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 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

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 dorsal 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.[3] 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...
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

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**Table 1: Classic classification system**

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
<th>Feeders</th>
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<tbody>
<tr>
<td>Type I</td>
<td>Dural AVF in the dorsal sleeve of the nerve root</td>
<td>Radicular arteries</td>
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<tr>
<td>Type II</td>
<td>Intramedullary pial AVM</td>
<td>Anterior and posterior spinal arteries</td>
</tr>
<tr>
<td>Type III</td>
<td>Extensive juvenile AVM</td>
<td>Radicular, and anterior and posterior spinal arteries</td>
</tr>
<tr>
<td>Type IV</td>
<td>Intradural perimedullary AVF</td>
<td>Anterior and posterior spinal arteries</td>
</tr>
</tbody>
</table>

AVM – Arteriovenous malformations; AVF – Arteriovenous fistulas

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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)
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 A VF 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.,[3] 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[4] 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.\cite{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,\cite{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>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
<th>Involved vessels</th>
<th>Kim and Spetzler classification\cite{1}</th>
<th>Bulsara et al. modification\cite{3}</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type I</td>
<td>Intradural radicular AVF</td>
<td>Radicular feeding artery drains into intradural venous plexus at the level of dural sleeve of the nerve root</td>
<td>Intradural dorsal AVF</td>
<td>Extramedullary AVF-divided into dorsal and ventral</td>
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<tr>
<td>Type I A</td>
<td></td>
<td>Dorsal venous plexus</td>
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<td></td>
<td></td>
<td>Ventral venous plexus</td>
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<tr>
<td>Type I B</td>
<td></td>
<td>Anterior or posterior spinal artery drains into ventral or dorsal intradural venous plexus, respectively</td>
<td>Intradural ventral AVF-divided into A, B, and C</td>
<td>Intramedullary AVM</td>
</tr>
<tr>
<td>Type I I A</td>
<td>Small AVF with single feeder</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Type I I B</td>
<td>Intermediate AVF with major feeder from the anterior spinal artery and minor feeders at the level of the fistula</td>
<td></td>
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<tr>
<td>Type I I C</td>
<td>Giant AVF</td>
<td></td>
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<tr>
<td>Type III</td>
<td>Extradural-intradural AVM</td>
<td>AVMs are invested along the somite level, involving skin, muscle, bone, spinal nerves, and spinal cord</td>
<td>Extramedullary AVM</td>
<td>Extensive juvenile malformation</td>
</tr>
<tr>
<td>Type IV</td>
<td>Intramedullary AVM</td>
<td>Have single or multiple feeders from branches of the anterior and posterior spinal arteries</td>
<td>Intramedullary AVM-divided into compact and diffuse</td>
<td>Perimedullary AVF-involving the anterior spinal artery</td>
</tr>
<tr>
<td>Type IV A</td>
<td>Compact</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Type IV B</td>
<td>Diffuse</td>
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<tr>
<td>Type V</td>
<td>Conus medullaris AVM</td>
<td>Multiple feeders from branches of the anterior and posterior spinal arteries</td>
<td>Conus medullaris AVM</td>
<td></td>
</tr>
</tbody>
</table>

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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.)\cite{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.

**References**