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Original Article

Type I Spinal Arteriovenous Fistula with Ventral Intradural Venous Drainage: A Proposal of a Modified Classification

Abstract

Objectives: Spinal arteriovenous fistula (AVF) is the most common spinal vascular lesion and constitutes an abnormal communication between a feeder artery and a draining vein. Arterialization of the venous plexus leads to venous hypertension; consequent edema and congestion of the spinal cord are associated with progressive neurological decline. **Patients and Methods:** In this report, we describe two unique cases of type I cervical spinal AVF, in which a radiculomeningeal artery forms an intradural fistula that drains into the ventral venous plexus. **Results:** Both patients underwent surgical obliteration of the fistula with complete occlusion confirmed on postoperative angiography. **Conclusion:** Both cases do not fit into the current classification scheme. A modified classification is proposed.

Keywords: *Arteriovenous, arteriovenous fistulas, classification, fistula, spinal, Type I, venous plexus, ventral*

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Introduction

Spinal arteriovenous fistulas (AVFs) are the most common vascular lesions in the spinal cord. Nonetheless, they are generally rare and underdiagnosed. They constitute an abnormal communication between a feeder artery and a draining vein. Spinal AVFs have been classified into four types on the basis of the anatomy of the arterial supply and venous drainage [Table 1]. In Type I, a radiculomeningeal artery forms an abnormal communication with the intradural dorsal venous plexus at the level of the dural sleeve of the nerve root. Arterialization of the venous plexus leads to venous hypertension and stasis.[1,2] Consequent edema and congestion of the spinal cord are associated with progressive neurological decline.[2] On rare occasions, an intracranial subarachnoid hemorrhage can occur if the venous plexus drains cranially. Thus, early detection and prompt treatment of these lesions are warranted.

In this report, we describe two unique cases of Type I spinal AVF, in which a radiculomeningeal artery forms an intradural fistula that drains into the ventral venous plexus. To the best of our knowledge, one of our cases is the only reported case of its kind in the literature.[3]

Patient, Methods and Results

Case 1

A 68-year-old Caucasian female with a history of recurrent left carotid artery stenosis treated by carotid endarterectomy was found to have incidental left spinal AVF on follow-up computed tomography (CT) angiography [Figure 1A]. The patient had a previous history of stroke with residual right hemiparesis, but no new neurological symptoms. On digital subtraction (DS) angiography, a vascular structure corresponding to Type I spinal AVF was found at the level of C3–C4. The fistula was supplied by a radiculomeningeal branch of the ascending cervical artery and drained into the ventral venous plexus in both the cranial and caudal directions [Figure 1B].

Because of concerns about the progressive nature of spinal AVF, the patient underwent surgical disconnection through standard C3–C4 laminectomy. Following durotomy and gentle medial retraction of the spinal cord, an AVF with a large arterialized vein was seen on the ventral surface on the spinal cord [Figure 1C]. Bipolar cautery and microscissors were used to coagulate and disconnect the fistula, which appeared to be intradural along the C4 dorsal root [Figure 1D]. Indocyanine green was

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used to document complete obliteration of the fistula. Postoperative DS angiography on the same day revealed complete obliteration with no residual filling [Figure 1E]. Few days later, she was discharged to a rehabilitation center. On the last follow-up in clinic, she did not have significant disability and was able to carry out all usual duties and activities (modified Rankin Scale (mRS) =1).

Case 2

A 68-year-old Caucasian female presented with progressive numbness, weakness, and difficulty with ambulation. The patient had reported neck pain and left arm pain 2 months earlier. On physical examination, hemiparesis and decreased proprioception involving the left side were noted, and there was decreased pain and touch sensation involving the right leg. She also displayed a Hoffman's reflex, and clonus on the left side. These findings are consistent with left-sided Brown-Sequard syndrome. CT angiography showed a vascular lesion on the ventral surface of spinal cord at the level of C6 [Figure 2A]. Magnetic resonance imaging showed that the vascular lesion compressed the left side of the spinal cord and was associated with T2 signal enhancement from the cervicomedullary junction down to the mid-thoracic region [Figure 2B]. DS angiography showed a Type I spinal AVF supplied by a radiculomeningeal branch of the right vertebral artery and draining into the dilated ventral venous plexus. Both the thyrocervical and costocervical trunks also contributed to the fistula, but with no involvement of the anterior spinal artery [Figure 2C]. During the same procedure, the feeder artery was embolized using Onyx. Nevertheless, the patient's neurological examination remained stable, and CT angiography revealed persistent filling of the venous plexus, although this was significantly diminished. Therefore, the patient underwent surgical disconnection through standard C6–C7 laminectomy. Following durotomy, the AVF was identified along with arterialized veins at the ventral nerve root and ventral

Table 1: Classic classification system

Type	Description	Feeders
Type I	Dural AVF in the dorsal sleeve of the nerve root	Radicular arteries
Type II	Intramedullary pial AVM	Anterior and posterior spinal arteries
Type III	Extensive juvenile AVM	Radicular, and anterior and posterior spinal arteries
Type IV	Intradural perimedullary AVF	Anterior and posterior spinal arteries

AVM – Arteriovenous malformations; AVF – Arteriovenous fistulas

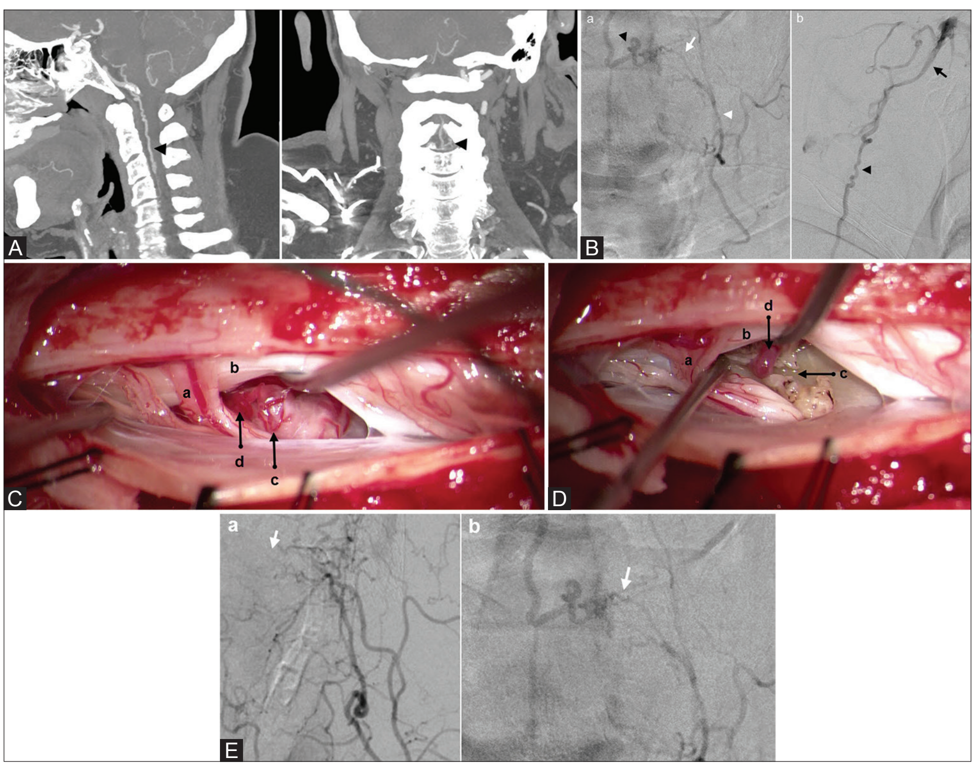

Figure 1: (A) Computed tomography angiography showing a tortuous dilated vein (black arrowhead) on the ventral surface of the spinal cord. (B) Preoperative digital subtraction angiography showing; (a) radiculomeningeal branch (white arrow) arising from the ascending cervical artery (white arrowhead) forming a fistula that drains into a tortuous dilated vein (black arrowhead in a and b) on the ventral surface of the spinal cord. The vein drains caudally and cranially into the posterior cranial fossa (black arrow). (C) Intraoperative image showing the arteriovenous fistula with tortuous dilated vein on the ventral surface of the spinal cord. (a) C4 dorsal nerve root; (b) Gentle retraction of the spinal cord; (c) arteriovenous fistula; (d) draining vein. (D) Intraoperative image showing disconnected arteriovenous fistula. (a) C4 dorsal nerve root; (b) Gentle retraction of the spinal cord; (c) Disconnected arteriovenous fistula; (d) Stump of the draining vein. (E) Postoperative digital subtraction angiography (a) showing complete absence of the fistula, compared to preoperative digital subtraction angiography (b)

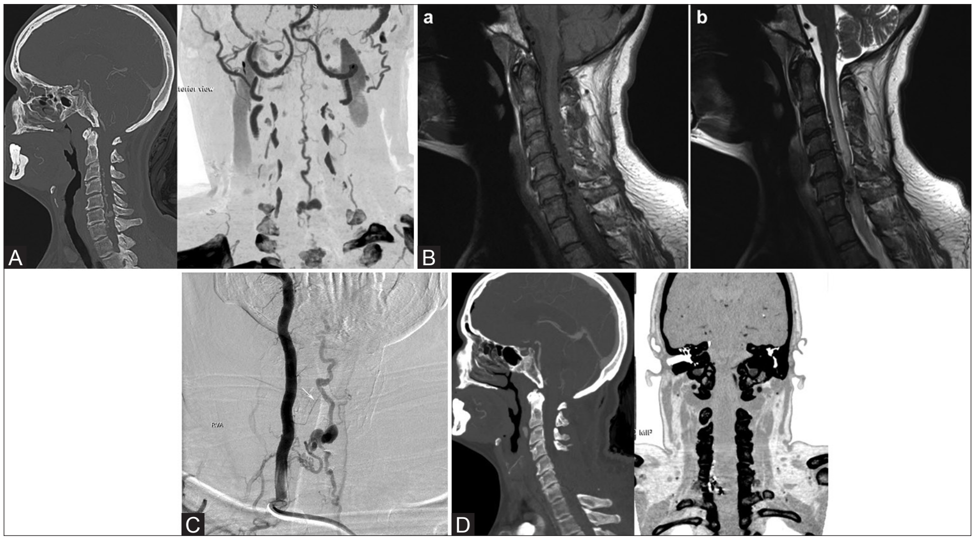

Figure 2: (A) Computed tomography angiography showing a tortuous dilated vein on the ventral surface of the spinal cord. (B) Sagittal T1- (a) and T2- (b) weighted magnetic resonance imaging scan demonstrating spinal cord compression by dilated ventral veins. (C) Digital subtraction angiography showing that the most prominent supply to the lesion arises from a radiculomeningeal branch of the right vertebral artery. (D) Postoperative computed tomography angiography showing no further filling of the fistula

surface of the spinal cord. A temporary clip was placed on the lesion near the nerve root, and then, a permanent clip was used to occlude the fistula. CT angiography performed on the following day showed complete obliteration of the fistula [Figure 2D]. After recovering for few days in the hospital, she was transferred to a rehabilitation center. She made good recovery on reassessment in clinic few months later (from mRS 4-2).

Discussion

Before the advent of angiography, spinal AVF could only be diagnosed intraoperatively during surgical exploration in patients with myelopathy. With the advent of angiography during the 1960s, diagnosis and treatment of these vascular lesions were revolutionized. In 1977, Kendall and Logue[4] reported the first case series with angiographic examination of spinal AVFs. They located the lesions at the level of dural sleeve of the nerve root and demonstrated drainage into the venous plexus around the spinal cord. Their description correlated with Type I AVF in the current classification.

Type I is the most common type of spinal AVF. They are low flow and low pressure vascular lesions located within the dural sleeve of the nerve root. They are supplied by radicular arteries and drain into the dorsal venous plexus. They are mostly found in males between the ages of 50–70 years and are predominantly located in the thoracolumbar region.[2]

Myelopathy results from venous congestion and decreased arterial supply to the spinal cord. It can also result from spinal cord or spinal nerve root compression by a dilated venous plexus. In contrast to arteriovenous malformations (AVMs), spinal hemorrhage is almost never seen in these patients.[2] Intracranial subarachnoid hemorrhage can also occur on rare occasions when the vein drains cranially. The progressive nature of the disease with deterioration of clinical symptoms warrants expedited treatment. Spontaneous regression and resolution have not been reported.

The primary goal of treatment is to achieve complete occlusion of the fistula. This can be done by either surgical obliteration or endovascular embolization. Although no definitive treatment of spinal AVF has yet been established, surgical obliteration has been the treatment of choice for Type I spinal AVFs. The ease of exposure and direct visualization of the vascular anatomy facilitate complete obliteration.[1,2] During surgery, the intradural component of the arterialized veins is identified and disconnected as they enter the spinal canal. This can be performed either through coagulation and separation or clip placement. The postoperative obliteration rate of up to 98%[5] compares favorably with endovascular embolization, which varies from 30% to 90% depending on whether the proximal vein is penetrated.[6]

Classifications

The first angiographic classification of spinal arteriovenous lesions was proposed by Di Chiro *et al*.[7] and Baker *et al*.,[8] who divided them into three types: extramedullary, glomus intramedullary, and juvenile intramedullary. The extramedullary type was later renamed dural AVF. In 1986, Heros *et al*.[9] added a fourth type to the classification scheme. Several classifications have emerged since based either on the genetic origin, the location of the draining veins, or the anatomy of the involved arteries and veins, which is most widely used classification system is based on the anatomy of the arteries and veins involved. The most recent modification by Kim and Spetzler[1] divides spinal arteriovenous lesions into intradural dorsal AVF, intradural ventral AVF, extradural-intradural AVM, intramedullary AVM, and conus medullaris AVM.

In this study, we report two cases with a Type I spinal DAVF that drains into the ventral, rather than the dorsal, intradural venous plexus. The fistulas were supplied by a radiculomeningeal branch of the ascending cervical artery and vertebral artery, respectively, with no anterior spinal artery involvement. These cases do not precisely fit into the current classification scheme, which makes no distinction on the basis of the location of the draining venous plexus. Bulsara *et al*.[3] previously proposed a modification of the classic classification in which Type I is divided into dorsal and ventral subtypes depending on where the venous plexus predominantly draining the fistula is located. With the two cases reported here, we find this modification to be necessary. However, to avoid confusion with the "intradural ventral AVF" type described by Kim and Spetzler,[1] further modifications based on the precise location of the fistula (radicular versus pial) are suggested [Table 2]:

- Intradural dorsal AVF to be changed to intradural radicular AVF and to be subdivided into:

Table 2: Modified classification system

Type	Description	Involved vessels	Kim and Spetzler classification[1]	Bulsara *et al*. modification[3]
Type I	Intradural radicular AVF	Radicular feeding artery drains into intradural venous plexus at the level of dural sleeve of the nerve root	Intradural dorsal AVF	Extramedullary AVF-divided into dorsal and ventral
Type IA		Dorsal venous plexus		
Type IB		Ventral venous plexus		
Type II	Intradural pial AVF	Anterior or posterior spinal artery drains into ventral or dorsal intradural venous plexus, respectively	Intradural ventral AVF-divided into A, B, and C	Intramedullary AVM
Type IIA		Small AVF with single feeder		
Type IIB		Intermediate AVF with major feeder from the anterior spinal artery and minor feeders at the level of the fistula		
Type IIC		Giant AVF		
Type III	Extradural-intradural AVM	AVMs are invested along the somite level, involving skin, muscle, bone, spinal nerves, and spinal cord	Extradural-intradural AVM	Extensive juvenile malformation
Type IV	Intramedullary AVM	Have single or multiple feeders from branches of the anterior and posterior spinal arteries	Intramedullary AVM-divided into compact and diffuse	Perimedullary AVF-involving the anterior spinal artery
Type IVA		Compact		
Type IVB		Diffuse		
Type V	Conus medullaris AVM	Multiple feeders from branches of the anterior and posterior spinal arteries	Conus medullaris AVM	

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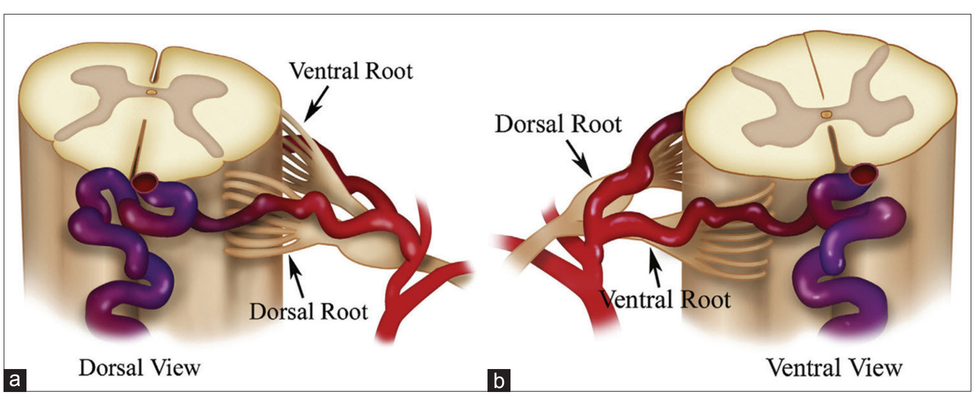

Figure 3: New proposed Type I subclassification as type IA (a) and Type IB (b) (reprinted with permission from: Bulsara *et al*.)[3]

A-Dorsal venous plexus, and B-Ventral venous plexus [Figure 3]
- Intradural ventral AVF to be changed to intradural pial AVF. The latter term is also more accurate as the arterial supply to the fistula can be derived from the anterior or posterior spinal arteries, and draining into the ventral or dorsal venous plexus, respectively[10]
- The different types will be labeled Types I–V.

Conclusion

Despite being the most commonly encountered spinal vascular malformation, spinal AVFs are rare vascular lesions that can lead to significant neurological deterioration. Therefore, early detection and prompt treatment are warranted. Several classification systems have been developed to assist clinical judgment. In this study, we report two cases with Type 1 AVF with ventral drainage, which do not fit into the current classification scheme. A modified classification is proposed with the hope to validate this classification scheme in the future.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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