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Case Report

A case of spontaneous direct vertebral artery - External vertebral venous plexus fistula in the upper cervical portion

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ABSTRACT

Background: Spontaneous direct vertebral artery-external vertebral venous plexus (VA-EVVP) fistula is a rare disease that presents in patients with neurofibromatosis type 1 (NF-1) or trauma.

Case Description: An 82-year-old female patient with no neurological deficits or trauma presented to our hospital with right hemianopsia. Head magnetic resonance imaging (MRI) revealed left occipital cerebral infarction and magnetic resonance angiography demonstrated high signal intensity in the left transverse sinus (TS). The attending doctor diagnosed an old infarction on the left occipital lobe and dural arteriovenous fistula (AVF) in the TS. After 3 years after the first diagnosis, her new attending doctor re-checked the MRI and performed digital subtraction angiography (DSA). The DSA examination revealed a single-hole AVF between the vertebral artery and external vertebral plexus at the C2 level, which was diagnosed as upper cervical VA-EVVP. The patient presented with tinnitus due to a high-flow VA-EVVP fistula, so we performed coil embolization of the fistula under general anesthesia using a double-catheter technique and achieved subtotal embolization, which diminished the intracranial reflux. The 6-month follow-up DSA image revealed complete obliteration of the AVF.

Conclusion: We report a rare case of upper cervical VA-EVVP fistula in a patient with no history of trauma and relevant medical conditions. Coil embolization of the fistula was performed using a combination of balloon-assisted and double-catheter techniques. Although the patient showed residual shunt flow after the intervention, follow-up DSA revealed complete obliteration. These findings should provide novel insights for the treatment strategy against VA-EVVP fistula.

Keywords: Arteriovenous fistula, Coil embolization, Spontaneous, direct vertebral artery-external vertebral venous plexus fistula

INTRODUCTION

Spontaneous direct vertebral artery-external vertebral venous plexus (VA-EVVP) fistula is a rare entity that presents as an arteriovenous fistula (AVF) between the vertebral artery (VA) and the adjacent venous plexus.[4] VA-EVVP fistula is usually caused by trauma but may be occasionally caused by neurofibromatosis type-1 (NF-1), fibromuscular dysplasia (FMD), Ehler-Danlos syndrome, or Marfan syndrome.[1,13,14] The goal of treatment for VA-EVVP fistula is complete occlusion of the high-flow fistula by endovascular occlusion or surgical ligation. Here, we report

a rare case of spontaneous VA-EVVP fistula with high-flow reflux to the intracranial venous system. We performed endovascular treatment using a combination of balloon-assisted and double-catheter techniques, and the fistula was found to be obliterated in the follow-up examination performed 3 months after the treatment.

CASE REPORT

An 82-year-old female patient presented to our outpatient department with right-lower quadrantanopia and tinnitus. Her medical history included episodes of depression, hypertension, and no congenital disease. Head magnetic resonance imaging (MRI) revealed an old infarction in the left medial occipital lobe, and time of flight (TOF) imaging demonstrated high intensity in the left transverse sinus (TS) [Figures 1a and b]. Cervical MRI showed no T2 high intensity in the spinal cord. The attending doctor diagnosed TS dural AVF. After 3 years after the first diagnosis, the next attending doctor noticed that the high intensity in the sinus was contiguous with the suboccipital cavernous sinus or marginal sinus. Subsequently, digital subtraction angiography (DSA) was performed, and it showed AVF between the left VA at the C3 level and the external vertebral venous plexus (EVVP) with a 3.6-mm varix [Figure 2a]. The VA exhibited tortuosity, ectasia, and irregularity. The fistula was not fed by collateral flow from the external carotid artery and deep cervical artery. The ascending venous drainage root extended to the TS through the suboccipital cavernous sinus or jugular vein, and the other venous drainage root extended into the contralateral EVVP through the internal vertebral venous plexus (IVVP) [Figures 2b-d]. The left distal VA showed decreased flow due to the arteriovenous (AV) shunt. The patient reported tinnitus, so we decided to perform endovascular occlusion of the VA-EVVP fistula.

Endovascular treatment was performed under general anesthesia. An 8-Fr OPTIMO catheter (Tokai Medical Products, Aichi, Japan) was guided into the left VA using a 6-Fr JB2 catheter and a 0.035-inch guiding wire. An intermediate catheter (NAVIEN; Medtronic, Minneapolis, MN, USA) was then advanced into the distal VA. A microcatheter (Excelsior SL-10; Stryker, Kalamazoo, MI, USA) was inserted into the lateral EVVP using a microwire (SynchroSELECT soft; Stryker). Another microcatheter (Phenom 17; Medtronic) was placed in the varix of the EVVP near the fistula using a microwire. A balloon catheter (Scepter XC: Terumo, Tokyo, Japan) was placed in the VA where the fistula was present [Figure 3a]. Coil embolization using Phenom 17 was performed with the balloon-assisted technique, and 10 coils were deployed in the lateral EVVP with the varix [Figure 3b]. The fistula was mostly occluded while maintaining patency of the left VA, but a residual AV shunt was noted [Figure 3c]. The distal VA flow was greater than that in the preoperative phase, and the reflux to the suboccipital cavernous sinus and TS disappeared.

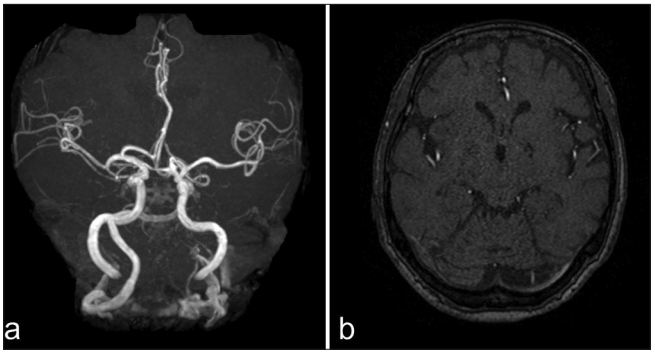

Figure 1: (a) Initial head magnetic resonance angiography showing a high signal in the transverse sinus, basilar plexus, internal jugular vein, and suboccipital cavernous sinus. (b) Time of flight imaging showing a high signal in the transverse sinus.

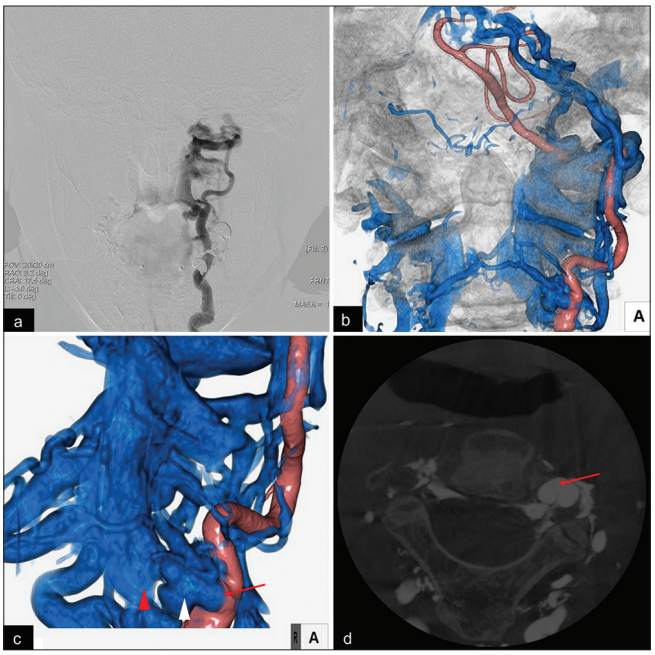

Figure 2: (a) Left vertebral artery (VA) angiography revealing vertebral artery- external vertebral venous plexus (VA-EVVP) fistula at the C2 level and venous drainage into the external VVP (EVVP), internal VVP (IVVP), internal jugular vein, and transverse sinus. (b, c, d) Three-dimensional rotational angiography and cone-beam computed tomography of the VA showing a single hole of a large fistula (arrow), EVVP (white arrowhead), and IVVP (red arrowhead).

Head MRI on the following day showed no remarkable infarction and intracranial venous reflux, and the patient's tinnitus disappeared. She was discharged on post-operative day 7. The 3-month follow-up DSA scan demonstrated

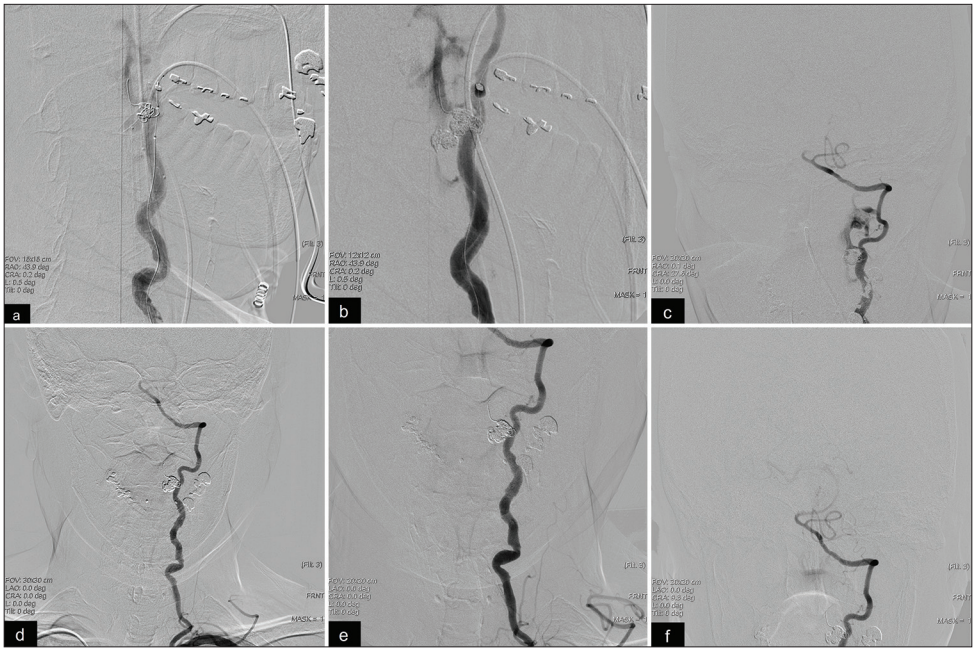

Figure 3: (a) Microcatheters were introduced into the EVVP (white arrowhead) and IVVP (red arrowhead). The arrow indicates the balloon catheter in the left VA. (b) Coil embolization by a combination of balloon-assisted and double-catheter techniques. (c) VA angiography after coil embolization. (d,e,f) VA angiography at three months after treatment showing complete occlusion of the fistula.

complete occlusion of the VA-EVVP fistula with patency of the left VA [Figures 3d-f].

DISCUSSION

VA-EVVP fistula is a rare disease caused by trauma, iatrogenic complications, or congenital disorders.[9] The most common cause of VA-EVVP fistula is neck trauma or iatrogenic injuries, such as those caused by surgery or percutaneous internal jugular vein cannulation.[2,6] On the other hand, spontaneous VA-EVVP fistula is often associated with congenital disorders such as NF-1, FMD, Ehler-Danlos syndrome, and Marfan syndrome.[5,6] Although VA-EVVP fistula due to trauma occurs at the lower cervical level, spontaneous VA-EVVP fistula is observed at the upper cervical level.[16] In the present case, the patient showed no remarkable medical history accompanying the VA-EVVP fistula, such as congenital disorders, trauma, and iatrogenic injuries.

VA-EVVP fistula is a rare disorder that presents with a single hole and a high-flow shunt between the VA and vertebral venous plexus (VVP).[5,14] In general, the only feeding artery to the fistula is the VA, but other arteries, such as the contralateral VA, internal carotid arteries on both sides, occipital artery, deep cervical artery, and ascending cervical artery, may contribute to this shunt.[2,5,10,14,16] The draining vein is mainly the VVP, and one of the VVPs is the IVVP, which exists in the spinal canal. This is the reason why most patients experience neck pain (40%), radiculopathy (30%), radiculomyelopathy (28%), and bruit (>50%). In this case, only bruit was noted, and the patient showed no symptoms related to the spinal cord and root despite the increased flow and expansion of the IVVP.

Due to the rare incidence of VA-EVVP fistula, its clinical course has not been established. While the appearance of symptoms such as numbness, neck pain, and gait disturbances can easily inform the decision to perform treatment for VA-EVVP fistula, treatment decisions for cases showing asymptomatic fistulas or fistulas with only bruit require more careful consideration. In comparison with other fistulas, the VA-EVVP fistula rarely shows spontaneous thrombosis.[12] Iampreechakul *et al*. reported that partial surgery resulted in spontaneous obliteration of VA-EVVP fistulas associated with FMD.[11] In the present case, we performed endovascular treatment, which achieved subtotal occlusion with partial residual shunt flow and subsequently resulted in complete occlusion. Although complete occlusion is the primary goal of surgery or endovascular treatment for VA-EVVP fistula, our findings indicate that partial obliteration may also yield good outcomes, especially in patients showing VA-EVVP fistulas with lower risks and complication rates.

Various methods to treat VA-EVVP fistula, including surgery and endovascular treatment, have been reported to date. Endovascular treatment accounts for the majority of cases and surgery is limited to cases showing unsuccessful endovascular treatment.[7] Although the surgical techniques involve proximal and distal ligation or only proximal ligation, neither strategy can preserve the ipsilateral VA.[7,8,15] Surgical ligation is also associated with the risks of exposure difficulty and extensive bleeding. Thus, endovascular occlusion has become the preferred strategy for the treatment of VA-EVVP fistula. In contrast to surgical treatment, endovascular treatment can preserve the VA using detachable balloons, detachable coils, liquid embolic agents, and covered stents.[3] However, due to the high-flow shunt in the fistula or the appearance of vascular-related events due to hemodynamic changes, complete occlusion without parent artery occlusion is difficult in many cases. In our case, partial embolization with balloon-assisted and double-catheter techniques was effective. The main high blood flow through the fistula, such as the flow from the ascending and contralateral drainage root through the EVVP and IVVP, was diminished, and the residual shunt flow was drained in only the EVVP, which was not the main drainage, facilitating thrombosis in the residual fistula. Therefore, in selective embolization, the main drainage root should be obliterated using balloon-assisted or double-catheter techniques.

Preservation of VA patency in endovascular treatment of VA-EVVP fistula is also challenging, and some reports have recommended the use of assisted techniques to preserve VA patency.[3,6,16] In our case, placement of the balloon catheter in the VA may have helped prevent coil herniation into the parent artery. Although coil embolization by a simple technique is the most popular method, no strategies have been established to prevent coil herniation, which may easily occur in VA-EVVP fistulas with a large hole and may result in inadequate embolization. The double-catheter technique is also useful to prevent dangerous drainage into the vein in the spinal canal. In fact, we performed additional embolization after the first obliteration of the varix in the EVVP due to residual drainage into the IVVP. In our case, the combination of balloon catheter-assisted and double-catheter techniques was successful and resulted in good patient outcomes.

CONCLUSION

We report a rare case of spontaneous VA-EVVP fistula in a patient with no history of trauma, iatrogenic injuries, and congenital disease. DSA was performed due to the non-specific symptoms and the difficulty of diagnosis with only a head MRI. Coil embolization using a combination of balloon-assisted and double-catheter techniques finally resulted in complete obliteration of the fistula.

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

REFERENCES

1. Bahar S, Chiras J, Carpena JP, Meder JF, Bories J. Spontaneous vertebro-vertebral arterio-venous fistula associated with fibro-muscular dysplasia. Report of two cases. Neuroradiology 1984;26:45-9.
2. Briganti F, Tedeschi E, Leone G, Marseglia M, Cicala D, Giamundo M, *et al.* Endovascular treatment of vertebro-vertebral arteriovenous fistula. A report of three cases and literature review. Neuroradiol J 2013;26:339-46.
3. Chen C, Wu Y, Zhao K, Duan G, Liu J, Huang Q. Endovascular treatment of vertebro-vertebral arteriovenous fistula in neurofibromatosis type I: A report of two cases and literature review with a focus on endovascular treatment. Clin Neurol Neurosurg 2021;207:106806.
4. Deans WR, Bloch S, Leibrock L, Berman BM, Skultety FM. Arteriovenous fistula in patients with neurofibromatosis. Radiology 1982;144:103-7.
5. Gao P, Chen Y, Zhang H, Zhang P, Ling F. Vertebral arteriovenous fistulae (AVF) in neurofibromatosis type 1: A report of two cases. Turk Neurosurg 2013;23:289-93.
6. Geng J, Hu P, Ma Y, Zhang P, Zhang H. Endovascular treatment of V3 segment vertebro-vertebral arteriovenous fistula with Willis covered stent: Case report and literature review. Interv Neuroradiol 2019;25:97-101.
7. Gobin YP, Duckwiler GR, Viñuela F. Direct arteriovenous fistulas (carotid-cavernous and vertebral-venous). Diagnosis and intervention. Neuroimaging Clin N Am 1998;8:425-43.
8. Halbach VV, Higashida RT, Hieshima GB. Treatment of vertebral arteriovenous fistulas. AJR Am J Roentgenol 1988;150:405-12.
9. Honarmand AR, Ansari SA, Alden TD, Soltanolkotabi M,

Schoeneman SE, Hurley MC, *et al.* Endovascular management of pediatric high-flow vertebro-vertebral fistula with reversed basilar artery flow. A case report and review of the literature. Interv Neuroradiol 2013;19:215-21.
10. Hori Y, Goto K, Ogata N, Uda K. Diagnosis and endovascular treatment of vertebral arteriovenous fistulas in neurofibromatosis type 1. Interv Neuroradiol 2000;6:239-50.
11. Iampreechakul P, Siriwimonmas S. Spontaneous obliteration of spontaneous vertebral arteriovenous fistula associated with fibromuscular dysplasia after partial surgery: A case report. Interv Neuroradiol 2016;22:717-27.
12. Lasjaunias P, Ter Brugge KG, Berenstein A. Surgical neuroangiography. Berlin: Springer; 2006.
13. Narayana RV, Pati R, Dalai S. Endovascular management of spontaneous vertebrovertebral arteriovenous fistula associated with neurofibromatosis 1. Indian J Radiol Imaging 2015;25:18-20.
14. Paolini S, Colonnese C, Galasso V, Morace R, Tola S, Esposito V, *et al.* Extradural arteriovenous fistulas involving the vertebral artery in neurofibromatosis Type 1: Case report. J Neurosurg Spine 2008;8:181-5.
15. Sher MH, Meyer NI, Lenhardt HF, Trummer MJ. Arteriovenous fistula involving the vertebral artery: Report of three cases. Ann Surg 1966;163:408-13.
16. Uneda A, Suzuki K, Okubo S, Hirashita K, Yunoki M, Yoshino K. Neurofibromatosis Type 1-associated extracranial vertebral artery aneurysm complicated by vertebral arteriovenous fistula after rupture: Case report and literature review. World Neurosurg 2016;96:609.e13-8.

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