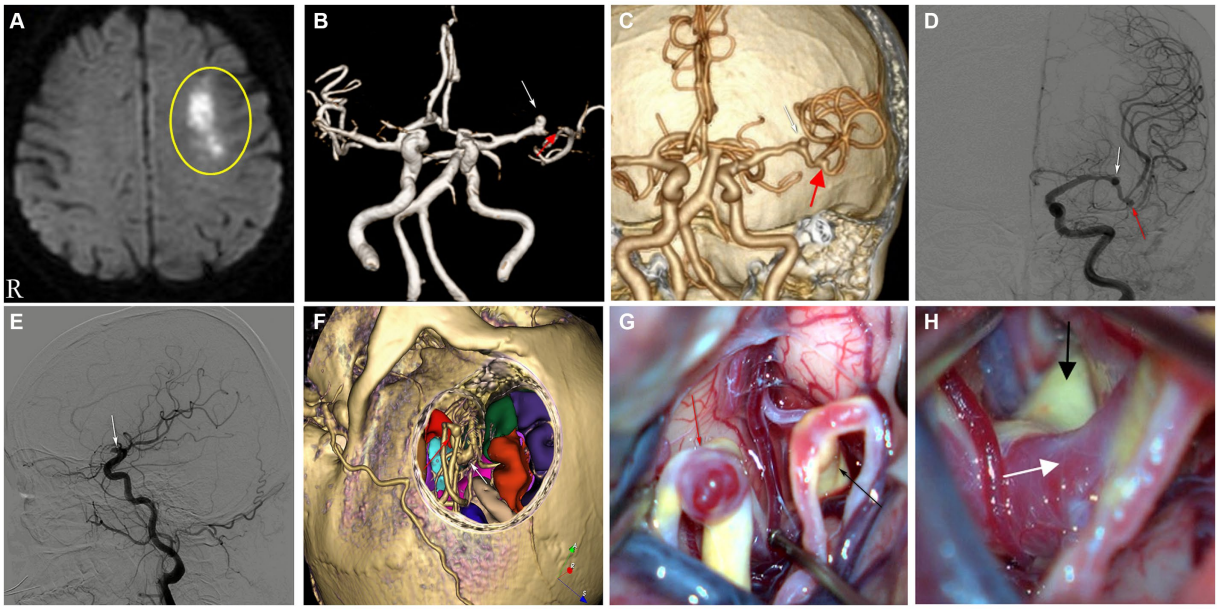

FIGURE 1
(A) DWI showing acute infarct lesion in the anterior segment of the left frontal hemi-oval center. **(B)** MRA showing aneurysm of the left MCA-M1 bifurcation anterior segment (white arrow) and MCA bifurcation showing a defect (red arrow). **(C,D)** CTA and DSA (orthostatic), aneurysm of the anterior segment of the M1 bifurcation of the left MCA (white arrow), MCA bifurcation aneurysm (red arrow). **(E)** DSA (lateral), aneurysm of the anterior segment of the M1 bifurcation of the left MCA (white arrow). **(F)** Preoperative surgical simulation showing aneurysm of the left MCA-M1 bifurcation anterior segment (white arrow), showing aneurysm of the left MCA bifurcation (red arrow). **(G)** Presence of an intact vessel branch in the M1 bifurcation anterior segment with significant atherosclerosis (black arrow) and aneurysm of the left MCA bifurcation (red arrow). **(H)** The left MCA-M1 bifurcation anterior segment trunk with marked atherosclerosis (black arrow), the branch vessel initiation segment there, which is a false positive aneurysm, shown by cerebrovascular imaging (white arrow).

This case had many unusual features, including the clinical presentation, the structural features of the arterial occlusion, and the anatomy of the MCA branches and combined MCA-M1 bifurcation and MCA bifurcation aneurysms, which contributed to the initial misdiagnosis. Spontaneous intracranial subarachnoid hemorrhage induced by a ruptured middle cerebral artery aneurysm is common, and the choice of craniotomy or endovascular intervention is safe and effective, but the latter has a higher rate of asymptomatic thromboembolic events, more frequently in the setting of acute subarachnoid hemorrhage (7). To date, no reports of false positive aneurysms on angiography have been reviewed in the literature on spontaneous subarachnoid hemorrhage.

In contrast, a meta-analysis of unruptured MCA aneurysms shows that a surgical clipping aneurysm remains highly safe and effective. The efficacy and safety of endovascular treatment of unruptured MCA aneurysms continue to improve; however, it has a low occlusion rate (8).

MRA, CTA, and DSA are commonly used to detect intracranial aneurysms. One study found that the sensitivity of MRA for detecting intracranial aneurysms was 95% and the specificity was 89%, false negative and false positive aneurysms detected on MRA were mainly located at the skull base and MCA (9). At present, DSA is commonly used internationally as the "gold standard" for the diagnosis of intracranial aneurysms, especially 3D-DSA, which has high sensitivity and specificity for the diagnosis of small aneurysms (10). Therefore, cerebrovascular CTA and DSA are more accurate in the detection of intracranial aneurysms. In this case, cerebrovascular CTA and DSA were performed successively, both of which revealed the presence of two aneurysms: left MCA-M1 bifurcation and MCA bifurcation aneurysms. However, during the craniotomy, we found that the patient's left MCA-M1 bifurcation, which was actually a branch vessel of the MCA, showed severe atherosclerotic changes, and during the disease progression, the vessel was occluded, so both CTA and DSA failed to provide us with the whole vessel morphology, and the angiography only showed the morphology of the stump after vessel occlusion, which looked very much like an aneurysm, thus leading to misdiagnosis.

Vascular stumps that are misdiagnosed as aneurysms are mostly located in the posterior circulation and MCA. Only one case report of a cerebral angiography of a 70 years-old man revealed an aneurysm at the intersection of the anterior communicating artery and the A1-A2 segment of the right anterior cerebral artery, which was found to be a tapered duplication of the A1 segment of the right anterior cerebral artery by craniotomy and, thus a false positive aneurysm (11).

To the best of our knowledge, there are no reports of false positive aneurysms on angiographic imaging of the MCA-M1 bifurcation, and our case is the first of its kind. Yu et al. (3) reported on a 57 years-old man with recurrent right-sided weakness and aphasia, who was diagnosed with an aneurysm at the left MCA bifurcation on both CTA and DSA, with Moyamoya phenomenon in the vicinity. However, the "aneurysm" was found to be the stump of an occluded vessel during craniotomy. Lee et al. (1) reported a 26 years-old man with recurrent left-sided hemiparesis and an aneurysm at the right MCA bifurcation on both CTA and DSA. However, three bifurcations of the middle cerebral artery with normal upper and lower trunk vessels and significant atherosclerosis in the middle trunk branch vessels were found during craniotomy, so it was postulated that the "aneurysm" shown in the imaging was only the stump of the middle trunk branch after occlusion. Park et al. (2) reported two cases of occluded vessel stumps at MCA bifurcations that showed false positive aneurysms on angiography. Case 1 was a 52 years-old woman with no symptoms; a left middle cerebral artery aneurysm was found by MRA and CTA, along with moyamoya phenomenon near the aneurysm. Case 2 was a 62 years-old woman with years of non-specific headaches and mild hypertension; a right middle cerebral artery aneurysm was found by MRA and DSA. The false positive aneurysms reported above were all diagnosed intraoperatively as vascular stumps.

In addition, there were two cases of MCA aneurysms diagnosed by angiography that were confirmed to be vascular stumps after conservative treatment (12, 13). In one of these cases, after the patient had to opt for conservative treatment for personal reasons, it was detected on a review of the cerebral angiogram that the pre-treatment diagnosis was incorrect (12). These are the rare cases that can be searched so far, indicating the lack of awareness among neurologists that intracranial vascular stumps may present as aneurysmal patterns on vascular imaging.

In summary, there is a total of six cases of false positive middle cerebral artery aneurysms reported prior to our report, with patients undergoing DSA angiography with vascular stumps located at the bifurcation or trifurcation of the middle cerebral artery. Four of these cases had symptoms of cerebral ischemia, and the other two cases were found during screening for cerebrovascular disease. In contrast, the false positive aneurysm (vascular stump) in our case was located in the anterior segment of the M1 bifurcation of the MCA, a highly confusing location, and interestingly, there was also a true aneurysm at the M1 bifurcation of the MCA, which could easily be misdiagnosed as a multiple aneurysm of the MCA.

This case highlights that the traditional cerebral vascular imaging tools, namely, MRA, CTA, and DSA, are not absolutely reliable for the diagnosis of intracranial aneurysms. Neurologists need to take into account that atherosclerosis or arterial entrapment can lead to occlusion or stenosis of the vessel and the formation of a vascular stump, resulting in a false positive aneurysm on angiography. If such patients are misdiagnosed and endovascular interventional embolization or craniotomy is performed, it may lead to adverse outcomes.

Kuribara et al. (14) utilized 3D-fast imaging employing steady-state acquisition (FIESTA) and MRA fusion imaging on the occluded MCA's distal segment, revealing vascular details beyond the occlusion, up to the M3 segment. Similarly, Ozaki et al. (15) combined 3D-T2 sampling perfection with application-optimized contrast using different flip angle evolution (SPACE) and MRA, providing a visualization of the occluded artery by merging the flow void effect in T2-SPACE with its MRA image. These MR fusion techniques may be instrumental in delineating obstructed vessels before mechanical thrombectomy (MT) for acute cerebrovascular occlusions. In addition, high-resolution vessel wall imaging (HRVWI) has been used to evaluate intracranial vascular pathologies such as intracranial atherosclerosis, occlusion, Moyamoya disease, vasculitis, and reversible cerebral vasoconstriction syndrome, and is useful for evaluating the vessel wall in the presence of stenosis (16).

In our case, if the abovementioned examinations had been perfected preoperatively, the branch stenosis originating from the anterior segment of the M1 bifurcation of the MCA may have been detected, thus avoiding misdiagnosis of it as an aneurysm. A literature

search revealed that there are no relevant studies utilizing the abovementioned examinations as a means of identifying false positive aneurysms in vascular stumps.

Conclusion

While infrequent, false positive aneurysms identified by vascular imaging warrant consideration by neurologists. This is particularly pertinent in cases where patients exhibit unruptured aneurysms in atypical locations concomitant with other ischemic cerebrovascular pathologies, including cerebral infarction or Moyamoya disease. A judicious selection of therapeutic strategies is imperative. In addition, MR fusion techniques may be helpful in the differential diagnosis of false positive aneurysms.

Data availability statement

The original contributions presented in the study are included in the article/supplementary material, further inquiries can be directed to the corresponding author.

Ethics statement

The studies involving humans were approved by Hospital of Chengdu University of Traditional Chinese Medicine. The studies were conducted in accordance with the local legislation and institutional requirements. The participants provided their written informed consent to participate in this study. Written informed consent was obtained from the individual(s) for the publication of any potentially identifiable images or data included in this article.

Author contributions

WX: Writing – original draft. XH: Writing – original draft. DL: Conceptualization, Data curation, Writing – review & editing. DY: Supervision, Writing – review & editing.

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Cerebral venous sinus stenting and jugular bulb embolization for pulsatile tinnitus: A case report

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Background: Pulsatile tinnitus (PT) is a rare form of tinnitus that aligns with the heartbeat. It is typically brought on by lesions with significant vascularity, which produce aberrant sound conduction and increase the risk of mental health issues and hearing loss. Venous PT is more prevalent than arterial PT. Open procedures or interventional procedures can be used to treat PT. We present here a case of PT caused by venous luminal stenosis combined with jugular bulb (JB) malformation, which was improved by stenting and JB embolization.

Case presentation: A 59-year-old woman presented with long-term tinnitus consistent with heart rhythm and hearing loss, accompanied by anxiety, insomnia, and depression. The results of brain MRV, CT, and DSA showed stenosis of the right sigmoid sinus and high jugular bulb (JB) with dehiscence of the JB wall. The patient saw a significant improvement in PT symptoms following sigmoid sinus stenting and spring coil embolization of the high JB, following the diagnosis of PT. The patient had no PT recurrence for the course of the 31-month follow-up period.

Conclusion: In the present PT case, there was a simultaneous onset of the right sigmoid sinus stenosis and the high JB with the JB wall abnormalities. Sigmoid sinus stenting and spring coil embolization of high JB may be a treatment for the PT, but the prevention of post-stenting complications is still an issue that requires great attention and needs further study.

KEYWORDS

pulsatile tinnitus, jugular bulb, sigmoid sinus, stenting, spring coil embolization

1 Introduction

Pulsatile tinnitus (PT) is a form of tinnitus characterized by the perception of a rhythmic sound that can be attributed to various identifiable causes (1). This condition typically manifests unilaterally and is caused by alterations in vascular hemodynamics or abnormal conduction of normal sounds. PT can have a significant impact on the patient's overall well-being, often leading to sleep disturbances, anxiety, depression, and in extreme cases, suicidal ideation (2). Furthermore, PT can serve as an early indicator of an underlying and potentially serious medical condition.

Common vascular causes of PT include arterial anomalies, venous anomalies, and arteriovenous fistulae, among which venous PT(VPT) includes idiopathic intracranial hypertension(IIH), venous sinus stenosis(VSS), sigmoid sinus wall abnormalities(SSWA),

jugular Vein and other venous malformations (3). VPT can be ameliorated by open procedures such as surface reconstruction/plastic surgery or endovascular interventions such as stenting (4). However, the relatively few reports and pathologic knowledge of venous PT predispose to misdiagnosis and underdiagnosis of PT, which in turn affects therapeutic decisions. Therefore, the etiology and imaging diagnosis of venous PT is crucial (5).

In this case study, we encountered a patient who experienced PT as a result of both VSS and high jugular bulb(JB) issues. The patient underwent successful treatment through the implantation of a sigmoid sinus stenting and subsequent JB embolization. Notably, there was no recurrence of symptoms during the 31-month follow-up period. This report presents the first documented instance of the successful combination of sigmoid sinus stent implantation and JB embolization for the treatment of PT. We report this case in the hope of giving neurosurgeons some references about the diagnosis and treatment of VPT.

2 Case description

The patient, a 59-year-old female, presented with bilateral symmetrical low-key tinnitus and accompanying hearing loss three years ago. Over the past year, she experienced a progressively worsening blowing or running sound in her right ear, resembling a "murmur," along with synchronous with her heartbeat rhythm. Concurrently, her original symptoms worsened, manifesting as slightly decreased visual acuity, insomnia, anxiety, depression, and suicidal ideation. She had previously been diagnosed with sudden deafness and received microcirculation treatment but did not experience any improvement.

The patient reported that the persistent tinnitus in his right ear was seriously affecting his normal life and urgently needed treatment. The patient had suffered from hypertension and hyperlipidemia in the past and was taking medication regularly to keep his blood pressure and blood lipids within normal limits, with no history of other illnesses, no history of allergies to medications or food, and no history of such hereditary diseases in his family. We assessed the patient's tinnitus, sleep and anxiety levels using the Tinnitus Handicap Inventory(THI), the Pittsburgh Sleep Quality Index (PSQI) and the Hamilton Depression Scale(HAMD), with the following results: THI: 56/100 points (Grade 3); PSQI: 14/21 points; and HAMD: 10 points, confirming moderate tinnitus handicap, sleep disorder, and mild anxiety state.

After admission, we performed neurological examinations: (1) intracranial pressure was normal; (2) otoscopy: bilateral external auditory canals were patent, tympanic membranes were intact, grayish-white in color, and no congestion or fluid flatness was seen; the hearing loss was observed in both ears by pure tone audiometry, and tympanic ventricular conductance mapping showed a pattern of 3C; (3) A computed tomography (CT) scan of the right ear showed: normal right sigmoid sinus wall and stenosis at the junction of the right internal jugular vein(IJV) and the sigmoid sinus, as well as a high JB on the right accompanied with Jugular bulb wall dehiscence (JBWD) (Figure 1). Digital subtraction angiography (DSA) showed moderate-to-severe sigmoid sinus stenosis(SSS) at its junction with the IJV (stenosis of approximately 50%–70%) and a high JB (Figure 1), and distal venous sinus manometry was performed. Initially, we thought that the PT in the right ear might be due to the SSS increasing the blood flow velocity creating a vortex in the high JB, so that the PT sound entered the inner ear. Two other evidences supported our diagnosis: first, during DSA venography, the contrast catheter passed through the stenosis site, and when the pressure was measured at the distal end, it changed the direction of blood flow at the site, and the venous vortex flow was reduced, and the patient's PT disappeared; second, by the right side neck-pressure test, when the pressure was increased to a certain degree, the PT disappeared on the patient's right side after the venous reflux was blocked, and the left side hearing was not changed. However, the results of subsequent interventions confirmed more than just what was previously stated.

3 Therapeutic intervention

Following discussions among the neurosurgeons, it was determined that the initial course of action should involve endovascular treatment of the SSS. DSA was performed under local anesthesia to confirm the presence of severe stenosis at the junction of the right sigmoid sinus and the IJV, including a right high JB as depicted in Figure 2. A 6F Long Sheath (Penumbra, California, USA) was inserted into the right femoral vein and positioned at the beginning of the right IJV, below the high JB. The stenosis was successfully crossed using a Synchro 0.014-in micro guidewire (Stryker, Michigan, USA), followed by the placement of a Neuro RX 2.50×12 mm balloon (Sinomed, Tianjin, China) across the stenosis. The balloon was then inflated to a pressure of 6 atm, resulting in an improvement of the stenosis. Subsequently, the balloon was withdrawn and a Solitaire AB 6×30 mm stent (Medtronic, Minnesota, USA) was deployed at the target site through an Excelsior XT-27 microcatheter (Stryker, Michigan, USA). Post-stenting DSA revealed the disappearance of the stenosis, with contrast-medium observed in the right high JB as shown in Figure 2. The patient reported a reduction in post-treatment pain intensity by approximately 70% to 80%.

Regrettably, the patient experienced a recurrence of PT on the third day following the stenting procedure. The second DSA showed no thrombosis in the stent at the sigmoid sinus, good improvement of stenosis, and no recurrence of stenosis with smooth blood flow (Figure 2). Therefore, we considered that even if the SSS was improved, the blood flow velocity in the sigmoid sinus was only transiently improved, eddy currents still appeared in the high JB, and the jugular bulb wall rupture due to the high JB caused the patient's PT to recur. To address this, a second intervention known as stent-assisted spring coiling embolization was performed. The procedure involved the use of general anesthesia and a 6F Long Sheath (Penumbra, California, USA) to introduce an Excelsior SL-10 microcatheter (Stryker, Michigan, USA) into the right transverse sinus, confirming that the microcatheter was located in the sigmoid sinus stent. A Precise RX 7×40 mm stent (Cordis, Florida, USA) was then deployed at the junction of the right sigmoid sinus and the IJV along a Transend 300 cm guidewire (Stryker, Michigan, USA). The microcatheter was adjusted to enter the high JB, where an 8×30 mm spring coil (Stryker, Michigan, USA) was positioned to create a basket-like structure. Subsequently, four intracranially

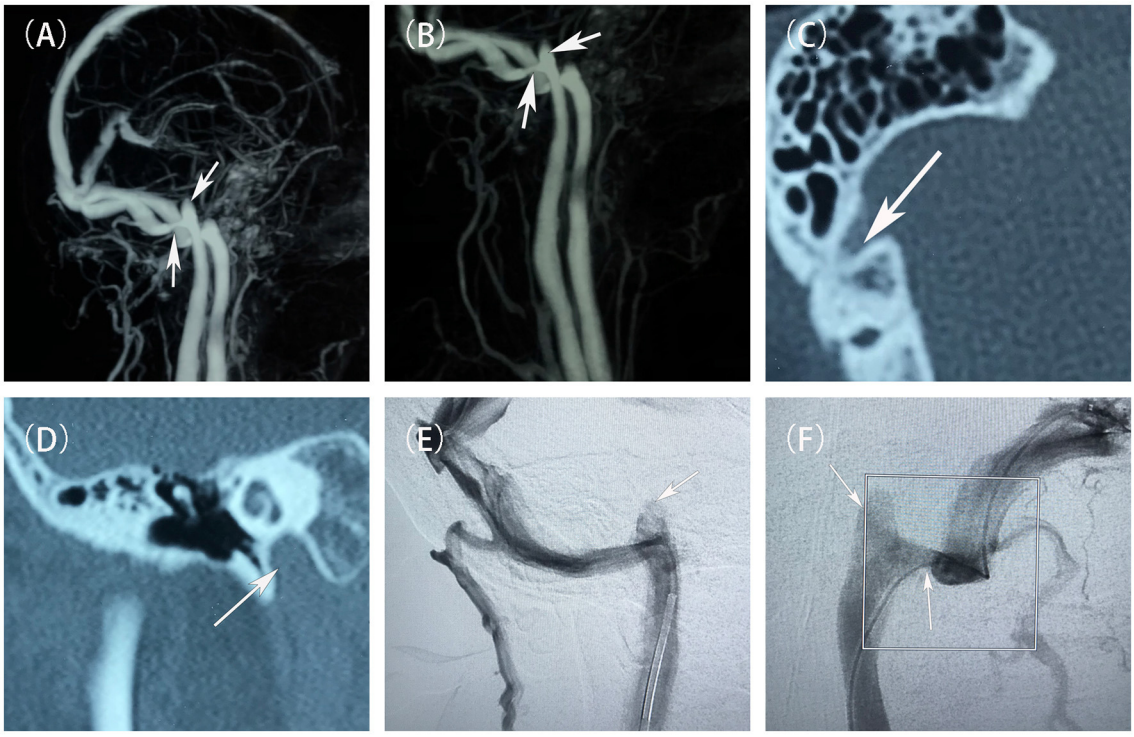

FIGURE 1
Preoperative evaluation and diagnostic imaging of the patient. **(A, B)** Pre-treatment MRV scans showed the stenosis at the junction of the right sigmoid sinus and the internal jugular vein, and a high jugular bulb (white arrow). **(C)** Pre-treatment non-contrast CT scans showed a branch of the mastoid vein emanating from the right sigmoid sinus. **(D)** Pre-treatment non-contrast CT scans demonstrated the right high jugular vein bulb invaded the inner ear structures. **(E, F)** Pre-treatment DSAs showed that a narrowing of the sigmoid sinus at its junction with the internal jugular vein, and a high jugular bulb (white arrow).

detachable spring coils (8×40 mm, 7×40 mm, 6×60 mm, 6×20 mm, Gachy, Shanghai, China) were introduced to embolize the High JB. Post-intervention DSA imaging revealed smooth blood flow within the stent and no contrast medium in the right high JB (Figure 2). The patient reported the disappearance of their PT after awakening from anesthesia. Postoperative anticoagulation treatment was administered, consisting of Dabigatran (Boehringer-Ingelheim, Shanghai, China) 150 mg twice daily for the first 6 months, followed by a reduced dosage of 75 mg for the subsequent 6 months.

4 Follow-up and outcomes

At the 12-month and 31-month follow-up examinations, the patient's right PT has not been recurrent. Both DSA images demonstrated unobstructed blood flow within the stent and complete occlusion of the high JB (Figure 2). THI scores at the two follow-ups were 12/100 (Grade 1) and 10/100 (Grade 1). The timeline of the present case (Figure 3).

5 Discussion

Causes of synchronous PT can be categorized as vascular or non-vascular. Vascular causes can be further divided into arterial, venous, and arteriovenous causes. Examples of vascular causes include carotid artery aneurysms, dural venous sinus anomalies, and arteriovenous fistulas. Venous abnormalities are more commonly associated with PT than other etiologies (3). Approximately 10% of PTs are attributed to venous lumen (or sinus) abnormalities (5). In a study by Walters, venous abnormalities were found in 11 out of 18 PT patients, including high or dehiscent jugular bulbs, dural venous sinus anomalies, and venous thrombosis. Compression of the ipsilateral IJV can improve PT symptoms and indicate an association with venous anomalies (6).

In this particular case, the patient initially reported PT with a heartbeat rhythm, suggesting a possible association with vascular abnormalities. After conducting a right neck compression test and DSA, we believe that the patient's PT may be due to SSS. Unilateral SSS usually causes ipsilateral PT, and it would be interesting to find a papillary bone defect on CT (3), whereas SSWA is often associated with stenosis of the sigmoid sinus, and by compression of the ipsilateral IJV, the ipsilateral PT is diminished, and there is no abnormality on the other side. SSS and SSWA usually occur on the right side because the right side is often the area of dominant drainage. Venous stenosis creates turbulent blood flow, resulting in a sound. The pathophysiological mechanisms behind this sound may be related to trans-stenotic gradient pressure (7, 8). The accelerated flow velocity caused by SSS may lead to SSWA because of higher pressures on the vessel wall and lower wall shear stress (9). SSWA amplifies the abnormal flow sounds due to SSS, further

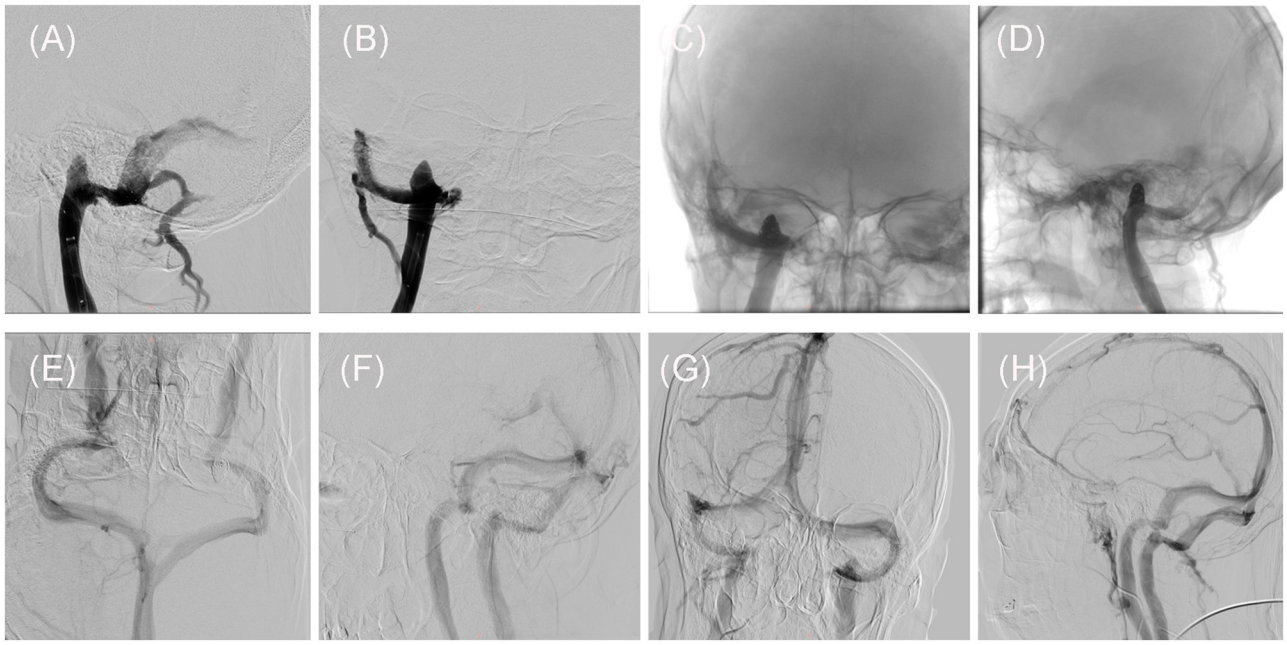

FIGURE 2
Intraoperative and post-treatment follow-up DSAs of the patient. **(A, B)** Intraoperative DSA showed that the stenosis has been improved by placement of the SAB stent, but the high jugular bulb remains on Dec. 28, 2020. **(C, D)** Intraoperative DSA demonstrated the presence of the spring coils within the high jugular bulb, on Jan. 04, 2021. **(E, F)** The first follow-up DSA showed that the stent at stenosis and the high jugular bulb embolized by the spring coils, on Jan. 17, 2022. **(G, H)** The second follow-up DSA showed that the stent at stenosis and the high jugular bulb embolized by the spring coils, on Aug. 14, 2023.

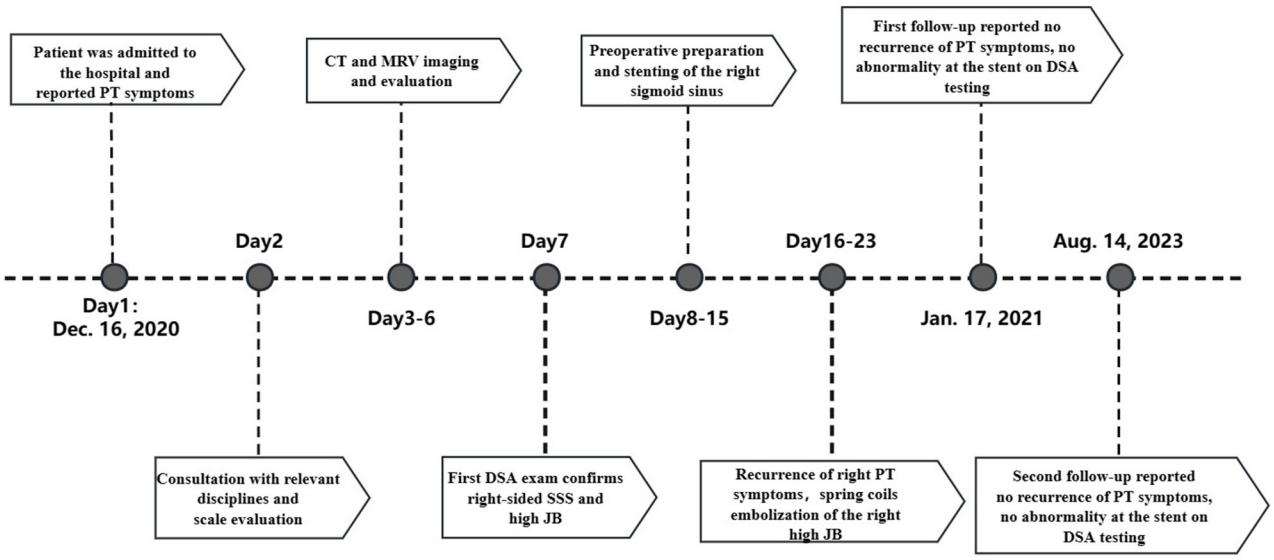

FIGURE 3
The timeline of the present case.

promoting PT. Some studies have suggested that it is SSWA that is the main determinant of PT occurrence (10). We believe that the PT in this case was caused by SSS. Because our careful review of all of the patient's imaging did not reveal anything about SSWA, we only identified SSS. Therefore, we hoped that by the first stent placement in SSS, by improving stenosis, decreasing blood flow velocity, and utilizing the shielding effect of the stent to decrease vascular tone transmission, we would temporarily leave the high JB untreated, as it has been shown that PT disappeared in some patients after improving stenosis while leaving the high JB untreated (11). In this case, the PT immediately disappeared after the first procedure, confirming the relationship between SSS and the PT.

However, the patient's PT reappeared after the third postoperative day when the stent was normal, suggesting that the PT was not only due to SSS but also possibly due to high JB and JBWD. Improvement of stenosis by stent placement alone was not sufficient to eliminate the PT symptoms in this patient. The top of the wall of the JB protruding into the internal auditory canal within 2 mm was considered a high JB. Similarly, a high JB generates an unstable vortex that produces a characteristic whistle sound and leads to oscillations in pressure fields (12). The absence of adventitia in the JB amplifies unusual sounds, which are then transmitted to the inner ear through the temporal bone, cochlea, and other structures. A high JB also obstructs venous blood return in the inner ear, causing edema of the auditory hair cells. This leads to abnormal discharge of the hair cells and irreversible damage to cochlear function, resulting in symptoms such as tinnitus and hearing loss. On the one hand, the eddy currents formed by the high JB can damage the vessel walls of the JB and on the other hand, the venous outflow laterality may generate a force that acts on the high JB, making it susceptible to mural fracture (13). In this way, the sound and shock produced by the blood vortex are transmitted through the JBWD into the inner ear or mastoid cavity. In this case, high-resolution CT showed that the JB had invaded the structures of the inner ear and caused JBWD. We embolized the high JB with spring coils so that no more blood flow passed through it, which prevented the production of an abnormal blood sound, and also prevented further structural damage and hearing damage due to the constant impingement of blood flow on the vessel wall of the JB through the blocking effect of the spring coils.

It is difficult to distinguish between SSWA and JBWD in terms of clinical symptoms, both may present with abnormal mastoid noises, hearing loss, vertigo, etc. However, the two can be differentiated by imaging tests such as high-resolution CT, where SSWA may be demonstrated as irregular sigmoid sinus bone wall or adjacent cranial cortical changes or thinning or disappearance of the sigmoid sinus superficial cortical bone wall or mastoid bubbles with direct sigmoid sinus contact, and JBWD may be demonstrated by high JB invasion into the inner ear structures and an incomplete thin bony jugular plate between the JB and the middle ear cavity (14). The special feature of this case is the coexistence of SSS and JBWD (Figure 1) and the different results of the two interventional procedures confirm that both etiologies contributed to the development of PT in the patient at the same time.

Reconstruction of the sigmoid sinus wall or the wall of the JB through some biomaterials such as bone cement or autologous soft tissues such as temporal fascia may also be a better approach, but one may face extensive thrombosis and IIH, and the autologous soft tissues may not be able to withstand the pressure of the blood flow (15), reconstruction needs to be comprehensively addressed at every point of the dehiscence, and any omission may result in the need for a second reconstructive surgery, and in addition, reconstructive surgery is an open procedure, which may cause more damage to the surrounding normal tissues. Embolization of the sigmoid sinus via spring coils or gelatin sponges is also a method of relieving PT symptoms, but it may lead to increased pressure in the dural sinus causing IIH as well as papilledema, which affects vision (16). Placement of a stent to improve SSS is a safer method that avoids the continuous washout of blood against the vessel wall, improves the direction of abnormal blood flow, and reduces the vibration of the vessel, while the stent acts as a shield to reduce the conduction of abnormal sounds, and also avoids the development of IHH because there is no loss of drainage from the normal sigmoid sinus (17–19).

However, intravenous stenting is a complex procedure with a significant complication rate, including in-stent stenosis, hematoma, in-stent thrombosis, and bracket detachment (20). To prevent thrombotic complications, guidelines state that anticoagulation needs to be resumed as soon as possible after neurointervention, with dual antiplatelet therapy(DAPT) being the preferred choice, and that the same patients need lifelong oral anticoagulants to avoid stent-related thromboembolism (21). We believe that DAPT anticoagulation should be used for 12 months after venous stenting (in the absence of coagulation abnormalities), Clopidogrel 75 mg plus Aspirin 100 mg is recommended. Also lifelong anticoagulants such as Dabigatran. This does increase the burden on the patient. In-stent restenosis(ISR) is also a problem that needs to be faced, as stent implantation causes damage to the vessel wall, which in turn causes an inflammatory response that can contribute to ISR, as evidenced by the fact that 3 out of 133 patients with stenting of the IJV and cerebral venous sinus developed intraluminal restenosis at an average follow-up of 33 months (22), therefore in-stent restenosis needs to be taken very seriously and needs further long-term follow-up is needed.

6 Conclusion

In conclusion, this case study suggests that PT may be related to changes in blood flow and pressure due to SSS and the high JB. A dehiscence in the wall of the JB or the wall of the sigmoid sinus may be crucial for the development of PT. Endovascular therapy may be the safer way to treat PT. The prevention of post-stenting complications and the effects of stenting on patients' hemodynamics require sustained attention. Noninvasive imaging of the intracranial vasculature can help identify possible causes of PT and localize the target vessel. By improving the hemodynamics of the vessel, the diagnosis and treatment of PT might be performed. However, this patient still experiences unresolved symptoms of other forms of tinnitus, which require further investigation to determine the underlying cause and find a resolution.

Data availability statement

The original contributions presented in the study are included in the article/supplementary material, further inquiries can be directed to the corresponding author.

Ethics statement

Written informed consent was obtained from the individual(s), and minor(s)' legal guardian/next of kin, for the publication of any potentially identifiable images or data included in this article.

Author contributions

MX: Writing – original draft, Writing – review & editing. XD: Resources, Writing – review & editing. CZ: Resources, Writing – review & editing. TZ: Writing – review & editing. GW: Funding acquisition, Supervision, Writing – review & editing.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Endovascular treatment of epidural arteriovenous fistula associated with sacral arteriovenous malformation: case report

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Spinal epidural arteriovenous fistulas with sacral arteriovenous malformation (AVM) are a rare type of spinal arteriovenous fistulas. There are two varieties of spinal epidural arteriovenous fistulas (SEDAVFs), with type 1 involving intradural venous drainage and type 2 not involving intradural venous drainage. We present a case of transarterial embolization for type 1 SEDAVFs with sacral AVM. Within 8 months, a 14-year-old boy presented with progressively weaker lower extremities and bladder-bowel dysfunction. Magnetic resonance imaging (MRI) of the whole spine revealed thoracic spinal cord congestion, a single dilated flow void running from the lumbosacral area to the conus medullaris, and continuing cranial draining up to the C5 level via the perimedullary vein. Filling of the venous sac through a preferential feeder after embolizing the AVM nidus was performed. After 3 months, the clinical follow-up showed improvement of motoric function, although mild. Endovascular treatment for SEDAVF type 1 might have achieved total obliteration without any procedural complications. Nevertheless, it can be very challenging due to multiple feeders and the presence of an AVM nidus like in this case. However, the most difficult thing in fistula cases is establishing the diagnosis and finding the fistula point. Early treatment is required, due to the fact that longstanding lesions could cause irreversible damage.

KEYWORDS

arteriovenous fistula, arteriovenous malformation, embolization, endovascular, epidural, spinal

Introduction

Spinal vascular malformations are classified in a variety of ways based on their anatomical location and characteristics. The two most prevalent kinds of spinal vascular malformations are spinal dural arteriovenous fistulas (SDAVFs) and spinal epidural arteriovenous fistulas (SEDAVFs). It is important to distinguish between dural and epidural fistula based on their clinical presentations and preferred approach to therapy, as SEDAVFs are poorly understood and may be mistaken for SDAVFs, according to the literature. The arteriovenous shunt in SEDAVFs is positioned in the epidural space and fills it, whereas the arteriovenous shunt in SDAVFs is located inside the dural sheath of the nerve root and drains directly into an intradural vein without filling the epidural space (1). However, because of the limited number of cases, understanding of the etiology, demography, pathophysiology, and treatment approach for these problems are also limited (2).

Modification of the previous classification of arteriovenous malformations was done by Kim and Spetzler (3) in 2002 and later in 2006. In 2011, Rangel-Castilla categorized extradural AVFs into three types: A, B1, and B2, which may or may not have intradural venous drainage and may or may not have neurological impairments (Table 1) (4). The shunted epidural venous pouch in SEDAVFs can be found ventrally, laterally, or dorsally. To distinguish such venous pouch locations, the interpedicle line was used as a bone structural reference. In ventral and dorsal SEDAVFs, the shunted pouch is located medial to the medial interpedicle line and on the ventral side and dorsal side, respectively, of the spinal canal in the lateral view (1). The shunted pouch in lateral SEDAVFs is placed lateral to the medial interpedicle line on the anterior view.

SEDAVF symptoms at first are frequently vague. Along with symmetrical or asymmetrical sensory symptoms including paresthesia in one or both feet, diffuse or patchy sensory loss can occur. SEDAVFs can present with symptoms secondary to compressive symptoms, or can present secondary to congestive myelopathy and can manifest with both symptoms. Radicular pain can be developed secondary to compressive symptoms. Although micturition and defecation disturbances can happen early in the disease, both often appear in the late stage. Therefore, as a marker for the location of the venous pouch, the pouch's position may also be connected to the symptoms. Vascular engorgement causes a significant mass effect on the nerve root in the ventral or lateral side, which can manifest as benign symptoms such as radiculopathy (5, 6). Nevertheless, in contrast to when the venous pouch position dorsally located, the author believes that compressive symptoms are less frequently presented due to the position being farther from the nerve root.

Lumbosacral fistulas are frequently associated with a fistula in the ventral epidural region that drains into epidural veins as well as a filum terminale vein (7). As a result, when a fistula develops and drains into the intradural venous drainage, it raises the medullary venous pressure by allowing arterial blood to reflux into the intradural veins, causing progressive venous congested myelopathy (VCM) or both symptoms (8, 9). However, the mechanisms of myelopathy can be due to venous hypertension, mechanical compression, and vascular steal effects (2, 10); the elimination of these AVFs is the goal of treatment (11).

The key to treatment in spinal AVFs is the level location and understanding of the angioarchitecture. AV shunts, such as common spinal DAVFs, that are situated in the sacrum or filum terminale can be treated by surgery, an endovascular approach, or both. The chosen course of treatment has been surgery, which has a higher complete obliteration rate (12). Regardless of whether the shunt is dural or epidural, the dilated filum terminale vein, which connects with subarachnoid veins around the cord, can be a sign of where the fistulous point is located in the lumbosacral region (7, 13–15). Nonetheless, due to variations in the angioarchitecture of the shunts, treatment needs to be individualized. A multidisciplinary approach is crucial in the treatment of complex neurovascular diseases, where the combination of endovascular therapy and surgery plays an increasingly significant role.

Although research on sacral EDAVFs is scarce, it is known that the lateral sacral artery (LSA), middle sacral artery, or iliolumbar artery (ILA) are typically responsible for supplying sacral AVFs (16). However, the research mainly consists of case reports, particularly when there is an association with other abnormalities, such as AVM, as in this case, where the fistula point does not go directly to the shunt point but rather feeds the AVM from multiple feeders before draining into epidural veins and the filum terminale vein. Identification and treatment of these lesions are difficult due to the association of abnormalities and the intricate angioarchitecture of these lesions. We were unable to find any case reports with the same abnormalities, to the best of our knowledge. As a result, we describe a case of sacral EDAVFs connected to a sacral AVM that was fed by multiple levels of the LSA and the sacral branch of the ILA.

TABLE 1 Classification of spinal AV shunts reported by Kim and Spetzler (3) and Rangel-Castilla et al. (4).

Kim and Spetzler (3)	Rangel-Castilla et al. (4)
AVF types:	Extradural AVF
Extradural Intradural Dorsal A: Single arterial feeder B: Multiple arterial feeders Ventral A: Small B: Medium C: Large	A: With intradural venous drainage B1: Without intradural venous drainage B2: Without intradural venous drainage, without neurological deficits
AVM types:	
Extradural-Intradural Intradural Intramedullary Intramedullary-extramedullary Conus medullaris	

AVF, arteriovenous fistula; AVM, arteriovenous malformation.

Case presentation

A 14-year-old boy experienced left radicular pain that started developing 3 months earlier along with progressive bilateral foot and leg weakness for 8 months. Reviewing his medical history revealed no significant trauma or systemic illness. During a neurological examination, it was noted that the strength of the left and right leg muscles below the knee, dorsi, and plantar flexion had decreased (grade 1) and that there was still slight movement in the iliopsoas, hamstring, and quadricep muscles (grade 2). Pathological reflexes were absent. Spasticity of the lower extremities and muscle wasting at the left and right gastrocnemius were noted. Sensory disturbances below thoracal 12 and decreased pinprick and light touch sensation were detected in the left foot more than the right side. In the last 3 months, his symptoms worsened, he lost control over his bladder and bowel, and became bedridden. At the time of admission, the patient's Aminoff-Logue disability score was G5M3B1, and intermittent urinary catheterization was carried out.

Radiological findings

A contrast-enhanced MRI of the whole spine showed a single dilated flow void extending from the lumbosacral region to the conus medullaris and continuing cranial drainage up to C5 level through the perimedullary vein, as well as diffuse abnormal central-cord T2-weighted image hyperintensity that suggested spinal cord congestion extending from the level of the thoracic region (Figure 1A). Contrast-enhanced MR angiography (CE-MRA) showed perimedullary vein engorgement drainage, cranially from the filum terminale until the cervical level (Figure 1B). The Spinal AVM was impressed in the left sacral region, with enlargement of the venous pouch before it entered the intradural region (Figure 1C). The filum terminale vein must be used for any sacral dural AV shunt draining in the direction of the spinal cord. Therefore, it makes sense to suggest that the presence of an engorged and tortuous intradural filum terminale vein may be a sign of a sacral AV fistula.

A complete diagnostic spinal angiogram, including the iliolumbal artery (ILA), lateral sacral arteries (LSA), and medial sacral artery (MSA) was performed to find the fistula point. From the angiogram, a venous pouch was seen in the left L5-S1 region that was associated with sacral AVM that was fed by multiple arteries with early venous drainage into the filum terminale and drainage to the perimedullary vein cranially, consistent with an SEDAVF type 1 (Figure 1D). After multidisciplinary team meetings and discussion with the patient, transarterial embolization was performed.

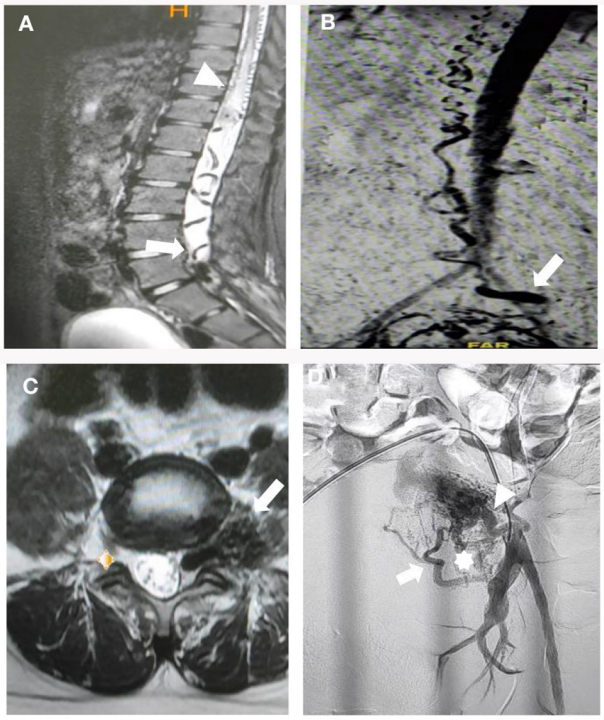

FIGURE 1
Preoperative multimodal neuroimaging evaluation. **(A)** Diffuse abnormal central-cord T2-weighted image hyperintensity that suggested spinal cord congestion extending from the level of the thoracic region, seen on magnetic resonance imaging (MRI) with contrast, along with a single dilated flow void that ran from the lumbosacral area to the conus medullaris (arrow). **(B)** Contrast-enhanced MR angiography (CE-MRA) showed a venous pouch at the lumbosacral region, with drainage cranially from the filum terminale until the cervical level (arrow). **(C)** Sacral arteriovenous malformation (AVM) was impressed in the left sacral region, with enlargement of the venous pouch before it enters the intradural region (arrow). **(D)** Oblique projection of angiography from the left internal iliac artery demonstrating that a venous pouch was seen at the left L5-S1 region that was associated with sacral AVM that was fed by hypertrophied lateral sacral arteries (LSA) S1 (asterisk), S2 (arrow), and the sacral branch of the iliolumbal artery (ILA) (arrow head).

Endovascular treatment

The procedure was carried out under general anesthesia, and the left internal iliac artery was selectively catheterized with a 5-Fr vertebral diagnostic catheter (Terumo, Tokyo, Japan). Each feeder of the AVM from the LSA and ILA was selected using a headway 17 microcatheter (microvention, Aliso Viejo, CA) and a Marathon microcatheter (ev3 Inc.) under fluoroscopic and roadmap guidance. Detachable hypersoft coils (microvention, Aliso Viejo, CA) were placed in additional feeders to reduce collateral inflow from having better penetration and to prevent the liquid materials from refluxing to unwanted locations. The sandwich technique was carried out when onyx 18 (ec3, Irvine, Calif) was injected using a double lumen ballon scepter C microcatheter (microvention, Aliso Viejo, CA). Finally, a guide catheter dextrose 5% push technique was used to inject N-butyl-2-cyanoacrylate-NBCA (Histoacryl; B. Braun, Melsungen, Germany) mixed with Lipiodol (Guerbet, Roissy, France) in a 1:3 ratio through the remaining small, tortuous arterial feeders until the venous pouch and draining vein were filled.

Discussion

The concomitant presence of multiple spinal vascular diseases in a single patient is very rare. We report a case of sacral EDAVFs supplied by the multiple segments of the LSA and ILA that are idiopathic. Surprisingly, the shunt is not directly toward the drainage vein, which we believe is the filum terminale; however, all the feeders form a nidus that is consistent with the features of AVM in the sacral region. To the best of our knowledge, we could not find any similar cases that have been reported to date in the literature with the same angioarchitecture pattern. Since the feeding artery did not provide a direct shunt to the vein in our case, it cannot be accepted that the fistula-induced myelopathy is caused by a steal phenomenon from the arteries to the fistula. Instead, it is believed that arterialization of the veins increases the venous pressure in the perimedullary venous system, and that the decreased intramedullary arteriovenous pressure gradient causes hypoxic myelopathy.

SEDAVFs have been described as ventral epidural shunts or ventral epidural AVFs in the classification system for spinal arteriovenous shunt diseases (1, 9). The absence of a horizontal T-sign, which is a typical sign of SDAVFs, is not present in SEDAVFs. In addition, SEDAVFs more frequently occur at lumbar spinal levels

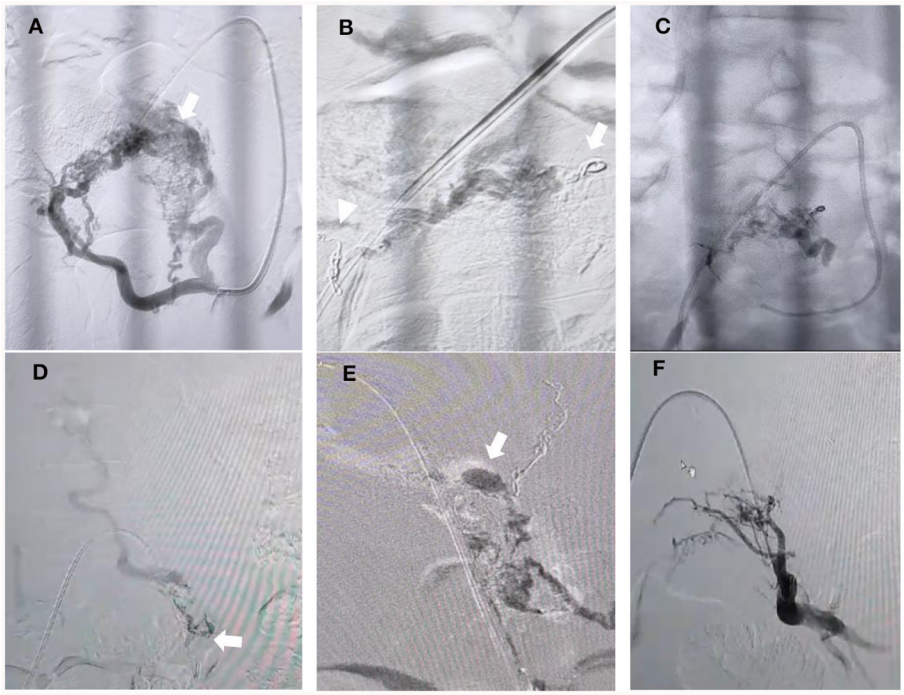

FIGURE 2
Embolization process. **(A)** Oblique projection of selective angiography from left lateral sacral arteries (LSA) showing that hypertrophied arteries of s2 and s1 fed the nidus AVM, which converged to the shunted pouch in the left ventro-lateral epidural side (arrow). **(B)** Coils in the lumbal branch of the ILA (arrow) and S2 branch of the LSA were performed (arrowhead). **(C)** Final cast after "sandwich technique" Onyx injection was performed over 20 minutes from the main feeder from the S1 branch of the LSA but could not penetrate more distal to the venous pouch. **(D)** Microcatheter injection was performed from the left remaining branch of the LSA but could not advance more distal close to the fistulous side due to the very small and tortuous branch (arrow). **(E)** N-butyl-2-cyanoacrylate (NBCA) injection using the guide catheter dextrose push technique was performed until there was filling of the venous pouch and the draining vein (arrow). **(F)** Final angiography from the internal iliac artery showing complete obliteration of the AVM and fistula.

than SDAVFs and are typically located in the ventral epidural space with multiple feeders. SEDAVFs are brought on by the epidural arterial branches of the ascending cervical, vertebral, intercostal, lumbar, or sacroiliac arteries directly arterializing the epidural venous plexus (1). Sacral EDAVFs are typically located in the ventral epidural space, with the arterialized vein in the root sleeve being distant from the fistula (7).

Contrast-enhanced MR angiography (CE-MRA) provides adequate visualization of the perimedullary and lumbar draining veins and facilitates subsequent digital subtraction angiography (DSA) examinations in the vast majority of cases. The filum terminale is frequently dilated in suspicious lumbosacral region fistulas, which is a helpful sign. However, for accurate fistula localization, DSA remains the gold standard diagnostic tool (14). In our case, the left ventro-lateral epidural side's shunted pouch was fed by a number of feeders from the left sacral branch of the ILA and LSA at the S2 and S1 levels (Figure 2A). The AVFs drained retrogradely through the epidural venous pouch into the filum terminale and drained to the perimedullary vein cranially, consistent with sacral EDAVFs.

Although it has been reported that epidural exposure at the actual fistula site is unnecessary and residual epidural arteriovenous shunting appears to be harmless because it does not produce venous hypertension, a surgical approach to the arterialized venous pouch is risky because the venous pouch is usually located at the ventral epidural space and may require destabilization of vertebral structures to be able to safely manage the malformation (5, 7). So far, the optimal management strategy of SDAVFs has remained unclear (17), especially in our case, due to the fistula associated with the AVM nidus. After multidisciplinary meetings, endovascular treatment was performed, and surgery will be performed if the endovascular approach fails to close the fistula.

The recurrence may have happened when an endovascular approach was chosen, even after the liquid embolic agents partially reached the draining vein. In order to prevent the fistula from recanalizing as a result of the recruitment of more arterial afferents, endovascular treatment of sacral EDAVFs with intradural venous drainage focuses on occluding the epidural venous pouch and the proximal intradural draining vein. For cases with a large epidural venous pouch fed by multiple feeders, complete filling of the embolic materials in the entire venous pouch and draining veins is frequently difficult (6, 18). Nevertheless, complete obliteration can be achieved by a surgical approach; however, concomitant lesions can be challenging (19). During surgery, likely spinal AVM in the common regions requires the inverse approach—the draining vein has to be preserved as much as possible until most or all of the arterial feeders to the nidus are obliterated to prevent rupture (17).

Selecting the optimal liquid embolic agents is crucial, each with its own advantages and disadvantages if the endovascular approach is selected. In earlier reports, particles have the advantages of

embolization, particularly prior to operation; however, the agents have a high failure rate over the long term (20). In treating SDAVFs in their study population, Larsen et al. (21) found that liquid embolization using N-butyl-cyanoacrylate (n-BCA) or glue was just as successful as surgical ligation. In fact, when the n-BCA penetrates into the venous side of the nidus, 85% of the fistula is permanently occluded (22). However, when there are multiple artery supplies, it may be challenging to penetrate the venous end with liquid embolic agents, especially if glue is used as it hardens when it comes into contact with blood. Other liquid materials that have been used with success include onyx, squid, and PHIL. Given that each liquid embolic material has its own set of limitations, operator familiarity is very crucial when selecting it. Penetration of the fistula and reflux are still the main challenges with onyx and squid, but forward flow is easier to attain with PHIL. However, PHIL has less radio-opacity than other agents because it is an iodinated contrast-based embolic material (23).

The preferential flow, plug and push, and filling the venous sac techniques have all been mentioned as effective ways to embolize SEDAVFs (24). Nevertheless, when the fistula has a nidus and multiple feeders, it is difficult to know which feeder has dominant flow directed to the pouch due to competing flow from each. Before performing embolization, we placed some coils in the lumbar branch of the ILA and one of the branches of the LSA (S2 level) to decrease the competing flow so as to reduce reflux to an unwanted location and penetrate the feeders that drain dominantly to the venous pouch (Figure 2B). Unfortunately, after embolization from the main trunk of the LSA was performed, the onyx could not penetrate more distal to the venous pouch (Figure 2C). We identified the small branch from the left LSA that fed the fistula, which became more obvious than before and could not be seen at the beginning due to the competing flow. The microcatheter was advanced to the branch but could not advance more distally close to the fistulous side due to the very small and tortuous branch (Figure 2D). Therefore, we decided to inject glue from a microcatheter using the guide catheter dextrose push technique during glue injection to avoid early polymerization and improve distal penetration to the draining vein and venous pouch (Figure 2E). A control angiogram from the internal iliac artery showed that there was no longer an AVM, and fistula drainage was seen (Figure 2F).

Regarding clinical outcomes, after 3 months, improvements in sensoric and motoric strength were noted (grade 2), with an Aminoff-Logue disability score of G4M2B1. The reversibility of the spinal lesions caused by spinal venous congestion myelopathy may be related to complete recovery. When preoperative deficits are mild, clinical outcomes are favorable, particularly in terms of motor and sensory function. Early treatment is therefore necessary because persistent lesions may result in spinal cord damage that is irreversible. However, the limitation of this study is that only one case has been reported, and physicians need to understand the pathophysiology, angiographic, and clinical aspects of the disease in order to determine the optimal treatment for the patient. More similar cases are needed.

Conclusion

Sacral EDAVFs are uncommon and present a number of diagnostic and treatment challenges. They may have bilateral arterial supplies from the LSA and frequently have multiple feeders, which may affect the results of embolization with liquid embolic agents. Successful management of these lesions necessitates a thorough understanding of the region's variable arterial supply patterns. Misdiagnosis and diagnostic delays are common. In selected patients, prompt diagnosis and treatment are likely to result in better neurological outcomes. To ensure prompt therapy, patients with lower thoracic myelopathy of unknown etiology should have sacral EDAVFs considered in the differential diagnosis. The symptoms can be characterized by the particular anatomy of sacral intradural venous drainage through the filum terminale vein caudally, resulting in venous hypertension in the spinal cord. CE-MRA may be a very useful diagnostic imaging tool to assist in locating the fistula point, especially when a dilated filum terminale is present.

Data availability statement

The original contributions presented in the study are included in the article/supplementary material, further inquiries can be directed to the corresponding author.

Ethics statement

Written informed consent was obtained from the individual(s), and minor(s)' legal guardian/next of kin, for the publication of any potentially identifiable images or data included in this article.

Author contributions

AA: Conceptualization, Data curation, Formal analysis, Funding acquisition, Investigation, Resources, Visualization, Writing – original draft, Writing – review & editing.

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Conflict of interest

The author declares that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Dural arteriovenous fistula and sinus thrombosis presenting as parkinsonism and dementia: a case report with literature review

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Introduction: Dural arteriovenous fistula (DAVF) is an uncommon malformation involving an abnormal connection between dural arteries, or the pachymeningeal branches of cerebral arteries, and dural veins. Its exact pathogenesis remains elusive. Known potential triggers for DAVF include cerebral venous sinus thrombosis (CVST), trauma, ear infections, and cranial surgeries. Due to its rarity and diverse clinical presentations, diagnosing DAVF can be a challenge.

Case description: We present a case of DAVF associated with CVST, manifesting as rapidly advancing parkinsonism accompanied by dementia over a month. Brain magnetic resonance imaging (MRI) revealed bilateral symmetric T2 hyperintensities in the basal ganglia and brain stem. Cerebral angiography further confirmed a fistula between the torcular herophili and the transverse-sigmoid sinuses. Despite strong recommendations for transvenous embolization of the fistula, the patient declined the procedure. The anticoagulant therapy and symptomatic treatments administered did not yield any improvement in the patient's condition. Additionally, we reviewed 27 DAVF-derived parkinsonism and dementia cases.

Conclusion: DAVF must be considered in the differential diagnosis of cases of rapidly progressive parkinsonism with concurrent dementia. Given its potential for treatment and reversibility, timely diagnosis and intervention for DAVF are paramount.

KEYWORDS

DAVF, parkinsonism, dementia, CVST, Wernicke's encephalopathy

1 Introduction

Dural arteriovenous fistula (DAVF) is a rare vascular anomaly characterized by an abnormal connection between dural arteries, or pachymeningeal branches of cerebral arteries, and dural veins. Typically manifesting later in life, DAVF can be triggered by factors such as cerebral venous sinus thrombosis (CVST), trauma, ear infections, and cranial surgeries (1). It is believed that these factors can lead to venous thrombosis. DAVF patients may exhibit a range of symptoms, from pulsatile tinnitus and ophthalmoplegia to acute confusion, cognitive impairment, parkinsonism, and specific neurological deficits. The clinical and radiographic presentations are largely determined by the pattern of venous drainage. The primary treatment approach is endovascular

embolization, but surgical intervention and stereotactic radiosurgery serve as viable alternatives (1). Given that DAVF is treatable and many symptoms are reversible, early diagnosis and intervention can significantly enhance a patient's prognosis and reduce long-term disabilities. In this context, we present a unique case of a patient with rapidly progressing parkinsonism and dementia attributable to DAVF who was misdiagnosed as having Wernicke's encephalopathy.

1.1 Case description

A 59-year-old man came in with worsening and consistent gait difficulties and signs of dementia over the past month. He had not experienced any headaches, double vision, nausea, vomiting, or delirium. His medical history was mostly unremarkable, except for a head injury approximately 1 year ago. Additionally, he had a long history of alcohol consumption. He was initially evaluated at a local hospital, where brain magnetic resonance imaging (MRI) revealed bilateral symmetrical lesions in the brain stem and basal ganglia. Due to the patient's prolonged history of alcohol abuse and progressive cognitive decline, Wernicke's encephalopathy was initially suspected at the previous hospital. The patient received high-dose thiamin (300 mg/day, administered via intramuscular injection) for 2 weeks. However, his condition continued to deteriorate, leading to his referral to our neurology department.

Upon examination, he took short steps, showed symmetrical rigidity in all four limbs, general slowness of movement, with mild increased muscle tone and normal strength, hyperreflexia, diminished speech, a soft voice, and cognitive decline without lamination (scoring 18/30 on the Mini-Mental State Examination, especially in attention, calculation, and execution ability). He also exhibited urinary incontinence, and a vascular murmur was audible behind his ears. Laboratory tests, including those for blood count, infections, coagulation function, protein C and S levels, lupus anticoagulant, autoimmune markers (anti-neuronal antibodies, anti-nuclear antibodies, antibodies to extractable nuclear antigen, anti-neutrophil cytoplasmic antibody, anti-cardiolipin antibodies, ds-DNA antibodies, and autoimmune encephalitis antibodies), tumor markers, thromboelastogram, anti-aquaporin 4 antibody, anti-myelin oligodendrocyte glycoprotein antibody, anti-glial fibrillary acidic protein antibody, oligoclonal bands, and metabolic screens, all returned normal results. His blood D-dimer level was measured at 0.73 μg/mL (0–0.55 μg/mL). A lumbar puncture showed that the cerebrospinal fluid pressure exceeded 400mmH2O. Analysis of this fluid indicated normal white cell counts, glucose, and chloride levels, with a slightly elevated protein level of 592.3 mg/L. There were no signs of infection, inflammation, or bleeding.

An MRI of the brain revealed bilateral symmetrical lesions in areas such as the pontine, pontine arms, and basal ganglia. These lesions showed restricted diffusion on diffusion-weighted imaging and no enhancement (as seen in Figures 1A–F). Additionally, magnetic

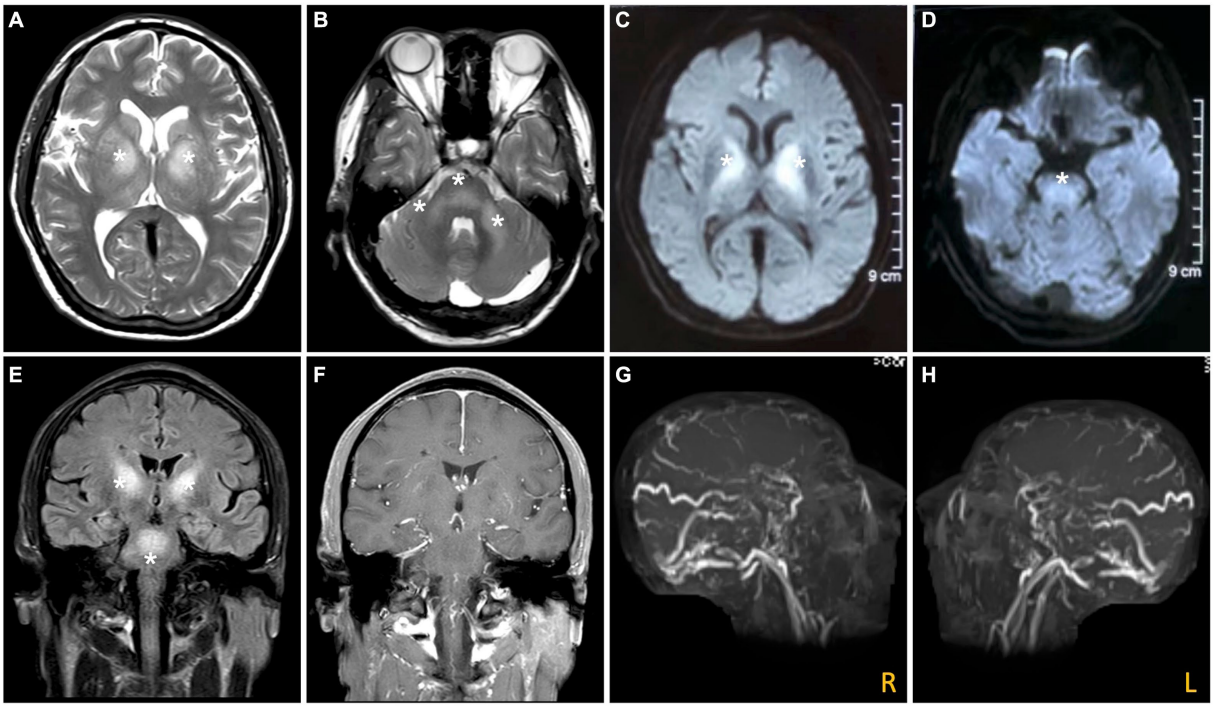

FIGURE 1
Brain MRI and MRV. The axial section of the T2-weighted image **(A,B)** and the diffusion-weighted image **(C,D)** reveal bilateral symmetrical hyperintensities in the basal ganglia and brain stem (white asterisk). The lesions present with hyperintensity on fluid attenuated inversion recovery (FLAIR) images **(E)**, and no enhancement on enhancing MRI **(F)**. The MRV **(G,H)** indicates a filling defect in the right transverse, left sigmoid, and superior sagittal sinuses.

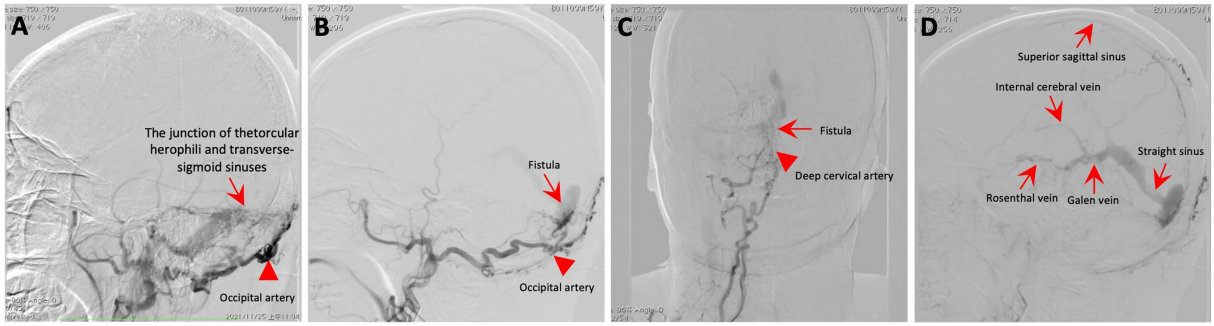

FIGURE 2
Cerebral angiography. **(A–C)** The angiogram showcases a fistula (red arrow) connecting the torcular herophili to the transverse-sigmoid sinuses, with blood supply from the occipital artery (red arrowhead) and the deep cervical artery (red arrowhead). **(D)** The DAVF drained into the straight sinus, which flowed in a retrograde direction into the vein of Galen, as well as into the internal cerebral veins and basal vein of Rosenthal.

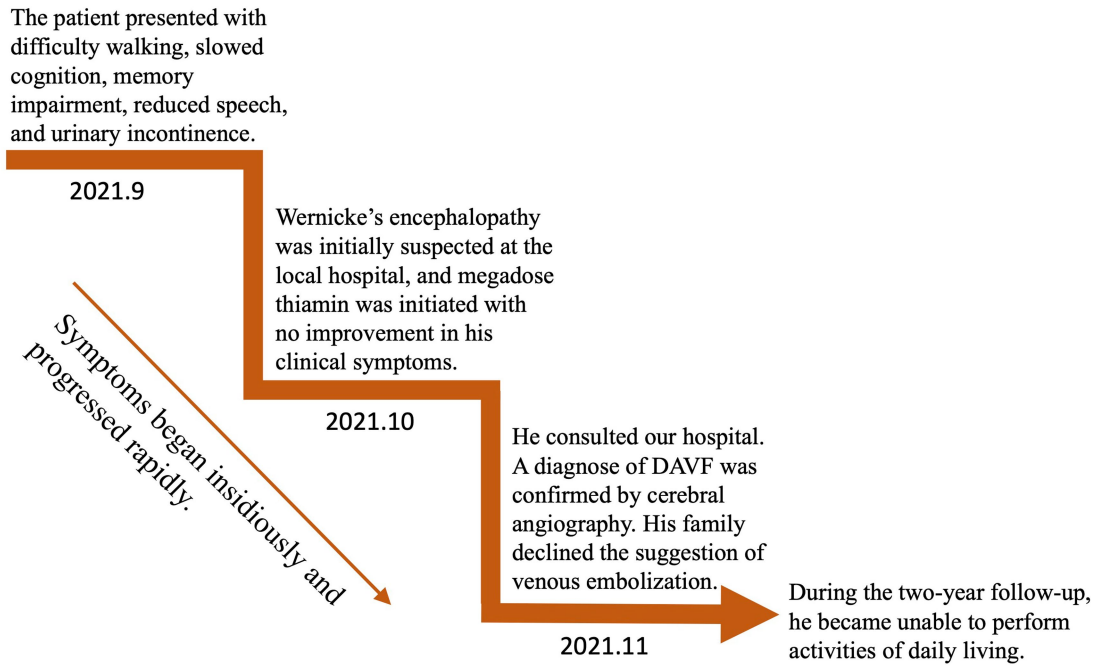

FIGURE 3
Time course of symptoms, diagnosis, and treatment.

resonance venography (MRV) highlighted dilated veins and a filling defect, pointing to possible sinus thrombosis in the superior sagittal sinus (as seen in Figures 1G,H). Cerebral angiography further confirmed thrombosis in the vein of Galen and identified a DAVF between the torcular herophili and transverse-sigmoid sinuses, fed by the occipital artery and arteria cervicalis profunda (as shown in Figure 2). The DAVF drained into the straight sinus, which flowed in a retrograde direction into the vein of Galen, as well as into the internal cerebral veins and basal vein of Rosenthal. Given that DAVFs are treatable and their symptoms can be reversed, it was strongly recommended that the patient undergo transvenous embolization of the fistula. However, the son of the patient declined for personal reasons. Even with subcutaneous injections of low molecular weight heparin (5000AXaIU) every 12 h and symptomatic treatments, including levodopa, there was no improvement in his clinical symptoms. The time course of symptoms, diagnosis, and treatment is shown in Figure 3.

2 Discussion

DAVF refers to abnormal shunts between dural arteries and structures such as dural venous sinuses, meningeal veins, or cortical veins. The exact cause of DAVF remains elusive. While

many cases appear without any known cause (idiopathic), there are reports suggesting links to CVST, trauma, ear infections, venous hypertension, and prior cranial surgeries. The co-occurrence of DAVF with CVST is relatively rare (2, 3). In our study, we document a case where DAVF is seen alongside CVST. DAVF leads to retrograde blood flow into the straight sinus and the vein of Galen. Thrombosis in the straight sinus and the vein of Galen further exacerbates venous pressure, causing reflux obstruction and insufficient perfusion, subsequently leading to infarction in the corresponding drainage area of brain tissue. Actually, the exact relationship between CVST and DAVF is not well-defined, leading to debates about whether DAVF precipitates CVST or emerges as a consequence of it. Sinus thrombosis can increase venous pressure, which might trigger the development of DAVF by activating dural arteriovenous pathways. This potentially can lead to ischemia because of backward venous flow and blood stagnation. Additionally, the turbulent blood flow observed in DAVF might contribute to thrombus formation (3).

Clinical manifestations linked to DAVF vary widely and are influenced by the pattern of venous drainage. These can range from pulsatile tinnitus and ophthalmoplegia to acute confusion, rapidly advancing dementia, parkinsonism, seizures, cerebellar symptoms, and specific neurological deficits. Notably, DAVF patients with cortical venous drainage and venous ectasia are at a heightened risk of hemorrhage (4). Instances of DAVF patients exhibiting parkinsonism, with or without cognitive dysfunction, are infrequent. Research indicates that parkinsonism in DAVF patients tends to be more prevalent in older male patients, averaging approximately 63 years of age, with cases spanning from 40 to 81 years. The time from initial symptoms to DAVF diagnosis has been documented to range between 1 week and 3 years (5). In this study, we reviewed 27 DAVF patients who have parkinsonism with cognitive dysfunction (Table 1). Of these patients, 18 were male, with an average age of 65 years, ranging from 40 to 87 years old. While the precise mechanism remains unclear, the prevailing theory is that reduced blood flow in the frontal lobes and a perfusion defect in the basal ganglia may be contributing factors to the onset of DAVF-related parkinsonism (5, 7). There is growing evidence suggesting a specific vascular mechanism whereby thalamic damage leads to subacute encephalopathy or swiftly progressing dementia (25). In the case we observed, cognitive impairments included slowed thinking, difficulties with calculations and concentration, memory loss, executive dysfunction, and diminished speech, suggesting dysfunction in the basal ganglia or frontal lobe.

Given the patient's rapidly advancing parkinsonism and dementia, combined with a long history of alcohol abuse and bilateral symmetrical lesions in the brain stem and basal ganglia as revealed by MRI, various conditions such as Wernicke's encephalopathy, central pontine myelinolysis, extrapontine myelinolysis, hepatic encephalopathy, Creutzfeldt–Jakob disease, and glioma must be considered (3). Extensive diagnostic efforts were made to rule out these alternatives (as seen in Figure 4). Vitamin B1 levels were not checked because the patient had already been receiving high doses of thiamine for several days. Wernicke's encephalopathy is more likely to involve the medial thalamus, hypothalamus, periaqueductal gray matter, and the areas surrounding the third and fourth ventricles on MRI. However, the patient does not present with oculomotor paralysis or psychiatric symptoms, and high-dose thiamine treatment has been ineffective, which does not support the diagnosis of Wernicke's encephalopathy. Extrapontine myelinolysis is more likely to affect the bilateral striatum, and clinical symptoms often outweigh the radiological changes. Additionally, there is no history of electrolyte disturbances in the patient. Non-invasive vascular imaging techniques, including CT and MRI, could offer clues about the presence of DAVF. In this case, retrograde venous drainage can lead to increased venous pressure, resulting in early vascular-origin edema, which may progress to venous infarction characterized by cytotoxic edema. The MRI lesions primarily concentrate in the basal ganglia region, with relatively preserved midbrain areas, correlating with the susceptibility of deep brain tissues to restricted outflow. Moreover, we acknowledge that, in our case, it cannot be conclusively determined whether the involvement of the deep pontine is caused by the known venous thrombus. Cerebral angiography assists in identifying the arterial feeders and venous outflow of the fistula. The Cognard classification system, commonly utilized to categorize DAVF, separates it into five types based on venous drainage patterns (26). In this particular case, according to the angiograph, the DAVF drains into the straight sinus, with filling of the Galen, internal cerebral, and Rosenthal veins, without cortical venous drainage, unveiling a Cognard type IIa DAVF. However, in this classification, there is no detailed subtyping for reflux into deep veins. In future classifications, refinement based on deep venous drainage may have a more positive significance for prognosis assessment and surgical selection.

In the management of non-Parkinson's disease (PD) patients with parkinsonism, the treatment involves removing precipitating factors, managing the underlying primary disease, and additionally administering medications used in the treatment of PD, such as amantadine, levodopa, dopamine receptor agonists, monoamine oxidase-B inhibitors, and catechol-O-methyl transferase inhibitors, among others. However, these medications may not provide significant improvement in symptoms. Indeed, damage due to edema or ischemia could be reversible. Early intervention through endovascular embolization, aiming for complete obliteration of the fistula, is highly recommended for DAVF patients. Prior research has shown that the vast majority of DAVF patients experienced significant symptom alleviation, with some even achieving total symptom resolution after undergoing endovascular embolization treatments (27). Moreover, clinical improvement correlates with radiographic improvement. Venous sinus thrombosis may further increase venous pressure. Therefore, in patients without surgery, anticoagulant therapy may potentially reduce venous pressure and thus exert a protective effect. However, the role of anticoagulation in untreated DAVF combined with thrombosis remains unclear. In our case, the patient's family opted against transvenous embolization of the fistula. Consequently, despite receiving anticoagulant therapy and symptomatic treatments, the patient showed no clinical improvement.

We recognize that our case report has certain limitations. First, despite conducting cerebral angiography, we cannot definitively confirm a causal relationship, as the patient did not undergo treatment for the malformation, followed by recovery.

TABLE 1 Cases with DAVF presenting with progressive dementia and parkinsonism.

No.	Author (Year)	Age/ gender	Neurologic signs (except dementia and parkinsonism)	CT/MR imaging change	Angiography location/drainage	Treatment	Outcome
1	Mastsuda S. et al. (6)	55/M	Headache	White matter, left thalamus	Right sigmoid sinus/ superior sagittal sinus, straight sinus	TAE	Improved
2	Mastsuda S. et al. (6)	78/M	Blurred vision	White matter, subarachnoid space	Right sigmoid sinus/ superior sagittal sinus, straight sinus, frontal cortical veins	TAE	Improved
3	Mastsuda S. et al. (6)	69/F	Tinnitus, headache	White matter, subarachnoid space	Left sigmoid sinus/ transverse sinus, superior sagittal sinus, straight sinus, cortical veins	TAE	Unimproved
4	Lee PH. et al. (7)	60/F	Normal	White matter	Left transverse-sigmoid sinus/superior sagittal sinus, cortical venous	TAE	Improved
5	Chan HY. et al. (8)	77/M	Double incontinence	Left temporal and occipital lobes	Transverse-sigmoid sinus/ vein of Labbe and other cortical veins	TAE	Improved
6	Kajitsni M. et al. (9)	75/M	Transient loss of consciousness after slow deep breathing	Frontal lobes and basal ganglia	Transverse-sigmoid sinus/ superior sagittal sinus	TAE	Improved
7	Miura S. et al. (10)	65/M	Diplopia, tinnitus, ataxia	Basal ganglia, white matter	Left transverse-sigmoid sinus/superior sagittal sinus, straight sinus	TAE	Improved
8	Norgueira RG. et al. (11)	79/M	Tinnitus, hearing loss, vertigo	Serpiginous flow voids on the right mesial temporal lobe	Left transverse sinus/ superior sagittal vein, straight sinus, basal vein of Rosenthal	TAE, surgery, and TVE	Improved
9	Netravathi M. et al. (12)	54/M	Headache	Thalamus, globus pallidus, basal ganglia, white matter	Torcula/straight sinus	TAE	Minimal change
10	Netravathi M. et al. (12)	40/M	Urinary incontinence	Gray and white matter	Superior sagittal sinus/ retrograde sinus flow and cortical venous reflux	Unsuccessful TAE and TVE	Deteriorated
11	Shahar T. et al. (13)	59/M	Limitation in right and up-gaze, vertical nystagmus on up-gaze	Right occipital lobe, lenticular nuclei	Straight sinus/vein of Galen, vein of Rosenthal, internal cerebral veins	TAE	Improved
12	Hattori T. et al. (14)	52/F	General fatigue, urinary incontinence, emesis, tinnitus	Whiter matter, basal ganglia	Transverse-sigmoid sinus/ superior sagittal sinus, straight sinus	TVE	Improved
13	Fujii H. et al. (5)	69/M	Normal	Frontal lobes, basal ganglia	Superior sagittal sinus/ND	TVE	Improved
14	Jagtap SA. et al. (15)	73/F	Myoclonic jerks	Transverse sigmoid junction	bilateral transverse sinus-sigmoid sinus junction/ND	No	Died
15	Luo Y. et al. (16)	54/M	Urinary incontinence	Flow void clusters at the inner part of the left temporal lobe	Right transverse-sigmoid sinus/ straight sinus, the right transverse sinus	No treatment	No change

(Continued)

TABLE 1 (Continued)

No.	Author (Year)	Age/ gender	Neurologic signs (except dementia and parkinsonism)	CT/MR imaging change	Angiography location/drainage	Treatment	Outcome
16	Luo Y. et al. (16)	75/M	Headache	Dilated vein on the left temporal cortex	Left transverse-sigmoid sinus/straight sinus, left temporal cortical veins	No treatment	Died
17	Ma C. et al. (17)	62/M	Weakness, apathy, urinary incontinence	Normal	Left temporal region/ superior sagittal sinus	TAE	Improved
18	Enofe I. et al. (18)	82/F	Seizures	White matter	Transverse sinus/torcular Herophili, superior sagittal sinus, and transverse-sigmoid sinuses	TAE	Improved
19	Lai J. et al. (19)	62/M	Headache, apathy, disorientation, visual hallucinations, generalized status epilepticus	White matter	Right transverse, sigmoid sinus, torcular sinuses, superior sagittal sinus/ straight sinus, vein of Galen, pterygoid plexus, cortical veins	TVE	Improved
20	Lai J. et al. (19)	65/F	Tinnitus, headache, visual and auditory hallucinations, ataxia, myoclonus	White matter	Right transverse-sigmoid sinus, right Sigmoid sinus/ left transverse sinus, straight sinus, vein of Galen	TVE	Improved
21	Pu J. et al. (20)	51/M	Normal	Lenticular nuclei, white matter	Straight sinus/vein of Galen, vein of Rosenthal, internal cerebral veins	TAE	Improved
22	Gopinath M. et al. (21)	45/F	Normal	White matter	Torcular sinus, superior sagittal sinus/ND	TAE	Improved
23	Tominaga A. et al. (22)	87/F	Depression, disturbance of consciousness	Brainstem, left cerebellar peduncle	left transverse sinus/left superior petrosal sinus	TAE	Improved
24	Prosperini L. et al. (23)	84/M	Delirium, poor speech fluency	White matter	Left transverse sinus/left occipital artery	TAE	Improved
25	Xie J. et al. (24)	70/F	Normal	White matter	Upper and lower sagittal sinuses, bilateral transverse sinuses/ND	No treatment	Deteriorated
26	Xie J. et al. (24)	67/M	Pain and discomfort in the occiput and back of the neck, increased lethargy	Thalamus	Internal cerebral vein, vein of Galen, straight sinus/ND	No treatment	Died
27	Present case	59/M	Normal	Brain stem, basal ganglia	Transverse-sigmoid sinuses/straight sinus, vein of Galen, vein of internal cerebral, vein of Rosenthal	No treatment	Deteriorated

F, female; M, male; TAE, Transarterial embolization; TVE, Transvenous embolization; ND, Not described.

Second, our report is limited to a single case. It is important that future reports include a series of cases to shed light on the relationship between CVST and DAVF. Additionally, functional experiments should be conducted to better understand the underlying mechanisms.

In conclusion, we present a classic case of DAVF characterized by rapidly progressing parkinsonism accompanied by dementia. Cerebral angiography was instrumental in identifying the arterial feeders and venous outflow of the fistula. It is crucial to consider DAVF as a potential underlying cause for rapidly progressing parkinsonism with dementia. As the condition is treatable and its effects potentially reversible, prompt diagnosis and intervention for DAVF are of paramount importance.

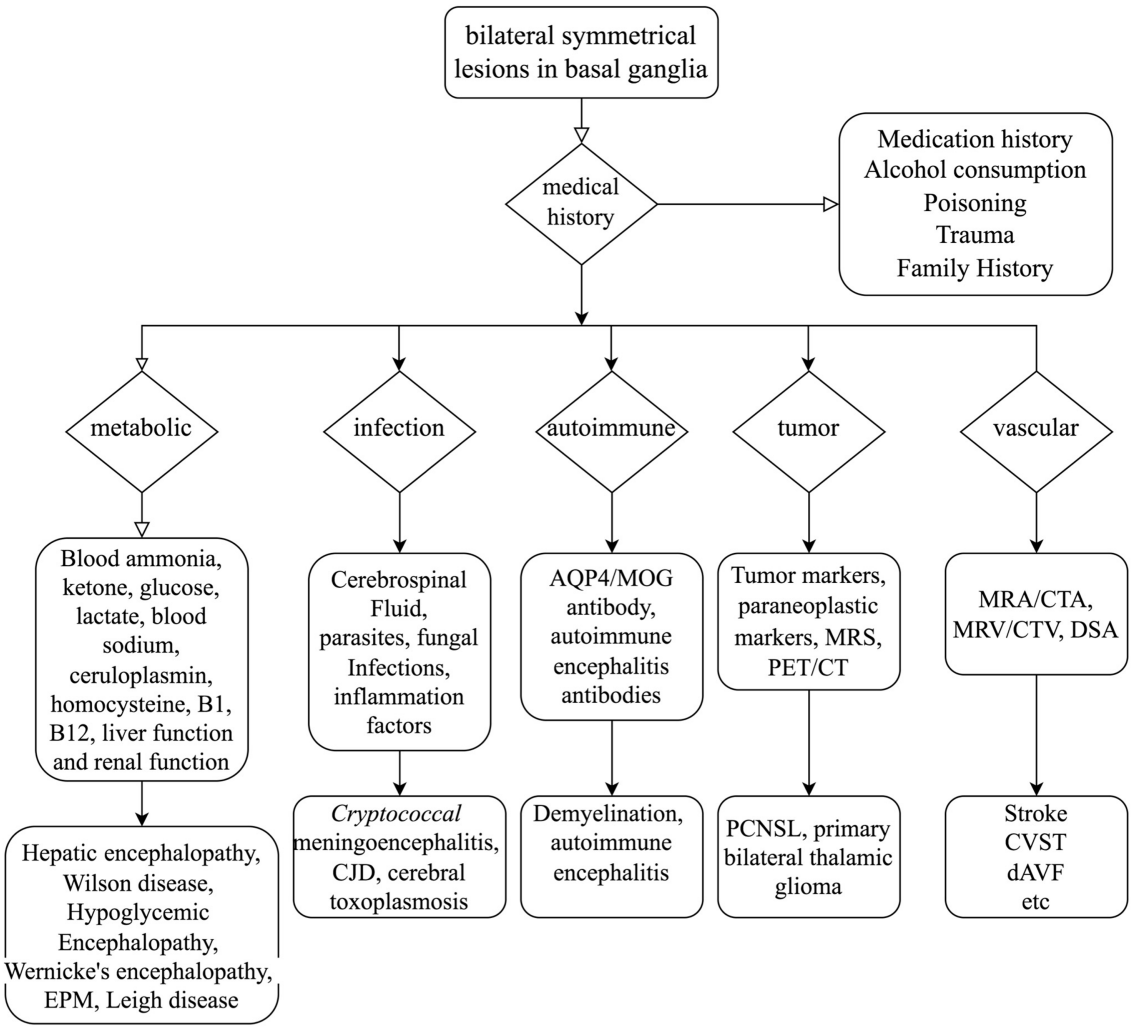

FIGURE 4
Mind flow chart for the differential diagnosis of bilateral symmetrical lesions in the basal ganglia. AQP4, Aquaporin-4; MOG, myelin oligodendrocyte glycoprotein; MRS, magnetic resonance spectroscopy; PET/CT, positron emission tomography/computed tomography; MRA, magnetic resonance angiography; CTA, computed tomography angiography; MRV, magnetic resonance venography; CTV, computed tomography venography; DSA, digital subtraction angiography; EPM, extrapontine myelinolysis; CJD, Creutzfeldt–Jakob disease; PCNSL, primary lymphomas of the central nervous system; CVST, cerebral venous sinus thrombosis; dAVF, dural arteriovenous fistula.

2.1 Patient perspective

The son of the patient says, "I noticed that over a month ago, my father began to walk slowly, hunching over with a diminished stride, taking small, shuffling steps. Additionally, he has become less responsive, his voice has lowered, and he's lost control of his bladder and bowels. We sought medical attention at our local hospital and underwent some tests. Given my father's long history of alcohol consumption, the local hospital treated him for alcohol poisoning. However, not only did his symptoms not improve, they actually worsened. We then consulted the Department of Neurology at the Second Xiangya Hospital. After a cerebral angiogram, the doctor diagnosed him with DAVF and recommended surgery. But the operation can be complicated. We worried about the safety of the surgery as well as a considerable expense, so we could not proceed with the operation."

Data availability statement

The datasets presented in this article are not readily available because of ethical and privacy restrictions. Requests to access the datasets should be directed to the corresponding author.

Ethics statement

Ethical review and approval was not required for the study on human participants in accordance with the local legislation and institutional requirements. Written informed consent from the patients/participants or patients/participants' legal guardian/next of kin was not required to participate in this study in accordance with the national legislation and the institutional requirements. Written informed consent was obtained from the individual(s) for

the publication of any potentially identifiable images or data included in this article.

Author contributions

RT: Writing – original draft, Writing – review & editing. QC: Writing – review & editing. LQ: Writing – original draft, Writing – review & editing.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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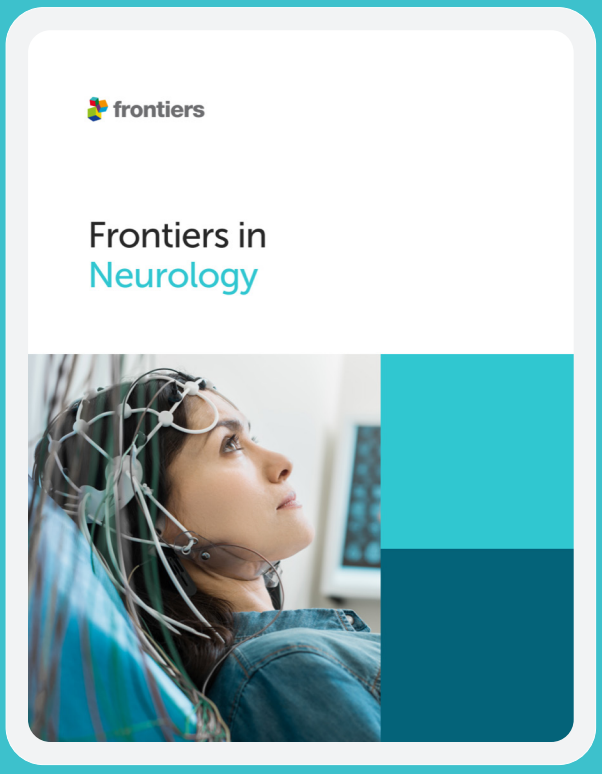

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