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Case Report

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Isolated Brainstem Congestion Caused By Craniocervical Junction Arteriovenous Fistula

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ABSTRACT

Although arteriovenous fistulas (AVFs) at the craniocervical junction (CCJAVFs) are rare, they often develop into a subarachnoid haemorrhage when they have an ascending venous drainage, or cause venous congestion of the spinal cord with descending venous drainage. Isolated brainstem lesions due to CCJAVF are extremely rare, and, to our knowledge, the vascular architectural features that could cause such lesions are unknown. We present a case of CCJAVF manifesting as isolated brainstem congestion and review the literature on the vessel architecture of these rare lesions.

A 64-year-old man was admitted to our hospital with gradually worsening nausea, dysphagia, double vision, grogginess, and gait disturbances. On admission, the patient showed dysarthria, horizontal ocular nystagmus to the left, paresis of cranial nerves IX and X, and ataxia on the right side. Magnetic resonance imaging (MRI) revealed an isolated lesion in the medulla. Cerebral angiography (CAG) showed a CCJAVF with the coexistence of intradural AVF and dural AVF, fed by the right first cervical radiculomedullary, right vertebral, and intradural posterior inferior cerebellar arteries, which were drained by the anterior spinal vein in an ascending direction. The patient underwent direct surgery to occlude dural and intradural fistulas. Postoperatively, the patient returned to work with full recovery from the neurological deficits via rehabilitation. MRI revealed vanishing brainstem congestion, and CAG revealed complete disappearance of the AVF.

CCJAVFs with venous drainage around the brainstem, regardless of their direction (ascending or descending), can cause isolated brainstem congestion, although this condition is rare.

KEYWORDS: Arteriovenous fistula (AVF), Brainstem congestion, The craniocervical junction (CCJ), Drainage

ABBREVIATIONS: AVFs: Arteriovenous fistulas, **CAG:** Cerebral angiography, **CCJAVFs:** Arteriovenous fistulas at the craniocervical junction, **CT:** Computed tomography, **MRI:** Magnetic resonance imaging, **PICA:** Posterior inferior cerebellar artery, **SAH:** Subarachnoid haemorrhage, **VA:** Vertebral artery

INTRODUCTION

The craniocervical junction (CCJ), the area from the foramen magnum to the axis level, has complicated structures, and arteriovenous fistulas at the craniocervical junction (CCJAVFs) are rare (9). Among arteriovenous fistulas (AVFs) in the skull or spinal cord region, CCJAVFs are reported in 1–2% of cases (8). They reportedly develop subarachnoid haemorrhage (SAH) or venous congestion of the spinal cord. Manifestation of brainstem ischemia/dysfunction localised to the brainstem is particularly rare. Myelopathy is present in 37–38% of CCFAVFs, while SAH and brainstem dysfunction occur in 35-45% and 3-8% of cases, respectively (12,15). Therefore, its rarity sometimes leads to an incorrect diagnosis of another brainstem or spinal cord disease, especially in cases of lesions localised to the brain stem (1,11,14).

Although it is widely accepted that ascending venous drainage is associated with haemorrhagic presentation and descending venous drainage is associated with venous congestion of the

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spinal cord (6,7,10), knowledge regarding the architectural information that causes brainstem congestion is lacking.

Herein, we present a case of CCJAVF with isolated brainstem congestion and review the literature on the vessel architecture of these rare lesions.

CASE DESCRIPTION

A 64-year-old man was admitted to our hospital with gradually worsening nausea, dysphagia, double vision, grogginess, and gait disturbances. His medical history was unremarkable, except for mild cervical disc herniation, which was treated conservatively. On admission, a neurological examination revealed dysarthria, horizontal ocular nystagmus to the left, paresis of cranial nerves IX and X with curtain signs to the right side, and an absent gag reflex. Ataxia was observed on the right side.

Although head computed tomography (CT) performed on admission revealed no abnormal findings, T2-weighted magnetic resonance imaging (MRI) showed a hyperintense lesion in the medulla (Figure 1A) and a thin meandering blood vessel around the medulla oblongata (Figure 1B). However, no signal was observed on MRA at the same site. Subsequent contrast-enhanced MRI revealed a pool of contrast medium in veins located around the medulla (Figure 1C). Cerebral angiography (CAG) showed CCJAVFs, with the coexistence of intradural AVF and dural AVF at the CCJ, which was fed by the right first cervical radiculomedullary artery, right vertebral artery (VA), and intradural posterior inferior cerebellar artery (PICA), and drained by the anterior spinal vein with ascending venous drainage (Figure 2A).

The patient underwent direct surgery on the 4th day of hospitalisation. We carefully examined the abnormal vessels during surgery and compared their anatomical architecture with the operative findings. We occluded the intradural feeding artery just before the shunt point and dural shunting immediately after passing through the dura mater (Figure 3A, B). Finally, we cauterised the dilated draining veins as far as possible (Figure 3C). CAG performed three days after surgery showed the complete disappearance of the AVFs. The patient's symptoms gradually improved, and MRI performed three months after surgery showed the disappearance of brainstem congestion. Subsequently, he returned to work three months after the procedure, fully recovering from his neurological deficits. At the 1-year follow-up evaluation, CAG showed no AVF recurrence (Figure 2B).

This study was conducted following the Declaration of Helsinki, and informed consent for publication was obtained from all patients. Our institutional review board approved this study (approval number: 510).

DISCUSSION

This case report provides important clinical suggestions. First, CCFAVFs rarely cause isolated brainstem congestions. Although recent reviews have shown that CCJAVFs often develop brainstem congestion, most brainstem lesions are continuous from the upper spinal cord. Thus, lesions limited to the brainstem, as in this case, are extremely rare (10,13). Only 11 cases, including the present case, have been reported in the literature (Table I).

Although isolated brainstem congestion due to CCFAVFs is rare, it is important to consider CCJAVFs as a causative disease, as patients presenting with venous congestion reportedly show more permanent neurological deficits than patients presenting with SAH (10). Consequently, early diagnosis and surgical obliteration are mandatory. However,

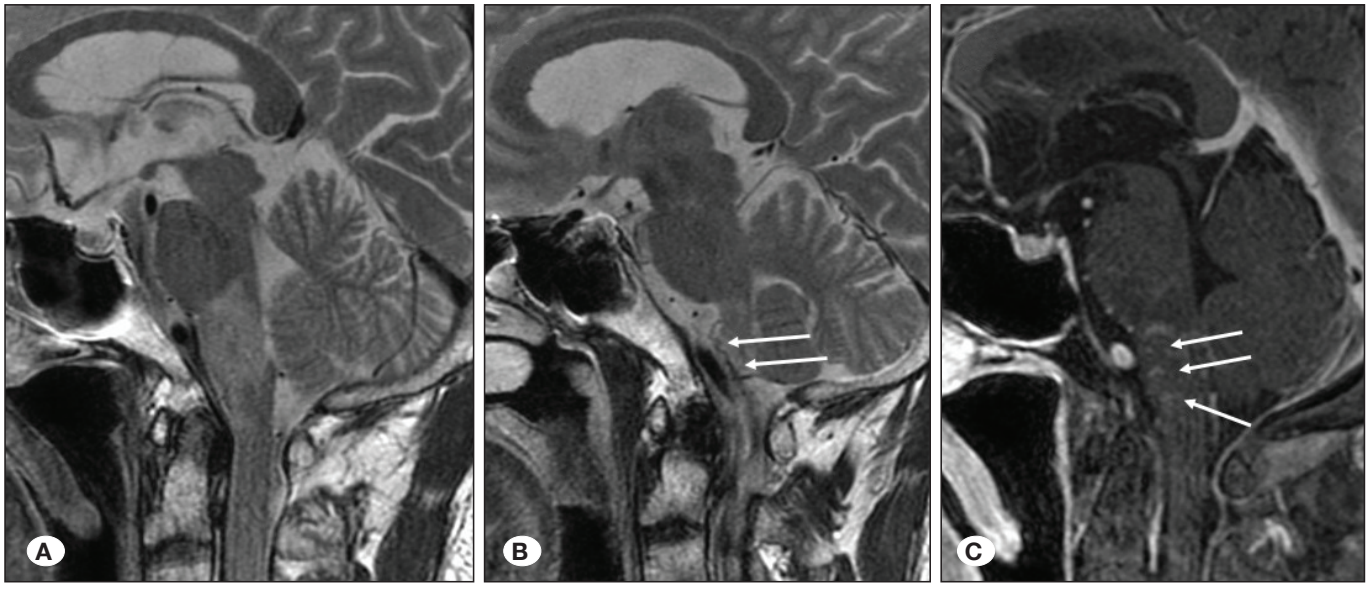

Figure 1: Magnetic resonance imaging (MRI) before surgery. **A)** Initial sagittal T2-weighted imaging (T2WI) showing a high-intensity area in the medulla oblongata. **B)** Sagittal T2WI MRI showing thin meandering vessels on the surface of the medulla oblongata (arrows). **C)** Sagittal T1WI with Gd enhancement showing a pool of contrast medium in the veins located around the medulla oblongata.

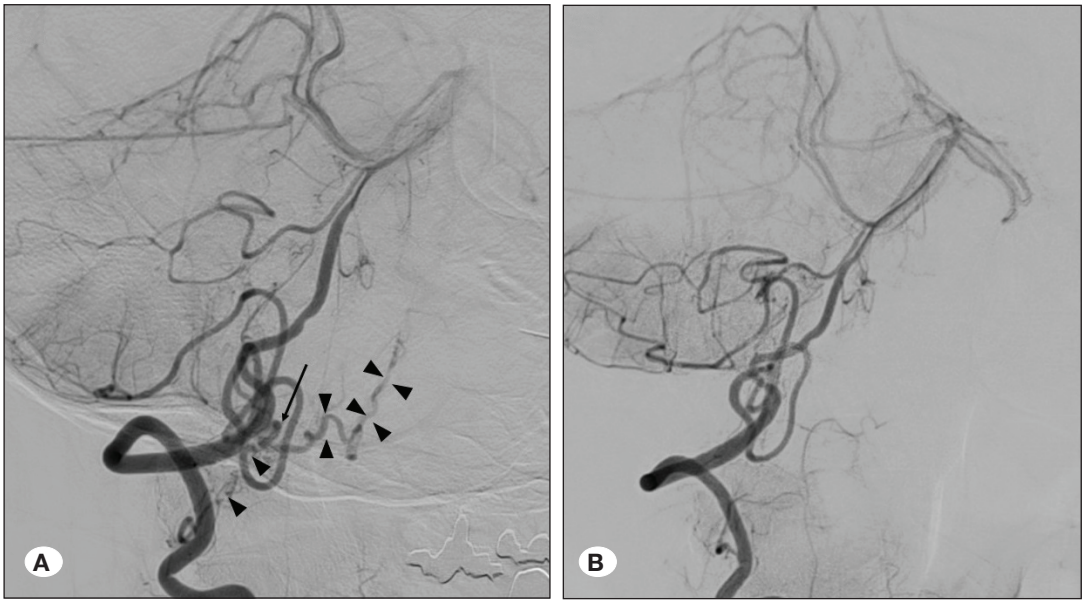

Figure 2: Vertebral angiography; right anterior oblique 45 degrees. **A)** Preoperative right vertebral angiography showing two dural arteriovenous fistulas fed by the right first cervical radiculomedullary artery and right vertebral artery (single arrowheads), with one intradural fistula fed by the posterior inferior cerebellar artery (PICA) (arrow) with anterior spinal vein drainage (paired arrowheads). **B)** Postoperative right vertebral angiography showing complete resolution of the AVFs.

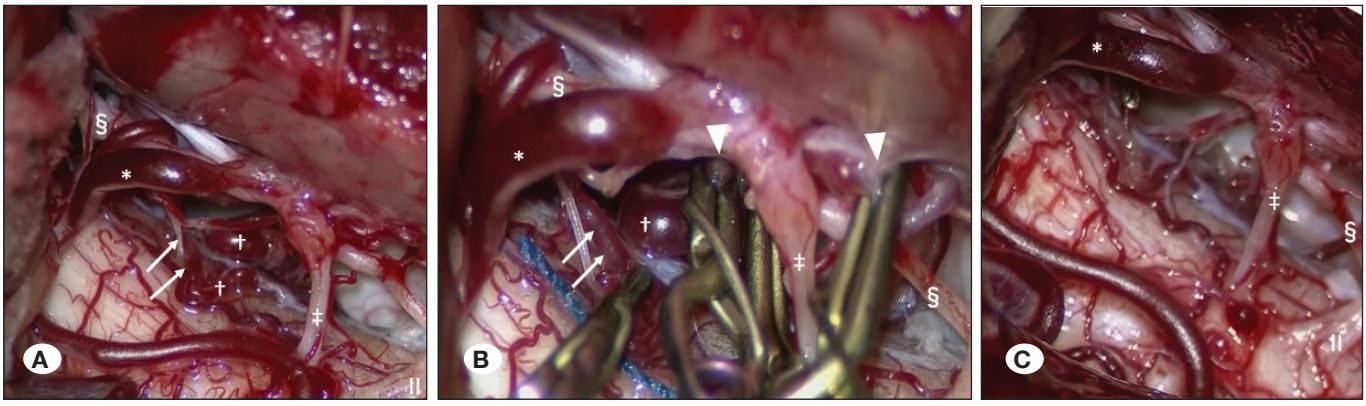

Figure 3: Intraoperative photographs of direct the disconnection of the fistulas of the craniocervical junction via right suboccipital craniotomy (right posterolateral views). **A)** Intraoperative photograph showing that the intradural AVF fed by the posterior inferior cerebellar artery (PICA; arrows) is connected to the dilated draining veins. **B)** The intradural AVF fed by the PICA is occluded, and the dural AVF is ligated immediately after penetrating the dura mater using temporary clips. **C)** Final view of the procedure showing disconnection of all three AVFs with coagulation and disappearance of the abnormally dilated vein. *posterior inferior cerebellar artery (PICA), † dilated anterior spinal vein, ‡ first cervical (C1) dorsal root, § spinal root of accessory nerves, and II second cervical (C2) dorsal root.

brainstem congestion complicated by CCJAVFs might often lead to delayed or false diagnoses (such as tumours, demyelination, and inflammation) and subsequent improper management (1,11,14). The table shows six of the 11 cases had delayed or missed diagnoses.

Venous drainage direction cannot always be used to determine the presentation of onset, SAH, or spinal congestion. The Table shows that CCJAVFs with venous drainage around the brainstem, regardless of their direction (ascending or descending), may cause isolated congestion in the brainstem. The architectural features of CCFAVFs that can cause isolated lesions in the brainstem are classified into two patterns: one is fed from the ECA and has descending drainage (The table; Patients no. 1–6), and the other is fed from the VA (including the cervical radiculomedullary artery) and has ascending drainage (The table; Patients no. 7–11). Although it has been widely accepted that ascending venous drainage is associated with haemorrhagic presentation and descending venous drainage is associated with venous congestion of the spinal cord (6,7,10). The table, including our case, demonstrates that CCJAFSs with ascending venous drainage with the feeding artery from the RMA of the VA can also present isolated brainstem congestion.

Recognising the co-existence of dural AVF (DAVF) and intradural AVF (perimedullary AVF) is important for developing surgical strategies. In our case, we identified the coexistence of intradural AVF from the detailed interpretation of the preoperative CAG. We interrupted the abnormal feeding

Table I: The Summary of Reported Cases of Isolated Brainstem Lesion due to CCJAVFs

Patient No.	Age/ Sex	Congestion location	Feeding artery	Draining direction	Delayed-/mis-diagnosis	Treatment	Author	Year
1	46/M	Pons, Medulla	NMB of APhA	Descending	Yes	Emboli	Wiesmann	2000
2	68/M	Medulla	NMB of APhA	Descending	NA	Emboli	Wang HC	2009
3	72/M	Medulla, Flocculus	NMB of APhA	Bidirection	Yes	Emboli	Copelan	2018
4	76/M	Pons, Medulla, inferior cerebellar peduncle	NMB of APhA, branch of the OA petrosquamous branch of MMA, petrous branch of PAA	Descending	Yes	Emboli, OS	Roelz	2015
5	63/M	Medulla	OA, APhA	Descending	None	Emboli	Takahashi	2018
6	66/M	Pons, Medulla	OA, VA	Bidirection	Yes	Emboli	Chen	2019
7	38/M	Pons, Medulla	RMA of VA	Bidirection	Yes	OS	Wang XC	2018
8	53/M	Medulla	RMA of VA	Bidirection	Yes	OS	Wang XC	2018
9	54/F	Pons	RMA of VA	Ascending	None	OS	Kulwin	2012
10	46/F	Pons, Medulla	RMA of VA	Bidirection	None	Emboli	Wu	2014
11	64/M	Medulla	RMA of VA, PICA	Ascending	None	OS	Present Case	

VA: *Vertebral artery,* ***PAA:*** *Posterior auricular artery,* ***OA:*** *Occipital artery,* ***APhA:*** *Ascending pharyngeal artery,* ***MMA:*** *Middle meningial artery,* ***PICA:*** *Posterior inferior cerebellar artery,* ***RMA:*** *Radiculameningial artery,* ***NMB of APhA:*** *Neuromeningial branch of APhA,* ***PMA of VA:*** *Posterior meningeal artery,* ***NA:*** *Not applicable,* ***Emboli:*** *Endovascular embolization,* ***OS:*** *Open surgical disconnection*

artery during the operation, which led to improved curability. Recent reports have highlighted the coexistence of intradural AVFs and CCJAVFs (2,4,5), which may be more common than previously believed. Although accurate identification of the shunt site in the CCF area is often difficult, especially when AVFs contain microfistulas or coexist with high-flow shunts (3), it is important to not overlook coexisting intradural AVFs preoperatively to ensure the disappearance of the shunt point in the surgical procedure and prevent the postoperative recurrence of CCJAVFs.

CONCLUSION

Although rare, we should be aware of CCFAVFs that can cause isolated brainstem congestion when they have venous drainage around the brainstem, regardless of their direction.

AUTHORSHIP CONTRIBUTION

Study conception and design: SY

Data collection: SY, HO

Analysis and interpretation of results: SY

Draft manuscript preparation: SY

All authors (SY, HO) reviewed the results and approved the final version of the manuscript.

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