Spinal dural arteriovenous shunt (dAVS) is the most common type of spinal vascular malformation, constituting approximately 70% of all spinal arteriovenous malformations. Clinical manifestations include progressive congestive myelopathy and subarachnoid hemorrhage. Intramedullary hemorrhage in relation to spinal dAVS is extremely rare and has been reported only once in the English literature. Here, we report a case of thoracic dAVS initially presenting with intermittent, stabbing left-sided chest pain, as well as gradual worsening of paresthesia, and later complicated by the sudden onset of intramedullary hemorrhage. We evaluated the patient using MRI at different time points. Importantly, before the hemorrhage occurred, T2-weighted MRI demonstrated a high-intensity spot in the spinal cord around a wedged portion of draining veins. Given the clinical course and intraoperative findings in this case, the imaging finding was considered to indicate increased transluminal pressure leading to rupture of the varix-like structure and intramedullary hemorrhage.

Case Report

History and Examination. A 49-year-old man was referred to our department with complaints of paresthesia of the chest, trunk, and bilateral lower limbs as well as difficulty in walking. He also had a history of intermittent stabbing chest pain, which was so intense that he had required admissions to another hospital to rule out angina pectoris. He needed a cane for walking long distances because of increased paresthesia over the preceding few months.

On admission, neurological examination revealed reduced superficial and deep sensation in areas below the T-6 dermatome. Muscle weakness was not apparent in the patient's extremities. There was no Romberg sign and no pathological reflexes. The cremasteric reflex was present on the right side and absent on the left side. Patellar and Achilles tendon reflexes were slightly diminished bilaterally.

Magnetic resonance imaging showed a flow-related signal void on the posterior surface of the spinal cord (Fig. 1). On T2-weighted MRI, hyperintense lesions were noted but limited to the T-6 level where a portion of the draining vein was noted to be tortuous and wedged into the dorsal surface of the spinal cord (Fig. 1C). Selective spinal angiography revealed a dAVS located at the T-6 level with a varix-like structure of the draining veins. The varix was noted to be tortuous and wedged into the dorsal surface of the spinal cord. Given the clinical course and intraoperative findings in this case, the imaging finding was considered to indicate increased transluminal pressure leading to rupture of the varix-like structure and intramedullary hemorrhage.

Key Words: hematomyelia • intramedullary hemorrhage • myelopathy • spinal dural arteriovenous shunt • spinal arteriovenous malformation • varix

Abbreviations used in this paper: dAVS = dural arteriovenous shunt; SAH = subarachnoid hemorrhage.

This article contains some figures that are displayed in color online but in black-and-white in the print edition.
Spinal dAVS with intramedullary hemorrhage

Indocyanine green videoangiography detected a shunt at the left T-6 root sleeve (Fig. 2). A preoperative diagnosis of thoracic dAVS was made, and the patient was scheduled for elective surgery.

While the patient waited for the surgical intervention, he experienced the sudden onset of back pain followed by exacerbation of paresthesia and muscle weakness in the left leg. He was not on anticoagulation or antiplatelet therapy. At the ictus, he was lying and resting in bed. No fluctuation in systemic blood pressure was noted before the hemorrhagic event. He was unable to stand up and was admitted emergently to our hospital. Although he could move his right leg against resistance, muscle strength in his left leg was 2 or lower on manual muscle testing. His superficial and deep sensation was graded as 50% bilaterally below the T-6 dermatome. Surprisingly, MRI revealed an intramedullary hemorrhage at T-6. The wedged portion of the draining vein was now dilated to form a varix-like structure (Fig. 3). High-intensity areas in the spinal cord, cranial and caudal to the hematoma, were now noticeable on T2-weighted images. A rupture of the varix was considered to be a potential cause of intramedullary hemorrhage and acute neurological deterioration.

Operation. The patient underwent surgical intervention on the following day after preoperative evaluations. With the patient prone, motor and sensory evoked potentials were monitored as described previously. Partial hemi-laminotomies of the left T-5 and T-6 laminae were performed for exposure. Following dural incision, a pedicle of the draining vein was recognized as arising from the left T-6 root sleeve (Fig. 4A and B). Soon after interruption of the draining vein, the arterialized vein turned purple. Indocyanine green videoangiography was used to confirm remission of the arterialized venous flow in the dilated veins. As we dissected and separated the veins from the dorsal spinal cord, a dilated varix-like portion was found protruding into the spinal cord parenchyma and facing a cavity in the hematoma. The varix was caught between thick pial membranes (Fig. 4C and D). We cut these membranous tissues and excised the varix for histological analysis. Careful inspection under an operative microscope did not reveal any other vascular malformation (Fig. 4E and F). Having considered that the location of this varix-like structure was identical to what preoperative MRI had indicated, we confirmed that the rupture of this varix-like structure was the source of intramedullary hemorrhage. No changes in the amplitude and latency of the motor and sensory evoked potentials were recorded during the operation.

Pathological Findings. Histological examination revealed hemosiderin deposition and fibrin attachment on the surface of the varix, which supported the contention that a rupture of the varix was the cause of the intramedullary hemorrhage (Fig. 5).

Postoperative Course. Postoperative MRI confirmed that the intramedullary hematoma was completely removed. Flow-related signal voids of the dilated venous structures disappeared, and T2 hyperintensity lesions were remarkably diminished. Immediately after the operation, the patient’s chest pain subsided. His motor weakness showed gradual improvement. One month after surgery,
muscle strength in his left leg was 4 on manual muscle testing, and he could walk with a cane. However, his post-operative neurological recovery was limited thereafter. Two years after surgery, paresthesia and sensory disturbances remained in his trunk, back, and lower extremities. He still required a cane for ambulation because of left leg weakness, and he needed self-catheterization for voiding urine.

**Fig. 2.** Digital subtraction angiograms (A–C) of the left T-6 intercostal artery, anteroposterior view, and a reconstructed image (D). The arteriovenous shunt was localized (black arrow) at the left T-6 dural root sleeve. Ascending and descending draining veins were visualized. No other spinal vascular malformations were detected. A wedged portion of the draining vein (arrowheads) was noted as an oval-shaped structure.

**Fig. 3.** Magnetic resonance images obtained after the onset of intramedullary hemorrhage. A T2-weighted image (A) and T1-weighted image (B) showing the intramedullary hematoma at T-6 (asterisks). Hyperintense lesions were apparent along the spinal cord on the T2-weighted image. Gadolinium-enhanced T1-weighted image (C) revealing enlargement of the previously wedged portion of the draining vein protruding into the hematomyelia (arrow).
Spinal dAVS with intramedullary hemorrhage

Fig. 4. Intraoperative photographs including those taken with indocyanine green videoangiography. After durotomy, a pedicle of the draining vein was located at the left T-6 root sleeve (arrowheads in A and B). Following disconnection of the draining veins, the vessels turned purple (C and D). A varix-like structure was caught underneath the pial membranes (arrow in C). Following dissection of the membranes, the embedded varix was exposed (arrow in D) and excised from the hematoxia cavity. Inside the cavity, there was no other source of bleeding (arrows in E and F). No arterial blood flow was detected inside of the cavity or in the draining veins according to indocyanine green videoangiography (F).

Discussion

In the last 10 years, we have treated 42 cases of spinal dAVS using microsurgery or endovascular interventions. The currently reported case is the only one to present with intramedullary hemorrhage. Five cases have presented with SAH, and the shunts were localized in the cervical spine or craniocervical junction in all of these cases.

In spinal dAVS, arteriovenous shunts often locate at the dural root sleeve.13 They were categorized as dorsal intradural arteriovenous fistulas in a system proposed by Spetzler et al.17 Increased intravascular pressure is transmitted from radiculomeningeal arteries to the radicular veins and the venous system of the spinal cord.5 It causes stagnation of the venous outflow from the spinal cord, resulting in intramedullary venous hypertension and ischemic insult to the spinal cord.5 Thus, patients suffer progressive congestive myelopathy, which occurs most frequently in the thoracolumbar region.8,16

Subarachnoid hemorrhage is encountered as another clinical presentation of this disease, especially when shunts are located in the craniocervical junction or the cervical spine.7,10,16 A pattern of venous outflow has been reported as significant in determining the clinical manifestation of the disease.7 More specifically, venous drainage ascending cranially may pose an increased risk of hemorrhage. In such cases, accelerated venous blood flow through the draining veins and focal dilation to form a venous sac10 or varix-like pouch were reported.9

Considering a case report of lumbar dAVS presenting with SAH, common features of hemorrhagic spinal dAVS may include an accelerated rate of arteriovenous circulation and an altered configuration of the radiculospinal vein.10

In the presented case, successive MR images demonstrated formation of the varix from a wedged portion of the draining vein. In this context, we speculated that increased venous hypertension had led to the formation of a varix. Furthermore, we confirmed, based on intraoperative and histological findings, that the hemorrhage occurred where the varix was protruding into the spinal cord. The reason that the increased venous pressure was focused in the varix and caused an intramedullary hemorrhage but not SAH remains to be elucidated. As shown in Fig. 2C, venous drainage was equally distributed to cranial and caudal directions. Thus, the same theory of SAH in a cervical or craniocervical junction dAVS cannot be applied in this case.

Interestingly, it was intraoperatively confirmed that the varix was caught between pial membranes and protruded into the spinal cord parenchyma. Given this unique configuration, we speculate that intraluminal pressure may have been acutely and heterogeneously increased inside the varix, resulting in a rupture. The walls of the varix, where the hemorrhage occurred, lacked elastic fibers and tended to be fragile.

Dorsal spinal veins predominantly run in subarachnoid spaces.12 However, our case is an extremely rare example in which the draining veins of the dAVS ran underneath the pia mater, which may also explain the rarity of intramedullary hemorrhage in spinal dAVS.

As demonstrated here, angioarchitecture of the draining veins, especially the relationship to the spinal cord parenchyma, could be key in predicting the formation and rupture of a varix along the draining veins. One important finding in preoperative MRI was the T2 hyperintensity areas around the protruded portion of the perimedullary vein in the spinal cord parenchyma (Fig. 1C). As reported previously, perifocal T2 hyperintensity indicated increased transluminal pressure in a case of intracranial arteriovenous malformation.1 In that report, intramedullary aneurysm formation was apparent in the center of the T2 hyperintensity area on angiograms. Because of the increased risk of hemorrhage, urgent resection of the arteriovenous malformation as well as the intranidal aneurysm was per-
formed, which resulted in satisfactory results. In our case, enlargement of the varix and hemorrhage occurred in the exact location where the abnormality appeared on the T2-weighted MRI image. Thus, we speculate that intraparenchymal T2 hyperintensity around the varix wall may have indicated an increment of intraluminal pressure and predicted the hemorrhagic event.

The clinical presentation of this case was also distinct from that of typical congestive myelopathy associated with midthoracic spinal dAVS. The patient complained of paresthesia and difficulty in walking. However, neurological examination did not reveal motor weakness, increased deep tendon reflexes, or pathological reflexes. Given the lack of objective abnormality in the neurological examinations, we did not consider the case to require urgent surgical intervention, and the intramedullary hemorrhage occurred before the treatment. However, when accompanied by clinical signs of spinal cord dysfunction, sudden and stabbing pain should be considered with caution, since it may indicate hemorrhage due to spinal arteriovenous malformation. Given the limited number of similar reports, whether the stabbing pain preceded the intramedullary hemorrhage or the SAH remains to be determined.

The patient suffered acute and severe neurological deterioration and limited postoperative functional recovery as a result of the hemorrhage. Given the devastating clinical course of our patient, it is important to recognize the possibility of such hemorrhagic events in patients with thoracic spinal dAVS.

Conclusions

We describe a case of thoracic dAVS that presented with intramedullary hemorrhage with preceding intermittent stabbing chest pains and paresthesia. A subpial wedge of the draining veins and intraparenchymal T2 hyperintensity around the dilated venous structure could be a key finding in preoperative MRI to predict the formation and rupture of a varix. It is important to recognize that intramedullary hemorrhage, although it is rare, can occur in association with thoracic dAVS, because it could dramatically degrade the clinical course and neurological outcomes of patients.

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 preparation include the following. Conception and design: Endo, Narisawa, Tominaga. Acquisition of data: Endo, Narisawa, Watanabe, Takahashi. Analysis and interpretation of data: Endo, Sato. Drafting the article: Endo, Narisawa, 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: Endo. Administrative/technical/material support: Takahashi, Tominaga. Study supervision: Takahashi, Tominaga.

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Manuscript submitted April 21, 2013. Accepted December 5, 2013. Please include this information when citing this paper: published online January 10, 2014; DOI: 10.3171/2013.12.SPINE12163.

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