The craniocervical junction is a complex structure including the lower cranial and upper spinal nerves; caudal brainstem and rostral spinal cord; VA and its branches; veins and dural sinuses around the foramen magnum; and ligaments and muscles uniting the atlas, axis, and occipital bone. Thus, AVFs at the craniocervical junction should belong to a category distinct from lesions in other spinal regions, given such peculiar characteristics as high-flow state, frequent hemorrhagic presentation, or association with other vascular anomalies. However, the etiology, pathogenesis, and natural course of AVFs at the craniocervical junction are poorly understood, because of their rarity.

Here, we describe 9 cases of concurrent DAVF and PAVF at the craniocervical junction, with the goal of elucidating the clinical and angiographic characteristics of such lesions. All patients underwent detailed selective angiography.
giography and open surgery. We confirmed angiographic findings in the operative field. The similarity of the angiographic architecture and the close relationship between DAVFs and PAVFs at the craniocervical junction led us to propose that these lesions are pathogenetically linked.

Methods

Arteriovenous fistulas at the craniocervical junction were defined as a communication between arterial feeders and a radicular or leptomeningeal vein with an identifiable shunt located between the foramen magnum and C-2 level. The shunting point was located at the site where the caliber of the vessels changed. All diagnoses were made using selective angiography.

A retrospective review of 24 consecutive cases with AVFs at the craniocervical junction, treated at our institute between January 2000 and December 2011, revealed 9 patients who fulfilled the study entry criteria of concurrent dural and intradural AVFs revealed operatively or angiographically. Our institutional review board did not require informed consent for participation in this study because our analysis relied on the information obtained as part of the routine clinical care of patients. The evaluations included a detailed medical history, full neurologic examination, imaging findings, intraoperative findings, and outcome 6 months after treatment.

Angiography Studies

All patients underwent selective angiography of internal and external carotid arteries, VAs, and thyrocervical and costocervical arteries. Evaluation of the angiographic images included feeding arteries, fistulas, drainage routes, and accompanying angiographic features such as aneurysms or other vascular diseases. Oblique views were systematically included in our protocol and were obtained at the best angle to disclose the fistulous compartments. We also performed rotational spin angiography to acquire 3D volume-rendering images by using an Advantage 4.4 workstation (GE Healthcare) for detailed understanding of the angioarchitecture.

Surgical Procedure

All patients in the series underwent a suboccipital craniotomy and C-1 laminectomy, followed by longitudinal linear incision of the dura mater. Somatosensory and motor evoked potentials were monitored throughout surgery. After dissecting the arachnoid membrane, lesions on the ventrolateral side were exposed by gently rotating the spinal cord, with dentate traction stitches in some cases. This technique provides exposure of the ipsilateral half of the ventral surface of the cord and anterior spinal artery. Intraoperative DSA or ICG videoangiography was performed to delineate intraoperative vascular anatomy. Perimedullary arteriovenous fistulas as well as associated aneurysms were coagulated and dissected, and the draining veins of DAVFs were interrupted at the point where these vessels entered the intradural space. Intraoperative angiograms were assessed to confirm obliteration of the fistulous connection and restoration of normal blood flow.

Follow-Up Analysis

In the follow-up examination, radiological studies were performed using either MRI or DSA 6 months after the last surgical intervention. Thereafter, clinical and MRI follow-up were continued annually. The mean follow-up period among all 9 patients was 38.4 months (range 12–96 months). The Glasgow Outcome Scale score and the incidence of recurrent hemorrhage were recorded.

Illustrative Cases

Case 1

A 59-year-old man presented with SAH 1 month before admission to our institute. His Glasgow Coma Scale score was 15, with no focal neurological deficit upon arrival at our hospital. Right VA angiography demonstrated a DA VF and PAVF at the craniocervical junction (Fig. 1). Venous reflux from the DAVF drained totally into the right C-1 radicular vein joining the anterior spinal vein, directing rostrally and caudally. The PAVF, which was fed by the anterior spinal artery and the radiculopial artery arising at the C-2 level of the right VA and penetrating the dura at the C-1 level, drained into the same radicular vein. Aneurysmal dilations were found on the radiculopial feeding artery in the vicinity of the PAVF. Surgical intervention consisted of suboccipital craniotomy with C-1 laminectomy. After gentle rotation of the spinal cord with dentate traction stitches, the PAVF was seen on the right anterolateral surface of the cord (Fig. 2A–D). The abnormal arteriovenous connection and aneurysms with a fibrin cap were coagulated and dissected. After dissecting the PAVF, an abnormal dilated radicular vein, emerging from the dura just inferior to the point at which the VA penetrated, was identified by an arterial flow pattern on ICG videoangiography (Fig. 2E and F). After electrocoagulation and transection of the origin from the dura mater, complete occlusion of the DAVF was verified by ICG videoangiography. Postoperative angiography showed complete disappearance of both the DAVF and PAVF. The patient returned to normal activities of daily life without any neurological deficits.

Case 4

A 62-year-old man experienced the sudden onset of severe headache. On admission, his Glasgow Coma Scale score was 14 (E3V5M6), and physical examination revealed no focal neurological deficit. Computed tomography showed SAH localized mainly in the posterior fossa. A DA VF, revealed on right VA angiography, drained into the anterior spinal vein via the right C-1 radicular artery (Fig. 3A and B). Left VA angiography demonstrated a PAVF fed by the anterior spinal artery at the level of the craniocervical junction (Fig. 3C). The PAVF drained into the same right C-1 radicular vein. A suboccipital craniotomy with a C-1 laminectomy revealed that the PAVF was situated on the right anterolateral surface of the cord, and the abnormal arterialized radicular vein originated from the dura just inferior to the point at which the right VA penetrated the dura (Fig. 3D and E). Indocyanine green videoangiography confirmed this angioarchitecture (Fig.
3F). The PAVF was coagulated and dissected, and the arterialized draining vessel was then coagulated and interrupted. The patient’s postoperative course was uneventful, and angiographic studies obtained 6 months after surgical treatment demonstrated complete disappearance of the shunts (Fig. 4).

**Results**

Clinical and angiographic characteristics in this series are summarized in Table 1. Nine cases were identified in 5 men and 4 women. The mean age was 66.3 years with a range of 59–82 years. Patients reported no history of neck trauma or disease, such as fibromuscular dysplasia or neurofibromatosis. All cases presented with an SAH classified as Hunt and Hess Grade II.8

Radiological findings revealed 7 cases of concurrent DAVF and PAVF on the same side (right, 5 cases; left, 2 cases) and at the same spinal level (C-1 level, 6 cases; C-2 level, 1 case), and 1 case (Case 2) of DAVF at the right C-2 level and PAVF at the right C-1 level. Another patient (Case 6) had 3 AVFs, a DAVF, and a PAVF located at the right C-1 level as well as a DAVF at the left C-2 level. Seven (70%) of 10 DAVFs were located at the C-1 level, exactly where the VA penetrates the dura. No contribution to the DAVF was found from the external carotid, thyrocervical, or costocervical branches. All DAVFs except one were fed by a reticular network of radiculomeningeal arteries mainly originating from an enlarged radicular artery arising at the same side and same spinal level. In the patient in Case 3, the DAVF at the C-1 level was fed by radiculomeningeal arteries mainly originating from an enlarged radicular artery at the C-2 level. All PAVFs were located on the ventrolateral surface of the cord. The feeding vessels of the PAVFs were both the radiculopial arteries and the anterior spinal artery in 8 (88.9%) of 9 cases and the radiculopial arteries alone in 1 case. The DAVFs drained into the anterior or anterolateral spinal vein via a radicular vein, and the main drainage route was rostrally directed in 5 cases (55.6%), bidirectionally in 3 (33.3%), and caudally in 1 (11.1%). All of the concurrent DAVFs and PAVFs shared this main drainage route, in which shunted blood flow was directed from the DAVF toward the PAVF, with 1 exception. In the patient in Case 6, the right VA angiography showed that the shunted blood flow from the DAVF and PAVF at the right C-1 level drained into the anterior spinal vein in the rostral direction and, at the same time, caudally before connecting to the left C-2 radicular vein. Left VA angiography demonstrated that the second DAVF developed at the point at which the left C-2 radicular vein crossed the dura. Shunting blood flow from the left C-2 DAVF drained into the epidural venous plexus with little reflux to the radicular vein.
Open surgery was performed about 1 month after the onset of SAH in all cases. Ruptured arterial aneurysms were detected in 8 (88.9%) of 9 cases. In the patient in Case 7, ruptured venous ectasia was detected at the perimedullary fistulous point. The bleeding point existed in the vicinity of the PAVF rather than the DAVF in all cases. Histological evaluation of the aneurysms demonstrated an arterial wall without evidence of infection or collagen disease.

The postoperative course was uneventful, with no neurological deficit in any of the patients. No recurrence was observed in the 4 patients who underwent postoperative angiography, and no rebleeding event was observed during the follow-up period in all 9 patients.

Figure 5 illustrates the angioarchitecture of each lesion, presenting the best exposure view of the angiography with reference to intraoperative findings.

Discussion
In this study we described 9 cases of concurrent DAVF and PAVF at the craniocervical junction. The cases shared 3 key clinical and angiographic characteristics: coexistence of DAVF and PAVF on the same side, common drainage route directing ventrally, and hemorrhagic presentation. The PAVFs were always located on the anterolateral surface of the spinal cord. The bleeding point was found in the vicinity of the PAVFs. These findings indicate that common pathomechanical factors can play a role in the development of these peculiar angioar-
Multiple AVFs at the craniocervical junction

Architecture. Thus, we believe that these cases should be grouped, studied, and discussed together.

Spinal cord arteriovenous malformations represent a heterogeneous group of vascular anomalies. These lesions have been classified into 4 types: Type I, AVF between a dural branch of the spinal ramus of a radicular artery and an intradural medullary vein (known as DAVFs); Type II, intramedullary glomus malformations; Type III, extensive juvenile malformations, often extending to paraspinal structures; and Type IV, intradural PAVFs. Type IV lesions have been further classified into 3 subtypes: Type IVa, simple extramedullary fistulas fed by a single arterial branch; Type IVb, intermediate-sized fistulas with multiple, dilated arterial feeders; and Type IVc, giant multipediculated fistulas.

Dural Arteriovenous Fistulas at the Cranio-Cervical Junction

Spinal DAVFs are known to be acquired lesions located inside the dura mater close to the spinal nerve root where the arterial blood from a radiculomeningeal artery enters a radicular vein. Most shunts are found in the thoracolumbar region, whereas high cervical lesions (at the level of the foramen magnum) occur in 2% of patients. Cranio-Cervical DAVFs are distinguished by a propensity to cause SAH, which occurs in 34%–45% of cases.

The mechanism underlying the hemorrhagic presentation of cranio-cervical DAVF is presumed to involve venous hypertension, but it is not entirely understood. Aviv et al. noted that the presence of varices of the draining veins was significantly more common in patients with...
craniocervical DAVFs with hemorrhagic presentation than in patients with a nonhemorrhagic presentation. Kai et al.\textsuperscript{10} concluded that cephalad or intracranially directed venous drainage was significantly associated with hemorrhagic presentation.

Despite increased pressure, the flow in drainage veins is generally slow in spinal DAVFs. However, accelerated venous blood flow has been observed in spinal DAVFs with a hemorrhagic presentation. The rate of arteriovenous circulation was faster (1.0 seconds) than in patients presenting with congestive myelopathy (1.5–2.0 seconds).\textsuperscript{13} In the craniocervical junction, the relief of venous drainage into collateral veins of the posterior fossa (that is, ascending or rostral venous drainage) can accelerate drainage flow enough to induce high-flow venopathy of the draining vein (for example, venous ectasia with varix formation\textsuperscript{4,10,12} and/or venous steal effect\textsuperscript{16,17}). The presence of varices may be a consequence (“a symptom”) of the high-flow venopathy and not necessarily a cause of hemorrhage. This idea is supported by findings in our current study, in which 8 patients had a bleeding point in an arterial aneurysm arising from the feeding artery to the PAVF, not in a varix.

Perimedullary Arteriovenous Fistulas at the Craniocervical Junction

Direct communication between an enlarged spinal artery and vein distinguishes PAVFs, which compose approximately 10%–20% of all spinal arteriovenous shunts. The fistulous communication is always in the pia mater on the surface of the spinal cord.\textsuperscript{5} The anatomical distribution of the shunt along the long axis of the spine is bimodal, predominantly at the conus medullaris and, to a lesser extent, in the upper cervical region.\textsuperscript{2}

Perimedullary arteriovenous fistulas have been considered congenital in nature, although there is little evidence that diagnosed lesions in adults are present in the same form at birth.\textsuperscript{18} In addition, some cases of de novo PAVF have been reported in the brain\textsuperscript{9,16,24,25,27,31} and spine.\textsuperscript{33} Patients with acquired PAVFs in moyamoya disease\textsuperscript{31} and after cerebral vein thrombosis\textsuperscript{24,25} suggest that tissue hypoxia is an important etiological factor. Hypoxia is a powerful trigger for the upregulation of angiogenic factors.\textsuperscript{32}

Association of DAVFs and PAVFs

Concurrence of DAVFs and PAVFs is exceedingly rare in the spine, although the incidence of concurrent spinal DAVFs is presumed to be approximately 2%.\textsuperscript{15,26} There are only 3 published reports of spinal DAVF associated with spinal PAVF.\textsuperscript{3,14,23} All of these lesions were located in the thoracolumbar region, and all patients presented with congestive myelopathy. However, Kim et al.\textsuperscript{11} reported that 5 of 12 patients showed an additional complex vascular abnormality associated with a cervical spinal DAVF. Coincidental lesions consisted of a spinal/brain arteriovenous malformation (2 cases) or a dural/epidural AVF (3 cases), including a case of DAVF at the craniocervical junction with a coincident PAVF. This

<table>
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<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
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<th>Varix</th>
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<td>bidirectional</td>
<td>ASV</td>
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</tr>
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<td>78, M</td>
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<td>ALSV</td>
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<td>lt C-1</td>
<td>RPA, ASA</td>
<td>rostral</td>
<td>ALSV</td>
</tr>
</tbody>
</table>

* All patients presented with SAH, and all patients had a good recovery. Abbreviations: AA = arterial aneurysm; ALSV = anterolateral spinal vein; ASA = anterior spinal artery; ASV = anterior spinal vein; RPA = radiculopial artery.

Fig. 4. Case 4. Right (left) and left (right) VA angiograms, antero-posterior views, obtained 6 months after the operation, showing the disappearance of the AVFs and abnormal veins.
PAVF drained into the rostral radicular vein shared by the DAVF and harbored a ruptured aneurysm, similar to findings in our study. Lastfogel et al.\textsuperscript{19} described a case of cervical DAVF in a patient presenting with hemorrhage, which was intraoperatively shown to have another smaller fistulous connection.

These recent reports, as well as our current study, highlight the fact that craniocervical DAVFs and PAVFs may coexist more frequently than generally thought and suggest that the incidence of concurrent AVFs is underestimated, because complete spinal or cerebral angiography is not systematically performed.\textsuperscript{3} It is reasonable to conclude that early progression to comprehensive vascular imaging studies in the diagnostic algorithm may be indicated to assess for the presence of such simultaneous lesions. Our study underscores the importance of performing complete craniocervical angiography to delineate the angioarchitecture of the lesions in detail.

Possible Pathogenesis of Concurrent DAVFs and PAVFs

The relationship between the 2 different AVFs described herein remains unclear. We were unable to dis-
cern whether their simultaneous occurrence was purely a coincidence. The possible pathomechanisms of concurrent DAVFs and PAVFs are as follows.

**Dural Arteriovenous Fistulas Inducing PAVFs.** Venous hypertension and/or increased flow of blood into the low-resistance venous system can promote the growth of microscopic arteriovenous shunts, which are found within the vasavorum of normal parenchymeninges, and stimulate the release of angiogenic factors in experimental models and in intracranial lesions. Because these induced PAVFs seem to be located preferentially at certain parts of affected perimedullary veins, there may be additional regional factors involved. Anatomical factors and hemodynamic situations in the vicinity of the affected perimedullary vein dictate the location of these induced PAVFs, which form upstream from the high-flow DAVFs. The induced PAVFs can cause secondary arterial changes, such as an aneurysm related to flow in the shunt, formed in the arterial feeder supplying the shunts. Given that DAVFs at the craniocervical junction are probably embryological homologs of the intracranial DAVFs draining into the petrosal vein or bridging veins of the posterior cranial fossa, we postulate that relatively high-flow DAVFs cause a venous steal effect or venous hypoxia due to venous hypertension that, in a suitable combination of anatomical and hemodynamic factors, induces PAVF at the craniocervical junction.

**Perimedullary Arteriovenous Fistulas Inducing DAVFs.** The possibility of PAVFs inducing DAVFs has been proposed, particularly in some high-flow PAVFs associated with DAVFs upstream from their drainage in the cranium. However, this is not the situation in our series, which involves high-flow DAVFs closely related anatomically to the PAVFs upstream from the DAVFs. The dominant shunts in our cases were DAVFs rather than PAVFs.

**Spinal Arteriovenous Metameric Syndrome.** Rodesch et al. classified spinal cord arteriovenous malformations into 3 main groups according to their morphological features: genetic hereditary lesions, genetic nonhereditary lesions, and single lesions. Patients with genetic nonhereditary lesions exhibited shared metameric links, typically present with multiple shunts of the spinal cord, nerve root, bone, and paraspinal, subcutaneous, and skin tissue. Although 8 patients in our series had DAVFs and PAVFs at the same metameric levels, no patient had other arteriovenous malformations in bone or connective or skin tissue.

**Treatment of AVFs**

Although the possible treatment for AVFs at the craniocervical junction is either surgical or endovascular, open surgery has been predominantly chosen. For transarterial embolization, the feeding arteries are usually small and tortuous and arise directly from the VA, presenting a high risk of embolic complication. For transvenous embolization, there is no feasible venous access route to the small draining veins of this type of fistula. On the other hand, microsurgical electrocoagulation and disconnection of the fistulas are regarded as an effective as well as the most reliable method of treatment. A microsurgical procedure is also a great advantage in detecting a bleeding point in the operative field. For patients who present with hemorrhagic, coincidental vascular lesions, the main goal of treatment should always include, first of all, eradication of the symptomatic lesion.

**Study Limitations**

Limitations of our study include the small number of patients and its retrospective approach. Our database of AVFs at the craniocervical junction may include a higher proportion of cases presenting with hemorrhage than in previous reports, because our institutes predominantly manage patients with hemorrhagic stroke. Future evaluations using a large number of patients are warranted to address the characteristics of these peculiar lesions.

**Conclusions**

We described 9 patients with concurrent DAVFs and PAVFs at the craniocervical junction who had presented with SAH. The similarity of these fistulas’ angiographic appearance leads us to propose that common pathogenetic mechanisms may exist. Our series underscores the importance of meticulous angiographic investigation in cases of suspected AVFs at the craniocervical junction. We believe that the pathophysiological mechanisms and anatomical features of these lesions represent unique situations that must be recognized angiographically to plan appropriate management.

**Disclosure**

The authors report no conflicts of interest concerning the materials or methods used in this study or the findings specified in this paper. Author contributions to the study and manuscript preparation include the following. Conception and design: Sato, Tominaga. Acquisition of data: all authors. Analysis and interpretation of data: Sato, Tominaga. Drafting the article: Sato. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Sato. Administrative/technical/material support: Sato, Endo. Study supervision: Tominaga.

**References**

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