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Case Report

Unique vascular structures of a radicular arteriovenous fistula at the craniocervical junction along the first cervical spinal nerve: A case report

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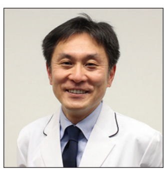

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ABSTRACT

Background: An arteriovenous fistula (AVF) at the craniocervical junction (CCJ) is a rare vascular malformation. Definitive diagnosis and curative treatment of CCJ AVF are challenging.

Case Description: A 77-year-old man presented with subarachnoid hemorrhage. Cerebral angiography showed an AVF at the CCJ, which drained into a radicular vein. The lesion was fed by a vertebral artery, anterior and lateral spinal arteries (LSAs), and the occipital artery (OA). There were two unique structures: the LSA originating from the posterior inferior cerebellar artery of the extracranial V3 segment and the OA feeding the shunt. Curative treatment involved two steps: endovascular embolization of feeders using Onyx and surgical shunt disconnection. Feeding arteries were blackened by Onyx, which helped identify the location of the shunt. The shunt was located behind the first cervical (C1) spinal nerve, and the draining vein was confirmed on the deep side of the nerve. A clip was applied to the draining vein distal to the shunt. Tiny vessels supplying the shunt were then coagulated referring to blackened arteries.

Conclusion: A radicular AVF at the CCJ along the C1 spinal nerve had unique vascular structures. Definitive diagnosis and curative treatment were achieved by combining endovascular embolization using Onyx and direct surgery.

Keywords: Craniocervical junction, Direct surgery, First cervical spinal nerve, Onyx, Radicular arteriovenous fistula

INTRODUCTION

Arteriovenous fistulas (AVFs) at the craniocervical junction (CCJ) are rare vascular malformations, accounting for approximately 2% of spinal AVFs.[1,3] Typical manifestations of the lesions are subarachnoid hemorrhage or myelopathy induced by chronic venous congestion.[2,3,14,15] Preoperative diagnosis of CCJ AVFs is challenging because the anatomy of the neurovascular structures is complex. CCJ AVFs are diagnosed based on the location of the AV shunt (intradural, dural, and epidural), feeding arteries (spinal cord or dural artery), and draining veins. Locations of AV shunts are divided into intradural (on the spinal cord or on the spinal nerves) or dural (on the inner surface of the dura mater) and epidural (on the outer surface of the dura mater). CCJ AVFs are classified into five types, based on angioarchitecture, as dural AVF, radicular AVF, epidural AVF with pial feeders, epidural AVF, or perimedullary AVF.[2] Radicular AVFs are defined as those fed by radicular and/or meningeal arteries that drain into radicular veins through AV shunts located on the spinal nerve roots. In particular, radicular AVFs at the CCJ along the first cervical (C1) spinal nerve are a rare and unique subtype.[3] Definitive diagnosis is difficult by imaging findings alone and the treatment strategy remains unclear.

A case of a radicular AVF at the CCJ along the C1 spinal nerve with unique vascular structures is described. Definitive diagnosis and curative treatment were achieved by combining endovascular embolization using Onyx and direct surgery.

CASE DESCRIPTION

A 77-year-old man presented with sudden-onset headache and consciousness disturbance. His Glasgow coma scale was E3V4M5. Computed tomography (CT) showed massive subarachnoid hemorrhage in the posterior cranial fossa, especially around the medulla [Figure 1a]. CT angiography did not show any apparent ruptured aneurysms, although abnormal blood vessels around the right vertebral artery (VA) at the C1 level were detected [Figures 1b-d]. The right VA angiography showed an AVF at the CCJ, which was supplied mainly by the right VA and the anterior spinal artery (ASA), and it drained into a radicular vein [Figure 1e]. On the other hand, the left VA angiography did not demonstrate vascular lesions [Figure 1f]. In detailed assessments of the vascular structures using super-selective angiography, the right lateral spinal artery (LSA) originated from the right posterior inferior cerebellar artery (PICA) and was feeding the shunt [Figures 2a and b]. Interestingly, a branch of the right occipital artery (OA) was also feeding the shunt [Figures 2c and d]. On super-selective angiography with contrast agent injections from both the LSA and the OA at the same time, the flow of each was confirmed to supply the same shunt [Figures 2e and f]. A radicular AVF at the CCJ was suspected from the imaging findings. Two-step treatment was planned: decreasing blood flow by endovascular embolization (first-step), followed by curative surgical shunt resection (second-step).

Endovascular embolization of the LSA and the OA feeders was performed using Onyx under general anesthesia. After embolization, the right VA angiography showed residual

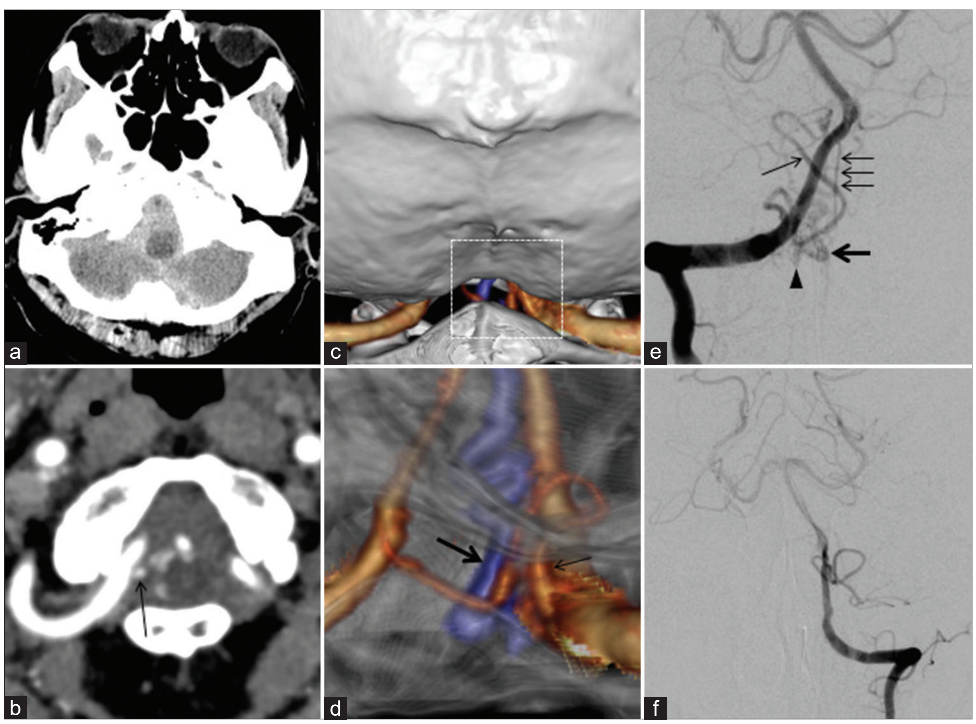

Figure 1: Computed tomography shows subarachnoid hemorrhage in the posterior cranial fossa (a). Computed tomography angiography shows abnormal blood vessels around the right vertebral artery (VA) at the C1 level (b-d). d represents an enlargement of the white dot square part of c. Right VA angiography shows an arteriovenous fistula draining into a radicular vein (e). Left VA angiography does not demonstrate vascular lesions (f). (Arrow: Right posterior inferior cerebellar artery, Thick arrow: Draining vein, Arrowhead: Shunt, Arrows: Anterior spinal artery).

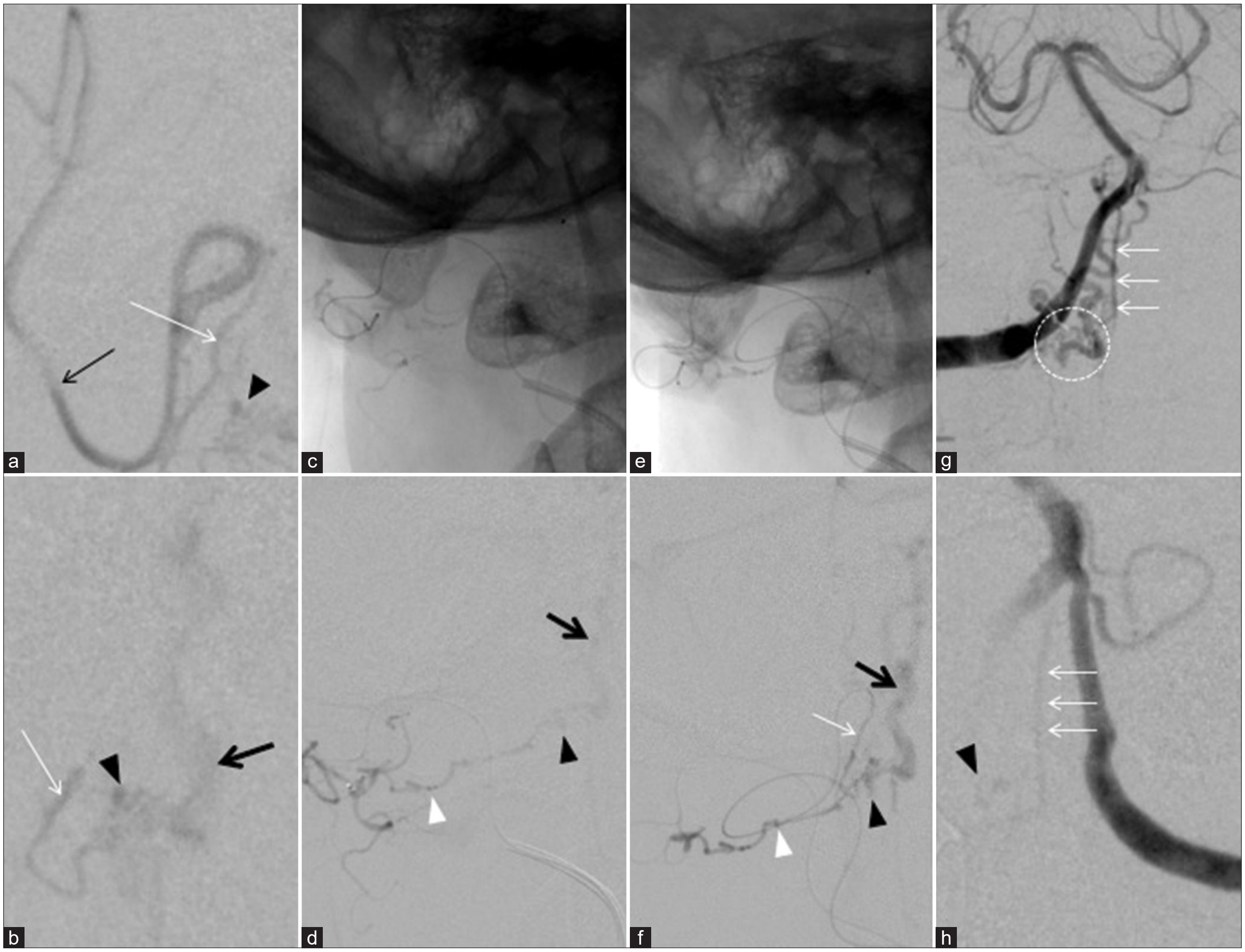

Figure 2: Super-selective angiography shows that the right lateral spinal artery (LSA) originates from the right posterior inferior cerebellar artery and as a feeder (a and b). Super-selective angiography shows the occipital artery (OA) as a feeder (c and d). Super-selective angiography of both the LSA and the OA demonstrates each feeder supplying the same shunt (e and f). After embolization, right vertebral artery (VA) angiography shows residual flows from the right VA and the anterior spinal artery (ASA) (g). Left VA angiography also demonstrates the residual flow from the ASA (h). (White arrow: LSA, White arrows: ASA, White arrowhead: OA, Arrow: Posterior inferior cerebellar artery, Arrowhead: shunt, Thick arrow: Draining vein, Dot circle: Radiculomeningeal arteries from the right VA).

feeder flows from radiculomeningeal arteries of the right VA and spinal pial arteries of the ASA [Figure 2g]. The left VA angiography also confirmed the residual flows of the ASA [Figure 2h]. The day after first-step treatment, surgical shunt resection was performed via sub-occipital craniotomy with partial atlas osteotomy. Intraoperative photographs showed the PICA penetrating dura mater from the extradural space, and feeding arteries from the LSA were blackened by Onyx [Figure 3a]. The shunt was located behind the right C1 spinal nerve, although it was not fully exposed because bleeding was encountered when trying to peel it off. A red draining vein was confirmed at the cranial deep side of the C1 spinal nerve [Figure 3b]. On intraoperative indocyanine green angiography, the draining vein was visible in the arterial phase [Figure 3c]. Judging from all imaging and intraoperative findings, the AV shunt was located along the C1 spinal nerve, fed by radiculomeningeal arteries from the VA, spinal arteries from both the LSA and ASA, and branches of the OA, and it drained into a radicular vein. Therefore, the definitive diagnosis was a radicular AVF at the CCJ along the C1 spinal nerve. A clip was applied to the draining vein distal to the shunt [Figure 3d]. Motor evoked potential monitoring did not show any changes. Then, tiny vessels around the shunt and the draining vein were coagulated referring to the blackened arteries. Finally, disappearance of abnormal vessels was confirmed by repeat indocyanine green angiography. Postoperative cerebral angiography showed disappearance of the lesion [Figure 4]. The patient was discharged without apparent neurological deficits and there has been no recurrence for more than 3 years.

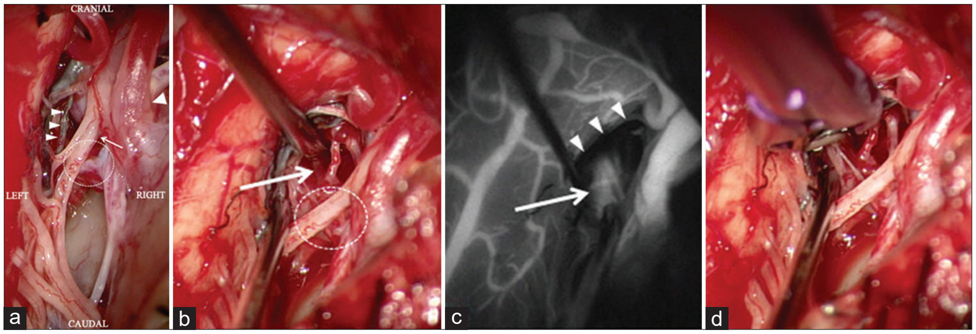

Figure 3: Intraoperative photographs show the posterior inferior cerebellar artery penetrating the dura mater, the lateral spinal artery (LSA) is blackened by Onyx, and the shunt is located behind the first cervical (C1) spinal nerve (a). Another intraoperative photograph from a different angle shows a draining vein at the cranial deep side of the C1 spinal nerve (b). Intraoperative indocyanine green angiography shows that the draining vein is visible in the arterial phase (c). A clip is applied to the draining vein distal to the shunt (d). (Arrow: C1 spinal nerve, Arrowhead: Posterior inferior cerebellar artery, Arrowheads: LSA blackened by Onyx, Dot circle: Location of the shunt, Thick arrow: Draining vein).

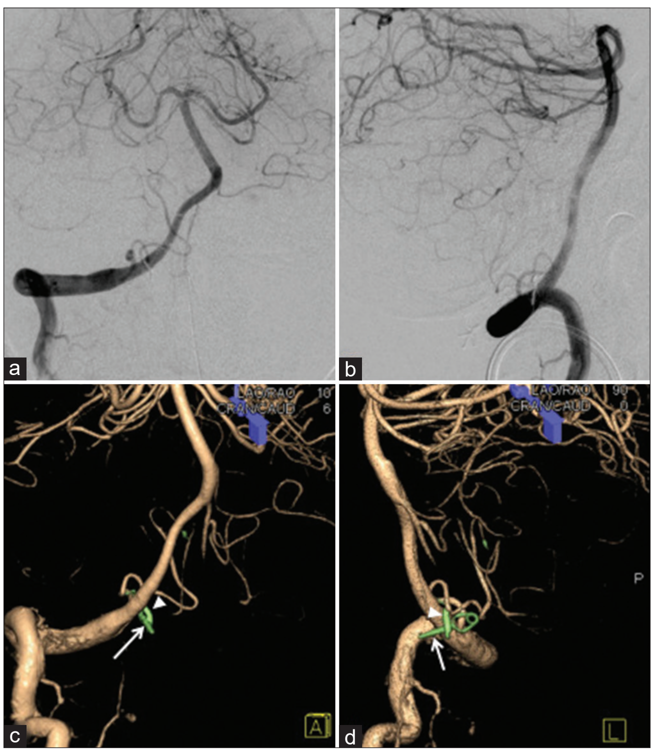

Figure 4: Postoperative right vertebral artery angiography shows disappearance of the lesion (a and c: Anteriorposterior, b and d: Lateral views). (Arrow: Clip, arrowhead: Onyx).

DISCUSSION

CCF AVFs were once described as dural AVFs in the 1990s.[7] However, the lesions are now interpreted as the coincidental development of a dural AVF and a perimedullary AVF.[5,11] The most common presentation of CCJ AVFs was subarachnoid or intramedullary hemorrhage.[2,14,15] Historically, an ascending drainage route into an intracranial vein and venous varices was considered to be associated with a risk of hemorrhage.[5,7,14,15] Recently, a large, multicenter, and cohort study assessed the details of the angioarchitecture of CCJ AVFs and associations with clinical presentations, including 54 patients with a total of 59 lesions.[2] The proportions of the five types were dural AVF (37%), radicular AVF (29%), epidural AVF with pial feeders (14%), epidural AVF (10%), and perimedullary AVF (10%). AV shunts were fed by radiculomeningeal arteries from the VA (98%) and the spinal pial artery (63%) from the ASA and/or LSA. Angiographic characteristics associated with hemorrhagic presentations were: ASA as a feeder; presence of aneurysmal dilatation of feeders; and radicular AVF.[2]

Cerebral angiography and rotational CT angiography are the gold standard for diagnosis and can demonstrate small feeding arteries and venous varices.[3] Radicular AVF at the CCJ along the C1 spinal nerve root may be overlooked on routine initial cerebral angiography if bilateral VA injection is not performed.[3] Contralateral VA angiography would be omitted when confirming retrograde filling of the distal contralateral VA to the level of the PICA on a selective ipsilateral VA injection. In the present case, the right VA angiography showed the lesion, although left VA angiography did not. Super-selective angiography is essential for assessing vascular structures accurately and making a diagnosis of CCJ AVF.[2] Moreover, the technique of super-selective angiography contrast agent injections from multiple feeders at the same time may be useful to confirm each flow supplying the same shunt.

In the present case, the radicular AVF at the CCJ along the C1 spinal nerve had two unique vascular structures. The first was a feeder of the LSA originating from the PICA of the extracranial V3 segment. The unique structure of the

PICA penetrating dura mater and the LSA originating from the proximal cranial loop of the PICA was confirmed by angiography and direct surgery. The PICA usually originates at the intracranial V4 segment, although about 5% to 20% originate at the extracranial V3 segment.[4,6] The anatomical variations of the VA and PICA are related to variations of the LSA.[8,9,12] The origins of the LSA are classified into the PICA, intradural VA, or anastomotic artery associated with the C1 nerve root.[8] The second unique vascular structure was the OA feeding the shunt. The external carotid artery as a feeder of CCJ AVFs was seen in 12 of 59 lesions (12%). In radicular AVF, only one was seen in 17 lesions (6%).[2]

Early treatment after initial hemorrhage will provide better outcomes, because the rebleeding rate of CCJ AVFs was 8.3%.[10,16]

Curative treatment of CCJ AVFs involves disconnecting or interrupting the connection between the shunt and draining vein.[2,3,16] In large case series, treatment methods were reported to be direct surgery only (47%), endovascular embolization only (25%), both direct surgery and endovascular embolization (14%), and conservative management (14%).[2] Glue or coils were used as the embolic materials. Despite advances in endovascular techniques, endovascular embolization alone of CCJ AVFs may not be feasible or safe.[3,13,16] In the present case, preoperative endovascular embolization was performed using Onyx to reduce shunt flows. CCJ AVFs supplied by not only dural but also pial arteries have many tiny feeders around the shunt and the draining vein. Injection of Onyx makes such tiny feeders also turn black and facilitates clear visualization of the anatomical structures during direct surgery. Intraoperative photograph showed the blackened feeders, which was very informative for identifying the locations of the shunt and draining vein. Therefore, two-step treatment involving endovascular embolization using Onyx (first step) and then disconnecting the shunt and draining vein by direct surgery (second step) is a useful and safe choice for curative treatment of CCJ AVFs.

CONCLUSION

A case of a radicular AVF at the CCJ along the C1 spinal nerve presenting with subarachnoid hemorrhage was described. The lesion had two unique vascular structures, a feeder of the LSA originating from the PICA of the extracranial V3 segment and the OA feeding the shunt. Definitive diagnosis and curative treatment were achieved by combining endovascular embolization and direct surgery. Preoperative injections of Onyx into the main feeders were effective in reducing blood flow and allowed clear visualization of the anatomical structures around the shunt and the draining vein during direct surgery.

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.

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