

Dural arteriovenous fistula causing progressive myelopathy: Limited drainage to the perimedullary veins of the cervical area alone

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ABSTRACT

Intracranial dural arteriovenous fistulas (DAVFs) are uncommon cerebrovascular malformations that usually drain into a sinus or cortical veins. Depending on the pattern of the venous drainage, various clinical symptoms can be present, and most of them are ocular and/or intracranial symptoms, such as cranial nerve paralysis, intracerebral hemorrhage (ICH), subarachnoid hemorrhage (SAH), and intracranial hypertension. DAVFs that drain from a cortical to a perimedullary vein and show venous congestion that results in progressive myelopathy are extremely rare lesions. These symptoms are most commonly associated with an abnormal drainage pattern of Adamkiewicz's artery or when the venous drainage descends toward the thoracolumbar level. We report a case of DAVFs with drainage limited to the cerebellar and perimedullary veins of the cervical area causing progressive myelopathy.

1. Introduction

Intracranial dural arteriovenous fistulas (DAVFs) are composed of a meshwork of arteriovenous shunts located within the intracranial dura, usually drain into a sinus or cortical vein, and account for 15 % of all cerebrovascular malformations [1]. Their etiology still remains a matter of speculation, but in most cases, DAVFs are considered to be acquired, probably as a result of dural sinuses or venous thrombosis, and can be located anywhere in the dura [1,2]. Their angioarchitecture is that of an "arteriolo-venous fistula," with multiple arteries converging into a single venous structure [2]. DAVFs are mainly supplied by the dural artery, but aggressive, high-flow DAVFs can make reflux to the pial and cortical branches [3,4]. Depending on the pattern of the venous drainage, DAVFs are classified into several types including those that drain into a sinus with or without flow restriction and those that drain into cortical veins either directly or by reflux from a sinus [3,4]. The clinical symptoms of DAVFs can be varied, and the venous drainage of DAVFs is responsible for most signs and symptoms [3]. The symptoms can range from minor symptoms such as bruit, cranial nerve paralysis, and ocular symptoms to progressive symptoms such as intracerebral hemorrhage (ICH), subarachnoid hemorrhage (SAH), and intracranial hypertension [3,4]. DAVFs with cortical to perimedullary venous drainage are very rare [3–5]. Perimedullary venous drainage can cause venous congestion, which results in progressive myelopathy. When the venous drainage is

limited to the cervical cord, patients generally have no myelopathy; myelopathy is more commonly present when there is abnormal drainage pattern of Adamkiewicz's artery or when the venous drainage descends toward the thoracolumbar level [4,5]. Herein, we report a case of a posterior fossa DAVF causing progressive myelopathy even though the perimedullary venous drainage was limited to the cervical level and discuss the management, pathophysiological mechanisms, and treatment.

2. Case report

A 57-year-old man developed a gait disturbance with weakness in both legs for several weeks. A few days before admission, the patient could not walk on his own due to progressive ataxia of the lower extremities and developed severe dysphagia. A brain and cervical MRI/A revealed a DAVF in the torcula area and an enlarged drainage vein involving the cerebellar cortical and perimedullary veins at the cervical spinal cord level. At the thoracic level, there was no perimedullary vein enlargement or reflux (Fig. 1). A conventional cerebral angiogram showed DAVFs in the torcula area that were supplied by numerous branches of occipital artery and middle meningeal artery. In the fistula area in the location of the reflux and abnormal drainage vein, the feeding arteries were very corrugated on both sides and had a screw shape with many twists (Fig. 2). Spinal angiography was not performed

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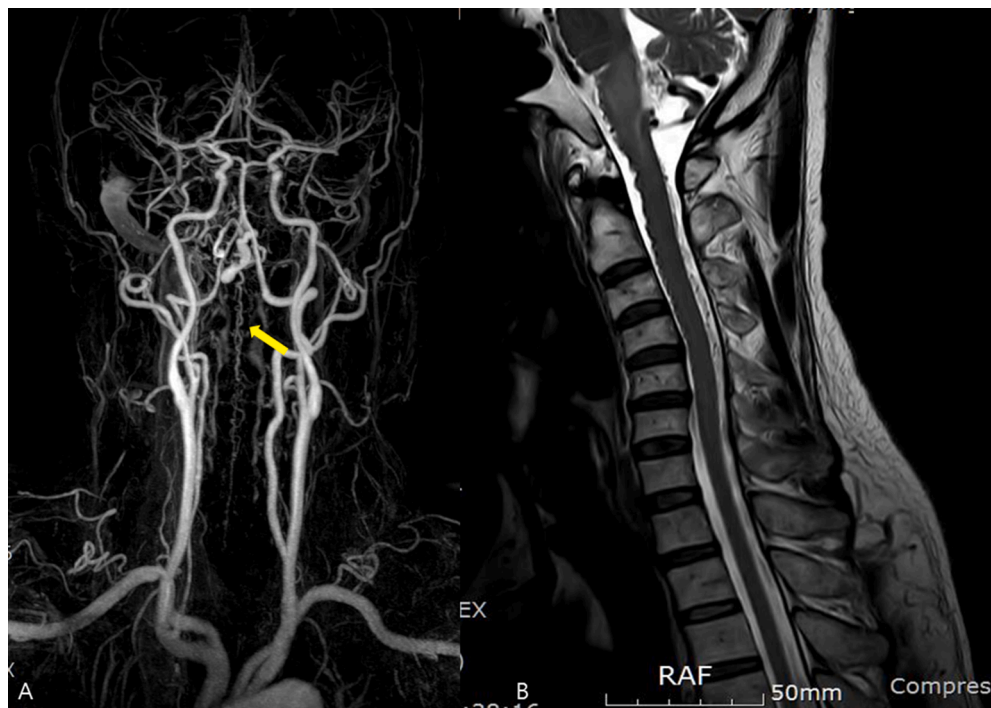


Fig. 1. (A) Brain and cervical MRI/MRA revealed a DAVF in the torcula area and an enlarged drainage vein involving the cerebellar cortical and perimedullary veins at the cervical spinal cord level (yellow arrow). (B) At the thoracic level, there was no perimedullary vein enlargement and reflux.

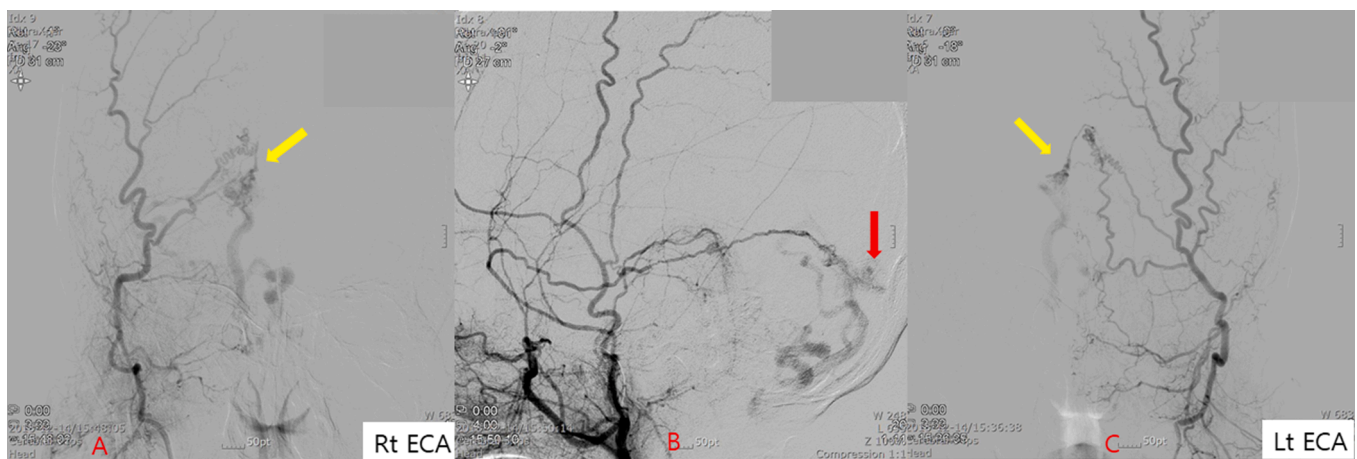


Fig. 2. (A-C) Cerebral angiogram showing DAVFs in the torcula area supplied by numerous branches of the occipital artery and middle meningeal artery (yellow arrow: from the occipital artery, red arrow: from the middle meningeal artery).

because intracranial DAVFs were confirmed, and a spinal MRI showed no perimedullary drainage or enlargement at the thoracic level such as in the Adamkiewicz artery. Endovascular treatment with Onyx was planned. There were numerous feeding arteries on both sides; thus, if the fistula portion was not completely obliterated at one time by approaching from one side, the treatment was planned to be divided into two sessions (left and right). During endovascular treatment on left side, the exact fistula point could not be accessed by the microcatheter because of the severe tortuosity of the vessels. At first, the microcatheter approached as close as possible to the fistula point through the most dominant feeding artery (branch of the occipital artery), and embolization with Onyx was performed. The embolic material did not completely obliterate the fistula point, and reflux of Onyx occurred because of the severely tortuous vessels. After occlusion of the main feeding arteries, subsequent embolization of the other feeding arteries was performed sequentially. With the dominant feeding arteries

occluded, new feeding arteries that were not seen previously on the cerebral angiogram were noted. All of these new arteries were also embolized in turn, and eventually all the feeding arteries were occluded. The final angiogram showed no abnormal connections or venous reflux. The DAVF was completely eliminated on the left side (Fig. 3). After treatment of only the left-sided lesion, the patient's neurological symptoms completely resolved. After two weeks, embolization of the right-sided lesion was performed using the same method used for the left side. Fistulas and reflux were not seen on angiograms of both external carotids after the final treatment (Fig. 4).

3. Discussion

The most common type of spinal arteriovenous malformation is a spinal dural fistulas with perimedullary venous drainage [6]. These malformations occur most commonly in the thoracic or thoracolumbar

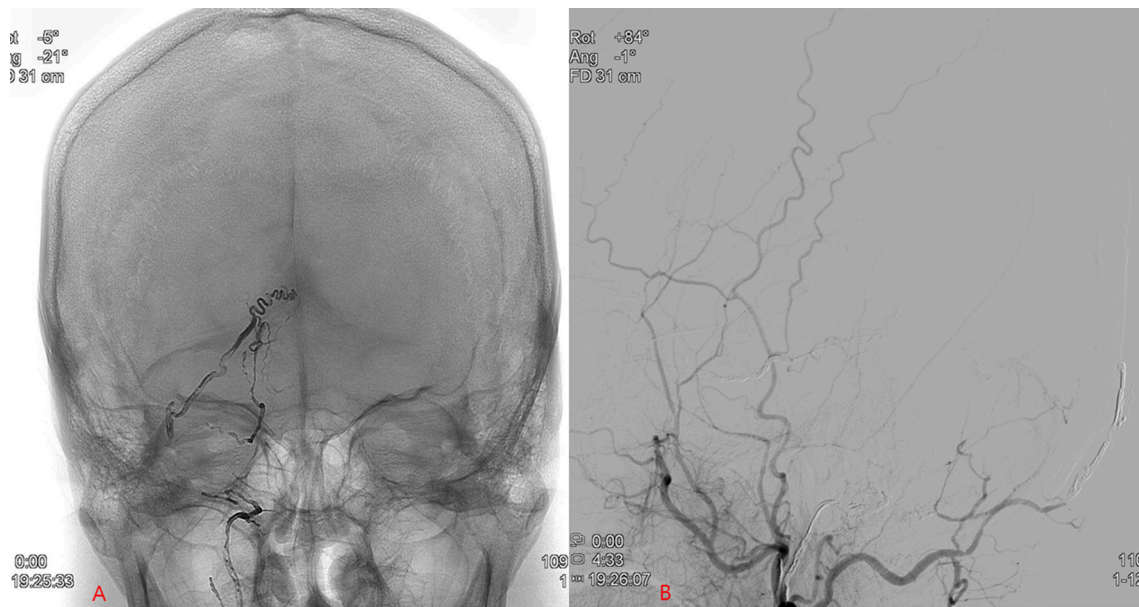


Fig. 3. (A and B) All of feeding arteries were embolized in turn. An angiogram showed no abnormal connection and venous reflux. The DAVFs were completely eliminated on the right side.



Fig. 4. After two weeks, embolization of the left-sided lesion was performed using the same method used for the right side. (A and B) Fistulas and reflux were not seen on the left external carotid angiogram after the final treatment.

spine. Moreover, the lower thoracic and lumbar spinal cord is more vulnerable to hemodynamic changes, and the initial symptoms of spinal DAVFs usually reflect dysfunction of the lower part of the spinal cord, namely, the conus medullaris [6,7]. In contrast to these spinal DAVFs, intracranial DAVFs with perimedullary drainage are rare. In a survey of previous literature, we found 39 reported cases (Table 1) [5,7,8,17]. A classification system for intracranial DAVFs according to their venous drainage was introduced in 1978 by Djindjian and Merland [3]. Depending on the pattern of venous drainage, they are classified into several types including those that drain into a sinus with or without flow restriction and those that drain into cortical veins either directly or by reflux from a sinus [3,4,18]. The original classification system included three types (types I, II, and III). Later, two additional types were defined because their clinical presentation was so uncommon: type IV (DAVF

with a giant venous varix) and type V (DAVF with spinal venous drainage) (Table 2 and Fig. 5) [3,4]. In subsequent reports, the clinical characteristics, prognosis, and treatment options of these five DAVF types were described more precisely based on these classifications [4].

The etiology of these rare malformations remains a matter of speculation; a sinus thrombosis has been uncommonly reported [13]. The blood flow through the fistula runs via an anastomotic pathway between the pontomesencephalic venous system and both anterior and posterior perimedullary veins to the coronal venous plexus, which becomes dilated and tortuous [11]. The pressure in the veins can reach approximately 70 % or more of systemic arterial blood pressure [19]. Drainage of DAVFs through the enlarged spinal perimedullary veins can be followed distally along the entire spinal cord. In general, there are numerous medullary radicular veins draining the blood from the spinal

Table 1
Characteristics of the reported 39 cases.

Author	No. of cases	Symptoms and signs	Site of shunt	Treatment modality
Brunereau et al [5]	12	Transient aphasia, SAH, myelopathy	Tentorium, foramen magnum, petrous transverse sinus, torcular region sinus	Treatment modality not reported
Askasawa et al [7]	1	Myelopathy	Foramen magnum	Embolization
Bousson et al [8]	1	Myelopathy	Tentorium	Embolization
Bret et al [9]	1	Myelopathy	Tentorium	Surgical clipping
Chen et al [10]	2	Myelopathy	Torcular region	Surgical clipping
Gobin et al [11]	5	Myelopathy	Petrous region, tentorium, transverse sinus	Embolization, surgical clipping
Hahnel et al [12]	1	Myelopathy	Petrous sinus	Embolization
Ricolfi et al [13]	5	Myelopathy	Tentorium, petrous sinus, cavernous sinus, sigmoid sinus	Embolization, surgical clipping
Trop et al [14]	1	Myelopathy	Foramen magnum	Surgical clipping
Versari et al [15]	2	Myelopathy	Petrous sinus, sigmoid sinus	Surgical clipping
Wiesmann et al [16]	1	Myelopathy	No exact description	Embolization
Wrobel et al [17]	3	Myelopathy	Petrous apex, tentorium	Surgical clipping
Christof Renner et al [20]	1	Myelopathy	Tentorium	Surgical clipping
Partington et al [22]	3	Myelopathy	Tentorium, foramen magnum	Surgical clipping

cord to the epidural space in the cervical and thoracic areas. However, if the medullary venous system is not enough to drain into the radicular vein, a fistula can cause stagnation of the blood flow out of intramedullary veins that have no valves. Thus, the intramedullary veins are directly affected by increased pressure in the perimedullary venous system, leading to reduced perfusion and microcirculation. This may lead to progressive functional impairment or spontaneous thrombosis, as in Foix-Alajouanine syndrome [8,10,13,19]. To date, the reason why drainage to the radial vein has failed remains unknown; some authors believe that degenerative changes of the spine may reduce the potential reservoir of draining veins [14]. It has been reported that myelopathy occurs when extensive ascending or descending perimedullary venous drainage involves the thoracic or thoracolumbar levels, such as abnormal venous drainage of the artery of Adamkiewicz in spinal or intracranial DAVFs. In spinal or intracranial DAVFs without myelopathy, the common angiographic characteristic is that the spinal perimedullary drainage veins can be followed down to the cervical spinal cord only. At the cervical level, the perimedullary veins drain into the epidural veins via a medullary radicular vein. This drainage prevents spinal cord venous hypertension [5]. In reviewing previous reports [5,7,8,17], our patient is one of few patients with an intracranial DAVF causing progressive myelopathy with perimedullary venous drainage in only the cervical area without abnormal venous congestion at the thoracolumbar level such as in the Adamkiewicz artery.

Only Trop et al [14] and Renner et al [20] reported similar patients

in 1998 and 2006, respectively. Compared with previous cases, our patients also showed similar MRI findings of expansion of venous congestion limited to the cervical and cervicothoracic levels. Thus, we think that the opinion of Willinsky et al [21] that a normal angiogram of the Adamkiewicz artery rules out a DAVF is not always applicable. In particular, Christof Renner [20] postulated that an abnormality of the spinal cord that is limited to the cervical/cervicothoracic level is highly suspected to be an intracranial DAVF. It should be kept in mind that DAVFs with drainage limited to the cervical level may cause progressive myelopathy.

The treatment of choice remains controversial. Endovascular or conventional surgery can be treatment options. Many clinical factors must be considered for treatment. The experience and proficiency of the surgeon, the morphological characteristics of the lesion such as the accessibility to the fistula during endovascular treatment, where the feeding artery originates, how many feeding arteries there are, and how the fistula is drained must all be considered. Therefore, uniform recommendations cannot be given. In our case, considering the multiple diffuse fistulas, there were many limitations with regard to conventional surgery. Endovascular treatment was not easy, but successful treatment was achieved by occlusion of all visible feeding arteries and the fistula portion.

4. Conclusion

The spinal cord vasculature is very complex. The reason for venous stagnation causing myelopathy and lack of drainage into the epidural venous system still remains unclear. DAVFs with drainage limited to the cervical level may cause progressive myelopathy. A normal angiogram of the Adamkiewicz artery does not rule out a DAVF. If there is cervical myelopathy without a degenerative or traumatic spinal lesion and the vascular abnormality is limited to the cervical/cervicothoracic level, an intracranial DAVF may be considered. In treatment, uniform standard recommendations have not yet been established. Before determining the treatment modality, the characteristics of the lesion should be well documented through accurate, precise examinations, and many clinical

Table 2
Modified classification of intracranial DAVFs; original classification by Djindjian and Merland.

Type	Description
I	Drainage into a sinus without restriction
II	Drainage into a sinus with flow restriction and reflux into other sinuses or cortical veins
III	Drainage into a cortical vein
IV	Drainage into a cortical vein with a large varix
V	Spinal perimedullary venous drainage

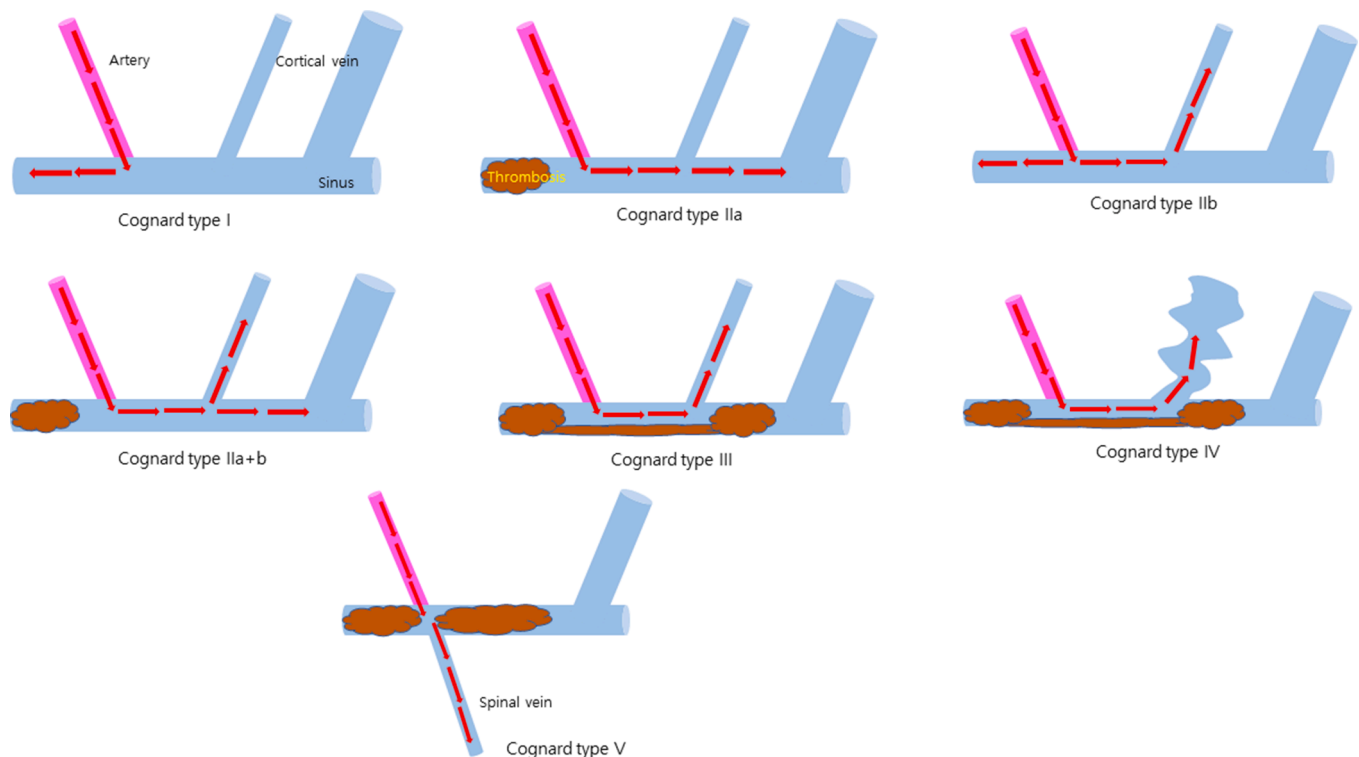


Fig. 5. Cognard classification of intracranial DAVFs.

factors must be considered.

5. Dural arteriovenous fistula causing progressive myelopathy

Limited Drainage to the Perimedullary Veins of the Cervical Area Alone.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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References

- [1] S. M. Reflections upon the nature and management of intracranial and intraspinal vascular malformations and fistulae. *J Neurosurg* 1994. [Epub ahead of print].
- [2] Houdart E, Gobin YP, Casasco A, et al. A proposed angiographic classification of intracranial arteriovenous fistulae and malformations. *Neuroradiology* 10.1007/BF00588376.
- [3] Djindjian R. Super-selective arteriography of the external carotid artery. *J Neuropathol Exp Neurol* 10.1097/00005072-197811000-00012.
- [4] Cognard C, Gobin YP, Pierot L, et al. Cerebral dural arteriovenous fistulas: Clinical and angiographic correlation with a revised classification of venous drainage. *Radiology* 10.1148/radiology.194.3.7862961.
- [5] Brunereau L, Gobin YP, Meder JF, et al. Intracranial dural arteriovenous fistulas with spinal venous drainage: Relation between clinical presentation and angiographic findings. *Am J Neuroradiol* 1996. [Epub ahead of print].
- [6] Hurst RW, Kenyon LC, Lavi E, et al. Spinal dural arteriovenous fistula: The pathology of venous hypertensive myelopathy. *Neurology* 10.1212/WNL.45.7.1309.
- [7] Asakawa H, Yanaka K, Fujita K, et al. Intracranial dural arteriovenous fistula showing diffuse MR enhancement of the spinal cord - Case report and review of the literature. *Surg Neurol* 10.1016/S0090-3019(02)00861-3.
- [8] Bousson V, Brunereau L, Vahedi K, et al. Intracranial dural fistula as a cause of diffuse MR enhancement of the cervical spinal cord. *J Neurol Neurosurg Psychiatry* 10.1136/jnnp.67.2.227.
- [9] Bret P, Salzmann M, Bascoulegue Y, et al. Dural arteriovenous fistula of the posterior fossa draining into the spinal medullary veins—an unusual cause of myelopathy: Case report. *Neurosurgery* 10.1227/00006123-199411000-00026.
- [10] Chen CJ, Chen CM, Lin TK. Enhanced cervical MRI in identifying intracranial dural arteriovenous fistulae with spinal perimedullary venous drainage. *Neuroradiology* 10.1007/s002340050609.
- [11] Gobin YP, Rogopoulos A, Aymard A, et al. Endovascular treatment of intracranial dural arteriovenous fistulas with spinal perimedullary venous drainage. *J Neurosurg* 10.3171/jns.1992.77.5.0718.
- [12] Hähnel S, Jansen O, Geletnek K. MR appearance of an intracranial dural arteriovenous fistula leading to cervical myelopathy. *Neurology* 10.1212/WNL.51.4.1131.
- [13] Ricolfi F, Manelfe C, Meder JF, et al. Intracranial dural arteriovenous fistulae with perimedullary venous drainage. Anatomical, clinical and therapeutic considerations. *Neuroradiology* 10.1007/s002340050846.
- [14] Trop R, Roy D, Raymond J, et al. Craniocervical dural fistula associated with cervical myelopathy: Angiographic demonstration of normal venous drainage of the thoracolumbar cord does not rule out diagnosis. *Am J Neuroradiol* 1998. [Epub ahead of print].
- [15] Versari PP, D'Aliberti G, Talamonti G, et al. Progressive myelopathy caused by intracranial dural arteriovenous fistula: Report of two cases and review of the literature. *Neurosurgery* 10.1227/00006123-199311000-00023.
- [16] Wiesmann M, Padovan CS, Pfister HW, et al. Intracranial dural arteriovenous fistula with spinal medullary venous drainage. *Eur Radiol* 10.1007/s003300000382.
- [17] CWrobel CJ, Oldfield EH, Di Chiro G, et al. Myelopathy due to intracranial dural arteriovenous fistulas draining intrathecally into spinal medullary veins. *J Neurosurg* 10.3171/jns.1988.69.6.0934.
- [18] Halbach V V., Higashida RT, Hieshima GB, et al. Treatment of dural arteriovenous malformations involving the superior sagittal sinus. *Am J Neuroradiol* 1988. [Epub ahead of print].
- [19] Hassler W, Thron A, Grote EH. Hemodynamics of spinal dural arteriovenous fistulas. An intraoperative study. *J Neurosurg* 10.3171/jns.1989.70.3.0360.
- [20] Renner C, Helm J, Roth H, et al. Intracranial dural arteriovenous fistula associated with progressive cervical myelopathy and normal venous drainage of the thoracolumbar cord: case report and review of the literature. *Surg Neurol* 10.1016/j.surneu.2005.06.022.
- [21] Willinsky R, Lasjaunias P, Terbrugge K, et al. Angiography in the investigation of spinal dural arteriovenous fistula - A protocol with application of the venous phase. *Neuroradiology* 10.1007/BF00588560.
- [22] Partington MD, Rüfenacht DA, Marsh WR, et al. Cranial and sacral dural arteriovenous fistulas as a cause of myelopathy. *J Neurosurg* 1992. [Epub ahead of print].