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Asymptomatic spinal dural arteriovenous fistulas: pathomechanical considerations

Clinical article

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Object. Spinal dural arteriovenous fistulas (SDAVFs) consist of a shunt with converging feeding vessels arising from radiculomeningeal arteries and draining retrogradely via a radicular vein into the perimedullary veins, thereby causing progressive myelopathy due to venous hypertension in the spinal cord. The purpose of this study was to evaluate the hypothesis that the obstruction of radicular venous outlets could be an additional factor inducing symptomatic venous hypertension due to a decreased outflow in SDAVFs.

Methods. The authors compared the clinical and imaging findings in patients with asymptomatic SDAVFs identified incidentally at the upper thoracic region with the findings in symptomatic patients who harbored SDAVFs at the same level.

Results. All symptomatic patients presented with medullary dysfunction. The mean age of patients with asymptomatic SDAVF was 51.5 years, approximately 10 years younger than the patients with symptomatic SDAVF (64.1 years old). Despite the existence of dilated perimedullary vessels in the dorsal side of the spinal cord in all patients, the spinal cord edema seen in symptomatic patients was not detected on the MR images obtained in patients with asymptomatic SDAVF. The spinal angiograms of the asymptomatic patients distinctively demonstrated early radicular venous outflow from affected perimedullary veins to the extradural venous plexus as a potential alternate route for the venous hypertension to be released.

Conclusions. Obstruction of the radicular venous outflow could be an important factor in inducing spinal congestive edema due to venous hypertension, as well as subsequent clinical symptoms of SDAVFs.
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Key Words • **dural arteriovenous fistula** • **radicular vein** • **spinal cord** • **symptom** • **venous outflow** • **vascular disorder**

Spinal dural arteriovenous fistulas are the most frequent vascular malformation in the spine and account for approximately 70% of all spinal vascular malformations.[8,17] At least 80% of patients are male, and more than 66% of patients are in their 6th or 7th decade of life, indicating a preponderance of male and older patients.[6] Generally thought to be an acquired lesion, this fistula is located within the dura mater where the arterial blood from radiculomeningeal arteries enters a radicular vein.[10,11,13] The arterialized venous system will lead to a decrease in drainage of the normal spinal veins and an increase in spinal venous pressure, and thus to venous spinal congestion and myelopathy.[10,11,13] Therefore, the clinical presentation of SDAVFs nearly always includes symptoms of medullary dysfunction.[2,19] Although the pathomechanism by which these shunts form has not been elucidated, it has been hypothesized that once a shunt is present, the progressive fibrosis or thrombosis of radicular venous outlets could be an important factor inducing venous hypertension due to a decreased outflow.[3] However, this concept has never been proved in vivo given that until now no asymptomatic patients with SDAVF have been reported.

Here, we evaluated this hypothesis by investigating 2 patients with asymptomatic SDAVF at the upper thoracic level, in whom we were able to demonstrate early radicular venous outflow as a potential alternate route for the venous hypertension to be released. We compared clinical and radiological features between these asymptomatic patients and all patients in our database who had a fistula located at the same region to assess the pathomechanism of SDAVFs and to determine the imaging characteristics of a symptomatic lesion.

Abbreviations used in this paper: ASA = anterior spinal artery; SDAVF = spinal dural arteriovenous fistula.

Methods

Institutional review board approval was obtained along with a waiver of informed consent given the retrospective nature of the study.

A retrospective review of 32 consecutive patients with SDAVFs, who had been admitted to Toronto Western Hospital since 2004, revealed 10 patients with fistulas located in the upper thoracic region (T1–6). Eight of 10 patients presented with progressive neurological symptoms, while SDAVFs in 2 patients were incidentally identified through neurological investigations conducted for unrelated reasons. Evaluations included a detailed medical history, full neurological examination, and imaging findings. All diagnoses were confirmed with digital subtraction angiography.

Digital subtraction angiograms were evaluated for feeders, shunt location, drainage route, and circulation time through the ASA. The T2-weighted MRI studies were evaluated for the location and range of abnormal flow void signals, as well as cord signal abnormalities indicating congestive edema. Patient age, sex, and clinical presentation were assessed.

Illustrative Cases

Case 9

This 50-year-old woman underwent contrast-enhanced neck CT for assessment of a known follicular lymphoma. Computed tomography revealed prominent, enhancing serpiginous vessels along the posterior aspect of the cervical spinal cord. She had no neurological deficit.

On T2-weighted MR imaging, serpentine perimedullary hypointense signals were documented, indicating dilated perimedullary veins posterior to the spinal cord from the C-5 to the T-3 level (Fig. 1A). There was no evidence of an intramedullary nidus and no abnormal signal of the cord to suggest venous congestion. Spinal angiography was performed with the injection of contrast agent into the cervical and thoracic segmental arteries bilaterally. After injection of the common trunk of the T-3 and T-4 segmental arteries on the left side, we observed a shunt without an intervening nidus. This shunt was fed by the radiculomeningeal artery of T-3 and drained into the radicular vein and then into enlarged and tortuous posterior perimedullary veins, ascending up to the cervical region where they drained into the epidural sinus via a widely patent left C-2 radicular vein (Fig. 1B–D). When injecting the right vertebral artery, supply to the ASA was documented. There was no evidence for stagnation of contrast in the late phases of this angiographic run, indicating normal arteriovenous transit time of the cord supply.

Case 10

A 52-year-old man was admitted to our hospital with multiple complaints of diffuse back pain and headache. On examination, he had no neurological deficit.

Sagittal T2-weighted MRI revealed prominent perimedullary vessels seen over the dorsal aspect of the spinal cord from C-4 to the upper thoracic region (Fig. 2A). There was no evidence for congestive edema within the cord. On spinal angiography, injection of the right T-4 segmental artery demonstrated a shunt at the level of the right T-4 pedicle supplied by the radiculomeningeal artery (Fig. 2B). This shunt drained into the radicular vein and then into markedly enlarged and tortuous posterior perimedullary veins, which extended cranially up to the cervical spine and inferiorly toward the lower thoracic region (Fig. 2C). Early venous outflow to the epidural plexus at the upper cervical region was documented (Fig. 2D).

Results

The characteristics of patients in this study are summarized in Table 1. Eight cases with symptomatic SDVFs at the upper thoracic region (T1–6) were identified in our database. Only men, whose mean age was 64.1 years (range 48–83 years), were afflicted with symptomatic DAVF. Symptoms included paraparesis at the lower limbs (7 patients), sphincter disturbance (5 patients), and intolerable numbness at the lower limbs (1 patient). The mean interval between the onset of symptoms and a conclusive diagnosis was 19.8 months. One man and 1 woman, with a mean age of 51.5 years, had asymptomatic SDAVFs at the same region.

Abnormal hyperintensity ranging from the lower thoracic region to the conus in the spinal cord was present on T2-weighted MRI in all patients with symptomatic SDAVF. There was no intramedullary abnormality in the spinal cord of either of the patients with asymptomatic SDAVF.

Spinal angiography revealed shunts at the level of T-1 (2 cases), T-3 (1 case), T-4 (2 cases), T-5 (2 cases), and T-6 (3 cases) in the right (7 cases) or left side (3 cases). Stasis of contrast material in the radiculomedullary arteries, especially the ASA, in the venous phase, indicating venous congestion of the spinal cord, was seen in all symptomatic patients in whom segmental arteries supplying the ASA were injected (6 of 8 cases) but not in the asymptomatic patients. The main intradural venous drainage routes were located at the dorsal site of the cord in all cases in this study. However, in the symptomatic cases, affected perimedullary veins tended to drain caudally (4 of 8 cases), whereas the shunt flow mainly drained into ascending perimedullary veins in the asymptomatic cases. Extradural drainage through radicular veins at the cervical region was documented in both asymptomatic cases but was not found in the symptomatic cases. Figure 3 features radiological images of an illustrative case with symptomatic SDAVF at the upper thoracic region.

Discussion

In this study, we compared the radiological and clinical features between symptomatic and asymptomatic patients with SDVAFs in the same spinal region. Unlike those with symptomatic SDAVFs, patients with asymptomatic SDAVFs did not present with spinal cord edema caused by venous hypertension, despite the existence of dilated perimedullary veins in the dorsal site of the spine. Their shunts drained mainly into ascending perimedullary veins, which opened to the external paravertebral ve-

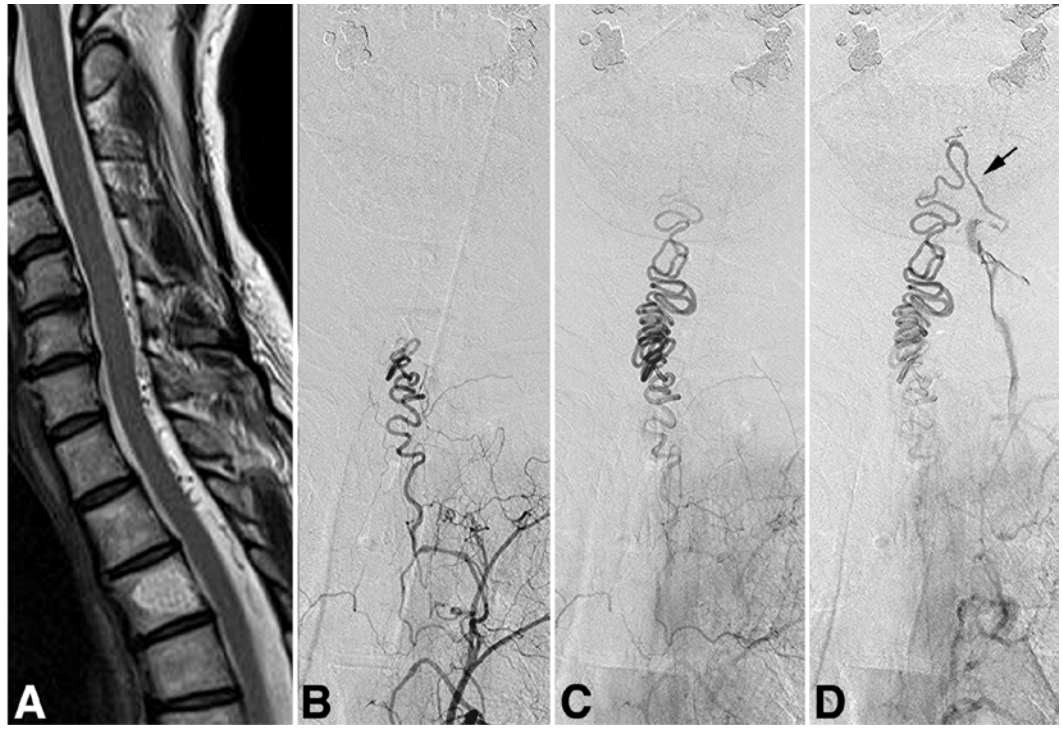

Fig. 1. Case 9. Sagittal T2-weighted MR image **(A)** of the cervicothoracic spine demonstrating abnormal flow void signals in the dorsal subarachnoid space from the C-5 to the T-3 level. There is no abnormal signal in the spinal cord. Anteroposterior angiograms of the left T3–4 segmental artery demonstrating a shunt fed by the T-3 radiculomeningeal artery **(B)**, draining via a radicular vein into ascending perimedullary veins **(C)**, which opened to the external paravertebral plexus via the left C-2 radicular vein (**D**, *arrow*).

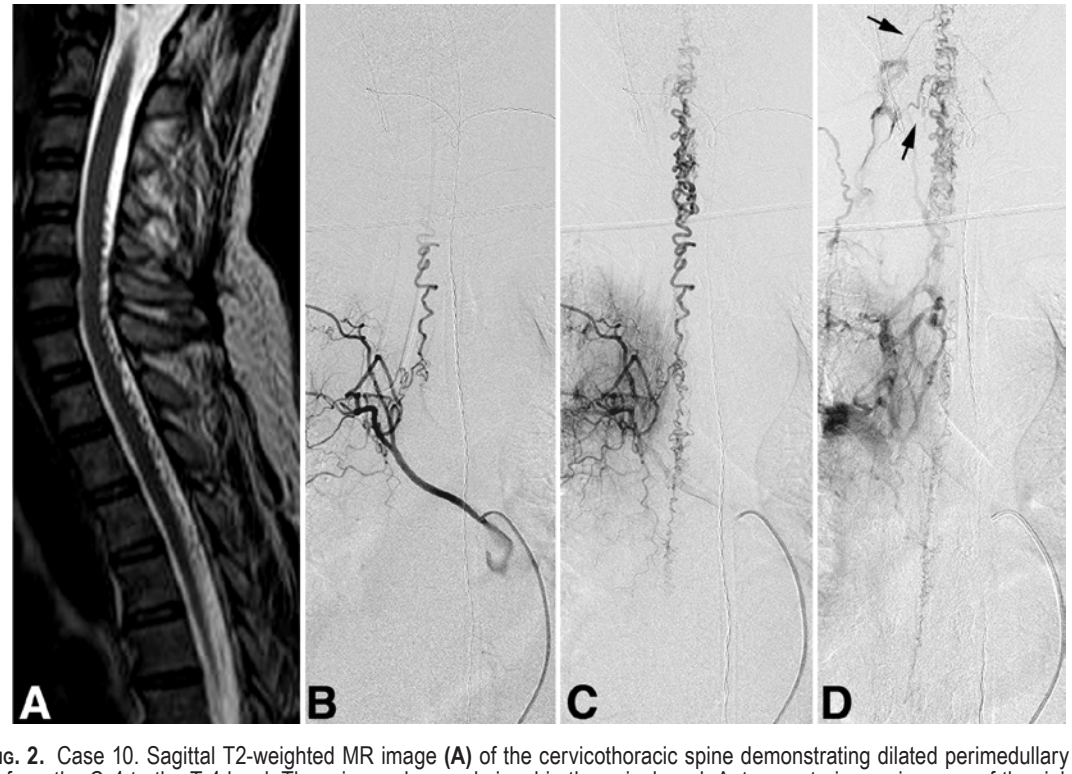

Fig. 2. Case 10. Sagittal T2-weighted MR image **(A)** of the cervicothoracic spine demonstrating dilated perimedullary vessels from the C-4 to the T-4 level. There is no abnormal signal in the spinal cord. Anteroposterior angiograms of the right T-4 segmental artery revealing shunting of the right T-4 radiculomeningeal artery into a radicular vein **(B)**, which drained mainly into cranially ascending as well as caudally descending perimedullary veins **(C)**. The cranially ascending perimedullary veins opened to the external paravertebral plexus via the right C-2 and C-4 radicular veins (**D**, *arrows*).

TABLE 1: Summary of characteristics in 10 patients with SDAVFs at the upper thoracic levels of T1–6*

Case No.	Age (yrs), Sex	Symptom	Duration (mos)†	T2W Imaging			Shunt Level	Feeder		ASA Stagnation	Draining PMV				Extradural Drainage	
				Yes/No	Top	End		Site	No.		Ventral/Dorsal	Direction‡	Top	End	Yes/No	Level
symptomatic																
1	48, M	paraparesis, sphinc dist	3	yes	T-7	T-11	T-1	lt	1	yes	dorsal	asc/desc	C-2	L-1	no	
2	66, M	numbness	2	yes	T-8	L-1	T-6	rt	1	yes	dorsal	desc	T-6	T-12	no	
3	58, M	paraparesis	60	yes	T-7	T-10	T-6	rt	2	yes	dorsal	asc/desc	T-5	T-10	no	
4	71, M	paraparesis, sphinc dist	24	yes	T-8	T-12	T-6	rt	1	yes	dorsal	desc	T-6	T-11	no	
5	73, M	paraparesis, sphinc dist	12	yes	T-6	L-1	T-5	rt	1	NA	dorsal	desc	T-4	L-1	no	
6	83, M	paraparesis	30	NA§	NA	NA	T-1	rt	1	yes	dorsal	asc/desc	T-1	T-8	no	
7	55, M	paraparesis, sphinc dist	9	yes	T-8	L-1	T-5	lt	2	yes	dorsal	asc/desc	C-1	T-8	no	
8	59, M	paraparesis, sphinc dist	18	yes	T-12	L-1	T-4	rt	1	NA	dorsal	desc	T-3	L-1	no	
asymptomatic																
9	50, F	none	0	no			T-3	lt	2	no	dorsal	asc	C-5	T-3	yes	C-2
10	52, M	none	0	no			T-4	rt	2	no	dorsal	asc/desc	C-4	T-4	yes	C-2, C-4

* asc = ascending; desc = descending; NA = data not available; PMV = perimedullary vein; sphinc dist = sphincter disturbance; T2W = T2-weighted.
† Interval between the onset of initial symptoms and time of diagnostic angiography.
‡ Direction of draining perimedullary veins is described as ascending, descending, or bidirectional (asc/desc).
§ Data not available because of previous laminoplasty with syringosubarachnoid shunt placement.

nous plexus via the upper cervical region, as an alternate venous outflow route; in symptomatic patients no drainage into radicular veins was visualized. In addition, there was no evidence of contrast stagnation in the ASA in the asymptomatic patients, whereas in the symptomatic patients contrast material was seen to linger in the ASA into the late venous phase. These findings indicated that the symptomatic venous congestion due to venous hypertension might be induced by the obstruction of alternate venous outflow routes from the arterialized perimedullary veins.

It is presumed that SDAVF is an acquired disease, although the exact etiology is not known. Tadié et al.[23] performed microangiography studies of normal radicular veins and demonstrated constriction at the point at which the veins crossed the dura. The constriction acts as a valve that prevents the transmission of pressure into the intradural venous system. The fistula has to be located at or beyond this point, which has been confirmed histologically.[1] Once a fistula has formed within the dura, increased medullary venous pressure is transmitted through the coronal venous plexus into the cord via the radial veins. This will lead to an increase in spinal venous pressure that in turn diminishes the arteriovenous pressure gradient and leads to decreased drainage of the normal spinal veins and thus to venous congestion with intramedullary edema.[5,7] This congestion results in chronic hypoxia and progressive myelopathy.[14] Merland et al.[17] and Hassler et al.[4] expanded on this theory and described additional insufficient venous drainage in symptomatic SDAVFs due to a lack of normal draining radicular veins.

It is believed that the number of radicular veins diminishes due to progressive fibrosis or thrombosis related to aging, a process that may be accelerated in the presence of an arteriovenous shunt.[16] Two arguments support this assumption: in cranial DAVFs angiographic changes can occur with progressive and spontaneous occlusion of previously patent venous outflows.[9,21] In addition, the presence of 1 or 2 thrombosed and fibrotic emissary veins in the spine was previously reported during routine dissection.[18] This peculiar arrangement of the spinal venous system can create an additional natural obstacle in cases of abnormally arterialized radicular veins. Spinal DAVFs, which drain into the longitudinal venous system, could worsen this anatomical arrangement and produce retrograde congestion within the cord.[16] The reflux in these lesions may create an irreversible hemodynamic situation that can lead to further stagnation and thrombosis in the venous system. This progressive venous outflow obstruction was also reported in intracranial DAVFs,[21,22] and recent data demonstrated that 16.1% of DAVFs, which were followed up without treatment, presented spontaneous angiographic conversion mainly due to venous outflow occlusion.[9] Thus, in the present study, it may be of interest that the average age of patients with asymptomatic SDAVFs with maintained venous outflow was approximately 10 years younger than the age of patients with symptomatic SDAVFs, all of whom lost their venous out-

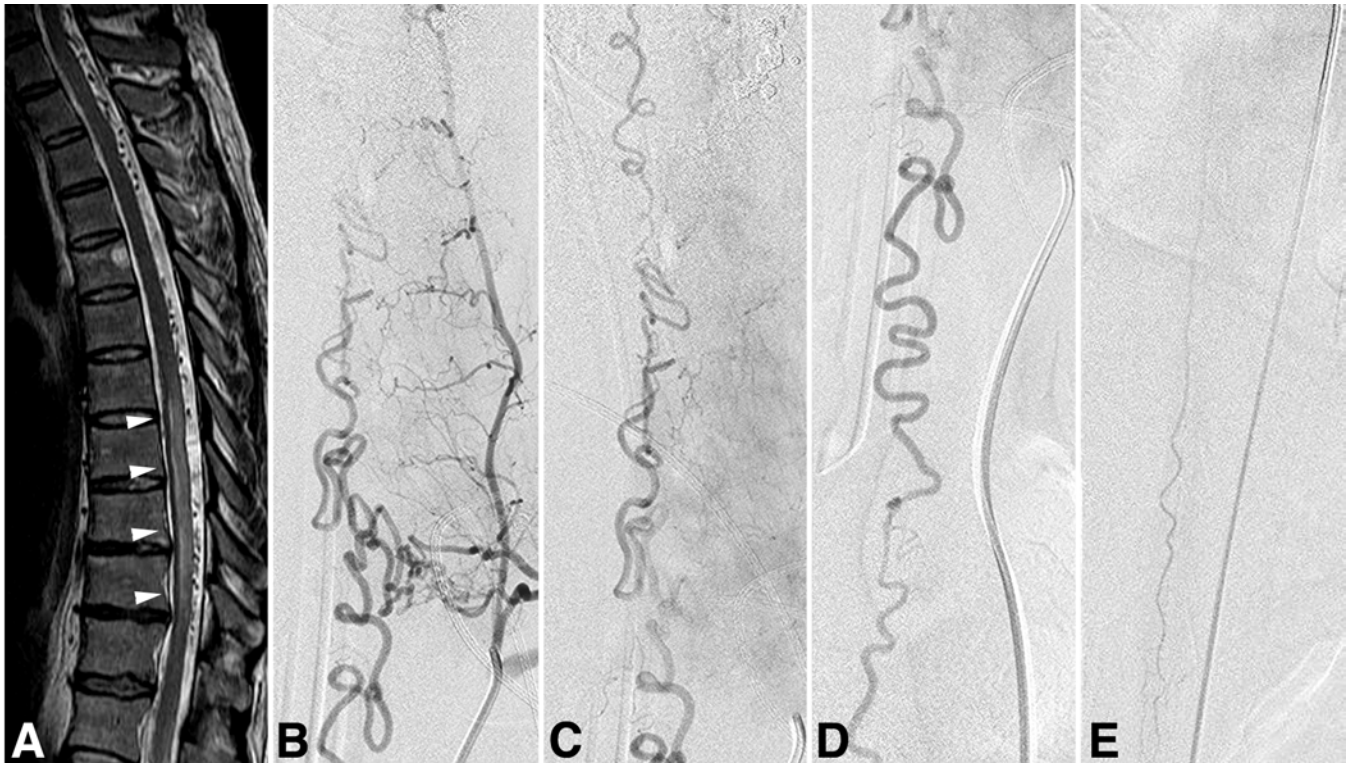

Fig. 3. Case 1. Sagittal T2-weighted MR image **(A)** demonstrating an intramedullary hyperintensity between T-7 and T-11 *(arrowheads)*, as well as serpentine perimedullary hypointensity signals. Anteroposterior angiograms (**B**, early phase; **C** and **D**, late phase) of the left T-1 segmental artery revealing a shunt fed by the T-1 radiculomeningeal artery (B), draining via a radicular vein into ascending perimedullary veins up to the upper cervical level (C) and descending perimedullary veins down to the thoracolumbar level (D). Stasis of contrast material in the affected perimedullary veins is noticed in the late venous phase (C and D). Anteroposterior angiogram **(E)** obtained in the late venous phase of the left T-8 segmental artery, demonstrating stasis of contrast material in the anterior spinal artery at the thoracolumbar level, indicating venous congestion of the spinal cord.

flow routes. These findings indicate that the progressive occlusion of radicular veins as alternate venous outflow routes, which can occur spontaneously or be induced by a shunt, will induce clinical symptoms in patients with SDAVFs.

Because the thoracolumbar region originally has relatively fewer venous outflow channels as compared with other spinal regions,[23] the venous congestive edema is likely to be transmitted in a caudocranial direction throughout the spinal cord. This may explain why the first symptoms of myelopathy sometimes reflect dysfunction of the conus medullaris, even though the fistula is located at the upper thoracic region.[12] Neither the location of pathological vessels nor the intramedullary imaging findings seem to be related to the location of the fistula.[15]

The indication to treat an asymptomatic SDAVF is controversial because its natural history remains unknown, although we believe that such fistulas will become symptomatic with time. However, asymptomatic SDAVFs with venous outflow to the extradural space can be monitored with close follow-up imaging studies and neurological examination. Once the lesion becomes symptomatic, however, early treatment is paramount since prognosis depends on the duration of symptoms before treatment and pretreatment disability.[19]

Limitations of this study include the small number of patients and the retrospective approach. We did not assess the quality of abnormal hyperintensity on T2-weighted MRI of the spinal cord. The measurement of the apparent diffusion coefficient, a quantitative parameter of the tissue water diffusion, may be useful for assessing the severity of venous congestive edema.[20] We compared the clinical and radiological features between asymptomatic and symptomatic SDAVFs located in the upper thoracic level only. A prospective approach with a large number of patients with asymptomatic SDAVFs, including lesions located in any other spinal levels, and young patients is warranted for clarifying the natural history of SDAVFs.

Conclusions

We described 2 cases of asymptomatic SDAVFs, which preserved their radicular venous outflow routes to the epi- and extradural spaces. Obstruction of the radicular venous outflow could be an important factor in inducing spinal congestive edema due to venous hypertension as well as subsequent clinical symptoms of SDAVFs.

Disclosure

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 to the study and manuscript prepara-

tion include the following. Conception and design: Krings, Sato. Acquisition of data: all authors. Analysis and interpretation of data: Sato. 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: Krings. Study supervision: TerBrugge.

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