



Long-standing tentorial dural arteriovenous fistula presenting as progressively worsening cervical myelopathy by degenerative cervical intervertebral disc: a case report

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Received: 3 April 2019 / Accepted: 24 May 2019 / Published online: 30 May 2019
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Abstract

We report a rare case of long-standing asymptomatic tentorial dural arteriovenous fistula (DAVF) presenting as symptomatic progressive cervical myelopathy which was caused by the protrusion of degenerative cervical intervertebral discs into the spinal canal and compression of the anterior spinal vein. The cervical spinal canal was narrowed, and the spinal cord was compressed anteriorly by protrusion of cervical intervertebral discs at the C6–7 level. The intervertebral discs compressed the draining vein of the tentorial DAVF (the anterior spinal vein) so venous congestion of the spinal cord resulted in cervical myelopathy only above the C6 level.

Keywords Cervical myelopathy · Dural arteriovenous fistula

Introduction

Intracranial dural arteriovenous fistulas (DAVFs) account for 10–15% of all intracranial vascular malformations [9]. A significant number of DAVFs remain clinically silent or spontaneously involute, so the true incidence may be much higher. In addition, their etiology still remains unclear and a matter of speculation; thrombosis of the dural sinuses is inconsistently reported [6]. DAVFs can occur anywhere within the intracranial dura mater and consist of numerous tiny connections between branches of dural arteries and veins or a venous sinus [7].

DAVFs with perimedullary venous drainage are very rare lesions. Their clinical presentation spans subarachnoid hemorrhage to progressive myelopathy [11] although true perimedullary spinal fistulas, i.e. anterior DAVF, do not cause bleeding but venous congestion only. Similar to the more

frequent spinal DAVFs, intracranial DAVFs lead to venous congestion of the spinal cord [1–5, 8, 10, 12–14]. Tentorial DAVFs with spinal venous drainage causing progressive cervical myelopathy are much rarer. Here, we report a case of long-standing asymptomatic tentorial DAVF which became symptomatic progressive cervical myelopathy through the protrusion of degenerative cervical intervertebral discs into the spinal canal, resulting in compression of the anterior spinal vein.

Case report

A 66-year-old male with quadriplegia was admitted, and the patient's status became progressively worse to include quadriplegia with urinary incontinence. His cervical spine magnetic resonance image (MRI) showed a definite high signal intensity from C1 to C6 on the T2-weighted image (Fig. 1a). In addition, abnormal dilatations of the vessel in the spinal canal were found. Below the C6 level, the spinal cord was normal without MRI signal changes. Digital subtraction angiography (DSA) was performed and showed DAVF from tentorial branches of the meningohypophyseal trunk (MHT) on the right internal carotid artery, and the feeders were only from the marginal tentorial arteries of the MHT. The DAVF drained into the petrous vein, then to the anterior pontomesencephalic vein and, finally, to the anterior spinal vein (Fig. 1b, c, and d). By reviewing old radiographic images, we discovered that the

This article is part of the Topical Collection on *Vascular Neurosurgery - Other*

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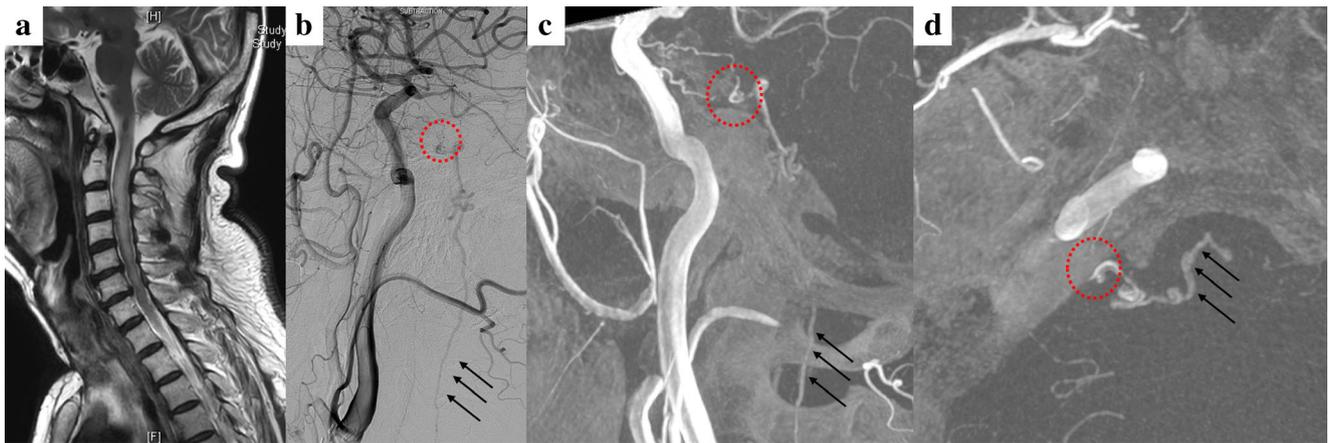


Fig. 1 **a** Cervical spine magnetic resonance image showed a definite high signal intensity from C1 to C6 on the T2-weighted image. Below the C6 level, there was a normal spinal cord without signal changes. **b** Digital subtraction angiography showed dural arteriovenous fistula (red dotted circle). Feeders were only from the marginal tentorial arteries of the meningohypophyseal trunk on the right internal carotid artery. The fistula

drained into the petrous vein, then to the anterior pontomesencephalic vein and, finally, to the anterior spinal vein (black arrows). Flat-panel detectors computed tomography (VasoCT) showed the fistula (red dotted circle) and draining veins (black arrows) in relation with the skull on **c** sagittal and **d** axial images

DAVF had been present previously, yet had been asymptomatic. We retrospectively found that there had been a dilatation of the anterior spinal vein on contrast-enhanced cervical computed tomography from 4 years ago when the patient underwent an operation to remove tonsillar cancer. Additionally, greater dilatation of the anterior spinal vein was observed on a neck MRI from 1 year ago (Fig. 2a). However, at presentation, the cervical spinal canal was narrowed, and the spinal cord was compressed anteriorly by protrusion of cervical intervertebral discs at the level of C5–6 and C6–7. The intervertebral discs compressed the draining vein of the tentorial DAVF (the anterior spinal vein), aggravating venous congestion of the spinal cord and resulting in cervical myelopathy only above the C6 level (Fig. 2b and c). The feeders were very small and seemed to be difficult candidates for embolization. Through a lateral suboccipital approach, the fistula on the tentorium was coagulated and

ligated by clipping (Fig. 3). There were no procedure-related complications. Postoperative DSA showed complete obliteration of the fistula (Fig. 4a). The cervical myelopathy improved on the postoperative MRI (Fig. 4b), and the 1-year follow-up MRI showed normal cord signals (Fig. 4c). Presently, the patient walks independently, but partial urinary incontinence remains.

Discussion

In the present case, we reported that an asymptomatic DAVF with drainage to the anterior spinal vein could become progressively symptomatic, resulting in cervical myelopathy, due to degenerative cervical intervertebral discs protruding into the spinal canal and compressing the anterior spinal vein.

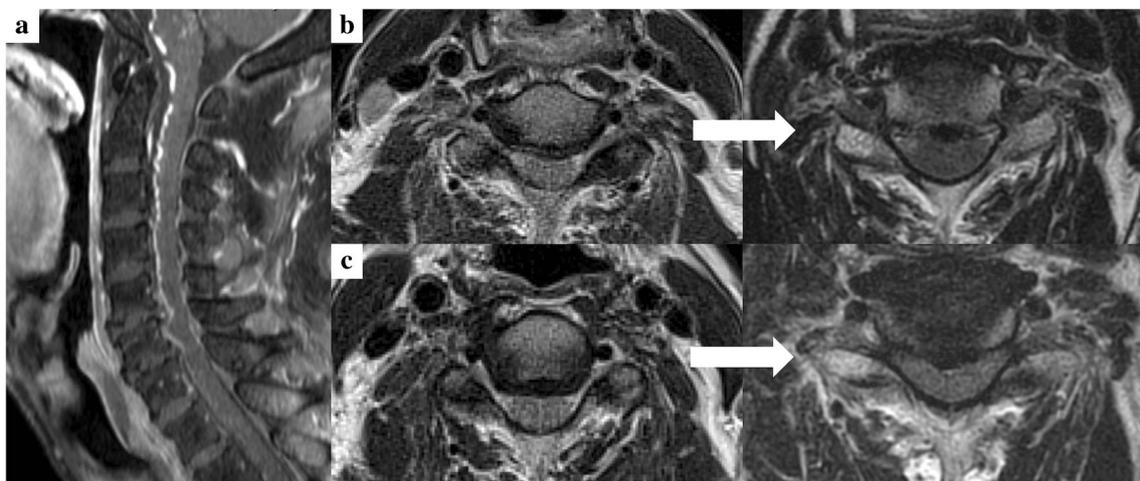
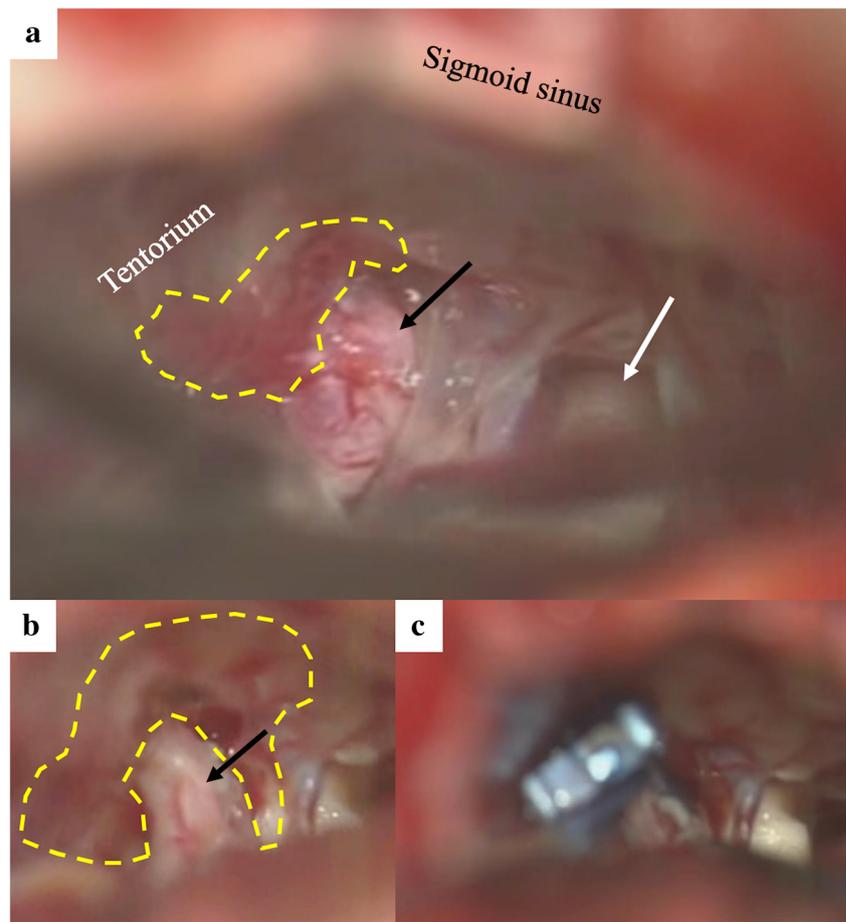


Fig. 2 **a** Asymptomatic dilatation of the anterior spinal vein on the neck magnetic resonance image 1 year prior to presentation. The cervical spinal canal subsequently narrowed, and the spinal cord was compressed anteriorly by protrusion of cervical intervertebral discs at the level of **b** C5–6 and **c** C6–7

Fig. 3 Through the lateral suboccipital approach, **a** the fistulae on the tentorium (yellow dotted line) and the petrosal vein (black arrow) were seen. A white arrow indicates the trigeminal nerve. The fistulae were **b** coagulated (yellow dotted line) and **c** ligated by clipping



Since the first report in 1988, there have been 14 reported cases of tentorial DAVFs that presented with progressive cervical myelopathy (Table 1). All patients were male, and the mean age of onset was 54.4 years. Feeders of DAVFs were mainly from the MHT of the internal carotid artery. Symptoms were usually progressive, but an acute onset of neurological disorders was seen in some cases. Among the reported cases, it was speculated that degenerative changes of the spine might

reduce the potential reservoir of draining veins. However, the real cause has not been reported yet. Furthermore, there is no reported case in which degenerative cervical disc herniation aggravated the decrement of venous drainage, causing progressive cervical myelopathy.

Spinal myelopathy can be rarely caused by cranial DAVFs. Venous congestion of the spinal cord can be produced by any cranial or spinal DAVFs that gain access to the venous system

Fig. 4 **a** Postoperative digital subtraction angiography showed complete obliteration of the fistula. Cervical myelopathy improved on **b** the postoperative magnetic resonance image (MRI) and **c** the 1 year follow-up MRI showed normal cord signals

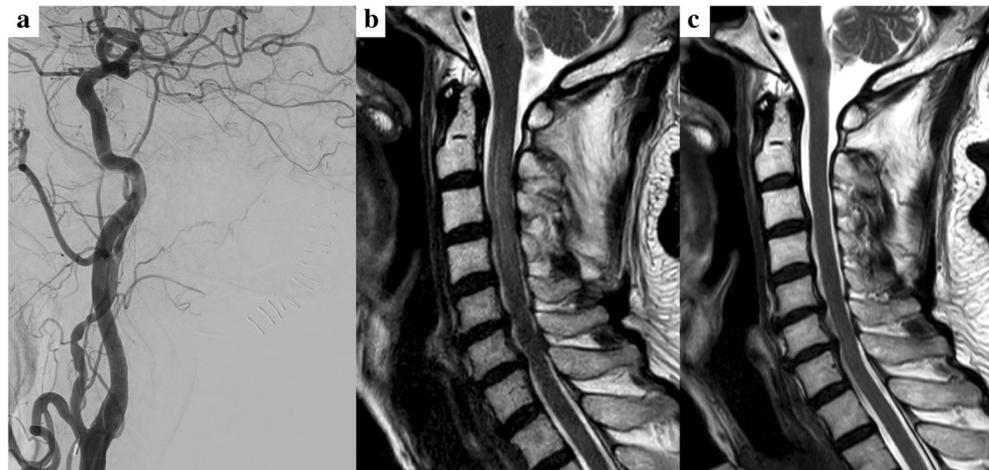


Table 1 List of the 15 tentorial dural arteriovenous fistulae presented by cervical myelopathy including the present case

Authors	Age	Sex	Symptoms and signs	Feeders	Site of shunt	Treatment
Wrobel et al. [14]	68	M	Myelopathy	Branches from MHT	Tentorium	Microsurgery
	42	M	Myelopathy	Branches from MHT	Tentorium	Microsurgery
Gobin et al. [5]	53	M	Myelopathy	Branches from MHT	Tentorium	Embolization
Partington et al. [10]	40	M	Myelopathy	Branches from MHT	Tentorium	Microsurgery and embolization
Versari et al. [13]	50	M	Myelopathy	Branches from MHT	Tentorium	Microsurgery
Bret et al. [2]	31	M	Myelopathy	Branches from MHT	Tentorium	Microsurgery
Brunereau et al. [3]	53	M	Myelopathy	Branches from MHT	Tentorium	Microsurgery
	69	M	Myelopathy	Branches from MHT	Tentorium	Microsurgery
Earnst et al. [4]	71	M	Myelopathy	Branches from MHT	Tentorium	Microsurgery
Bousson et al. [1]	36	M	Myelopathy	Branches from MHT	Tentorium	Embolization
Ricolfi et al. [12]	69	M	Myelopathy	Branches from MHT	Tentorium	Embolization
	53	M	Myelopathy	Branches from MHT	Tentorium	Embolization
Kalamangalam et al. [8]	68	M	Myelopathy	Branches from MHT	Tentorium	Embolization
Renner et al. [11]	58	M	Myelopathy	Branches from MHT	Tentorium	Microsurgery
Kwon et al. (the present case)	66	M	Myelopathy	Branches from MHT	Tentorium	Microsurgery

M male; MHT meningohipophyseal trunk

of the spinal cord and produce elevated pressure in the coronal venous plexus or the medullary veins. This is the mechanism for the pathophysiology of cord dysfunction in patients with DAVFs, and it can elicit progressive myelopathy. Spinal cord ischemia caused by arterial steal cannot occur in these DAVFs since they are low-flow fistulae. The only potential mechanism for the production of neurological deficits is supported by the enlargement and bright signal of the cord on the MRI. Thus, interruption of a cranial DAVF that drains into spinal veins permits recovery of the myelopathy.

Brunereau et al. [3] reported 6 of 12 patients with intracranial DAVFs draining in the perimedullary venous system who had cerebral symptoms and signs without myelopathy. The main symptoms were subarachnoid hemorrhage of the posterior fossa. The angiograms of these patients showed spinal drainages that could only be followed to the cervical level where the veins drained into the epidural venous system via a medullary-radicular vein, not the fistula perimedullary drainage. As a rebuttal, this phenomenon implied that missing myelopathy in those patients might have existed. Thus, degenerative changes in the spine might aggravate venous drainage into the perimedullary vein.

The proper diagnosis is challenging because symptoms are mostly nonspecific and prevalence is rare. Patients with a suspected spinal arteriovenous malformation usually undergo spinal angiography. However, angiographic studies must include the cranial vasculature when spinal studies are normal or if the abnormality on MRI is maximal in the upper spinal cord [8]. In this case, it was very difficult to differentiate whether cervical intervertebral disc protrusion or ischemia of

the anterior spinal vein caused the myelopathy. Cervical myelopathy is predominantly caused by spinal cord compression at the cervical level. In the present case, degenerative cervical intervertebral discs protruding into the spinal canal compressed the anterior spinal vein. Our case is one of the rarest and true cases of degenerative changes of the spine reducing the potential reservoir of draining veins. Our case shows the importance of cervical spine imaging for diagnosing this rare cerebrovascular disease. High-resolution imaging, including computed tomography, MRI, and DSA, should be done to rule out these cerebrovascular diseases. Congestion and swelling of the blood vessels and signal changes of the spinal cord could be helpful markers for the initial screening and diagnosis.

There are two treatment options for intracranial DAVFs with perimedullary venous drainage: microsurgical clipping with coagulation and endovascular embolization. The goal of treatment is to proximally occlude the draining vein as it exits the arteriovenous shunt by embolization, microsurgery, or both. Fistula accessibility could be a key for the proper selection of treatment options, and the preference of the neurointerventionist or neurosurgeon might be another factor. In the present case, small marginal tentorial arteries of the MHT were the only feeders, and the DAVF drained into the petrous vein, then to the anterior pontomesencephalic vein and, finally, to the anterior spinal vein. Thus, neither the transvenous approach nor the transarterial approach for endovascular embolization could be performed due to vessel inaccessibility. However, a lateral suboccipital-retrosigmoid approach led us to the petrous vein and tentorium, so we performed microsurgical coagulation and ligation (by clipping) to treat the DAVF.

Conclusion

By compressing the anterior spinal vein, degenerative cervical intervertebral discs protruding into the spinal canal could aggravate symptoms of a tentorial DAVF draining to the anterior spinal vein. In the present case, microsurgical coagulation and ligation cured the DAVF.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval All procedures performed in the studies involving human participants were in accordance with the ethical standards of our Institutional Review Board with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Patient consent The patient has consented to the submission of the case report for submission to the journal.

References

- Bousson V, Brunereau L, Vahedi K, Chapot R (1999) Intracranial dural fistula as a cause of diffuse MR enhancement of the cervical spinal cord. *J Neurol Neurosurg Psychiatry* 67:227–230
- Bret P, Salzmann M, Bascoulergue Y, Guyotat J (1994) Dural arteriovenous fistula of the posterior fossa draining into the spinal medullary veins—an unusual cause of myelopathy: case report. *Neurosurgery* 35:965–968
- Brunereau L, Gobin YP, Meder JF, Cognard C, Tubiana JM, Merland JJ (1996) Intracranial dural arteriovenous fistulas with spinal venous drainage: relation between clinical presentation and angiographic findings. *AJNR Am J Neuroradiol* 17:1549–1954
- Ernst RJ, Gaskill-Shiple M, Tomsick TA, Hall LC, Tew JM Jr, Yeh HS (1997) Cervical myelopathy associated with intracranial dural arteriovenous fistula: MR findings before and after treatment. *AJNR Am J Neuroradiol* 18(7):1330–1334
- Gobin YP, Rogopoulos A, Aymard A, Khayata M, Reizine D, Chiras J, Merland JJ (1992) Endovascular treatment of intracranial dural arteriovenous fistulas with spinal perimedullary venous drainage. *J Neurosurg* 77:718–723
- Ha SY, Kwon YS, Kim BM, Kim DI, Kim DJ (2012) Clinical and angiographic characteristics of multiple dural arteriovenous shunts. *AJNR Am J Neuroradiol* 33(9):1691–1695
- Halbach VV, Higashida RT, Hieshima GB, Mehninger CM, Hardin CW (1989) Transvenous embolization of dural fistulas involving the transverse and sigmoid sinuses. *AJNR Am J Neuroradiol* 10: 385–392
- Kalamangalam GP, Bhattacharya J, Teasdale E, Thomas M (2002) Myelopathy from intracranial dural arteriovenous fistula. *J Neurol Neurosurg Psychiatry* 72:816–818
- Luciani A, Houdart E, Mounayer C, Saint Maurice JP, Merland JJ (2001) Spontaneous closure of dural arteriovenous fistulas: report of three cases and review of the literature. *AJNR Am J Neuroradiol* 22:992–996
- Partington MD, Rufenacht DA, Marsh WR, Piepgras DG (1992) Cranial and sacral dural arteriovenous fistulas as a cause of myelopathy. *J Neurosurg* 76:615–622
- Renner C, Helm J, Roth H, Meixensberger J (2006) 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* 65(5):506–510
- Ricolfi F, Manelfè C, Meder JF, Arrue P, Decq P, Brugieres P, Cognard C, Gaston A (1999) Intracranial dural arteriovenous fistulae with perimedullary venous drainage. Anatomical, clinical and therapeutic considerations. *Neuroradiology* 41:803–812
- Versari PP, D’Aliberti G, Talamonti G, Boccardi E, Collice M (1993) Progressive myelopathy caused by intracranial dural arteriovenous fistula: report of two cases and review of the literature. *Neurosurgery* 33:914–918
- Wrobel C, Oldfield EH, Di Chiro G, Tarlov EC, Baker RA, Oppman JL (1988) Myelopathy due to intracranial dural arteriovenous fistulas draining intrathecally into spinal medullary veins, report of three cases. *J Neurosurg* 69:934–939

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