

## Subarachnoid hemorrhage due to a craniocervical junction arteriovenous fistula associated with thrombus formation in the internal jugular vein: illustrative case

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**BACKGROUND** A craniocervical junction arteriovenous fistula (CCJAVF) is a rare vascular malformation, and its etiology remains unclear. Here, to the best of the authors' knowledge, they present the first case of CCJAVF associated with thrombus formation in the ipsilateral internal jugular vein.

**OBSERVATIONS** An 80-year-old man presented with a sudden occipital headache. Computed tomography revealed a subarachnoid hemorrhage surrounding the brainstem and upper cervical cord. Digital subtraction angiography showed a CCJAVF fed by the left C2 radiculomeningeal artery with ascending intracranial drainage and epidural plexus. After endovascular treatment, the authors retrospectively found that his ipsilateral internal jugular vein and innominate vein were occluded with a huge thrombus at admission.

**LESSONS** This case suggested a restricted antegrade venous flow due to thrombus-induced progressive retrograde intracranial drainage causing hemorrhage. Venous hypertension should be considered one of the causes of hemorrhage due to CCJAVF as well as intracranial arteriovenous fistulas.

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**KEYWORDS** arteriovenous fistula; craniocervical junction; internal jugular vein; subarachnoid hemorrhage; venous thrombosis

A craniocervical junction arteriovenous fistula (CCJAVF) is a rare vascular malformation with various draining patterns.<sup>1–4</sup> Because of its rarity, the etiology remains uncertain, and the clinical course is sometimes unpredictable.<sup>5,6</sup> Herein we report a case of a subarachnoid hemorrhage (SAH) caused by CCJAVF associated with internal jugular vein (IJV) occlusion.

### Illustrative Case

An 80-year-old man presented with a sudden occipital headache with vomiting and was transferred to our institution. The patient's laboratory data indicated no coagulant disorders, and he had no history of trauma, cancer, or intravenous catheter placement. Computed tomography (CT) revealed an SAH surrounding the brainstem and upper cervical cord (Fig. 1). There were no positive findings in his initial neurological examination, but respiratory disturbance and tetraparesis appeared after admission and worsened rapidly without

abnormal signals in the spinal cord on magnetic resonance imaging (Fig. 2, left). No intracranial aneurysms were detected, but MR angiography demonstrated arterial dilatation adjacent to the craniocervical junction (Fig. 2, right). Digital subtraction angiography showed an AVF at the craniocervical junction fed by the left C2 radiculomeningeal artery with drainage to the epidural venous plexus (Fig. 3A–C). The fistula also had ascending intracranial drainage, and we recognized this draining vein as the cause of the SAH.

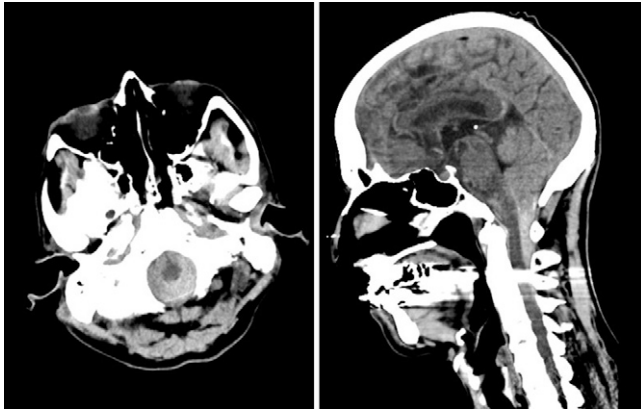
At first, we attempted to perform direct surgery to achieve radical and secure treatment, but the patient was intolerant of the prone and lateral position during the surgery because of severe aspiration pneumonia. The patient received transarterial embolization after his respiratory condition became stable. Under general anesthesia, a 7-Fr guiding catheter was navigated to the left vertebral artery via the femoral approach. After a Marathon microcatheter (Covidien) was introduced to dilated C2 radiculomeningeal artery, Scepter XC balloon (Microvention) was placed in the vertebral artery just distal to the radiculomeningeal artery. We could

**ABBREVIATIONS** AVF = arteriovenous fistula; CCJAVF = craniocervical junction AVF; CT = computed tomography; IJV = internal jugular vein; MR = magnetic resonance; SAH = subarachnoid hemorrhage.

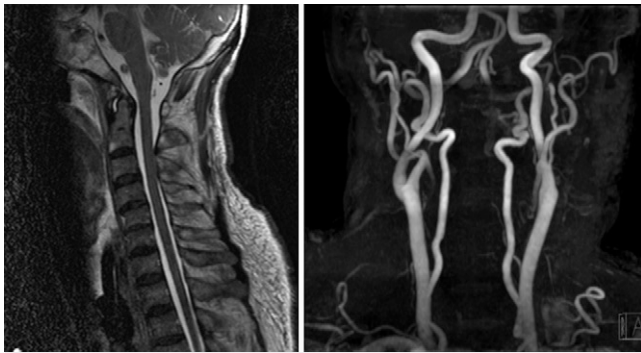
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**FIG. 1.** Initial axial (left) and sagittal (right) head CT scans show an SAH surrounding the medulla and upper cervical cord.



**FIG. 2.** Preoperative sagittal T2-weighted MR image of the cervical spine shows no abnormal signals suspicious for myelopathy or infarction on the spinal cord (left). MR angiography demonstrates arterial dilatation near the craniocervical junction (right).

not place the microcatheter distal site of the feeder because the feeder was tortuous for catheterization; therefore, the balloon was temporarily inflated during injection of 33% n-butyl cyanoacrylate glue to avoid glue

reflux to the vertebral artery. The intracranial drainage disappeared, and the shunt flow for extradural space was dramatically reduced after the glue penetrated adjacent to the shunting site (Fig. 3D).

The postoperative course was uneventful, and respiratory disturbance and tetraparesis improved significantly. Postoperative CT angiography showed no residual intracranial drainage although a huge thrombus was detected in the left IJV after surgery (Fig. 4A and B). We retrospectively found that the left IJV and left innominate vein were already occluded on admission (Fig. 4C and D). Anticoagulation therapy was initiated, and the thrombus gradually dissolved. The patient was completely recovered 3 months after surgery.

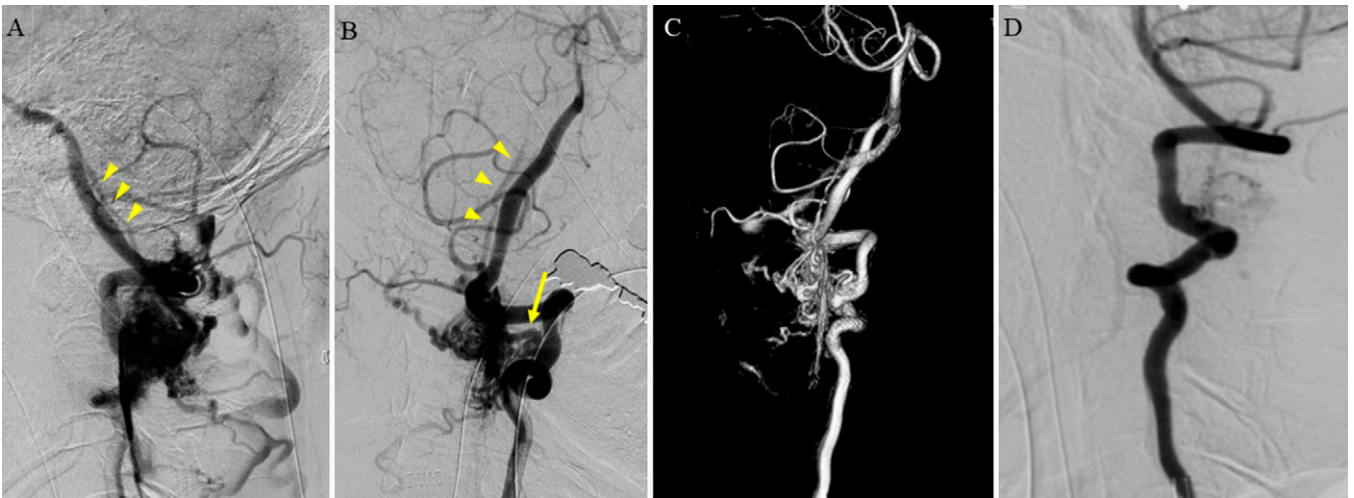
## Discussion

### Observations

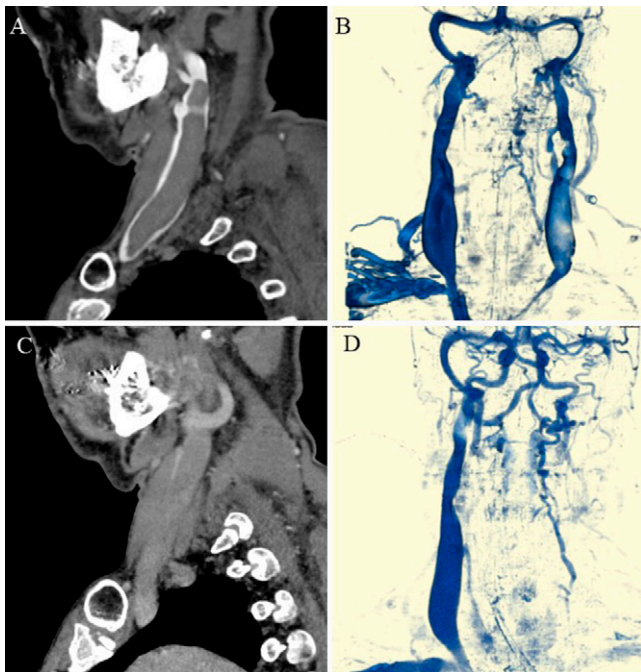
A craniocervical junction is a rare AVF lesion that accounts for 1% to 2% of intracranial or spinal AVFs<sup>1,4</sup> and mainly occurs in middle-aged men.<sup>1,2,6,7</sup> CCJAVFs have a wide range of clinical presentations, including acute SAH,<sup>8–11</sup> myelopathy,<sup>12</sup> intramedullary hemorrhage,<sup>2</sup> brainstem dysfunction,<sup>6,13–15</sup> and radiculopathy.<sup>3</sup> Our patient had respiratory disorders and tetraparesis after admission and completely recovered after treatment. The patient's upper cervical cord dysfunction was probably due to compression by the SAH at the craniocervical junction rather than myelopathy because there were no abnormal signals in the spinal cord.

CCJAVFs are usually fed by radiculomeningeal arteries and meningeal branches from vertebral arteries, occipital arteries, ascending pharyngeal arteries, and anterior spinal arteries.<sup>1,3,4,11,16</sup> They mainly drain into the medullary vein, coronal venous plexus, intracranial venous system, anterior spinal vein, and epidural plexus.<sup>1,3,4,11,17</sup> In previous reports, risk factors for bleeding, including intracranial drainage, varix, feeder aneurysm, and feeders from the anterior spinal arteries, have been mentioned.<sup>1–3,11,16</sup> Epidural drainage has been reported to infrequently cause a hemorrhage.<sup>2</sup>

The treatment for CCJAVF remains controversial. Most previous reports discussed direct surgery because the feeders were small and tortuous for endovascular embolization.<sup>3,4</sup> CCJAVFs fed by the occipital arteries or meningeal branches from the vertebral arteries are sometimes successfully treated by endovascular embolization;<sup>18</sup> however,



**FIG. 3.** Lateral (A), right oblique (B), and three-dimensional reconstruction (C). Digital subtraction angiography of the left vertebral artery shows CCJAVF from the radiculomeningeal artery (arrow) and ascending intracranial drainage (arrowheads) and drainers for epidural venous plexus. The postoperative image (D) reveals the disappearance of intracranial drainage and flow reduction of drainage for the venous plexus.



**FIG. 4.** Postoperative CT angiography demonstrates a huge thrombus formation in the left IJV (A and B). Retrospectively, the initial CT angiography shows occlusion of the left innominate vein and IJV (C and D).

Some intracranial AVFs are associated with trauma, inflammation, history of craniotomy, or dural sinus thrombosis.<sup>19,20</sup> Venous hypertension caused by sinus thrombosis is considered to restrict antegrade venous flow and lead to the opening of small capillary vessels or activation of vascular growth factors.<sup>20,21</sup> However, the mechanism underlying CCJAVF formation remains unclear because of its rarity. There are several reports of CCJAVFs following cervical spine fracture,<sup>22,23</sup> infection, and surgical procedure.<sup>23,24</sup> Several studies have suggested that venous hypertension may be associated with CCJAVF.<sup>2,17</sup> Another possible etiology is thrombosis or congestion of the internal dural vein due to bone fracture or postoperative fibrosis causing an abnormal shunt.<sup>22</sup> In the present case, SAH was probably caused by ascending intracranial drainage, but the AVF mainly drained into the epidural plexus. We assume that the thrombus in the IJV and innominate vein restricted the antegrade venous flow out because the external vertebral venous plexus at the cervical level plexus mainly joins the innominate vein. Paravertebral venous hypertension may have caused an increase in the abnormal shunt flow and bleeding from intracranial drainage.

To date, this case appears to be the first report of CCJAVF related to thrombus formation in the IJV; only four previous cases of intracranial AVFs after IJV occlusion or stenosis have been reported (Table 1).<sup>21,25–27</sup> The AVF site is relatively common in the transverse or sigmoid sinus. The interval from IJV occlusion or stenosis to AVF formation ranges widely (4 months to 5 years). In our case, the duration of the IJV occlusion was probably not long because the thrombus was not organized and diminished with anticoagulation therapy. Therefore, we assume that the AVF had already been formed before IJV occlusion; however, the thrombus in the IJV caused an increase in intracranial drainage flow and led to SAH.

misembolization of normal vessels can cause severe disabilities when the feeding arteries are from the radiculomeningeal artery or vertebral arteries.<sup>15</sup> In the current case, we were able to prevent the glue from flowing through the vertebral arteries with distal balloon protection.

### Lessons

To the best of our knowledge, this was the first case of SAH due to CCJAVF associated with thrombus formation in the IJV.

**TABLE 1. Summary of arteriovenous fistula associated with occlusion or stenosis of internal jugular vein**

Case No.	Authors & Year	Age (yrs)/ Sex	Initial Symptoms	Primary Findings on CT or MRI	Past History	IVJ Stenosis/ Occlusion	Duration*	Site of SVF	Treatment	Prognosis
1	Matsuyama et al., 1997 <sup>26</sup>	32/F	HA, pulsatile tinnitus	Mass in suprasellar region	Hormonal therapy for infertility	Stenosis	—	Lt TS-SS	TAE (PVA)	GR
2	Ngerageza et al., 2016 <sup>25</sup>	72/M	None (incidental diagnosis)	ND	Tongue cancer removal & ligation of IJV	Occlusion (ligation)	5 yrs	Lt TS	Direct surgery	GR
3	Fudaba et al., 2017 <sup>27</sup>	74/M	Unconsciousness	Cerebellar hemorrhage, epidural hematoma	Tongue cancer removal & ligation of IJV	Occlusion (ligation)	4 mos	Lt TS, rt SS (de novo dAVF at SSS & rt TS after 6 mos)	TVE (TAE for de novo dAVF)	GR
4	Suzuki et al., 2020 <sup>21</sup>	77/F	Pulsatile tinnitus	ND except for AVF	Head injury 3 mos before admission	Stenosis due to elongated styloid process	—	Rt hypoglossal canal (multiple shunt)	TVE & TAE	GR
5	Present case	80/M	Sudden occipital HA	SAH surrounding upper cervical cord	Nothing	Idiopathic occlusion	—	C2 dura	TAE	GR

GR = good recovery; HA = headache; MRI = magnetic resonance imaging; ND = not described; SS = sigmoid sinus; TAE = transarterial embolization; TS = transverse sinus; TVE = transvenous embolization; — = unknown.

\* Duration from IJV occlusion or stenosis.

Venous hypertension may have caused the fast retrograde venous flow of intracranial drainage and bleeding. Although the prevalence of CCJAVF is low, clinicians should be aware of the coexistence of venous thrombus associated with bleeding or venous congestion as well as intracranial AVFs.

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## Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

## Author Contributions

Conception and design: Morofuji, Shiozaki, Kawahara, Tsutsumi. Acquisition of data: Shiozaki, Kutsuna. Analysis and interpretation of data: Morofuji, Shiozaki, Uchida. Drafting the article: Morofuji, Shiozaki, Haraguchi. Critically revising the article: Ono, Tsutsumi. Reviewed submitted version of manuscript: Morofuji, Ono, Tsutsumi. Approved the final version of the manuscript on behalf of all authors: Morofuji. Study supervision: Tsutsumi.

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